

Psychoneurotic Profiles Of Thalassemia.

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الخلاصة

مرض فقر الدم البحري مرض مزمن واحد أسباب الإعاقة التي تعود لمضاعفاته المتعددة الجسدية منها والنفسية على المريض. يشغل العلاج الجسدي الحيز الأكبر من الاهتمام في حين لا يحض الجانب النفسي إلا على النزر اليسير من الاهتمام المطلوب إن التشخيص المبكر والعلاج المناسب للأمراض النفسية التي يعاني منها مرضى فقر الدم البحري قد يساهما في تحسين نتائج العلاج المرجوة ويرتقى بنوعية حياة المريض.

هدف البحث تعيين السجل العصابي والنفسي لمرضى فقر الدم البحري وأكثر الأعراض العصابية والنفسية لديهم أخذت عينتين متماثلتين من ألناحية ألعمرية والجنسية أحدهما تمثل مرضى فقر الدم ألبحري والأخرى تمثل أفراد أصحاء من ألمرض. تتألف كل عينة من ثمانين فردا. تم تقييم الإجابات كل على حدة لمعرفة وجود أو عدم وجود مرض نفسي باستخدام قياس كرون كرسب لاختبار التيارات العصابية المعرب، ظهر من نتيجة البحث إن نسبة الأعراض النفسية لدى مرضى فقر الدم البحري كانت 87.5% بينما كانت نسبة الأمراض النفسية لدى العينة السليمة من مرض فقر الدم البحري كانت 19.2% كانت نسب الاضطرابات الرحامية ،ألرهاب،القلق ،الوسواس والاكتناب لدى مرضى فقر الدم البحري هي بالتعاقب 85.5% 88.8% والاكتناب 19.5% والاكتناب ققر الدم البحري هي بالتعاقب 85.5% و 88.8% و الاكتناب النفسي فقر الدم البحري في مجتمعنا كبيرة أن الشعور بالدوار وضيق النفس هو أكثر الأعراض النفسية شيوعا لدى مرضى فقر الدم البحري المتقيم النفسي وجديرة بالاهتمام مما يتطلب وجوب خضوع جميع مرضى فقر الدم البحري للتقييم النفسي وشمول الجوانب النفسية والاجتماعية بالعلاج اللازم و إذا ما تم ذلك سيكون له انعكاساته وشمول الجوانب النفسية العلاج ونوعية الحياة التي يحيياها المرضي.

Abstract

Background: Thalassemia is chronic disabling disease which have both physical and psychological consequences. Thalassemic patients are receiving medical care that is the focus of major attention with lesser focus on the psychiatric aspect of the illness. Early diagnosis and appropriate management of psychiatric disorders among thalassemic patients may improve the outcome, prognosis and overall quality of patients life.

Objectives: To determine psychoneurotic profiles in thalassemic patients and to find out rates of anxiety, phobia, obsession, somatization, depression & hysteria.

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Method: The sample of the study was consisted of case group and control group. Each of the two groups include 80 thalassemic patients and 80 normal people respectively. The self-rating inventory Crown Crisp Experiential Index(CCEI) has been used to study their psychoneurotic profiles.

Results: The study revealed that the frequency of psychiatric morbidity was significantly high among thalassemic patients. The rate of psychiatric symptoms was 87.5% vs.21.25% among thalassemic free people. The rate of somatization (92.5%), phobia (88.8%), anxiety (85%), obsession (83.8), hysteria (81.3%) and depression (76.3). The most common psychiatric symptom was dizziness and shortness of breath and the least common one was feeling loss of sympathy with others.

Conclusions: The psychiatric morbidity of thalassemic patients is remarkable and suggest that all patients with thalassemia should undergo psychiatric assessment and Psychosocial aspects need to be addressed in the overall treatment of children with thalassemia.

Introduction

Thalassemia(Cooley's anemia)(Mediterranean anemia) is an inherited partial or complete impairment of hemoglobin synthesis, or Thalassemia syndromes are genetic disorders of alpha and beta globins production that are prevalent among patients from the Mediterranean, Middle East, India, Africa and Southeast Asia.

Thalassemia was first described by Cooley and Lee in 1952 in several Italian children as a severe anemia with spleen and liver enlargement, skin discoloration, and bony changes. (1) The following are the etiological types of thalassemia

A-Beta- thalassemia major.

