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Original Article

ASSESSMENT OF QUALITY OF LIFE, COMPLICATIONS AND POST-TRANSFUSION ADVERSE REACTIONS IN THALASSEMIA PEDIATRIC IN TERTIARY CARE HOSPITAL

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ABSTRACT

Objective: To assess the quality of life, investigate complications and adverse transfusion reactions post blood transfusion in thalassemia pediatric.

Methods: A prospective, observational study was conducted over a period of six months in the pediatric department of Bharati hospital and research center, Pune. Thalassemia patient profile form was used for collecting demographic details, to record adverse transfusion reactions and complications. Using PedsQL™ (Pediatric Quality of Life Inventory) questionnaire thalassemia patients, and the control group was interviewed to assess their quality of life.

Results: Out of 39 patients who were enrolled, 28 patients were above the age of 3 y; their overall quality of life score was significantly lower in thalassemia pediatric than healthy control. School function was a most affected domain. 53.84% had post-transfusion fever, 5.1% of respiratory distress while 41.06% had myalgia, swelling at IV site, headache, and vomiting. Out of 39 patients, 24 patients examined for complications; 33.34% had hepatosplenomegaly, 25% had infections, 16.66% had splenomegaly, 13.88% had cardiac complications, 8.34% had bone deformity, and 2.78% had growth failure.

Conclusion: It was found that there was a significant decrease in the quality of life domains in thalassemia pediatric compared to control group. School domain was most affected. Hepatosplenomegaly is one of the leading complications observed in this study. Most common transfusion-related reaction was post-transfusion fever. Public awareness and preventive measures such as pre-marital and prenatal testing should be done to eliminate this fatal disease.

Keywords: Thalassemia, Quality of life, PedsQL™, Adverse transfusion reactions, Complication in Thalassemia.

INTRODUCTION

Thalassemia is one of the major hemoglobinopathies among the population all around the world. It is a single-gene hereditary disorder in a human. It is caused due to decreased or absent amount of globin chain of hemoglobin [1]. It occurs when there is an abnormality or mutation in one of the genes involved in hemoglobin production, which is inherited from both parents [2]. The average incidence of ß-thalassemia trait in India is 3.3% with 1–2 per 1,000 couples being at risk of having an affected offspring each year. The incidence of β-thalassemia in different regions of India varies from 3% to 17%, with a mean prevalence of 4% [3]. It is very high among certain communities such as Sindhis and Punjabis from Northern India, Bhanushalis, Kutchis, Lohanas from Gujarat, Mahars, Neobuddhists, Kolis and Agris from Maharashtra, Gowdas and Lingayats from Karnataka etc. and certain tribes in the northern, western and eastern parts, with lower incidence in the southern tribes [4]. Patients with beta thalassemia major usually become symptomatic by about 4–6 mo of age and without transfusion support, progressive anemia causes decreased growth and development, heart failure, and eventually premature death. Without transfusions, very few patients survive beyond 5 y of age [5]. The management of thalassemia includes regular blood transfusion, iron chelation therapy, and appropriate management of co-morbidities [6]. The more thalassemia major develops into a chronic disease with an expanding life expectancy, the more patient quality of life is becoming an important dimension of care. It is a potential life-threatening condition causing a substantial disturbance in education and social activities [7]. Based on literature survey, it was observed that few studies were conducted on the effect of thalassemia on the quality of life in patients and also fewer studies were available to address, document adverse transfusion reactions and complications associated with thalassemia in India. The positive impact of treatment diminishes, especially in terms of health-related quality of life if they interfere with daily activities or are less tolerable e. g. regular blood transfusion that requires frequent hospital visits. As children are less able to voice their concern and are more vulnerable than adults, the assessment of health-related quality of life in children is essential for the provision of proper care, since it helps in identifying the impact of disease and treatment from the children's perspective. Even if in recent years the innovative research strategies for thalassemia major have greatly increased life expectancy which varies between 25-55 y, the fear of the disease itself and possible complications of invasive medical procedures, still affect the quality of life of patients. Also, it is important to assess long-term disease complication and adverse transfusion reactions due to blood transfusions, which will further help in improving the quality of life.

Therefore, the objective of the present study is to assess the quality of life in beta thalassemia pediatric using PedsQL™ version 4.0 (Pediatric Quality of Life Inventory) questionnaires, to identify and assess the types of complications and adverse transfusion reactions.

MATERIALS AND METHODS

The study was carried out for 6 mo duration at Bharati Medical Hospital, and Research Centre, Dhankawadi, Pune and an Institutional Ethical Committee clearance was obtained with an ethics vide letter no. BVDU/MC/76. The study was conducted among children and adolescent with beta thalassemia major who received blood transfusion regularly at Bharati hospital. A random sample of healthy school going children of age 2-18 y was selected as the control group for comparing the quality of life.

