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# A Cross-sectional Study to Evaluate the Quality of Life among Children Diagnosed with Thalassemia Major in Gujarat

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## Abstract

**Background** Thalassemia, a chronic condition that lacks an effective treatment, significantly impairs a child's quality of life (QoL), affecting academic performance and various dimensions such as physical, emotional, and social functioning. The objectives of this study are to evaluate the QoL of children with thalassemia major and to investigate the relationship with selected demographic variables of children. This study addresses the crucial need to assess the QoL among children living with thalassemia major. By evaluating QoL, health care professionals can tailor treatment approaches to improve the lives of children affected by thalassemia major.

**Study Design and Methods** This cross-sectional analytical study was conducted at the district division of the Indian Red Cross in Ahmedabad, Gujarat. The consecutive sampling technique was used. The study included 117 children with β-thalassemia major. The QoL of children aged 8 to 18 years was evaluated in September 2021 using the Short Form-36 (SF-36) questionnaire. Statistical analysis involved mean, standard deviation, and chi-square test.

**Results** The mean age of the participants was $14.9 \pm 3.1$ years. The majority of the included children (56%) received blood transfusions for 15 days. The mean total score of SF-36 was $68.62 \pm 12.96$. Out of the total number of children, 55 had a score of more than 70, 50 had a score between 50 and 70, and 12 had a poor QoL with a score less than 50. The chi-square analysis reveals a statistically significant association between gender perception and QoL ($p = 0.033$). Similarly, a notable association is noted between monthly family income and QoL ($p = 0.037$).

**Conclusion** This study revealed that the QoL in children diagnosed with thalassemia major is compromised. QoL assessment should be initiated for all children with thalassemia to identify and implement essential interventions that focus on the affected domains.

**Keywords**
- β-thalassemia
- QoL
- thalassemia major
- quality of life
- children

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## Introduction

Thalassemia major presents a significant public health challenge in Southeast Asia.[1] A considerable number of individuals with thalassemia major and carriers have been documented. This phenomenon is associated with the prevalence of consanguineous marriages in these regions.[2] The incidence of thalassemia varies across different geographical regions. Recent estimates indicate that in India, 0.37 out of every 1,000 fetuses are affected by hemoglobin disorders.[3] Children with thalassemia major undergo a demanding treatment regimen that commences in infancy, involving regular blood transfusions and iron chelation therapy.[4] Currently, patients with this condition have increased life expectancies due to advancements in treatment. However, prolonged life expectancies come with challenges such as frequent blood transfusions, daily iron chelator administration, complications, and potential splenectomy. Children with thalassemia major often experience poor nutritional status, leading to various health issues that can impede their overall development. Ensuring adequate nutrition is essential for effectively managing their health.[5] These obstacles significantly impact the quality of life (QoL) of children, affecting their mental, physical, emotional, social, and educational well-being.[6,7] Hence, the present investigation aims to assess the health-related QoL (HRQoL) in children diagnosed with thalassemia major using the Short Form-36 (SF-36) questionnaire and identify the factors associated with it.[8] The objectives of this study were to evaluate the QoL in children diagnosed with thalassemia major and investigate the relationship between selected demographic variables and the QoL of children with thalassemia major.

## Study Design and Methods

This study was an analytical cross-sectional investigation involving children diagnosed with thalassemia major, aged 8 to 18 years. A consecutive sampling, nonprobability sampling technique was employed to select children diagnosed with thalassemia major. Specifically, children attending the district branch of the Indian Red Cross Society, Gujarat, for blood transfusion during the study period and expressing willingness to participate were chosen. The total number of children included in this study was 117, determined by power analysis. Inclusion criteria required participants to be able to speak Gujarati and English. Exclusion criteria involved children with impaired cognitive function or severe clinical conditions that hindered their ability to participate. Recruitment of participants began in September 2021. Data were collected directly from children aged 8 to 18 years, with parents sometimes present during data collection.

The Persian version of the SF-36 questionnaire, previously translated from English and validated, was utilized in this study.[9–11] The SF-36 questionnaire is a standardized tool. Section I includes demographic data, and the Gujarati version of the tool was validated by four nursing experts, two doctors, and one Gujarati editor. Reliability was calculated using Cronbach's α method. The SF-36 serves as a self-administered tool for scoring QoL, comprising eight scales and two aspects. These scales encompass physical functioning, role-physical, role-emotional, energy-fatigue, emotional well-being, social functioning, pain, general health, physical health, and mental health. The primary five scales contribute to the physical health element, while the last three scales form the mental health element.[12,13]

It has previously been shown that the SF-36 questionnaire can be completed through an interview, by phone, or by computerized administration, even though it is designed as a self-administered questionnaire.[14] The SF-36 Health Survey measures HRQoL across eight domains: physical functioning, role-physical, bodily pain, general health, vitality, social functioning, role-emotional, and mental health. Each domain is scored separately based on responses to specific questions, and the scores are then converted to a 0 to 100 scale. Higher scores indicate better health status or fewer limitations. The scoring process involves recoding specific items, summing up scores, and transforming raw scores. This method provides a comprehensive profile of an individual's HRQoL, with each domain score interpreted independently. Demographic variables of patients, such as age, gender, age at disease onset, age at the first blood transfusion, frequency of blood transfusion, and splenectomy, were recorded.