B-Beta-thalassemia intermedia.

C-Sub clinical thalassemia trait.

D-Hemoglobin H disease.

E-Hemoglobin Bart's disease. (2)

Clinical Presentation:

In beta thalassemia the excess alpha globin chains are unstable and precipitate within the cell leading to variety of clinical manifestations. Infants with beta thalassemia major are well at birth with symptoms emerging as fetal hemoglobin levels decline around 6 months of age. Pallor, irritability, growth retardation, abdominal swelling due to hepatosplenomegaly and jaundice indicate the presence of severe hemolytic anemia. Shortly after, symptoms



associated with ineffective erthryopoiesis such as bony abnormalities and abnormal skeletal development follow. Because of the anemia, there is also an increase in iron absorption, with toxicity leading to further complications. Many of the complications of thalassemia seen in developed countries today are the result of iron overload. Growth retardation is a consequence of severe anemia. There are numerous cardiopulmonary complications, including congestive heart failure and pericarditis, hepatobiliary disease, and hepatitis. (3)

Evaluation/diagnosis:

In major thalassemia, diagnosis is made by hemoglobin electrophoresi.

However, deletion of two alpha globin genes often mistaken for iron-deficiency anemia because patients will have a normal hemoglobin electrophoresis. The diagnosis of iron deficiency must be eliminated and the diagnosis presumed or confirmed by DNA testing. Deletion of three alpha globin chains is identified by hemoglobin electrophoresis. Beta thalassemia can always be diagnosed by hemoglobin electrophoresis. Those infants born with Hydrops Fetalis, a negative Coombs test and morphologic abnormalities seen on smear rule out hemolytic diseases due to blood group incompatibility. (4)

Psychological aspects:

Children with thalassemia are often a bit thin. They go through normal puberty, though often two or three years later than their "normal" friends. Usually a women will need regular transfusions during pregnancy, not so much for herself, but to help the baby to grow normally. If parents are over-protective, the child feels that he must be in constant danger, and because he can't see what the danger is, he becomes anxious about every little thing. We always recommend parents to treat their thalassemic child as normally as possible.

To help children feel secure, it is important to answer all their questions truthfully. The biggest problem for thalassemic during adolescent and young adult life is to continue taking the pump regularly. Older patients often find themselves torn between despair of the "old thalassemia", which was a fatal disease, and hope of the "new thalassemia", a disease that can be controlled, with a normal future, but requires a continual struggle. So it is only natural for a thalassemic's mood to swing rather suddenly. (5) Culture and education play a major role in illness experiences. The consequence

of thalassemia is extremely stressful, and patients face a variety of physical, psychological, and social problems. ⁽⁶⁾In thalassemic any complication they develop is usually due to adverse effects of treatment (transfusion & chelating). ⁽⁷⁾

In thalassemia treatment its self is stressful to patients because of the need for frequent injections and transfusions. Transfusion usually of red blood cells rather than whole blood. The frequency of transfusion is variable depending upon the severity of anemia but it is usually every four weeks.⁽⁸⁾

Chelating agents such as desferrioxamine (desferal) which is widely used for chronic iron overload from multiple transfusions. It is administered 20-40 mg/kg by S.C. infusion pump daily and 500-1000 mg I.V. infusion in separate solution along with each unit of blood transfused. $^{(9)}$

6-Most patients do not need more than psychological support, reassurance and help to improve their adjustment to illness. Psychiatrist should look carefully for symptoms which are strongly associated with definite psychiatric disorder such as persistent deliberate destructiveness, aggression leading to injury, deliberate self harm, inappropriate sexual behavior, fire setting, social disinhibition, persistent isolation and withdrawal, bizarre behavior& hallucinations and delusions. (10)

Neurosis:

Neurosis is chronic or recurrent non psychotic disorder characterized mainly by anxiety which is experienced or expressed directly or is altered through defense mechanisms. It appears as symptom such as an obsession, a compulsion, a phobia or a sexual dysfunction.

