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Quality of life in thalassemia major patients in an Iranian district

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Abstract: Beta thalassemia major is a chronic inherited disease which leaves a lot of physical and mental effects on the individual and family. Over time the intensive and disabling complications of the disease affect body organs and tissues, reduce the individual's efficiency, and ultimately reduce the quality of life. The present research was carried out with the objective of a study on quality of life in thalassenia major patients referring to thalassemia center of Boeir-Ahmad. This research is a descriptive- analytical study which was carried out on 72 thalassemia patients referring to thalassemia center of Boeir-Ahmad. The patient's quality of life was studied using an Iranian version of short questionnaire with 36 options on quality of life. Patient's gender, education, age, Job, medical history, familial background, blood group, RH type and facial deformities were evaluated and reported. Data are reported using descriptive statistics such as frequency, mean and standard deviations and also inferential statistics. The age mean and standard deviation (SD) of participants were 20.5± 4.7 years, 65% were single, and 40% with mild to moderate face deformities. The findings in the research showed the quality of life was relatively desirable in 52 % of the patients, undesirable in 19% and desirable in 29%. Also the findings showed that the emotional role is most affected aspect of quality of life. Mental health and general quality of life did not show significant correlation with level of education, sex, economic status, type of follow- up, type of complications. A significant correlation was observed only with facial changes ($p<0.001$). undesirable and relatively desirable quality of life and also the high prevalence of facial deformities are significant health problems in Boirahmad thalassemia patients**. So** we suggest applying appropriate educational plans and psychiatric consultations and also more and better medical services and rehabilitations to reduce the complications of the disease in these patients.
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1. Introduction

Beta Thalassemia major is an inherited hypochromic microcytic anemia which results from abnormality in the structure of beta hemoglobin chain. Based on severity of genetic defect, there are three types of beta Thalassemia including beta Thalassemia minor, inter media and major (Fergus, 2002). Each year, about 100,000 children might be born with severe forms of the disease over the world (Bateman and Wisconsin, 2005). It is also the most prevalent genetic disease in Iran and statistics released by The Iranian Foundation for special disease reported 220000 patients suffering from Thalassemia major (Charity foundation for special disease, 2003). Thalassemia prevalence in Iran varies in terms of geographical zones, so that the highest prevalence is on the Caspian Sea shores and the Persian Gulf. Guilan, Mazandaran, Hormozgan, Khuzestan, and Kohgiluyeh Boer Ahmad are Iranian provinces with the highest prevalence rates (Vichinsky et al; 2005). The disease is associated with severe and long-term complications including anemia, splenomegaly and facial and bone deformities (Borgna-Pignatti et al; 2005).

Also concern over premature death may result in anger, despair, isolation and sever tension in these patients (Ismail et al; 2006). These patients have to get lifetime medical treatment that result in numerous social, psychiatric and economic problems for them (Pakbaz et al; 2005). Generally the disease is diagnosed in childhood, and medical care should starts as soon as the diagnosis is confirmed. Unpleasant, frequent and longitudinal treatment processes, beside their complications, result in increasing psychosocial problems in these patients (Jarman and Oberklaid, 1990).

Consequently a consensus conference of international experts has stated that health related quality of life instruments should include physical, social and emotional functioning as well as

perceptions of overall quality of life or general life satisfaction (R.Berzon and Hays, 1993). Compared with healthy individuals, thalassemia patients have more serious psycho – social problems. Medical treatment should be along with emotional and psycho – social supports to improve quality of life and prevent irreversible complications in these patients (Aydin et al; 1997).

Boirahmad is a province in south-east of Iran and because of prevalent familial marriages; thalassemia is an important health problem in this county. However no data is available on the quality of life in the thalassemic patients living in Boirahmad. The present research was carried out with the objective of study on quality of life in thalassemia major patients referring to thalassemia center of Boirahmad.

