



IMPACT OF THALASSEMIA MAJOR ON PATIENTS; FAMILIES IN SOUTH PUNJAB, PAKISTAN

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ABSTRACT... Objectives: The objectives of this study were to examine the level of awareness of the parents regarding Thalassemia Major; the cost of treatment of Thalassemia Major Patients and the sufferings it brings to the families; the social problems faced by patients' families; to identify the barriers patients' families face in the treatment of Thalassemia child. **Study Design:** Descriptive study. **Setting:** Four Thalassemic Centers (i) Thalassemia / Hemophilia Centre, The Children's Hospital & the Institute of Child Health Multan (ii) Fatimid Foundation Multan (iii) Amna Blood Foundation (iv) Minhajul Quran Multan. **Period:** January-2013 to June-2013. **Methods:** A sample of 500 respondents was drawn from the total population and structured interview schedule was administered. Data were analyzed and interpreted by using SPSS (Statistical Package for Social Sciences) 19.0 version software. The structured interview was discussed with two experts of the Sociology Department and two Senior Doctors (>8 years' experiences) working in the Thalassemia centers and Government Health Institutions. It was revised to incorporate recommended improvements. Descriptive and inferential statistics were applied to analyze the data that includes: frequency, percent, mean, standard deviations. **Results:** Data indicated that 100(20.0%) respondents were patients' father while 329(65.8%) were mothers and 71(14.2%) were close relatives. Of the 500 respondents, 306(61.2%) were married to their first cousins, 91(18.2%) of the respondents were married to their second cousins, 34(6.8%) of the respondents married in distance relatives and 69(13.8%) of the respondents married out of family. **Conclusion:** The study summarized that the health care providers should be encouraged to talk about Thalassemia as a public health problem in Pakistan and should enhance the public awareness to eliminate the Thalassemia in Pakistan.

Key word: Impact; Thalassemia Major; Patient Families

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INTRODUCTION

Thalassemia major is an inherited hematological disorder leading to anemia in affected children. It is an autosomal recessive disease in which chromosome 11 is involved. It affects synthesis of the B globin chain of hemoglobin, which is either decreased or absent, leading to an early turnover of Red Blood Cell (RBC). When one of the beta globin chain genes is normal and other abnormal, it is Thalassemia minor. If both the genes are involved and disorder presents later in life (older children and adults), it is called as Thalassemia intermedia. If both the genes are involved and disorder is manifested early in life (infants) it is called as Thalassemia major which is a more aggressive disease. Abnormal shaped RBCs are rapidly destroyed by the reticuloendothelial system, particularly the spleen leading to micro

crystal hypo chronic anemia and iron overload.¹ Patients of Thalassemia cannot have an adequate hemoglobin in their bodies and they consequently suffer from anemia. The victims of Thalassemia cannot produce red cells in their bodies and need a regular blood transfusion for survival. It is an inherent disease rather than infectious and does not affect by living, sleeping, eating and breathing but it passes from parents to children.² Thalassemia major is the most common genetic disorder which causes a great genetic problem for mankind throughout the world.³

Thalassemia is preventable, but unlikely, it is a serious health problem in Pakistan; approximately 5 to 6 million children are suffering from this disease and carrying its symptoms.⁴

Over 5000 Thalassaemia homozygotes are born every year⁵ and 6% people are carrying the genes of Thalassaemia due to abnormal production of hemoglobin.⁶ These numbers are growing rapidly due to insufficient awareness and educational campaigns. Unfortunately, most of the parents with Thalassaemia traits never come to know that they are carrying the genes as Thalassaemia carriers the main source of the disease.⁶ The disease can only be diagnosed through blood investigation, but people seem to be afraid to get the carrier test due to societal norms and another reason for not going under screening process, adverse implications in marriage prospects, unawareness to disease is another reason for not getting screened. Thalassaemia poses tremendous physical problem in the affected children, as Thalassaemia children depend on blood transfusions and need to be treated the rest of their lives.^{7,8,9} The disease also causes some other medical problems to the patients, i.e. has pale color, Thalassaemia faces, exhaust and cannot do hard work.^{10,11} Moreover, the disease psychologically affects the patients; feeling of helplessness, un-acceptance, loss and grief.^{12,13,14,15, 16} Similarly, Thalassaemia is a major disease and its complications carry a social and financial impact on patients' families, which result in emotional burden, proper management of disease, its financial constraint and complexity with social integration.