The study protocol received approval from the registered ethics committee of Rudraksh Hospital and the Medical Secretary of the Indian Red Cross Society, district branch. Informed consent and assent were obtained from each child and parent after providing a comprehensive explanation of the study's purpose, potential impact, and data confidentiality.

Data collection was conducted with the assistance of a predesigned and pretested tool through interviews with the study participants to gather information on their sociodemographic profile. Information on the clinicotherapeutic profile was obtained through interviews with the study participants and by reviewing the existing patient records (including data on ongoing treatment). HRQoL was evaluated using the SF-36 Health Survey. Statistical analysis employed mean, standard deviation, chi-square, and independent samples *t*-test.

## Results

Overall, we included 117 children, comprising 62 males (53%) and 55 females (47%), with a mean age of $14.8 \pm 3.2$ years (range: 8–18 years).

►**Supplementary Table S1** (online only) illustrates the demographic and clinical features of children diagnosed with thalassemia major. The sample of 117 individuals comprises 53% males and 47% females. Age distribution is varied, with 39% aged 16 to 18 years. The majority (72%) experienced disease onset between 0 and 5 years of age, and 79% had their first blood transfusion within the same age range. Educationally, 35% of the population reached secondary education, and 24% attained higher education. Family income exceeds 30,000 INR for 45% of the sample. Blood transfusions are most frequently administered biweekly (56%). Deferasirox is the most common iron chelator, accounting for 56% of cases. Only 20% underwent a splenectomy, while 61% did not have any surgeries. Among the total children, 55 individuals

 

(31.93%) achieved a score exceeding 70, indicating a good QoL, while 12 individuals (5.88%) recorded a score below 50, indicating a poor QoL.

►**Table 1** presents a statistically significant relationship between the QoL and selected demographic variables among children diagnosed with thalassemia major. The chi-square analysis reveals a statistically significant association between gender perception and QoL among children with β-thalassemia major ($p = 0.033$). Similarly, a notable connection is noted between monthly family income and QoL ($p = 0.037$). However, no statistically significant association was identified between the QoL of children with β-thalassemia major and other sociodemographic variables.

## Discussion

In this study, we assessed the QoL of children diagnosed with thalassemia major using the SF-36 questionnaire. This tool is widely recognized for its effectiveness in assessing QoL across diverse populations and various medical conditions.

**Table 1** Association among QoL and sociodemographic variables

| Variables | | Pretest overall QoL | | | Total | Chi-square | *p*-Value |
|---|---|---|---|---|---|---|---|
| | | Good >70 score | Average 50–70 score | Poor <50 Score | | | |
| Gender | Male | 33 | 20 | 9 | 62 | 6.805 | 0.033 |
| | Female | 22 | 30 | 3 | 55 | | |
| Age (y) | 8–10 | 12 | 11 | 1 | 24 | 4.053 | 0.852 |
| | 10–12 | 6 | 5 | 1 | 12 | | |
| | 12–14 | 5 | 5 | 3 | 13 | | |
| | 14–16 | 9 | 10 | 3 | 22 | | |
| | 16–18 | 23 | 19 | 4 | 46 | | |
| Age of onset (y) | Below 5 | 41 | 36 | 7 | 84 | 2.720 | 0.843 |
| | 5–10 | 9 | 10 | 3 | 22 | | |
| | 10–15 | 4 | 2 | 1 | 7 | | |
| | Above 15 | 1 | 2 | 1 | 4 | | |
| Age of first blood transfusion (y) | Below 5 | 45 | 40 | 7 | 92 | 7.595 | 0.269 |
| | 5–10 | 8 | 7 | 2 | 17 | | |
| | 10–15 | 1 | 2 | 2 | 5 | | |
| | Above 15 | 1 | 1 | 1 | 3 | | |
| Education status | Illiterate | 5 | 7 | 3 | 15 | 5.560 | 0.474 |
| | Primary education | 18 | 13 | 2 | 33 | | |
| | Secondary education | 19 | 16 | 6 | 41 | | |
| | Higher secondary education | 13 | 14 | 1 | 28 | | |
| Family income | 10,000 INR or less | 7 | 4 | 4 | 15 | 13.366 | 0.037 |
| | 10,000–20,000 INR | 9 | 4 | 3 | 16 | | |
| | 20,000–30,000 INR | 18 | 12 | 3 | 33 | | |
| | Above 30,000 INR | 21 | 30 | 2 | 53 | | |
| Blood transfusion cycle (wk) | 2 | 25 | 31 | 9 | 65 | 5.528 | 0.237 |
| | 3 | 22 | 12 | 2 | 36 | | |
| | 4 | 8 | 7 | 1 | 16 | | |
| Chelator agent | Deferoxamine | 22 | 14 | 5 | 41 | 1.937 | 0.747 |
| | Deferasirox | 28 | 31 | 6 | 65 | | |
| | Other | 5 | 5 | 1 | 11 | | |
| Surgery | Splenectomy | 12 | 8 | 3 | 23 | 1.514 | 0.824 |
| | Other | 9 | 11 | 3 | 23 | | |
| | No any | 34 | 31 | 6 | 71 | | |

