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Original Artical

Study on Management Adherence of Parents of Thalassemia Patient

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Abstract

Background: Thalassemia 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. **Objective:** To find out the management adherence of parents with Thalassemia children **Materials and Methods:** A cross sectional, descriptive study was conducted in department of Pediatrics in TMSS Medical College and Rafatullah Community Hospital, Bogura, Bangladesh from May 2018 to April 2019. Parents were interviewed through a pretested questionnaire to assess their knowledge, awareness, and the practices in management adherence as they follow in regard to the transfusion, treatment, vaccination and prevention of thalassemia. **Results:** About 66% patients were males, 34% were females. Family history of thalassemia was present in 26% of the cases and history of consanguineous marriage was present in 7% of the cases. Seventy six percent of the parents were aware of the prenatal diagnosis that could be performed to prevent the birth of thalassemia children ($P<0.05$). About 82% of the parents were undergone for prenatal diagnosis and it was found more in middle income family. So, socioeconomic status-wise difference in attitude towards prenatal diagnosis ($P=.001$) was found statistically significant. Maximum number of parents who gave 6-10 correct answers regarding management adherence were coming for regular follow up, monitoring ferritin levels of their children, whose children were using chelating agents and were screened for HIV, HBsAg and HCV i.e. 52%, 56.2%, 56.7%, 61.8%,58.9% and 60% ($P<0.05$). So relation of knowledge and management adherence was found statistically significant.

Conclusion: A community based educational efforts, social and behavior change to increase awareness against Thalassemia should be developed in Bangladesh based on the findings of this study. Control of thalassemia requires management adherence of the individual Thalassemia parents as well as a community based educational effort to increase the awareness of this problem.

Keywords: Thalassemia, Transfusion, Management adherence.

Introduction

B-thalassemia is an autosomal recessive single gene disorder characterized by defective production of hemoglobin and excessive destruction of Red Blood Cells. Hemoglobin is formed of four protein subunits, normally two α and two β. Genetic mutation in the gene encoding for β subunits of proteins, results in reduced or totally absent synthesis of β globin chain leading to the formation of abnormal hemoglobin or even to the absence of β hemoglobin. 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. 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] Thalassemia exists in three 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 at least 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 that causes severe microcytic, hypochromic anemia leading to early transfusion therapy. Children usually become symptomatic from profound weakness and cardiac decompensation during the latter half of first year.[3] 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 haemolyticfacies (maxilla hyperplasia, flat nasal bridge, frontal bossing) or pathological fractures may develop. Management of patients includes regular blood transfusions to keep haemoglobin (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. 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 thalassemia child. The emotional affliction and financial anguish with the agony and apprehension of the future of their children is put forth by this study.

Materials and Methods

Study design and setting: A cross sectional descriptive study was conducted in the Department of Pediatrics of TMSS Medical College and Rafatullah Community Hospital, Bogura from May 2018 to April 2019. Parents of Thalassemia major patients were interviewed by pretested structured questionnaire for knowledge, attitude, and practices in management adherence of Thalassemia parents in a language best understood by them. Parents who did not give consent were excluded from the study.

Sample collection: Parents of 100 patients who were coming regularly to have blood transfusion from Department of pediatrics, TMSS Medical College and Rafatullah Community Hospital, Bogura.

Inclusion criteria: All the patients coming for regular blood transfusion in the Pediatrics department from 0 year to 15 years of age and submitted the written consent from parents to participate in the study.

Exclusion criteria: Those patients who do not give written consent and are not coming for regular transfusion in the centre during the period of study

Statistical methods: The variables were compared by Chi Square test and results so obtained were analysed using SPSS version 21.0 and level of significance was determined as its 'P' value with $P<0.05$ taken as statistically significant.

Scoring System: The patients were divided into four groups according to age i.e. 0-4 years; 5-9 years; 10-14 years and 15 years and into five groups according to socio-economic status of the patients applying Kuppuswamy Scale. The patients were also divided into three groups on the basis of number of questions regarding knowledge correctly answered i.e. 0-5; 6-10 and 11-15.

Results

Table-I: Distribution of the patients according to their bio-demographic characteristics

Characteristics	Value	
	n	Percentage
Age groups (months)		
0-4	24	24
5-6	35	35
10-14	29	29
15	12	12
Total	100	100
Sex		
Male	66	66
Female	34	34
Total	100	100

Table I shows distribution of patients according to their bio-demographic characteristics where 66% patients were males and 35% were females. About 24% of the patients were in between 0-4 year's age group, 35% of the patients were in 5-6 years of age group, 29% patients were belonged to 10-14 year age group and rest 12% of the patients were 15 years of age.