An information sheet was explained to parents. Assent was taken from parents of both patients and control group who were enrolled in the study. Patient's demographic data and clinical data were noted down in a specially designed thalassemia patient profile form.

Patients and their parents were interviewed about the patient's quality of life and if they experience any adverse reactions post blood transfusions when they visit for subsequent blood transfusion. The interview was carried out by administering PedsQL™ version 4.0 questionnaires for assessing their quality of life. The questionnaire had choices "never", "almost never", "sometimes", "often', and "almost always", with corresponding scores of 100, 75, 50, 25, and 0. Each patient was given a score from 0-100, with higher scores indicating better health or a higher level of function. At the end of the study, the questionnaire was analyzed, and adverse transfusion reactions were documented. The same procedure was followed to interview the children from the control about their quality of life.

Data was analyzed by Microsoft Excel 2007 result was calculated in terms of percentage, mean, standard deviation (SD) and was also used for graphical representation. The quality of life comparison between thalassemia pediatrics and control group was analyzed using one-way ANOVA for which p>0.05 shows a significant difference between thalassemia pediatrics and control group.

RESULTS

In a study period of six months total number of patients who were screened for beta thalassemia major were 48 patients, out of which 39 patients were enrolled for the study based on inclusion criteria, 2 patients were excluded as they did not meet the inclusion criteria, while 7 patients left blood transfusion follow-up from Bharati Hospital, as it was not feasible for them to continue the blood transfusion follow-up in the hospital.

Among 39 patients, 28 were male and 11 were female with the mean age of 6 y±4.32. 14 (35.90%) patients belonged to the age group of 1-3 y, followed by 9 (23.08%) each in age group of 4-6 y and 7-9 y respectively while 7 (17.94%) were in the age group of 10-18years. It was observed that 9 (23.07%) were in pre-primary, 10 (25.65%) in primary, 6 (15.38%) in secondary, 11 (28.21%) were not attending school as they were under the age of 3 y, while 3 (7.69%) left school as they were not comfortable with the school environment. To evaluate the quality of life in patients with beta thalassemia major, a control group of 28 was introduced, who were in the age range of 1–18 y. Among them, 18 (64.28%) were male and 10 (35.72%) were female. 14 (50%) were in the age group of 4-6 y. Children from the control group were randomly selected among school going, children. Most of them were from primary education level (46.43%) (table 1).

From the data collected it was found that most of the patients had blood group B+ve 14 (35.90%), followed by O+ve 10 (25.65%), A+ve 8 (20.52%), AB+ve 4 (10.25%), B-ve 2 (5.12%) and AB-ve 1 (2.56%) (table 2).

In the present study, it was observed that 20 (51.28%) had a consanguineous marriage while 19 (48.72%) had non-consanguineous marriage (fig. 1).

It was observed that 23 patients (58.98%) were diagnosed among the age group of 3-6 mo, 6 (15.39%) among the age group of 7-9 mo, 5 (12.82%) with age greater than 1 y, followed by patients with age group of 10-12 mo 4 (10.25%), 1 patient of age less than 3 mo (2.56%) (fig. 2).

From the data collected it was observed that patients complained of pallor, fever, cough, coryza, weakness at the time of diagnosis. Other symptoms which were seen are chills, irritability, abdominal pain, vomiting, decreased oral intake, edema, breathlessness, diarrhea, headache, body ache, joint pain (table 3).

It was found that about 21 patients (53.84%) had Hb between ranges of 2.0-4.9 g/dl followed by 15 patients (38.46%) had Hb between the range of 5.0-7.9 g/dl and 3 patients (7.70%) had Hb less than 8.0 g/dl at the age of diagnosis. (fig. 3).

The data showed that 9 patients (23.10%) of thalassemia children had parents who were both carriers. In 1 patient (2.55%) only 1 of the parent was a carrier and in 29 (74.35%) parents were not screened for beta thalassemia (table 4).

The data collected showed that among 39 patients, 15 (38.46%) had no siblings while 13 (33.34%) patients had siblings who were not affected while 5 patients (28.20%) had siblings who were affected with beta thalassemia (fig. 4).

Comparison data showed that there was a significant difference of quality of life between thalassemia pediatrics and control group. Assessments of four quality of life domains in thalassemia pediatrics vs. control group showed the following: physical function 55.45 vs. 89.23 (p value= 0.9930), emotional function 62.32 vs. 92.85 (0.9752), social function 62.14 vs. 92.85 (p value= 0.9332) and school function 46.42 vs. 85.53 (p value= 0.9992). One way ANOVA test was used to compare thalassemia pediatrics and control group and it was found to be p>0.05. This shows that there was a significant difference between thalassemia pediatrics and control group.

