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Original Research Article

Knowledge, practice and experiences of parents with a thalassemic child

Amit Saxena, Mumtaz Sharif*, Sadaf Siddiqui, Swati Singh

Department of Pediatrics, D Y Patil Medical College, Nerul, Navi Mumbai, Maharashtra, India

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***Correspondence:**
Dr. Mumtaz Sharif,
E-mail: Mumtaz23.75@gmail.com

ABSTRACT

Background: Thalassaemia is a chronic disorder requiring lifelong transfusions and medications causing emotional and financial burden to the family. This study was done to assess the knowledge and awareness of parents having a thalassemic child and to ameliorate their experiences in the upbringing of their child.
Methods: A cross sectional, descriptive study was conducted in Pediatric Ward of a Tertiary care Hospital in Navi Mumbai from May-August 2016 in which parents of 40 beta Thalassemia major children between age group of 6 months to 15 years were enrolled. Parents were interviewed through a questionnaire to assess their knowledge, awareness, the practices they follow in regard to the transfusion, treatment, vaccination and prevention of thalassemia. The study also focuses on the emotional suffering of the parents.
Results: 47.5% of parents were aware of thalassemia being a genetic disorder.62.5% were aware of regular blood transfusion. 23 parents were aware of the regular medications taken by their children. 42.5% parents had adequate knowledge of the optional vaccines to be taken. 90% parents followed a good practice of getting their child for regular blood transfusion and 92.5% gave them regular medicines.31 parents would periodically get their child investigated but 65% of parents lacked in giving optional vaccines to the child.60% agreed to undergo MTP while only 45% knew about the antenatal detection.
Conclusions: Majority of parents followed good practice and had a positive attitude, though the knowledge and awareness about the disease was inadequate.

Keywords: Experiences, Knowledge, Thalassemia

INTRODUCTION

Thalassemia is a hereditary haemoglobinopathy resulting from the absence or reduced synthesis of either alpha or beta globin chain. Depending upon the globin chain involvement, thalassemia is categorized into Alpha-thalassemia and Beta thalassemia. Beta thalassemia is further classified as beta-thalassemia major, intermedia and minor, on the basis of clinical severity. The worldwide prevalence of annually affected conceptions with Beta thalassemia is 42,409 cases with annual births worldwide being 1,28,667,000.[1] High prevalence is present in populations in the Mediterranean, Middle-East, Transcaucasia, Central Asia, Indian subcontinent, and Far East. The South-east Asia alone accounts for 21,693 annually affected conceptions with annual births being 38,139,000. Every year, 10,000 children with Thalassemia are born in India[1].The prevalence of thalassemia is found to be very high among certain communities such as Sindhis and Punjabis from Northern India, Bhanushalis, Kutchis, Lohanas from Gujarat, Mahars, Kolis and Agris from Maharashtra, etc.

Thalassemia exists in 3 forms: Thalassemia trait or the asymptomatic carrier stage – carry a single beta globin mutation and are generally asymptomatic except for

microcytosis and mild anemia.[2] Thalassemia intermedia– have atleast 1 beta $^{+}$ thalassemia mutation and is less severe phenotype such that they do not require regular transfusions. Thalassemia major-In β thalassemia major, the production of β-globin chains is severely impaired, because both β-globin genes are mutated (homozygous state). The severe imbalance of globin chain synthesis and production results in ineffective erythropoiesis, severe microcytic, hypochromic anemia leading to early transfusion therapy.

