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Quality of life in patients with thalassemia

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Abstract

Background: Thalassemia major is a chronic disease that presents as a range of serious clinical and psychological challenges. The effect of thalassemia on physical health can lead to deformity, growth retardation and delayed puberty. We conducted this study to assess quality of life of thalassemia major patients registered in thalassemia treatment centers.

Methods: This descriptive cross-sectional study was done in TTC (Thalassemia Treatment Center) from November 2021 to December 2021 after ethical approval from Institutional Research Committee. A total of 120 thalassemia patients ranging from 5 to 28 years of age brought for blood transfusion were included. Patients having impaired cognitive functions and those having multiple comorbidities were excluded. A modified version of WHO QOL BREF questionnaire was developed to assess the physical, psychological, environmental and social aspects of Quality of Life. Data was entered on SPSS v 22 and descriptive statistics were applied.

Results: Out of 120 patients surveyed, majority were females (55.83 %) with a mean age of 10.22± 4.5 years. Forty four percent of the patients were males with mean age of 11.42± 5.19 years. Quality of life of these patients was found to be 68.8%. In females it was 69.34 % and in males it was 68.15%. Quality of life was significantly associated with psychological ($p<0.05$), social ($p=0.009$) and physical ($p<0.05$) factors. Poorer quality of life was found against environmental (financial resources, transport, healthcare) aspects (63.13%). Adverse impact of thalassemia was also found on physical (67.88%) and psychological (68.82%) quality of life of patients. Social Quality of life was found to be (92.3%).

Conclusion: Poor psychological support along with other environmental and physical aspects lead to lower Quality of life of β thalassemia major

1. INTRODUCTION

Thalassemia as the most common genetic disorder worldwide is regarded as a serious problem in public health issues in the Mediterranean region (1). Iran is located in the geographical belt of thalassemia and it has been estimated that thalassemia carriers vary from one to ten percent (with a mean of 4.5%) in different parts of Iran (2). Although, morbidity and mortality of the thalassemia major has been reduced significantly in the light of modern medical treatment, however, it could influence diverse aspects of patients' lives. Some aspects of thalassemia major and its associated complications are expected to impact on the QOL. the diagnosis and treatment of the thalassemia major could have an impact on family stability and family dynamics and bone deformities and short stature could induce poor self-image. Also, frequent hospital visits for transfusion, nightly subcutaneous infusions, delayed or absent sexual development and impaired fertility and complications such as heart disease, bone disease, diabetes, infections and Uncertainties about the future and difficulties in long-term planning could be mentioned as a result of thalassemia major (3). The World Health Organization (WHO) defined the quality of life as: "An individual perception of their

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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 in a complex way by the person's physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment (4). There are few inconsistent studies about the QOL in patients with thalassemia major and most of these investigations obtained the results based on interviews with patients, careers, doctors and nurses with the focus on coping strategies and they did not mention control group. Previous investigations showed that treatment and cultural differences did not have a major effect on the QOL (5). Also, results indicated that patients had moderately impaired overall health and overall QOL (6) and other serious hemoglobinopathies such as sickle cell disease (SCD) might induced poor QOL (7-10) However, there might be differences in the domains affected as well as the extent of variation across specific chronic disorders (11) Quality of life of patients with thalassemia has been reported in Europe and the United States; however, the results show variations due to cultural and socio-economic differences in the targeted populations (12). Quality of Life of individuals with thalassemia is influenced by a number of factors such as burden of having a chronic disease, physical appearance, treatment modalities including multiple visits to the hospitals for transfusions, complications of the disease, uncertainty regarding the future and expectation of early death (13,14). There has been improvement in HRQOL in thalassemic patients in developed countries but there are very few published studies on QOL of thalassemic patients in developing countries such as Pakistan where the prevalence is quite high. This study was therefore designed to assess the Quality of Life of thalassemic patients in physical, psychological, social and environmental domains of their lives.