2. Materials and Methods

A hundred of thalassemia major patients over the 14 years of age, who referred to thalassemia center of Yasouj, participated in the study. Data was collected interviewing and filling in questionnaire. Demographic data, (age, gender, marital status and economic status), regularity in treatment follow up, type and severity of clinical complications, level of education, history of Despheral use, the common transfusion blood product, RH type, history of familiar thalassemia, consanguinity and facial deformities were recorded in a data sheet. Quality of life was assessed using a standardized Iranian questionnaire. In this questionnaire the patient's quality of life is evaluated using 36 questions, in eight sub-scales including physical function, physical role, bodily pain, general health, vitality, social function, emotional role, and mental health(Montazeri et al; 2005). The questions were scored based on a 1 to 5 Lickert scale.

The eight, scales were summed up into two more general domains named mental health and physical health. The physical health domain included physical role, physical pain, physical function and general health and the mental health domain included vitality, emotional role, social function, and mental health.

Quality of life scores were transformed in percent of maximum attainable point and categorized in three groups; group with impaired quality of life, group with relatively desirable and desirable quality of life. Also quality of life scores are reported as mean ± standard deviation of computed percent. Data were analyzed using SPSS version 15. Binomial variables were compared using U. Man Whitney; categorical variables were compared using Kruskal Wallis test.

3. Results

Socio- economic characteristics of cases are presented in table 1. Mean age of the cases was 20.± 4.7 years and 65% of them were female (n=47). Table 2 shows mean and standard deviation of different aspects of patient's quality of life. Findings showed that 19% of cases had impaired quality of life and 52% had relatively desirable and 29% of cases had desirable quality of life. The study showed that 19% of thalassemia cases had impaired quality of life, and 52% had relatively desirable and 29% with desirable quality of life.

Painful and prolonged treatments of thalassemia disease, exposes the patients to various type of social, emotional and behavioral problems, which deteriorate patients quality of life. Hadi et al, in their study on Shiraz thalassemia patients showed that thalassemia regardless of its severity affect different aspects of patient's quality of life (Hadi et al; 1990). similar findings were reported on quality of life and mental health in thalassemia patients by Khani et al (khani et al; 2009). Mikelli et al also reported lower quality of life in patients with thalassemia major compared with their handicapped peers (Mikelli and Tsiantis, 2004).

Our findings showed that bodily pain was the most affected domain of quality of life in these patients, while the healthiest domain was related to emotional role. Khani et al in their study on over 15 years old thalassemia patients of Mazandaran reported desirable physical health in the patients (khani et al; 2009) But Alavi et al showed physical health as the most problematic domain in 8-18 years old Shahrkord thalassemia patients(alavi et al; 2007). However in a study on over 14 years old thalassemia patients of Tehran, Tabarsi et al, reported mental health as the most affected quality of life domain (Tabarsi et al; 2006). Hadi also reported physical health a main problem in Shiraz thalassemia patients (Hadi et al; 1990).

In this research the status of emotional health was shown to be impaired. It is likely that in addition to physical and functional problems, the thalassemia patients might be faced to other circumstances such as marriage, higher education and getting appropriate job, which cause many mental problems in these patients(Kashani et al; 1989).

In a study by Pour Movahed, it was shown that thalassmia patients have a high anexity compared with their peers age (Pourmovahed et al; 2003). Also Haqshenas reported that thalassemia children have higher psychiatric disorders including anxiety and depression compared with other chronic disabilities (Haghshenas and Zamani, 1997). These problems might ultimately lead to despair and reduced social functions. In this research, general quality of life and

physical health of the patients participating in the research did not show a statistically significant association with level of education, marital status, place of residence, family history of thalassemia and economic status, and the only significant association was with facial deformity.

Lack of medical equipment's, distance from medical centers, costs of treatment and diagnosis, are reported factors that causes economic problems in these patients (Karimov and Asadov, 1995). It is reported that in a group of thalassemia patients 35% had high and 49% had moderate economic problems (Tabarsi et al; 2006).

