Indian J Pediatr (March 2017) 84(3):188–194
DOI 10.1007/s12098-016-2267-z

ORIGINAL ARTICLE

Quality of Life in Children with Thalassemia and their Caregivers in India

Sapna Sharma[1] · **Bageshree Seth**[1] · **Prashant Jawade**[2] · **Madhavi Ingale**[3] · **Maninder Singh Setia**[4]

Received: 20 May 2016 / Accepted: 17 November 2016 / Published online: 7 December 2016

Abstract
Objectives To assess and compare the Quality of Life (QOL) of children with beta-thalassemia major on regular transfusion therapy with normal children, and of the caregivers of children with beta-thalassemia major to that of caregivers of normal children.
Methods A cross-sectional comparison of QOL in 75 thalassemic and 80 non-thalassemic children was conducted using the PedsQL™ 4.0 generic core scale. Also self-rated health was assessed in their caregivers using Short Form-36 Health Survey.
Results The total QOL score according to child-self report [83.7 (10.8) *vs*. 97.6 (3.3); $p < 0.001$] and parent-proxy report [84.2 (11.9) *vs*. 96.7 (3.5); $p < 0.001$] was significantly lower in cases as compared with controls. It was found that a significantly higher proportion of caregivers of cases reported poor health compared with caregivers of controls (29.2% *vs*. 2.5%, $p < 0.001$). Even after adjusting for age, sex, socio-economic status, and total QOL score by the parent, it was found that caregivers of thalassemic children were significantly more likely to report poor health compared with those of controls (odds ratio: 15.8, 95% confidence intervals: 2.8–89.9).
Conclusions Health Related QOL is significantly affected in children with beta-thalassemia major on regular transfusion across all age groups, gender and socio-economic classes and also in their caregivers.

Keywords Thalassemia · Quality of life · Peds QOL generic core scale · Caregivers

✉ Bageshree Seth
bageshrees@rediffmail.com

[1] Department of Pediatrics, Mahatma Gandhi Mission's Medical College, Sector –1, Kamothe, Navi-Mumbai, Maharashtra 410209, India

[2] Navi-Mumbai Municipal Corporation General Hospital, Sector 10A, Vashi, Navi-Mumbai, Maharashtra, India

[3] Thalassemia Day Care Centre, Navi-Mumbai Municipal Corporation General Hospital, Sector 10A, Vashi, Navi-Mumbai, Maharashtra, India

[4] MGM Institute of Health Sciences, Sector –1, Kamothe, Navi-Mumbai, Maharashtra, India

Introduction

The worldwide prevalence of annually affected conceptions with Beta-Thalassemia is 42,409 cases with annual births worldwide being 1,28,667,000. The South East Asia alone accounts for 21,693 annually affected conceptions with annual births being 38,139,000 [1]. Every year, 10,000 children with thalassemia are born in India [2].

While significant advances have been made in the medical management of thalassemia, this development has not been matched by progress in rehabilitation of thalassemia patients [3]. Various Quality of Life (QOL) studies have been conducted worldwide for thalassemia [4–6] but these studies may not reflect the picture in India. Literature review of paediatric journals revealed a relative lack of studies that address QOL among Indian children with thalassemia.

The therapeutic regime of thalassemia is complex, lifelong and inconvenient, requiring repeated hospitalisations and blood transfusions, which often affects the child's physical and mental health adversely [7]. For chronic diseases such as thalassemia, where a cure is not affordable to most and treatment may be prolonged, Health-related Quality of Life (HRQOL) is likely to be an essential outcome when

considering options for treatment for individual patients and the allocation of healthcare resources [4].

As children are less able to voice their concerns and are more vulnerable than adults, the assessment of HRQOL in children is essential for the provision of proper care, since it helps in identifying the impact of the disease and treatment from children's perspective. A better understanding of the factors associated with HRQOL among children and adolescents with thalassemia could have a direct effect on the development of more suitable clinical, counselling and social support programs to enhance treatment outcomes, especially in terms of HRQOL of these patients.

This study aims to bridge the void in the information available on the QOL of Indian children with beta-thalassemia major. The study also explores the impact of the disease on the caregivers.

Material and Methods

The present study is a prospective comparative study conducted in the Pediatric day care centre and outpatient department of Mahatma Gandhi Mission's Medical College and Hospital and Navi-Mumbai Municipal Corporation (NMMC) Hospital, Navi-Mumbai, India during April through September 2014.

