International Journal of Community Medicine and Public Health
Lohare BS et al. Int J Community Med Public Health. 2021 Sep;8(9):4513-4517
http://www.ijcmph.com
pISSN 2394-6032 | eISSN 2394-6040

DOI: https://dx.doi.org/10.18203/2394-6040.ijcmph20213561

Original Research Article

A cross-sectional study to assess quality of life in 2-18 years children and adolescent living with beta thalassemia major registered at comprehensive thalassemia care centre attached to tertiary care hospital in a metropolitan city

Basvaraj S. Lohare[1]*, Seema S. B. Gokhe[2]

[1]Department of Public Health, Government of Maharashtra, Mumbai, Maharashtra
[2]Department of Community Medicine, LTMMC & GH Sion, Mumbai, Maharashtra, India

Received: 08 July 2021
Accepted: 13 August 2021

***Correspondence:**
Dr. Basvaraj S. Lohare,
E-mail: medbasu@gmail.com

ABSTRACT

Background: Thalassemia is a serious inherited blood disorder. Children and adolescent living with thalassemia major require lifelong repeated blood transfusions and costly medicines for their survival. In India, β-thalassemia prevalence is 3-4%. Children are basic pillars of society although they are the most vulnerable part of society in view of nutrition, education and social life.
Methods: Cross-sectional, observational study was conducted at comprehensive thalassemia care centre attached to tertiary care hospital in a metropolitan city. 76 children and adolescents living with beta thalassemia major aged between 2 to 18 years registered for blood transfusion were included.
Results: The mean age of the study subjects was 13.81±6.71 years, 63.2% were males. Majority of study subjects belonged to Hindu religion. 46.3% study subjects were in primary school, 84.21% parents of study subjects gave the history of non-consangious marriage. 92.1% of the study subjects' parents and their relatives were not aware of history of thalassemia in the family. Genetic counselling was offered to 71.1%. Age of onset of disease was 0-6 months amongst 64.5% of children. 53.9% children had B positive blood group. Total health summary score, emotional health summary score, social health summary score and psychosocial health summary score was better in age group 2-4 years. School health summary score was very low in all age groups.
Conclusions: To improve the quality of life of children and adolescent living with beta thalassemia and to reduce catastrophic out pocket expenditure from parents of study subjects, there is need of developing comprehensive thalassemia care center at medical colleges/district hospitals for preventive and promotive health care of children and adolescents.

Keywords: Quality of life, Beta thalassemia major, Children, Adolescents

INTRODUCTION

Beta–thalassemia major is an autosomal recessive disorder which is the most commonly encountered single gene disorder in India. Beta thalassemia major is preventable genetic disorder that in its severe forms is associated with chronic life impairing and life-threatening diseases with inherent serious health sequelae that can lead to disability or death. More specifically, it is a disorder of the haemoglobin molecule inside the red blood cells. Genetic disorders where both genes are required to be abnormal for the disease to manifest are called autosomal recessive disorder. It is an inherited genetic disease, a disease passed from parents to children through the genes. The two main types of thalassaemia are beta (β) and alpha (α) (Figure 1 and 2).[1,2] Both affect the production of normal

haemoglobin – a key constituent of human red blood cells.[1] Beta thalassemia major is defined as a serious inherited disorder in which the bone marrow cannot form sufficient red blood cells. Some cells get distorted within the bone marrow. Normal cell survives for 120 days. In thalassemia, red cell survival is reduced.

Between births and three to six months, baby with beta thalassemia major will seems normal and quite healthy. Then baby will begin to show symptoms of anaemia (they became pale) there may be shortness of breath, jaundice and an enlarged spleen.[2] Children and adolescent living with thalassemia major require lifelong repeated blood transfusions and costly medicines for their survival. Frequency of blood transfusion varies in individual case from 2 weeks to 4 weeks. Besides these transfusions, patient living with this disorder requires chelation therapy, including desferal injections which are costly, that need to be infused over a period of several hours. That means the patient has to keep the injection and the infusion pump attached to the body over a period of ten-twelve hours, several days a week.