Due to numerous mental and physical problems these patients are not often successful in continuing their education (Dipalma et al; 1998). With higher education and information, these patients can solve their problems easier, and consequently they will be healthier than uneducated patients (Hadi et al; 1990). Self-care training and educating the patients about complications, signs and symptoms of thalassemia would improve their quality of life (Kasper et al; 2005). In this study facial deformity was the most important factor related to physical health and quality of life. Similar findings are reported by other studies (Tsiantis et al; 1996). Khani Also showed 64.9% of the thalassemia patients suffered from mood and mental disorders that were related to thalassemia deformities (khani et al 2009). Generally, physical deformities of thalassemia extensively expose patients to emotional problems during adulthood and can lower quality of life in these patients (Kashani et al; 1989).

Table 1: social and individual characteristics of Thalacemic patients

STATISTICAL INDEX VARIABLE		FREQUENCY	PERCENT
Sex	female	47	65
	male	25	35
Place of residence	town	44	61
	village	28	39
education	illiterate	5	7
	Primary schoool	5	7
	Guidance school	19	26
	High school	17	24
	diploma	16	22
	Academic	10	14
Marital status	Single	69	96
	Married	3	4
Age of diagnosis	Birth to 6 months	7	10
	6 months to one year	10	14
	Two years to 5 years	16	22
	5 years an higher	39	54
familial Background	yes	28	39
	No	44	61
consanguinity	Yes	45	62.5
	No	27	37.5
Blood group	A	28	39
	B	8	11
	Ab	2	3
	O	34	47
RH Type	Positive	69	96
	Negative	3	4
common transfusion	Condensed globule	11	15
	Washed globule	59	82
	Ordered blood	2	3
Previous Despheral use	Yes	60	83
	No	12	17
Method of Despheral injection	Subcutaneous by pomp	60	83
	Only serum despheral	2	3
	Muscular injection	5	7

	Subcutaneous injection Pomp and serum	**5**	**7**
injection complications	**Topical reaction and pain**	**44**	**51**
	Painful injection	**18**	**25**
	Sensitivity to medicine	**8**	**5**
Facial deformity	**No**	**12**	**16.6**
	Mild	**29**	**40.2**
	Moderate	**28**	**38.8**
	Serious	**3**	**4.1**
follow- up	**Irregular**	**27**	**37.5**
	Regular	**45**	**62.5**
Job status	**House work house wife**	**6**	**8**
	Pupil	**24**	**33**
	Student	**10**	**14**
	Employed	**2**	**3**
	Unemployed	**30**	**42**
family size	**Up to 3**	**4**	**6**
	4-5	**25**	**35**
	6-7	**30**	**42**
	8 and higher	**13**	**18**
Fathers job status	Unemployed	17	**24**
	Retired	16	22
	Pensioned	9	13
	Farmer	6	8
	Employer	6	8
	Worker	18	25
Insurance status	Protective	2	3
	Medical service	37	52
	Social security	24	33
	Armed forces	8	88
	Without insurance	1	1

Table 2: Mean and standard deviation of the marks of quality of life (General and in components of physical and mental health) in thalassrmia patients of Boir Ahmad.

Statistical index variable	frequency	Mean±SD	Confidence interval	minimum	maximum
Quality of life general	72	62.6±16.5	58.8-66.5	14.58	93.75
Physical health	72	64.6±16.5	60.8-68.5	27.8	91.2
Psychological health	72	59.4±18.2	55.1-63.7	12.5	*94.4*

Table 3: Mean and standard deviation of the marks of the eight domains of SF-36 in the thalassemia major patients of Boir-Ahmad.

