

## Knowledge, Attitude and Practices of Parents of Thalassemic Children Undergoing Treatment at Thalassemia Day Care Centre of Pediatrics Department, Govt. Medical College & Rajindra Hospital, Patiala

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**Abstract :** This study was conducted on parents of 100 patients of thalassemia who were coming regularly for blood transfusion in thalassemia day care centre run by Patiala Thalassemic Children Welfare Association at Rajindra Hospital, Patiala from January 2011 to Nov 2012, done to explore the knowledge of the parents of thalassemic children, their attitude towards the prevention of birth and practices followed by them in relation to the treatment adherence. The parents were interviewed regarding their knowledge about the inheritance and types of thalassemia, prenatal diagnosis and various treatment options available and practices followed by them on a pre-designed questionnaire.

### INTRODUCTION

$\beta$ -thalassemia is an autosomal recessive single gene disorder characterized by defective production of hemoglobin (Hb) and excessive destruction of Red Blood Cells. Hemoglobin is formed of four protein subunits, normally two  $\alpha$  and two  $\beta$ . Genetic mutation in the gene encoding for  $\beta$  subunits of proteins, results in reduced or totally absent synthesis of  $\beta$  globin chain<sup>1</sup>.

In  $\beta$ -thalassemia, there is excess of  $\alpha$ -globin chains relative to  $\beta$  and  $\gamma$  globin chains and  $\alpha$  globin tetramers are formed. These inclusions interact with the red cell membrane and shorten red cell survival, leading to anemia and increased erythroid production.

Mainstay of therapy of thalassemia major is transfusion therapy and management of its complications.

Repeated transfusions can lead to complications associated with iron overload which affects the heart, liver and endocrine glands mainly. Iron build-up is evaluated by measuring serum ferritin levels. When ferritin levels rise above 1000  $\mu\text{g/L}$ , it's the time to start iron chelation therapy<sup>1</sup>. Definitive therapy available for thalassemia major which gives permanent cure is bone marrow transplantation<sup>3</sup>.

It has been estimated that more than 1,00,000 people are born every year all over the world with thalassemia, 10,000 are in India alone. Prevalence of thalassemia in India is 3.3% in general population and 8-15% among certain communities and religions such as Sindhis, Punjabis, Khatri from North, Bengalis, Jains and Muslims<sup>2,4,5</sup>.

$\beta$  thalassemia is a chronic illness that causes excessive psychological burden on children and their families<sup>6</sup>. A psychosocial support aimed at reducing emotional distress, improving compliance of chelation therapy, and strengthening the coping strategies for a better integration in daily life, is therefore necessary<sup>1</sup>.

Improvement of quality of patient care, reinforcement of medical

education and enhanced efforts by clinical staff to provide practical knowledge to patients with thalassemia major should improve patient's adherence to treatment<sup>7</sup>. Although there is need to increase the community awareness of thalassemia; there is possibility that prenatal diagnosis and pregnancy termination will be accepted for the prevention of thalassemia<sup>8</sup>.

### MATERIAL AND METHODS

The study was conducted on parents of 100 patients of thalassemia who were coming regularly for blood transfusion in Thalassemia Day Care Centre run by Patiala Thalassemic Children Welfare Association at Rajindra Hospital, Patiala after taking approval from ethical committee. Diagnosed cases of thalassemia major were included. Questionnaire was drafted to suit the adult population and interview technique was adopted to fill the questionnaire by explaining to mother and father; data thus obtained was analysed statistically.

### RESULTS

The patients were divided into 4 groups according to age, i.e. 0-4 years; 5-9 years; 10-14 years;  $\geq 15$  years; further into 5 groups according to socio economic status i.e. I-upper class, II-upper middle, III-lower middle, IV-upper lower, V-lower; after applying Kuppuswamy scale, into 3 groups on the basis of number of questions of knowledge correctly answered i.e. 0-5, 6-10, 11-15. The variables were compared by Chi square test and results so obtained were analyzed using SPSS version 15 and level of significance was determined as its 'p' value with  $p < 0.05$  taken as statistically significant.

