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Quality Of Life in Children with Thalassemia Major

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ABSTRACT Thalassemia is one of the common single gene disorders in India. Although advances in diagnosis and treatment have reduced the morbidity and mortality in patients with Thalassemia, the quality of life of the patients is commonly affected severely. The present study attampts to determine the quality of life of children with thalassemia major and compare it with that of children without any chronic disease. A case control study to compare the quality of life was carried out in the Thalassemia day care centre and pediatric wards of a tertiary care hospital. The PedsQL 4.0 generic core scale was used for assessing the quality of life of the children. 28 children with thalassemia and on regular follow up were part of the study with similar number of controls. The groups were matched demographically. The quality of life in all parameters was lower in the study group as compared to the control group.

KEYWORDS : .

Thalassemia is the most common single gene disorder in India. It is a heterogeneous group of inherited disorders of hemoglobin synthesis.[1] Thalassemia is an autosomal recessive disease originating from countries of the Mediterranean region. Deficiencies in globin chain synthesis may lead to severe anemia requiring regular blood transfusions and iron chelation therapy starting at an early age.[2]

Although, morbidity and mortality of the thalassemia major has been reduced significantly in the light of modern medical treatment, however, it could influence diverse aspects of patients' lives.[3] Patients with transfusion-dependent thalassemia are expected to have an unfavourable quality of life due to multiple factors.[4] Some aspects of thalassemia major and its associated complications are expected to impact on the QOL.[3] The World Health Organization (WHO) defined the quality of life as : "An individual perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad ranging concept affected in a complex way by the person's physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment.[5] In the present study, we aimed to assess the Quality of life in patients with thalassemia major and compare them with a population of children without any chronic disease.

Materials and methods

A descriptive case control study was conducted over a period of 4 months at a Thalassemia Day Care Centre of a tertiary level children's hospital for assessment of quality of life of children suffering from Thalassemia Major and comparison of the same with non thalassemic children of similar age group.

Study group:

All eligible children (age 2–15 years) diagnosed and registered for treatment and follow-up for Thalassemia Major and regularly attending the Thalassemia clinic and whose parents' consented for the study were included in the study.

Exclusion criteria were other forms of Thalassemia and refusal to participate in the study.

Control group:

Children admitted to the hospital wards for reasons other than Thalassemia of similar age group as the study group were included as controls. Children suffering from any chronic diseases / congenital anomalies were excluded.

The children and parents were interviewed in a single session while the children were undergoing transfusion in the centre. Controls were interviewed along with the parents at the time of their discharge from the hospital. Socio demographic and clinical characteristics were collected from interview and existing medical records.

The PedsQL 4.0 generic core scale developed by Varni et al.[6] was used for assessing the quality of life of the children. This 23-item scale was used to measure the core dimensions of health that is physical, emotional and social, as well as role (school) functioning with developmentally appropriate forms for ages 2–4, 5–7, 8–12 and 13–18 years. Each item is on 5-point rating scales from 0 to 4, labeled "Never/Almost never/Sometimes/Often/ Almost always". *Psychosocial Health Summary Score* was computed as the sum of the items over the number of items answered in the Emotional, Social, and School Functioning Scales together. The *Physical Health Summary Score* was the same as the Physical Functioning Scale Score. For the *Total Scale Score,* the mean was calculated by sum of all the items over the number of items answered on all the Scales.

Permission was taken from MAPI Research Institute, France prior to using the instrument. Written informed consent of the parents/guardians and assent of the children was obtained. Data analysis was done using Microsoft excel Data was expressed as mean. The differences between groups were analyzed by unpaired t-test

Results:

A total of 28 children were included in the study group and an equal number of healthy children were included in the control group. The demographic characteristics were similar in the study group and the control group.

Table 1: Demographic characteristics of study population

Parameter		Study group	Control group
Mean Age		7.77 (2.77)	7.14 (2.59)
Gender	Male	16	15
	Female	12	13
Age Ranges	2-4	3	2
	5-7	9	11
	8-12	14	13
	13-18	2	2

Table 2 lists mean scores of the four PedsQL 4.0 subscales and their summarised Psychosocial Health Summary and Total Summary Score for the thalassaemia patients and the differences of these scores compared to the healthy controls. The Psychosocial Health

Summary is the average of the Emotional, Social and School Functioning, and the Total Summary Score is the average scores of all items.

Physical functioning, social functioning and school functioning of thalassemia patients were lower than the healthy controls. The emotional functioning of the patients was also slightly lower than the healthy controls. The total psychosocial health summary score as well as the total summary score were also less in the study group as compared with the control group.

Table 2 Quality of Life scores for study group and control group.

Domain	Study group	Control group
Physical functioning	69.5	81.3
Social functioning	73.7	80.2
Emotional functioning	62.8	79.2
School functioning	60.3	77.1
Psychosocial health summary	65.6	78.8
Total Summary Score	66.5	79.4

Discussion:

It is important to assess the quality of life in children especially in children with chronic illness such as thalassaemia. If they survive the illness, children have longer lives to lead compared to adults and also are less able to voice their concerns. Children also tend to be more vulnerable than adults.[7]. Impact of chronic diseases like thalassemia and its treatment on the children can be be better identified by the use of self report health-related quality of life questionnaires in their assessment. The children's perspective is better brought out with these questionnaires.[8] The PedsQL 4.0 Generic Core Scale self report incorporates the dimensions necessary for measuring the Health Related Quality of Life (HRQoL) of paediatric population. It has also been tested for validity and reliability in both physically healthy paediatric populations and in paediatric acute and chronic health conditions.[9,10] Hence it was chosen by the team as the research instrument. A few studies have used PedsQL 4.0 to assess HRQoL in thalassemia patients from developing countries.[2].

