Original Research Article

Quality of life in children with thalassemia major following up at a tertiary care center in India (GOTQoL)

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Received: 03 November 2018

Accepted: 29 November 2018

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ABSTRACT

Background: This study aims to measure the quality of life (QoL) scores in children with thalassemia major following up at a tertiary care center for routine blood transfusion in comparison to healthy children.

Methods: A case control study design was adopted, wherein on the QoL of 36 children with thalassemia in the age group 5 to 18 following up for blood transfusion at Goa Medical College, was measured using PedsQL™ 4.0. This was compared to the QoL in age and gender matched healthy children from a government school. A higher score on a subscale indicates better quality of life on this instrument.

Results: The children with thalassemia had lower mean scores on physical (67.85 vs 84.24; $P < 0.001$), social (78.34 vs 87.95; $P = 0.002$) and school (62.64 vs 79.48; $P < 0.001$) functioning subscales compared to the healthy children. They also had lower mean psychosocial summary score (73.32 vs 82.01; $P = 0.003$) and total health summary score (71.95 vs 82.57; $P < 0.001$). The physical functioning subscale had significantly higher score among the children who were on chelation and also among the ones whose parents reported as being informed about the condition.

Conclusions: The children with thalassemia have poor QoL in physical, social and school functioning domains. Improvement in QoL requires consolidated efforts on part of doctors, parents, school authorities and policy makers. These patients should be provided with low cost-effective chelation therapy. The parents need to be counselled about this disease by the treating team.

Keywords: Goa, India, PedsQL 4.0, Quality of Life, Thalassemia major

INTRODUCTION

Thalassemia syndrome is one of the common inherited blood disorders. It is characterized by genetic defect in production of alpha or beta globin chains and is inherited in an autosomal recessive manner. Thalassemia major is the homozygous form of deficiency of beta globin chain synthesis which results in a severe transfusion dependent anemia and which becomes apparent in first six months. This disease is a major public health problem in

Mediterranean, the Middle East, Indian subcontinent and the South East Asia.¹ According to Modell and Darlison estimates published in 2008, there are 6,583 children with β thalassemia annually conceived in India, out of which 3,227 are β thalassemia major.² This chronic condition by itself is debilitating and subsequent iron overload makes them prone to complications like heart failure, hypogonadism, hypothyroidism and diabetes. Together it severely affects the child's physical competence and scholastic performance. The bone deformities and typical

facial appearance negatively impacts their self-esteem, giving them always the feeling of being different affecting their social relationships and psychological well-being.

Over the years, with regular transfusions and effective chelation, authors have been able to achieve longer survival rates.³ But these gains will be of less value if they are not accompanied with a better quality of life (QoL). Measuring QoL is not an easy task, especially in children, but there is evidence that school-age children can self-report on their health which provides a viable means of understanding internal experiences of health and distress.⁴ In India, much of the research regarding this has been centered on qualitative aspects of it. In 2007, Roy and Chatterjee published in-depth interviews of 36 adolescent thalassemic patient respondents (ATPRs) from the age group 9 to 17 from West Bengal and found that the consequence of thalassemia is extremely stressful, and patients face a variety of physical, psychological and social problems.⁵ The primary objective of this study is to report the effect of this disease and its treatment on the health related QoL of these patients from this part of India. Authors will also try to understand the factors affecting the QoL which can help us arrive at the possible interventions that can improve it. The study will attempt to fill the lacunae in the literature regarding this area and contribute to the global pool of similar studies. The results of parent proxy questionnaire will be reported in a separate manuscript.

METHODS

This study was conducted in the Pediatric ward of Goa Medical College, which is a tertiary care hospital in Goa, where children with thalassemia would follow up for routine blood transfusion. The Goa Medical College has been providing free services, including blood transfusions for children with thalassemia. The study period was from November 2012 to February 2013. Goa is a small state on the west coast of India with a population of 1.46 million (census 2011).⁶ The hospital caters to patients from Goa and neighboring districts from states of Maharashtra and Karnataka as well.

Inclusion criteria

- The inclusion criteria for this study were children in age group 5 to 18, diagnosed with thalassemia major, registered and following up at the hospital for at least past three months.

Exclusion criteria

- Children who were cognitively impaired and where parents or the child refused to consent/assent for the study were excluded.