In this study, 25% of the patients were aged between 0-4 yrs, 28% between 10-14 yrs, 13% above 15 yrs of age. Mean age of the patients was 8.36yrs. 66% were males and 44% were females. 38% of patients were from upper middle class and 38% were from upper lower class. 21% patients belonged to lower middle class, 2% to lower class and only 1% belonged to upper class. 57% of the patients were from urban area and 43% were from rural area. In this study maximum number of parents was from khatri community i.e. 41%, followed by jatts (9%), bazigars (9%) and banias (7%). Family history of thalassemia was positive in 23% of the cases and history of consanguineous marriage was present in 4% cases only.

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All the patients were diagnosed within 3yrs of age. 41% of the parents knew that thalassemia is a genetic disorder, 26% of the parents knew that there are three types of thalassemia, 57% knew the role of consanguinity and 76% of the parents had knowledge about the prenatal diagnosis that could be performed to prevent the birth of thalassemic children.

51% of the parents knew that this disease was manageable. 77% of parents knew that blood transfusion was needed for growth of the child and to maintain hemoglobin levels and 61% of the parents knew the role of the ferritin levels. 60% of the parents knew the role of chelation therapy, 43% knew about deferiprone, 48% about deferasirox and only 5% had knowledge about desferrioxamine. Seventy three percent knew about bone marrow transplantation, 42% knew about the surgery i.e. splenectomy and only 10% knew about the role of hydroxyurea. Maximum no. of patients whose parents knew about importance of ferritin levels, role of chelation therapy and bone marrow transplantation below the 5-9 yrs age group. Those who knew about DES had children above 15yrs of age. Knowledge of the parents regarding importance of ferritin levels, role of chelation therapy and option of the DES, BMT was related to age of the patient significantly.

Knowledge about the disorder, prenatal diagnosis, curability of the disease, chelation therapy treatment options, was not different significantly in relation to the sex of the patient. But knowledge about availability of deferiprone was more among the parents of male children and it was found statistically significant.

Maximum percentage of parents who knew about inheritance of disease was 42.9%; 3 types of thalassemia 69.2%; role of consanguinity 47.4%; prenatal diagnosis 46.1%; curability of the disease 62.7% and need for blood transfusion 42.9% were from upper middle class. Knowledge of the parents about these variables according to their socio-economic status was found to be statistically significant.

Maximum number of parents who knew the importance of ferritin level was 52.5%; role of chelation therapy 50%; options of DFP 60%; DFR 54.2%; about bone marrow transplantation 50.7%; surgery 61.9%; and role of hydroxyurea 70%; were from upper middle class. The difference in knowledge of the parents was found to be statistically significant in relation to their socio-economic status.

Maximum percentage of parents (40.6%) who underwent prenatal diagnosis had children of 10-14 years of age. Maximum number of those who were willing for abortion of thalassemia affected foetus and who were adopting family planning method i.e. 34.8% and 38.8% had children between 5-9 years of age. Difference in the attitude of the parents for adopting family planning method in relation to the age of the patients was statistically significant.

Amongst those who underwent prenatal diagnosis was 68.8% and agreed to with performing abortion of thalassemia affected fetus and those who were adopting any family planning method 64.1%, 62.7% were parents of male children. As overall % age of male patients is more than that of female patients attitude of parents towards prevention of disease in relation with sex of the patients was not found to be statistically significant.

Amongst those who underwent prenatal diagnosis, agreed for abortion of thalassemia affected foetus and adopted family planning method, 68.8%, 59.9% and 62.7% were from urban area, respectively. Attitude of parents towards prevention of disease in

relation to geographical area was not found to be statistically significant.

Maximum number of the parents who were coming for regular follow up, monitoring of ferritin levels of their children, whose children were using chelating agents, getting screened for HIV, HBsAg and HCV were 39.2%, 45.2%, 45.9%, 38.4%, 49% and 48%, respectively from the upper middle class. Difference between treatment adherence of the parents in terms of regular follow up, monitoring ferritin levels, using chelating therapy and screening for HBsAg of their children in relation to their socio-economic status was found to be statistically significant.

Maximum number of parents who were coming for regular follow up, monitoring ferritin levels of their children, were using chelating agents and were screened for HIV, HBsAg and HCV i.e. 48.5%, 56.2%, 56.7%, 51.2%, 58.8%, 60% were parents who gave 6-10 correct answers. Also 100%, 90.5%, 85.7%, 80.9%, 71.4%, 66.7%, 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 and HCV respectively. So relation of Knowledge and treatment adherence was found statistically significant.

