

Impact of Thalassaemia on Quality of Life

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Abstract

Objective: To determine the impact of beta thalassaemia major on health related quality of life in terms of physical and psychosocial aspects of thalassaemia children and to compare their perception on quality of life with their parents' perception.

Duration and Place of Study: Thalassaemia centre Sir Ganga Ram Hospital, Lahore from July 2014 to December 2014.

Study Design: Cross sectional study

Methodology: Diagnosed cases of beta thalassaemia major aged 2-18 years on regular blood transfusion and their parents were included in the study. Patients with beta thalassaemia minor and intermedia, alpha thalassaemia and other haemoglobinopathies were excluded from the study. PedsQL™ 4.0 generic core scale questionnaire translated into Urdu was used with permission of MAPI Research Institute, Lyon, France.

Results: Total 266 patients of 2-18 years of age and their parents were recruited. Male children and adolescents were 169 (63%) and 97(37%) were female. Their mean age at diagnosis of beta thalassaemia major was 10.43±12.02 months. Children receiving one blood transfusion per month were 129 (49%) while 137 (51%) were receiving two or more than two blood transfusions per month. In general, Parents reported lower PedsQL 4.0 score as compare to their thalassaemic children. Psychosocial health summary score was 75.37± 25.79 versus 70.73 ± 23.16 with pvalue 0.04. Mean score for emotional functioning was 75.38 ±28.89 versus 67.31± 23.51 (p= 0.00). No significant association of age, gender and number of blood transfusion was found on perception of health related quality of life.

Conclusion: Beta thalassaemia major impairs patient's health related quality of life regarding physical and psychosocial aspects. Parents scored lower as compare to their children at psychosocial and emotional health parameters.

Key Words: PedsQL 4.0, Psychosocial, Quality of life, Thalassaemia

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Introduction

Thalassaemia is a group of inherited blood disorder of worldwide distribution. It is common in Mediterranean region.¹ In Pakistan carrier rate of Thalassaemia is 5-7%.² In low and middle income countries annually 50-100 thousand deaths occur due to thalassaemia.³ This chronic illness burdens the affected families as well as

the economy of a country.⁴

Thalassaemia is caused by deficient or absent alpha or beta chains due to genetic mutation. Gene mutation leads to synthesis of abnormal hemoglobin. This abnormal hemoglobin has impaired oxygen carrying capacity in red blood cells. Severity of disease depends

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upon the nature of mutation. Beta (β) thalassaemia is broadly classified into minor, intermedia and major. Thalassaemia carrier parents are usually asymptomatic and remain undiagnosed. They transmit the disease to their offspring in autosomal recessive fashion resulting in β -thalassaemia major. Thalassaemia major presents in infancy with severe microcytic anaemia requiring frequent regular blood transfusion. Transfusions lead to iron accumulation in the body and iron chelation therapy is recommended in these patients. Thalassaemia survivors have skeletal deformities, growth retardation, and delayed puberty, cardiovascular and renal complication.⁵⁻⁷

This chronic illness impairs quality of life by disrupting physical activities, school performance and social life of the sufferers. The affected families face financial, cultural, social and logistic problems in treatment. Hence, affected children and their parents need continuous psychosocial and economic support.⁹ Preventive measures in the form of premarital screening and prenatal diagnosis should be adopted by the societies to eradicate the disease.⁸

In literature, mostly parent's perspective has been evaluated regarding psychosocial problems of their thalassaemic children. Very few studied both parent's and patient's point of view on health-related quality of life.¹⁰ Pediatric Quality of Life Inventory (PedsQL)TM 4.0 generic core scale is a useful multidimensional tool for assessment of quality of life in chronic illness like thalassaemia. So we conducted a study using Peds QL4.0 scale from patient's self-report and parent's proxy report. Aim of study was to determine the impact of β thalassaemia major on quality of life in terms of physical and psychosocial aspects of thalassaemia children and adolescents. We compared the patient's and parent's perception on health related quality of life.