In Diagnostic and Statistical Manual of Mental Disorders 1V edition(DSM-IV) neurosis is defined as: a mental disorder in which the predominant disturbance is a symptom or group of symptoms that is distressing to the individual and is recognized by him or her as unacceptable and alien (ego-dystonic), the reality testing is grossly intact, behavior does not actively violate gross social norms, the disturbance is relatively enduring or recurrent without treatment and is not limited to a transitory reaction to stressors and there is no demonstrable organic etiology or factor.

Bio-social theory defined neurosis as a poor ability to adapt to ones environment, an inability to change one's life patterns, and the inability to develop a richer, more complex, more satisfying personality.

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Anxiety is a response to a threat that is unknown, internal, vague or conflictual. (11)

Anxiety is the subjective experience of unpleasant tension, uneasiness and distress that accompanies psychic threat or conflict.

Origin of neurosis:

- 1-W.Cullen (1710-1790) was the first to use the term neurosis to denote diseases without fever or localized pathology.
- 2-R.Whytt (1714-1766) divided neurosis into hysteria, hypochondriasis & nervous exhaustion (neurasthenia).
- 3-Sigmund Freud(1856-1939) In 1893 stated that memories of trauma can be responsible for hysterical symptoms)
- 4- Jean-Martin Charcot (1825-1893) regarded neurosis as brain disease.
- 5-Jean-Paul Sartre (1905-1980) stated that neurosis is an escape from freedom which is the key to maintaining psychological health⁽¹²⁾.
- 6-Ivan Petrovitch Pavlov stated that all of the human race can be separated into several types: artists, thinkers, and a middle type. The chronic pathological deviations from the normal are expressed in the excitable and in the weak type by a mild form of neurosis. Neurasthenia is a form of general weakness, occurring in the middle human type. Hysteria is the result of general weakness in the artistic type; psychasthenia a product of weakness in the thinking type. (13) The term remain useful as simple descriptors, to indicate disorders that are often comparatively mild and usually associated with some form of anxiety or marked tension, especially if the specific disorder can not yet be determined. (14)

Common symptoms of neurosis:

Neuroses with anxiety as the chief symptom are common: around 16% of the population are affected by some form of an anxiety illness at any one time. In mild depressive disorder there are symptoms which can be broadly categorized as "neurotic. These include anxiety, phobias, and obsessional symptoms. In addition to these symptoms, people with mild depressive disorders will also have a degree of low mood, lack of energy, and irritability. Biological (physical) symptoms such as poor appetite and weight loss etc, may be found, but are usually much less severe than those which occur in people with major depressive disorder. (15)

The other common symptoms of neurosis are an emotional disturbances that is disproportionately severe or prolonged distressing alterations in bodily sensations(autonomic & somatic), altered cognition such as novelty & uncertainty, changes in self-image & self-awareness, thoughts of self-reassurance& obsession, avoidance &ritual behaviors. In addition the disorder can not entirely be explained by the presence of medical conditions, psychosis, depressive mood states or personality disorders. (16)

Epidemiology:

Neurosis is common .It affect at least 15% of the adult population in UK and at least 50% of people suffering from neurotic symptoms.

The prevalence of neurotic disorders in UK from national psychiatric morbidity(NPM)of house hold survey involve the following non specific neurotic disorder (NSND) 7.7%, generalized anxiety disorder(GAD) 3.1%, depressive episodes 2.1%,all phobias 1.1,obsessive compulsive disorder(OCD)1.2% panic disorder 0.8%,any neurotic or mood disorder 16%.

In epidemiological catchment area study (ECA) result include the following any anxiety disorder 12.6%,GAD 3.7%,all phobic disorders 10.9%,agoraphobia 5.8%,panic disorder 1.3%.OCD 2.1%,depressive episode 5.7%.

The United States national co morbidity study(NCS) obtained the following results any anxiety disorder 17.2%,GAD 3.1%,all phobic disorders 11.9%, panic disorder 2.3% & agoraphobia 2.8% & depressive episode 10.3%.The results of WHO-psychological problems in general health study (WHO-PPGHC) include any anxiety disorder 10.2%,GAD 7.9%, panic disorder 1.1%, agoraphobia 1.5% depressive episode 10.4%. (16)

The co morbidity between psychiatric disorders and thalassemia:

Thalassemia impose persistent stress on the youngsters and their caregivers which in fact, caregivers may be more affected than the youngsters themselves because of a better appreciation of the magnitude of the illness and its treatment with resultant increased the risk for anxiety and mood disorders. (17) Fatalism positively correlated with somatization and depression. (18)

Thalassemic patients will face many stresses in their whole life, including frequent blood sampling for laboratory tests, multiple transfusions and frequent subcutaneous injections of chelator drugs, which altogether will make the patient susceptible to psychiatric disorders such as depression.



