Health-Related Quality of Life in Transfusion-Dependent Thalassemia Major Patients and Associated Factors in Dubai, UAE, 2011

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Abstract

Background: Exploring knowledge about and associated factors of health-related quality of life (HRQOL) among patients with thalassemia is essential in developing more suitable clinical, counseling, and social support programs to improve treatment outcomes of these patients.

Objectives: To investigate knowledge about HRQOL in transfusion-dependent thalassemia major patients, and to identify the socio-demographic and disease related factors that affect their quality of life (QOL).

Methods: A cross-sectional study was conducted to assess the QOL of 279 transfusion-dependent thalassemia major patients (≥ 13 years) who had no co-morbidities other than disease or transfusion related complications and factors affecting it. The PedsQL 4.0 Generic Core questionnaire was administered. It consists of 23 items, and has physical, emotional, social, school and work dimensions.

Results: Overall QOL mean score (±SD) was 78.88 (± 13.14). Out of 5 predictors affecting the total score (type of diagnosis, annual serum ferritin, number of complications, diabetes mellitus and (hypogonadism) sexual growth complications) it was found that the total summary score was positively predicted by diagnosis (p=0.018); the total quality of life in thalassemia intermediate was better than thalassemia major patients, and it was negatively affected by mean annual serum ferritin level (p=0.014); the higher the serum ferritin level the lower the total QOL score.

Conclusion: The thalassemia patient’s quality of life at Dubai Thalassemia Center didn’t appear different from others around the world. It was affected by serum ferritin, number of complications, type of iron chelators and professional job. While cardiac complication impacted the physical dimension negatively, diabetes has impacted the psychosocial dimension. These data may help in implementing interventions focused on the affected dimensions.

Key words: Quality of Life, transfusion dependent, thalassemia major
Background

Quality of life (QOL) is a broad ranging concept affected in a complex way by the person’s physical health, psychological state, level of independence, social relationships, personal beliefs and relationship with the salient features of his/her environment(1). It is considered an important index of effective treatment. An assessment of QOL differs from other forms of medical assessment in that it focuses on the individuals’ own views of their well-being and assesses other aspects of life, giving a more holistic view of wellbeing(2).

Thalassemia does not only affect patients’ physical functioning, emotional functioning, social functioning and school functioning(3-9) but also their ability to find partners, establish family (due to infertility), find a career and obtain social support, so patients with transfusion dependent thalassemia tend to have impaired health-related quality of life (HRQOL)(7).

It is now possible for a thalassemia patient to have a near normal life span with a good HRQOL(10) with the availability of better transfusion regimen, iron chelating therapy, proper management of complications and good supportive care. As a result, attention has shifted to the wellbeing of the patients with thalassemia. HRQOL should now be considered an important index of effective health care in thalassemia.

There is scarcity in published work on evaluation of QOL in thalassemia patients(2,11,12). Research on thalassemia, in general, starts from hard outcome measures that are based on the medical aspect of the disease and on the impact of regular blood transfusion as well as chelating agents’ maintenance treatment on the patients QOL, with limited attention to thalassemia individuals’ own perspectives and values about their life. Attention to treatment outcomes should no longer be restricted to the reduction and elimination of direct health-consequences, but should start from a broad perspective, including the improvement of thalassemia patients’ physical and mental health, their social functioning and overall well-being(13) since QOL of thalassemia patients is a crucial point that needs to be placed in the forefront in their management, and also clients’ subjective perspectives on QOL that are seldom heard need to be investigated for their usefulness as a central assessment and outcome measure in the care and support provided for them.

Objectives

1. To measure the total quality of life of the adult and adolescent thalassemia patients in Dubai thalassemia center.
2. To identify some of the contributing factors affecting the quality of life of thalassemia patients in Dubai thalassemia center which include socio-demographic and disease related factors.