In the present study, it was observed that overall the quality of life score was less in thalassemia pediatrics indicating that their quality of life was affected due to the disease and among all quality of life domains; school function was the most affected domain (fig. 5).

From the data collected it was found that most common blood transfusion reactions were fever, respiratory distress while other blood transfusion reactions which didn't occur commonly were headache, vomiting, swelling at injection site, tachypnea (table 5).

It was observed that 20 patients (51.28 %) came for blood transfusion when their pre-transfusion Hb was in between 5-7.9 g/dl while there were 13 patients (33.34 %) whose pre-transfusion Hb was in between 8-10.9 g/dl, 6 patients (15.38 %) had pre-transfusion Hb less than 5 g/dl (fig. 6).

The present study showed that 27 patients (69.23 %) came for blood transfusion once a month that is they were regular with their transfusions, 8 patients (20.15 %) came within 15 d, whereas 4 patients (10.26 %) were irregular with their blood transfusions (table 6).

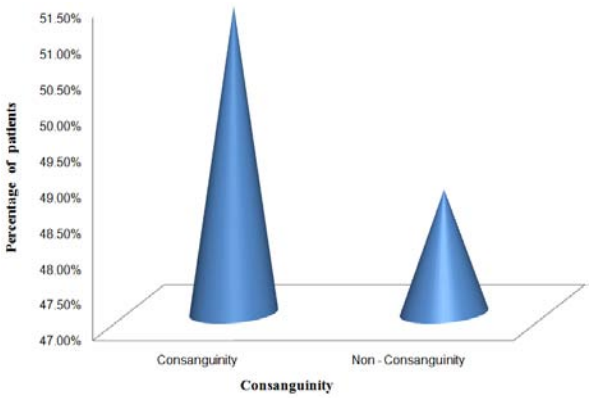

Fig. 1: Consanguinity

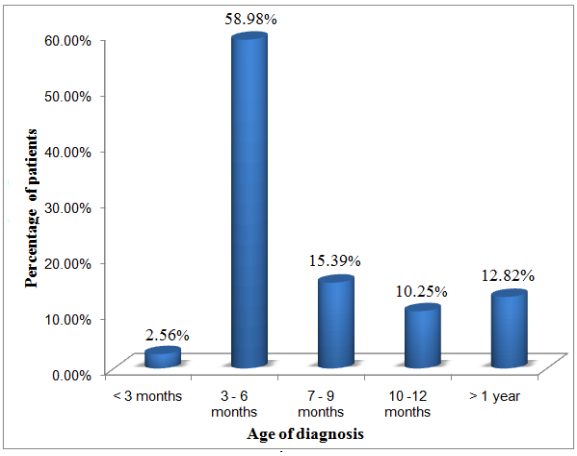

Fig. 2: Age of diagnosis

In the present study it was found that among 39 patients; 11 (28.20%) had serum ferritin levels more than 1000 ng/ml, 3 patients (7.70%) showed less than 1000 ng/ml and for 25 patients (64.10%) reports regarding serum ferritin level were not available (fig. 7).

From the collected data it was observed that 18 patients (46.53%) were started on chelation therapy based on their serum ferritin level to avoid iron overload in patients caused due to blood transfusions (table 7).

The collected data showed that 21 patients (53.85 %) were prescribed with supportive therapy like calcium supplement and folic acid while 18 patients (46.15 %) were not on any supportive therapy (table 8).

Chronic blood transfusion can lead to many complications which require regular monitoring. In this study, it was observed that only 24 patients (61.54%) had their regular check-up was done (fig. 8).

Among 24 patients who had been examined for complications, it was found that 12 patients (33.34%) had hepatosplenomegaly, followed by infection in 9 patients (25%), splenomegaly in 6 patients (16.66%), cardiac complications in 5 patients (13.88%), bone deformity in 3 patients (8.34%) and growth failure in 1 patient (2.78%). (fig. 9).

