ISSN 2515-8260 Volume 10, Issue 2, 2023

SOCIO-DEMOGRAPHIC PROFILE AND FACTORS AFFECTING QUALITY OF LIFE IN THALASSEMIA MAJOR PATIENTS - A CROSS -SECTIONAL STUDY

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Abstract

Introduction: Beta thalassemia pose a significant health burden in India. The average prevalence of its carriers is 3–4% which translates to 35 to 45 million carriers in our multi-ethnic, culturally, and linguistically diverse population of 1.21 billion people according to the Census of India 2011. Quality of life (QOL) assessments are important and are frequently required when evaluating health outcomes.[1]

Aims: To assess the socio-demographic profile and the factors affecting quality of life in thalassemia major patients.

Material and Methods: The present study was a cross-sectional study. This study was conducted from 1st April 2021 to 31st March 2022 at Rajindra Hospital, Patiala. 113 participants were included for study. Pediatric Quality of Life Inventory™ (PedsQL™) 4.0 Generic Core Scale was employed for measuring quality of life.

Results: 31(27.4%) participants were 5-7 years of age, 35 (31.0%) participants were 8-12 years of age, and 47 (41.6%) participants were 13-18 years of age. QOL scores in different domains of PedsQL scale classified by age groups was found to be **statistically significant** amongst both child self-report and parent proxy- report.

Conclusion: Evidence continues to accumulate on the importance of HRQOL in all aspects of patient care. QOL scores in different domains of PedsQL scale classified by age groups was found to be **statistically significant** amongst both child self-report and parent proxy-report. QOL scores in different domains of PedsQL scale classified by age at diagnosis was not found to be statistically significant amongst both child self -report and parent proxy-report except for mean school functioning domain in parent -proxy report where it was found to be **statistically significant (p=0.041)**.

Keywords: Quality of life, Thalassemia Major and PedsQL 4.0

ISSN 2515-8260 Volume 10, Issue 2, 2023

Introduction

Thalassemia is a serious public health hazard throughout the Mediterranean region, Africa, the Middle East, the Indian subcontinent, and South-East Asia, with a prevalence ranging from 2% to 25%.[2] Thalassemia affects around 4.4 out of every 10,000 live births globally. Every year, 50,000 to 100,000 children in low- to middle-income countries die from thalassemia major, and around 7% of the global population is a carrier of a hemoglobin disorder.[3]

Beta thalassemia pose a significant health burden in India. The average prevalence of its carriers is 3–4% which translates to 35 to 45 million carriers in our multi-ethnic, culturally, and linguistically diverse population of 1.21 billion people according to the Census of India 2011. Several ethnic groups have a much higher prevalence (4–17%).[4]

Roy and Chatterjee published in-depth interviews with 36 adolescent thalassemic patient responders (ATPRs) from West Bengal, ranging in age from 9 to 17. They determined that thalassemia is extremely stressful, and that children face a variety of physiological, psychological, and social problems.

Many studies across the world show that with the help of existing treatments, life expectancy in persons with thalassemia has grown and mortality rates have dropped. Frequent blood transfusions and iron-chelating treatments (ICTs) have increased the life expectancy of children with Thalassemia Major dramatically, (Brittenham et al.[5] 1994) yet their standard of living remains low. Quality of life (QOL) assessments are important and are frequently required when evaluating health outcomes[1.]

The PedsQL 4.0 Generic Core Scales are multidimensional child self-report and parent proxy-report scales designed to be integrated with the PedsQL Disease-Specific Modules. The PedsQL 4.0 Generic Core Scales contain 23 items suited for healthy school and community groups as well as pediatric populations with acute and chronic health concerns.[6]

The purpose of this study was to provide an overall picture of thalassemic children's health-related quality of life and the factors that impact it. It was also intended to learn about patients' and parents' views of illness progression and QOL.

MATERIAL AND METHODS

STUDY DESIGN

A cross- sectional study was conducted at Rajindra Hospital, Patiala.

INCLUSION CRITERIA

Patients diagnosed with thalassemia major between 5 and 18 years of age reporting to thalassemia ward Rajindra Hospital, Patiala.