Subjects and Methods

A cross sectional study design was conducted on transfusion dependent thalassemia patients who are on follow-up at the Dubai Thalassemia Centre. All thalassemia patients of ≥ 13 years in the Dubai Thalassemia Center (n=279) as (adolescents and adults) were subjected to interview questionnaire which included 4 sections: socio-demographic data, disease related data, assessment of patient self-perception of health, and assessment of QOL. QOL Inventory version 4 (Peds QL 4.0 Generic Core Scales) is a modular instrument for measuring HRQOL divided into 6

<table>
<thead>
<tr>
<th>Domain / Item (No = 279)</th>
<th>Almost always No (%)</th>
<th>Often No (%)</th>
<th>Sometimes No (%)</th>
<th>Almost never No (%)</th>
<th>Never No (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Walking</td>
<td>2 (0.7)</td>
<td>13 (4.7)</td>
<td>22 (7.9)</td>
<td>17 (6.1)</td>
<td>225 (80.6)</td>
</tr>
<tr>
<td>Running</td>
<td>20 (7.2)</td>
<td>20 (7.2)</td>
<td>62 (22.2)</td>
<td>30 (10.8)</td>
<td>147 (52.7)</td>
</tr>
<tr>
<td>Making activity</td>
<td>19 (6.8)</td>
<td>14 (5)</td>
<td>36 (12.9)</td>
<td>38 (13.6)</td>
<td>172 (61.6)</td>
</tr>
<tr>
<td>Lifting something</td>
<td>15 (5.4)</td>
<td>25 (9)</td>
<td>45 (16.1)</td>
<td>34 (12.2)</td>
<td>160 (57.3)</td>
</tr>
<tr>
<td>Taking a bath</td>
<td>4 (1.4)</td>
<td>-</td>
<td>-</td>
<td>6 (2.2)</td>
<td>269 (96.4)</td>
</tr>
<tr>
<td>Doing chores</td>
<td>3 (1.1)</td>
<td>6 (2.2)</td>
<td>15 (5.4)</td>
<td>24 (8.6)</td>
<td>231 (82.8)</td>
</tr>
<tr>
<td>No pain</td>
<td>8 (2.9)</td>
<td>28 (10)</td>
<td>74 (26.3)</td>
<td>46 (16.5)</td>
<td>123 (44.1)</td>
</tr>
<tr>
<td>Having energy</td>
<td>5 (1.8)</td>
<td>32 (11.5)</td>
<td>78 (28)</td>
<td>37 (13.3)</td>
<td>127 (45.5)</td>
</tr>
</tbody>
</table>

Table 1: The frequency of the physical domain items of the quality of life in thalassemia patients attending Dubai Thalassemia Center DHA 2011
different age groups (2-4, 5-7, 8-12, 13-18, 19-25 and ≥ 26 years). User agreement was signed with MAPI Research Institute, Lyon, France prior to using the tool. It's a multidimensional tool of child self-report, parent proxy-report and adult scales. It consists of 23 items applicable for healthy school and community populations, as well as pediatric populations with acute and chronic health conditions. The questionnaire dimensions are: Physical, Emotional, Social, and School/Work.

**Results**

Table 1 illustrates the physical functioning of the studied thalassemia patients. It was found that 96.4% of the patients never had problems taking a bath alone. As regards walking and doing chores, 80.6% & 82.2% respectively never had problems. As regards activities and exercises 61.6% never had problems. According to the patients, 57.3% never had problems lifting something heavy. On the other hand, 52.7% of the studied thalassemia children never had problems running. Regarding energy and pain, 45.5% and 44.1% respectively of the patients never had problems.

