Research Article

Awareness among parents of children with thalassemia major from Western India

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Abstract

Background: Parental awareness is the best way to prevent thalassemia in children. Parental knowledge regarding transfusion and chelation helps to improve the quality of life of children with thalassemia.

Objective: To determine awareness among parents of children with thalassemia major regarding the transfusion practices and iron chelation.

Materials and Methods: This cross-sectional study was conducted at the thalassemia ward of KT Children Hospital and PDU Medical College, Rajkot, Gujarat, India, from August to October 2012. Parents of children with thalassemia, receiving blood transfusion from PDU Medical College Blood Bank, were interviewed using a predesigned questionnaire. Informed verbal and written consent was obtained from these parents. Questions regarding duration of illness, awareness about blood screening, mode of transmission of the disease, and knowledge and practices about prevention and treatment of the disease were asked.

Result: A total of 110 caretakers were questioned. A majority of them were from low socioeconomic class and 18% were illiterate. Among the parents of children undergoing transfusion, only 15.8% knew the importance of blood screening. Although all parents knew about iron overload, but only 48% of the children were receiving iron chelation therapy adequately. Despite of the fact that 15% were aware that thalassemia is an inherited disorder, family screening of sibling and antenatal diagnosis in subsequent pregnancies were done in only 5.8% and 5% respectively.

Conclusion: Awareness of the parents regarding the disease was inadequate. General public and parents of children with thalassemia should be sensitized in this regard.

KEY WORDS: Thalassemia major, blood transfusion, awareness

Introduction

Thalassemia is one of the most common inherited gene disorders in India. It is estimated that every year around 8,000–10,000 children are born with thalassemia in our country.[1] Further, thalassemia is a serious disease causing severe anemia, ineffective erythropoiesis, extramedullary hematopoiesis, and iron overload resulting from transfusion and increased iron absorption.[2,3] The definitive treatment of thalassemia is bone marrow transplantation, which is beyond the reach of many patients in this country.[4]

The best way to reduce the burden of thalassemia is prevention. However, the quality of life of children with thalassemia should be improved. There are different strategies to prevent thalassemia, which include parental awareness, population screening, genetic counseling, and prenatal diagnosis.[5–7] Creating awareness and educating parents proved 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 population screening and prenatal diagnosis of thalassemia, but there is paucity of data

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Table 1: Demographic profile of patients with thalassemia (n = 110)

Age (years)	Male	Female
<5	15 (13.63%)	6 (5.45%)
5–10	15 (13.63%)	10 (9.09%)
10–15	25 (22.72%)	8 (7.27%)
>15	21 (19.09%)	10 (9.09%)
Parental education	**Mother**	**Father**
Illiterate	21 (19.09%)	19 (17.27%)
Matriculate	71 (65.54%)	68 (61.18%)
Below matriculation	18 (16.36%)	23 (20.90%)
Consanguineous marriage	14 (12.71%)	
H/o thalassemia in relatives	9 (8.18%)	
H/o thalassemia in siblings	4 (3.36%)	

on thalassemia awareness. Moreover, awareness is found to be low in parents of children with thalassemia. Therefore, this study was undertaken to assess the awareness of parents of children with thalassemia.

Materials and Methods

This cross-sectional study was carried out at the thalassemia ward of KT Children Hospital and PDU Medical College, Rajkot, Gujarat, India, from August to October 2012. The subjects were parents of children with thalassemia coming to thalassemia ward of P.D.U Medical College Blood Bank for blood transfusion which is a registered blood bank that provides blood and blood products to all patients.

The study protocol was approved by the ethical committee of the PDU Medical College. At the time of initiating the study, each participant was informed about the study protocol and written consent was obtained from each. A single interviewer administrated the predesigned and pretested questionnaire to parents and explained about the questions.

The questions consisted of demographic profile of patients, parental education, history of consanguineous marriage, and thalassemia in relatives. Further, awareness regarding transfusion practices, other modalities of treatment in thalassemia, iron overload, and iron chelation was also assessed.

Statistical Analysis

All the data were entered in SPSS software, version 16, and evaluated. Results have been presented in the form of frequencies and percentages where applicable.

Result

A total of 110 caretakers were interviewed. The mean age of patients was 10.86 years, with ages ranging from 2 to 18 years. There was a slight predilection for males, which accounted for 69% of the patients. Total 18% subjects were illiterate, and consanguinity was positive in 12.7%. Family history of thalassemia was present in 8.18%, and 3.36% were having positive history of thalassemia in siblings [Table 1].