Children with beta-thalassemia major in the age group of 2–18 y who were receiving regular blood transfusion and their caregivers were taken as cases. Beta-thalassemia children who suffered from any other major illness were excluded from the study. Children in the age group of 2–18 y, not having beta-thalassemia major, who were attending the OPD for minor ailments and their caregivers were controls in the study. Children suffering from any major medical or surgical illnesses were excluded from the study.

The sample size consisted of 75 beta-thalassemia major children and 80 controls along with their caregivers.

Informed consent was taken from the caregivers of the participants in the study. The demographic details were obtained from the participants using the case study form. The Kuppuswamy's socioeconomic status scale revised for 2012 was used for the socioeconomic status classification. After having received requisite permissions from MAPI trust, Pediatric Quality of Life Inventory (PedsQL) Generic core scale 4.0 questionnaire was used for assessing the QOL of children with beta- thalassemia major as well as normal children.

The pediatric quality of life inventory (PedsQL™) questionnaire was administered to cases and controls (Child-Self Report) as well as their caregivers (Parent-Proxy Report). For children 2–4 y, only the parents answered the questionnaire. This questionnaire comprises of 5 essential core domains namely: Physical Functioning, Emotional Functioning, Social Functioning, School Functioning and Psychosocial Functioning for Pediatric HRQOL measurement [8, 9]. The Psychosocial Health Summary Score represents the sum of items answered in the Emotional, Social and School functioning scales [10]. Each item is rated on a 5-point Likert scale from 0 (Never) to 4 (Almost always). The child self and parent proxy-reports of the PedsQL measurement model are sensitive to cognitive development and contain appropriate forms for children aged 5–7, 8–12 and 13–18 y and their parents.

The self-rated health of caregivers was assessed by using Short Form 36-version 2 (SF- 36v2) questionnaire. The caregivers of both cases and controls were administered SF-36v2 questionnaire after obtaining necessary permissions from Quality Metric Incorporation. The General Health domain of this questionnaire was used to assess their quality of life. The primary outcome for this was self-rated poor health. Self-rated health is reported on a five point scale: excellent/very good/good/fair/ poor. The authors classified anyone reporting fair/poor health as poor health and all others were considered good health [11].

The authors calculated the mean and standard deviation for continuous variables and proportions for the categorical variables. The means were compared using the unpaired t-test between the two groups and the proportions were compared using the chi square test. The authors also used logistic regression models for multivariate analysis. The outcome for this analysis was self-rated poor health (as described above). The primary explanatory variable was caregiver of thalassemic child/non-thalassemic child. The other variables included in the multivariate model were: age, sex of the child, socio-economic status, and total parent QOL score. The authors built unadjusted and adjusted models for these analyses.

The study was approved by the Institutional Ethics Committee of MGM Medical College.

Results

The mean (SD) age of cases was 7.4 (4.7) y and of controls was 7.1 (3.3) y. Out of 75 cases interviewed, 43 (57.3) were males and 32 (42.6) were females. Out of 80 controls, 41 (51.2) were males and 39 (48.7) were females. The demographic characteristics of the cases and controls are listed in Table 1.

The total QOL score according to child-self report [83.7 (10.8) *vs*. 97.6 (3.3); $p < 0.001$] and parent-proxy report [84.2 (11.9) *vs*. 96.7 (3.5); $p < 0.001$] was significantly lower in cases as compared with controls. The scores in each of the domains of physical, emotional, school and psychosocial functioning of cases was significantly lower as compared with controls in both child-self report as well as parent-proxy report. There was no significant difference in scores between

Table 1 Demographic and clinical characteristics of 155 children in Mumbai, India

Characteristic	Cases $n = 75$ N (%)	Controls $n = 80$ N (%)	*P* value
Age			
2–4 y	27 (36)	18 (22.5)	
5–7 y	18 (24)	31 (38.75)	
8–12 y	15 (20)	25 (31.25)	
13–18 y	15 (20)	6 (7.50)	0.009
Sex			
Male	43 (57.3)	41 (51.2)	
Female	32 (42.6)	39 (48.7)	
Transfusion transmitted infections			
HIV status	0	0	
Hepatitis B	0	0	
Hepatitis C	2 (2.67)	0	0.264
Attending school			
Yes	51 (68)	75 (93.75)	
No	24 (32)	5 (6.25)	
Socio-economic status			
Upper (I)	7 (9.33)	16 (20.25)	
Upper-Middle (II)	15 (20)	24 (30.38)	
Lower-Middle (III)	32 (42.67)	30 (37.97)	
Upper-Lower (IV)	21 (28)	9 (11.39)	0.016
Age at diagnosis in months; median (range)	8 (2 to 48)		
Age at first transfusion in months; median (range)	6 (2 to 48)		

cases and controls in the domain of social functioning in child report; however, the mean scores were significantly lower in the cases compared with controls in the parent-proxy report. The scores of child-self report and parent proxy report are shown in Table 2 and Table 3 respectively.