All the patients satisfying the inclusion-exclusion criteria were included. A case control study design was adopted

to compare the impact of the thalassemia on the quality of life of the participants. Controls were matched for gender and age and selected from a school, situated in the same district as the medical college.

Study instruments

The main instrument for this study was Pediatric Quality of Life Inventory™ (PedsQL™) 4.0 Generic Core Scales. This questionnaire has been used in QoL studies among children worldwide. 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 2-4, 5-7, 8-12 and 13-18 years). It measures the amount of problem as how much in past four weeks over a five-point Likert scale ranging from 0 (never) to 4 (almost always) and three point for young children. There are 23 questions covering domains of physical, emotional, social and school functioning. The raw scores are transformed into standardized scores and then it becomes a unidirectional positive scale, i.e., a higher value over a scale of 0-100 means a better quality of life.⁷ Good internal consistency and validity in children with chronic health conditions as well as in healthy children and adolescents has been shown by the original PedsQL.⁸ In India, the Hindi version of PedsQL was validated by Sebi Das et al.⁹ In a separate study it the instrument has shown to differentiate between HIV infected and uninfected children.¹⁰ Because some of our patients knew only Konkani (the regional language of state of Goa), the instrument was translated into Konkani (Devanagiri and Roman script). This followed the guidelines prescribed by Mapi Institute for translation to generate a version conceptually equivalent to the original version, as well as clear and easy to understand. The forward translation was done by two native speakers of Konkani language and bilingual in the source language, this was followed by backward translation by an independent translator. The final version so prepared was tested in a pilot study. The reports produced at each stage and the final translated version were mailed to the author of the instrument, who approved its use for this study.

Authors also collected general socio demographic and anthropometric data, medical history, transfusion and chelation status, routine laboratory test results and asked two closed ended questions to the parents. The first question was “Do they consider themselves fully informed about the condition” and the next one asked, “if they knew about bone marrow transplantation”.

Ethics: The study was approved by the Institutional Ethics Committee at Goa Medical College. Informed consent was taken after providing information about the study. Informed consent was signed by the parent for children and an assent was taken from the child. They were informed that they can skip any question they feel uncomfortable answering and that the services that they

receive in this hospital will not be affected by their refusal to participate or by what response they give to these questions. Strict confidentiality was maintained throughout the study. Permission from the school authorities was also taken.

Data collection: Patients in age group of 5 to 8 were asked questions by trained interviewers. Patients in age group of 8 to 12 and 13 to 18, were either asked to fill by themselves if they could do so or were interviewed otherwise. They were asked to participate in this study during their waiting period for getting blood transfusion. While collecting data from healthy children who acted as controls for this study, a preliminary physical examination was performed and any child with history of acute or chronic illness was excluded. These children from respective age groups were either interviewed or asked to fill by themselves.

Statistical analysis

The collected data was entered into Microsoft Excel 2007 and then subsequently analyzed in SPSS (Statistical Package for the Social Sciences) version 19.0. The collected scores were transformed according to the scoring guidelines. To calculate the summary score for each subscale, the sum of the items was divided by the number of items answered. But if more than 50% of the items in the scale are missing, the scale scores were not computed. To calculate the total health summary, score the sum of all the items was divided by the number of items answered on all the scales and for psychosocial health summary score the sum of emotional, social and school were divided by the number of items answered on all these scales.¹¹ The data collected was not normal and hence non-parametric tests, Mann Whitney U test where the independent variable had two groups and Kruskal-Wallis H test where independent variable had three or more groups were used for analysis. A p value <0.05 was considered statistically significant.

RESULTS

Quality of life of children following up at the tertiary care hospital were studied by adopting a case control study design. During the study period, 62 children came for regular blood transfusion. After applying filters for age, diagnosis and consent, 52 children in the age group 3-18 satisfied the inclusion-exclusion criteria. Parents of these children were asked to fill the parent proxy PedsQL questionnaires. Out of these children, 36 were in the age group 5-18, and child proxy questionnaire was filled for/by all of them, the response rate was 100%. There were 22 (61.1 %) male children and 14 (38.9 %) females. The control group was selected from a school in the same district as the medical college. The control arm consisted of 50 children with similar sex distribution as those with thalassemia-30 (60.0 %) males and 20 (40.0 %) females, as presented in Table 2. The age group distribution in each category was also similar.

Table 1: Socio-demographic characteristics of the children with thalassemia.