Statistical index Dimensions of quality of life		Mean±SD	Confidence interval 95%	First group*		Second group**	
				frequency	percent	frequency	**percent**
Physical health	Physical functioning	70.4±24.1	64.7-76.04	31	43	**41**	**57**
	Physical role	56.8±29.7	49.8-63.8	38	53	**34**	**47**
	Bodily pain	73.1±19.9	68.4-77.7	27	37	**45**	**63**
	General health	58.4±19.2	53.9-62.9	37	51	**35**	**49**
Mental health	Vitality	58.2±17.6	54.1-62.4	35	53	**37**	**47**
	Social functioning	67.5±24.9	61.7-73.4	43	54	**29**	**46**
	role Emotional	55.8±33	48.03-63.5	37	46	**35**	**54**
	Mental health	56.1±20.1	51.4-60.9	37	51	**35**	***49***

*** – First group:** marks lowerthan the average of the research community.
****-Second group:** marks higher that the average of the research community

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Mean score in eight domains of quality of life is shown in Table 3. The highest score in quality of life was related to bodily pain (73.1±19.9), while the lowest score was in role emotion (55.8±33.) domain. Except for face deformities and complication severity, we didn't find any significant relation between quality of life scores and other variables.

Findings related to demographic characteristics are shown in Tabe 4.

Table 4: Marks (status) of general quality of life in thalassemia patients of Boyer Ahmad in terms of individual-socialcharacteristics.

Statistical index Variable		*General quality of life* standard deviation± average	*Confidence interval 95%*
Sex	female	64.2±16.7	59.3-69.1
	male	59.7±15.9	53.1-66.3
Residence	town	64.4±17.3	59.1-69.6
	village	60±15	54.1-65.8
education	illiterate	52.5±9.6	40.6-64.4
	p.school	55.1±8.5	44.5-65.7
	Guidance school	59.7±19.7	50.2-69.2
	High school	64.4±16.1	56.1-72.6
	diploma	63.6±16.6	54.7-72.4
	associate	73.5±14.4	55.6-91.3
	B.S	71.8±10.9	58.3-85.2
Age of diagnosis	Birth to 6 months	63.3±15.3	58.4-68.3
	6 months to one year	60.4±15.2	52.3-68.5
	Two years to 5 years	62.8±23.6	45.9-79.6
	5 years an higher	63.9±17.3	47.9-79.8
familial Background	yes	61.5±17.4	54.7-68.2
	No	63.1±16.1	58.1-68.1
Consanguinity	Yes	62.7±18.1	57.2-68.1
	No	62.6±13.6	57.2-68.02
Blood group	A	58.6±14.4	53-64.2
	B	65±14.2	53.1-76.8
	Ab	70.5±24.1	*
	O	65±18.1	58.7-71.3
common transfusion	Condensed globule	65.05±15.9	54.4-75.7
	Washed globule	62.2±16.9	57.8-66.6
	Frozen blood	62.8±3.4	*
Background of despheral injection	Yes	63.1±16.8	58.7-67.4
	No	60.5±15	51-70
Method of despheral injection	Subcutaneous by pomp	63.5±16.3	59.3-67.7
	Subcutaneous injection Pomp and serum	61.7±14.1	44.1-79.2
Difficulties of injection	Topical reaction and pain	62.04±16.6	57-67.1
	Painful injection	63.8±17	55.3-72.2
	Sensitivity to medicine	61.5±18.04	46.5-76.6
Facial change	No	69.1±17.2	58.1-80.1
	Mild	67.02±17.9	60.2-73.8
	Moderate	55.4±12.2	50.7-60.1
	Serious	62.1±15.9	*

* -Because of insufficient sample volume performance of statistical test was not possible.

4. Discussion

This research indicated that quality of life in a significant number of thalassemia patients in Yasuj is impaired and their emotional role and mental health is most affected quality of life domain, compared with other domains. Also the thalassemia complications are strongly related to patient's quality of life. Therefore to reduce disease complications, appropriate programs such as educating self-care procedures, for improving physical, mental and social problems should be planned.

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