RESEARCH ARTICLE

# Quality of life among beta-thalassemic major children presenting at Federal Government Hospital Islamabad, Pakistan

Gul Mehar Javaid Bukhari


## Abstract

**Objective:** To assess the quality of life among beta-thalassemic major children in a tertiary care setting.

**Method:** The cross-sectional descriptive study was conducted at the Federal Government Hospital, Islamabad, Pakistan, from October to December, 2020, and comprised beta-thalassemic major children aged 7-13 years. Socio-demographic information was collected using a questionnaire, while the quality of life was assessed using a pretested tool with Cronbach's alpha value 0.855. The data was analysed using SPSS 25.

**Results:** Of the 87 subjects, 47(54%) were males and 40(46%) were females. The overall mean age was 10.71±1.99 years. The mean quality of scale score was 50.24±18.88. Poor quality of life was found among 33(37.9%) children. The quality of life had significant association with age 7-9 years, male gender and blood transfusion frequency 2 or more ($p$<0.05). The adjusted odds were also significant with age and blood transfusion frequency ($p$<0.05). The overall mean score was significantly related within age groups and frequency of blood transfusion ($p$<0.05), whereas physical and emotional domains were significant with age ($p$<0.05), while the four domains of physical, psychological, social and educational were associated with frequency of blood transfusion ($p$<0.05).

**Conclusion:** Quality of life among thalassemic children was found to be considerably low. The physical and emotional domains need to be focussed upon for improving the quality of life. Measures should be taken to avoid the increased need of blood transfusions through treatment compliance.


**Keywords**: Beta-thalassemia major, Quality of life, QoL, Blood transfusion. (JPMA 72: 2241; 2022)

**DOI: https://doi.org/10.47391/JPMA.5248**

## Introduction

Thalassemia is a genetically transmitted disease that primarily affects the formation of haemoglobin (Hb) globin chains. In the Mediterranean, the Middle East, the Indian subcontinent, and in Southeast Asia, this disease is a major public health issue.[1] The prevalence across different regions of the world showed that gene mutation is responsible for the condition among Americans 0-3%, in Europe 0-19%, in sub-Saharan Africa 0-12%, in Eastern Mediterranean region 2-18% and in Southeast Asia 0-11%.[2] Pakistan has beta-thalassemia major (βTM) gene carriers (5-7%) as well as 9.8 million people present in the general population, making the disease the most widespread hereditary haemoglobinopathies.[3] The enrollment figures at various treatment centres in the country accounts for approximately 50,000 patients.[4] The transfusion-dependent thalassemic children in Pakistan are estimated to be 100,000 and various studies have described overall prevalence rate of 5-7%.[5] Owing to the chronic nature of certain diseases, such as βTM, not only the significance of survival is important for the patients but also their social as well as psychological functioning that must be given importance.[6] There are several other symptoms that add to the severe nature of their condition, like proper growth patterns, anaemia, bone disorders and enlargement of liver and spleen.[7] The treatment with transfusions for the βTM children include increased infections, iron overload as well as antibody formations. To remove excessive iron from the body, chelation therapy must be initiated after regular transfusions for preventing morbidities and also to increase life expectancy.[8,9] Mona Hamdy et al. showed that mostly patients (93.6%) had monthly blood transfusion frequency. There was insignificant difference found with respect to gender while the "general health perception" domain was mostly affected and "vitality" was the least affected.[10] Safe transfusions increase quality of life (QOL) among βTM patients. The average life span among such patients is still very short at almost 10 years.[11] The disease can be prevented through simple measures, but in developing countries like Pakistan, the prevalence is increasing considerably along with reduced treatment facilities in the country and results in poor QOL.[10,11] TM is a blood disorder that affects patients and their families psychologically. It is usually diagnosed at birth or within the first 6-12 months of life. Because children must undergo recurrent transfusions and iron chelation therapy, it is a constant source of distress for them. Social isolation, low self-esteem, low academic achievement, and school absenteeism are all substantial psychological difficulties for the patients. Those with thalassemia do not express their emotions and keep

Department of Community Medicine, Federal Medical College, Islamabad, Pakistan.

