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Research Article

Health Related Quality of Life of Children with Thalassemia Major in Two Selected Hospitals in Sri Lanka

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Abstract
Introduction: Thalassemia is an inherited hemoglobin disorder. There are two types; thalassemia major and minor. Thalassemia major directly affects children's physical, emotional, social and school functions. Health Related Quality of Life (HRQoL) is an important indicator to assess the health of human. Methods: A cross sectional study was conducted using 60 children (age 2-12 years) with thalassemia major who attended clinics in two selected hospitals in Sri Lanka from July to September 2018. Demographic data and risk factors were collected using a self-administered questionnaire. HRQoL data were collected using the Pediatric Quality of Life Inventory Version 4.0 Generic Core Scale. Data were analyzed using SPSS. Results: Mean values of the total functioning scores of physical, emotional, social and school were 89.42 (±9.20), 87.83 (±10.43), 95.00 (±7.53) and 78.75 (±14.13) respectively. Age of the children was significantly associated with emotional functioning (p=0.046), school functioning (p=0.023), psychosocial health (p=0.021) and the total summary score (p=0.006). Total number of blood transfusions per year of the patient was significantly associated with physical functioning (p=0.002), psychosocial health (p=0.045) and total summary score (p=0.034). Conclusions: Age and the total number of blood transfusions per year significantly affect the HRQoL of children with thalassemia major. Gender, type of iron chelation and age at first blood transfusion did not affect HRQoL of the children.

Keywords: HRQoL, Thalassemia major, Haemoglobin disorder

Introduction

Thalassemia is a genetic hemoglobin disorder which mainly occurs as a result of high frequency of consanguineous marriages. Thalassemia can be broadly classified into α, β, δβ, and εδβ depending on the haemoglobin structure [1]. Globally, more than 330,000 infants are born annually with haemoglobin disorders. Out of them, 17% suffer from thalassemia [2]. The β-thalassemia is the highest existing type of thalassemia carrieracross the world (1.5% of the global population). Approximately, half of thesecarriers originate from South East Asia [3]. One third of the Hemoglobin E (HbE) thalassemia is presented with β-thalassemia [4]. In Sri Lanka, according to an island wide survey held in 2017, there were approximately 2000

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patients suffering from severe thalassemia [5]. These patients are distributed all over the island but the majority of them have been reported in the North Western Province and the North Central Province in Sri Lanka [6].

There are several types of treatments used for thalassemia disease including blood transfusion, iron chelation therapy and Hematopoietic Stem Cell Transplantation (HSCT). The allogenic HSCT is the only treatment mode for the definitive cure of thalassemia major [5]. However, this method is a highly intensive treatment mode and it is affected by many factors [7].

In Sri Lanka, most of the patients with thalassemia are mainly treated by blood transfusions and iron chelation therapy. As a result of these efficient treatment methods, thalassemia patients have shown a significant improvement of the survival rate in the country during the last decades. These patients are basically managed by government hospitals, which have safe blood transfusion facilities. The main thalassemia center of Sri Lanka is located in Kurunegala and it is supported and guided by a team of international experts under the supervision of the Ministry of Health [8].

The nature is different when the disease is presented in school children. It is a critical illness among school children and mainly affects school performance. According to the World Bank report 2015, Sri Lanka is a middle income country and due to many financial constraints, most patients cannot afford the costs of treatment [9]. Therefore, monthly income of the family has a direct impact on the quality of life (QoL) of these patients. Due to these facts, the assessment of the effects of this disease on their Health-Related Quality of Life (HRQoL) is essential.

The HRQoL measurement in paediatrics is used in healthcare settings to; facilitate the patient-physician communication, improve patient/parent satisfaction, identify hidden morbidities and assist in clinical decision making [10]. The Paediatric Quality of Life Inventory Version 4.0 (PedsQL 4.0) is a multidimensional model used to measure the paediatric quality of life. It is a useful tool to assess the HRQoL in paediatric patients with thalassemia. It captures physical functioning, emotional functioning, social functioning and school functioning of children [11].

