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Original Research Article

Assessment of quality of life in transfusion dependent thalassemic children - need to address parents/care givers

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ABSTRACT

Background: Thalassemia is an autosomal recessive congenital disease caused by the reduced or absent beta globin chain synthesis of hemoglobin tetramer. The degree of imbalance between alpha and non alpha globin chains determines the severity of clinical manifestations. The disabling nature of the disease and chronic therapy affects the normal life causing psychosocial burden. Overall patient's life, such as education, free-time, physical activities, skills, capabilities, and family adjustment is affected. The effects of which often result in psychological, emotional and social compromise. Health-Related Quality of Life (HRQoL) measurement is a multidimensional concept that focuses on the impact of the disease and its treatment on the well being of an individual.
Methods: A descriptive observational hospital based study was conducted over a period of 3 months. Transfusion dependency in thalassemic children aged between 5 years and 18 years was the inclusion criteria. Thalassemic children having debilitating illnesses unrelated to thalassemia were excluded. Quality of life was assessed using Pediatric Quality of Life Inventory (PedsQL™ 4.0)[4]. The tool assesses the quality of life in five domains: physical functioning (PF: 8 items), psychosocial functioning (sum of emotional, social and school functioning), emotional functioning (EF: 5 items), social functioning (SF: 5 items) and school functioning (SC: 5 items).
Results: Total of 125 children were enrolled with a mean age of 9.4±4.6 years (age range 5-18 yrs). According to the PedsQL questionnaire, the quality of life was similarly assessed by both parents and children. The total mean QoL score of the parents was 72.36±11.47 and of the children was 77.63±14.17. Emotional, school and psycho-social function were significantly affected according to both child and parents without statistical significance.
Conclusions: Thalassaemia patients and their parents require lifelong psychological support for prevention of mental health issues. By increasing the awareness and knowledge levels of the parents, we can help sick children in developing countries to get the best care locally and to thus improve HRQoL.

Keywords: HRQoL, Mental health issues, Transfusion dependent thalassemia

INTRODUCTION

Thalassemia is an autosomal recessive congenital disease caused by the reduced or absent beta globin chain synthesis of hemoglobin tetramer. The degree of imbalance between alpha and non alpha globin chains determines the severity of clinical manifestations.[1] Thalassemia has its origin from mediterranean countries and is now present worldwide with high prevalence in mediterranean, middle east and central Asian countries.[2] According to various reports, 7500-12,000 β-thalassaemia major babies would be born in India each year.[3-5] Children with transfusion dependent beta thalassemia present with progressive pallor, failure to

thrive and splenomegaly. Regular blood transfusions and iron chelation to counter iron overload state secondary to blood transfusions form the mainstay of treatment. Bone marrow transplant remains the curative treatment. Hospital appointments or admissions for regular monthly blood transfusion and/or treatment of complications make children to skip school leading to school absenteeism compromising their self-identity and making them increasingly dependent on others.[6] The disabling nature of the disease and chronic therapy affects the normal life causing psychosocial burden. Overall patient's life, such as education, free-time, physical activities, skills, capabilities, and family adjustment is affected. The effects of which often result in psychological, emotional and social compromise.[2] Health-Related Quality of Life (HRQoL) measurement is a multidimensional concept that focuses on the impact of the disease and its treatment on the well-being of an individual. The measures are seen as ways of capturing patients' perspectives of their disease and treatment, their perceived need for health care and their preferences for treatment and disease outcomes.[7] Various studies have shown that all patients with thalassemia should undergo QOL assessment so that interventions focused on the affected domain can be implemented.[8,9] Given the paucity of research in this area and the need to rely on additional evidence-based data to further improve patient care, we did a pilot study to evaluate the quality of life in children with transfusion dependent thalassemia attending our centre. Considering that previous studies of childhood illness have shown that parents ratings of their child's HRQoL tend to be lower and possibly reflect parental distress.[10,11] We also evaluated the differences between HRQoL reports of the children and their parent-proxies.

METHODS

A cross sectional observational hospital based pilot study Study was conducted for the period of 3 months from June 2019 to September 2019 were included in the study.

Inclusion criteria

Transfusion dependency in consented thalassemic children aged between 5 years and 18 years.

Exclusion criteria

Thalassemic children having debilitating illnesses unrelated to thalassemia.