Countries like Cyprus and Turkey have adopted the policy of stopping birth of new Thalassaemic children. In Iran, blood test for pre-marriage and the genetic counseling have been made mandatory by the Government. This strategy has produced good results in preventing the Thalassaemia. Observing the socioeconomic scenario and the low rate of literacy in Pakistan, we need a workable strategy to prevent the disease. If we look at the literature, a limited number of studies have been carried out in Pakistan, but all studies reported that Thalassaemia not only affects the child's health, but also devastates the socioeconomic condition of the families and leaves overwhelming psychological and financial burden on patients' families^{17, 18, 19}. But previously no study has been carried out in the population of

Southern Punjab, Pakistan, therefore this study is carried out to fill the gaps in the literature and to contribute to it. The study will help policy makers, philanthropist, social worker, and society member and health professionals to understand the impact of Thalassaemia major on patients' families.

OBJECTIVES OF THE STUDY

The Present study is based on the following objectives:

1. To describe the belief of parents having Thalassaemia child.
2. To evaluate the awareness and knowledge of parents regarding Thalassaemia disease.
3. To identify the socioeconomic impact of the Thalassaemia major on patients' families.
4. To identify the barriers patients' families face.
5. To suggest the recommendation for Thalassaemia prevention.

MATERIALS AND METHODS

The study was carried out in the four Thalassaemia Centers situated in Multan, Southern Punjab, Pakistan. The study population comprised of the parents of registered Thalassaemia major patients at the (1) The Children's Hospital and the Institute of Child Health Multan, (2) Fatimid Blood Foundation, (3) Minhaj-UI-Quran, and (4) Amina Blood Foundation Multan. Interview schedule was developed after accessing the situation and process of blood transfusion in these Centers. Interview schedule was discussed with two senior doctors (>8 years' experience) working at Thalassaemia Centers. It was revised to incorporate the recommended improvements. A five and four point Likert scale was used to procure the opinion of respondents about every statement that asked in the questionnaire. A Likert scale measures the level of agreement and frequency of use, i.e. it shows the opinion of a person that up to what extent he agrees or disagrees with the question. The most common scales which have been 5=Very Often, 4= Often, 3= Sometimes, 2= Seldom, 1= Never and 4= To a Great Extent, 3= To Some extent, 2= Very Little, 1= Not at all, used in this study by the researcher. A Pilot study was carried out at the completion of 15 questionnaires, to check the validity and reliability

of the interview. Data was analyzed statistically by means of Portable IBM SPSS Statistics (Statistical Package for the Social Sciences) version 19. The researcher used descriptive statistics, which include frequency, percentage, mean and standard deviation. Respondents' anonymity and confidentiality were ensured. Informed consent was obtained from participants. During the data collection process, the purpose of the study was explained to respondents in detail. In case of their questions, they were answered calmly. Respondents voluntarily participated in this study. Respondents were also briefed and encouraged to ask for the additional information or questions.

RESULTS

Respondents age was broad, with 25(5.0%) between 15- 20 years, 107(21.4%) from 21-25 years, 111(22.2%) from 26-30 years, 134(26.8%) from 31-35 years, 53(10.6%) from 36-40 years, 40(8.0%) from 41-45 years, 14(2.8%) from 46-50 years, 9(1.8%) from 51-55 years and 7(1.4%) between 56-60 years. Most of the respondents 134(26.8%) were from the age group ranging 31-35 years, followed by 111(22.2%) respondents from age group 26-30. While the less number of respondents 7(1.4%) was from the age group 56-60 years. Demographic information shows that 119(23.8%) were male and 381(76.2%) were female. Data indicated that 100(20.0%) respondents were patients' father while 329(65.8%) were mothers and 71(14.2%) were close relatives. Of the 500 respondents, 224(48.8%) were from rural areas and 256(51.2%) respondents were from the urban areas. Analysis shows that 234(46.8%) respondents were literate and 266(53.2%) were illiterate. The results depicted that illiterate respondents were more in numbers. Illiteracy among the families might stress on understanding and awareness of the chronic disease like Thalassemia. Of the total respondents, 5(1.0%) were unemployed, 12(2.4%) were working in the business sector, 47(9.4%) were doing private work, 4(.8%) were government employees, 79(15.8%) were laborers, and 353(70.6%) were unemployed. This indicates that the majority of the (70.6%) respondents were unemployed. Amongst the

total respondents 34(6.8%) have the monthly family income were between Rs.1000 to 3000, 99 (19.8%) respondents were Rs. 3100 to 5000, 168(33.6%) were Rs. 5100 to 7000, 106(21.2%) were 7100 to 9000 and 93(18.6%) respondents have the monthly income above Rs. 9000.