High incidences of thalassemia exist in Southeast Asian countries, especially in India. It is estimated that 3.9% of the existing population in India carries the thalassaemic gene. In India, β-thalassemia is prevalent across the country, with an average frequency of carriers being 3-4%.[3-5] Prevalence in Maharashtra in different district is 1-6%. It is estimated that about 10,000 to 15,000 babies with beta thalassemia major are born every year.[3] In Mumbai prevalence of beta thalassemia carrier is 2.25-2.95%.[3]

A higher frequency has been observed in certain communities, such as Sindhis, Punjabis, Guajarati's, Bengalis, Mahars, Kolis, Saraswats, Lohanas and Gaurs.[5,6] It is estimated that about 10000-15000 babies with thalassemia major (TM) are born every year.[9] The only cure available for these children with thalassemia major is bone marrow transplantation (BMT) that is hematopoietic stem cell transplant (HSCT). However, this can help only a few patients because of cost, paucity of BMT centres, or non-availability of a suitable HLA matched donor. Therefore, the mainstay of treatment is a regimen of regular blood transfusions followed by adequately monitored iron chelation therapy to remove the excessive iron overload-as a consequence of the multiple blood transfusions. Thus, it is a transfusion dependent disorder and places a great burden on healthcare services.

Children are basic pillars of society although they are the most vulnerable part of society in view of nutrition, education and social life. With increasing control of communicable disease, inherited abnormalities are assuming a proportionately greater importance. As in India there are few studies carried out to assess how to improve the quality of life and found factors improving the quality of life were control of iron overload, management of co-morbidities and fewer hospital visits. The present study was conducted to assess the quality of life of children and adolescent living with beta thalassemia major which will help policy makers to design interventions to reduce burden of beta thalassemia children and need of establishment of prevention and control programme for thalassemia.

METHODS

It was a cross-sectional, observational study conducted at comprehensive thalassemia care center attached to tertiary care hospital in a metropolitan city. 76 children and adolescent living with beta thalassemia major aged between 2 to 18 years registered for blood transfusion were included in the study and study duration was 18 months. Census enumeration method of sampling was used for data collection. A predesigned pretested semi structured interview schedule questionnaire was prepared in accordance with study objectives. The questionnaire for study consists of part A which include socio-demographic data and part B include to access the quality of life of study subjects using PaedsQol 4.0 generic core scale questionnaire. This 23 items scale is to be used to measure the core dimensions for pediatric health-related quality of life (HRQoL) measurement that is physical functioning (8 items), emotional functioning (5 items), social functioning (5 items), school functioning (5 items) 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 labelled, never/almost never/sometimes/often/ almost always, 5-point Likert scale from 0 (never) to 4 (almost always).

Permission is taken from MAPI Research Institute, Lyon, France prior to using the instrument and Marathi, Hindi translation of same is obtained. The questionnaire was prepared in English and interview was conducted in the language in which they best understood. The approval of ethical clearance was sought from Institutional scientific review committee and ethical committee prior starting the study.

RESULTS

In present study, the age of study subjects ranged from 2-18 years. Maximum 22 (28.9%) belonged to age group of 8-12 years and 48 (63.2%) were males. The mean age of the study subjects was 13.81±6.71 years. Majority (77.6%) of study subjects belonged to Hindu religion. 11.84% children were not eligible for school, 23.6% children were in pre-primary school, whereas 40.7% study subjects were in primary school, 14.9% children were in secondary school and 11.9% children were in higher-secondary school (Table 1).

With reference to the marital status of parents, it was observed that majority (84.21%) of parents of study subjects gave the history of non-consangious marriage, whereas rest gave history of consangious marriage. Only 7.9% of the study subjects' parents and their relatives were aware of history of thalassemia in family, whereas 92.1%

of the study subjects' parents and their relatives were not aware of history of thalassemia in the family. It is observed that thalassemia status of both father and mother was thalassemia minor i.e. 76 (100%). It was observed that post marital genetic counselling was offered to only 71.1% parents of study subjects (Table 1).

Table 1: Distribution of study subjects according to socio-demographic profile.