EXCLUSION CRITERIA

1) Patients having impaired cognitive function as the condition limited their ability to participate in the study.

ISSN 2515-8260 Volume 10, Issue 2, 2023

2) Parents or the child refused consent/assent for the study were excluded.

SAMPLE SIZE

All the patients satisfying the inclusion-exclusion criteria were enrolled. During the study period of one calendar year starting from 1st April 2021 to 31st March 2022 ,113 participants were included for study.

SAMPLING TECHNIQUE

Thalassemia major patients aged between 5 to 18 years reporting to thalassemia ward for blood transfusion were enrolled consecutively for the study. It is a type of consecutive random sampling**.**

ETHICAL CONSIDERATIONS

Before the start of study due clearance was obtained from the institutional ethics committee. A written informed consent of the parents and assent of the children was obtained. The participants were assured that the confidentiality will be maintained.

DATA COLLECTION

A validated structured proforma containing 2 sections was used to collect data. It was filled by face-to-face interview with the participants and their parents while they were waiting for blood transfusion.

The first section contained questions on socio-demographic profile and history of their disease. Second section was used to assess quality of life. After giving introductory instructions data was collected via self-administrated questionnaire except for the young children (5-7 years), the questionnaire was administered by reading the instructions and each item word by word for the child. The age-appropriate Pediatric Quality of Life Inventory™ (PedsQL™) 4.0 Generic Core Scale (Indian version) was available in both English and vernacular language (Hindi & Punjabi). Parents and children completed the questionnaires independently of one another.

STUDY INSTRUMENT

Pediatric Quality of Life Inventory™ (PedsQL™) 4.0 Generic Core Scale was employed for measuring quality of life. A user agreement was signed with the Mapi Research Institute in Lyon, France for its use. It has two parallel reporting mechanisms, one is child self-reports (age ranges 5-7, 8-12 and 13-18 years) and the other parent proxy-reports (age ranges 5-7, 8-12 and 13-18 years). There are 23 questions covering domains of physical, emotional, social, and school functioning. It asks how much of a problem a particular thing has been for patients during past four weeks over a five-point Likert scale ranging from 0 (never) to 4 (almost always) for all categories except a three-point Likert scale ranging from

ISSN 2515-8260 Volume 10, Issue 2, 2023

0 (Not at all), 2 (Sometimes) and 4 (A lot) was used for the Young Child (ages 5-7) child report.

Items on the PedsQL^{TM} 4.0 Scales were inverted, scored, and linearly converted from an 0-4 scale to a 0-100 scale as follows: 0=100, 1=75, 2=50, 3=25, and 4=0. A higher value over a scale of 0-100 means a better quality of life.

ANALYSIS

The data thus generated was entered into a Microsoft excel spreadsheet and then analyzed by Epi-info CDC Atlanta version7.2.2.1.6. Analysis of the data was done using both descriptive statistics and inferential statistics. Data had been summarized as mean and standard deviation for continuous variables, for categorical variables count and percentages was used. The QOL data collected was not normally distributed and hence non-parametric test, Kruskal Wallis H test where independent variable had three or more groups was used for analysis. A p value of < 0.05 was considered statistically significant.

Results

Table No.1 depicts frequency and percentage distribution of participants according to socio-demographic variables. Out of 113 participants, most of participants (31.0%) were 13-18 years of age with male preponderance, hailing from urban area (50.4%), Hindu by religion (49.6%) with more than 1/3rd (38.1%) studying in primary class.

