# Quality of life among adults with beta-thalassemia major in western Saudi Arabia

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## ABSTRACT

**Objectives:** To assess the quality of life in the thalassemia adult patients and clarify how effective the management is of these patients and whether a change in care is warranted.

**Methods:** In this cross-sectional study, adult thalassemia patients (≥18 years) of both genders, attending the day care unit in King Abdulaziz University Hospital, Jeedah, Saudi Arabia were surveyed using SF-36 questionnaire. Data were collected between October 2012 and December 2012. The questions highlighted 3 health status scales; physical functioning (PF), emotional functioning (EF), and social functioning (SF). Scores were analyzed using SPSS.

**Results:** Forty-eight adults were surveyed (mean±SD: 26.02±5.56). These were made up of 60.4% males and 41.7% were Saudis. The frequency of blood transfusion was every 3 weeks in 81.3% of patients, but 18.8% were having transfusions less frequently. Half of our sampled patients were splenectomized (54.2%). The PF score for the total sample was 61.4 (SD=22.7), the SF score was 75 (SD=26.4) and the EF score was 69.7 (SD= 21.6); the SF and EF scores were lower in females and non-Saudis compared to male Saudis.

**Conclusion:** The PF score in our sample was low compared to other regional studies; the SF and EF scores were low in females and non-Saudis.

Coming from the Greek word *Thalassa* meaning sea, a reference to the Mediterranean sea around which the disease was originally described and is prevalent, thalassemia is a commonly inherited blood disorder. The adult hemoglobin (HbA1) is a tetramer of 2 alpha and 2 beta chains. The alpha chain is coded for by 2 genes on chromosome 16, while the beta chain by only one gene on chromosome 11. It is in these genes that the mutations leading to the thalassemia syndromes take place, leading to either a drop in the production or absence of the corresponding chain; alpha-thalassemia and beta-thalassemia.[1,2]

The prevalence of thalassemia in Saudi Arabia, especially on the western coast, is in no need to be underscored. With a study published in 2007 reporting 3.4% of 488,315 premarital screenings done have a thalassemia trait, while 0.07% had the thalassemia disease; focused mostly in the western, southwestern, and eastern parts of Saudi Arabia.[3] As regular blood transfusions and iron chelation are being a standard of care in thalassemia, the life expectancy of thalassemia patients has increased significantly. With more patients reaching adulthood, thalassemia is now a chronic disease having its burden on a population different from what it was a while back.[4,5] That and the biopsychosocial model, an approach now taken up everywhere in medicine, necessitate the need to evaluate the quality of life in these patients.[4,6] Congruent to the biopsychosocial model, the tools used must appraise the physical, social, and emotional/psychological well-being of the patient. These carefully-crafted questionnaires attempt to quantify these aspects in the patient's life and therefore provide us with reproducible data that assess the outcome of thalassemia management and assess its burden on patients' lives. Also, this data should provide the healthcare system with indicators whether to mobilize more resources to better manage these diseases or change the format of the management, including one that's more holistic and more inclusive of the social and psychological aspects of patients' lives. In this study, we aim to assess the quality of life in thalassemia adult patients in the western region of Saudi Arabia.

**Methods.** This is a cross-sectional study of adult patients (≥18 years) with major beta-thalassemia of both genders attending the Day Care Unit in King Abdulaziz University Hospital, Jeddah, Saudi Arabia. Data were collected between October 2012 and December 2012. The total sample was 48 adults. No sample size calculations were carried out prior to the study; all patients attending the day care unit who met the aforementioned criteria and agreed to participate were included in the study.

An ethical approval was obtained from King Abdulaziz University Hospital, and written informed consent were obtained from all patients. Retrieved data included demographics (age, gender, and nationality), educational level (elementary, middle school, high school, and postgraduate degree), employment status, marital status. Also retrieved were the variables of frequency of blood transfusions since diagnosis,

splenectomy status. The questionnaire used in our study was the RAND 36 item Short Form Health Survey (SF-36) in 2 versions: English and Arabic. The questionnaire was either self-administered or if the patient were illiterate or unable due to other health related difficulties, the questionnaire was filled as an interview conducted with the patient by a trained interviewer. As for the Arabic translation provided for this questionnaire, its validity, and reliability had been proven on adults in Saudi Arabia.[7]

The SF-36 questionnaire used in our study is widely used for its applicability and reproducibility. It is a generic questionnaire, and the version we used covered 6 domains; Physical functioning, Social functioning, emotional functioning, Severity of pain, level of energy, and general health.[8] Using the SF-36 model, each patient is given a score from 0 to 100 with higher scores indicating better health or a higher level of function. The patients' answers are presented as a profile of scores calculated for each scale. Two measures affect these scales, mainly the Physical and Mental Health Scores.[8]

***Statistical analysis.*** Data entry and analysis were performed using the Statistical Package of Social Sciences version 20.0.0 (SPSS Inc., Chicago, IL, USA). Descriptive statistics were used to calculate the different frequencies. Comparison between different groups for each score was carried out using the unpaired t-test. *P*-value of less than 0.05 was considered statistically significant.

