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Original Research Article

HEALTH RELATED QUALITY OF LIFE AND ECONOMIC BURDEN IN CHILDREN WITH BETA THALASSAEMIA MAJOR AGED 2-18YEARS

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Abstract

Background: Presence of chronic disease like thalassemia places tremendous psychosocial and economic burden on the patient and family. Hence assessment of quality of life in children is important. Scarcity of data on psychosocial life aspects of thalassemics limits us from implementing proper intervention strategies. Objective: 1. Assess Health Related Quality of Life in Beta thalassaemic patients 2.Assess knowledge of parents about prenatal diagnosis 3.Evaluate need for free chelating drug. **Materials and Methods:** Cross sectional hospital based study including 50 patients receiving regular blood transfusion. Structured interviews were carried out with each of the subjects and parents/caretakers using PedsQL4.0generic scale score for assessing quality of life in physical, emotional, social and school functioning. Questions relating to economic burden and knowledge of prenatal diagnosis were included. **Results:** Adverse impact of thalassaemia was perceived in all domains of quality of life by both patient and caretakers. 81% of study population donot have an idea of prenatal diagnosis or carrier state diagnosis in other siblings. Medication and transport cost accounted for most of the expenditure for each transfusion. **Conclusion:** Low scores in all domains of HRQL indicates need for psychosocial support. Inherited nature of disease is not clear in most of the families indicating need for genetic counseling. High economic burden indicates need for free medications and transport allowance.

INTRODUCTION

Beta – Thalassaemia (βthal) is genetic blood disorder and is most challenging hematologic disorder characterized by partial or no production of beta globin chains which form part of structure of hemoglobin in red blood cells. βthal is an increasingly serious public health problem throughout Indian subcontinent, Mediterranean region, Middleeast and Southeast asia.[1]

Children with βthal appear well at birth but develop progressive worsening anemia. If left untreated, this results in early deaths.[2,3] Patients with beta βthal major need regular transfusions for survival. Children have to receive blood transfusions, frequency depending on severity & iron overload requires chelation therapy.[2,3,4] Health related quality of life (HRQL) is affected in children undergoing therapy. In developing countries like India, main cause of death from βthal is non compliance due to economic and psychosocial factors.[5]

Beta Thalassemia challenges individual at physical, social, emotional, cognitive levels and disrupts quality of life. Frequency of therapy leads to financial burden for family.[6]

HRQL is a multi dimensional concept that focuses on impact of disease and its treatment on well being of individual. Measures are - capturing patients perception of disease and treatment, perceived need for health care, preferences for treatment and disease outcomes.[7] Most research involves interviews with patients, caregivers, nurses and doctors and focus is on coping strategies for thalassemia patients and their parents, attitudes and expectation of these patients, counseling strategies and screening programs.

Our study uses PedsQL4.0[8] generic core scales for assessing HRQL in βthal children (2-18 years) in four domains – Physical, Emotional, Social and school functioning. The study was also conducted to evaluate burden, to assess knowledge among affected families and to find out their attitude to prenatal diagnosis.

MATERIALS AND METHODS

Cross sectional study conducted on 65 βthal major patients receiving regular blood transfusions at Kasturba medical college hospital(KMCH), and Regional advanced pediatric care center(RAPCC), Mangalore during a period from March 2011 to June