Children usually become symptomatic from profound weakness and cardiac decompensation during the later half of first year. Transfusions begin in the 2nd month to 2nd year of life but rarely later. Affected children fail to thrive and become progressively pale. Bone deformities secondary to marrow expansion, hepatosplenomegaly, cachexia, classical haemolytic facies (maxilla hyperplasia, flat nasal bridge, frontal bossing) or pathological fractures may develop. Management of patients includes regular blood transfusions to keep Hb above 9-10gm/dl, iron chelation therapy to reduce iron overload, surgical interventions like splenectomy. Allogenic hematopoietic transplantation is the definitive curative treatment available till date.[4]

Since the past three decades, regular blood transfusions and iron chelation have transformed thalassemia from a rapidly fatal disease in early childhood to a chronic disease compatible with prolonged life. Today life expectancy of a thalassemic varies between 25-55 years, depending on the compliance with medical treatment. Despite increased life expectancy, complications keep arising which are mainly transfusion induced haemosiderosis. These include heart disease (heart failure and arrhythmias), chronic liver hepatitis, which can evolve in cirrhosis and, rarely, hepatocellular carcinoma, endocrinal problems (hypogonadism, hypothyroidism, diabetes, hypoparathyroidism), stunted growth, osteoporosis, risk of serious infections like HIV, HCV, HbsAg contracted through blood transfusions. The long-term treatment and management of thalassemia is complex, lifelong and involves lot of inconvenience for the parents in the form of repeated hospitalisations, blood transfusions, chelation therapy, which often affects their physical and mental health adversely.

The best way to reduce the burden of thalassemia is prevention. There are different strategies to prevent thalassemia, which include parental awareness, population screening, genetic counselling, and prenatal diagnosis. Creating awareness and educating parents can prove to be cost-effective in the prevention of the disease and improvement of quality of life of patients with thalassemia. In our country, there are studies on general population about screening and prenatal diagnosis of thalassemia, but very few studies emphasize on the awareness and experiences of parents raising a thalassemic child. The emotional affliction and financial anguish with the agony and apprehension of the future of their children is put forth by this study.

METHODS

This was a cross sectional descriptive study conducted in the Pediatric ward between May to August 2016, where parents of 40 beta major Thalassemic patients were interviewed in a pre-structured questionnaire for knowledge, attitude and practice in a language best understood by them. Parents who did not give consent were excluded from the study. Data was analysed using relevant statistical tests.

RESULTS

This study included interviewing 40 parents of thalassemia major children aged 6 months to 15 years. Of these, 25 were males (62.5%) and 15 (37.5%) were females.

On testing knowledge about thalassemia, 47.5% of total parents were aware of thalassemia being a genetic disorder and 55% knew that it is caused by consanguineous marriage. Nearly half (45%) of parents were aware of thalassemia being detected antenatally but a large number of parents (55%) did not know of any permanent cure for it. Majority of parents (62.5%) understood the need and advantages of regular blood transfusion. While only 40% could appreciate the need for regular investigation necessary in thalassemia, 23 parents (57.5%) were aware of the regular medications taken by their children. 57.5% parents did not have adequate knowledge of the optional vaccines to be taken. Majority of parents (62.5%) could not comprehend the side effects of blood transfusion and 57.5% did not know of side effects of chelation therapy.

Regarding attitude, majority of parents 72.5% did not consider them as a burden even though 65% felt there was a lot of family expenditure on this child. Almost 62.5% of parents felt that they had an emotional turmoil in raising their child. Majority 80% felt that the disease would hamper their child's education and adolescence. After interviewing, 60% agreed that if given an option for prenatal diagnosis for early detection of thalassemia, they would undergo relevant tests in next pregnancy. Also, a large group (67.5%) were convinced to undergo medical termination of pregnancy if they carried a thalassemic child. Almost 65% of parents had informed family and society about their child's disease but almost 35% had inhibitions about disclosing it to the family and society.

Almost 90% parents followed a good practice of getting their child for regular blood transfusion and 92.5% gave them regular medicines (including chelation therapy). 31 parents (77.5%) would periodically get their child investigated but 65% of parents lacked in giving optional vaccines to the child. Almost (27.5%) of parents had undergone antenatal diagnosis for next pregnancy. All

parents encouraged their children to lead a normal life even after being affected by this disease.

