SOCIAL IMPACT OF THALASSEMIA MAJOR ON PATIENTS' FAMILIES

Syed Hassan Raza[1], Kamran Ishfaq[2*], Hina Fazal[3]

[1]MS Sociology, National College of Business Admistration & Economics, PAKISTAN,
dewanhassanbukhari@gmail.com

[2] Social Welfare Officer, The Children's Hospital & the Institute of Child Health Multan, PAKISTAN,
kamranishfaqchc@gmail.com

[3]Lecturer, Department of Sociology, Bahauddin Zakariya University Multan, Pakistan,
hinabaluch652@gmail.com

*Corresponding author

ABSTRACT

The objectives of this study were to find out the social impact of Thalassemia major on patients' families; to describe the awareness among the parents regarding Thalassemia Major; the cost of treatment of Thalassemia Major and the sufferings it brings to the families; the social problems faced by patients' families. The study population consisted of the parents of Thalassemia major children, visited to the (i) Minhaj UL Quran (ii) Amina Blood Foundation (iii) Fatimid Blood Foundation Multan and (iv) Thalassemia / Hemophilia Centers of The Children's Hospital & the Institute of Child Health Multan. A sample of 500 respondents was drawn from the total population and structured interview was administrated. Data were analyzed and interpreted by using SPSS (Statistical Package for Social Sciences) 19.0 version software. A structured interview was conducted for data collection. Interview schedule was constructed after the detailed literature review and also accessing the situation of peripheries in District Multan. The interview discussed with two experts of Sociology and two senior doctors (>8 years' experience) working in the Thalassemia centers, Government and Private 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, one way ANOVA and Pearson correlation. 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.

Keywords: Impact, Thalassemia, Major, Patients' Families

1. INTRODUCTION

Thalassemia is an inherited blood disorder which is passed from parents to their children. This disorder results in excessive destruction of red blood cells and there is no effective treatment. Patients require lifelong blood transfusion, usually started within 6 to 12 months of birth of the patient, which on the other hand has its own complications. It is a chronic disease that manifests so early in life that it leads to psychological and social problems for parents. The prevalence of thalassemia is very high in Pakistan. Around 5000 children are diagnosed every year. Thalassemia is a most important health problem in Pakistan. Thalassemia major patients depend on the regular blood transfusion and in Pakistan blood is in very short supply and blood banking facilities are no up to the mark (Ansari, Shamsi, 2010). It is a preventable disease demonstrated in countries like Grace, Cyprus and Italy. They were amongst the first to establish a successful national program resulting in significant reduction in the births of affected Children. In Pakistan prenatal diagnosis and carrier detection is available over a decade, but its use remains limited due to lack of public awareness and the cost involved. The religious and cultural scenario in Pakistan is such that consanguineous marriages are very common (Ahmad, Saleem, Modell,

ISBN: 978-605-64453-2-3

Petrous, 2002). There is no concept of premarital screening or counseling of individuals with the family history of the disease. Furthermore antenatal diagnosis is not widely available. The concept of termination of pregnancy is an ethical and religious issue in the community. The only cure is bone marrow transplant, which it is beyond the resources of a large segment of the population (Rehman, Lodhi, 2004).

1.1. KINDS OF THALASSEMIA

There are two kinds of Thalassemia. They are Thalassemia major and Thalassemia minor (which is also called Cooley's anemia).

1.1.1 THALASSEMIA MINOR

Thalassemia minor is also known as Thalassemia traits 'Carriers' Thalassemia. Most of the people having Thalassemia traits are not known that they are suffering with Thalassemia minor. It is the only probe through a special investigation of blood test or if the one's child suffering from Thalassemia major. The Thalassemia trait presented at the time of birth. It remains the same for the whole life and can be passed from parents to their children. The Thalassemia comes down from parents to their children in the following ways.

- If one parent has the beta Thalassemia feature, it is a one in two (50%) chance that each of their children will have beta Thalassemia traits.
- If both parents put up with Thalassemia trait in each pregnancy, there is a likelihood of each pregnancy, having a one in four (25%) that child will become the disease of Thalassemia major.
- If both have normal blood all children are normal.