Table No. 1 SOCIO DEMOGRAPHIC PROFILE OF CHILDREN WITH THALASSEMIA MAJOR

Age Groups (In Years)	Frequency	Percentage (%)
5-7	31	27.4
8-12	35	31.0
13-18	47	41.6
Gender		
Female	48	42.5%
Male	65	57.5%
Residence		
Rural	56	49.6%
Urban	57	50.4%
Religion		
Hindu	56	49.6%
Muslim	2	1.8%
Sikh	55	48.7%
Educational status		

Pre-Primary	9	8.0%
Primary	43	38.1%
Middle	24	21.2%
Secondary School And above	37	32.7%

FIGURE 1: DISTRIBUTION OF PARTICIPANTS ACCORDING TO THALASSEMIA STATUS OF SIBLING (n=113)

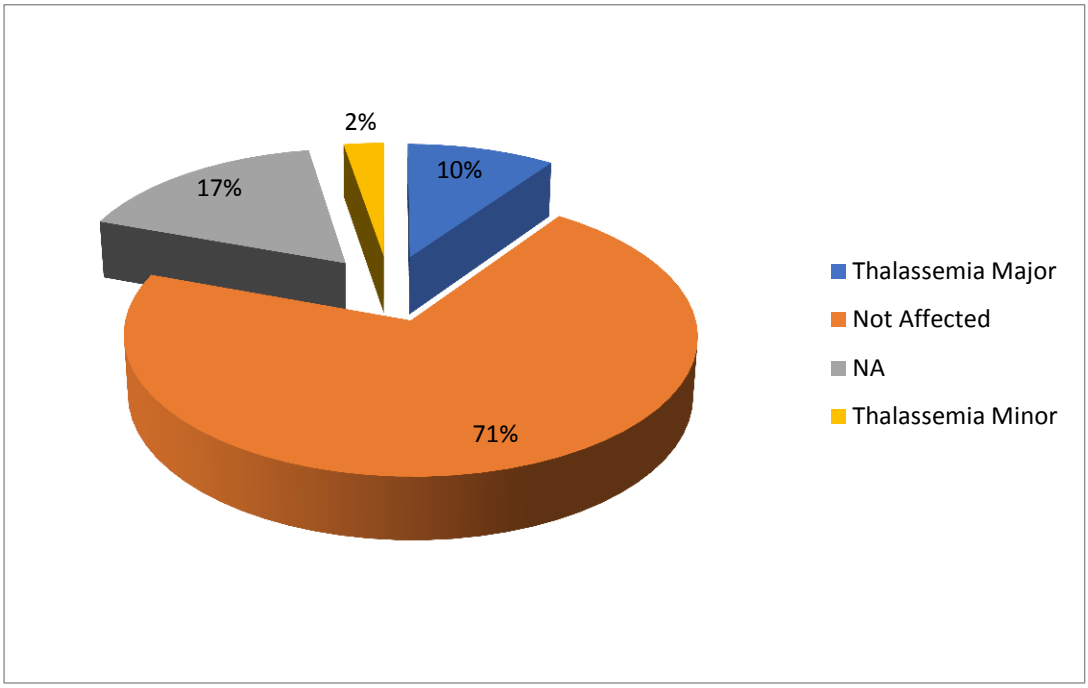

Figure 1 shows that 11 (9.7%) participants had Thalassemia Major affected sibling and 3 (2.7%) participants had siblings with Thalassemia Minor.

TABLE 2: DISTRIBUTION OF PARTICIPANTS ACCORDING TO AGE AT DIAGNOSIS AND TREATMENT VARIABLES (n=113)

Table No.2 shows that 32 (28.3%) participants were <6 months, 48 (42.5%) participants were 6-12 months old while 13 (11.5%) participants had >2 years age at first diagnosis/transfusion.64.6% participants were having 3 weeks blood transfusion interval. Higher number of participants (84.1%) were >2 years of age at start of chelation.

Age at Diagnosis/transfusion	Frequency	Percentage (%)
<6months	32	28.3%
6-12 months	48	42.5%
1-2 years	20	17.7%
>2 years	13	11.5%
Interval of Transfusion		

2 weeks or less	30	26.5%
3 weeks	73	64.6%
4 weeks	9	8.0%
5 weeks or more	1	0.9%
Age at start of chelation		
<1 year	1	0.9%
1-2 year	16	14.2%
>2 year	95	84.1%
Don't Know	1	0.9%

Table 3: Association of QOL score vis-a-vis Age groups (Both child self-report and parent proxy report)