2. MATERIALS AND METHODS:

It was a cross sectional study conducted in the Thalassemia Treatment Center of Holy Family November 2021 to December 2021 after taking ethical approval from Institutional Research Committee. Non-probability Convenient sampling was done and 100 patients were included in this study. Patients ranging in age from 10 to 30 years diagnosed with thalassemia major who were on regular blood transfusions were included. Patients with impaired cognitive skills; those with multiple comorbidities and those unwilling to participate were excluded. A modified version of WHO QOL BREF questionnaire was used. The questionnaire was translated into Urdu prior to the data collection. A pilot study was conducted on 10 patients; statistics were applied on that data and then questionnaire was administered to the participants. The questionnaire included 20 questions that were developed to evaluate the Quality of Life in 4 domains (physical, psychological, environmental and social). Physical Quality of Life was assessed by questions related to pain, sleep and tiredness. Psychological QOL was assessed using questions related to depression, negative feelings, confidence in treatment, satisfaction and concentration. Social QOL include relationship with family members and loneliness. Environmental QOL included financial difficulties, healthcare and transport difficulties. Likert scale 5 was used to score individual questions. Scores were scaled in a positive direction i.e.; higher score denotes higher quality of life. The mean score of questions within each domain was used to calculate the domain score. Domain scores were then summed to get the overall Quality of Life score. Data were entered on SPSS v22. Frequencie were calculated for age, gender, residence and educational status. (Table III) Percentages were calculated for Quality-of-life scores. (Table I) T-test and one-way

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ANOVA test were applied to find statistical significance. P-valu <0.05 was considered statistically significant.

3. RESULTS

Section I: Demographic Data

A total of 100 patients were included in this study; fifty-seven were females with a mean age of 15.22±4.5 years whereas 43 were males with a mean age of 16.42 ± 5.19 years. Forty five percent of the patients lived in rural areas. All of the patients were transfusion dependent. fifteen percent were receiving once weekly transfusions, 55% twice a month, 6% thrice a month and 24% were receiving once monthly transfusions. Blood groups of the participants were as: A+ve (20%), B+ve (40%), AB+ve (10%), O+ve (10%), A-ve (8%), B-ve (2%), AB-ve (8%), O-ve (2%). Quality of life was calculated to be 69.8%. It was 70.34% in females and 69.15% in males. Quality of life in its four domains was calculated as; Physical QOL (65.88%), psychological (70.82%), social (93.3%) and environmental (65.13%). Patients living in urban areas had a mean quality of life score of 72% and those living in rural areas had mean score of 67%.

QOL	SCORE
Physical	65.88%
Psychological	70.82%
Environmental	65.13%
Social	93.3%
Overall	73.78%

Table I: Quality of life in Thalassemia Patients

Section II: Quality of Life Data

While assessing physical QOL, it was observed that 72.8% of the patient's endured pain and 36.8% of the total suffered from extreme amount of pain during their course of treatment. Significant association was found for pain (p=0.013) with overall quality of life. Sleep difficulties were found in about 31 .8% of the patients, 41% of these suffered from extreme amount of sleep difficulties. Majority of patients said that they get too tired at times and are unable to carry out routine chores. Sixteen percent of patients suffered from extreme tiredness during the course of their treatment. Similarly, psychological QOL showed depression, anxiety and lack of confidence in the patients. About 18.5% of the patients had no confidence in success of their treatment and another 26.16% had very little confidence. Social QOL was found to be better than other domains. Considering financial difficulties, 62 out of 100 suffered from financial difficulties and 25.7% of these suffered from severe financial crisis during the transfusions. Disease affected the education of patients; 31% of the total could never study, 55 .5% studied up to primary school, 7.3% in secondary school and only 3.2% studied beyond secondary school. Seventeen percent of the patients were not confident about the success of the treatment they were receiving. Sixty three percent of the patients were worried about their disease and had low confidence in success of their treatment. Sixty one percent of the patients had negative feelings and 48.16% had marked feelings of depression and sadness. Significant association was found for pain (p=0.01), distress due to pain (p<0.01),

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tiredness (p=0.009), sleeping difficulties (p=0.07), worry due to disease (p=0.01), depression or sadness (p<0.05), anxiety and negative thoughts (p<0.01), satisfaction with health (p=<0.01), satisfaction with life (p=0.08).

