Research Article

A study on transfusion transmitted infections (TTIs), transfusion-related complications, and quality of life among the beta-thalassemia major patients in Jamnagar district

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Abstract

Background: Thalassemia is considered the most common genetic disorder worldwide. The condition affects economical and psychosocial quality of life of whole family broadly. It is a serious public health issue throughout Indian subcontinent and southeast Asia.

Objective: (1) To study the prevalence of transfusion transmitted infections (TTIs) in patients of beta-thalassemia major who are on transfusion therapy, (2) to assess the complications of iron overload and complications owing to chelation therapy, and (3) to study psychosocial and economic burden on patients' family.

Materials and Methods: This cross-sectional study was conducted in Thalassemia ward of Pediatric Department from December 2010 to December 2011. All the patients of beta thalassemia major on blood transfusion therapy were included in the study. Preformed, pretested questionnaire was used to interview the patients and their caretakers**.** Data entry and analysis were done using Microsoft Excel 2007 and SPSS software, version 17.

Result: Prevalence of human immunodeficiency virus (HIV), hepatitis B virus (HBV), and hepatitis C virus (HCV) was 3.95%, 2.25%, and 2.25%, respectively. Six in every 10 patients ever have experienced from blood transfusion reactions. Almost 40% patients had left the studies owing to illness or they have not gone to school ever.

Conclusion: Even after introduction of sensitive screening test, prevalence of TTIs is more in patients than general population. Quality of life is affected economically and psychosocially.

KEY WORDS: Transfusion transmitted infections (TTIs), consanguinity, iron overload, sickness absenteeism, splenectomy

Introduction

Beta-thalassemia major is an autosomal recessive disease characterized by severe hemolysis. Thalassemia is regarded as the most common genetic disorder globally. About 3% of people worldwide carry beta-thalassemia gene. It is a critical public health problem throughout the Mediterranean region, the Middle East, the Indian subcontinent, and Southeast Asia.[1,2]

The study area is an area with high prevalence of beta-thalassemia major patients. Thalassemia major that is treated incompletely is a possibly deadly disorder.[3]

In the last 30 years, introduction of regular blood transfusion therapy and effective iron chelating therapy with desferrioxamine have markedly improved the life expectancy of beta-thalassemia patients.[4–6] Transfusion-dependent patients are more prone to acquiring various transfusion transmitted infections (TTIs) such as hepatitis B virus (HBV), hepatitis C virus (HCV), and human immunodeficiency virus (HIV).[7]

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The grouping of transfusion and chelation therapy has considerably prolonged the life expectancy of these patients, thus transforming thalassemia from a rapidly lethal illness of childhood to a chronic illness compatible with a prolonged life. On the other hand, frequent blood transfusions leading to iron overload and the chronic nature of the disease have contributed to a whole new spectrum of complications in adolescents and young adults presenting thalassemia major.[8]

The organ dysfunctions owing to iron overload following repeated Blood Transfusion (BTs) are also a major problem. The condition affects the economical and psychosocial quality of life of the whole family broadly.

With this background, this study has been conducted to find out the prevalence of TTIs, transfusion-related complications and quality of life of beta-thalassemia major patients.

Materials and Methods

A cross-sectional study was conducted in Thalassemia ward of Pediatric Department from September 2010 to February 2011. Patients of beta-thalassemia major on blood transfusion therapy were included in the study. A total of 210 patients were on blood transfusion therapy in Thalassemia ward. All patients included in the study were on blood transfusion for more than 1 year. So, among them, 177 beta-thalassemia major patients were interviewed. Others refused to participate in the study and there were drop outs. Preformed, pretested questionnaire was used to interview the patients and their caretakers. Details regarding the duration of blood transfusion, TTIs, any complication ever occurred owing to blood transfusion, stress to caretakers, economical burden of treatment on family, etc. were asked. To see the quality of life of patient, effect of disease on patients' education had been asked in detail. Detailed clinical examination, including anthropometric measurements and other clinical parameters relevant to the study, was also carried out. Blood of the patients who had not been tested for their HIV, hepatitis B surface antigen (HBsAg), and HCV status over the past 1 year was sent for the same. Results of the laboratory tests and the clinical examination were subsequently analyzed. A verbal consent was taken from the caretakers of the patients. Ethical clearance from the ethical committee was taken before conducting the study.