Some studies demonstrate that 80% of thalassemia major patients at least suffer from one psychiatric disorder. In contrast some other studied revealed that thalassemic patient will cope with life difficulties in better way. (19)

Thalassemic adolescents were having higher scores in neuroticism. Some behavioral problems are also found to exist along with neurotic manifestations. There remains a need to improve the management of thalassemia in terms of psychological aspects in order to improve the mental health of this group. Many neurotic symptomatologies were found in patients with thalassemia in different surveys. Depressive moods and anxiety were diagnosed in children with thalassemia major in a study conducted in Italy. Same results were found in a similar study conducted in Singapore. It is also observed that subjects with chronic illness have limiting their daily life, experience more depression than those without chronic illness. Another study conducted in Utrecht, Netherlands points to the depressive coping styles in thalassemic adolescents. (20)

Aims of the study

1-To determine the psychoneurotic profiles among thalassemic patients.

2-To identify the commonest psychiatric symptom in thalassemic patients.

The Methods

A-Subjects:

One hundred thalassemic patients were asked to fill the questionnaire. Of the 100 patients that met inclusion criteria (age between12years to 28 years& diagnosed as thalassemic), 80 consented to take part in this study.

We have complete data on all of them. Non-participants did not differ significantly from participants in terms of demographics or illness characteristics. The reasons of their nonparticipation include that it was time consuming or they were tired enough to answer 48 questions.

The study was conducted during the period from 2nd January 2008 to 10th May 2008 in the Hereditary Blood Diseases Care Centre in Basra city.

The sample was selected randomly by attending in Hereditary Blood Diseases Care Centre. The exclusion from the sample include only those who had concurrent diagnosis of psychotic disorders or substance use disorders. There were forty six females and thirty four males, and the sample was heterogeneous regarding chronicity of the disease and the presence or absence of its complications.

The cases sample was selected on the basis of having a diagnosis of thalassemia and all patients had a special follow up cards. Verbal consent was obtained from all sample members. Thirty questionnaire was filled by the patient's companions or by the researcher because of their difficulties in reading.

Eighty students from Shat Al-Arab secondary school for boys and girls and from Basrah Medical College according to age and sex characters of that of the case sample form the control sample group.

B-Assessment:

The crown crisp Experiential Inventory (the Middlesex hospital questionnaire) (CCEI). The Crown Crisp Experiential Inventory is a general mental health screening measure. It has six subscales, free-floating anxiety, phobic anxiety, obsession, somatic anxiety, depression & hysteria. Test-retest reliability has been clearly demonstrated & is reported in the test manual at over 0.68 for all six subscales. The CCEI was included to allow the assessment of criterion validity. (21). CCEI has been used to investigate variety of specific problems. Its relative popularity relates to its genuine fulfillment of need for brief, easily administered scored personality test with psychiatric base and useful for testing clinically devised hypothesis particularly in psychosomatic field. (22). The CCEI was translated into every day Arabic from English by two psychiatrist, a psychologist and a teacher well versed in both languages. meaning. (23)

C-Statistical analysis:

Statistical analysis was performed by two statisticians using SPSS 11 for windows. Microsoft office Excel 2003 was used for selective analysis of the six subscales data. After confirming that key variables were normally distributed, Chi-square test was used to determine statistical significance of our findings. All values were presented as numbers and percentages except for age where mean and standard deviation appeared to be more appropriate.



Results

Table -1: mean ages of cases and control.

	Cases(thalassemia)	Control	P value
No. of	80	80	0.719
persons			
Mean	17.41	17.63	
SD	4.09	3.78	

The range of age for both cases and control was12 years-28 years. As presented in the table, the means of cases and control groups were semi identical to each others. No significant statistical difference was found between the case group & control group.

Table -2: Comparison of cases and control regarding sex.