Table-II: Distribution of the patients according to their socioeconomic characteristics

Characteristics	Value	
	n	Percentage
Area of living		
Rural	32	32
Semi urban	12	12
Urban	56	56
Total	100	100
Socioeconomic status (SES)		
Upper	02	2
Upper middle	37	37
Lower middle	21	21
Upper lower	37	37
Lower	03	3
Total	100	100

Table II shows that 32% of the patients were from rural area, 12% patients lived in semi urban area, and 56% patients were from urban areas. Again, socioeconomic status of these patients was about 37% of the patient belonged to upper middle class and upper lower class respectively. On the other hand, 21% patients SES were lower middle class, 3% patient's lower class and only 2% patient's status was upper class.

Table- III: Distribution of the Thalassemic patients according to the age at the time of their diagnosis

Thalassemia diagnosed at age in months	n	Percentage
0-6	65	65
7-12	28	28
13-36	07	07
Total	100	100

Table III depicts the distribution of the patients according to the age at the time of their diagnosis. About 65% of the cases the age of diagnosis of Thalasssemia was between 0-6 months, 28% of cases between 7-12 months, and 07% of the cases were diagnosed between 1-3 years of age. About 07% of the patients were diagnosed within 3 years of age.

Table-IV: Distribution of the patients according to their family history of Thalassemia and consaguineous marriage of parents

Characteristics		Male n (%)	Female n (%)	Total (%)
Family History				
Thalassemia	Present	14(52)	12(48)	26
	Absent	50(69.5)	22(30.6)	72
Consanguineous Marriage	Present	3(42.8)	4(57.1)	07
	Absent	63(67.7)	30(32.2)	93
Total		66	34	100

Table IV shows distribution of the patients according to their family history of Thalassemia and consaguineous marriage of parents where family history of Thalassemia was present in 26% of the cases and absent in 72% of patients. Again, history of consanguineous marriage was present in 7% of the cases and rest 93% had no history

Table-V: Knowledge and awareness of the parents about Thalassemia disease and its treatment options in relation with age of the Thalassemia patient

Age group / knowledge and awareness	Age in years in months				Total	Statistical Analysis	
	0-4	5-9	10-14	>15	n		
	24	35	29	12	100	χ^2	P
	n (%)	n (%)	n (%)	n (%)			
Type of b lood disorder	11(25.6)	16 (37.2)	9 (20.9)	7 (16.3)	43	7.855	.04
Genetic disorder	10(1 1.9)	38 (45.2)	25(29.8)	13(15.5)	84	9.809	.021
3 Types of Thalassemia	3(10.7)	5(17.8)	16(57.1)	4 (14.3)	28	8.8 14	.016
Role of Consaguiniy	8 (14.0)	16(28.1)	15(26.3)	18(31.6)	57	9.965	.012
Prenatal Diagnosis	13 (17.1)	27(35.5)	20(26.3)	16(21.1)	76	7.408	0.4 5
Is the disease manageable	7 (13.7)	20(39.2)	14(27.5)	10(19.6)	51	9.632	.02 1
Need for BT	20(26)	28(36.4)	19(24.7)	10(13.05)	77	8.3 20	0.2 7
Importance of Ferritin Levels	8 (13.1)	22(36.1)	19 (31.1)	12 (19.7)	61	14. 654	.00 3
Role of chelation therapy	9 (14.1)	22 (34.4)	20 (31.2)	14(21.9)	64	10. 560	.01 3
BMT	14(19.2)	26(35.6)	20(27.4)	13(17.8)	73	8.5 17	.03 1
Role of Hydroxyurea	4 (4 8)	2 (16.7)	3 (20)	3 (25)	12	2.8 13	.510

BT – Blood Transfusion, DFP- Deferiprone, DFR- Deferasirox, DES- Desferrioxamine, BMT – Bone marrow transfusion.