Patient characteristics (n=36)	N (%) / Mean (SD)
Gender	
Male	22 (61.1)
Female	14 (38.9)
Age	
<8	11.66 (4.73)
≥8 and <13	10 (27.8)
≥13	16 (44.4)
Religion	
Hindu	19 (52.8)
Muslim	13 (36.1)
Christian	04 (11.1)
Birth order	
First	21 (58.3)
Second	10 (27.8)
Third or more	05 (13.9)
Family history	
Yes	08 (22.2)
No	28 (77.8)
Consanguinity	
Yes	08 (22.2)
No	28 (77.8)
Family income in INR (n=34)	
<5000	10
5000-14999	16
15000+	8

Table 2: Clinical characteristics of children with thalassemia.

Patient characteristics	N (%) / Mean (SD)
Age at diagnosis (in months)	7.37 (6.52)
Age at first transfusion (in months)	7.86 (6.76)
Pretransfusion Hb (on this visit)	
< 7.5000	10 (27.8)
≥7.5 and <9	09 (25.02)
≥9	17 (47.25)
Transfusion frequency	
Once a month	24 (66.7)
Twice a month	12 (33.3)
Chelation	
Yes	25 (69.4)
No	11 (30.6)
Type of chelation	
Oral	23 (63.9)
Subcutaneous	02 (5.6)
Parents consider themselves informed about the condition	
Yes	18 (50)
No	16 (44.4)
Not sure	1 (2.8)
Parents know about bone marrow transplantation	
Yes	14 (38.9)
No	17 (47.2)
Not sure	4 (11.1)

Table 3: Characteristics of the participants in the control group.

Control characteristics (n=50)	N (%) / Mean (SD)
Gender	
Male	30 (60.0)
Female	20 (40.0)
Age	11.28 (3.23)
<8	12 (24)
≥8 and <13	16 (32)
≥13	22 (44)

On asking about family history of same disease 8 (22.0 %) patients gave a positive family history. Interestingly, in Table 1 authors see similar number 8 (22.0%) patients born out of consanguineous marriage.

However, only one patient was positive for both these findings. Parents of two children chose not to divulge

information regarding their family income. Twenty-five (69.4%) children were on chelation, out of which only two were on subcutaneous and rest on oral (Table 2).

The total QoL score was found to be (71.95±14.24) with the highest in the emotional (78.96±17.8) and social (78.34±15.26) domains followed by physical (67.85±20.27) and lastly lowest in scholastic domain (62.64±18.54). The score for psychosocial domain was (73.32±14.37). Mann-Whitney test U test was run to determine if there were differences in quality of life scores between healthy (control) children and children with thalassemia.

As displayed in Table 4, there was a significant difference in all domains except emotional. The total quality of life score for healthy children (Mdn=83.05) was higher than the children with thalassemia (Mdn=75.55), U=492, z =-3.572, p <0.001.

Table 4: Quality of life scores in children with thalassemia (cases) vs healthy participants (controls).

Quality of life subscale	Mean (SD)	Median	Mean ranks	Mann-Whitney U	z statistic	P-value
Physical functioning						
Cases	67.85 (20.27)	70.31	30.17	420	-4.212	<0.001
Controls	84.24 (14.42)	87.50	53.1			
Emotional functioning						
Cases	78.96 (17.8)	80.00	44.88	850.5	-0.437	0.662
Controls	78.6 (14.85)	80.00	42.51			
Social functioning						
Cases	78.34 (15.26)	80.00	33.53	541	-3.172	0.002
Controls	87.95 (11.75)	90.00	50.68			
School functioning						
Cases	62.64 (18.54)	70.00	30.65	437.5	-4.069	<0.001
Controls	79.48 (14.77)	80.00	52.75			
Psychosocial health summary score						
Cases	73.32 (14.37)	73.33	34.69	583	-2.779	0.005
Controls	82.01 (9.85)	82.08	49.84			
Total health summary score						
Cases	71.95 (14.24)	75.55	32.17	492	-3.572	<0.001
Control	82.57 (9.81)	83.05	51.66			

The quality of life scores classified by characteristics of the patients are presented in Table 5 and 6. A statistically significant result was found in physical domain between the children with thalassemia who were on chelation (72.7±19.24) than the ones who were not (57.82±22.48), p=0.024.

Parent's knowledge about the disease was associated with higher scores in physical domain but not in the other domains. Compared to the knowledge about the disease where majority (50%) considered themselves informed

about the condition, majority (47.2%) were not aware about bone marrow transplantation.

