Original Article

A cross-sectional study of awareness and practices regarding thalassemia among parents of thalassemic children

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Abstract

Background: This cross-sectional study was carried out in thalassemia ward of Rajindra Hospital, Patiala, among the parents of thalassemic children to determine awareness about side effects and complications of blood transfusion therapy, other treatment options, nature of disease, and food practices of transfusion-dependent patients. The study was carried out using a predesigned questionnaire and 118 parents participated in the study. About 50.84% patients belonged to the Sikh community, 45.76% patients practiced Hindu religion, and only 3.38% of the patients were Muslim. This study shows that 87.29% parents do not know how the disease is spread. About 55.93% have no knowledge about iron-containing food should not be included in the diet of transfusion-dependent patients. About 86.44% parents believed they had no role in transmission of the disease to their child, 79.66% parents do not understand the importance of screening before marriage, and 95.76% parents do not know about alternative treatment options. This study wants to shine light about the inadequate and superficial knowledge of thalassemia among general public and how awareness of the disease will bring down the incidence rates. **Aims:** The main objective of the study is to determine the degree of awareness of the disease, their knowledge of complications of blood transfusion therapy, and other treatment options among the parents of the children with thalassemia who are currently on blood transfusion therapy. **Subjects and Methods:** This cross-sectional study was conducted in thalassemia ward of Rajindra Hospital, Patiala from June 2018 to November 2018. After informed verbal consent was ensured, parents of the patients were interviewed using a questionnaire as the patients received blood transfusion. Questions include prevention, progression, cause, and spread, of the disease. The questions also include side effects and complications of blood transfusion therapy and other treatment options available. **Statistics Used:** Continuous variables were summarized as mean and standard deviation and categorical variables as proportion (%). Percentage and frequency was used wherever applicable. **Results:** Parents of about 118 patients were interviewed out of which 74.57% parents were illiterate and only 25.42% were literate. About 50.84% of the patients were Sikh, 45.76% were Hindu, and only 3.38% patients were Muslims. About 71.19% of the parents had no knowledge about the prevention of the disease, and 87.29% of the parents did not know mechanism of spread. Despite having transfusion-dependent children, only 44.07% of the parents restricted iron-containing food from the diet of their children. About 72.05% of the patients have inadequate information about risk of hepatitis B, hepatitis C, and HIV due to blood transfusions and only 21.29% of the patients understand the importance of hepatitis B vaccine. **Conclusion:** Awareness among both literate and illiterate parents was inadequate and sensitization among general public and parents of thalasemmic children should be initiated.

Keywords: Blood transfusion therapy, chelation therapy, iron overload, thalassemia, transfusion screening

Introduction

Thalassemia is the genetically transferred group of autosomal recessive diseases which are characterized by decreased production globin chain of hemoglobin.[1] The disease is fairly

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variable in presentation ranging from asymptomatic to in utero death.[2]

The burden of thalassemia across India is massive translating up to 35–45 million carriers and the prevalence being extremely variable across India about 0–10.5% predisposing some ethnic groups and subgroups more than others.[3] Screening in Ludhiana has shown very high carrier rate about 3.96%, making it one of the highest carrier rates in India.[3]

With so many carriers screening and awareness of thalassemia, it becomes pivotal in order to decrease its incidence across the country.

The current problem India faces is its high carrier number and the only way to bring down the crippling number of thalassemia cases is through increased awareness of the disease. This study was done in order to evaluate the awareness among the care takers of thalassemic children.

Subjects and Methods

The study was carried out in the thalassemia ward of Rajindra Hospital, Patiala for 6 months from June 2018 till November 2018.

The patients receive blood transfusion in the thalassemia ward and the parents of these patients were interviewed with the help of a predesigned questionnaire after the verbal consent had been ensured.

The questions involved the knowledge of the disease progression, complications, inheritance, consanguinity among parents, iron-containing food, screening before marriage or pregnancy, complications and side effects of blood transfusion therapy, and other treatment options.

All data were documented in excel sheets and then evaluated; the results were evaluated in the form of percentage and frequency.