Maximum number (56.7%) of parents adopting family planning methods were those who answered 6-10 number of questions regarding knowledge. Difference between attitudes of parents towards prevention of disease in relation to knowledge regarding disease was statistically significant. Only relation of knowledge about need for blood transfusion and chelating therapy in parents was statistically significant with presence of tamper tantrums. Relation of knowledge about treatment options available and the effect on growth, behavior and studies was not found statistically significant.

## DISCUSSION

In this study, 66% patients were males and 34% were females. Mean age of the patients was 8.36 years. This finding is in concordance with study done by Shukr et al<sup>9</sup>. Majority of the parents belong to middle class. This finding is not in concordance with a study done by Arif et al<sup>10</sup>. There is no study available in literature to make geographical distribution wise comparison.

In our study, family history of thalassemia was present in 23% of the cases and history of consanguineous marriages was present in 4% of the cases. This finding is not in concordance with study conducted by Arif et al<sup>10</sup>.

In our study 100% were diagnosed within 3 years of age. Similar result seen in study conducted by Upadhyay et al<sup>11</sup>. This shows that maximum number of patients of thalassemia major became symptomatic in infancy and the rest in 1<sup>st</sup> 3 years of life.

In our study 41% of the parents knew regarding what kind of blood disorder it is, its types and role of consanguinity & Similar result seen in study by Arif et al<sup>10</sup>, Bandyopadhyay et al<sup>12</sup>. In our study 76% of the parents knew about prenatal diagnosis. This is in concordance with study done by Ishak et al<sup>13</sup> & Shukr et al<sup>9</sup>. In our study 43%, 48%, 5% parents knew the option of deferiprone, deferasirox and desferrioxamine. Knowledge regarding desferrioxamine was present in 55% of the parents in Arif et al<sup>10</sup> study. A significant difference was found in the knowledge of parents from rural of urban area regarding the all aspects of disease

studied. No such study is available in literature.

In our study, 32% of the parents underwent prenatal diagnosis. Similar results were found in study done by Shukar et al<sup>9</sup> and 92% of the parents were willing for abortion of thalassemia affected pregnancy. Similar results were shown in studies of Ahmad<sup>14</sup> and Karimi et al<sup>15</sup> and 67% of the parents were currently practising contraception. This result is similar to the study done Han et al<sup>8</sup>.

In this study, parents who brought their children for regular follow up, getting ferritin levels done, using chelation therapy, getting viral markers done regularly had their children >15 years old, from upper middle class and from Khatri community.

Positive attitude towards prevention of disease in relation to knowledge about disease, was found significant.

#### Attitude of parents towards improving the quality of life of the patients in relation to Knowledge about treatment options available.

In our study, relation of knowledge of chelation therapy and effect of disease on behavior of the child; knowledge about need for blood transfusion, chelation therapy and presence of temper tantrums, were correlated significantly. Knowledge about the treatment options available and effect on growth, behavior and studies was not found significant. No study in the literature has correlated the knowledge about treatment options and quality of life of the patients.

In our study, 36% of the patients had effect of disease on their behavior. Similar results were shown in study conducted at by Shaligram et al<sup>16</sup>, Canatan D. et al<sup>17</sup>. Psychosocial support aimed at reducing emotional distress, improving the compliance to chelation therapy, and strengthening the coping strategies for a better integrity in daily life, is therefore necessary.

#### CONCLUSION

Knowledge of the parents of thalassemic children was inadequate as only 21% parents gave 11-15 correct answers regarding

knowledge. Features like knowledge of the parents regarding importance of ferritin levels, role of chelation therapy and bone marrow transplantation were correlated to age of the patients, significantly. No significant difference was found in knowledge of parents in relation to sex of the patients. A significant correlation was found in the knowledge of parents and socioeconomic status. Significant correlation has been found between knowledge about disease, treatment options and prenatal diagnosis with treatment adherence. More positive attitude of parents towards prevention of disease was found in parents who answered maximum number of questions correctly.

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