1.1.2. THALASSEMIA MAJOR

Thalassemia major begins in the early childhood. The major Thalassemia children are normal at the birth, but anemic between the age of three and eighteen months. These children need regular medical treatment and transfused every three to four weeks with the component of blood called red blood cells. These children are regular transfusion dependent. So the regular transfusions cause the accumulated iron in the body which is first stored in the liver. If the children with Thalassemia major are not treated, they usually die between the age of one and eight years. These patients have to be treated the whole time their lives.

1.1.3. BONE MARROW TRANSPLANTATION

The expensive treatment for the patient of Thalassemia major is bone-marrow transplantation. The procedure of bone-marrow transplantation is very expensive, costing more than two million rupees. It is necessary that to have full compatible donor with tissues that match the Thalassemia patient exactly. Most likely the donors can be a brother or a sister of Thalassemia major patient.

2. OBJECTIVES OF THE STUDY

The Present study is based on the following objectives:

1. To describe the social impact of Thalassemia major on patients' families.
2. To evaluate the awareness and the knowledge of patients' parents about Thalassemia disease.
3. To suggest recommendation for the prevention of Thalassemia.

3. MATERIAL & METHODS

The population of this study is comprised of the parents of registered Thalassemia Major Patients at four Thalassemic centers; (1) The Children's Hospital and the Institute of Child Health Multan (CH&ICH), (2) Fatimid Blood Foundation (FBF), (3) Minhaj-UL-Quran (MUQ), and (4) Amina Blood Foundation Multan (ABF). Interview schedule was developed after accessing the situation of Thalassemia Centers and the population in Multan and also conducting a detailed relevant literature review. The Interview schedule was discussed with two senior doctors (>8 years' experience) working at Thalassemia 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,

ISBN: 978-605-64453-2-3

i.e. it shows the opinion of a person that up to what extent he agrees or disagrees with the question. The most common scale which are 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, was used in this study by the researcher. 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, percent, mean and standard deviation.

4. 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%) were 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. The data show that 53(10.6%) respondents were of primary level 53(10.6%) were on the middle level, 59(11.8%) were matriculates, 25(5.0%) were intermediate, 38(7.6%) were graduate, 15(3.0%) were Master and 257(51.4%) were illiterate. It was concluded that the majority of the participants were not educated adequately. 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. It is examined that, the greater part of the respondents (70.6%) was unemployed. The lower level of education can be a cause of unemployment. Of the 500 respondents, 9.4% were working in the private sector and .8% in the government sector. To some extent the people in government sector get some assistance to meet the medical expenses. The minority, (15.8%) were laborers the most vulnerable because of costly medical cure for Thalassemia. Therefore, they are unable to bear the medical expenditure. It is observed that they only get a schedule work and are not capable to afford the medical costs. Amongst the total respondents 34(6.8%) have the monthly family income Rs.1000-3000 while 99 (19.8%) have the family monthly income to Rs. 3100-5000, 168(33.6%) have the monthly family income Rs. 5100-7000, 106(21.2%) have the monthly income 7100-9000 and 93(18.6%) respondents have the monthly income above Rs. 9000.