Table 1: Demographic characteristics of beta thalassemia major patients and control group

Characteristics	Thalassemia patients	Percentage (%)	Control N= 28	Percentage (%)
Gender				
Male	28	71.80	18	64.28
Female	11	28.20	10	35.72
Age (y)				
1-3	14	35.90	5	17.86
4-6	9	23.08	14	50
7-9	9	23.08	2	7.14
10-12	2	5.13	2	7.14
13-15	4	10.25	4	14.28
16-18	1	2.56	1	3.58
Education levels of children				
Preprimary	9	23.07	9	32.15
Primary	10	25.65	13	46.43
Secondary	6	15.38	6	21.42
Left school	3	7.69	0	00.00
Not attending school yet	11	28.21	0	00.00

Table 2: Blood group distribution in thalassemia patients

Blood group	Number of patients	Percentage (%)
A+	8	20.52
B+	14	35.90
AB+	4	10.25
O+	10	25.65
B-	2	5.12
AB-	1	2.56

Table 3: Symptoms during diagnosis

Symptoms	Number of patients
Pallor	28
Fever	25
Cough and Coryza	15
Weakness	14
Chills	3
Irritability	3
Abdominal pain	3
Vomiting	3
Decreased oral intake	3
Diarrhea	3
Edema	2
Breathlessness	2
Headache	1
Body ache	1
Joint pain	1
Others	2

Table 4: Parent as carrier

Parent as carrier	Parents screened	Percentage (%)
Not screened	29	74.35
2 Carrier	9	23.10
1 Carrier	1	2.55

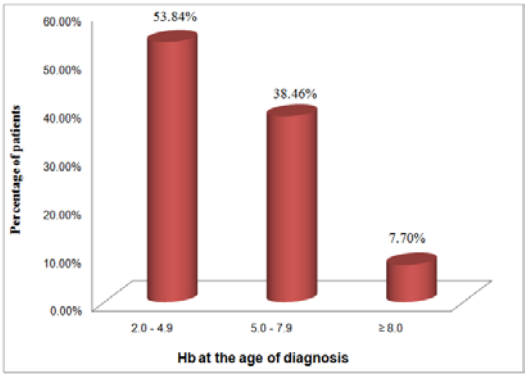

Fig. 3:-Hb at the age of diagnosis

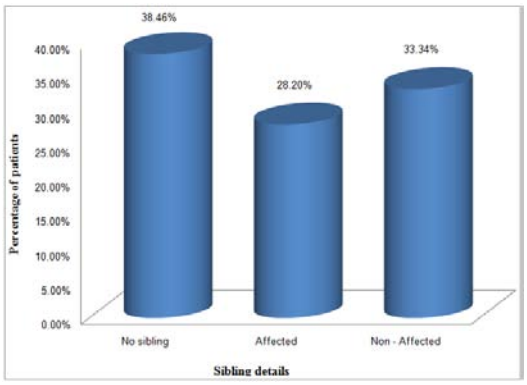

Fig. 4: Siblings details

Quality of life of thalassemia pediatrics

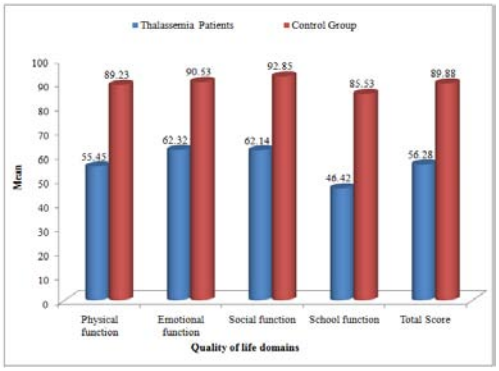

Fig. 5: Differences in quality of life between thalassemia pediatrics and control group

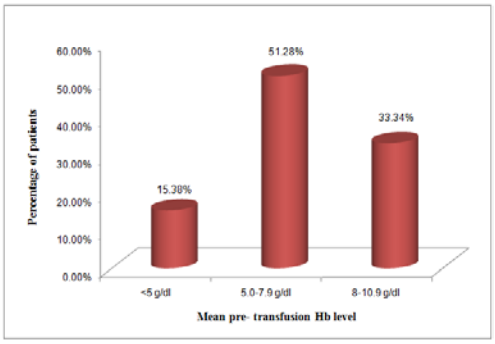

Fig. 6: Mean pre-transfusion Hb level

Blood transfusion details of thalassemia pediatrics

Table 5: Transfusion details of thalassemia pediatrics for duration of six months