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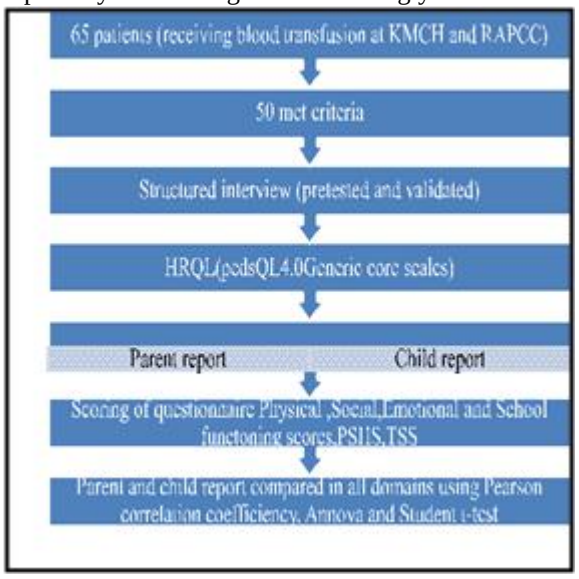
2011. 50 of them who met inclusion criteria formed the study group. The power of study is 95% .Patients and caretakers were asked to complete pretested questionnaire after necessary detailing. Questionnaire included assessing HRQL, questions relating to knowledge of prenatal diagnosis and economic burden. (medication, transport and loss of wages).

HRQL assessed withPedsQL4.0Generic core scales. This instrument has 23 items designed to measure core dimensions of health as delineated by WHO. PedsQL4.0 encompasses essential core domains for paediatric HRQL measurement– Physical, emotional, social and school functioning. It consists of developmentally appropriate forms for ages 2-4, 5-7, 8- 12,13-18 Years.

Questionnaire was translated to local language (Kannada) and then pretested. A total of 65 patients were receiving regular transfusions in KMCH & RAPCC out of which 50 met inclusion criteria and Questionnaire was answered by parent and child separately and scoring done accordingly.



On PedsQL Generic Core Scales, for ease of interpretability, items are reverse scored and linearly transformed to a 0-100 scale, so that higher scores indicate better HRQL i.e., 0=100, 1=75, 2=50, 3=25, 4=0. After reverse scoring mean score in each domain is calculated as sum of items over number of items answered in each domain separately. This accounts for missing data. If more than 50% of items in scale are missing, the Scale Score is not computed. Summary scores-

1. Psychosocial Health Summary Score(PSHS) - Computed as sum of items over number of items answered in Emotional, Social, and School Functioning Scales
2. Total Scale Score(TSS) - Computed as sum of all items over number of items answered on all Scales

A questionnaire was framed to assess knowledge of parents about prenatal diagnosis and to evaluate need for free chelating drug to βthal patients. Data was coded and analyzed using SPSS package (SPSS Inc., Chicago, IL, USA, Ver 11.5). Parent and child reports are compared using Pearson correlation co efficiency. Annova test and Student t-test are used to compare parent and child report for different age groups. Other data were analyzed using descriptive statistics. Level of significance was $P < 0.05$.

RESULTS

Of 65 children receiving treatment at KMCH &RAPCC Mangalore 50 were recruited after fulfilling inclusion criteria Mean age of cases is 8.47years. 31(62%) males and 19(38%) females.

Table 1: Health related quality of life scoring

	Physical functioning	Emotional functioning	Social functioning	School functioning	PSHS	TSS
PARENT	66.16	51.9	77	71.7	66.14	66.68
CHILD	65.42	81.94	80.83	74.31	77.81	73.92

PedsQL scoring for each subclass and PSHS and TSS. In children mean physical, emotional, social and school functioning scores- 65.42%, 81.94%, 80.83%, 74.31% and PSHS and TSS are 77.81% and 73.92%. Among parents mean physical, emotional, social and school functioning scores- 66.16%, 51.9%, 77%, 71.7% respectively and PSHS and TSS are 66.14% and 66.68%. Table 1.

Table 2: Correlation between parent and child report

	Physical functioning child	Emotional functioning child	Social functioning child	School functioning child	Psychosocial health summary child	Total summary score child
Physicalfunctioning r parent p N	.569 0.001vhs 36					
Emotional functioning r parent p N		.227 .183 36				
Social functioning r			.513			

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parent N	p			0.001vhs 36			
School functioning parent N	r p				.481 0.003 hs 36		
Psycho social health Summary parent N	r p					.396 0.017 sig 36	
Total summary score parent N	r p						.449 0.006 hs 36

vhs- very highly significant, hs- highly significant, sig- significant
r- correlation, p – significance, N – total number

Table 2 shows correlation between parent and child report. When child and parent report were compared using Pearson correlation coefficient correlation is very highly significant in social (0.001) and physical functioning (0.001), highly significant in school functioning(0.003) and TSS(0.006), significant in PSHS(0.017).