Table 2 illustrates the emotional, social and school/work functioning domains of the studied thalassemic patients. According to the patients, it was found that 70.3% of the patients never felt afraid. In relation to having sleeping trouble, 54.1% % never had this problem. Regarding sadness, 49.1% never had this problem. Regarding worry and angry, 42.3% and 30.8% respectively of the

<table>
<thead>
<tr>
<th>Domain / Item (No =279)</th>
<th>Almost always No (%)</th>
<th>Often No (%)</th>
<th>Sometimes No (%)</th>
<th>Almost never No (%)</th>
<th>Never No (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Emotional</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Being afraid</td>
<td>3 (1.1)</td>
<td>6 (2.2)</td>
<td>43 (15.4)</td>
<td>31 (11.1)</td>
<td>196 (70.3)</td>
</tr>
<tr>
<td>Being sad</td>
<td>8 (2.9)</td>
<td>30 (10.8)</td>
<td>70 (25.1)</td>
<td>34 (12.2)</td>
<td>137(49.1)</td>
</tr>
<tr>
<td>Being angry</td>
<td>23 (8.2)</td>
<td>50 (17.9)</td>
<td>87 (31.2)</td>
<td>33 (11.8)</td>
<td>86 (30.8)</td>
</tr>
<tr>
<td>Sleeping trouble</td>
<td>28 (10)</td>
<td>27 (9.7)</td>
<td>41 (14.7)</td>
<td>32 (11.5)</td>
<td>151 (54.1)</td>
</tr>
<tr>
<td>Being worried</td>
<td>34 (12.2)</td>
<td>30 (10.8)</td>
<td>58 (21.1)</td>
<td>38 (13.6)</td>
<td>118 (42.3)</td>
</tr>
<tr>
<td><strong>Social</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Getting along with others</td>
<td>2 (0.7)</td>
<td>9 (3.2)</td>
<td>10 (3.6)</td>
<td>26 (9.3)</td>
<td>232 (83.2)</td>
</tr>
<tr>
<td>Others refuse them</td>
<td>-</td>
<td>4 (1.4)</td>
<td>12 (4.3)</td>
<td>17 (6.1)</td>
<td>246 (88.2)</td>
</tr>
<tr>
<td>Teasing from others</td>
<td>5 (1.8)</td>
<td>11 (3.9)</td>
<td>21 (7.5)</td>
<td>25 (9)</td>
<td>217 (77.8)</td>
</tr>
<tr>
<td>Can't do things as others</td>
<td>8 (2.9)</td>
<td>21 (7.5)</td>
<td>41 (14.7)</td>
<td>26 (9.3)</td>
<td>183 (65.6)</td>
</tr>
<tr>
<td>Keeping up with others</td>
<td>3 (1.1)</td>
<td>6 (2.2)</td>
<td>15 (5.4)</td>
<td>21 (7.5)</td>
<td>234 (83.9)</td>
</tr>
<tr>
<td><strong>School/work</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pay attention in class/work</td>
<td>7 (2.5)</td>
<td>17 (6.1)</td>
<td>48 (17.2)</td>
<td>29 (10.4)</td>
<td>178 (63.8)</td>
</tr>
<tr>
<td>Forgetting things</td>
<td>12 (4.3)</td>
<td>45 (16.1)</td>
<td>83 (29.7)</td>
<td>51 (18.3)</td>
<td>88 (31.5)</td>
</tr>
<tr>
<td>School work/ work</td>
<td>4 (1.4)</td>
<td>13 (4.7)</td>
<td>34 (12.2)</td>
<td>41 (14.7)</td>
<td>178 (67)</td>
</tr>
<tr>
<td>Missing school/work due to illness</td>
<td>21 (7.5)</td>
<td>16 (5.7)</td>
<td>53 (19)</td>
<td>40 (14.3)</td>
<td>74 (26.5)</td>
</tr>
</tbody>
</table>

Table 2: The frequency of the psychosocial domain items of the quality of life in thalassemia patients attending Dubai Thalassemia Center DHA 2011
studied patients never had this problem.

Patients who never had problems regarding acceptance of themselves from others as friends constituted 88.2%. Regarding keeping up with others and getting along with others, 83.9% and 83.2% respectively of patients never had this problem. Patients who were not teased by others constituted 77.8%. In relation to the ability of doing things the same as others, 65.6% of patients never had this problem.

Regarding keeping up with school/work, 67% of patients never had this problem. Regarding paying attention in class/work, according to patients it was shown that 63.8% of patients never had this problem. Regarding missing school/work to go to hospital, 33% of patients had this problem. In relation to forgetting things, 29.7% of patients sometimes had this problem. In relation to missing school/work due to illness, 26.5% of patients never had this problem. The first three items related to school or work functioning have a cognitive component, while the others are more related to physical aspects.