Data entry and analysis were done using Microsoft Excel 2007 and SPSS software, version 17. The χ^2-test was used to see the statistical significance of the test at 95% confidence interval at 5% level of significance.

Result

Table 1 shows that the study group included 62.14% male and only 37.86% female subjects. Almost one-third (59.33%) of the children were aged 5–15 years. As the age increases, the proportion of children decreases. Only 6.78% children were in the age group of 20–26 years.

Figure 1 shows that one-fourth (25.43%) of the patients belonged to Vankar community. Second large group belonged to Muslim community (21.47%), followed by Ahir (16.39%) and Bhanushali communities (6.78).

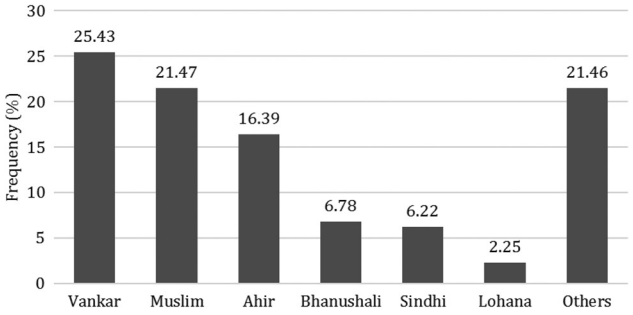

Figure 1: Distribution of patients according to their caste.

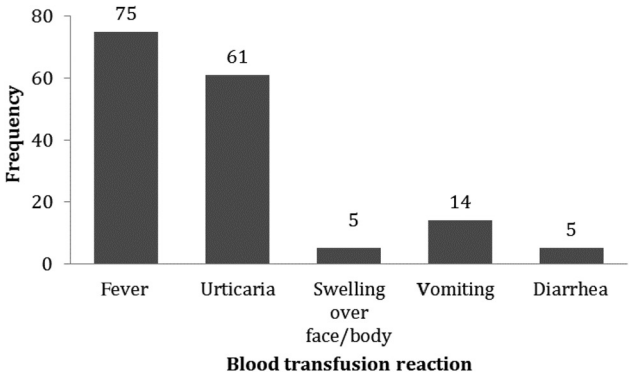

Figure 2: Number of patients who have ever been affected by blood transfusion reaction* (*n* = yes, 112; no, 65).

*Multiple responses.

About 42.37% patients showed history of consanguineous marriages of their parents and 22.03% patients with siblings of beta-thalassemia major.

Figure 2 shows that almost two-thirds (63.28%) of the children have experienced any kind of blood transfusion reaction during the treatment period. Among the patients who experienced reactions, 67% presented fever with rigors, 55% urticaria, and others presented swelling, vomiting, or diarrhea.

Table 2 shows that, among the 177 patients, 8.47% were affected from TTIs. Among them, HIV contributed 3.95% while 2.26% each of HBV and HCV.

Table 3 shows that 21.46% patients showed cardiac complications, 13% endocrinal, and 19.77% have undergone splenectomy. Table 3 also shows clearly that as the age increases, the complications also increase (linear association). Maximum proportion of complication was seen among the patients in the age group of 20–26 years.

Table 4 shows that there is a statistically significant association between consumption of chelating therapy and complications owing to iron overload. Those who were on chelating therapy showed only 1.13% complications when compared with those who were not taking chelating therapy (36.16%).