Patient no.	Mean pre-transfusion Hb (g/dl)	Number of transfusion in six months	Frequency of transfusion (in a month)	Transfused blood volume (ml)	Number of bags given	Reactions during transfusion	Frequency of reactions
1.	8	4	Once	540	1	Fever	1
2.	8.4	6	Once	1476	1	Fever	1
3.	8.4	6	Once	2346	1	No	-
4.	8.2	11	Twice	5680	2	Fever,	4
5.	8.4	5	Once	746	1	No	-
6.	7.5	6	Once	2872	1	Fever, Post RD, myalgia, Abd. Dist	2
7.	7.3	9	Twice	3420	1	Fever	4
8.	4.3	4	Once	1460	1	Fever, lower back pain	1
9.	6.9	5	Once	1435	1	Fever	3
10.	5.8	6	Once	5190	2	Headache, fever	2
11.	4.3	4	Once	2360	1	Fever, swelling at injection site	1
12.	6.2	6	Once	3194	1	Fever	1
13.	5.1	6	Once	3780	1	Fever	1
14.	4.6	8	Twice	3550	2	Fever and shivering	2
15.	9.13	5	Once	798	1	No	-
16.	8.2	8	Twice	4831	2	No	-
17.	4.54	5	Once	2544	1	No	-
18.	7.81	5	Once	1685	1	Fever	1
19.	9.18	7	Once	586	1	Fever	2
20.	7.9	7	Once	1668	1	No	-
21.	5.1	5	Once	3240	2	Vomiting	1
22.	7.6	5	Once	2430	1	Fever	1
23.		EXCLUDED					
24.*	5	2	Once	104	1	Fever	1

25.	8	8	Twice	2710	1	No	-
26.	7.81	6	Once	1729	1	Fever	1
27.	7.91	3	Every 2 mo	3640	2	No	-
28.	7.1	11	Twice	1540	1	No	-
29.	9.01	6	Once	1030	1	No	-
30.	6.13	6	Once	900	1	No	-
31.	6.96	5	Once	1602	1	Fever, shivering	2
32.	8.51	8	Once	2065	1	No	-
33.	8.4	4	Once	498	1	Fever	1
34.	5.87	4	Once	1066	2	No	-
35.	7.76	3	Once	569	1	No	-
36.	9.06	6	Once	949	1	No	-
37.	4.04	6	Twice	1562	1	Tachypnea, RD	2
38.		EXCLUDED					
39.*	6.5	2	Once	120	1	Fever	1
40.*	8.8	4	Once	428	1	No	-
41.*	3.8	2	Once	570	2	Fever	1

* Newly diagnosed, RD-Respiratory distress, Abd. dist.-Abdominal distension

Table 6: Frequency of blood transfusion

Frequency of blood transfusion	Number of patients	Percentage (%)
1 mo	27	69.23
15 d	8	20.51
Irregular	4	10.26

Table 7: Chelation therapy in thalassemia patients

Chelation therapy	Started	Not started
Number of patients	18	21
Percentage	46.15%	53.85%

Supportive therapy in thalassemia pediatrics

Table 8: Supportive therapy in thalassemia pediatrics

Supportive therapy	Number of patients	Percentage (%)
Yes	21	53.85
No	18	46.15

Management of iron overload due to blood transfusion

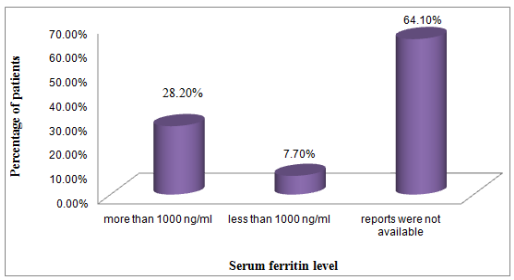

Fig. 7: Serum ferritin level in thalassemia patients

Complications associated with beta thalassemia major

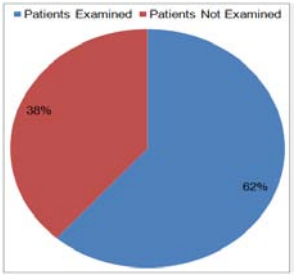

Fig. 8: Patients examined for complications

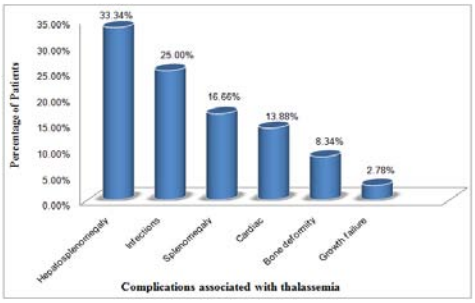

Fig. 9: Complications associated with thalassemia

DISCUSSION

Thalassemia major is characterized by inadequate globin chain synthesis, leading to ineffective erythropoiesis, severe anemia and chronic disease in adulthood.

The present study conducted for duration of 6 mo included 39 Beta-thalassemia patients selected from Pediatric ward. Among which 71.80 % were male and 28.20 % were female which is comparable to other studies which showed a higher occurrence among males than in females [8]. On the contrary, others study showed there was non-statistically significant difference observed between male and female thalassemia patients [9]. This difference in thalassemia patients (males more affected than females) is noteworthy and deserves further investigation considering thalassemia as a single gene disease transmitted by recessive mode of inheritance [10].