Cases(thala	Control			P value	
	No.	%	No.	%	0.437
Male	34	42.5%	36	45%	
Female	46	57.5%	44	55%	
Total	80	100%	80	100%	

As presented in the table ,there were 46(57.5%) females and 34(42.5%) males out of 80 thalassemic patients forming the cases group. There was insignificant statistical difference between the case group &the control group.

Table-3: Comparison of cases and controls regarding psychiatric morbidity (neurosis).

	Cases(thalassemia)		Controls		P value
	No.	%	No.	%	0.001
Without psychiatric	10	12.5%	63	78.75	
symptoms					
With psychiatric	70	87.5%	17	21.25	
symptoms					
Total	80	100%	80	100%	

As presented in the table, there were 70(87.5%) cases out of 80 thalassemic patients who had psychiatric symptoms. There was a significant statistical difference between the case group & the control group , i.e. psychiatric symptoms were significantly more frequent among thalassemic patients than persons who were free from thalassemia.

Table -4: The symptoms among thalassemic patients with psychiatric symptoms.

co	ommon symptoms	No.	%		No.	%
1	Dizziness& shortness	79	98.8%	Feeling of loss of	15	18.8%
	of breath			sympathy with		
				others		
2	Enjoyment of attention	73	91.3%	Reduced sexual	20	25%
				interest		
3	Worry when relatives	73	91.3%	Acting	23	28.8%
	are late					
4	Cleanliness	72	90%	Panicky in crowds	24	30%
5	Much effort Life	71	88.8%	Special effort to	26	32.5%
				face up difficulty		

The most common symptom was shortness of breath (98.8%). The least common symptom was feeling of loss of sympathy with others (18.8%).

Table -5: comparison of cases and controls regarding phobia.

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		Cases(thalassemia)	Contro	ols	P value
	No.	%	No.	%	0.000
Without phobia	9	11.3%	73	91.25	
With phobia	71	88.8%	7	8.75	
Total	80	100%	80	100%	

As presented in the table there were 71 cases out of 80 thalassemic patients had phobia. There was significant statistical difference between the case group &the control group "i.e. phobia was significantly more frequent among thalassemic patients than non thalassemic individuals.

Table -6: Comparison of cases and controls regarding somatization.

	Cases(thalassemia)		Controls		P value
	No.	%	No.	%	0.001
Without somatization	6	7.5%	64	80	
With somatization	74	92.5%	16	20	
Total	80	100%	80	100%	

As presented in the table, there were 74 cases out of 80 thalassemic patients who had somatic symptoms. There was a significant statistical difference between the case group &the control group, i.e. somatization was significantly more frequent among thalassemic patients than nonthalassemic individuals.

Table -7: Comparison of cases and controls regarding hysteria.

•	Cases(Thalassemia)		Cont	rols	P value
	No.	%	No.	%	0.001
Without hysteria	15	18.8%	67	83.75	
With hysteria	65	81.3%	13	16.25	
Total	80	100%	80	100%	

As presented in the table, there were 65 cases out of 80 thalassemic patients who had hysteria. There was a significant statistical difference found between the cases group &the control group. Hysteria was significantly more frequent among thalassemic patients than persons without thalassemia.

Table -8: Comparison of cases and controls regarding anxiety.

	Cases(Thalassemia)		Controls		P value
	No.	%	No.	%	0.000
Without anxiety	12	15%	76	95	
With anxiety	68	85%	4	5	
Total	80	100%	80	100%	

As presented in the table, there were 68 cases out of 80 thalassemic patients who had anxiety. There was significant statistical difference between the cases group & the control group.

Anxiety was significantly more frequent among thalassemic patients than those with out thalassemia.

Table -9: comparison of cases and controls regarding obsession.

	Cases(Tl	nalassemia)	Contr	ols	P value
	No.	%	No.	%	0.000
Without obsession	13	16.3%	77	96.25	
With obsession	67	83.8%	3	3.75	
Total	80	100%	80	100%	

As presented in the table ,there were 67 cases out of 80 thalassemic patients had obsession. There was significant statistical difference between the case group & the control group.

Obsession was significantly more frequent among thalassemic patients than those with out thalassemia.

Tuble 10. Comparison of cases and controls regarding depression.								
	Cases(Thalassemia)		Contr	ols	P value			
	No.	%	No.	%	0.000			
Without	19	23.8%	77	96.75				
depression								
With depression	61	76.3%	3	3.75				
Total	80	100%	80	100%				

Table -10: Comparison of cases and controls regarding depression.