There is no previously published research on HRQoL of children with thalassemia in Sri Lanka. Therefore, this study was designed to determine the effect of thalassemia and its treatment on the HRQoL of children having thalassemia major, in terms of physical functioning, psychosocial functioning, emotional functioning, and school functioning sub-scales.

Methods

Study design, subjects and settings

This is a cross sectional study. The research was carried out at the Lady Ridgeway Hospital for Children (Teaching) in Colombo, Sri Lanka and the North Colombo Teaching Hospital at Ragama, Sri Lanka. The children (age between 2-12 years) with thalassemia major who attended the clinics at the above two institutions from July to September 2018 were enrolled for the study.

The age of the subject was calculated from the date of birth to the date of interview and they were categorized in to two groups; age between 2-7 years and age between 8-12 years. The patients/their guardians who gave consent were included and those who had no competence in English and Sinhala language were excluded. Study subjects with severe clinical conditions were also excluded.

Sample size

Sample size was calculated by using the following equation [12].

$$n = \frac{z^2\sigma^2}{d^2}$$

n= Sample size of population is infinite
Z= Critical value according to the confidence interval
(confidence interval is 95%, critical value is 1.96)
σ= The mean (SD) of total summary score was 76.67 (11.4)
d= Absolute error or precision. It was taken as 3

Study instruments

The data on HRQoL of the children (n=60) were collected by using PedsQL 4.0 (in Sinhala version) [13], which consists of parent proxy reports (age ranges 2-4, 5-7, 8-12 and 13-18 years) parallel to child self-reports (age ranges 5-7, 8-12 and 13-18 years). The study instrument was pretested using 10 randomly selected children who belonged to this age category.

The questionnaire consisted of 23 questions which evaluated the QoL in four domains including; physical functioning, emotional functioning, social functioning and school functioning. The factors considered under each domain is given in Table 1.

All responses given by the parents/guardians of the child with thalassemia were rated as 0, 1, 2, 3 and 4 for never, almost never, sometimes, often and almost respectively. It was scored as 0–100 (0=100, 1=75, 2=50, 3=25, 4=0). The higher scores denoted by higher QoL of patients.

Data collection

The demographic data and risk factors were collected using a self-administered questionnaire. This self-administered questionnaire was in both Sinhala and English languages and filled by the parent/guardian of the study subject. The clinical data of the patients were perused from the clinical files after obtaining permission from the relevant authorities.

Data analysis

Data were analyzed using MS Excel and Statistical Package for Social Sciences (SPSS) version 20. Total summary scores were computed by adding the scores for each function in the four domains and dividing it by the total number of functions in four domains.

Psychosocial health score was computed as summation of the all items over the number of items answered on emotional functioning, social functioning and school functioning domains. Total summary score was computed as a summation of all the items over the number of items answered on all the scales.

An independent sample t-test was used to find the associations between the demographic factors and the HRQoL. ANOVA was used to find the associations between the clinical factors and the HRQoL. The p value <0.05 was considered as significant.

Ethical approval

The study commenced after obtaining the ethical approval from the Ethical Review Committee of the Faculty of Allied Health Sciences, University of Ruhuna, Galle, Sri Lanka (Ref No:14.02.2018:037) and the Ethical Review Committee of the Lady Ridgeway Hospital for Children (Teaching), Colombo. Permission was also obtained from the Director of North Colombo Teaching Hospital, Ragama, Sri Lanka prior to commencing the study. Before enrolling the study subjects, written informed consent was obtained from the parent/guardian of each child with thalassemia.