In this study frequency of patients with blood group, B+ve were higher (35.90 %) followed by patients with O+ve (25.65 %) while A+ve consisted of 20.52 % and AB+ve was 10.25 %. A similar study carried on beta-thalassemia patients showed a higher number of patients with blood group O+ve and lower number of patients with blood group B+ve. There was no significant difference found in blood group A+ve and AB+ve. This was in contrast with study conducted by Sharaf Kh H *et al.* which showed there was no effect of blood group on patients with Beta-thalassemia major [11].

The study reported that highest percentage of Beta-thalassemia major patients (50%) was in between the age grouped4-6 y. This result was comparable to the study conducted on the prevalence of beta-thalassemia by Qurat-ul-Ain *et al.* [10]. There is no correlation between age group and occurrence of Beta thalassemia major. Also, age group can vary from hospital to hospital.

Consanguinity seems to play an essential role in causing Beta thalassemia major in the patients; in this study, it was observed that 51.28% of patients were the product of the consanguineous marriage. A similar study conducted by Dorouddhi M *et al.* reported 49.5% of patients were the product of consanguineous marriage [9]. There was one more study carried by Adaay *et al.* which showed 71.4% of the patients were the product of consanguineous marriage [8]. The reason for same could be that the autosomal recessive trait will be more common in progeny of consanguineous parents since they have greater chance of inheriting identical copies of the mutant gene. Many studies have shown that consanguineous marriage plays a significant role in the expression of beta thalassemia in patients.

In this study, it was observed that most of the patients (58.98%) were diagnosed with beta thalassemia major in the age range between 3-6 mo. The above results are consistent with study conducted by Kirti Grow *et al.* which reported that 51% of patients were diagnosed with beta thalassemia major before 8 mo of age [12]. A study conducted by Masyitah Sri Wahyuni *et al.* reported that more than half of the subjects were diagnosed with thalassemia at the age of less than 2 y, although they had been anemic before the age of 2 [13].

Symptoms of fever, pallor, cough, coryza, weakness were observed during diagnosis in our study. Similar clinical features of beta thalassemia major were reported in various studies conducted by P Sandhya Rani *et al.* and G Shanthi *et al.* respectively [3, 14]. Other symptoms which were not commonly seen were chills, irritability, abdominal pain, vomiting, decreased oral intake, diarrhea, edema, breathlessness, headache, body ache, joint pain were observed in this study. Symptoms like fatigue, facial bone deformities, protruding abdomen, yellowish discoloration of the skin and dark urine are among the other symptoms reported in a study [3]. Symptoms may depend upon the severity of the affected hemoglobin chain.

It was found that of all thalassemic children's parents, 23.10% had both parents as carriers, 1% had 1 parent as a carrier and in 74.35% of parents was not screened for beta thalassemia major. Similar results were found in a study conducted by Masyitah Sri Wahyuni *et al.* [13]. This could be because the marriage of two carrier parents results in 25% chance of producing thalassemic offspring, 50% chance of producing a carrier who can pass on the disease if he/she marries another carrier and a 25% chance of normal offspring.

In this study incidence of beta thalassemia major among siblings was 28.20%. A similar study was carried out by Tazeen Majeed *et al.* which reported the incidence of beta thalassemia trait among 58% of siblings [15]. A study conducted in Karachi reported 62.2% siblings as beta thalassemia carriers in immediate family members of patients [16].

The negative impact of the disease and its treatment can affect the quality of life in patients. In this study when the quality of life of thalassemic children was compared to control group it was found that the result was significantly lower. Age at onset of anemia, age at first transfusion, irregular iron chelation therapy and low pre-transfusion hemoglobin levels were factors significantly affecting health-related quality of life. Various studies have reported that thalassemia pediatric had significantly lower health-related quality of life in all dimensions compared to their healthy counterparts [7, 13]. Also, it was observed that among all four domains of quality of life, school function was a most affected domain. This may be because thalassemia children are frequently absent from school since they routinely go to the hospital for blood transfusion. Also one of the reasons for academic performance could be that thalassemia patients may face deprecatory remarks from their peers or teachers. A school environment that includes verbal abuse and less peer support for ill children may be a problem. Insufficient knowledge of teachers about the illness and the inability to spend adequate time with chronically ill children present barriers to the integration of the chronically ill child in a classroom situation.

In the absence of adequate psychosocial intervention, chronically ill children may remain outside scope of formal education, something that in turn increases their isolation and misery which further affects the social life of the thalassemia patients. There is a need to develop a link between the patients, school officials, the family and the physician as they play an important role in helping children overcome this problem.