The total QOL score was significantly lower among cases in all age groups as compared with their controls as shown in Table 2. The mean score on total quality of life of cases were lower than controls in both males and females. The mean score on total quality of life was lower among cases of all socio-economic classes as compared with controls. The lowest score in the child self report was obtained in the domain of school functioning. However, in the age group of 13–18 y, no significant difference was observed in the mean scores obtained by the cases 80.8(16.4) and controls 82.5(11.7) in the domain of emotional functioning.

The total QOL score on parent- proxy report was significantly lower among cases as compared with controls in all age groups. The mean scores on total quality of life of cases were lower than controls in males and females. The mean score on total quality of life were lower among cases of all socio-economic classes as compared with controls.

While 29 caregivers of thalassemic patients reported poor health, only two caregivers of controls reported poor health. Thus, it was found that a significantly higher proportion of caregivers of cases reported poor health compared with caregivers of controls (29.2% [22/75] *vs*. 2.5% [2/80], $p < 0.001$). Even after adjusting for age, sex, socio-economic status, and total QOL score by the parent, it was found that caregivers of thalassemic children were significantly more likely to report poor health compared with those of controls (odds ratio: 15.8, 95% confidence intervals: 2.8–89.9) (Table 4).

Discussion

The HRQOL of 75 children with β thalassemia major and their caregivers was assessed using the PedsQL 4.0 questionnaire and SF 36 questionnaire respectively and compared with that of children without thalassemia or any other chronic medical or surgical disease and their caregivers in this study [12, 13]. The authors wish to highlight that this is one of the few Indian studies [14, 15] that compares the HRQOL in Indian children with beta-thalassemia major and normal children using PedsQL 4.0 questionnaire. However, worldwide, there are a few studies which have used this questionnaire [4–6, 16–21]. Very few studies have compared the QOL of children with thalassemia with either normal children or with their normal siblings as controls [4, 16, 22, 23].

HRQOL of children with beta-thalassemia major was significantly lower as compared with controls both in child-self and parent-proxy report. This is congruent with other studies reviewed by authors, which have compared QOL of thalassemic children with healthy children or siblings [4, 16]. HRQOL in the domains of physical, emotional, school and psychosocial functioning was significantly lower in child-self report as compared with controls, which is in concurrence with other studies [6]. A study done by Ismail et al., 2006 in Malaysian children with thalassemia using PedsQL 4.0 reported low scores in three domains namely physical, social and school functioning which were statistically significant as compared with controls. However, they did not report a significant difference in the domain of emotional functioning between thalassemic children and healthy controls [4].

In the index study, in the child-self report, the lowest scores were obtained in the domain of school functioning. This is in concurrence with studies who have all reported lowest scores in the school functioning domain in the child self report [4, 6, 16, 17, 23]. This could be attributed to the fact that children with beta-thalassemia major have to visit the day care centre for blood transfusions once every 3–4 wk. School absenteeism and coping up with the class work missed may be an additional problem. Thus, it may be important to introduce flexible times (such as on Saturdays, Sundays, and holidays). Low hemoglobin levels in children with beta-thalassemia major is associated

Table 2 Values from PEDSQL 4.0 Child-Self Report in 110 children (aged 5–18 y), Mumbai, India