Methodology

This study is a cross-sectional study. It was conducted at thalassaemia centre of Sir Ganga Ram Hospital, Lahore from July to December 2014. Ethical approval was obtained from Ethical review Board Sir Ganga Ram Hospital (No.1166 I.E.R.B). Diagnosed cases of

beta thalassaemia major of 2-18 years of age on regular blood transfusions were included in the study. Patients with beta thalassaemia minor, intermedia, alpha thalassaemia and other haemoglobinopathies were excluded from the study. Step parents and grandparents were also excluded.

Pediatric Quality of Life Inventory(PedsQL)TM 4.0 generic core scale questionnaire translated into Urdu was used with permission of MAPI Research Institute, Lyon, France. The questionnaire was used for patients of five to eighteen years as a self report and for their parents as a proxy report. For patients of 2-4 years, questionnaire was answered by parents only. Written consent was obtained from parents and those thalassaemia patients who approached 18 years of age. The questionnaire contained 23 items, designed to explore physical, emotional, social and school functioning of children and adolescents. Privacy of participants and confidentiality of data was ensured.

All responses were measured using five points rating scale ranging from 0-4 (never a problem to almost always a problem). To reverse score, transform the 0-4 scale items to 0-100 as 0=100, 1=75, 2=50, 3=25 and 4=0. If more than 50% of the items in the scale were missing, scale score was not computed. Thus a total score was obtained. A higher score indicates better quality of life.

Data was analyzed using SPSS 20. Qualitative demographic variables like gender were calculated as percents. Quantitative variables like age at diagnosis, number of blood transfusions per month and PedsQL 4.0 scores were measured using mean and standard deviation. Parents' and children's PedsQL 4.0 scores were compared to assess the agreement in perception of quality of health. Independent group's t-test between means was applied and P value ≤ 0.05 was considered clinically significant.

Results

Total 266 patients of 2-18 years of age and their parents were recruited. Their demographic characteristics are shown in table I. Mean age of children at diagnosis of β thalassaemia major was

Table I: Demographic Characteristics

Age(years)		2-4	5-7	8-12	13-18	Total	%age
Gender	Male	27	57	51	34	169	63%
	Female	23	20	40	14	97	37%
No. of blood transfusions/ month	1/month	45	55	21	08	129	49%
	2 or > 2/month	05	28	58	46	137	51%

10.43±12.02 months.

Mean PedsQL 4.0 score obtained by thalassaemia children of age 5-18 years in their self and their parental proxy report is shown in table 2. There is significant difference in parents' proxy report and children's self report in psychosocial health summary and emotional functioning. Parents' perception of health

related quality of life of 2-4 years old children is shown in table II.

Comparison of PedsQL 4.0 total summary score among thalassaemia children receiving one blood transfusion per month and two or more than two blood transfusions per month is shown in table III. Similarly, comparison of perception of quality of life between male and female gender and age groups is shown in table III. No significant association was found.

Discussion

Thalassaemia is an inherited disorder which compromise quality of life due to its physical, psychological, social and economic implications. PedsQL4.0 total summary score in our study was 68.58±26.9. Same was found by Ismail et al who compared the health-related quality of life of thalassaemia patients with healthy participants.¹¹

In contrast, Jordanian children reported low summary score (59.2±16.3)¹² and it was comparatively high in Thai children (78.5± 2.1).¹³ The difference may be due to different in racial, cultural and social values.

Thalassaemia children have low energy level due to impaired oxygen transport which hampers their daily physical activity. Mean PedsQL 4.0 score on physical performance was 61.86±28.08.

Table II: Paeds QL 4.0 Score

PedsQL 4.0 Domains	5-18 years Peds QL 4.0 Score			2-4 years PedsQL 4.0 Score
	Patients' self report Mean±SD n= 216	Parents' Proxy report Mean±SD n= 216	P value	Parents' Proxy report
Physical Functioning	61.86±28.08	58.04 ±29.86	0.17	61.56±26.52
Psychosocial Health	75.37 ±25.70	70.73 ±23.16	0.04	71.85 ±25.38
• Emotional Functioning	75.38 ±28.89	67.31±23.51	0.00	69.34±27.26
• Social Functioning	84.65± 22.90	81.59 ±20.37	0.14	82.15± 24.64
• School functioning	66.08 ±25.59	69.49 ±23.16	0.14	64.07 ±24.26
Total Summary Score	68.59± 26.9	64.38 ±26.51	0.10	66.70 ±25.95