As thalassemia is a type of anemia, improper oxygenation of cells can affect normal physical functions. This can lead to hindrance in patient's day to day activities. The chronic nature of the disease, regular visit to hospitals for blood transfusions and decreased physical functions may have an influence on the emotions of the patients. This can make them more vulnerable to mental illnesses like anxiety and depression.

In this study, we found that most of the patients had their mean pre-transfusion Hb in the range of 5-7.9 gm/dl. The results of our study are comparable to the studies conducted by Masyitah Sri Wahyuni *et al.* and Mohaisen H Adaay *et al.* [13, 8]. These results of low Hb among patients can be explained by the limited health education of the parents about the disease, so that, blood transfusion was used only when the patient showed clinical symptoms caused by severe anemia or simply just to sustain life.

Beta-thalassemia major is a transfusion dependent anemia requiring a lifelong blood transfusion for the afflicted patients to stay alive. In this study, it was observed that 69.23% came for blood transfusion once a month. The frequency of blood transfusion depends upon the age, severity of anemia, the rate of erythropoiesis, pre-transfusion hemoglobin level, blood type of the patient and availability of the blood.

Commonly occurring adverse transfusion reactions in our study was fever, respiratory distress, swelling at the IV site, vomiting, headache, lower back pain, myalgia abdominal distension, and tachypnea. Fever during transfusion is thought to be caused by recipient antibodies reacting with white cell antigens or white cell fragments in the blood product or due to cytokines which accumulate in the blood product during storage. It is important to distinguish from fever due to the patient's underlying disease or infection (check pre-transfusion temperature). Fever may be the initial symptom in a more serious reaction such as bacterial contamination or hemolytic reaction. Myalgia, headache, and lower back pain can be associated with fever.

Chronic blood transfusions can lead to iron overload in the body. It is necessary to monitor it regularly to avoid complication associated with iron overload. Serum ferritin is a good and practical method to monitor iron overload. Its level should be maintained below 1000 ng/ml or soon it will get accumulated, causing various cardiac, hepatic and endocrine complications.[9]In this study it was found that 28.20% of patients had serum ferritin above 1000 ng/ml while reports for 64.10% of patient's reports were not available. This is because the reports of the thalassemic children were not maintained properly by the parents. Also, there were some patients in whom the serum ferritin test was not conducted; it could be due to low socioeconomic status of the patients.

Based upon the serum ferritin level (>1000 ng/ml), chelation therapy is initiated. In this study, 46.15% of patients were started on chelation therapy, and they were commonly prescribed with

deferasirox. The reason for prescribing this chelation therapy could be that it can be taken orally once daily and the patients are more compliant with the regimen as compared to other chelation therapy. Chelation therapy is initiated based on the number of transfusions a patient is receiving and whether the therapeutic goal is to decrease or maintain body iron levels [3].

The combination of transfusion and chelation therapy has dramatically extended the life expectancy of these patients, thus transforming thalassemia from a rapidly fatal disease 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 suffering from thalassemia major. Complications which were observed in this study was hepatosplenomegaly (33.34%) and infections like HIV and sepsis (25%) followed by cardiac complication and bone deformity. A similar study was carried out by Kirti Grow *et al.* which reported growth failure as commonly occurring complication followed by cardiac and endocrine complications [12]. Hepatomegaly can be related to significant extramedullary hematopoiesis. Frequent blood transfusion can lead to iron overload, especially in the liver. The liver has a large capacity to produce proteins, which bind the iron and store it in the form of ferritin and hemosiderin. Therefore, it can produce severe iron overload [17]. Splenomegaly (enlarged spleen) is common in thalassemia major due to the high rate of hemolysis (red blood cell destruction). This takes place because the spleen sees the defective red cell of the thalassemic's as deficient and the transfused red cells as invaders (much the same as with host vs. graft disease) and removes them from circulation. This hyperactivity of the spleen results in splenomegaly due to the amount of blood cells that are filtered out and also by a physical increase in size of the spleen so it can handle what it sees as a heavy load of blood cells that need to be filtered out.

In this study, 53.85% were started on supportive therapy that is folic acid and calcium supplements. Folic acid is given because folic acid deficiency is a common complication in patients with beta thalassemia, mainly because of the extreme demand associated with the severe expansion of bone marrow, also poor absorption and intake can further contribute to its deficiency [18]. A calcium supplement is given to prevent osteoporosis, which is one of the complications of the disease [6]. This could be because of progressive marrow expansion, direct iron toxicity on osteoblasts due to iron overload, as well as liver disease. Furthermore, iron chelation therapy can be correlated with bone deformities.