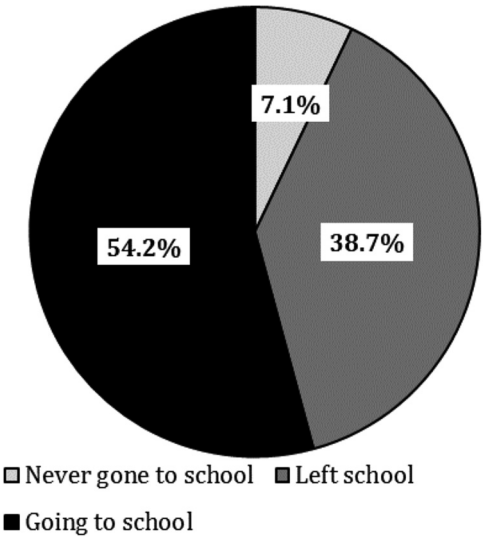

Figure 3: Patient's status of schooling (*N* = 142, age>5 years).

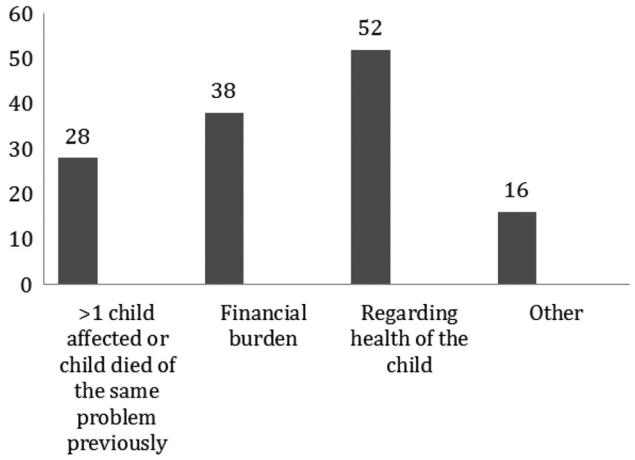

Figure 4: Reasons for stress among the parents of patients (*n* = 131)*.

*Multiple responses.

The average cost of treatment per patient per month was Rs. 4,090, which included direct medical cost (drugs, investigations, etc.), direct nonmedical cost (travelling, food, and staying cost), and hidden cost (loss of daily wages of parents/caretaker). This amount is an economic burden on the family, which affects the quality of life of people.

Table 1: Age- and sex-wise distribution of patients

Age group (years)	Male (%)	Female (%)	Total (%)
1–5	21 (11.87)	14 (7.90)	35 (19.77)
5–10	35 (19.78)	17 (9.60)	52 (29.38)
10–15	32 (18.07)	21 (11.88)	53 (29.95)
15–20	18 (10.17)	7 (3.96)	25 (14.13)
20–26	4 (2.26)	8 (4.52)	12 (6.78)
Total	110 (62.14)	67 (37.86)	177 (100)

Table 2: Presence of TTIs among the patients

TTI	Male	Female	Total (%)
HIV	2	5	7 (3.95)
HBV	3	1	4 (2.26)
HCV	3	1	4 (2.26)
Total	8 (4.52)	7 (3.95)	15 (8.47)

Table 3: Complications owing to iron overload among the beta-thalassemia major patients*

Age group (years)	Cardiovascular complications (%)	Endocrinal complications (%)	Splenectomized (%)
1–5 (*n* = 35)	1 (2.85)	0	0
5–10 (*n* = 52)	3 (5.76)	1 (1.92)	4 (7.69)
10–15 (*n* = 53)	18 (33.96)	8 (15.1)	14 (26.41)
15–20 (*n* = 25)	9 (36)	9 (36)	12 (48)
20–26 (*n* = 12)	7 (58.33)	5 (41.46)	5 (41.46)
Total (*n* = 177)	38 (21.46)	23 (13)	35 (19.77)

*Multiple responses.

Table 4: Association between chelating therapy and complications owing to iron overload

Taking chelating agent	Presence of complication		Total
	Yes	No	
Yes	2 (1.13)	13 (7.34)	15 (8.47)
No	64 (36.16)	98 (55.37)	162 (91.53)
Total	66 (37.29)	111 (62.71)	*N* = 177 (100)

$\chi^2 = 4.022$, $p = 0.045$.

Figure 3 shows that only half (54.2%) of the children are attending the school, while 38.7% have left the school owing to the disease. About 7.1% children have never gone to school.