Table 2: Awareness regarding transfusion practices and iron chelation (n = 110)

Pre-BT Hb done	
Yes	42 (38.18%)
No	11 (10%)
Sometime	57 (51.81%)
Knowledge about BT-related reactions	
Yes	70 (63.36%)
No	40 (36.63%)
Knowledge regarding leukocyte filter	
Yes	9 (8.18%)
No	101 (91.8%)
Hepatitis B immunization	
Complete	47 (42.72%)
Incomplete	63 (57.27%)
Knowledge regarding bone marrow transplant	
Yes	9 (8.18%)
No	101 (91.81%)
Knowledge regarding splenectomy	
Yes	73 (66.36%)
No	37 (33.63%)
Knowledge about iron overload	
Yes	110 (100%)
No	0 (0%)
Iron chelation taken	
Yes	53 (48.18%)
No	57 (51.18%)
Which iron chelation	
Deferasirox	52 (47.27%)
Deferiprone	1 (0.9%)

BT, blood transfusion; Hb, hemoglobin.

Among study subjects, knowledge regarding the role of pre–blood transfusion Hb was present in 38.18%, whereas regarding blood transfusion reaction was present in 63.36%. Only 8.18% subjects were aware of leukocyte filter during transfusion and bone marrow transplantation. Most of the parents were aware about iron overload, but only 48.18% children were receiving iron chelation [Table 2].

Discussion

We report data regarding awareness among 110 parents of children with thalassemia major from tertiary care hospital in India. The findings of our study are important as awareness of parents is important for prevention as well as proper management of thalassemia major in their children.[8]

Thalassemia is a genetic disease and there is no sex predilection, but in this study, male (69%) outnumbered female. Similar observations were found in other studies from India, Bangladesh, and Pakistan.[9–11] On the contrary, one study from Pakistan found a significantly higher number of females with thalassemia major.[12] The higher number of males in our study may be because our study was hospital based and males seek more attention than females.

In our study, the level of education was low among study subjects and almost 20% parents of children with thalassemia were found to be illiterate. If parents of children with thalassemia would have higher level of education, they can better understand the importance of prenatal diagnosis.[13]

In this study we found that 12.7% children with thalassemia were born from consanguineous marriage. The frequency of consanguineous marriage in our study was much less as compared to that from a study conducted in Pakistan where cousin marriages were found to be high due to religious and cultural beliefs.[14,15] The prevalence of -thalassemia increased with consanguineous marriage as it is an autosomal recessive disorder. A study from Iran found significant association between β-thalassemia and first cousin marriage.[16] In our study, we also found 8.1% patients with thalassemia were having a family history of thalassemia and 3.3% were having siblings with thalassemia. This may be due to lack of proper knowledge about the disease and not opting for genetic testing during pregnancy.

Pre–blood transfusion Hb should be kept around 9–10 g/dl in patients with -thalassemia. If Hb of child is less, he or she will develop features of extramedullary hemopoisis and growth will adversely affected.[17] In our study, only about one-third patients regularly get their Hb levels assessed before transfusion, which may be due to lack of knowledge and counseling regarding the importance of pre–blood transfusion Hb.

Transfusion-related reaction such as febrile and urticarial rashes are well known in patients with thalassemia, which is why transfusion of washed, leukocyte-depleted RBC is recommended for all patients.[18] In our study, about two-third of patients knew about transfusion-related reactions and very few were aware of leukocyte filter. Hepatitis B vaccine should be given to all patients with thalassemia before transfusion to prevent transfusion-transmitted hepatitis B.[18] In our study, only 42% patients were completely immunized for hepatitis B. This may be because hepatitis B vaccine is not available with government supply in our hospital.

Stem cell transplantation reported 80%–95% event-free survival in thalassemia, but the most important strategy for prevention is prenatal diagnosis.[19] There was lack of knowledge regarding prenatal diagnosis among parents and affordability was less. Splenectomy should be considered in patients with poor growth and features of extramedullary hemopoiesis. It helps in decreasing the transfusion requirement by improving the Hb level.[20,21] About two-third parents in our study were aware of splenectomy.

In thalassemia, there is increased risk of iron overload due to regular transfusion and increased iron absorption from the gastrointestinal tract.[22,23] There are three major iron chelators present till date.[24,25] Parents should be provided information about iron chelation. In our study, although all parents knew about iron chelation, only about half were giving it to their children because of the cost and affordability. The government in our state has recently started providing iron chelation free of cost to all patients, so condition might improve in future. Most of our patients with thalassemia were using deferasirox as iron chelator because of better compliance. Moreover, its side effects are less and are also being provided free of cost in our hospital.

The major limitation of our study was that it was a hospital-based single-center cross-sectional study. The awareness of parents from other thalassemia centers and community could not be evaluated.

Conclusion

We conclude that awareness regarding thalassemia is inadequate in parents. Parents of children with thalassemia should be sensitized about the disease. There is a need for creating awareness among families with thalassemia and the general public through mass media, booklets, lectures, video, etc., so that the burden of thalassemia in the community can be reduced and children with thalassemia may have a better life.

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