Among the total participants, 38(7.6%) participants had one child, 58(11.6%) two children, 83(16.6%) three children, 74(14.8%) four children, 88(17.6%) of the respondents had five children, 46(9.2%) six children, 17(3.4%) seven children and 96(19.2%) respondents had more than seven children. It is detected that the commonly, 88 respondents keep five Children. The data shows that 152(30.4%) respondents had more than one Thalassemia major child in the family while 348(69.6%) of the respondents were not have more than one Thalassemia child in the family. Of the total respondents 201(40.2%) living under the joint family system and 299(59.8%) under the nuclear family system. Among the total respondents, 83(16.6%) of the respondents visited to the Thalassemia centers weekly for the purpose of their children transfusion, 214(42.8%) of the respondents visited Thalassemia centers after fifteen days and 168(33.6%) visited on a monthly basis. Among them, 18(3.6%) of the respondents received blood for their Thalassemia children through Government Health Institutions, 40(8.0%) of the respondents received blood from private and government hospitals, while 59(11.8%) received blood with the support of their relatives and majority 383(76.6%) of the respondents received from their friends.

Figure 1. describes the distribution of respondents by their marriage types. Of the total respondents, 306(61.2%) were married to their first cousins, 91(18.2%) of the respondents were married to their second cousins, 34(6.8%) of the respondents married in distance relatives and 69(13.8%) of the respondents married out of family. It is observed that 306(61.2%) couples married endogamous with 1st cousins. It further shows that mostly the marriage with the first cousin leads to the Thalassemia Major in the family.

Of the 500 respondents, 29(5.8%) had knowledge

that Thalassemia is an inherited disease while majority 471(94.2%) had no knowledge about it. Among the total respondents, 48(9.6%) of the respondents went the process of carrier test and 452(90.4%) did not get the carrier test.

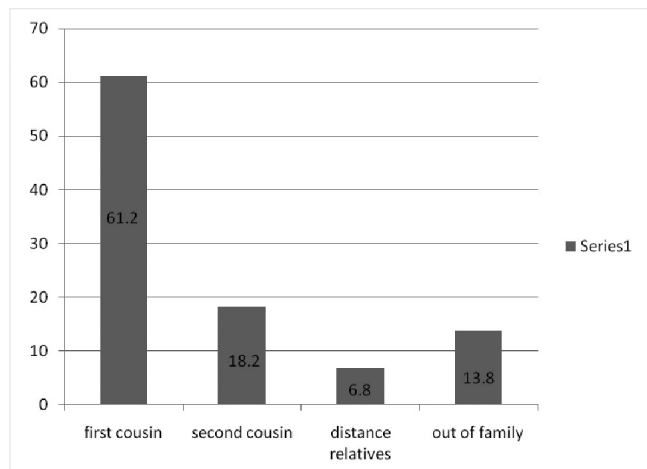


Figure-1. Type of Marriage

Respondents were asked a question regarding the difference between government and private hospital treatments. The majority of the respondents 'somewhat' see a difference between the treatment of government and private hospitals ($\mu=2.71$). The respondents were asked questions whether they face any reaction from in-laws. They face 'somewhat' reaction from their in-laws ($\mu=2.56$). Respondents were asked a question regarding how they keep balance between normal and affected children. The majority of the respondents do balance 'very little' between normal and effected children ($\mu=1.84$). In order to know respondents tendency towards family planning, it reveals that their tendency towards family planning were 'very little' ($\mu=1.72$). (table-I).