Quality of life was assessed using Pediatric Quality of Life Inventory (PedsQL™ 4.0).[4]

The tool assesses the quality of life in five domains: physical (PF: 8 items), psychosocial (sum of emotional, social and school functioning), emotional (EF: 5 items), social (SF: 5 items) and school (SC: 5 items).

Each item is rated on a 5- point Likert scale from 0 (Never) to 4 (Almost always) and items are reversed scored and linearly transformed to a 0-100 scale as follows: 0=100, 1=75, 2=50, 3=25, 4=0.

Data were computed and analyzed by and SPSS (Statistical Package for the Social Sciences) program version 19.0. General characteristics of the patients were presented in terms of percentage, mean, and standard deviation and median for data not normally distributed. For QoL, both total HRQoL score and physical, emotional, social, school achievement and psychological scores were presented in terms of mean and standard deviation.

RESULTS

One hundred twenty five transfusion dependent thalassemia children were included between the age group of 5-18 years. Male babies outnumbered female babies (male:female :: 73:52). Children were divided into three age groups 5-7 yrs, 8-12 yrs, 13-18 yrs. 34 babies contributed to 5-7 yrs age group, we had 51 babies in 8-12 yrs, 40 babies in 13-18 yrs.

Table 1: Demographic characteristics of the participants (n=125).

Variable		Value	Percentage
Gender	Male	73	58.4
	Female	52	41.6
Age group (years)	Young children 5-7	34	27.2
	8-12	51	40.8
	13-18	40	32.0
Residence	Urban	75	60.0
	Rural	50	40.0

On recording the quality of life score by children and parents, following results were obtained. Mean physical score according to child was 81.57±16.33, parent recorded 77.63±16.44 (p=0.45). For the social score was 73.68±13.1 in children, parents recorded 68.42±14.04 (p=0.23). Emotional score as per children was 70.63±16.44, parents reported score was 66.68±15.1 (p=0.40). Score in school domain in children was 67.1±16.77, parents reported score was 60.52±15.17 (p=0.19). Regarding psycho social domain, Child reported score was 65.78±12.38, parent reported score was 63.15±12.82 (p=0.52). Total score was 77.63±14.17 in children and parents recorded score was 72.36±11.47. Physical and social domains were significantly higher reported by both child and parents as well as total HRQoL score, though statistically insignificant. Emotional, school and psycho-social function were significantly affected according to both child and parents without statistical significance.

Table 2: Children and parents total PedsQL score and single scale scores comparison.

Questionnaire PedsQL	Questionnaire version	Mean	SD	Wilcoxon test	P value
Total /Sum	Child	77.6316	14.17827	Z= -1.25	0.21
	Parent	72.3684	11.47079		
Physical	Child	73.6842	13.10663	Z=-1.19	0.23
	Parent	68.4211	14.04879		
Emotional	Child	77.6316	16.44591	Z=-0.83	0.40
	Parent	73.6842	13.10663		
Social	Child	81.5789	16.33441	Z= -0.75	0.45
	Parent	77.6316	16.44591		
School	Child	67.1053	16.77596	Z= -1.29	0.19
	Parent	60.5263	15.17442		
Psychosocial function	Child	65.7895	12.38987	Z= -0.65	0.52
	Parent	63.1579	12.82473		

DISCUSSION

In the present study, assesing the QoL in transfusion dependent thalassemia children showed the impact of transfusion dependent thalassemia on the patients performance in different aspects of life.

Psychosocial performance was the significantly affected domain. Followed by school and emotional domains. Similar reports were found in study by Sachith Mettananda, Adriana Ismail.[12,13]

On comparing parents and child self reporting, it was observed that parents score was low compared to child's score.

It would appear that parents tend to consider HRQoL of their children more compromised in domains dealing with interpersonal relationships (Social) rather than those concerning physical impairment. i.e., Physical functioning is better than social function. Similar results were seen in study by Surapolchai P, Caocci et al.[14,2]

A possible explanation may be that parents of thalassemic children unconsciously project their pessimistic feelings onto their children's functioning.

Low school domain score signifies the role of healthcare providers, counsellors and school teachers in helping children to overcome this problem.