Table 1: Factors associated with HRQoL of patients with thalassemia

Domain	Factors
Physical functioning	1: Walking more than one block 2: Running 3: Participating in sports activity or exercise 4: Lifting something heavy 5: Taking a bath or shower by himself or herself 6: Doing chores around the house 7: Having hurts or aches 8: Low energy level
Emotional functioning	1: Feeling afraid or scared 2: Feeling sad or blue 3: Feeling angry 4: Having trouble in sleeping 5: Worrying about what will happen to him or her
Social functioning	1: Getting along with other children 2: Other kids not wanting to be his or her friend 3: Getting teased by other children 4: Not able to do things that other children of his or her age can do 5: Keeping up when playing with other children
School functioning	1: Paying attention in class 2: Forgetting things 3: Keeping up with schoolwork 4: Missing school because of not feeling well 5: Missing school to go to the doctor or hospital

Results

Demographic, risk and clinical factors of the 60 thalassemia children incorporated in to the study are presented in Table 2. The mean age of the population was 8.48±3.08 years. The majority of the patients were aged between 8-12 years and were males (55%). Only 30.0% of them had a family history of thalassemia and the majority of the parents (86.7%) had no consanguineous marriages. The commonest blood group of the study population was O positive (45%) and there were no patients with AB negative blood.

Out of the total study population, the majority of them (88%) did not have past surgical histories. Only 32% had a past history of allergy. Six treatment modalities were identified among the study population. Out of them, the majority of the patients received a combination of desferasirox and folic acid. It was found that 50.0% of patients had received desferasirox as an oral iron chelator. Only 13.3% of patients did not receive any iron chelator as their treatment.

Table 2: Descriptive data of the demographic factors, risk factors and clinical data of the study subjects (n=60)

	n (%)		n (%)
Demographic factors			
Age		*Gender*	
2-7 years	25 (41.7)	Male	33 (55.0)
8-12 years	35 (58.3)	Female	27 (45.0)
Risk factors			
Patient with family history		*Consanguineous marriage*	
Presence	18 (30.0)	Yes	8 (13.3)
Absence	42 (70.0)	No	52 (86.7)
Clinical factors			
Blood group		*Treatment patterns*	
A +	8 (13.3)	None	4 (6.7)
A -	1 (1.7)	Desferasirox	4 (6.7)
B +	13 (21.7)	Desferasirox + folic acid	27 (45.0)
B -	1 (1.7)	Desferasirox + folic acid +	20 (33.3)
AB +	6 (10.0)	desferoxamine	
AB -	0 (0)	Folic acid	3 (5.0)
O +	27 (45.0)	Desferasirox + folic acid +	2 (3.3)
		desferoxamine + other drugs	
Past surgical history			
Yes	7 (11.7)	*Total number of blood transfusions per year*	
No	53 (88.3)	7	7 (11.7)
		9	4 (6.7)
History of allergy		13	32 (53.3)
Yes	19 (31.7)	17	17 (28.3)
No	41 (68.3)		

n- Number, %- Percentage

All the patients were depended on blood transfusion. The majority of the patients had undergone 13 blood transfusions per year. A few (n=4) had undergone blood transfusions nine times per year.

The mean values of the total physical, emotional, social and school functioning scores of this study population were 89.43±9.21, 87.83±10.43, 95.00±7.53 and 78.75±14.13 respectively.

The association between HRQoL scores of children and the demographic features were computed (Table 3). The age of the children with thalassemia was significantly associated with the emotional functioning (p=0.046), school functioning (p=0.023), psychosocial health (p=0.021) and total summary scores (p=0.006). It was not significantly associated with physical (p=0.119) and social functioning (p=0.731) scores. Gender was not associated with a single factor, which is associated with the HRQoL of the study subjects (p>0.05).

The association between the QoL scores of children and the clinical factors are explained in Table 4. The iron chelation therapy was not significantly associated with the factors which determine the HRQoL of thalassemia patients. The total number of blood transfusions per year was significantly associated with the physical functioning (p=0.002), psychosocial health score (p=0.045) and total summary (p=0.034) score. It had no significant association with the QoL scores and age at first blood transfusion.