CONCLUSION

Health-related quality of life of thalassemia children was found to be significantly lower than the control group and also adverse transfusion reactions and complications were observed in our study conducted at Bharati Hospital. By increasing the awareness and knowledge levels of the parents, it can be helpful to get better care locally and thus improve the quality of life in thalassemia children. Cognitive-Behavioral Family Therapy (CBFT) can be an effective psychological approach to children with beta thalassemia major, capable of increasing compliance to treatment, lessening the emotional burden of disease and improving the quality of life of children. It is important to identify and monitor transfusion-related adverse reactions so that we can improve the care given to the patients. Early detection of associated complications and adverse transfusion reactions in beta-thalassemia patients would be quite helpful to reduce the burden of disease through preventive measures. Creating awareness, genetic counseling, premarital and prenatal screening can be the best preventive measures that can be effective in the near future to avoid the occurrence of the disease in children.

CONFLICT OF INTERESTS

Declared none

REFERENCES

1. Amrita Panja, Tapan K Ghosh, Anupam Basu. Genetics of thalassemia in Indian population. J Community Nutrition Health 2012;1:39-45.
2. Thalassemias. Available from: http://www.nhlbi.nih.gov/book/export/html/4874. [Last accessed on 12 Sep 2014].
3. P Sandhya Rani, S Vijayakumar, N Chandana. Beta–thalassemia: mini review. Int J Pharmacol Res 2013;3:71- 9.
4. Kirti Grow, Minakshi Vashist, Shiksha Sharma. Beta thalassemia in india: current status and the challenges ahead. Int J Pharm Pharm Sci 2014;6:28-33.
5. Farzana Sayani, Molly Warner, John Wu. Guidelines for the Clinical Care of Patients with Thalassemia in Canada. Canada: Anemia Institute for Research and Education; 2009. Available from: http://www.thalassemia.ca/wp-content/uploads/ Thalassemia-Guidelines_LR.pdf. [Last accessed on 10 Sep 2014].
6. Hira Tahir, Syeda Amna Shahid, Khawaja Tahir Mahmood. Complications in thalassemia patients receiving blood transfusion. J Biomed Sci Res 2011;3:339- 46.
7. Kirti Grow, Minakshi Vashist, Shiksha Sharma. Associated complications in beta-thalassemia patients. IOSR J Pharm 2013;3:22-5.
8. Melody J Cunningham, Eric A Macklin, Ellis J Neufeld. Complications of beta-thalassemia major in North America. Am Soc Haematol 2004;104:34-9.
9. Mohaisen H Adaay, Moayed M Al-Anzy, Khudair A Al-Tikriti. Some observations on the occurrence of β-thalassemia in mosul. Iraq Joinerysoft Microsoft Pinpoint 2011;9:270-4.
10. Mehrnoosh Doroudchi, Ali Akbar Asadi Pooya. Thalassemia major and consanguinity in shiraz city, Iran. Turk J Hematol 2004;21:127-30.
11. Qurat-ul-Ain, Muhammad Hassan, Shahid Mahboob Rana. Prevalence of beta-thalassemic patients associated with consanguinity and anti-HCV-antibody positivity–a cross-sectional study. Pak J Zool 2011;43:29-36.
12. Mohammadreza Sattari, Dorna Sheykhi, Alireza Nikanfar. The adverse effects of thalassemia treatments including blood transfusion and main pharmacological therapies. Pharm Sci 2013;18:199-204.
13. Giovanni Caocci, Fabio Efficace, Francesca Ciotti. Health-related quality of life in Middle Eastern children with beta-thalassemia. BMC Blood Disord 2012;12:1-7.
14. Sharaf KhH, Moayad M Y Al Anzym, Mustaffa NG. Certain hematological values of the beta thalassemia major among mosul population. Tikrit J Pure Sci 2006;24:1-6.
15. G Shanthi, D Balasubramaniam, R Srinivasan. Clinical and demographical studies of beta thalassemia in tamilnadu. Res J Pharm Biol Chem Sci 2013;4:952- 6.
16. Tazeen Majeed, Ujala Nayyar, Mohammed Adil Akhther. Frequency of β-thalassemia trait in families of thalassemia major patients, Lahore. J Ayub Med College Abbottabad 2013;25:58-60.
17. Ansari SH, Nida Baig, Tahir Sultan Shamsi. Screening immediate family members for carrier identification and counseling: a cost-effective and practical approach. J Pak Med Assoc 2012;62:1314–7.
18. Hashemizadeh H, Noori R, Kolagari SH. Assessment hepatomegaly and liver enzymes in 100 patients with beta thalassemia major in mashhad, Iran. Iran J Haematol Onco 2012;2:171-7.