DISCUSSION

This study interviewed parents of 40 children aged 6 months to 15 years with a mean age of 5.34 years suffering with beta thalassemia major who visited our hospital for regular check-up and blood transfusion between May-August 2016. This was a prospective cross-sectional Hospital based durational study. Parents were interviewed in the language best understood by them. Majority of parents (78%) belonged to the lower socioeconomic strata and had primary or secondary schooling.

In current study, we had a male preponderance with 25 males comprising 62.5% and 15 were females i.e. 37.5%. Thalassemia is an inherited autosomal recessive disorder, seen equally in males and females. A study by Goyal JP et al 3 and Bandyopadhyay B et al showed male preponderance similar to present study.[4] However, one of the studies conducted in Pakistan by Arif F showed slight female preponderance.[5]

Regarding knowledge about thalassemia, parents were asked questions pertaining to transmission of disease, prevention, treatment and medications involved. Nearly half of them (47.5%) were aware that thalassemia is a genetic disorder and 55% had knowledge that it is common amongst consanguineous marriage. This number seems very low to the authors as the burden bearers of the disease themselves amongst whom the awareness is of utmost importance, are not aware of the mode of inheritance. Arif F et al have showed similar lack of knowledge among parents regarding transmission of thalassemia.[5] On inquiring about antenatal detection, only 45% of parents were aware of thalassemia being detected antenatally whereas in the study by Ali S et al, 74% of parents were aware of antenatal detection of thalassemia which was much higher than present study.[6] In the developed countries much attention has always been directed to the prevention of disease by detection of thalassemia carriers and marriage counselling. By using this prevention programme in Sardinia, the incidence of thalassemia patients has decreased from 1:250 live births to 1:1000 live births.[7] Similarly, in Cyprus, the incidence of thalassemia major cases dropped by 96%.[8]

Sixty two percent of parents were aware of the need for regular transfusion and only 40% could appreciate the need for regular investigation needed to detect the complications in thalassemia including increased iron overload depicted by high ferritin levels, hypothyroidism, diabetes, heart disease, osteopenia, viral infections such as hepatitis B and C due to repeated transfusions. Thalassemic children need to take iron chelators like oral deferasirox or deferiprone, subcutaneous desferrioxamine to reduce iron overload. In present cohort, 92.5% of parents gave their children regular chelators but only 23 parents (57.5%) were aware of the purpose of regular medications taken by their children. Similar lack of awarenesss was seen with vaccination as only 43% of parents had knowledge of vaccination needed by their child. On asking about bone marrow transplant, which is the definitive permanent cure for thalassemia till date but an expensive treatment option, a large number of parents (55%) were not aware of this treatment.

For testing the attitude, parents were questioned about issues like finances, emotional outlook, prenatal testing. Almost 62.5% of parents in present study had an emotional turmoil in the upbringing of their children. They were stressed about the physical health as well as the psychosocial outcome of their children owing to the disease. They had a number of issues to address from the best choice of treatment for their child to the emotional responses their child has, living with a chronic disease. The same has been reflected in the study by Sharma S, Seth B et al which focussed on the emotional stress faced by parents in raising their thalassemic child and they found significantly higher proportion of poor health in caregivers than the control group.[1] A study done by Ismail A et al in thalassemic children in Malaysia revealed that the quality of life of thalassemia patients is indeed much lower than the quality of life of healthy controls regardless of age, gender, ethnicity.[9] Majority of parents in this study, (80%) felt disease would hamper child's education as also seen in the study mentioned above. Seventy two percent of parents did not consider their child as a burden emotionally but almost 65% were burdened financially as reflected in the study by by S Mallik in a tertiary care centre in Kolkata where it was observed that 70% of the families had to spend up to 20% of their yearly income for treatment of thalassemia.[10]

Almost 60% agreed to undergo prenatal testing, if they were given an option for prenatal diagnosis. Ali S et al observed that a large number of parents were aware of prenatal screening in thalassemics in her study but only few opted due to religious beliefs. In this study, 67.5% parents agreed to undergo MTP if they were positive for a thalassemic child which is much higher than the study by Basu M in a tertiary care centre in Kolkata where 42% of parents agreed for MTP.[11] A majority of parents (65%) had informed family and society about their child's disease and supported their children in every aspect.