Among the total participants, 38(7.6%) have a single child, 58(11.6%) of the respondents have two children and the 83(16.6%) of the respondents have three children, 74(14.8%) of respondents have four children, 88(17.6%) of the respondents had five children, 46(9.2%) of the respondents have six children, 17(3.4%) of the respondents have seven children and 96(19.2%) of the respondents have more than seven children. It is detected that the commonly, 88 respondents keep five Children. The data showd that 152(30.4%) respondents have more than one Thalassemia major child in the family while 348(69.6%) of the respondents were not having more than one Thalassemia child in the family. Of the total respondents 345(69.0%) respondents have a single Thalassemia major child and while 155(31.0%) of the respondents have two Thalassemia Major affected children in their family. Almost the parents who had taken a Thalassemia child major they knew about Thalassemia and its nature. Of the 500 respondents, the majority (69.0%) has one Thalassemic child. It convinced them to accept the decision of preventing the birth of another Thalassemic child. Of the total respondents 201 (40.2%) lives under the joint family system and 299 (59.8%) were living under the nuclear family system. Among the total respondents, 83(16.6%) of the respondents weekly visited to the Thalassemia centers for the purpose of their children transfusion while 214(42.8%) of the respondents visits Thalassemia centers after fifteen days; 168(33.6%) of the respondents visits for their children transfusion on a monthly basis. Only 35(7.0%) respondents visited to the Thalassemia centers more than monthly. Among them, 18(3.6%) of the respondents received blood for their Thalassemia children through Government Health Institutions, 40(8.0%) of the respondents get help through both private and government hospitals, while 59(11.8%) received blood with the support of their relatives and majority 383(76.6%) of the respondents have the only source of their friends.

ISBN: 978-605-64453-2-3

Figure.1 shows the distribution of respondent's children by their other complications associated with their Thalassemia major. Among the total respondent's children, 9(1.8%) suffered from other complications of heart, 13(2.6%) of the respondent's children suffer from liver disease, 5(1.0%) of the respondents children had diabetes problems, 7(1.4%) of the respondents had undergone with thyroid problems, 17(3.4%) of the respondent's Thalassemia Major children were suffering from other complication associated with growth hormones and 449(89.8%) respondent's children were not having any other complication associated with Thalassemia Major.

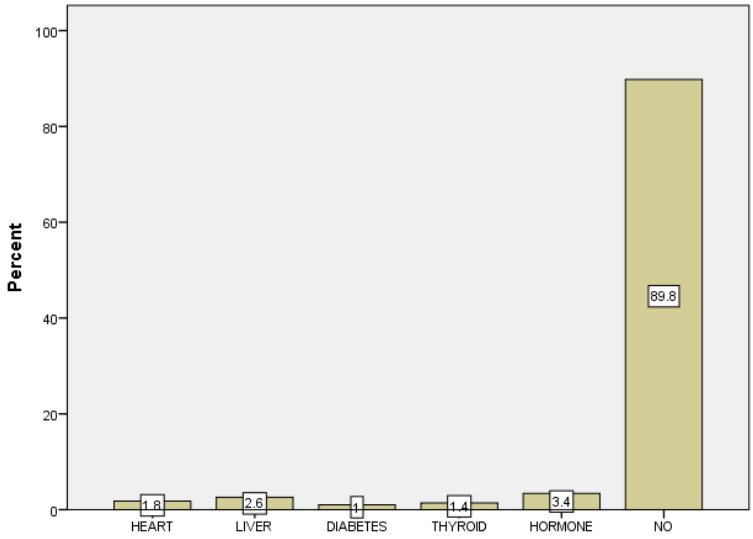

FIGURE 4.16 OTHER COMPLICATIONS ASSOCIATED

Figure 2.describes the distribution of respondents by their marriage type. 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 the family. From the above table 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.