Variables	Physical Functioning Mean (SD)		Emotional Functioning Mean (SD)		Social Functioning Mean (SD)		School Functioning Mean (SD)		Subtotal Functioning Mean (SD)		Total Functioning Mean (SD)	
	Case	Control	Case	Control	Case	Control	Case	Control	Case	Control	Case	Control
All	88.2** (19.5)	99.2 (2.4)	79.3** (17.5)	93.3 (9.8)	98.1 (7.5)	99.2 (2.7)	75.3** (13.5)	98.6 (3.6)	84.3** (9.6)	97.0 (4.1)	83.7** (10.8)	97.6 (3.3)
Age												
5–7 y	75.8** (24.0)	99.0 (2.8)	81.3** (20.8)	94.8 (7.0)	95.0 (12.6)	98.7 (3.4)	73.4** (14.4)	98.2 (4.2)	83.2** (12.0)	97.2 (3.7)	81.4** (13.4)	97.7 (3.2)
8–12 y	83.6** (15.9)	99.2 (2.1)	79.6** (10.7)	93.3 (11.6)	98.8 (3.0)	99.6 (2.1)	80.0** (8.8)	98.7 (3.4)	86.3** (5.0)	97.2 (4.5)	85.5** (6.3)	97.7 (3.7)
13–18 y	88.9* (13.0)	100.0 (0.0)	80.8 (16.4)	82.5 (11.7)	100.0[a] (0.0)	100.0[a] (0.0)	77.3** (8.3)	99.2 (2.0)	86.0* (6.1)	93.9 (4.3)	86.7* (6.1)	95.4 (3.2)
Sex												
Male	82.1** (18.9)	99.0 (2.8)	80.7** (15.7)	93.5 (10.0)	99.5 (2.0)	99.3 (2.5)	74.6** (14.7)	98.4 (3.7)	84.9** (8.6)	97.0 (4.1)	84.2** (9.9)	97.8 (3.4)
Female	82.4** (21.5)	99.4 (1.8)	77.9** (20.5)	93 (9.6)	95.8 (11.7)	99.0 (2.9)	76.4** (12.0)	98.8 (3.5)	83.5** (11.3)	96.9 (4.0)	83.1** (12.4)	97.6 (3.3)
SES												
Upper (I)	76.8* (29.7)	98.4 (3.8)	74.0* (16.7)	89.6 (9.4)	100.0 (0.0)	99.16 (2.8)	77.0** (6.7)	99.1 (2.8)	83.6** (6.3)	96.0 (3.1)	82.0** (10.4)	96.6 (2.7)
Upper-Middle (II)	76.7** (20.4)	99.6 (1.4)	77.7* (18.8)	90.5 (13.4)	100.0 (0.0)	98.9 (3.15)	73.5** (17.9)	99.2 (2.5)	84.1** (11.1)	96.2 (5.3)	82.1** (12.8)	97.1 (4.2)
Lower-Middle (III)	82.9** (15.8)	99.2 (2.1)	80.3** (13.8)	95.4 (6.6)	97.9 (3.4)	99.2 (2.8)	76.5** (14.4)	97.7 (4.6)	84.9** (8.5)	97.4 (3.9)	84.4** (8.0)	97.8 (3.3)
Upper-Lower (IV)	87.3 (20.2)	99.3 (2.1)	82.1* (21.5)	98.3 (5.0)	96.0 (13.0)	100.0 (0.0)	74.6** (12.1)	98.8 (3.3)	84.2** (11.4)	99.1 (1.9)	85.0** (13.1)	99.1 (1.7)

SES Socio-economic status

*$p < 0.01$, **$p < 0.001$

[a] *P* value not calculated

Table 3 Values from PEDS QL 4.0 Parent proxy report in 155 children (aged 2-18 y), Mumbai, India