Table V shows knowledge and awareness of the parents about Thalassemia disease and its treatment in relation with the age of the Thalassemia patient. It shows that 43% of the parents were aware of that thalassemia is a blood disorder, 84% of the parents had knowledge that it is a genetic disease and again it was known to 28% of the parents that there are three types of thalassemia. About 57%, 76% and 51% of parents were aware of the role of consanguinity,the prenatal diagnosis that could be performed to prevent the birth of thalassemia children ,the disease is manageable respectively. Again, Knowledge of the parents regarding need for blood transfusion, importance of ferritin levels, role of chelation therapy and bone marrow transplantation was found in 77%, 61%, 64%, and 73% of the parents. ($P<0.05$)

Table -VI: Attitude of parents towards preventive treatment option for Thalassemia in relation to socioeconomic status

Characteristic / Socioeconomic status	Undergone prenatal diagnosis	Agree with performing abortion	Adopting family planning method
	n (%)	n (%)	n (%)
Upper	07 (8.5)	19 (20.7)	02 (1.5)
Upper middle	36 (43.9)	37 (40.2)	26 (38.8)
Lower middle	18(21.9)	02 (1.1)	14 (20.9)
Upper lower	20(24.4)	33 (35.9)	25 (37.3)
Lower	01 (1.2)	02 (2.2)	01 (1.5)
Total (n=100)	82 (100)	93 (100)	68 (100)
$\chi 2$	17.922	2.389	.715
P value	.001	.547	.789

Table VI shows the attitude of parents towards preventive treatment option for Thalassemia in relation to socioeconomic status. It shows that 82% of the parents underwent prenatal diagnosis and socioeconomic status-wise difference in attitude towards prenatal diagnosis (P=.001) is statistically significant. Again, 93% of parents were willing for abortion of thalassemia affected fetus, and 68% of the parents were adopting family planning methods.

Table-VII: Treatment adherence of the parents in relation to their disease knowledge

No.of questions answered by parents regarding disease knowledge	No. of patients	Coming for regular follow up	Monitoring ferritin levels	Using chelating agents	Regularly screened for			Patient undergone splenectomy
					HIV	HBs Ag	HCV	
		n (%)	n (%)	n (%)	n (%)	n (%)	n (%)	n (%)
0-5	28	26(26.5)	15(20.5)	13(17.6)	8(11.8)	5(9.8)	6(12)	1 (20)
6-10	51	51(52)	41(56.2)	42(56.7)	42(61.8)	30(58.9)	30(60)	2 (40)
11-15	21	21(21.4)	17(23.3)	19(25.7)	18(26.5)	16(31.4)	14(28)	2 (40)
Total	100	98(100)	73(100)	74(100)	68(100)	51(100)	50(10)	5(100)
Statistcal analysis	$\chi 2$	9.057	13.956	39.292	8.177	18.470	20.310	1.802
	P	.35	.001	.000	.43	.000	.000	.387

Table-VII shows the treatment adherence of the parents in relation to their disease knowledge It shows that maximum number of parents who were coming for regular follow up, monitoring ferritin levels of their children, whose children were using chelating agents and were screened for HIV, HBsAg and HCV and splenectomy i.e. 52%, 56.2%, 56.7%, 61.8%,58.9%, 60% and 40%. About 51% parents were give 6-10 correct answers. It also shows that 100% i.e. 21/21, 80.9% i.e. 17/21, 90.5% i.e 19/21, 85.7% i.e 18/21, 76.2% i.e 16/21, 66.7% i.e 14/21 of parents who gave 11-15 correct answers regarding disease knowledge were coming for regular follow up, monitoring ferritin levels of their children, using chelating agents and getting their children screened for HIV, HBsAg, HCV respectively. Here, the relation of knowledge and treatment adherence was found statistically significant (P<0.05).

Discussion

In this study, the parents of hundred thalassemia parents were asked questions regarding knowledge about disease and various treatment options available, their attitude towards prevention of birth of thalassemia affected baby and practices followed by them as per pre-designed and pre-tested performa. In this study, 66% patients were males and 34% were females. This finding of male preponderance is in concordance with other studies done by Bandyopadhyay et al (2003); Shukr et al (2011); Yagnik (1997) and Sen et al (1994).[4, 5, 6, 7] In our study 24% of the children were in age group 0-4 years, 35% in 5-9 years, 29% were in 10-14Years and 12% were >15 years. In other studies done by Shukr et al (2011) and Ishaq et al (2012) mean age of the patients was 9.5 years. There is no data available in literature to make age group wise comparison. (Table -I).[5, 8] In our study, 32% of the patients were from rural area,12% patients from semi-urban area and 56% were from urban areas. In this study majority of the parents about 58% are belonged to middle class family. This finding is not in concordance with a study done by Arif et al (2008) where majority of the parents were from low socioeconomic status.[9] There is no study available in literature to make geographical distribution wise comparison. (Table II) In our study, 65% of Thalassemia cases were diagnosed with in 6 month of life. A study conducted by Upadhyay and Chatterjee (2009) shows similar results where 80% of the patients were diagnosed by the age of 3 year. This shows that maximum number of patients of thalassemia major became symptomatic in infancy and the rest in 1st 3 years of life. (Table III).[10] In our study, family history of thalassemia was present in 26% of the cases and