Table III: Association of Variables with PedsQL 4.0 Score

Association between No. of blood transfusions and PedsQL 4.0 Score

	No. of blood transfusion	PedsQL 4.0 (Mean ±SD)	P value
Parents Proxy report (2-4 years)	1/month	70.46 ±26.59	0.22
	2 or >2/month	55.23 ±23.05	
Patients Self Report (5-18 years)	1/month	70.61 ±27.75	0.81
	2 or >2/month	69.08 ±26.88	
Parents, Proxy Report (5-18 years)	1/month	66.39±25.47	1.0
	2 or >2/month	65.10± 26.96	

Association between gender and PedsQL 4.0 Score

Parents Proxy Report (2-4 years)	Male	63.54 ±29.33	0.41
	Female	70.11 ±26.04	
Patients Self Report (5-18 years)	Male	74.34 ±24.67	0.26
	Female	70.25± 27.73	
Parents, Proxy Report (5-18 years)	Male	65.84± 25.85	0.84
	Female	66.72± 29.29	

Relationship between age and PedsQL 4.0 Score

Parents Proxy Report	2-4 Years	66.70 ±25.95	0.57
	5-18 years	64.38 ±26.51	

It was 68.4 ± 27.2 in Middle Eastern children.¹⁰ Garaibeh et al reported lower physical performance (54.2 ± 15.1).¹² Data analysis from thalassaemic Malaysian children revealed 10-24% lower PedsQL 4.0 score in physical, social and school domain as compared to healthy children irrespective of their age, gender and ethnicity.¹¹

Psychosocial summary score of our participants was 75.37 ± 25.79 same was reported by Surapolchai et al and Thavorncharoensap et al.^{13,14} Quality of life among thalassaemia children, adolescents and their caregivers was studied in Malaysia and they found a lower mean psychosocial score (63.91 ± 14.65) as compare to our results.¹⁵ The difference in perception reported was may be due to varied level of awareness, available health facilities and proper resource utilization.

The children's response on quality of life was compared with parents' report on the quality life of their siblings. Overall parental PedsQL 4.0 score was lower in all domains and was statistically significant in psychosocial health ($p=0.04$) and emotional functioning ($p=0.00$). Findings are supported by previous studies.^{10, 15} The agreement between the parent's and child's perceptions may be influenced by the measured domain, age of child, parent's health related quality of life and parent-child interpersonal relationship.¹⁶ It may be due to parental distress due to financial burden and social downgrading.

Thalassaemia children require regular blood transfusion to meet the body demands. Literature review reveal that number of blood transfusions are inversely proportional to quality of life.^{13,15} It may be due to multiple hospital visits, missing school and social gatherings, financial constrains and logistic hurdles. Although total summary score in patients receiving one blood transfusion per month was higher than those receiving two or more than two blood transfusions but difference was statistically insignificant. It may be explained by the well-established joint family system and family support in our country that shares the burden of affected family.

We found no association of age and gender with health related quality of life. It is in concordance with previous studies.^{15,17} However borderline difference ($p=0.047$) was found in gender with better school functioning in Middle Eastern girls than boys.¹⁰

Thalassaemia is a preventable public health problem. Families should be supported to face the disease challenges. Awareness should be created regarding

nature of disease and available diagnostic and treatments facilities in the country. Door to door awareness campaigns must be launched on preventive strategies like premarital screening and prenatal diagnosis. Regular psychological consultations should be arranged for the patients and care givers to improve their quality of life.^{1,18}

Conclusion

β -thalassaemia major adversely affects physical and psychosocial health of children. Parents scored lower as compare to their children at psychosocial and emotional health parameters.

Abbreviations

Beta thalassaemia = β thalassaemia

Pediatric Quality of Life Inventory 4.0 generic core scale = PedsQL 4.0 scale

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