Variables	Physical Functioning Mean (SD)		Emotional Functioning Mean (SD)		Social Functioning Mean (SD)		School Functioning Mean (SD)		Subtotal Functioning Mean (SD)		Total Functioning Mean (SD)	
	Case	Control	Case	Control	Case	Control	Case	Control	Case	Control	Case	Control
All	84.1** (17.5)	98.7 (3.1)	74.6** (17.5)	90.6 (9.6)	95.0* (12.7)	99.3 (2.5)	82.2** (15.2)	98.4 (4.2)	84.1** (11.5)	96.0 (4.1)	84.2** (11.9)	96.7 (3.5)
Age												
2–4 y	88.0* (14.5)	98.6 (3.2)	74.6** (12.5)	89.7 (8.6)	91.6* (15.5)	99.4 (2.3)	87.5* (17.6)	99.5 (1.7)	83.4** (12.6)	95.6 (3.9)	84.8** (12.3)	96.4 (3.1)
5–7 y	82.0** (22.3)	98.5 (3.2)	76.6** (19.5)	92.2 (7.4)	94.4 (16.1)	98.8 (3.1)	74.6** (20.0)	97.9 (5.0)	82.1** (13.5)	96.3 (3.7)	82.0** (15)	96.8 (3.2)
8–12 y	80.0** (17.6)	98.75 (3.1)	79.6* (15.0)	91.4 (11.8)	99.0 (2.8)	99.6 (2.0)	85.4** (10.2)	98.2 (4.2)	88.0** (6.9)	96.4 (4.7)	86.0** (7.6)	97.0 (4.1)
13–18 y	84.0* (16.0)	100.0 (0.0)	66.8 (23.8)	80.0 (7.07)	97.8 (5.8)	100.0 (0.0)	86.6* (8.8)	98.7 (2.8)	83.7 (10.5)	92.9 (2.5)	83.8* (11.0)	94.7 (1.9)
Sex												
Male	87.0** (13.4)	98.2 (3.8)	79.5** (12.9)	90.0 (10.7)	96.6* (8.3)	99.4 (2.3)	82.7** (12.5)	98.3 (4.0)	86.6** (8.0)	95.7 (4.5)	86.8** (7.7)	96.4 (3.8)
Female	80.3** (21.4)	99.3 (1.9)	68.3** (20.6)	91.0 (8.3)	92.8* (16.9)	99.1 (2.8)	81.5** (18.8)	98.4 (4.3)	80.7** (14.4)	96.2 (3.7)	80.7** (15.2)	97.0 (3.0)
Socio-economic Status												
Upper (I)	87.1* (11.7)	97.7 (4.5)	77.8* (6.4)	87.3 (7.7)	100.0 (0.0)	98.6 (3.5)	89.6* (9.4)	99.1 (3.3)	89.1** (3.4)	94.8 (2.9)	88.7** (4.8)	95.5 (2.9)
Upper-Middle (II)	87.7** (8.8)	98.8 (2.7)	74.3** (15.1)	89.6 (11.5)	99.3 (2.6)	99.2 (2.8)	87.5** (6.6)	98.9 (3.1)	87.3** (7.0)	95.9 (4.6)	87.4** (6.7)	96.6 (3.8)
Lower-Middle (III)	80.9** (19.4)	98.9 (2.8)	72.7** (18.2)	91.3 (9.2)	92.9* (14.7)	99.5 (2.0)	77.3** (21.1)	97.4 (5.3)	81.4** (12.9)	95.9 (4.5)	81.4** (13.1)	96.7 (3.6)
Upper-Lower (IV)	85.3* (20.7)	99.3 (2.1)	76.6* (20.9)	96.1 (6.1)	93.3 (15.3)	100.0 (0.0)	82.1** (9.9)	98.6 (4.1)	84.1* (12.9)	98.2 (2.1)	84.4* (14.0)	98.5 (1.8)

*$p < 0.01$, **$p < 0.001$

Table 4 Unadjusted and adjusted logistic regression estimates for 'self-rated poor health' of caregivers of 155 children, Mumbai, India

Variables	Unadjusted estimates Odds ratio (95% confidence intervals)	Adjusted estimates Odds ratio (95% confidence intervals)
Type of caregiver		
Control	Reference	Reference
Case	16.1 (3.7–71.8)	15.8 (2.8–89.9)
Age (y)		
2–4	Reference	Reference
5–7	0.7 (0.2–2.8)	1.1 (0.2–4.8)
8–12	1.4 (0.4–5.0)	2.3 (0.5–9.7)
13–18	6.0 (1.7–21.4)	8.4 (1.8–40.2)
Sex		
Male	Reference	Reference
Female	1.2 (0.5–2.9)	1.8 (0.6–5.4)
Socio-economic status		
Upper	Reference	Reference
Upper-Middle	5.7 (0.7–42.7)	9.7 (0.8–120.5)
Lower-Middle	2.8 (0.3–24.1)	4.7 (0.4–61.1)
Upper-Lower	8.0 (0.9–69.5)	8.5 (0.7–110.8)
Total QOL Parent score	0.95 (0.91–0.99)	1.00 (0.95–1.05)

QOL Quality of Life

with a number of symptoms, such as fatigue, general weakness, and decreased mental alertness, which might lead to impaired HRQOL of the patients in the domain of school functioning.