The total QoL scores of the 60 patients with thalassemia are listed in Table 5. It was found that mean of the total health summary score was 87.98±6.29. The mean of the total psychosocial health summary score was 87.53±7.08. The highest mean was obtained for the total social functioning score (95.00±7.53) whereas, the lowest was obtained for the total school functioning score (78.75±14.13).

Table 3: Association between the HRQoL scores and the demographic features

		Values for the quality of life scores (Mean±SD)					
Age groups	n	1	2	3	4	5	6
2-7	25	91.63±7.92	91.0±10.3	95.4±8.15	83.6±12.54	90.0±7.02	90.56±5.18
8-12	35	87.86±9.85	85.57±10.06	94.71±7.17	75.28±14.34	85.76±6.68	86.14±6.44
p value		0.119	0.046	0.731	0.023	0.021	0.006
Gender							
Male	33	89.39±9.50	86.21±10.68	95.60±7.26	77.27±15.31	86.97±7.80	87.41±6.71
Female	27	89.47±9.01	89.81±9.95	94.25±7.93	80.56±12.58	88.21±6.18	88.68±5.80
p value		0.976	0.185	0.496	0.375	0.505	0.443

SD-Standard Deviation, n-number, p-significance

1-Physical functioning, 2-Emotional functioning, 3-Social functioning,

4-School functioning, 5-Psychosocial health score, 6-Total summary score

Table 4: Association between the QoL scores of thalassemic children and the clinical factors

		Values of the quality of life scores (Mean±SD)					
	n	**1**	**2**	**3**	**4**	**5**	**6**
Type of iron chelation							
None	8	92.97±8.31	89.36±9.03	98.12±3.72	83.75±25.73	90.42±10.86	91.30±7.16
Desferasirox	30	86.56±9.18	88.17±11.7	94.00±8.74	79.0±11.62	87.72±6.78	86.91±6.37
Desferoxamine	22	92.05±8.67	86.81±9.32	95.22±6.63	76.59±11.79	86.21±5.75	88.24±5.68
p value		0.051	0.818	0.389	0.474	0.354	0.213
Total number of blood transfusions per year							
7	7	99.11±1.52	87.14±11.85	95.71±7.31	80.0±21.60	87.62±10.09	91.61±6.72
9	4	85.16±12.07	80±7.07	92.5±11.90	62.5±15.54	78.33±4.08	80.70±5.20
13	32	90.33±9.02	87.66±10.92	95.15±7.98	80.31±12.94	87.70±6.06	88.62±5.52
17	17	84.74±7.48	90.29±9.27	95.0±6.12	79.12±10.93	89.31±6.94	87.01±6.64
p value		0.002	0.363	0.920	0.122	0.045	0.034
Age at first blood transfusion							
< 1 year	18	91.49±8.76	86.94±12.85	96.39±7.24	84.44±10.56	89.25±5.21	90.03±5.01
1-2 years	27	89.12±9.15	86.67±9.61	95.18±8.02	76.85±14.55	86.23±7.64	87.27±6.67
2-3 years	8	87.5±10.69	85.62±7.76	91.86±7.03	70.00±16.03	84.99±9.71	84.23±6.99
3-4 year	7	87.5±9.89	97.14±3.93	94.28±7.31	81.43±14.35	90.95±3.83	89.75±5.60
p value		0.672	0.09	0.828	0.077	0.202	0.129

SD-Standard Deviation, n-number, p-significance, 1-Physical functioning, 2-Emotional functioning, 3-Social functioning, 4-School functioning, 5-Psychosocial health score, 6-Total summary score

Table 5: Total QoL scores of the thalassemic children

Type of function	Score (Mean±SD)	Minimum Score	Maximum Score
Total physical functioning	89.42±09.20	71.9	100.0
Total emotional functioning	87.83±10.43	55.0	100.0
Total social functioning	95.00±07.53	70.0	100.0
Total school functioning	78.75±14.13	40.0	100.0
Total psychosocial health	87.52±07.08	73.3	100.0
Total summary score	87.98±06.29	76.1	100.0

SD- Standard Deviation

Discussion

Thalassemia is a life limiting and life threatening disease condition. Hence, awareness of the factors affecting the QoL of thalassemia patients is important to control and for subsequent management of the disease. In our study cohort, all the subjects presented with thalassemia major and were also blood transfusion dependents. The majority of the patients were at the age between 8-12 years and were males.