**Correspondence:** Gul Mehar Javaid Bukhari. e-mail: drgulmehar@gmail.com

them hidden because they are afraid of being rejected by others. For children, especially those with chronic illnesses, assessing health-related QOL is an essential indicator of health outcome.[12]

According to the World Health Organisation (WHO), QOL is regarded as the opinion of the individuals related to objectives, prospects, concerns and principles of their lives in the context of value and cultural systems. Social discriminations, transition of life and behavioural changes also threaten QOL.[12] A study showed that QOL score among physical domain was lower compared to the psychological domain. There was no significant difference between various domain scores.[13] According to a study in Rawalpindi, stress score came out to be 51.38% severe, 10.28% moderate and 38.34% mild.[14] The QOL among these patients remained very important index for better health that could also be defined as patient's personal perception related to their disease pattern and also the treatment availability affecting their daily life activities, including mental, physical, social and emotional performances.[15]

QOL among these patients is influenced by diagnosis methods and treatment offered, their complications, increased hospital admissions for blood, and the creeping doubts regarding other treatment options.[16] Another study reported an overall occurrence of βTM as 6% and QOL was considerably good in the studied group.[17]

The current study was planned to quantify QOL among βTM children in terms of physical, psychological, social and educational domains.

## Patients and Methods

The cross-sectional descriptive study was conducted from October to December, 2020, at the Thalassemia Centre of the Federal Government Hospital, Islamabad, Pakistan. The centre is a 1250-bed hospital in which 35 beds are deputed specifically for TM treatment. The centre was selected as it was the only tertiary level centre specialised for thalassemia treatment for children in the area.

After approval from the ethics review board of Shaheed Zulfiqar Ali Bhutto Medical University, Islamabad, the sample size was calculated using cross-sectional formula

$$(n = \text{Sample size} = \frac{Z_{1-\alpha/2}^{2}\, p(1-p)}{d^2}\ )^{18}$$

by taking population proportion (p) of βTM 6%,[17] 95% z value ($1.96^2 = 3.84$) and absolute precision (d) 5%.

The data was collected using non-probability convenience sampling.

Those included were children aged 7-13 years and had no known complication at the time of the study. Physically or mentally impaired children were excluded. Independent variables were socio-demographics, including age, gender, residence, and frequency of blood transfusion, while dependent variables were QOL scores and good/poor QOL.

After taking informed consent from the parents/guardians, data was collected using a pretested questionnaire that had 25 items distributed among 4 domains; physical, psychological, emotional and educational. Among the questions, 6 items were reverse-coded due to their positive intent. The pre-tested questionnaire had Cronbach's alpha value 0.855 indicating validity. The questionnaire was scored on a f-point Likert scale; 0 = "not at all", and 4 = "always". Overall score ranged 0-100. The operational definition for QOL constituted that the children with total scores ≥50 had poor QOL.

The collected data was analysed using SPSS 25. Chi-square test was used to see the association of QOL with other parameters. Independent t-test was used to compare means of total score and individual domain scores with sociodemographic data and frequency of blood transfusion. Logistic regression was applied for calculating adjusted odds ratio (AOR) with only socio-demographic variables being adjusted. 'Wald' $p<0.05$ was considered statistically significant. $P<0.05$ and $p<0.01$ were considered statistically significant and highly significant respectively.

## Results

Of the 87 subjects, 47(54%) were males and 40(46%) were females. The overall mean age was 10.71±1.99 years. There were 23(26.4%) children aged 7-9 years and 64(73.6%) were

**Table-1:** Association of quality of life with sociodemographic variables.

| Variables | Quality of Life | | OR (95% CI) | AOR (95% CI) |
|---|---|---|---|---|
| | **Poor** (%) | **Good** (%) | | |
| **Age** (years) | | | | |
| 7-9 | 15 (65.2) | 8 (34.8) | 4.79 (1.73-13.24)[a] | 6.05 (1.94-18.92)[a] |
| 10-13 (Ref) | 18 (28.1) | 46 (71.9) | | |
| **Gender** | | | | |
| Male | 23 (48.9) | 24 (51.1) | 2.87 (1.15-7.19)[a] | 2.31 (0.81-6.60) |
| Female (Ref) | 10 (25) | 30 (75) | | |
| **Blood Transfusion** | | | | |
| One per month (Ref) | 16 (26.2) | 45 (73.8) | 0.19 (0.07-0.51)[b] | 0.17 (0.06-0.51)[a] |
| Two or More per Month | 17 (65.4) | 09 (34.6) | | |
| **Address** | | | | |
| Rural (Ref) | 15 (39.5) | 23 (60.5) | 1.12 (0.47-2.69) | -- |
| Urban | 18 (36.7) | 31 (63.3) | | |
| **Total** | 33 (37.9) | 54 (62.1) | | |

[a]*p*-value<0.05; [b]*p*-value<0.01; OR: Odds ratio, CI: Confidence interval, AOR: Adjusted odds ratio.