Variable	Domain	Age group (In Years)	N	Mean Rank	Kruskal-Wallis H	df	Asymp. Sig.
CHILD	Mean Physical functioning	5-7	31	67.55	13.56	2	.001
		8-12	35	65.57			
		13-18	47	43.66			
	Mean Emotional functioning	5-7	31	80.06	32.29	2	<0.0001
		8-12	35	61.83			
		13-18	47	38.19			
	Mean social functioning	5-7	31	50.98	7.39	2	.025
		8-12	35	69.21			
		13-18	47	51.87			
	Mean School functioning	5-7	31	71.65	8.88	2	.012
		8-12	35	52.64			
		13-18	47	50.59			
	Total child	5-7	31	71.08	20.37	2	.000
		8-12	35	66.49			
		13-18	47	40.65			
PARENT	Mean Physical functioning	5-7	31	58.90	7.63	2	.022
		8-12	35	67.71			
		13-18	47	47.77			
	Mean Emotional functioning	5-7	31	67.71	11.658	2	.003
		8-12	35	64.00			
		13-18	47	44.72			
	Mean social functioning	5-7	31	53.29	8.320	2	.016
		8-12	35	69.81			
		13-18	47	49.90			
	Mean School functioning	5-7	31	72.34	12.424	2	.002
		8-12	35	58.13			

		13-18	47	46.04			
	Total parents	5-7	31	66.56	17.161	2	.000
		8-12	35	68.80			
		13-18	47	41.90			

In Table No.3, QOL scores in different domains of PedsQL classified by age groups were found to be **statistically significant** amongst both child self-report and parent proxy- report.

Table No. 4: Association of QOL score vis-a-vis Age at Diagnosis (Both child self-report and parent proxy report)

Variable	Domain	Age at Diagnosis	N	Mean Rank	Kruskal-Wallis H	df	Asymp. Sig.
CHILD	Mean Physical functioning	< 6 months	32	51.42	1.529	3	.675
		6 -12 months	48	60.26			
		1-2 years	20	56.25			
		>2 years	13	59.85			
	Mean Emotional functioning	< 6 months	32	49.09	7.607	3	.055
		6 -12 months	48	62.51			
		1-2 years	20	66.10			
		>2 years	13	42.12			
	Mean social functioning	< 6 months	32	52.84	3.005	3	.391
		6 -12 months	48	54.55			
		1-2 years	20	67.18			
		>2 years	13	60.62			
	Mean School functioning	< 6 months	32	47.89	4.335	3	.227
		6 -12 months	48	59.00			
		1-2 years	20	66.10			
		>2 years	13	58.04			
	Total score	< 6 months	32	48.02	4.869	3	.182
		6 -12 months	48	60.05			
		1-2 years	20	66.88			
		>2 years	13	52.65			
PARENT	Mean Physical functioning	< 6 months	32	54.41	3.734	3	.292
		6 -12 months	48	62.38			
		1-2 years	20	46.35			
		>2 years	13	59.92			
	Mean Emotional functioning	< 6 months	32	47.03	4.489	3	.213
		6 -12 months	48	62.47			
		1-2 years	20	59.65			
		>2 years	13	57.27			
	Mean social	< 6 months	32	56.45	.107	3	.991

	functioning	6 -12 months	48	57.01			
		1-2 years	20	56.18			
		>2 years	13	59.58			
	Mean School functioning	< 6 months	32	44.73	8.241	3	.041
		6 -12 months	48	60.15			
		1-2 years	20	69.85			
		>2 years	13	55.81			
	Total score	< 6 months	32	48.19	3.674	3	.299
		6 -12 months	48	62.49			
		1-2 years	20	57.43			
		>2 years	13	57.77			

In Table No.4, QOL scores in different domains of PedsQL scale classified by age at diagnosis was not found to be statistically significant amongst both child self -report and parent proxy- report except for mean school functioning domain in parent -proxy report where it was found to be statistically significant (p=0.041).

