



SOCIO-DEMOGRAPHIC DETERMINANTS; THALASSIMIC CHILDREN

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ABSTRACT... Background: Thalassaemic children require regular multiple blood transfusions. It leads to multiple complications such as blood born infections, iron over load (Haemosidrosis) and spleen enlargement. Thalassaemia transmission to future generations can be prevented and life of thalassaemic children can also be prolonged by proper devoted pre-marital and intra uterine life management and mitigation of fatal complications. **Objectives:** The objective of this study was to assess the socio-demographic determinants of thalassaemic children. **Study Design:** Descriptive study. **Setting:** Thalassaemia Centre Bahawal Victoria Hospital Bahawalpur. **Period:** One year from January 1, 2016 to December 31, 2016. **Material and Method:** Parents /guardians of 200 diagnosed children as thalassaemic, who were receiving blood transfusions were selected for interview. The data was collected by Pre- designed Performa to assess socio-demographic determinants. The data was analyzed to formulate hypothesis. **Results:** It was found that maximum number of patients coming for blood transfusion were from 7-9 years and 10 years and above contributing 30% each while 4-6 years were 24% showing that majority of people seeking for frequent blood transfusion service were over 4 years of age in this area. At least one thalassaemic child death was disclosed by 32 (16%) families. The birth order of the present thalassaemic child as 2nd one was told by 60 (30%) parents. There were 72 (36%) coming from distance <20 km which shows maximum utility of this center by local area population. It was found that 104 (52%) belong to poor socio-economic class. There were 188 (94%) children receiving blood transfusion from teaching hospital blood transfusion center where as 100 (50%) blood donors were unknown people. There were 104 (52%) children who were never checked for Hepatitis C virus markers while 116 (58%) mothers were illiterate. Regular use of iron chelating therapy was found to be 8% only. There were 128 (64%) marriages proposed by parents and 72 (36%) enforced by grand elders resulting 144 (72%) marriages with first cousins. There were 160 (80%) consanguineous marriages and 188 (94%) without premarital thalassaemia test. There were 142 (71%) new born deliveries without prenatal thalassaemia test. **Conclusion:** The enforced consanguineous marriages, lack of pre- marital and pre-natal diagnosis, poverty, illiteracy and lack of iron chelating agent support are the thalassaemia problem exaggerating factors. Reducing magnitude of these problems can mitigate over load and socio-demographic constraints of thalassaemic patients in our country.

Key word: Thalassaemia, marriages, chelating agent, pre-marital, carrier.

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INTRODUCTION

There are multiple blood anomalies. Thalassaemia is one of these abnormal hemoglobin related pathological conditions. Iron deposition in multiple organs of these children results in multiple clinical presentations and fatal complications of thalassaemia, depending upon body tissue of a system involved but anemia is the universal sign.¹ Multiple blood transfusions are highly risky for blood born infections transmission such as hepatitis B or C and HIV including blood parasitic diseases.² Iron is heavy metal and is part of

hemoglobin. Iron over load can be determined by serum ferritin level measurement. Awareness about iron deposition is essential to initialize chelating therapy at the earliest to mitigate the intensity of hemosidrosis effects.³

The thalassaemia major also called Beta-thalassaemia causes more severe clinical presentations of this abnormality. It is due to the most common heterogeneous single gene inherited disorder in beta globin chain of human hemoglobin resulting in defective functional

red blood cells.⁵ The condition is genetically transferred to next generations due to its carriers. However, ideal prevention carrier problem can be achieved by multi-disciplinary actions through a team and not by a single person.⁶ Life long multiple blood transfusions cause many behavioral problems in thalassaemic children. Proper early diagnosis and intervention can mitigate this problem.⁷ It is estimated that there are 5 to 7% carriers in population of Pakistan.⁸ Anxiety, depression and conduct problems are very evident among thalassaemic children.¹⁰

There is strong family bonding system in Pakistan. The marriages are not decided usually by the wish and likeness of young couples, particularly in rural areas. These are enforced commonly by family elders. Male and female new born babies are nominated by grand parents for marriage together to retain inherited properties within there families in rural areas. Similarly, consanguineous marriage is highly influenced by the elders and is common practice among residents of Arab civilization territories.¹¹ This tradition is favorable for onward transmission and clinical appearance of thalassaemia among the next generations. Forceful consanguineous marriages are too difficult to prevent and require reproductive health education programs for many decades.