Details	Frequency (n=76)	Percentage (%)
Age (years)		
2-4	18	23.7
5-7	18	23.7
8-12	22	28.9
13-18	18	23.7
Gender		
Male	48	63.2
Female	28	36.8
Religion		
Hindu	59	77.6
Muslim	15	19.7
Christian	1	1.3
Others	1	1.3
Educational status		
Children not eligible for school	09	11.84
Pre-primary	18	23.68
Primary	31	40.79
Secondary	10	13.16
Higher secondary	08	10.53
Type of marriage		
Consangious	12	15.79
Non-consangious	64	84.21
Awareness of family h/o thalassemia		
Yes	6	7.9
No	70	92.1
Genetic counselling of parents		
Yes	54	71.1
No	22	28.9

Maximum (64.5%) number of children their age of onset of disease was 0-6 months, followed by 7-12 months (23.7%), 13-18 months (5.3%), 19-24 months (3.9%) and >24 months (2.6%). The mean age of onset of disease was 8±6.81 months. 53.9% belonged to B positive followed by O positive (25%), A positive (15.8%) and AB positive (5.3%). Maximum (73.7%) of the study subjects required blood transfusion every 21 days, followed by (15.8%) study subjects require blood transfusion every 15 days and (10.5%) study subjects require blood transfusion every 30 days. 5.26% study subjects required splenectomy due to excessive enlargement of spleen. All the study subjects were completely immunised and received injection hepatitis A and B. Severe iron infiltration was observed amongst 19.74% patients as per confirmed from reports of magnetic resonance imaging (MRI) abdomen and chest (Table 2).

Table 2: Distribution of study subjects according to age of onset of disease & frequency of blood transfusion.

Details	Frequency (n=76)	Percentage (%)
Age of onset of disease in months		
0-6	49	64.5
7-12	18	23.7
13-18	4	5.3
19-24	3	3.9
>24	2	2.6
Blood group		
A positive	12	15.8
B positive	41	53.9
AB positive	4	5.3
O positive	19	25
Frequency of blood transfusion (days)		
15	12	15.8
21	56	73.7
30	8	10.5
Iron infiltration		
No infiltration	25	32.89
Mild infiltration	34	44.74
Moderate infiltration	2	2.63
Severe infiltration	15	19.74

Total health summary score was better in age group 2-4 years (72.42) as compared to other age groups. Physical health summary score was better in age group 2-4 years (79.69) as compared in the age group of 8-12 years. Emotional health summary score was better in age group 2-4 years (81.39) as compared to in the age group of 13-18 years. Social health summary score was better in the age group of 2-4 years (89.44). As the school going age starts from 5-7 years, it was observed that there was very less school health summary score observed in 2-4 years age group. In 8-12 years, Age group school health summary score was 45.45 better as compared to other age groups score. Overall school health summary score was very low in all age groups and it was most affected domain amongst all QoL health summary score. Psychosocial health summary score is better in age group 2-4 years (69.62). There is significant association between ages of study subjects with various domains of quality of life (Table 3).

Table 3: Distribution of study subjects according to domain of assessment of quality of life in different age groups.

Quality of life assessment scores	Age groups			
	2-4 years	5-7 years	8-12 years	13-18 years
Total health summary score	72.42	68.60	66.85	63.47
Physical health summary score	79.69	70.83	66.19	13.28
Emotional health summary score	81.39	78.06	76.59	70.83
Social health summary score	89.44	80	79.55	72.78
School health summary score	9.72	44.17	45.45	40.56
Psychosocial health summary score	69.62	65.97	64.14	61.42
Degree of freedom=9	Chi square value=27.94		P value=0.0009744	