Abbreviation: QoL, quality of life.

 

The SF-36 includes eight domains, offering a comprehensive health profile. We utilized the Persian-translated version, which has been validated in multiple studies involving normal populations, ensuring its reliability and accuracy in this context. The results provide insights into the impact of thalassemia major on children's QoL, aiding in the development of targeted interventions and support measures.[8,15]

The mean age of children in our study (14.8 years) is notably lower than that reported in other studies done by Adam (22.9 years), Hamdy et al (17 years), and Haghpanah et al (19.22 years).[14,16,17] The age difference in our sample highlights a younger demographic, which could impact the evaluation of the QoL in children with thalassemia major. Interestingly, the QoL score in our study (68.6) was higher than those reported in previous studies by Hamdy et al in Egypt (44.9) and Haghpanah et al in Iran (67.8).[16,17] This suggests that our participants, despite being younger, reported a better QoL. Factors contributing to this discrepancy could include differences in health care quality, disease management, socioeconomic conditions, or cultural perceptions of health and well-being across the study populations.

Moreover, our study revealed a significant association between monthly family income and QoL, consistent with findings by Hamdy et al. This underscores the impact of economic factors on health outcomes. However, unlike this study,[16] we found no statistically significant association between QoL and other demographic variables such as age, gender, or educational background. This suggests that financial stability may play a more critical role in influencing the QoL in children with thalassemia major.

## Conclusion

Most people with thalassemia suffer from a chronic condition without an effective treatment, which negatively impacts their QoL. This cross-sectional study assessed the QoL of 117 children with β-thalassemia major in Ahmedabad using the SF-36 questionnaire. Results showed a mean QoL score of 68.62, with significant associations between gender, family income, and QoL. The findings of this study revealed that, despite excluding critically ill children, many diagnosed with thalassemia major still experience compromised QoL. A statistically significant association was observed between the QoL of children with thalassemia and their gender, as well as their economic status. It underscores the imperative for intervention to enhance the QoL of these children.

### Implications for Research, Policy, and Practice

The study's mean QoL score of 68.62 for children with β-thalassemia major highlights several key implications. With many participants receiving regular blood transfusions, health care providers should implement comprehensive care strategies addressing both medical and psychosocial needs. Gender-sensitive and socioeconomic interventions are essential, as males scored higher in physical role and females in pain. Socioeconomic support, such as financial aid and educational resources, is crucial for improving outcomes. Policymakers should utilize these findings to assist low-income families and create comprehensive health care programs that improve the QoL and well-being of children with thalassemia. Future research in this area should aim to explore additional factors influencing the QoL of children with β-thalassemia major. Specifically, studies could investigate the impact of psychosocial support programs, such as counseling and peer support groups, on QoL outcomes. Furthermore, longitudinal research could assess how QoL changes over time in response to various interventions and life transitions, providing insights into long-term care needs. Additionally, qualitative research methods, such as interviews or focus groups, could provide a more profound insight into the lived experiences of children with thalassemia and their families. This could help in developing more precise and culturally sensitive interventions. Finally, comparative studies across various health care settings and regions could clarify differences in QoL outcomes and pinpoint best practices for enhancing overall well-being in this population. Improving the QoL for children with thalassemia major requires comprehensive medical interventions, including regular blood transfusions and iron chelation therapy to manage iron overload.[17] Nutritional support tailored to their specific needs is essential to address and prevent deficiencies. Psychosocial support, including counseling and support groups, can help children and their families cope with the emotional challenges of the condition. Educational programs about thalassemia for families and caregivers can empower them with the knowledge needed to provide optimal care.

#### Ethical Approval

The registered ethics committee at Rudraksh Hospital (ECR/1139/Inst/GJ/2018/RR-21) thoroughly reviewed and granted ethical approval for this study. All participants provided their consent.

#### Conflict of Interest

None declared.

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