This awareness was associated with higher scores in physical (p=0.046), social (p=0.013), scholastic (p=0.025) and eventually the total scores (p=0.008).

There was lack of any significant difference in quality of life scores with other factors (gender, age, religion, birth order, family income, family history, transfusion frequency).

Table 5: Quality of life scores classified by demographic characteristics of the patients with thalassemia.

Variables	Physical	Emotional	Social	Scholastic	Psychosocial	Total
Gender						
Male	69.32 (19.67)	77.73 (18.74)	78.18 (14.02)	57.5 (19.51)	71.14 (14.49)	70.69 (14.43)
Female	65.53 (21.73)	80.9 (16.69)	78.57 (17.59)	70.71 (13.99)	76.73 (14)	73.93 (14.22)
P-value	0.649	0.633	0.624	0.06	0.217	0.27
Age						
<8	66.25 (28.75)	74.5 (22.39)	70 (22.12)	58 (23)	67.5 (20)	67.19 (21.07)
≥8 and <13	68.31 (10.41)	85.25 (11.09)	79.5 (11.17)	65.5 (19.65)	76.75 (11.96)	74.64 (10.07)
≥13	68.56 (19.88)	77.82 (17.98)	82.81 (10.33)	63.75 (15.23)	74.8 (11.09)	73.24 (11.14)
P-value	0.87	0.475	0.246	0.687	0.567	0.815
Religion						
Hindu	69.84 (18.31)	81.58 (16.57)	83.68 (10.39)	68.16 (13.57)	77.81 (10.38)	75.82 (10.34)
Muslim	64.67 (24.66)	75.2 (20.02)	70 (18.26)	60 (21.7)	68.4 (17.71)	67.47 (17.87)
Christian	68.75 (16.74)	78.75 (18.43)	80 (15.82)	45 (19.58)	67.92 (15.36)	68.13 (15.64)
P-value	0.861	0.745	0.092	0.106	0.155	0.32
Birth order						
First	67.27 (21.17)	80.48 (15.14)	77.14 (16.85)	63.57 (16.6)	73.74 (13.52)	72.12 (13.36)
Second	73.62 (17.52)	81.75 (17.25)	83 (10.33)	66.5 (18.12)	77.09 (11.21)	76.22 (11.64)
Third or more	58.75 (21.81)	67 (27.3)	74 (17.11)	51 (26.08)	64 (21.56)	62.69 (20.58)
P-value	0.702	0.871	0.674	0.669	0.803	0.67
Family history						
Yes	63.29 (9.27)	77.5 (15.12)	81.25 (8.77)	63.13 (22.19)	73.96 (12.57)	71.29 (9.93)
No	69.15 (22.42)	79.38 (18.72)	77.5 (16.7)	62.5 (17.83)	73.13 (15.05)	72.14 (15.4)
P-value	0.17	0.616	0.673	0.909	0.924	0.621
Family income (n=33)						
<5000	64.07 (18.48)	81 (19.56)	75.5 (12.13)	59.5 (22.54)	72 (16.76)	70.02 (16.28)
5000-14999	66.8 (23.44)	75.47 (17.83)	76.88 (17.88)	60.31 (19.02)	70.89 (14.66)	69.87 (14.34)
15000+	76.4 (15.28)	85 (13.37)	83.75 (15.3)	72.5 (10)	80.42 (11.41)	79.42 (11.54)
P-value	0.519	0.473	0.449	0.348	0.269	0.25

Table 6: Quality of life scores classified by clinical characteristics of the patients with thalassemia.