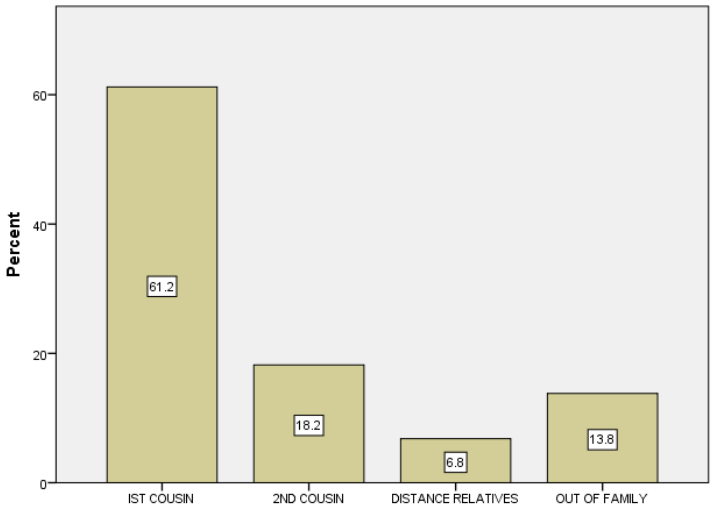

FIGURE 4.17 TYPE OF MARRIAGE

ISBN: 978-605-64453-2-3

Among the total respondents, 29(5.8%) have the knowledge that Thalassemia is inherited disease while majority 471(94.2%) respondents were not having the knowledge that thalassemia is an inherited disease. Pakistani Society is based one male dominated society and male always blame to their wives because they do not understand that Thalassemia is inherited and comes from both sides. Among the total respondents 48(9.6%) of the respondents undertook the carrier test and 452(90.4%) of the respondents did not take the career test. From the above table it is examined that (90.4%) of the respondents did not undertake the test because of lack of knowledge about test.

Respondents were asked a question regarding the difference between government and private hospital treatments. They 'somewhat' see a difference between the treatment of government and private hospitals (μ=2.71). Female respondents were asked questions whether they face any reaction from in-laws. They face 'somewhat' reaction from their in-laws (μ=2.56). Respondents were asked a question regarding how they keep balance between normal and affected children. They do balance 'very little' between normal and affected children (μ=1. 84). In order to know respondents tendency towards family planning, it reveal that their tendency towards family planning were 'very little' (μ=1.72). (table 1).

Parents' perception about general questions

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

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

The respondents were asked questions related their knowledge and treatments of the thalassemic child. They were asked about their level of knowledge regarding Thalassemic test, Table 4.51 shows that they were having knowledge 'to a great extent' about thalassemia test (μ=3. 87). In response to a question, respondents 'somewhat' agree that education of parents plays an important role in diagnosing a thalassemia disease in child (μ=2. 67). While they believe in rituals regarding child treatment is 'very litter' (μ=1. 79). (Table 2).

Parent's knowledge towards Thalassemia disease

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

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

ISBN: 978-605-64453-2-3

Respondents were asked multiple statements regarding their social life. Respondents 'somewhat' feel that their life is successful and messed-up because of the child's disease (μ=3. 10). They 'somewhat' feel hesitation while talking about the disease with others (μ=2.70). Respondent's feel 'very little' aggression because of the child's disease (μ=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 3).

Distribution of respondent's general questions

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

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

5. DISCUSSION

The study was carried out to explore the social impact of Thalassemia major on patients' families in Pakistan. The researcher used interview guide to achieve the objectives of this study. The research study was done in four Thalassemia Centers of the Southern Punjab. Which include (i) Thalassemia / Hemophilia Centre of the Children's Hospital & the Institute of Child Health Multan, (ii) Minhaj-ul-Quran, (iii) Amina blood Foundation and (iv) Fatimid Foundation Multan. The researcher reviewed the literature to explore the relevant topics and to become aware of the findings of the relevant studies conducted before in Pakistan and abroad. The researcher personally obtained the information on designed questionnaire from parents of the Thalassemia major patients through convenient sampling. The researcher had taken this step of gathering data from respondents to ensure the reliability and validity of the data collection tool and another reason was during pre-assessment of the population, researchers found out that the majority of the individuals from the population were illiterate. The data for the study was collected through interview schedule and 500 respondents were interviewed. Statistical Package for Social Sciences (SPSS) 19 version was used for the data analysis. Descriptive and inferential statistics were applied to analyze the data included: frequency distribution, percentages, bar charts, mean, and standard deviation. 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, but they were reluctant to have pre-marital screening and pre-natal diagnosis. The findings of this study correlate with other studies (Liem, Gilgour, Pelligra, Mason, Thompson, 2011; Arif, Fayyaz, Hamid, 2008; Zaman, Salahuddin, 2006) 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 the Thalassemic 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 families (Zaman, Salahuddin, 2006) 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 treatment similar finding that disease causes financial impact on patients' families (Sattari, Sheykhi, Nikanfar, Pourfeiz, Nazari, Dolatkhah, 2012). The average cost for treatment of a thalassemia patient is around 10,000