Section III: Association of Demographic Variable with QOL

Quality of life of patients was found to be affected by number of blood transfusions patients per month. Those receiving monthly blood transfusions had better Quality of scores as compared to those receiving biweekly. Quality of life scores of patients receiving blood transfusions once, twice, thrice and four times a month were found to be 72.16%, 71%, 67.09% and 69.8% respectively. Quality of life score was found to be significantly associated with transfusion interval of patients (p=0.005).

4. DISCUSSION

Quality of life is an indicator of overall wellbeing of an individual and has become an important tool to assess the impact of a chronic disease on various aspects of patients' life. There have been a limited number of studies on health-related quality of life of thalassemia patients. Thalassemia major is a condition that deleteriously affects the life activities of patients (15). This study found that patients suffering from thalassemia suffer from psychological stress and a number of physical impairments such as enduring severe amount of pain, tiredness and sleep difficulties. Thalassemia being a chronic disease restricts life activities, capabilities and relationships of patients leading to negative impact on physical, psychological and social quality of life. Regular blood transfusions are required to maintain normal hemoglobin levels in patients with thalassemia major. Patients' quality of life is strongly influenced by the number of transfusions they receive in a month; the transfusion interval. Greater the transfusion interval, better the quality-of-life scores and vice versa. Shorter transfusion interval is associated with complications related to iron overload. Such patients are at increased risk of organ damage such as heart failure. Multiple transfusions put a lot of financial burden on patients. Multiple hospital visits, non-availability of blood and transport difficulties add to the distress of patients. Those receiving once monthly transfusions had better quality of life scores as compared to those receiving multiple transfusion per month. In a study conducted in Egypt using Pediatric Quality of Life Inventory 4.0 Generic Core Scale, the mean quality of life score was 63.74(SD 13.2). Physical domain score was 58.46(SD 18) (16) Similarly, in another study conducted in Iraq, the lowest scores were encountered in physical domain at 61% (SD 13.37). (17) In these studies, physical domain scores are as compared to this study. This is due to the fact that the mean age of patients included was higher as compared to this study. With increase in age there is increase in complications associated with thalassemia major such as organ failure and thus lower quality of life scores. In contrast to studies conducted, significant association was found with residential address of patients. Patients living in rural areas had low quality of life scores. This can be explained by the fact that thalassemia treatment centers are very few in number and that too are located in big cities. Patients face transport difficulties as they travel to cities for blood transfusions. On the contrary, those living in urban areas have access to transfusion centers and face lesser difficulties so their quality of life was found to better. In a few studies conducted, it is shown that psychological and social aspects of life of thalassemia patients are affected adversely. This study showed that patients had negative feelings, anxiety and lack of confidence in the success of treatment. Patient suffer from psychological trauma

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that is attributed to the chronic nature of disease limiting the life of patients. Regarding family relationships, patients felt happy about their relationship with their family members. This may be due to the protective and caring attitude of parents towards their sick children. Education was also affected that is also attributable to the chronic nature of disease; limiting their normal study routine. Barrier to proper education is also attributed to the physical stresses such as extreme amount of pain and tiredness experienced by the patients. In this study, psychological QOL assessment showed that patients were uncertain about the success of their treatment. This study was done to determine the Quality of Life of thalassemia patients where there have been very few previous studies. This study focuses on the view that physical impairments, stresses, financial burden and problems with education make thalassemia patients vulnerable to psychological and trauma very early in their life.

5. CONCLUSION

Quality of life is significantly reduced in majority of the thalassemia patients. Apart from conventional therapy, psychological support programs should be developed to improve the Quality of Life of thalassemia patients.

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