METHODOLOGY

The descriptive study was carried out at thalassaemia centre, Bahawal Victoria Hospital Bahawalpur which is a tertiary care teaching hospital, during the period of one year from January 1, 2016 to December 31, 2016. Total number of 6757 blood transfusions was given to all thalassaemic children with average 23 transfusions daily. There were 8 to 15 newly diagnosed thalassaemia major patients every month. Sample size of 200 patients both new and old was taken by purposive convenience sample method. A Questionnaire Performa was designed for "face to face" interview to assess socio-demographic determinants of these patients from their parents or guardians seeking for their blood transfusion.

Parents of patients with thalassaemia confirmed,

diagnosed both new and old, registered and receiving blood transfusions were selected for interview after informed consent. Foster mother or father of adopted children and parents non-willing for interview were excluded from study. Those children who were very sick taking emergency management due to thalassaemia complications were also excluded because their parents or guardians were in severe mental stress. All patients who were taking blood transfusions from private blood banks/ hospitals were excluded.

Help of tetra lingual local paramedical staff and nurses was taken for translation and mitigation of language bias problems during interview. The study was carried out on self-finance bases getting no expenses from any donor organization. The help of field staff of Punjab thalassaemia prevention program was taken to study and understand socio-demographic determinants of thalassaemic patients and their solutions. Family pedigree drawn by field staff after thalassaemia carrier screening tests was also studied where ever available. The data was collected, condensed on data master sheet and analyzed to formulate hypothesis.

RESULTS

This study revealed that maximum number of patients coming for blood transfusion were from 7-9 years and 10 years and above contributing 30% each of them. However, 4-6 years age group was very close (24%) to the higher age patients. It has shown that people seeking for blood transfusion service for thalassaemic children were more commonly after 4 years of age in this area. No death by thalassaemia was reported from 148 (74%) families, but at least one child death incidence was disclosed by 32 (16%) families which is considerable death score among thalassaemic families as shown in Table-I.

The birth order of thalassaemic child as 2nd one was told by 60 (30%) parents. That means second baby has relatively more chance to be the sufferer. Similarly 72 (36%) were coming from distance <20 km which shows maximum utility of this center by local area population for blood transfusion facilities. It was found that

socioeconomic status / financial income was inversely proportional to the number of thalamic patients in family because majority of thalassaemic children, 104 (52%) were belonging to poor socio-economic class as shown in Table-I.

In spite of the fact that 188 (94%) children were receiving blood transfusion from teaching hospital blood transfusion center, it was disclosed that majority of blood donors, 100 (50%) were from unknown people which was very alarming. Treatment expenses were afforded by 192 (96%) parents only, showing no considerable financial support from other sources as depicted in Table-I. Viral hepatitis markers are very essential for these patients but 104 (52%) revealed that these were never checked. Maternal Illiteracy has been found to play major role for spread of thalassaemia as 116 (58%) mothers were illiterate. However, 76 (38%) fathers were secondary school level

literate. There were 180 (90%) families having 1-5 children alive in average but regular use of iron chelating therapy among their thalassaemic children was found to be 8% only, as shown on Table-I.