Table 3: Comparision of parent report for different age groups

		N	Mean	Std.deviation	Annova	P
Physicalfunctioning 5-8 >8	2-4 parent	14 13 23	72.24 67.46 63.61	15.766 15.208 15.198	1.381	0.261
Emotional functioning 5-8 >8	2-4 parent	14 13 23	52.50 50.00 53.04	25.019 18.371 24.943	0.703	0.930
Social functioning parent >8	2-4 5-8	14 13 23	85.50 78.08 73.48	7.314 11.094 17.018	3.459	0.04 sig
School functioning parent >8	2-4 5-8	4 13 23	74.75 73.46 70.22	11.786 10.682 18.246	0.260	0.772
Psycho social health Summary parent >8	2-4 5-8	14 13 23	67.64 66.77 64.87	11.043 7.247 16.052	0.220	0.803
Total summary score parent >8	2-4 5-8	14 13 23	69.64 67.08 64.22	8.635 8.139 15.886	0.843	0.437

sig – significant

Table 4: Tukey HSD (student t – test)

Dependent variable	(I)age group	(J)age group	Mean difference (I-J)	p
Social functioning parent	2-4	5-8	-7.42	0.334
>8			-12.02	0.03 sig
5-8	>8		-4.60	0.591

sig – significant

Table 3 and 4 shows comparison of parent report for different age groups using Annova test. There is decrease in score in all the domains as age increased but significant only in social functioning (0.04). When applied student t-test (TURKEY HSD) to social functioning report significant difference is between 2-4 year age group and >4years. There is no significant difference between 5-8 yrs and > 8years. When child report for different age groups were compared there is no significant difference in any domain.

25(50%) belonged to socioeconomic level 4 (modified Kuppuswamy classification). Mean economic burden is Rs/- 1677/month. All are receiving blood products for free. 9(18%) are receiving free medication. 39(78%) expressed medication and transport as main burden. 9(28.8%) parents knew about prenatal diagnosis and this was only after birth of affected child. 43(89.6%) were willing for prenatal diagnosis and 39(81.2%) were willing for abortion in case of affected fetus and others not willing in view of religious background.

DISCUSSION

Beta-Thalassaemia challenges the individual at physical, social, emotional, cognitive levels and disrupts the quality of life. Its frequent and complex treatment might also lead to financial burden for the individual and his/her family.[6]Although optimal medical management has reduced the difficulties faced by thalassaemics, the psychosocial problems faced by them are now of primary importance.

Presence of chronic disease like thalassemia places tremendous psychosocial burden on the patient and family. Hence assessment of quality of life in children is important. If they survive illness, children not only have longer lives to lead compared to adults, but they are less able to voice their concerns and are more vulnerable than adults. The scarcity of data on the psychosocial life aspects of thalassemics limits us from implementing proper

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intervention strategies. So we have selected this study to assess HRQL, economic burden on families with patients receiving regular transfusions.

The use of self report HRQL questionnaires in assessment of thalassaemia patients can help identify impact of disease and its associated treatments from children's perspective. A recent study found that children as young as six years old can adequately understand and accurately report their own health and well being.[9] Vincent and Higginson stressed that any measurement of the quality of life of children should include questions on physical, social, educational and psychological functioning of a child.[1]

Previous studies showed that all the domains are affected in thalassemic patients. Our study shows that thalassaemic patients scored low in all the domains – Physical, emotional, social and school functioning. The domain most affected is physical functioning as per the child report and emotional functioning as per parent report.