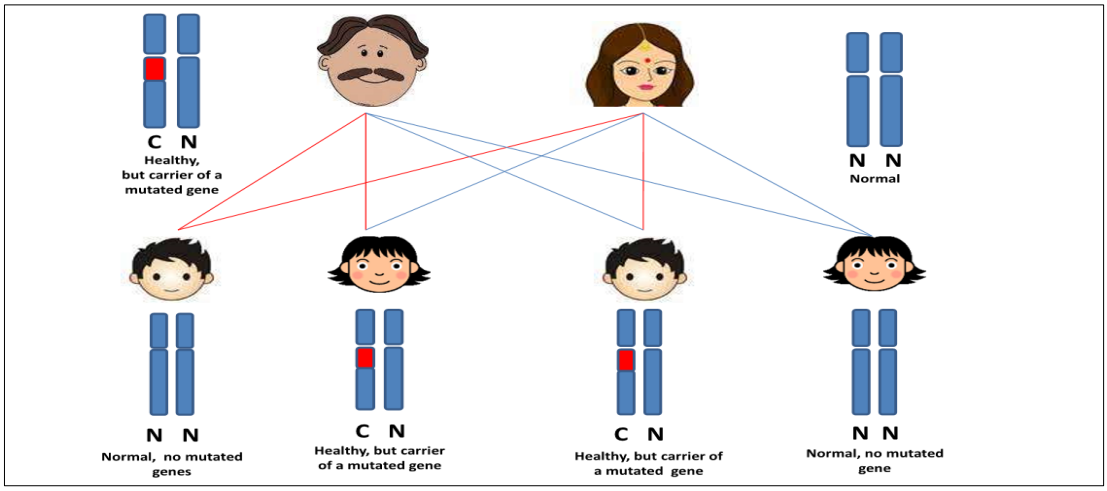

Figure 1: Inheritance and risk when one parent carries thalassemia allele.[2]

Each child has: 50% risk of inheriting one mutant allele (CN=carrier), and 50% chance of inheriting both normal alleles (NN=normal)

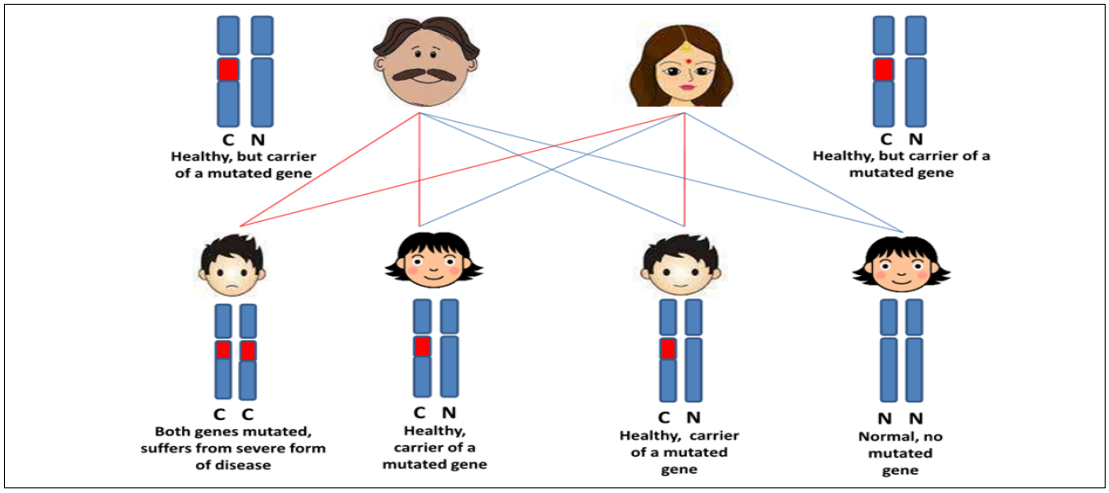

Figure 2: Inheritance and risk when both parent carries thalassemia allele.[2]

Each child has: 25% chance of inheriting of 2 normal genes (normal child), 50% chance of inheriting one altered gene and one normal gene (beta thalassemia carrier), and 25% chance of inheriting 2 altered genes, in this case both the gene carry mutation (beta thalassemia major)

DISCUSSION

There are 76 children and adolescents living with beta thalassemia major included in the study. 63.2% were males and 36.8% were females. The mean age of the study subjects was 13.81±6.71 years. Maximum 22 (28.9%) belonged to age group of 8-12 years. In the study by Dhirar et al, mean age of children was 8.69±4.98 years (range: 2–18 years).[6] Two-thirds 63.5% were boys and 36.5% were females. These observations are similar to present study. Similarly, in the study by Torcharus and Pankaew, mean age was 10.6 years; 61% were males and 39% were

females.[8] The mean age was 10.6 years. Present study shows that majority of study subjects belonged to Hindu religion i.e. 59 (77.6%) followed by Muslim religion i.e. 15 (19.7%) and Christians and others were equal i.e. 1 (1.3%) respectively. Similarly, in study by Dhirar et al majority of study subjects belonged to Hindu religion 71.4%, followed by Muslim religion 24.9% and others 3.7%.[6]

In present study, it is observed that, 11.84% children were not admitted in school, 26.9% children were admitted in pre-primary school, 46.3% study subjects were educated primary education, 14.9% children were educated secondary education and 11.9% children were educated higher-secondary education. Whereas in the study by Dhirar et al, total of 173 children were of school going age of which 89.5% were currently attending school and 8.6% were school dropouts.[6] A few children (1.7%) were never enrolled in school. The result is different from present study as they have calculated only school attending and dropouts. In the study by Torcharus and Pankaew et al, (80%) were in primary or secondary school.[8] These observations are similar to present study.