Marriages constraints were inquired and it revealed that 128 (64%) marriages were proposed by parents and 72 (36%) proposals were enforced by grand elders without like and dislike right giving to children. It is alarming constraint to control thalassaemia spread because 144 (72%) marriages were proposed within first cousins. There were 160 (80%) consanguineous marriages providing fertile ground for thalassaemia major genes expression while 188 (94%) marriages were done without premarital thalassaemia test. There were 142 (71%) new born deliveries without prenatal chorionic villous biopsy test for thalassaemia as shown in Table-I

Sr.	Variable	A	B	C	D
1	Age in years group	Birth-3 years 32 (16 %)	4-6 years 48 (24%)	7-9 years 60 (30%)	10 yrs & above 60 (30%)
2	Thalassaemic deaths already in family	Nil 148 (74%)	1 child 32 (16 %)	2 children 4 (2%)	>2 children 16 (8 %)
3	Birth order of this thalassaemic child	1 st 48 (24%)	2 nd 60 (30%)	3 rd 36 (18%)	4 th or above 56 (28%)
4	Home distance	<20 km 72 (36 %)	21-40 km 32 (16 %)	41-60 km 32 (16%)	>60 km 64 (32%)
5	Socioeconomics Income/ month	<20 Thousands 104 (52 %)	21-40 Thsds 60 (30%)	41-60 Thsds 20 (10%)	>60 Thsds 16 (8 %)
6	Who is blood donor	Parents 8 (4 %)	Relative 40 (20%)	Unknown 100 (50%)	Blood Bank 52 (26 %)
7	Blood transfusion frequent location	Primary H. C. Nil (0%)	2ndary H. C. Nil (0%)	Tertiary H.C. 12 (6%)	Teaching H.C. 188 (94%)
8	Treatment expenses Payer	Parents 192 (96%)	Government Nil (0 %)	Any NGO 4 (2%)	Relatives 4 (2 %)
9	Viral Hepatitis (Child) Markers Test	Never done 104 (52%)	Hep B Positive 4 (2%)	Hep C Pos+ 28 (14%)	Both Neg 64 (32%)
10	Mother Education	Illiterate 116 (58%)	Primary 16 (8%)	Secondary 52 (26%)	Higher 16 (8%)
11	Father Education	Illiterate 64 (32%)	Primary 28 (14%)	Secondary 76 (38%)	Higher 32 (16%)
12	Number of all children alive in family	A		B	
		1-5 children 180 (90%)	6-10 children 16 (8%)	C	
13	Receiving Iron Chelating therapy	Never 80 (40%)	Off /On 104 (52%)	>10 children 4 (2%)	
14	Who propose marriage in family	Forced, Grand elders 72 (36 %)	Love marriages Nil (0%)	Regularly 16 (8%)	
15	Cousins marriage of parents of this child			a = Yes	
				144 (72%)	
16	Consanguineous marriages in family			b = No	
17	Pre-marital thalassaemia screening ever done			56 (28%)	
18	Pre- natal thalassaemia screening ever done			40 (20%)	
19	Gender of thalassaemic children studied			188 (94%)	
				142 (71%)	
				a:M 128 (64%)	
				b: F 72 (36%)	

Table-I. Socio-demographic determinants of thalassaemic patients n=200

DISCUSSION

Thalassemia is one of the haemoglobinopathies having different sub types. This disease is spreading with enormous speed in our area. Thalassaemic children have multiple socio-demographic determinants. Iron overload (haemosiderosis) is fatal for them. All parents cannot afford chelating therapy expenses. The irregular, costly and insufficient supply problem of chelating agents, is increasing early death events of these children. The best portable infusion pump services can keep normal daily life activities of these children as depicted by Borgna-Pignatti C¹ but our study revealed only 16 (8%) patients who were using regular chelating therapy. There were 80 (40%) patients who have never received any chelating agent. It has been quoted by Tiwari A and Alisha AK³ that lack of awareness also play major role for use of chelating agent. There is urgent need of rationalization of chelating therapy.

There were 104 (52%) patients who have never been checked for Hepatitis C viral infection i.e. HCV status was unknown. It has been quoted in other literature¹ that thalassaemic patients get HCV infection commonly but our study revealed serious constraint of unknown viral hepatitis status among more than 50% cases. It was found that 28 (14%) patients were suffering from known hepatitis C infection but another study² have revealed that 15% children are infected with HCV which is very close to our finding.