Almost 90% parents came for regular blood transfusion most probably because as the haemoglobin of their child would fall, symptoms like fatigue and weakness would develop which compelled them to get their children for transfusion. In present study, 92.5% parents were giving regular chelators which was mostly oral deferasirox at our centre while Arif F et al saw only 10.3% parents giving chelation therapy which is low as compared to present study.

Thalassemia is an immunocompromised state and poses serious risk of infections and requires additional vaccines

for immunity. On inquiry, only 35% gave their children additional vaccines vaccines due to financial constraint and lack of knowledge to afford these vaccines. 77.5% of parents underwent regular investigations even though majority of them were not aware of their importance. Study by Arif F et al showed only 5% parents underwent antenatal diagnosis for next pregnancy while at present centre, nearly 27.5% parents had undergone it showing the need for more awareness regarding prenatal diagnosis, as we aim to achieve 100% of parents undergoing prenatal diagnosis for thalassemia.[3]

The highlighting point in present study is that all parents encouraged their children to lead a normal life even after being affected by this disease showing a scope of better management of thalassemics.

CONCLUSION

This study has focussed on the lack of knowledge among parents of thalassemic children about issues like prenatal awareness, additional vaccines, need for transfusion, chelators, etc but depicts their good outlook and mindset in treatment of their child. Parents may be emotionally or financially weakened but it has only impassioned them for the optimal treatment for their child. We have witnessed that the adequacy of knowledge and prenatal awareness had declined incidence of thalassemia in Cyprus drastically and we must take motivation from such places and take suitable measures to do so.

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REFERENCES

1. Sharma S, Seth B, Jawade P, Ingale M, Setia MS. Quality of life in children with thalassemia and their care givers in India. Indian J Pediatr. 2017;84(3):188-194.
2. Kliegman RM, Stanton B, St. Geme J, Schor NF. Nelson Textbook of Pediatrics. 20th ed. South Asian Edition. 2015;2:2352-3.
3. Goyal JP, Hpapani PT, Gagiya H. Awareness among parents of children with thalassemia major from Western India. Int J Med Sci Public Health. 2015;4:1356-13.
4. Bandyopadhyay B, Nandi S, Mitra K, Mandal PK, Mukhopadhyay S, Biswas AB. A comparative study on perceptions and practices among parents of thalassemic children attending two different institutions. Indian J Commun Med. 2003;28:128-132.
5. Fehmina A, Jabeen F, Ahmer H. Awareness among parents of children with thalassemia major. J Pak Med Assoc. 2008;58(11):621-4.
6. Ali S, Saffiullah, Malik F. Awareness of parents regarding beta thalassemia major disease. Khyber Med Univ J. 2015;7(2):72-5.
7. Cao A, Rosatelli C, Galanello R, Monni G, Olla G, Cossu P et al. The prevention of thalassemia in Sardinia. Clin Genet. 1989;36:277-85.
8. Buki MK, Qayum I, Siddiqui N. Prevalence and preventive measures for thalassemia in Hazara region of NWFP Pakistan. JAMC. 1998;10:28-31.
9. Ismail A, Campbell M, Ibrahim H, Jones G. Health related quality of life in Malaysian children with Thalassemia Health qual Life Outcomes. 2006;4:39.
10. Mallik S, Chatterjee C, Mandal PK, Sardar JC, Ghosh PN. Expenditure to treat thalassaemia: an experience at a tertiary care hospital in India. Iran J Public Health. 2010;39(1):78-84.
11. Basu M. A study on knowledge, attitude and practice about thalassemia among general population in outpatient department at a Tertiary Care Hospital of Kolkata. J Preven Medic Holistic Health. 2015;1(1):6-13.

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