An interesting finding was that social functioning was not significantly affected in the children with thalassemia according to child-self report as compared with controls in the index study. This finding varies from that of other studies. In the index study, scores in the social functioning domain in cases *vs*. controls were 98.1 (7.5) *vs*. 99.2 (2.7) with a *p* value of 0.258. In the study done by Ismail et al., 2006 scores in this domain were 73.29 (18.69) *vs*. 82.21 (15.29) with a *p* value of <0.05 [4]. Social functioning as assessed by PedsQL Generic Core Scale 4.0 assesses the social interaction with peers. The questions pertaining to this domain that were asked to the children were problems in getting along with other children, other kids not wanting to be his or her friend, getting teased by other children *etc*. Children with beta-thalassemia major, who are on regular blood transfusion therapy, as was the feature of present study group, do not "look" different in terms of physical appearance nor do they suffer from any physical handicap. They are therefore less likely to be subjected to social discrimination. This may explain the findings related to social functioning in index study.

In the parent proxy report, a significant difference in QOL was noted in all domains including social functioning between cases and controls.

Another interesting observation in both the child-self and parent-proxy report was pertaining to the age group 13–18 y. It was noted that QOL scores on emotional functioning domain were low in both reports in cases and it was not significantly different from controls who also reported low scores. Ismail et al. too have reported similar findings in the domain of emotional functioning, though not specifically in the 13–18 y age group. In the study done by Ismail et al., 2006 the score in the domain of emotional functioning of cases was 68.14 (16.75) and controls was 70.36 (16.86) with a *p* value of 0.349 [4]. One of the reasons could be that even the non-thalassemic teen-aged children may be having emotional problems like tiffs with parents, adolescent issues, peer pressure *etc*. and hence their emotional functioning was also affected. The emotional problems like sadness, mood swings, and temper tantrums in children with thalassemia may be overestimated by their parents which may be the reason for the low scores in the emotional functioning domain in the parent proxy report.

Most studies focus their attention on the patients suffering from chronic disease. Often the HRQOL of parents/caregivers caring for the patients is overlooked. Caregivers may suffer health issues themselves. Despite having health problems, they have to continue caring for the affected child. They may have emotional and psychosocial problems like stress and anxiety, grief, guilt, apprehension, fear of death of their child *etc*. Parents raising a child with beta thalassemia have many issues to address like choosing the best treatments for the child, financial burden, and how to deal with child's emotional responses to living with a chronic health problem. The present study corroborates the poor quality of life of caregivers as assessed by the SF 36v2 questionnaire. In the study conducted by Shaligram et al., 2007, 57% of the caregivers had psychiatric problems and QOL was affected in 50%. The greatest concerns of caregivers were regarding the future (91%), illness (80%) and finances (73%) [24]. In a study reported by Ali et al., 2012, psychological distress and coping strategies among parents of β-thalassemic patients showed that all the parents experienced severe parental stress. However, psychological distress was reported by 27 (67.5%) parents [25]. The QOL of thalassemic children may also be affected by therapy; thus, the QOL may be different in children who are well chelated compared with children who are not well chelated. Though, the authors did not address this in the manuscript, future studies should address this limitation.

To conclude, the quality of life of thalassemia patients is indeed much lower than the quality of life of healthy children regardless of age, gender, and household income. Thalassemia, being a chronic disease, negatively impacts various aspects of day to day functioning in the lives of affected children. In light of these findings, it is strongly recommended that counselling programs be incorporated in the routine care and treatment of thalassemic patients. There is a need of multi-disciplinary team in management of thalassemic patients; the team may include

the pediatricians, psychologists, and nutrition experts. In addition, health care services for children with thalassemia should be modified to become more patient-centered, flexible and comprehensive. These programs should help patients discuss and accept their illness, facilitate a normal lifestyle, and provide a link between patients and parents, school officials and the physician. Thus they will be helpful in improving the quality of life of thalassemia patients and their caregivers.

Contributions SS primary responsible for data collection; BS: Concept and data collection; PJ: Data collection and feedback on the concept & manuscript; MI: Data collection and feedback on the concept & manuscript; MSS: Concept, data analysis and feedback on manuscript. BS will act as guarantor for the paper.

Compliance with Ethical Standards

Conflict of Interest None.

Source of Funding ICMR grant (Short Term Studentship program); Reference ID −2014-00208.