As presented in the table, there were 61 cases out of 80 thalassemic patients who had depression. There was significant statistical difference between the case group & the control group.

Depression was significantly more frequent among thalassemic patients in comparison with those with out thalassemia.

Discustion

Thalassemia is chronic disease which have both physical and psychological consequences. Thalassemic patients are receiving medical care that is the focus of major attention with lesser focus on the psychiatric aspect of the illness. Early diagnosis and appropriate management of psychiatric disorders among thalassemic patients may improve the outcome, prognosis and overall quality of patients life.

The purpose of this study is to provide us with essential information about the psychological and psychiatric aspects of thalassemia that is a relatively common and disabling chronic illness by estimation of psychoneurotic profiles among thalassemic patients.

It is expected that thalassemia causes some psychiatric morbidity due to its early onset, hospitalization, long-term medication usage and restricted social life, being under an obligation to provide the compliance of the child to the treatment, and the burden of economical problems and shameful feelings. (24)

The range of sample age was 12 years-28years, the mean of age of cases group was 17.41years..Thalassemic patients consist of 46 (57.5%) of females and 34 (42.5%) of males.

According to our study the rate of psychiatric symptoms in thalassemic patients was 87.5 % versus 21.25% among control. The difference between the two groups was statistically significant as the P value was 0.001. This result is consistent with that of similar studies created in other countries such as that of Giuseppina Messina (80%)⁽²⁵⁾ and Ayden B.& Yaprak I. (80%)⁽²⁶⁾.

The results of A.Saini⁽⁵⁾, D.Shaligram⁽¹⁸⁾ and J.D.Moorjani ⁽²¹⁾ were also consistent with that of this study but the rates appeared to be

much lower. The rates of the last studies were 54%, 67% and 46% respectively. The higher rate in our study might be related to sociocultural factors, difference in chronicity, medical care provided, sample age range, subtype of thalassemia& their proportion in the sample size and presence or absence of physical complications.

In keeping with other studies somatization (92.5%) phobia (88.8%),anxiety(85%),obsession(83.8),hysteria(81.3%)and depression (76.3%) were the main findings.

Our study found higher rates of phobia, hysteria and particularly somatization than previous studies. Differences in socio-cultural factors may account for these findings, also our study included children who may express their psychological distress as somatization and phobia rather than as depressive symptoms which appear more likely to occur in older age as a result of growing insight into the nature of their illnesses. We have to put in our minds that complications of thalassemia and its treatment might be behind the high rate of somatization and to lesser extent to high rate of phobia.

The rates of anxiety(85%),obsession(83.8%) and depression(76%). were also high and inconsistent with that of previous studies that could be explained by sociocultural factors of our society and CCEI inability to differentiate between symptoms resulted from neurosis from that resulted from personality traits and might be the result of differences in measurement scales and cultural patterns of coping with disease

The rate of anxiety (85%) was higher than that of depression(76.3%) might be related to involvement of children and adolescents in our sample who tend to express their psychological difficulties in manner other than that of typical depression such as somatic symptoms, anxiety or phobia.

The most common symptoms in the present study was dizziness and shortness' of breath (somatization). It is consistent with that of some previous studies such as that of D. Shaligram & S.H. Husseini and inconsistent with other previous studies such as that of Ghanizadeh Ahmad where depressed mood was the most common symptom.

In brief, the present study reveals that, the psychiatric symptoms in thalassemic patients is remarkable and suggest that all patients with thalassemia should undergo psychiatric assessment and Psychosocial aspects need to be addressed in the overall treatment of patients with thalassemia to prevent the development of clinically manifest psychiatric disorder, so that early intervention will improve patients cooperation with treatment and improve their quality of life.

The recommendations:

- 1-It is recommended to continue researches about psychological and psychiatric aspects of thalassemia and to enlarge the sample size in the future.
- 2-It is better to use more than one method of assessment of the psychiatric aspects of thalassemia at the same time like administration of CCEI, General Health Questionnaire (GHQ) and clinical interview together in order to obtain more accurate results.
- 3-As known that thalassemia affect quality of life of patients⁽²⁸⁾, so that it is recommended to bring more attention toward this aspect.

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