Table 3 demonstrates the overall quality of life and the general health satisfaction of the thalassemic patients. The subjective self-perception of the thalassemia patients for their quality of life was highest at good score with 44.4% then at very good with 34.4% and least at very poor with 1.8%. While the self-perception of the thalassemia patients of their general health satisfaction was highest at satisfied with 52% then at very satisfied with 29.7% and least at very dissatisfied with 2.2%.

Table 4 demonstrates the descriptive statistics of the quality of life scale by domain comparing the domains together using the mean and standard deviation revealed that the mean was 82.01 (±16.889) in the physical domain and the psychosocial domain mean was 77.09 (±14.178). The highest mean of the psycho-social sub-domains was the social domain with 90.36 (±13.626) then the emotional domain with 71.67 (±21.761) and the least was the school or work domain with 69.19 (±20.272). The total quality of life mean was 78.88 (±13.14).

Table 5 demonstrates stepwise linear regressions of factors affecting QOL of thalassemia patients attending Dubai Thalassemia Center at Dubai Health Authority, 2011. The stepwise multiple linear regression analysis was applicable to examine factors affecting the physical summary scores; out of 5 predictors affecting the physical summary score (age, duration of the transfusion, annual serum ferritin, number of complications and heart failure) it was found that physical health summary score was negatively predicted by the Mean annual serum ferritin level (p=0.008); the higher the serum ferritin level the lower the physical QOL score. While the psychological summary score was positively predicted by the diagnosis (p=0.018); the psychosocial quality of life in thalassemia intermediate was better than thalassemia major patients, and it was negatively predicted by diabetes (p=0.025); the presence of diabetes will worsen the QOL. However, out of 5 predictors affecting the total score (type of diagnosis, annual serum ferritin, number of complications, diabetes mellitus and (hypogonadism) sexual growth complications) it was found that the total summary score was positively predicted by diagnosis (p=0.018); the total quality of life in thalassemia intermediate was better than thalassemia major patients, and it was negatively affected by mean annual serum ferritin level (p=0.014); the higher the serum ferritin level the lower the total QOL score.

Figure 1 Box and whisker plot (page 8) presentation of the QOL (total and domains) of thalassemia patients attending Dubai Thalassemia Center.
at Dubai Health Authority, 2011 is demonstrated. It was found that the mean (SD) of the Physical Summary Score was 82.01 (16.889), the median was 88 with minimum of 9 and maximum was 100; the first quartile (Q1) was 72 and the third quartile (Q3) was 94. While it was found that the mean (SD) of the Psychosocial Summary Score was 77.09 (14.178), the median was 79 with minimum of 32 and maximum was 100; the first quartile (Q1) was 70 and the third quartile (Q3) was 87. However it was found that the mean (SD) of the Total Score was 78.78 (13.196), the median was 81 with minimum of 25 and maximum was 100; the first quartile (Q1) was 71 and the third quartile (Q3) was 88.
Discussion
In spite of the fact that thalassemia has been a known disease for decades, the quality of life of the thalassemia patients is a still poorly studied and discovered area and with the great development and discoveries in the management modalities of the patients, their survival is now prolonged, so now the focus has shifted from prolonging life toward trying to give the patient a better QOL. This study examined the relationships of socio-demographic characteristics, disease related data and QOL among adult and adolescent thalassemia patients. It filled a gap in the literature related to adult and adolescent thalassemia patients and QOL. The findings are important for health care systems and providers. There have also been discussions among physicians and health researchers about the importance of patients' perspectives on their own health.

In this cross-sectional single-institution study of HRQOL with thalassemia in Dubai Thalassemia Center, the PedsQL 4.0 Generic Core Scale self-report was chosen as the research instrument as it incorporates the dimensions necessary for measuring the HRQOL of 5 different age groups in the population and has been tested for validity and reliability (14-16).