**Results.** In our study, we included 48 patients aged between 18 and 38 years of age (mean =26.02 and SD =5.56). Males made up 60.4% of our sample. 41.7% of patients were Saudis. 68.8% of patients had a positive family history of thalassemia. The frequency of blood transfusion was every 3 weeks in most of the patients (81.3%), but 18.7% were having transfusions less frequently. Half of our sampled patients were splenectomized (54.2%).

The general health score for the total sample was 54.3 (SD=15.7). The score was slightly higher in males than in females. Moreover, the score was lower in people receiving transfusion every 3 weeks than those who were transfused less frequently, while splenectomy status and nationality showed no difference with regards to the general health score. The physical functioning score (PF score) for the total sample was 61.4 (SD=22.7). In general, the PF score was not affected statistically with regards to any of the retrieved variables. The EF score for the total sample was 69.7 (SD=21.6). The EF score was significantly higher among Saudis and those getting blood transfusion every 2-3 months with a *p*-value of 0.02 for both. The SF score for the total sample was 75 (SD=26.4). The SF score was higher among males compared to females with a *p*-value of 0.01, but showed no statistical significance with regards to other variables. The level of energy for the total sample was 49.3 (SD= 20.5). Moreover, the severity of pain for the total sample was 69.4 (SD=29.2). For the above-mentioned scores, no variables were found to affect them statistically. The General Health score had a mean of 54.3 (SD=15.7) (**Tables 2 & 3**).

**Discussions.** In our sample of 48 thalassemia adult patients, 40.1% were Saudis; this is compared to a 21.7% in the other study assessing the quality of

**Table 1 -** Socio-demographic data retrieved before the short form health survey (SF-36) questionnaire was administered.

| Demographic data | n | (%) |
|---|---|---|
| *Age* | | |
| <25 | 19 | (39.6) |
| ≥25 | 29 | (60.4) |
| *Gender* | | |
| Male | 29 | (60.4) |
| Female | 19 | (39.6) |
| *Marital status* | | |
| Single | 43 | (89.6) |
| Married | 5 | (10.4) |
| *Nationality* | | |
| Saudi-Saudi | 20 | (41.7) |
| Non | 28 | (58.3) |
| *Level of education* | | |
| Illiterate | 5 | (10.4) |
| Primary | 9 | (18.8) |
| Intermediate | 7 | (14.6) |
| Secondary | 17 | (35.4) |
| University | 10 | (20.8) |
| *Occupation* | | |
| Working | 15 | (31.2) |
| Non-working | 33 | (68.8) |
| *Positive family history* | | |
| Yes | 33 | (68.8) |
| No | 15 | (31.2) |
| *Age of onset of thalassemia* | | |
| <6 months | 18 | (37.5) |
| 6 months - 5 years | 22 | (45.8) |
| >5 years | 8 | (16.7) |
| *Frequency of blood transfusion* | | |
| Every 3 weeks | 39 | (81.3) |
| ≥4 weeks | 9 | (18.7) |
| *Splenectomy* | | |
| Yes | 26 | (54.2) |
| No | 22 | (45.8) |

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life in thalassemia pediatric patients authored by the same group.[9] This might be due to the fact that with autonomy, adult Saudi patients transfer their care to another hospital closer to where they live, even if it meant having to pay for treatment. While for pediatric patients, especially with multiple affections in a single family being common, getting care in a public hospital seems wiser to the caregiver. In our literature review, a lack of similar studies to assess the quality of life in adult thalassemia patients stood clear. Although in the Middle East we encountered some that will be referenced here.

In our sample, a PF score of 61.4 was concluded; this is compared to 77.1 in a study done in Lebanon on thalassemia adult patients.[8] This may be attributed to the level of education, which eventually translates into better compliance and more adherence to follow-up. In the Lebanon study, which had the same sample size as our study (n=48), 33.3% had a college degree, compared to 20.8% in our sample. Also, this significant drop could be partly to the fact that 64.6% of patients in the Lebanese study were splenectomized, compared to 54.2% in our study.[8] Our PF score is also significantly lower than the one concluded in an Iranian study (PF=72.11).[10] Their sample was much larger (n=308), education level was similar to our sample, yet the PF score was significantly higher. Unfortunately, not many other variables were answered to in that study. Our study concluded a PF score that's lower than that concluded in aforementioned studies. We attribute that to the low socio-economic status in our sample (68.8% cited no work), which may be partly due to a selection bias; the fact that the university hospital is a public hospital offering services for free, therefore attracting that segment of the population. It is concluded that a low socioeconomic status is a predictor of a poor quality of life.[11] It is worth-mentioning here that an Italian study on a sample of 50 adult thalassemia patients concluded a PF score of 86.1 in 2009,[12] which is staggering compared to our PF score of 61.4; also 77.1 and 72.11 for the Lebanese and the Iranian studies, respectively. This is not a thing to marvel at with the WHO's World Health Report in 2000 ranking Italy second to France in the world in terms of healthcare.