Ranks	Statements	N	Mean	Std. Deviation
1	Difference between the treatment of Govt. and private hospitals	500	2.71	.964
2	Reaction from in-laws	500	2.56	.764
3	Balance between normal & affected children	500	1.84	1.080
4	Parents tendency towards family planning	500	1.72	.466

Table-I. Parents' perception about general questions

Score: 4= To a Great Extent, 3= somewhat, 2= Very Little, 1= Not at all

The respondents were asked questions related to their knowledge and treatments of the Thalassemic child. They were asked about their level of knowledge regarding Thalassemic test, Table 2 shows that they were having knowledge 'to a great extent' about Thalassemia test ($\mu=3.87$). In response to a question, respondents 'somewhat' agree that education of parents plays an important role in diagnosing of Thalassemia disease in child ($\mu=2.67$). While they believe in rituals regarding child treatment is 'very litter' ($\mu=1.79$). (Table-II).

Ranks	Statements	N	Mean	Std. Deviation
1	Level of parents' knowledge about test	500	3.87	.437
2	Education of parents plays an important role in diagnosing of child disease	500	2.67	.846
3	Believe in rituals regarding treatment	500	1.79	1.184

Table-II. Parent's knowledge towards Thalassemia disease

Score: 4= To a Great Extent, 3= Somewhat, 2= Very Little, 1= Not at all

Respondents were asked a set of questions regarding their social life. Respondents 'somewhat' feel that their life is unsuccessful and messed-up because of the child's disease ($\mu=3.10$). They 'somewhat' feel hesitation while talking about the disease with others ($\mu=2.70$). Respondent's feel 'very little' aggression because of the child's disease ($\mu=1.87$). They feel homelessness and mental stress because of the child's disease with a mean score 1.42 and 1.05

respectively (Table-III).

Ranks	Statements	N	Mean	Std. Deviation
1	Life is unsuccessful & messed-up because of child disease	500	3.10	.857
2	Hesitation while talking about the disease with others	500	2.70	1.000
3	Aggression because of child disease	500	1.87	.967
4	Homelessness feelings because of Childs disease	500	1.42	.719
5	Mental stress because of child disease	500	1.05	.240
6	Financial burden of child treatment	500	1.01	.155

Table-III. Distribution of respondent's general questions

Score: 4= To a Great Extent, 3= Somewhat, 2= Very Little, 1= Not at all

DISCUSSION

The study was carried out to explore the impact of Thalassemia major on patients' families in Pakistan. The researcher used interview guide to achieve the objectives of this study. The results of the study showed that the parents' understanding of disease were of very low level; they explained that cousin marriage was the root cause of this disease, on the other hand they were reluctant to have pre-marital screening and pre-natal diagnosis. The findings of this study correlate with other studies^{16,9,17,18,19} that inadequate knowledge, lack of pre-marital screening practices; illiteracy and unawareness were the main reasons in prevalence of the disease. Findings showed that the majority of Thalassemia children's parents were first cousins. The majority of mothers were illiterate. They were unfamiliar with any preventive measure during or before Thalassemia. Educated parents were taking preventive measures if a disease was presented in their families¹⁸ but on the other hand, due to the high rate of illiteracy majority was not aware of the preventive measures. The study also addressed some deep misconceptions which comprise Thalassemia is a blood cancer and blood transfusions is the only treatment for this disease. Such misconceptions obviously show the way to unnecessary anxieties. The disease has affected parental financial state very badly, the majority of the affected children's

families were poor and unable to afford the costly treatments. Similar findings have been found in a previous study conducted by Sattari et al.⁸ The average cost for treatment of a Thalassemia patient is around 10,000 PKR (100 USD) per month apart from blood arrangements, which is too high for a poor family³. Though, the Government is providing blood transfusion facilities; subsidize and free medicines in public teaching hospitals, which are only limited to big cities. Similarly, many philanthropists and NGO's are working together for the treatment of the disease, but this is not enough because the miseries of the poor patients are greater than the efforts made so far. If Thalassemic children are not treated in time, they can die between the ages of 1-8 years.⁶ Therefore, proper and timely treatment is essential for the survival of affected children.