history of consanguineous marriages was present in 07% of the cases. Naseri et al (1997) in their study on status of thalassemia in Iran also reported that 66% of thalassemia children had consanguineous parents. The difference being due to the fact that consanguinity is high in Muslims. (Table IV).[11] In our study 43% of the parents were known that it is a blood disorder, 84% of the parents had knowledge about genetic disease and 76% of the parents were aware of prenatal diagnosis. This is in concordance with study done by Ishak et al (2012) which reported that 76.5% parents knew about prenatal diagnosis.[8] In our study 51% of the parents were aware of that this disease is manageable. About 77% of parents were known that blood transfusion is needed for growth of the child and to maintain hemoglobin levels, 61% of the parents knew the role of ferritin levels and 62% of the parents knew the role of chelation therapy. A statistic significance was found in the knowledge of parents regarding the management of disease (P=.021), need for blood transfusion (P=.027), importance of ferritin levels (P=.003), role of chelation therapy (P=.013) and bone marrow transplantation (BMT) (P=.031). On other hand, knowledge of the parents regarding the inheritance and types of thalassemia, prenatal diagnosis, curability of the disease was also statistically significant in relation to the sex of the patient (P<0.05). Likewise, knowledge of the parents regarding use of deferiprone and desferrioxamine, splenectomy and role of hydroxyurea was not different statistically in the relation of the sex of their children. (Table V) In the present study, 82% of the parents underwent prenatal diagnosis (P<0.05). Similar results were found in study done by Shukr et al (2011).[5] In this study, 93% of the parents were willing for abortion of thalassemia affected pregnancy (P>0.05). Similar results were shown in studies of Ahmad (2007) where majority of the parents and Karimi et al (2010) where 86.7% of the parents were in favor of termination of the affected foetus. In our study majority of the parents were willing for termination of pregnancy with affected fetus because of poor outcome and prognosis. Again, in those who were not willing for abortion the reason was emotional and religious in majority of them.[12, 13] Maximum numbers of parents who underwent prenatal diagnosis was from upper middle class about 43.9%. Here, socioeconomic status wise difference in attitude towards prenatal diagnosis was found statistically significant (P=.001).There was no significant difference in the attitude of the parents towards prevention of disease like performing abortion and adopting family planning in relation to socioeconomic status as P values were >.05. (Table VI). A study by Department of Psychiatry, National Institute of Mental Health and Neurosciences, Bangalore, by Shaligram et al (2007) showed that 44% children had psychosocial problems.[14] Further research can be done regarding psychological issues of Thalassemic childen and their parents. Again, Zahed et al (2002) in their study showed that the recent emergence of oral iron chelation therapy with deferiprone, has helped to lessen the burden of desferrioxamine infusion and thus is improving compliance.[15] Table VII shows the division of parents giving correct answers for knowledge questionnaire into three groups i.e. 1st giving 0-5 correct answers, 2nd giving 6-10 correct answers, 3rd included parents who gave 11-15 correct answers. Then the knowledge was correlated with treatment adherence Maximum percentage of parents who gave 11-15 correct answers, were coming for regular follow up, monitoring ferritin levels of their children, giving chelating agents to their children, getting the screening for HIV, HBsAg and HCV was done. Relation between the treatment adherence and knowledge of the parents was found significant with P values =.35, .001, .000, .43, .000, .000 regarding regular follow up, monitoring of ferritin levels, use of chelating agents, screening for HIV, HBsAg and HCV. This is in concordance with study done by Lee et al (2009) the score for the patient's disease knowledge about thalassemia major was positively correlated with follow-up visit adherence (P<0.001) and with desferrioxamine infusion adherence (P<0.001). [16]

Conclusion

This study has focused on the lack of knowledge among parents of thalassemia 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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