References

1. Modell B, Darlison M. Global epidemiology of haemoglobin disorders and derived service indicators. Bull World Health Organ. 2008;86:480–7.
2. Parthasarathy A, Menon P. IAP textbook of pediatrics. New Delhi: Jaypee Bros. Medical Publishers; 2009.
3. Wahab I, Naznin M, Nora M, et al. Thalassaemia: a study on the perception of patients and family members. Med J Malays. 2011;66:326–34.
4. Ismail A, Campbell M, Ibrahim H, Jones G. Health related quality of life in Malaysian children with thalassaemia. Health Qual Life Outcomes. 2006;4:39.
5. Ismail M, Chun CY, Yusoff NAM, et al. Quality of life among thalassaemia children, adolescent and their caregivers. Sains Malaysiana. 2013;42:373–80.
6. Caocci G, Efficace F, Ciotti F, et al. Health related quality of life in middle eastern children with beta-thalassemia. BMC Hematol. 2012;12:6.
7. Olivieri N. The beta-thalassemias. N Engl J Med. 1999;341:99–109.
8. Varni J, Burwinkle T, Katz E, Meeske K, Dickinson P. The PedsQL™ in pediatric cancer. Cancer. 2002;94:2090–106.
9. Varni J, Burwinkle T, Seid M. The PedsQLTM 4.0 as a school population health measure: feasibility, reliability, and validity. Qual Life Res. 2006;15:203–15.
10. Varni J. PedsQL TM (Pediatric Quality of Life Inventory TM) [Internet]. Pedsql.org; 2013. Available at: http://www.pedsql.org/about_pedsql.html. Accessed 15 Dec 2013.
11. McHorney C, Ware J, Rachel Lu J, Sherbourne C. The MOS 36-item short-form health survey (SF-36): III. Tests of data quality, scaling assumptions, and reliability across diverse patient groups. Med Care. 1994;32:40–66.
12. Khairkar P, Malhotra S, Marwaha R. P01-378 - growing up with the families of B-thalassaemia major using an accelerated longitudinal design. Indian J Med Res. 2010;132:428–37.
13. Khurana A, Katyal S, Marwaha R. Psychosocial burden in thalassemia. Indian J Pediatr. 2006;73:877–80.
14. Gupta M, Jindal R. Quality of life in patients with thalassemia major. Int J Sci Res. 2016;5:41–2.
15. Soni S, Thawani R, Idhate T, Kalra M, Mahajan A. Health related quality of life in patients with transfusion- dependent thalassemia. Indian Pediatr. 2016;53:741–2.
16. Wahyuni MS, Ali M, Rosdiana N, Lubis B. Quality of life assessment of children with thalassemia. Paediatr Indones. 2011;51:163–9.
17. Thavorncharoensap M, Torcharus K, Nuchprayoon I, Riewpaiboon A, Indaratna K, Ubol BO. Factors affecting health-related quality of life in Thai children with thalassemia. BMC Blood Disord. 2010;10:1.
18. Salama H, Hussein H, Al Faisal W, et al. Health - related quality of life in transfusion - dependent thalassemia major patients and associated factors in Dubai, UAE, 2011. Middle East J Psychiatr Alzheimers. 2014;5:3–10.
19. Ayoub M, Radi S, Azab A, et al. Quality of life among children with beta-thalassemia major treated in western Saudi Arabia. Saudi Med J. 2013;34:1281–6.
20. Ismail A, Campbell MJ. Measuring the health related quality of life of Malaysian children with thalassemia: reliability and validity of PedsQL 4.0 generic score and SF36v2. J Statist Model Analyt. 2010;1:1–28.
21. Surapolchai P, Satayasai W, Sinlapamongkolkul P, Udomsubpayakul U. Biopsychosocial predictors of health-related quality of life in children with thalassemia in Thammasat University Hospital. J Med Assoc Thail. 2010;93:S65–75.
22. Baghianimoghadam M, Sharifirad G, Rahaei Z, Baghianimoghadam B, Heshmati H. Health related quality of life in children with thalassemia assessed on the basis of SF-20 questionnaire in Yazd, Iran: a case-control study. Cent Eur J Public Health. 2011;19:165–9.
23. Gharaibeh H, Gharaibeh MK. Factors influencing health-related quality of life of thalassaemic Jordanian children. Child Care Health Dev. 2011;38:211–8.
24. Shaligram D, Girimaji S, Chaturvedi S. Quality of life issues in caregivers of youngsters with thalassemia. Indian J Pediatr. 2007;74:275–8.
25. Ali S, Sabih F, Jehan S, Anwar M, Javed S. Psychological distress and coping strategies among parents of beta- thalassemia major patients. International conference on clean and green energy. Singapore: IACSIT Press; 2012. p. 124–8.