ISBN: 978-605-64453-2-3

PKR (100 USD) per month apart from blood arrangements, which is too high for a poor family (Khattak, Khan, 2004). 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 (Rehman, Batool, Qadir, 2002). Therefore, proper and timely treatment is essential for the survival of affected children. Thalassemia is a preventable disease and across all focus group discussions, 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 (Ishaq, Abid, Akhtar , Mahmood, 2012). 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. In an empirical study, an association between parents' education and Thalassemia was assessed. 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 (Zaman, Salahuddin, 2006). 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 (Caro, Ward, Green, Huybrechts, Arana , Wait, 2002). 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 is illiterate and the cousin marriage is another major factor. Mostly the population is illiterate and without proper health education it is impossible to know the exact nature of this deadly disease (Alswaidi , Brien, 2009). 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 thalassemia careers 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 effectiveness. 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 a prenatal diagnosis (Alswaidi, Brien, 2009). In Cyprus also screening had helped decrease the thalassemia cases from 1974 to 1978. In the early 1980s, screening was made essentially due to this only five affected births were occurring 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 controlling the blood supply would have been the larger problem (Cousens, Gaff, Metcalfe, Delatycki, 2010). In Taiwan the screening program 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 (Alswaidi, Brien, 2009).

6. CONCLUSION

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 said to be the carriers of thalassemia disease. About 5-8 percent population is suffering from thalassemia in Pakistan. The diagnosis of the disease is easier than the treatment. Only blood transfusion is required to sustain the patient's life, but due to

ISBN: 978-605-64453-2-3

improper screening of blood about 30,000 patients is affected with hepatitis during injecting blood. The Government must make it a law to stop thalassemia by conducting seminars for awareness.

7. REFERENCES

Ansari, S.H., Shamshi, T.S. (2010). Thalassemia Prevention Program, *Haematology Updates*, 23-28.

Ahmad, S., Saleem, M., Modell, B., Petrou, M. (2002). Screening extended families for genetic hemoglobin disorders in Pakistan. *England Journal of Medicine*, vol. 10, 1162-68.

Arif F, Fayyaz J, Hamid A.(2008) Awareness among parents of children with thalassemia major. *Journal of Pakistan Med ical Association*, vol,58(11):621-4.

Alswaidi FM, O'Brien SJ. (2009). Premarital screening programmes for haemoglobinopathies,HIV and hepatitis viruses: review and factors affecting their success. *Journal of medical screening*, vol,16(1):22-8.

Cousens NE, Gaff CL, Metcalfe SA, Delatycki MB. (2010). Carrier screening for beta-thalassaemia: a review of international practice. *European Journal of Human Genetics,* vol, 8(10):1077-83.

Khattak ST, Khan J. (2004). Heterozygous beta thalassemia in parents of children with beta thalassemia major. *Gomal Journal of Medical Sciences*, vol,4(2).

Lodhi, M.U., Lodhi, Y. (2004). Propects and future of conservative management of beta Thalassemia major in a developing country. *Pakistan Journal of Medical Sciences*, vol 20, 105-12.

Liem RI, Gilgour B, Pelligra SA, Mason M, Thompson AA. (2011) The Impact of Thalassemia on Southeast Asian and Asian Indian Families in The United States: A Qualitative Study. *childhood* ,11:12.

Sattari M, Sheykhi D, Nikanfar A, Pourfeiz A, Nazari M, Dolatkhah R. (2012). The financial and social impact of thalassemia and its treatment in iran.

Rehman S, Batool S, Qadir R. (2002). Socio-economic Status of Impact Thalassaemia Child on Families of Faisalabad District. Pakistan Journal of Applied Sciences, vol,2(2):202-5.

Zaman Q, Salahuddin M. (2006). Association between the Education and Thalassaemia: A Statistical Study. Pakistan Journal of Statistics and Operation Research, vol,2(2):103-10.

ISBN: 978-605-64453-2-3