CONCLUSION

Our pilot study showed the difference in self reporting of child and parent proxy reporting. It highlighted the significant effect of transfusion dependent thalassemia on psychosocial, school and emotional domains. Parent ratings of their child's HRQoL were lower for emotional functioning, psychosocial functioning and school functioning, suggesting the need to enhance the understanding and support of the parents. By increasing the awareness and knowledge levels of the parents, we can help sick children in developing countries to get the best care locally and to thus improve HRQoL.

Limitations of our study were small sample size, we obtained HRQoL meaurements in a cross-sectional manner we did not perform a complete cognitive or psychological evaluation based on normative scale scores, which would have been helpful in order to avoid confounding factors (i.e. cognitive, psychological or psychiatric problems in parents or children).

We conclude that Thalassaemia patients and their parents require lifelong psychological support for prevention of mental health issues. Several effective psychological strategies are available. Cognitive-Behavioural Family Therapy (CBFT) can be an effective psychological approach to children with beta-thalassaemia major, capable of increasing compliance to treatment, lessening the emotional burden of disease and improving the quality of life of caregivers as reported by Mazzone L et al.

Overall, HRQoL measurement should be part of therapy in children with chronic diseases and appropriate intervention can improve the quality of life of children and care givers.

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REFERENCES

1. Olivieri N, Weatherall DJ. Clinical aspects of β-thalassemia. In: Steinberg MH, Forget BG, Higgs DR, Nagel RL, editors. Disorders of hemoglobin, genetics, pathophysiology, and clinical management. Cambridge, England: Cambridge University. 2001;277–341.

2. Caocci G, Efficace F, Ciotti F, Roncarolo MG, Vacca A, Piras E, et al. Health related quality of life in Middle Eastern children with beta-thalassemia. BMC blood disorders. 2012;12(1):6.
3. Madan N, Sharma S, Sood SK, Colah R, Bhatia HM. Frequency of β-thalassemia and other hemoglobinopathies in northern and western India. Indian J Hum Genet. 2010;16:16-25.
4. Modell B, Petrou M. The problem of hemoglobinopathies in India. Indian J Haematol. 1983;1:1-5.
5. Choudhry VP, Upadhyay A. Thalassemia screening and control programme. In: Ghosh K, Colah R, editors. Control and management of thalassaemia and other hemoglobinopathies in the Indian subcontinent - Synoptic views. Mumbai: National Institute of Immunohaematol. 2008:36-44.
6. Atkin K, Ahmad W: Living a "normal life": young people coping with thalassemia major or sickel cell disorder. Soc Sci Med 2001, 53:615–626.
7. Bowling A. Current state of the art in quality of life measurement. In: Carr AJ, Higginson IJ, Robinson P, editors. Quality of life. London: BMJ Books; 2003.
8. Tefler P, Constantinidou G, Andreou P, Christou S, Modell B, Angastiniotis M. Quality of life in Thalassaemia. Annals N Y Aca Sci. 2005;1054:273–82.
9. Pakbaz Z, Treadwell M, Yamashita R, Quirolo K, Foote D, Quill L, et al. Quality of life in patients with Thalassaemia intermedia compared to Thalassaemia major. N Y Aca Sci. 2005;1054:457–61.
10. Speechley KN, Barrera M, Shaw AK, Morrison HI, Maunsell E: Health-related quality of life among child and adolescent survivors of childhood cancer. J Clin Oncol. 2006;24:2536–43.
11. Cremeens J, Eiser C, Blades M: Factors influencing agreement between child self-report and parent proxy-reports on the Pediatric Quality of Life Inventory 4.0 (PedsQL) generic core scales. Heal Qual Life Out. 2006;4:58.
12. Mettananda S, Pathiraja H, Peiris R, Bandara D, de Silva U, Mettananda C, et al. Health related quality of life among children with transfusion dependent β-thalassaemia major and haemoglobin E β-thalassaemia in Sri Lanka: a case control study. Health Qual Life Outcomes. 2019;17(1):137.
13. Ismail A, Campbell MJ, Ibrahim HM, Jones GL. Health Related Quality of Life in Malaysian children with thalassaemia. Health Qual Life Outcomes. 2006;4:39.
14. 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 Thai. 2010;93(Suppl 7):65–75.
15. Mazzone L, Battaglia L, Andreozzi F, Romeo MA, Mazzone D. Emotional impact in beta-thalassaemia major children following cognitive behavioural family therapy and quality of life of caregiving mothers. Clin Pract Epidemiol Ment Health. 2009;5(1):5.

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