As for the social functioning score (SF score), our sample averaged a score of 75, which is significantly higher than the SF score in the Iranian study (60.11),[11] and closer to the one concluded in the Italian study (81.7).[12] This relatively high SF score comes with 81.3% of our sample receiving transfusions every three weeks, and 18.7% receiving transfusions every 4 weeks or more. This also comes in spite of the fact that the Day Care Unit in the hospital does not offer services in the weekends, although the last statement is mitigated by the fact that 68.8% were unemployed. However, in our sample there was a statistically significant difference between males and females in their SF scores: males scored 82.2 and females scored 63.2 ($p$=0.01). This can be attributed to the gender roles in Saudi Arabia, especially those in traditional families where women play a minor social role and assume minor social responsibilities. We also noted a statistically significant difference of SF scores between Saudis and non-Saudis: Saudis scoring 83.8 and non-Saudis scoring 68.8. We can attribute this to the presence of more social contacts and circles, family and friends, which exceed that available for non-Saudi residents in Saudi Arabia.

The pattern in the EF score comes in tandem with SF score in our sample and in comparison with the other studies: our sample concluded an EF score of 69.7, compared to 56.22 in the Iranian study (n=10),

**Table 2 -** The mean and standard deviation of the different short form health survey (SF-36) scores.

| Score | Mean | SD |
|---|---|---|
| Physical functioning | 61.4 | 22.7 |
| Emotional functioning | 69.7 | 21.6 |
| Social functioning | 75.0 | 26.4 |
| Level of energy | 49.3 | 20.5 |
| Severity of pain | 69.4 | 29.2 |
| General Health | 54.3 | 15.7 |

**Table 3 -** The association of gender and nationality with the different short form health survey (SF-36) scores.

| Variables | PF score | EF score | SF score | Vitality (energy) | Pain | General health |
|---|---|---|---|---|---|---|
| *Gender* | | | | | | |
| Male | 59.5 (22.9) | 73.1 (19.9) | 82.8 (22.2) | 52.1 (18.6) | 74.3 (26.9) | 56.1 (16) |
| Female | 64.2 (22.7) | 64.4 (23.4) | 63.2 (28.4) | 45 (23) | 61.8 (31.6) | 51.5 (15.1) |
| *P*-value | 0.49 | 0.17 | 0.01 | 0.24 | 0.15 | 0.33 |
| *Nationality* | | | | | | |
| Saudi | 67.3 (22.1) | 78 (17.5) | 83.8 (20.3) | 52.5 (21.7) | 75.4 (27.3) | 53.8 (20.3) |
| Non-Saudi | 57.2 (22.5) | 63.8 (22.5) | 68.8 (28.8) | 47 (19.6) | 65.1 (30.1) | 54.7 (11.7) |
| *P*-value | 0.13 | 0.02 | 0.051 | 0.37 | 0.23 | 0.86 |

74.2 in the Italian study (n=12), and 71.7 in the Lebanese study (n=8). Our EF score is also significantly higher in males compared to females (73.1 and 64.4; $p$=0.17), also higher in Saudis compared to non-Saudis (78 and 63.8; $p$=0.02). We attribute these differences to the same reasons we cited for the difference in SF scores.

The level of energy (vitality) score in our sample was 49.3, compared to 54.31 in the Iranian study and 65.8 in the Italian study. This goes in tandem with the PF score, as expected. Being splenectomized or not, was not associated with any significant difference with any of the variables in our study. Plus, no studies to compare splenectomized and non-splenectomized patients in terms of quality of life were encountered in our literature review. It is worth mentioning here that bone marrow transplant is a way to cure Thalassemia, and a study comparing those transplanted patients over 20 years to the general population concluded a return to normal life style and a good health-related quality of life (HRQoL).[13] This provides us with an insight in the management of thalassemia patients.

In conclusion, a low PF score is noted, the PF score that takes up most of the SF-36 questionnaire. Here, it delineates the burden of thalassemia on adults despite managing those patients and should prompt initiatives to redress the issue. As for the SF and EF scores, our results show a positive outlook for male Saudis, but not for the female and non-Saudi segments of our sample. This should accentuate the need for a more holistic approach that encompasses the emotional and social wellbeing of thalassemia patients: patients could be given appointments to the psychiatric clinic and be offered the services of social workers in their hospitals. Special groups can be organized for thalassemia patients to better engage them socially and offer support. Like our study on pediatric thalassemia patients, we reiterate our appeal for an education program for these patients, one that render them more apt to deal with the physical, emotional, and social burdens of the disease.

We hope that our study will be used as a model for further studying to establish reproducible data that assess the progress carried out in the management of thalassemia in the kingdom, and be invoked in decisions when it comes to allocating resources considering the psychological and social dimensions of the disease and encouraging an interdisciplinary approach that includes the psychiatric and social departments in Saudi hospitals.

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