There has not been a previous study to measure the HRQOL of thalassemia patients in adolescent and adult ages in our region. This is the first study to provide evidence that the HRQOL of thalassemia patients is affected by the disease.

The QOL in this study showed that psychosocial health had a lower score than physical health. It is possible that their anxiety or depressive feeling, low self-confidence and esteem as well as frequent school/work missing or unemployment and suffering from chronically disability condition had a negative impact on patient's perspective. This finding is similar to a previous study done by Ismail A. et al; 2006 and Thavorncharoensap M. et al; 2010 (4, 8). But the current study scores were slightly higher than a recent report done by Wisai M. et al; 2009 and Pacharapan S. et al; 2010 (17, 18).

Direct comparisons to previous studies done by Wisai M. et al; 2009 and Pacharapan S. et al; 2010 (17, 18) found that the mean total Peds QLTM scores (SD) was found to be comparable to the total scores (TSS) obtained from this study. Nevertheless, the scores were found to have a significant difference, compared to Ismail A. et al; 2006 (4). It could be explained by the difference in disease severity, treatment of patient, differences in cultures, experiences and perspectives as well as a different age group, that contributed to different health perceptions and needs.

The emotional score in this study was found to be almost similar to other studies done by Varni et al; 2002(15) on oncology patients (acute lymphocytic leukemia, brain tumor, non Hodgkin lymphoma, Hodgkin lymphoma and other cancer) and Upton et al; 2005 (19) on asthma, cancer and diabetic patients. However it was higher than a study done by Ismail A, Dahlu M and El Dakhakhny A but it was lower than the studies done by Renifjell T, Thavorncharoensap M and Azarkeivan A. This can be contributed to the fact that this study has a wider age range than the others so young children may have difficulty expressing their emotions directly; fear of chronic diseases like thalassaemias that do not have a definite cure can be one of the important factors leading to these psychological reactions. Illness-induced anxieties and depression.
among these patients have been well-documented, Shaligram D et al; 2007, Roy T et al; 2007, Gharaiyeh H et al; 2009 and Khurana A et al; 2006 studies (6, 20-22).

The social score in this study was found to be the highest among all the assessed scores. This could be attributed to the fact that the patients adapted relatively well to their disease despite that some had reported an episode of being teased by others and difficulty to do the same things others do. Nevertheless difficulty in recalling episodes of stigmatization, may be due to refusal to declare they are facing a problem or that they refused to be treated differently so they don’t inform the surrounding people about their disease, play a role also in this. This finding contradicts a wide range of studies done by Ismail A. et al; 2006 and Thavorncharoensap M. et al; 2010 (4, 8) who reported it lower than in this study. However Azarkeivan A et al; 2009 and El Dakhakhny A M et al; 2011 reported it higher than the current study (11, 23).

The school or work score was the lowest scored in the QOL assessment done on the patients. This can be attributed to the fact that the patients miss their school or work to come for their transfusion which is done once every 3-4 weeks according to their hemoglobin level and to the amount of blood received on the last transfusion. This study was higher than studies done by Ismail A. et al; 2006 and Thavorncharoensap M. et al; 2010 (4, 8) but lower than others done by Cheuk D K L et al; 2008, El Dakhakhny A M et al; 2011 and Reinfjell T et al; 2006 (23-25).

Conclusion

1. QOL assessment promotes understanding of the disease and initiating intervention programs.
2. Physical and emotional dimensions of QOL assessment were the most affected dimensions with emotional being affected more by the predictors.
3. The school or work dimension was the lowest scored in the QOL assessment.

Recommendations

1. Establishing psychosocial and counseling programs aiming at helping patients discuss and accept their illness, facilitating a normal lifestyle, and providing a link between patients, school or work officials, the family and the physician may be helpful in alleviating these difficulties, especially academic performance.
2. Carrying out QOL assessment routinely for all thalassemia patients so that interventions focused on affected dimensions can be implemented;
3. Matched control analysis and QOL assessment is required for better understanding and improvement.

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