Our study have revealed that pre-marital and pre-natal screening for thalassemia detection were not done by 188 (94%) and 142 (71%) parents respectively. It is major socio-demographic problem responsible for rapid up surge of thalassemia spread in our area. Similarly Petrou M⁴ has also described that there is a little use of prenatal diagnosis in communities where high incidence rate is noted. We have found very strong preference of consanguineous marriages (80%) among first cousins (72%). It is very unfortunate that marriages proposed by parents were 64% and forced by grand elders were noted among 36% families. The like or dislike right of children has no value among these families. It is

mentioned in another study⁴ that 98% of at risk couples who were detected to be thalassaemic carrier very close to marriage ceremony proceed to marry.

There were 104 (52%) families from poor socioeconomic class having thalassaemic children. It is major socio-demographic problem because parents of more than half of thalassaemic children cannot afford costly therapeutic abortion and chelating therapy due to limitation of their resources. It is concordant with studies of other researchers⁴ also. The gender of thalassaemic children was inquired. It was found that there were 128 (64%) males and 72 (36%) females which correlate very closely with another study⁸ describing males to be 58.2% and females 42.8% cases. It may be most probably due to reproduction of male child preference for multiple reasons.

Our study revealed 116 (58%) mothers to be illiterate. However, Riaz H et al⁸ described 78.8% cases where education of care taker was below 10th grade level which is considerably higher than our finding. We have found father education more than 10th grade to be among 76 (38%) cases. Blood transfusions and all other diagnostic tests services should be decentralized. It will decrease the cost of treatment and family harassment in travelling as mentioned by Malik S and Chatterjee C⁹ also.

In families where access is poor, new health policies must be developed to tackle population diversities towards very high thalassemia prevalence which is emphasized by Szezepura A¹² also. Thalassemia control and prevention require joint multi-directional comprehensive efforts with full cooperation and participation of state health services and families.

LIMITATIONS OF STUDY

1. The sample size is relatively small.
2. Blood transfusion based HIV transmission to thalassaemic children has not been included.
3. Multiple social communities of different countries are required to make findings more

comprehensive.

CONCLUSION

Pre-marital carrier detection and therapeutic abortion of thalassemia major cases is essential to prevent this disease. Lack of prevention of enforced consanguineous marriages through influential local political and religious personals, poverty, illiteracy and lack of iron chelating agent support are the thalassemia problem exaggerating factors. Reducing these problems can mitigate thalassemia over load to very low level in our community.

RECOMMENDATIONS

Pre-marital thalassemia carrier's identification and prenatal intra uterine diagnostic services by the state health departments through thalassemia prevention program free of cost is essential. It is to identify thalassemia before appearance in the future generations and alert the parents to strengthen community awareness.

Therapeutic abortion of confirmed intra-uterine thalassemia major fetus and pre-marital community intervention action with consent of parents, elders and influential local political or religious community leaders must be emphasized to decrease this rapidly expanding disease in our community. Expenses for the purpose may be paid by the local government.

Consanguineous marriages may be discouraged slowly through constant reproductive health education and different effective psycho-social strategies.

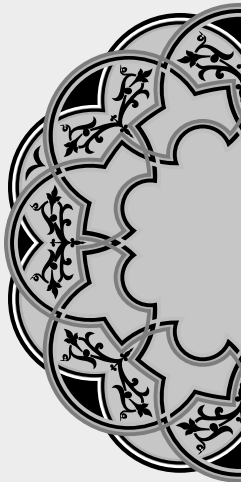
Iron chelating tablets and injections may be provided by health care delivery center free of cost to incorporate and register the maximum number of thalassemic children. Removal of burden of costly treatment from shoulders of thalassemic children's parents is highly recommended.

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“Being defeated is often a temporary conditions; giving up is what makes it permanent.”

Unknown

AUTHORSHIP AND CONTRIBUTION DECLARATION

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