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Sarah H. Siddiqui
Aga Khan University, sarah.hasan@aku.edu

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ORIGINAL ARTICLE

Quality of Life in Patients with Thalassemia Major in a Developing Country

Sarah H. Siddiqui[1], Ruba Ishtiaq[2], Faiza Sajid[3] and Raihan Sajid[4]

ABSTRACT

Objective: To determine the problems faced by thalassemic patients in their personal, psychological and social life.
Study Design: A cross-sectional multi-centre survey.
Place and Duration of Study: Karachi, Lahore and Quetta Centres of Fatimid Foundation, from October 2009 to October 2010.
Methodology: An indigenously developed Qualifty of Life (QoL) questionnaire modified from SF-36 questionnaire was administered to 101 transfusion dependent subjects suffering from thalassemia major. Variables were analyzed using SPSS version 15 for descriptive statistics.
Results: The mean age of the subjects was 10.5 years ranging from 6 - 21 years. Less than one third of the patients felt that their health was slightly worse as compared to last year. Forty five (44%) of the patients felt loneliness due to their disease. Parents of 36 (35.6%) of the children at times did not allow their children to play because of their disease. Twenty eight (27.7%) stated difficulty in mingling with children of their age. Seventy one (70.3%) of the patients reported that at some or all times they were worried about their future life and career while 70 (69.3%) admitted being taken extra care of by their friends and 56 (55.4%) by their teachers.
Conclusion: The quality of life of surveyed thalassemic patients was immensely affected. Having physical impairments, social stresses, financial burdens and problems with their education and career make them very much vulnerable to psychological trauma very early in their life. All of this creates a hindrance in their way of developing into autonomous functioning adults.

Key Words: *Quality of life. Thalassemia major. Social stress.*

INTRODUCTION

In spite of the recent modalities in the management of patients suffering from β-thalassemia major, those living in developing countries do not even receive the standard treatment.[1] For a chronic disease such as thalassaemia, where life long management is required, quality of life, and not just the survival of the patient has become an important aspect for the provision of complete health care.[2] The quality of life, which is an index of health care defined as patient's own assessment and outlook of his illness and accompanying treatment on his daily life including the domains of physical, psychological (including emotional and cognitive) and social functioning,[3] is often constraint in thalassemic patients.[4,5]

[1] *Department of Medicine, The Aga Khan University Hospital, Karachi.*
[2] *Department of Medicine, Civil Hospital, Karachi.*
[3] *Department of Biochemistry, Shaheed Mohtarma Benazir Bhutto Medical College, Lyari, Karachi.*
[4] *Department of Pathology, Al-Faisal University, Riyadh, Saudi Arabia.*

Correspondence: Dr. Faiza Sajid, B-50, Block-12, Gulistan-e-Jauhar, Karachi.
E-mail: faizataha@gmail.com

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The quality of life (QoL) of individuals with thalassemia major is influenced by a number of factors such as the impact of having a chronic disease, physical appearance[6] and burden of treatment modalities including frequent hospital visits for transfusion, nightly subcutaneous infusions of chelating agents,[7] complications of the disease,[4] uncertainties about the future and the expectation of early death resulting from the disease complications.[8]

Although there has been considerable improvement regarding Health Related Quality of Life (HRQoL) in developed countries, there are very few published studies on HRQoL of thalassemic children and adolescents in developing countries[1] inspite of the fact that thalassemias are most prevalent in the Asian developing countries.[9]

This is evident from the fact that after thorough literature search and to the best of authors' knowledge no published data was retrieved from Pakistan targeting the quality of life of thalassemia patients, which is an important component of overall health.

This study was planned to determine the problems faced by thalassemia patients with regards to their personal, psychological and social life.

METHODOLOGY

Data of a total of 101 patients suffering from thalassemia major was collected. It was a cross-sectional study

conducted at 3 centres of Fatimid Foundation, at Karachi, Lahore and Quetta, from October 2009 to October 2010. Convenient sampling was used. Children diagnosed with thalassemia major and on regular blood transfusions between the ages of 6 to 21 years were included in the study. Subjects not on regular blood transfusions, having thalassemia trait, haemoglobin S or C disease, chronic illnesses and mental retardation were excluded from the study.

A Thalassemia QoL questionnaire, modified from the SF-36 questionnaires, was designed and translated into Urdu. It was reviewed by a consultant Psychiatrist at The Aga Khan University and Hospital for validation.