Discussion

In terms of socio-demographic variables in the present study **(Table No.1),** majority (41.6%) participants were between the age of 13-18 years, followed by 31.0% in 8-12 years age group. **Alshamsi S et al[7] (2021)** conducted a study on a total of 68 children with transfusion-dependent thalassemia aged 2-18 years. Among them, 55.9% fell into the 13–18-year age group**.** This finding was in concordance with the present study**.** Higher number of participants were male (57.5%) in the present study. Male: Female Ratio was 1.4:1. Similar results were seen in a study by **Ankush A et al[8] (2018)**, in which majority (61.1%) were male children and 38.9 % were females. Majority of the participants were from urban area (50.4%) in the current study. This was concordant with the results as observed in the study done by **Tanveer T et al[9] (2018) which** 59.7% participants were living in urban areas**.**

In the present study**,** higher number of participants were Hindu (49.6%)**.** This was in agreement with the results as observed in the study done by **Ankush A et al[8] (2018)** in which majority of the participants (52.8%) were Hindu. In current study**,** most of the participants were in Primary class (38.1%) and 32.7% were in ≥ secondary school. It is in line with results of the study done by **Surapolchai P et al[10] (2010)** in which 31.7% and 32 % participants were in primary class and ≥ secondary school respectively.

In the present study **(Fig. No.1)**, majority of the participants (70.8%) had siblings' unaffected by thalassemia, similar study by **Tanveer T et al[9] (2018)** also reported that majority (78.7%) participants had no history of thalassemia affected sibling.

Table No. 2 depicted that majority of the participants were diagnosed between 6-12 months of age (42.5%). In a study done by **Nashwan AJ et al[11] (2018),** 90% patients were diagnosed between 6 -12 months age and **Caocci G et al[12] (2012)** also observed that beta thalassemia major was diagnosed at a median age of 8 months. These findings are in concordance with present study. However, in a study by **Ismail et al[13] (2018),** 75% of

participants had thalassemia diagnosis at <6 months of age. It might be due to increased awareness and better health seeking behavior of people in that study area.

Current study also depicted that around 60% participants had started blood transfusion from 6 months - 2 years. In a similar study done by **Caocci G et al[12] (2012)** median age at first transfusion was 11.5 months ranging from 2-60 months. Current study showed that significantly higher number of participants (64.6%) had 3 weeks interval of blood transfusion. A study by **Ayoub MD et al[14] (2013)** observed that most patients (84.8%) required three weekly blood transfusions. This was in concordance with present study. Discordant results were reported from the study conducted by **Ismail et al[13] (2018)** where the frequency of transfusion is every 2 weeks in majority (53%) participants. Individual response to anemia, status of spleen and degree of ineffective erythropoiesis can be few of the factors leading to this difference in figures. It was also observed that higher number of participants were >2 years of age at start of chelation (84.1%). In a study done by **Caocci G et al[12] (2012)** median age at start of iron chelation was 49 months which is almost in concordance with present study.

In **Table No. 3**, QOL scores in different domains of PedsQL scale classified by age groups was found to be **statistically significant** amongst both child self-report and parent proxy- report. Similarly, in a study done by **Surapolchai P et al[10] (2010)** age was significantly associated with physical functioning domain (p=0.047) and total summary score (p=0.035) in parent proxy report.

In **Table No. 4**, QOL scores in different domains of PedsQL scale classified by age at diagnosis was not found to be statistically significant amongst both child self -report and parent proxy- report except for mean school functioning domain in parent -proxy report where it was found to be **statistically significant (p=0.041)**.However, no statistically significant difference was observed with age at diagnosis in study done by **Caocci G et al[12] (2012)** in total summary score amongst both child self -report and parent proxy- report**.**

Conclusion

Although improving a patient's prognosis and overall survival may be the main goal of the modern treatment modalities, equally important to many patients is the quality of life.

The present study highlighted a unique perspective of HRQOL by evaluating both child self -report and parent -proxy reports. QOL scores in different domains of PedsQL scale classified by age groups was found to be **statistically significant** amongst both child self-report and parent proxy- report. QOL scores in different domains of PedsQL scale classified by age at diagnosis was not found to be statistically significant amongst both child self -report and parent proxy- report except for mean school functioning domain in parent -proxy report where it was found to be **statistically significant (p=0.041)**.

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