Most of the parents of the present study cohort were not of consanguineous marriages and the majority of them had no family history of thalassemia up to first and second degree relatives, though previous study has found that more than half of the parents of the studied children were relatives [14]. Therefore, in the current study cohort, the majority of the parents of the children with thalassemia can be considered as carriers.

As there are no previous studies done in Asian countries, the HRQoL of thalassemia major patients in the current study were assessed using the PedsQL Generic Score Scale were under four dimensions; physical, emotional, social and school functioning [12]. This is the first study which was conducted to assess the HRQoL of thalassemia patients in Sri Lanka using the aforesaid study tool.

Gender of the present study population had no significant association with the scores of any of the tested functioning domains or the total summary score of the current study. This finding is on par with the previous studies [12,15] and proves that the gender does not significantly affect the HRQoL of thalassemia children. In addition, age of the thalassemia patients in the current study was significantly associated with the emotional functioning (p=0.046), school functioning (p=0.023), psychosocial function (p=0.021) and total summary score (p=0.006). A study done in Thailand has found that age of thalassemia patients was significantly associated with the school and social functioning scores and had no significant association with the physical and emotional functioning score of the study

subjects [12]. Further, it confirmed that age was a significant predictor of HRQoL of children with thalassemia. This finding is substantiated by the findings of the present study.

The majority of the present study subjects had O positive blood and they had no experience with past surgeries and allergies. Out of the total study population, 45% of them had received desferasirox and folic acid as their medications. Most of the patients received desferasirox as the iron chelation therapy. The majority of the patients had 13 blood transfusions per year and most of the patients had received the first blood transfusion at one year of age. These clinical features are mainly affected the HRQoL of patients with thalassemia [16].

The age in which the first blood transfusion was made and the type of iron chelation therapy had no significant association with any of the scores determined in the present study. It revealed that these factors do not have any impact on the HRQoL of children with thalassemia similar to previous studies [11,17]. Further, all these patients were basically managed by the government sector hospitals where the blood transfusion facilities are available [8]. Therefore, this is a mark of the improvement of quality of care given to the thalassemia patients in Sri Lanka during the last few decades.

In the current study, the highest mean was obtained for the total social functioning score, however the lowest was obtained for the total school functioning score. In addition, psychosocial health score was lower than the total physical functioning score which is similar to the finding of a study conducted in Malaysia [18].

Limitations

The study was conducted only in two institutions. It might not represent the total population of the patients with thalassemia in our country. However, these two institutions are national referral centers for thalassemia patients in Sri Lanka. The current study is not a case control study and the HRQoL scores for healthy children at age between 2-12 years in Sri Lanka are not available. Therefore, the above study findings cannot be compared with the healthy children at same age categories and find out the most effective factor for the HRQoL of thalassemia patients in Sri Lanka. In the current study, HRQoL questionnaire was filled by the parent of the patient due to inability of reading, writing and understanding of questions given. Therefore, the rate given for each function does not reflect the children's own perception. This might affect the final results.

Conclusions

Total number of blood transfusions per year and the age of patient significantly affect the HRQoL of the children with thalassemia major. Therefore, these patients need understanding and support from the health-care providers, counsellors, and school teachers to overcome the associated problems and enhance their QoL. The gender, type of iron chelation therapy, and the age at first blood transfusion did not significantly affect the HRQoL of present thalassemia patients. Further, a significant number of parents in this cohort could be considered as carriers of the thalassemia disease.

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