Permission to carry out the study at different centres was obtained. The research associates visited different centres of Fatimid Foundation to approach the subjects (fulfilling the inclusion criteria) while they were undergoing their blood transfusion therapy. They explained the nature and reason for the research to the subjects and their parents. Those who were willing to participate in the study were administered the pre-formed thalassemia QoL questionnaire (with Urdu translation) after taking written consent. The subjects were asked to fill-up the questionnaire themselves and it was collected back by the research associates upon completion. The data was kept confidential and used only for statistical analysis using Statistical Package for Social Sciences (SPSS) version 15. Results were expressed as frequencies and percentages.

RESULTS

Data was collected from a total of 101 patients. Of these, 60 (59.4%) were males and 41 (41.5%) females. The mean age of the subjects was 10.5 years (ranging from 6 - 21 years). All of the subjects were transfusion dependent, with 35 (34.6%) of the patients needing once monthly transfusion, 36 (35.6%) twice monthly and 28 (27.7%) of patients requiring transfusion more than twice a month. No response noted for 2 (1.9%) of subjects.

Regarding the overall general health, the subjects were asked to self-evaluate the change in their health as compared to last year. Fifteen subjects (14.8%) felt that their health was much better as compared to last year, 21 (20.7%) felt their health to be slightly better than last year, 34 (33.6%) reported no change in their health, 18 (17.8%) of the patients felt that their health was slightly worse and 13 (12.8%) rated their health to be much worse as compared to last year.

Regarding psychological functioning, 29 (28.7%) of the patients reported that they often felt despair because of their illness, 27 (26.7%) often felt it as a burden on them, while 21 (20.8%) always remained irritated because of their disease. Forty five subjects (44.6%) of the patients had the feeling of loneliness secondary to their disease.

With regard to physical functioning, 25 (24.8%) of the patients stated that they sometimes feel weaker because of their disease. Around one quarter of the patients often could not participate in sports, while parents of 36 (35.6%) of the children at times did not allow their children to play because of their disease.

Table I.

	Yes	No	Never	Rarely	Sometimes	Often	Always	No response
Psychological aspect:								
I feel sad because of my disease	-	-	34 (33.7%)	3 (30%)	26 (25.7%)	32 (31.7%)	6 (5.9%)	0 (0.0%)
Disease is a burden for me	-	-	36 (35.6%)	0 (0.0%)	21 (21.8%)	27 (26.7%)	17 (16.8%)	0 (0.0%)
I feel angry/ irritated due to my disease	-	-	28 (27.7%)	2 (2.0%)	23 (22.8%)	27 (26.7%)	21 (20.8%)	0 (0.0%)
I feel lonely because of my disease	45 (44.6%)	41 (40.6%)	-	-	-	-	-	15 (14.8%)
I feel despair due to my disease	-	-	29 (28.7%)	2 (2.0%)	30 (29.7%)	29 (28.7%)	10 (9.9%)	1 (0.9%)
Physical aspect:								
I feel weaker because of my disease	-	-	41 (40.6%)	2 (2.0%)	25 (24.8%)	21 (20.8%)	12 (11.9%)	0 (0.0%)
Cannot participate in sports because of my disease	-	-	29 (28.7%)	6 (5.9%)	28 (27.7%)	25 (24.8%)	7 (6.9%)	6 (5.9%)
Social aspect:								
Have difficulty in mingling with other children	-	-	28 (27.7%)	3 (3.0%)	30 (29.7%)	28 (27.7%)	12 (11.9%)	0 (0.0%)
Can talk freely to my friends about my illness	66 (65.3%)	30 (29.7%)	-	-	-	-	-	5 (4.9%)
Hide my disease from my friends	31 (30.7%)	65 (64.4%)	-	-	-	-	-	5 (4.9%)
Friends take care and support me	70 (69.3%)	20 (19.8%)	-	-	-	-	-	11 (10.8%)
Family aspect:								
Parents have a different attitude compared to other siblings	-	-	12 (11.9%)	2 (2.0%)	10 (9.9%)	35 (34.7%)	42 (41.8%)	0 (0.0%)
Parents have financial problems due to my illness	-	-	23 (22.8%)	1 (1.0%)	20 (19.8%)	28 (27.7%)	27 (26.7%)	2 (1.9%)
Parents remain careful about me	-	-	6 (5.9%)	1 (1.0%)	11 (10.9%)	34 (33.7%)	45 (44.6%)	4 (3.9%)
Parents have problems because of me	-	-	15 (14.9%)	1 (1.0%)	21 (20.8%)	34 (33.7%)	24 (23.8%)	6 (5.9%)
Parents don't allow me to play due to illness	-	-	17 (16.8%)	6 (5.9%)	36 (35.6%)	28 (27.7%)	9 (8.9%)	5 (4.9%)
Educational aspect:								
School is affected because of my disease	-	-	12 (11.9%)	2 (2.0%)	32 (31.7%)	24 (23.8%)	9 (8.9%)	22 (21.7%)
Teachers are aware of my disease	67 (66.3%)	8 (7.9%)	-	-	-	-	-	26 (25.7%)
Teachers support me due to my illness	56 (55.4%)	16 (15.8%)	-	-	-	-	-	29 (28.7%)
I am worried about my future	-	-	28 (27.7%)	1 (1.0%)	27 (26.7%)	20 (19.8%)	23 (22.8%)	2 (1.9%)