Variables	Physical	Emotional	Social	Scholastic	Psychosocial	Total
Pre-transfusion Hb (on this visit)						
<7.5000	62.06 (17.87)	83 (19.89)	79.5 (13.43)	61.5 (22)	74.67 (16.98)	71.52 (15.35)
≥7.5 and <9	73.27 (16.02)	76.39 (17.82)	78.33 (14.15)	69.44 (14.68)	74.73 (12.62)	74.36 (12.39)
≥9	68.39 (23.52)	77.95 (17.22)	77.65 (17.52)	59.71 (18.33)	71.77 (14.32)	70.92 (15.15)
P-value	0.403	0.478	0.903	0.451	0.607	0.94
Transfusion frequency						
Once a month	68.49 (21.24)	74.38 (19.18)	75.83 (16.66)	61.46 (18.97)	70.56 (14.93)	70.04 (15.28)
Twice a month	66.56 (19.02)	88.13 (10.07)	83.33 (10.95)	65 (18.22)	78.82 (11.88)	75.76 (11.54)
P-value	0.579	0.043	0.224	0.447	0.122	0.33
Chelation						
Yes	72.7 (19.24)	78.3 (15.78)	77.6 (16.41)	63.2 (17.79)	73.04 (13.47)	72.95 (13.52)
No	56.82 (18.9)	80.46 (22.53)	80 (12.85)	61.36 (20.99)	73.94 (16.92)	69.66 (16.21)
P-value	0.024*	0.403	0.653	0.876	0.655	0.693
Parents consider themselves informed about the condition						
Yes	76.67 (12.22)	81.25 (14.36)	80.28 (13.01)	65 (17.58)	75.51 (11.45)	75.8 (10.42)
No/Not sure	57.82 (22.48)	77.66 (20.89)	77.81 (17.13)	60.94 (20.11)	72.14 (17.6)	68.56 (17.62)
P-value	0.013*	0.902	0.903	0.639	0.678	0.469
Parents know about bone marrow transplantation						
Yes	77.68 (13.26)	84.47 (14.82)	84.29 (14.4)	71.79 (16.83)	80.18 (12.2)	79.56 (11.04)
No/Not sure	62.44 (21.85)	76.67 (18.52)	74.29 (15.19)	56.19 (17.68)	69.05 (14.5)	67.4 (14.39)
P-value	0.046*	0.188	0.072	0.013*	0.025*	0.008*

* P-value<0.05

DISCUSSION

There is indeed a dearth of studies measuring QoL in thalassemia. With adequate search for similar studies in India in PubMed, which employs direct interviewing of children with thalassemia major and matched controls using PedsQL as the study instrument, this appears to be first of its kind study done in this country.

The number of children born out of consanguineous marriages were higher (22.2%) compared to the general prevalence of consanguineous marriages reported in NFHS-2 (14.4%) for state of Goa.¹² This finding is consistent with another study done in West Bengal on thalassemia patients, where the prevalence was found to be 22.7%, and is probably as expected for an autosomal recessive disease like Thalassemia.^{13,14}

The high scores in social domain and poor in scholastic are consistent with findings of Saha et al and also Khurana et al. In the later study where 80% reported no impact on social life and 70% expressed that thalassemia had an adverse impact on education, linked the observed lack of adverse effect on social life on close-knit nature of Indian families.^{13,15} The finding of poor scores in scholastic domain among all other domains has also been reported in studies from Thailand, Malaysia, Jordan and Saudi Arabia by Thavorncharoensap et al (67.89±15.92), Ismail et al (60.14±16.41), Gharaibeh et al (59.15±16.30) and Ayoub et al (54.3±24.3) respectively.¹⁶⁻¹⁹ The possible reason for this pertinent finding could be the inherent nature of the disease causing the child facing fatigue and activity intolerance to lag in academics and when this gets coupled with frequent absenteeism for taking blood transfusions, it eventually does take a toll on the child's overall school functioning. There is also evidence suggesting potential role of hemosiderosis on cognitive functioning in β thalassemia major.²⁰

Comparing findings of this study with other studies done in India where controls were used, the results of thalassemic children scoring significantly lower at all subscales is consistent with the finding of Ajij et al, although the study used WHO-BREF questionnaire and included only adolescents as subjects.²¹ Sachdeva et al assessed QoL of 26 children with thalassemia major from North India, where each child acted himself as control for their best possible QoL. They employed a modified Pediatric Cancer Quality of life scale as instrument and found QoL affected (<90%) in 88% of patients and severely affected (<70%) in 15% of patients.²²

Ismail et al also did not find any significant difference in emotional domain but Gharaibeh et al did find a significant difference albeit it was the lowest mean difference among all domains.^{17,18} In view of these findings and known psychosocial problems in children with thalassemia like depression, anxiety, impact on body image and self-esteem, the relationship seems to be complex.¹⁵ Tsiantis et al have argued about presence of

various adjustments and coping mechanisms in thalassemic children and their families toward emotional and behavioral problems.²³ The relationship between emotional problems of children with thalassemia and their quality of life certainly needs more research for better understanding.