Many left studies owing to ill health and those who were studying showed an average sickness absenteeism of 3 days/month. All the children who are going to school have sickness absenteeism: six in every 10, average days. Quality of life is affected as the disease is chronic and affecting the education of children.

Figure 4 shows that 74.01% parents exhibited stress owing to the disease in their children. The most common reason for stress among the parents was deteriorated health of the child (39.7%). Second large reason was financial burden (29%) on the family owing to expenses on the treatment of the affected child/children. About 21.4% parents were stressed owing to more than one child affected in the family or the death of child.

Discussion

The group of beta-thalassemia major patients studied included elder patients aged 26 years. This shows that, even with the blood transfusion and the chelation therapy, longetivity of these patients is at question.

Owing to consanguineous marriages, some castes are showing higher prevalence of the disease. Before few years, Sindhis and Lohanas were the communities with the highest prevalence of the disease in our study area, but our study revealed that both these castes presented 10 times less prevalence when compared with Vankars and Muslims. These may be owing to awareness regarding the disease and active preventive measures taken by these communities.

The prevalence of TTIs such as HCV, HIV, and HBsAg was 45%, 2%, and 2%, respectively in a study conducted by Neeraj et al.,[9] which shows quite high prevalence of HCV when compared with our study. While in a study by Ocak et al.[7] showed prevalence of HbSAg, HCV, and HIV to be 0.75%, 4.5%, and 0%, respectively, which is quite less than our study.

The anterior pituitary is particularly sensitive to iron overload, which disrupts hormonal secretion, leading to gonadal dysfunction. Thus, these patients show decreased gonadotropin reserves when compared with those of normal controls. This usually presents as delayed or absent puberty, primary or secondary amenorrhea, menstrual irregularities, and fertility problems later in life.[8]

In a study conducted by Li et al, of 138 patients evaluable for puberty, delayed puberty was observed in 53 (38.4%). Among 48 females aged older than 16 years, 13 (27.1%) did not have menarche, and three girls had menarche but subsequently developed secondary amenorrhea.[10] In this study also, 13% patients were showing delayed puberty, primary amenorrhea, and other endocrinal problems.

Cardiomyopathy was present in 35 (15.1%) patients with the median age of onset at 16 years. It was more common in older patients, and the median age of developing these endocrine complications was around 16 years, which is almost same as this study.[10]

Cardiac disorders and, most notably, left-sided heart failure are responsible for more than half of the deaths in these patients and are thus the main determinants of survival.[11] With the advent of chelation therapy, there has been a decline in the severity of these complications, but they still remain the leading cause of morbidity and mortality in these patients.

A high incidence of multiendocrine dysfunction has been reported in children, adolescents, and young adults with thalassemia major.[12]

The major step forward in improving survival and reducing complications was the introduction, in the 1960s, of the chelating agent deferoxamine, first as an intramuscular injection and later as a subcutaneous infusion. Two oral chelators, deferiprone and deferasirox, have more recently become available, making therapy easier and more efficacious. Compliance, although improved by the switch to oral therapy, still represents a problem and is the major obstacle to effective prevention, or limitation, of iron overload.[13]

In this study also, those who were on chelation therapy (1.13%) had quite less complications when compared with those who did not take any chelating therapy (36.16%).

Quality of life of these patients is affected in terms of deteriorated health, dissatisfaction toward physical appearance, effect on education, and economic burden of treatment on family.

Conclusion

Instead of sensitive screening of blood before transfusion, blood transfusion reactions and TTIs are more prevalent in beta-thalassemia patients than general population. Chelation therapy has helped in decreasing the transfusion-related complications and to increase the longetivity of life. Premarital testing for thalassemia status should be done. Screening of all the registered pregnant women for thalassemia status must be done. Parents should be counseled for prenatal testing of thalassemia. Financial assistance should be made available for chelation therapy for poor patients. Every patient should be vaccinated with hepatitis B vaccine. More sensitive screening tests should be used for HIV, HBV, and HCV. Proper precautions for safe transfusion practices should be adhered to.

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