Considering the social life of these patients, 28 (27.7%) stated that because of their disease, they often had difficulty in mingling with children of their age, 30 (29.7%) could not talk freely about their disease to their friends. However, 70 (69.3%) of the patients agreed that their friends take extra care of them because of their disease.

Financial issues have been well documented throughout the world for any chronic illness. Contrary to this, only 23 (22.8%) of these patients believed that their family never faced financial problems because of them. The rest were aware of the fact that financial burden increased to some extent on their families because of their disease.

Concerning the life of thalassemia patients at school, only 12 (11.9%) reported that their school was never affected because of their disease. Teachers of 67 (66.3%) were aware about their students' illness and 56 (55.4%) of these children were of the view that their teachers supported them in one way or the other, because of their disease.

Approximately 71 (70.3%) of the patients reported that at some or all times they were worried about their future life and career (Table I).

DISCUSSION

Beta-thalassemia major is a serious life-limiting and potentially life-threatening condition[10] that causes substantial disruption in education and social activities.[11] This study found that many of these patients suffered from sadness, anger and loneliness. This might be due to the chronicity of the disease state that influences patient's recreational activities, capabilities and peer as well as family relationships, culminating in anxiety, secluded behavior and depression.[12,13] The lowered psychosocial health score of thalassemia patients than those of healthy individuals is consistent with the findings of other studies also.[14-16]

This study indicated that around more than half of the children reported that their school was affected in one way or the other at some or most of the times. This may probably be due to their frequent hospital visits for blood transfusion, and nightly subcutaneous injections of iron chelation therapy causing disturbed sleep[17] and/or treatment of complications as was reported in various other studies.[1,18] Cantan *et al*. has suggested that missing school might be a contributing factor of these patients in pursuing higher education.[19] Continuous absenteeism from school contributes to poor academic performance which in turn might lead to psychosocial problems[20] such as isolation and depression.

Most of the parents in this study were found to be cautious about their thalassemic children. This over protective nature of parents of thalassemic children was also reported by Mazzone *et al*.[4]

It was also found that parents faced problems because of their suffering thalassemic children as was also stated in a recent study that parents avoided social get-together and even had strained relations with each other.[21] Also, the finding that only few children could participate regularly in sports without any hindrance indicates the stress which their parents experience due to their illness.[18,22]

In this study, more than one-third of the thalassemia patients have agreed that their parents do face financial problems because of their life-long illness. This is consistent with other studies which reported that Thalassemia patients are a source of socio-economic burden on their families and societies.[23,24]

It was found that most of the thalassemic patients were concerned about their future. This might be due to their fear of possible complications of the chronic illness and can significantly contribute to more psychosocial problems.[25]

This study, being the first in our country to assess the health related quality of life of children with a chronic disease such as thalassemia, focuses on the view that having physical impairments, social stresses, financial burdens and problems with their education and career make them very much vulnerable to psychological trauma very early in their life. All of this creates a hindrance in their way of developing into autonomous functioning adults. We recommend that the conventional medical therapy for these patients should be accompanied by a psychological support system so as to improve the quality of life of these patients so that they can also lead a near-to healthy and blissful life.

CONCLUSION

The quality of life of surveyed thalassemic patients was immensely affected. Having physical impairments, social stresses, financial burdens and problems with their education and career make them very much vulnerable to psychological trauma very early in their life. All of this creates a hindrance in their way of developing into autonomous functioning adults.

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