**Table-2:** Mean comparison of quality of life total score and domain scores with sociodemographic variables and frequency of blood transfusion.

| Domains Quality of Life | Mean Scores (Mean±SD) | | t-test/p-value |
|---|---|---|---|
| **Age** (Years) | 7-9 (n=23) | 10-13 (n=64) | |
| Total Score | 59.78±18.41 | 46.81±17.98 | 2.95/0.004[a] |
| Physical Domain Score | 16.04±6.63 | 12.52±6.02 | 2.35/0.021[a] |
| Psychosocial Domain Score | 14.83±6.44 | 11.67±6.62 | 1.97/0.052 |
| Educational Domain Score | 12.61±4.28 | 11.06±4.06 | 1.543/0.126 |
| Emotional Domain Score | 16.30±3.66 | 11.56±4.72 | 4.36/<0.01[b] |
| **Gender** | Male (n=47) | Female (n=40) | |
| Total Score | 53.87±19.23 | 45.98±17.76 | 1.977/0.051 |
| Physical Domain Score | 14.40±6.41 | 12.33±6.15 | 1.535/0.128 |
| Psychosocial Domain Score | 13.60±6.85 | 11.23±6.33 | 1.665/0.100 |
| Educational Domain Score | 11.87±4.54 | 11.00±3.64 | 0.976/0.332 |
| Emotional Domain Score | 14.00±4.77 | 11.43±4.77 | 2.51/0.014[a] |
| **Residence** | Rural (n=38) | Urban (n=49) | |
| Total Score | 49.61±18.72 | 50.73±19.18 | -0.275/0.784 |
| Physical Domain Score | 13.68±6.45 | 13.27±6.32 | 0.304/0.762 |
| Psychosocial Domain Score | 12.26±6.74 | 12.69±6.71 | -0.296/0.768 |
| Educational Domain Score | 11.42±3.48 | 11.51±4.65 | -0.102/0.919 |
| Emotional Domain Score | 12.24±5.07 | 13.27±4.80 | -0.967/0.336 |
| **Blood Transfusion Frequency** | One (n=61) | Two or More (n=26) | |
| Total Score | 45.74±17.06 | 60.81±19.03 | -3.643/<0.01[b] |
| Physical Domain Score | 11.98±6.06 | 16.88±5.71 | -3.51/<0.01[b] |
| Psychosocial Domain Score | 10.85±6.09 | 16.38±6.51 | -3.799/<0.01[b] |
| Educational Domain Score | 10.69±3.95 | 13.31±4.11 | -2.797/0.006[a] |
| Emotional Domain Score | 12.21±4.77 | 14.23±5.05 | -1.774/0.08 |

[a]*p*-value<0.05; [b]*p*-value<0.01. SD: Standard deviation.

aged 10-13 years age. The mean quality of scale score was 50.24±18.88 and individual domain score for physical aspects was 13.45±6.34, psychological aspect 12.51±6.68, educational domain 11.47±4.15 and emotional domain 12.82±4.92. Overall, 38(43.7%) children lived in rural areas and 49(56.3%) were from urban areas. The blood transfusion frequency was once per month in 61(70.1%) cases and two or more per month in 26(29.9%). Poor QOL was found among 33(37.9%) children.

There was significant association of QOL with age, gender and frequency of blood transfusion (Table 1)

Mean scores of total QOL and individual domains were significant associated with age and frequency of blood transfusion (Table 2).