Parent and child report correlated in all domains except for emotional functioning. This might be because of difference in perception of emotional functioning between parents and children and also mean age in our study is 8.5 years (Sd- 4.5),hence might not perceive emotional functioning as that of adults. Emotional functioning score is in a decreasing trend when compared for 5-8 and >8yrs child report but not statistically significant. Study by Adriana Ismail et al on 96 thalassemic patients did not show significant difference in emotional functioning when compared to healthy controls.[1] Interestingly, study by Tsiantis et al suggests that thalassaemia patients have their own coping strategies in dealing with their life.[10]

Physical functioning is affected the most of all domains. This might be due to direct and indirect effects of disease itself and treatment. There is scant available information on how thalassaemia affects physical functioning (sports, tiredness, etc). Study by Ratip et al 86% of 27 thalassaemic patients complained of tiredness and suboptimal performance in sports.[11] Our study findings are congruent with these.

School functioning of thalassaemic children is below 70%. This seems to prove that having to go to hospital for blood transfusions is one of the main reasons that thalassaemia patients are missing school and thus affecting HRQL.

Social functioning of thalassaemic children is 79%. The study also reveals that thalassaemia has different impact at different ages on social functioning parent report and is significant. Scores of all domains and PSHS and TSS decreased with increasing age but this decrease is not statistically significant except for the social domain parent report.

HRQL scores are decreased in all domains, physical, social, emotional, school functioning and also psychosocial health summary score and total summary score were low. These findings support previous studies on psychosocial aspects of thalassaemia that more psychosocial support should be given to thalassaemia patients.[12,13]

Most of the parents (19%) came to know about prenatal diagnosis only after the birth of first child. Only two of them had prenatal diagnosis done. The only reason for not getting a prenatal diagnosis done is economic constraint. 81 % of the study population do not have any idea about prenatal diagnosis or carrier state diagnosis in other siblings. This indicates the need for genetic counseling for families with affected children.

The economic burden including direct and indirect costs is Rs 1677, out of which most of the amount is spent for medications and transport. 50% belongs to lower socio economic class. Loss of wages is also an important factor contributing for the economic burden. Blood products are free and this support needs to be continued.

Those patients getting admitted in a private hospital are being charged for admission and for intravenous catheters and sets. This adds to the economic burden. Only 9 (18%) are receiving medications for free. As medication cost accounts for more than 2/3rd of the expenditure per month, medications if supplied free of cost will help decrease burden. Health authorities need to take an initiative to provide medicines for free of cost and provide for transport allowances. This is necessary as thalassaemic patients require lifelong chelation therapy and regular transfusions for a better quality of life.

Most distressing aspect for parents and their families is early death(46%) of the child and physical suffering(26%). The Psycho social health summary score, total summary score and emotional and social functioning score are low. These factors indicate that both patients and their families requires psychosocial support including promotion of a clear understanding of the disease.

Thalassaemia support groups are very important as they not only educate the patients and their families about disease but also provide patients with an opportunity to meet their peers and participate in social activities. These patients need the understanding and support especially from health authorities, policy makers, school authorities and society to provide them better quality of life. The need for psychosocial support is even higher in a developing country like India where the regular and expensive treatment can pose a financial burden as well for the families of the thalassaemics.

The results from our study which indicates the need for establishing a thalassemia unit in each district/hospital for a better quality of life are

- Low HRQL scores in all the domains
- Lack of knowledge about prenatal diagnosis and carrier state diagnosis
- Economic burden due to transport and medication
- Psychological and Physical stress.

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CONCLUSION

HRQL scores were low in all domains indicates the need for psychosocial support from health authorities, society and school authorities and also there is need for psychosocial support groups to improve the quality of life and help them in leading healthy, creative and fulfilling lives. Free blood products should be continued. As medications and transport are highest burden to the family free medication and transport allowance to be given by health authorities. Most of the families don't have knowledge about prenatal diagnosis and carrier state diagnosis in siblings of affected children, so there is need for genetic counseling.

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