Surapolchai et al found household income and type of payment were significantly associated with total score.²⁴ Authors did not find any such association probably because transfusions and other care were being provided free of cost at this center. Thavorncharoensap et al reported iron chelation therapy having a significant negative effect on QoL in the school functioning subscale and in contrast frequency of blood transfusion not significantly related to QoL.¹⁶ But this study with a sample size of 315 children with thalassemia had only 15 children with homozygous β thalassemia and included only subcutaneous injection treatment as chelation in questionnaire.

Whereas out of twenty-five, only two of our patients were on subcutaneous chelation. As patients with thalassemia major are diagnosed early and need frequent blood transfusions, this makes it imperative to study QoL scores separately in these patients. The significant difference in physical health subscale scores between the ones on chelation and others not, needs some caution while interpretation. While on one hand iron overload can cause complications like biventricular cardiomyopathy which itself can cause poor quality of life scores but so can 12 hourly five times a week subcutaneous iron chelation therapy. Caocci et al in a study of 60 children with thalassemia in Middle Eastern countries concluded that delayed start of iron chelation has a negative impact on children's HRQoL.²⁵ There is indeed a lack of independent studies measuring difference in QoL following chelation therapy and between oral vs subcutaneous. Oral chelation has been perceived with better QoL, but it's equally important that efficacy of chelation itself doesn't become a trade-off in the long run.²⁶ In fact, oral chelation is itself known to produce arthritis, abdominal pain, diarrhea and vomiting as side effects.²⁷ A lot of strides have been made in the direction for providing better care to children with thalassemia, ranging from first approval and marketing of deferiprone in India to inclusion of thalassemia care in 12th five year plan by the Working Group.^{28,29}

The answers to our two closed ended questions at the end of the general questionnaire shows the inadequate awareness about this disease and its potential treatment. The higher scores seen in children where parents were aware of bone marrow transplant could well be due to better educational background and socioeconomic status of the family. Although, authors did not find any significant difference between scores of children from different strata of family income, the study did not measure education status of parents. In other chronic pediatric diseases like epilepsy it has been shown that

knowledge of, perception of, and attitudes toward epilepsy affect the quality of life of children and of their parents.³⁰ In a country where in some kinships this disease is perceived to be corrupting blood (Rakter dosh), stigmatization is quite possible and only awareness among the patients, their family members and general public can break these cultural notions.³¹

The major limitation of this study was a small sample size, as it is restricted to children with Thalassemia Major. A larger sample size would have helped authors to perform reliable predictive analysis, assessing the factors affecting the quality of life on these children.

CONCLUSION

Thalassemia is a chronic debilitating disease and severely affects the quality of life. The children with thalassemia have poor QoL in physical, social and school functioning domains. Improvement in QoL requires consolidated efforts on part of doctors, parents, school authorities and policy makers. These patients should be provided with low cost-effective chelation therapy. The parents need to be counseled about this disease by the treating team. In developing countries like India, where bone marrow transplant is not a viable option for many, this makes a case for providing free or subsidized chelation therapy for these patients. Parents, school authorities and health care providers have to come together to ensure these children don't lag in their academics. Possible solutions at the hospital end could be like providing transfusion services on weekends to reduce frequent absenteeism.

The role of imparting better understanding among the parents regarding this disease cannot be emphasized any less. The first "complete talk" happens only when the child is diagnosed but it is important that it is revamped and advanced as the child grows, as it will help them in coping with the disease better and make the child him/herself understand it better. Doctors, counsellors and thalassemia societies have an important responsibility in this regard.

ACKNOWLEDGEMENTS

Authors would like to thank Purificasao Coutinho Memorial Project (Sangath) for their support, Department of Pediatrics, Goa Medical College for allowing to conduct this study, Dr. James Varni and Mapi Research Institute for giving permission to use Pediatric Quality of Life Inventory (PedsQL 4.0); Mr. Gurunath Pai and Dr. Kalyani Pai for forward translation of PedsQL in Konkani (Devanagari script), Dr. Joyce Barreto in Roman script, Dr. Anisha Deulker for backward translation; Dr. Shahin Amroskar and Dr. Joyce Barreto for their help during data collection from healthy school children and Dr. Lorraine D'Sa for her valuable inputs.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee, Goa Medical College, Goa, India

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Cite this article as: Ankush, Dias A, Silveira MP, Talwadker Y, Souza JP. Quality of life in children with thalassemia major following up at a tertiary care center in India (GOTQoL). *Int J Contemp Pediatr* 2019;6:168-75.