## Discussion

βTM is a chronic haematological genetic disorder under the umbrella of non-communicable diseases usually caused by inability to manufacture polypeptide glob chains and transmission of such disorders to subsequent generation under Mendelian rules.[19] The reduction in social activities negatively affects the patients and increases the chances of anxiety, compromises independence and leads to banishment fear. The major portion of lives of these patients is full of misery and pain; and also affects the care-givers' life. Such issues further degrade their QoL and require continuous care for the reduction of their disease-associated complications.[16]

The negative effects of disease on the mental as well as physical health of patients and their QOL requires effective role to be played by the family, the treating physicians and other organizations involved in thalassemia treatments. By improving such conditions, the QOL of patients could improve treatment outcomes.[19]

The present study was carried out in βTM children aged 7-13 years for their QOL using a self-designed questionnaire. The results obtained were similar to those reported earlier.[14] The highest scoring in physical domain pertaining poor QOL could be attributed to compromised activity levels due to low self-esteem among the diseased children. This thalassemic effect on the domain of physical health could be related to the effects of thalassemia as well as its treatment towards patients' physical health that might result in physical deformity, underdevelopment of muscles and bones as well as delayed puberty.[16] The psychological aspect had low scores, which was consistent with literature.[20,21] Although the extended family system is considered vital in our part of the world, it could be acquiring concerns and needs equally distributed towards healthy and the impaired children.[20] Besides, earlier studies have shown poor scores regarding school and education domains.[22-24] The psychological issues in children poses serious impact on their quality of life due to anxiety, depression, self-esteem and also relationship between these seem to be complex.[25] In the current study, the physical and emotional domain scores were significant with age. With respect to gender, the emotional domain score was statistically significant and among transfusion frequency, physical, psychological and educational domains were significant. The emotional domain was found non-significant by some studies.[23,24]

The argument about adjustments and coping strategies for thalassemic children along with their families must be present saliently for behavioural and emotional complications as these domains purely determine the QOL.[25] The QOL therefore is an important indicator of one's health and happiness.

The major limitation of the current study was its cross-sectional design and the fact that it was done at a single centre with a small sample size. Besides, recall bias may arise due to personal reporting from the respondents. Multi-centre, large-scale, longitudinal studies are needed to determine the exact role of domains towards overall QOL among thalassemic children.

## Conclusion

TM children showed an overall poor QOL. The improvement regarding QOL requires combined efforts from parents, school authorities, treating physicians and policy-makers.