In the present study, total health summary score was 72.42, physical health summary score was 79.69, emotional health summary score was 81.39, social health summary score was 89.44, and psychosocial health summary score was 69.62 which were better in age group 2-4 year. Very less school health summary score observed in 2-4 years age group. In the study by Dhirar et al, total mean QoL score of the children enrolled at the center was 82.0, physical QoL score 81.3, emotional Qol sore 75.4, social domain score 92.0, school functioning core 77.8 and psychosocial QoL score 82.5.[6] Whereas, in the study by Torcharus and Pankaew et al, total health summary scores were 74.35, physical functioning and psychological health were 72.32 and 75.44 respectively, emotional functioning score was 78.77, social functioning score was 85.40, school functioning score was 62.14.[8] These observations are similar to present study.

CONCLUSION

Development of screening and counselling program in premarital and post marital couple before having child, so as to avoid birth of child with thalassemia.

Recommendations

To improve the QoL of children and adolescent living with beta thalassemia and to reduce catastrophic out pocket expenditure from parents of study subjects, there is need of developing comprehensive thalassemia care centre at medical colleges/district hospitals for preventive and promotive health care of children and adolescents. To improve school attendance and performance extra classes should be arranged for missed period for children and adolescents living with thalassemia major. Operational screening programme for haemoglobinopathies detection in community and facilities level will reduce burden of disease.

ACKNOWLEDGEMENTS

Authors would like to thank study subjects and their parents who have given invaluable time without which the study would not have possible and they would like to extend thanks to Director and staff members at CTCC for their kind support.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. Hakeem GLA, Mousa SO, Moustafa AN, Mahgoob MH, Hassan EE. Health-related quality of life in pediatric and adolescent patients with transfusion-dependent ß-thalassemia in upper Egypt (single center study). Health Qual Life Outcomes. 2018;16(1):59.
2. Thalassemia – care and control in new millennium by Dr. V.P. Chaudhary, India.
3. Colah R, Italia K, Gorakshakar A. Burden of Thalassemia in India: The road map for control. Paediatr Haematol Oncol J. 2017;2:79-84.
4. Sandra SJ. Stress and Coping among Parents of Children Having Thalassemia. 2015;4(7).
5. Dhirar N, Khandekar J, Bachani D, Mahto D. Thalassemia Major: how do we improve quality of life? Springer Plus. 2016;5(1):5.
6. Torcharus K, Pankaew T. Health-related quality of life in Thai Thalassemic children treated with iron chelation. Southeast Asian Journal of Tropical Medicine and Public Health. 2011;42(4):951-9.
7. NHM Guidelines for Prevention and control of Hemoglobinopathies in India. 2016. Available at: https://www.eraktkosh.in/BLDAHIMS/bloodbank/aboutHemoglbinopathies.cnt. Accessed on 16 March 2021.
8. Mapi Research Trust - PedsQL™ Generic Core Scales (English, Hindi & Marathi) used to assess quality of life. Available at: https://eprovide.mapi-trust.org/instruments/pediatric-quality-of-life-inventory. Accessed on 16 March 2021.
9. NHM Guidelines for Prevention and control of Hemoglobinopathies in India. 2016. Available at: https://www.eraktkosh.in/BLDAHIMS/bloodbank/aboutHemoglbinopathies.cnt. Accessed on 16 March 2021.

Cite this article as: Lohare BS, Gokhe SSB. A cross-sectional study to assess quality of life in 2-18 years children and adolescent living with beta thalassemia major registered at comprehensive thalassemia care centre attached to tertiary care hospital in a metropolitan city. Int J Community Med Public Health 2021;8:4513-7.