Results

Parents of 118 patients were interviewed; mean age of these patients was about 9.4 ± 4.9 years with ages ranging from 0.5 to 18 years. About 35.5% of the patients were female and 64.4% patients were male showing a male predominance. About 50.84% of the patients belonged to the Sikh community, 45.76% patients were Hindu, and the rest 3.38% patients were Muslims.

Only quarter of the patients were literate (25.42%) and the rest were illiterate rounding up to 74.57% and nearly all marriages were a result of marrying outside the family (98.30%), while mere 1.69% marriages were a result of marriage between second cousins.

Out of all the parents only 28.81% were positive that the disease could be prevented, while the others either believed it could not be prevented (26.27%) or did not know if the disease could be prevented (44.92%).

Although 69.49% parents did not know how the disease is spread, and only 12.71% knew disease was spread genetically, other parents had variable beliefs with 11.02% believing that disease is spread through infectious agents, 5.08% believed that the disease is caused due to religious reasons, and 1.69% believed superstitious reasons for the spread of disease.

Most parents had no knowledge about different types of thalassemia (91.53%) and only 7.63% were certain that different types of thalassemia exist, whereas 0.85% parents believed that there are no different types of thalassemia.

Only 44.07% parents understood that iron-containing food was not good for children getting regular blood transfusion, while 46.61% did not have any clear knowledge if iron-containing food was good for their children getting blood transfusion or not, and 9.32% believed iron-containing food was good for their children.

The majority of parents (86.44%) believed that they had no role in transfer of disease to their children, whereas only 2.54% parents knew that their child inherited the genes from them and 11.02% had no knowledge if they transferred the gene to their child.

About 78.81% parents did not have any understanding if consanguinity has any role to play in the transmission of disease, and only 1.69% parents were certain that consanguinity increases the chances of disease in progeny, whereas 19.49% parents were certain that consanguinity has no role to play in incidence of disease.

Only 20.34% parents knew that thalassemia gene could be screened for before marriage and only 16.95% know that the disease can be screened for during pregnancy.

Most parents believe that blood transfusion is the only treatment about 77.12%, while only 6.78% knew about bone marrow transplantation therapy.

About half of the parents (57.63%) did not understand the importance of iron overload state and chelation therapy, and only 33.90% parents were knowledgeable about iron overload and chelation therapy accompanied with regular blood transfusion.

Almost more than half of the parents (55.93%) had no knowledge about blood transfusion reaction. And only 27.97% understood the risk of hepatitis B, hepatitis C, and HIV due to blood transfusions.

Only 21.19% patients were aware of importance of hepatitis B vaccination, while the rest either did not know (72.88%) or believed hepatitis B risk could not be decreased (5.93%).

Can this disease be prevented	Yes - 28.81%
	No - 26.27%
	Do not know - 44.92%
Belief about cause and spread of disease	Religious - 5.08%
	Genetic - 12.71%
	Infectious - 11.02%
	Superstitious - 1.69%
	Do not know - 69.49%
Is there any other type of thalassemia	Yes - 7.63%
	No - 0.85%
	Do not know - 91.53%
Is iron-containing food good for your child's health	Yes - 9.32%
	No - 44.07%
	Do not know - 46.61%
Do you have any role in transfer of disease to your child	Yes - 2.54%
	No -86.44%
	Do not know - 11.02%
Do you believe consanguinity has any role in its spread	Yes - 1.69%
	No - 19.49%
	Do not know - 78.81%
Can we detect thalassemia gene before marriage	Yes - 20.34%
	No - 17.80%
	Do not know - 61.86%
Can we detect this disease during pregnancy	Yes - 16.95%
	No - 6.78%
	Do not know - 76.27%
Is regular blood transfusion the only treatment	Yes - 77.12%
	No - 4.24%
	Do not know - 18.64%
Do you know about bone marrow transplantation therapy	Yes - 6.78%
	No - 50%
	Do not know - 43.22%
Do you know about iron overload state and chelation therapy	Yes - 33.90%
	No - 8.47%
	Do not know - 57.63%
Do you know about blood transfusion reactions	Yes - 35.59%
	No - 8.47%
	Do not know - 55.93%
Is there any risk of infections like hepatitis B, hepatitis C, and HIV, to your child, with repeated blood transfusions	Yes - 27.97%
	No - 10.17%
	Do not know - 61.88%
Is there any way by which we can decrease the chances of hepatitis B infection in your child	Yes - 21.19%
	No - 5.93%
	Do not know - 72.88%