**Disclaimer:** None.

**Conflict of interest:** None.

**Source of Funding:** None.

## References

1. Hisam A, Khan NUS, Tariq NA, Irfan H, Arif B, Noor M. Perceived stress and monetary burden among thalassemia patients and their caregivers. Pak J Med Sci 2018;34:901-6. doi: 10.12669/pjms.344.15420.
2. News Medical Life Sciences. Prevalence of Thalassemia. [Online] 2021 [Cited 2021 July 15]. Available from URL: https://www.news-medical.net/health/Thalassemia-Prevalence.aspx
3. Ehsan H, Wahab A, Anwer F, Iftikhar R, Yousaf MN. Prevalence of Transfusion Transmissible Infections in Beta-Thalassemia Major Patients in Pakistan: A Systematic Review. Cureus 2020;12:e10070. doi: 10.7759/cureus.10070.
4. Thalassemia Federation of Pakistan. What is thalassemia? [Online] 2021 [Cited 2021 July 15] Available from: http://tfp.org.pk/what-is-thalasseamia/.
5. Bibi A, Hassan M, Khan SU, Naeem MO, Asnath L. Assessment of the Quality of Life of Children with Beta Thalassemia in Peshawar District: A Cross Sectional Study. Pak J Med Res 2021;60:121-5.
6. Origa R. β-Thalassemia. Genet Med 2017;19:609-19. doi: 10.1038/gim.2016.173.
7. Mehr MA, Kalhorpour H, Shirani V, Seyed-Nezhad A, Kord A, Zainivand M. Study of quality of life from the perspective of parents and children with thalassemia major. Med Sci 2019;23:333-8.
8. Ansari SH, Parveen S, Siddiqui S, Perveen K, Ahmed G, Kaleem B, et al. Managing thalassemia in the developing world: an evidence-based approach for prevention, transfusion independency, and curative treatment with hematopoietic stem cell transplantation. Blood Adv 2018;2(Suppl 1):s42-5. doi: 10.1182/bloodadvances.2018GS112057.
9. Khan K, Zahoor S. Pattern of hemoglobinopathies on HPLC among patients referred to selected centers in Peshawar, Pakistan. Rawal Med J 2018;43:623-6.
10. Hamdy M, Draz IH, El Sayed IT, Ayyad AA, Salemd MR. Assessment of Quality of Life among Beta-Thalassemia Major Patients Attending the Hematology Outpatient Clinics at Cairo University Hospital. Macedonian J Med Sci 2021;9:156-60. doi: 10.3889/OAMJMS.2021.5692
11. Shah FT, Sayani F, Trompeter S, Drasar E, Piga A. Challenges of blood transfusions in β-thalassemia. Blood Rev 2019;37:e100588. doi: 10.1016/j.blre.2019.100588.
12. Ishfaq K, Diah NM, Ali J, Fayyaz B, Batool I. Psychosocial problems faced by thalassemia major patients of district Multan, Pakistan. Pak Pediatr J 2018;42:22-6.
13. Khodashenas M, Mardi P, Taherzadeh-Ghahfarokhi N, Tavakoli-Far B, Jamee M, Ghodrati N. Quality of Life and Related Paraclinical Factors in Iranian Patients with Transfusion-Dependent Thalassemia. J Environ Public Health 2021;2021:e2849163. doi: 10.1155/2021/2849163.
14. Yousafzai YM, Roghani A, Khan N, Shah I, Khan S, Taj AS. Quality of life and its determinants in transfusion dependent thalassemia. Pak J Physio 2018;14:64-7.
15. Ruggeri K, Garcia-Garzon E, Maguire Á, Matz S, Huppert FA. Well-being is more than happiness and life satisfaction: A multidimensional analysis of 21 countries. Health Qual Life Outcomes 2020;18:1-16. doi: 10.1186/s12955-020-01423-y
16. Tanveer T, Masud H, Butt ZA. Are people getting quality thalassemia care in twin cities of Pakistan? A comparison with international standards. Int J Qual Health Care 2018;30:200-7. doi: 10.1093/intqhc/mzx198
17. Yasmeen H, Hasnain S. Quality of Life of Pakistani Children with β-Thalassemia Major. Hemoglobin 2018;42:320-5. doi: 10.1080/03630269.2018.1553183.
18. Pourhoseingholi MA, Vahedi M, Rahimzadeh M. Sample size calculation in medical studies. Gastroenterol Hepatol Bed Bench 2013;6:14-7.
19. Adam S. Quality of life outcomes in thalassaemia patients in Saudi Arabia: a cross-sectional study. East Mediterr Health J 2019;25:887-95. doi: 10.26719/2019.25.12.887.
20. Ankush A, Silveira M, Talwadker Y, Souza J. Quality of life in children with thalassemia major following up at a tertiary care center in India (GOTQoL). Int J Contemporary Pediatr 2019;6:168-75. doi: 10.18203/2349-3291.ijcp20185203
21. Khan FU, Nawaz IA, Asif A, Rehman A. Quality Of Life Of Thalassemia Major Patients. J Rawalpindi Med Coll 2019; 23(Suppl 2):s7-10.
22. Madmoli M, Madmoli Y, Rahmati P, Adavi A, Yousefi N, Gheisari Z, et al. Quality of life and some related factors in patients with beta thalassemia major in Southwest Iran. J Client-Centered Nursing Care 2017;3:139-46. doi: 10.32598/jccnc.3.2.139.
23. Adam S, Afifi H, Thomas M, Magdy P, El-Kamah G. Quality of Life Outcomes in a Pediatric Thalassemia Population in Egypt. Hemoglobin 2017;41:16-20. doi: 10.1080/03630269.2017.1312434.
24. Yasmeen H, Hasnain S. Epidemiology and risk factors of transfusion transmitted infections in thalassemia major: a multicenter study in Pakistan. Hematol Transfus Cell Ther 2019;41:316-23. doi: 10.1016/j.htct.2019.03.008.
25. Dayasiri KC, Kulathilake A, Mudiyanse RM. Clinical profiles, medical complications and quality of life of children diagnosed with thalassaemia major in Peradeniya Teaching Hospital. Sri Lanka J Child Health 2018;47:112-7. doi: 10.4038/sljch.v47i2.8475