The present study highlights a lower quality of life in thalassemic children as compared to healthy controls. Thalassemia is one of chronic illness of childhood which affects physical well-being. At the same time it also compromises emotional and social well-being. In addition, school dropouts and school absenteeism are common in chronic diseases like Thalassemia. These are considered as a big problem in the care of chronic childhood illness like thalassemia in India.[11] Poor QoL in patients with thalassemia major can be explained as probably due to a combination of factors like having to live with a chronic disease as well as new challenges faced as the life expectancy in thalassemia improves with better treatment. Social and cultural factors greatly affect the quality of life in a country like India.[12]

Garaibeh et al. carried out a case control study in Jordanian children comparing the outcomes of PEDsQL obtained on a sample of 128 thalassemic children aged 8–18 years with those of 83 healthy children. HRQoL mean scores in all dimensions were significantly lower in the study group as compared to their healthy counterparts.[13] Ismail et al. studied a cohort of 78 Malaysian patients (mean age 11.9 years) and compared the scores to those of 235 healthy controls. There was a 10% to 24% reduction in the physical, social and school functioning domains of the patient as compared with the controls[14]

Limitations:

A complete cognitive or psychological evaluation, which would have helped to avoid factors like psychological or psychiatric problems in parents or children was not performed in the present study. Also the sample size of the study was small. Thus a larger study with a bigger sample size would help in confirming these findings.

Conclusion:

This study shows that the overall quality of life in thalassemic children was worse when compared to healthy children. Thalassemia patients and their parents require lifelong psychological support for prevention of mental health issues. Many useful and effective psychological strategies like Cognitive-Behavioural Family Therapy are available.[15] Their use may help the psychological approach to children with beta-thalassemia major. This may in turn aid in increasing the compliance to treatment as well as decreasing the emotional burden of disease. Eventually it will help to improve the quality of life of affected children as well as their caregivers.

References

1. Dhirar, N., Khandekar, J., Bachani, D., Mahto, D. (2016). Thalassemia Major: how do we improve quality of life? Springerplus. 2016. 28;5(1):1895. eCollection 2016.
2. Caocci G, Efficace F, Ciotti F, Roncarolo MG, Vacca A, Piras E, Littera R, Markous RS, Collins GS, Ciceri F, Mandelli F, Marktel S, La Nasa G. Health related quality of life in Middle Eastern children with beta-thalassemia. BMC Blood Disord. 2012. 22;12:6. doi: 10.1186/1471-2326-12-6
3. Ansari S, Baghersalimi A, Azarkeivan A, Nojomi M, Hassanzadeh Rad A. Quality of life in patients with thalassemia major. Iranian Journal of Pediatric Hematology and Oncology. 2014;4(2):57-63.
4. Soni S, Thawani R, Idhate T, Kalra M, Mahajan A. Health Related Quality of Life in Patients with Transfusion-dependent Thalassemia. Indian Pediatr. 2016. 8;53(8):741-2. Epub 2016 Jun 1.
5. World Health Organization.2005. Measuring Quality of Life. Available at :// www .who.int/evidence /assessment/QOL/index.htm.
6. Varni JW, Seid M, Rode C. The PedsQL: Measurement model for the pediatric quality of life inventory. Medical Care. 1999;37:126-39.
7. Ismail A, Campbell MJ, Ibrahim HM, Jones GL. Health related quality of life in Malaysian children with thalassaemia. Health and Quality of Life Outcomes. 2006;4:39. doi:10.1186/1477-7525-4-39.
8. Fayers P, Machin D: Quality of Life Assessment, Analysis and Interpretation. Chichester: John Wiley and Sons; 2001.
9. The PedsQL Measurement Model for the Pediatric Quality of Life Inventory. [http://www.pedsql.org/about_pedsql.html]
10. Varni JW, Burwinkle TM, Seid M: The PedsQL 4.0 as a school population measure: Feasibility, reliability, and validity. Quality of Life Research 2006, 15:203-215.
11. Mohit Gupta, Ritika Jindal, Quality of Life in Patients with Thalassemia Major. IJSR. 2016.5; 41-42
12. Gollo G, Savioli G, Balocco M, Venturino C, Boeri E, Costantini M, Forni GL. Changes in the quality of life of people with thalassemia major between 2001 and 2009. Patient Preference and Adherence 2013;7: 231– 36
13. Gharaibeh HF, Gharaibeh MK: Factors influencing health-related quality of life of thalassaemic Jordanian children. Child Care Health Dev 2012, 38:211–218.
14. 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.
15. Mazzone L, Battaglia L, Andreozzi F, Romeo MA, Mazzone D: Emotional impact in beta-thalassaemia major children following cognitivebehavioural family therapy and quality of life of caregiving mothers. Clin Pract Epidemiol Ment Health 2009, 5:5