Discussion

Thalassemia is a genetically transferred disease which is characterized by decreased in globin chain synthesis of hemoglobin leading to defective erythropoiesis and anemia.[1]

Pertaining to the large number of carriers in India and variable incidence across the nation,[3] the management of the disease and its prevention poses as the most important factor in a developing country. In India, every year 10,000 children are being born with thalassemia which approximately accounts for 10% of the total world incidence of thalassemia-affected children and 1 in 8 of thalassemia carriers lives in India.[4]

Even with patients with milder symptoms pertaining to thalassemia intermedia who do not require regular blood transfusion, there is increased risk of hypersplenism, such patients need to undergo surgical intervention and should be immunized against *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Neisseria meningitidis*. These patients may also have iron overload state despite not getting blood transfusions due to increased iron absorption from intestines and should be started on iron chelation therapy.[2]

Transfusion-related hemosiderosis and blood-borne infections pose a major threat to patients with thalassemia major who undergo regular blood transfusion therapy.[2] Studies have also shown that children with chronic diseases like thalassemia are more prone to emotional and psychiatric problems.[5]

Bone marrow transplantation is not a valid option due to financial constraints in developing country like India.

Henceforth, screening, prevention, and conservative therapy are the best choices. Awareness among parents about their carrier status and the treatment options and side effects of their thalassemic children are of utmost importance. Role of consanguineous marriages and resultant high rates of thalassemic burden on society needs to be addressed as well.[6]

Cost of prevention of the disease through screening is far less compared to the cost of the treatment of thalassemia and complications of a patient living for 50 years.[7] At present, the complete and only treatment available for thalassemia major is bone marrow transplantation, which only a few patients can afford. For supportive care and management of a child with thalassemia major, nearly 100,000–250,000 INR/year is required depending on the age and presence of complications.[8,9] Preventive measures such as mass awareness, screening, and disease education remain the best measures providing better financial, social, and health benefits.[10]

Repeated blood transfusions cause an increased risk of blood-borne infections and transfusion hemosiderosis. Lifelong transfusion therapy has considerable morbidity associated to it.[11] Every unit of blood given to a child increases the risk of these infections.[12]

Our present study indicates that about 72.03% parents had no idea about the risks of infection due to regular blood transfusions and only about 21.19% patients knew about hepatitis B vaccination.

These values are in accordance with other parts of the world[13] and reflect a very superficial understanding of the disease in the general public.

Despite the fact that frequent transfusions are tiresome, our study reflects that about 77.12% believe that transfusion is the only treatment option available and only about 6.78% patients

know about bone marrow transplantation. Hemosiderosis due to regular blood transfusion poses as a major threat in patients;[13] despite this our studies indicate that only 33.90% patients were aware of chelation therapy and its purpose, whereas 57.63% patients do not understand its importance. Only about 44.07% parents understand the importance of restricting food with high iron content and 46.61% parents do not know that food with high iron content should be restricted from the diet of children with thalassemia.

Our study shows that 86.44% of the parents believe that they had no role in transfer of the disease to their child and are unaware of their carrier status; this shows an underlying problem of lack of knowledge about the disease, its spread, and complications.

Our study concludes that the awareness of thalassemia among parents of children with thalassemia and general public is inadequate, which further poses a problem with prevention and decreasing incidence of the disease. Regular screening programs before marriage and antenatal, genetic counseling, mass media, national programs such as thalassemia status card especially in areas with high incidence, and collection of data of incidence and carriers need to be undertaken by the government and medical health-care professionals to bring down the burden of the disease on health services in India.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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