Thalassemia is a preventable disease, there is a need for anti Thalassemic educational exhibits and awareness programs. The only effective way to overcome the prevalence of Thalassemia is to stop the birth of Thalassemic children by making pre-marital screening compulsory for the general public and counseling to the affected families. Thalassemia has already been addressed successfully in many countries like Iran, Greece, Italy, and Cyprus by running educational campaigns; making people aware of the disease and highlighting its preventive measures to get rid of the disease.⁵ Prenatal diagnosis and carrier detection facility is available in Pakistan over a decade, but its use is still limited due to the lack of public knowledge and awareness. It is the need of time to realize the density of the disease and practical steps should be taken to reduce the prevalence of the disease. Therefore, it is important to make people aware of the disease, its complications and preventive measures.

It was found that educated parents were more worried and tended to cure the disease during pregnancy. But the majority of the parents in Pakistan are illiterate and they have little awareness about the disease. Moreover, cousin marriages are in vogue which tends to be a causative factor of the disease.¹⁸ When the symptoms of the disease

are treated, the social activities of the affected family members are restricted due to regular transfusion. Usually the patients cannot continue their studies and let off schools. In addition, the patients suffer from other complications such as heart, liver, diabetes, thyroid and hormones.¹⁹ The association between the education and Thalassemia assessed that parents' education is strongly linked to cure the disease especially that test during pregnancy. Because the majority of the parent's were illiterate and the cousin marriage is another major factor. Mostly the population were illiterate and without proper health education it is impossible to know the exact nature of this deadly disease.²⁰ Thalassemia major is a genetic disorder which causes a great genetic problem throughout the world. According to World Health Organization (WHO) in more than 60 Countries the number of the Thalassemic carriers patients reaches to 150 million.

In many countries screening has been made an integral part regarding the maternal and child health programs. It has helped in decreasing the incidence of Thalassemia effectively. In Italian city Sardinia in 1975 screening program was started. In 1995 it decreased the cases of beta Thalassemia from 1:250 to 1:4000. In Montreal, Canada, screening had caused the 95% decrease in the incidence of Thalassemia. The same results were observed in the region of France, Marseille, where 86% carrier partners were located some years before in a screening program in high school and later on all carriers couples were advised to get pre-natal diagnosis.²¹ In Cyprus also screening had helped decrease the Thalassemia cases from 1974 to 1978. In the early 1980s, screening was made essential due to this only five affected births were occurred between 1991 and 2001. And between 2002 to 2007 there was no Thalassemia case was observed. In Cyprus, before the screening parents faced the difficulties to arrange the blood supply and expensive medicines for the affected patients. So if the Thalassemic not had been controlled the blood supply would have been the larger problem.²² In Taiwan the screening programme resulted a noteworthy reduction in the incidence

of beta Thalassemia. Every year 20 Thalassemia children were born before the screening, but after screening this number comes down from three to six. In, Chinese City Guangdong, after screening only one child was born with beta Thalassemia and it happened due to a misdiagnosis. In many major countries of the world pre-natal diagnosis is accessible and abortion is an impediment approach.²¹

The current socioeconomic condition of the country reveals that Pakistan cannot adequately treat all of its Thalassemia. So, considering the gravity of the issue, Government of Punjab has initiated a Thalassemia Prevention Program, which has been implemented in the 18 districts of Punjab. In Pakistan the preventive measures are highly required to stop Thalassemia because almost 10 million people are suffering from carriers of Thalassemia disease. Through health education, awareness about the disease should be increased. Thalassemia counseling centres for the parents should be established in each Tehsil and district levels.

CONCLUSION

The present study concluded that the parents awareness regarding the Thalassemia was inadequate and patients continue to suffer a slow and painful course ultimately leading to death. Parental knowledge regarding the Thalassemia screening and prenatal diagnosis were limited. There is a need to understand the density of the problem and educate the general public and create awareness among Thalassemia families in order to reduce the burden of Thalassemia disease in Pakistan.

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“Happiness lies in the joy of achievement and the thrill of creative effort.”

Franklin D. Roosevelt



AUTHORSHIP AND CONTRIBUTION DECLARATION

Sr. #	Author-s Full Name	Contribution to the paper	Author=s Signature
1	Dr. Kamran Ishfaq	Main investigator, design the structure of the work and set the objectvies.	
2	Muhammad Shabbir Ch.	Find the relevant literature and review it.	
3	Salman Bin Naeem	Designed the questionnaire and gather the feed back from respondent.	
4	Safdar Hussain	Set the format, intent citation and compile a reference sight of the article.	