ORIGINAL ARTICLE

IMPACT OF THALASSEMIA CENTRE ON AWARENESS OF PARENTS OF THALASSEMIC PATIENTS ABOUT THE DISEASE: COMPARATIVE STUDY IN MUZAFFARABAD AND KOTLI DISTRICTS

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Background: Thalassemia major is a genetic blood disorder usually presenting in first year of life. Its management requires huge financial, technical, social and psychological support. However preventive measures and proper management reduce morbidity and mortality. It is of utmost importance to get an insight about knowledge of parents who play a vital role in care. In knowledge deficient groups, interventional strategies can help improve patient care. The objective of the study was to compare the awareness of parents of thalassemia patients attending AK CMH Muzaffarabad and thalassemia centre District Head quarter Hospital Kotli. **Methods:** This was a cross sectional study. Non probability convenient sampling was used to collect data by using pre-tested questionnaire. A questionnaire containing 32 items was used on 90 parents of children attending blood bank AK CMH Muzaffarabad and thalassemia centre District Head quarter Hospital Kotli. Data was analyzed on SPSS-21. Consenting parents of diagnosed cases of Thalassemia were included in study. **Results:** Study showed that overall acceptable knowledge about thalassemia was better in respondents from Kotli 32 (76.2%) as compared to Muzaffarabad 18 (41.9%) (p=0.01). When estimating correct responses in gender and parents with different educational background, significant difference (p=0.04) was found between percentage of correct answers in educated 58.14±5.01 compared to uneducated 51.08±10.9 respondents. **Conclusion:** Health facility having well established thalassemia centre has confirmed positive impact on awareness about Thalassemia inheritance, prevention and management. Thalassemia awareness programs, premarital screening and legislation are the need of the day.

Keywords: Thalassemia major, Thalassemia minor, Genetic counselling, Awareness raising programs

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INTRODUCTION

Thalassemia major is a genetic blood disorder that usually manifests in first year of life, patient develops hepatosplenomegaly, frontal bossing, maxillary prominence, severe anaemia and numerous other complications.[1] Disease can be diagnosed by blood tests, i.e., low MCV, low MCH and haemoglobin electrophoresis.[2] Treatment modalities include lifelong transfusions and chelation therapy.[3] Both modalities have complications. Bone marrow transplantation is the real answer[4] which is not affordable to most of the patients. Gene therapy is still an experimental tool[5]. In developing countries, majority of children remain undiagnosed, are misdiagnosed or die due to sub optimal treatment. Average life span of thalassemia children in Pakistan is 10 years.[6]

Thalassemia heterozygotes are also called carriers. Birth of thalassemia children can be prevented by providing genetic counseling.[2] Each year worldwide, around 60,000 symptomatic individuals are born out of 80–90 million thalassemia carriers.[7] In Pakistan 1–4 out of 1000 infants have the disease.[8,9] There is no National registry but it is estimated that annual reported cases of Beta-Thalassemia are more than 4,000. Estimated Beta-Thalassemia carrier rate is around 5.3%, with certain regional variation between 1.8 and 8.0%.[8–10] Incidence of beta thalassemia trait in Muzaffarabad is 5.6%,[11] Azad Kashmir is a remote mountainous area with difficult hilly terrains of 13,297 Km^2 and a population of 4,567,982 inhabitants. Consanguineous marriages are quiet common in Pakistani culture.[12–14] In Kashmiri culture it is even more common. Consanguinity in the presence of high gene frequency, and high birth rate, along with large population size constitute significant risk factors.[8]

Pakistani population is at high risk but has poor public knowledge about the disease.[15] Particular deficiencies include premarital screening, invasive[16] and non-invasive[17] prenatal diagnosis, and other prevention related issues.[10] Even people don't want to disclose the presence of disease in their families.[18] In some places carriers are socially isolated and have less chances of getting married.[19] Particularly abortion for a pregnancy affected with Thalassemia major is very difficult.[20] With increasing awareness, families can deal better and have positive attitude towards disease.[21]

Health care community is not adequately sensitized to deal with genetic counselling and treatment related issues.

Life span depends on quality of health care provided to patient.[22] Thalassemia population is young so family particularly parents, play the primary role as care provider. They must have adequate information about the disease process and proper management.[23] Knowledge of preventive strategies in general public can help reduce the birth of thalassemia children.[24] With this background information we wanted to compare the way various aspects of thalassemia are understood by the parents of patients suffering from thalassemia.

SUBJECTS AND METHODS

After ethical approval from Ethical review committee of AJK Medical College Muzaffarabad a pre-tested questionnaire[21–25] modified after permission was used to assess various aspects of thalassemia. Overall 32 items were included.

This was a cross sectional study for which simple convenient sampling was used to select the study participants from district Muzaffarabad and Kotli from June to December 2017. Consenting parents of diagnosed cases of Thalassemia major were included in study. Patients with any other diagnosis were excluded. Data was entered and analyzed using IBM SPSS-21. Categorical variable were described as frequency and percentages. Numerical variables were described as Mean±SD. The means were compared using independent sample *t*-test, and $p<0.05$ was considered statistically significant.

RESULTS

Ninety questionnaires were distributed to parents bringing their children for blood transfusion. Eighty five parents participated in the survey with 94.44% response rate. Forty three questionnaires were completed at Muzaffarabad while 42 were from Kotli. Forty one (48.2%) of the respondents were male and 44 (51.8%) were female. Most of the participants 75 (88.2%) were between the ages of 18–40 years. Majority of the accompanying parents 61 (71.8%) were educated. When estimating correct responses in gender and parents with different educational background, significant differences ($p=0.04$) were found between percentage of correct answers in educated (58.14±5.01) compared to uneducated (51.08±10.9) respondents. Mean percentage difference between genders was non-significant (male 56.8±11.6, female 54.05±10.94). Overall correct, incorrect and don't know responses are shown in Table-1.

Statistically significant ($p<0.05$) differences were found in various Thalassemia knowledge related questions and overall acceptable level of knowledge. More than 50% correct answers to the questions posed in thalassemia questionnaire were considered to be the acceptable level of knowledge regarding thalassemia. When the level of knowledge was compared, it was found that 18 (41.9%) respondents from Muzaffarabad and 32 (76.2%) respondents from Kotli had acceptable level of knowledge regarding thalassemia and the difference between the knowledge was statistically significant $p=0.01$ with residents of Kotli possessing more acceptable level of knowledge than residents of Muzaffarabad.

Table-1: Knowledge about Beta Thalassemia among parents (N=85) based on 32 questions [n (%)]

Assessment No		Correct	Incorrect	Don't Know
INHERITANCE RELATED				
1	Thalassemia is an inherited disorder	54 (63.5)	17 (20)	14 (16.5)
2	It is transmitted through genes	42 (49.4)	16 (18.8)	27 (31.8)
3	If both parents have Thalassemia major only then Thalassemia major baby is born	55 (64.7)	25 (29.4)	5 (5.9)
4	Thalassemia minor, can be present in siblings	53 (62.3)	26 (30.6)	6 (7.1)
5	Thalassemia is not an allergic reaction	25 (29.4)	50 (58.8)	10 (11.8)
6	Thalassemia is not a contagious disease	58 (68.2)	15 (17.6)	12 (14.2)
7	Thalassemia can skip a generation	53 (62.4)	14 (16.5)	18 (21.1)
8	Consanguinity is an important risk factor	74 (87.1)	8 (9.4)	3 (3.5)
9	Thalassemia is a disease of the blood	73 (85.9)	8 (9.4)	4 (4.7)
Group mean		55.4 (65.2)	18.6 (21.8)	11 (13)
PREVENTION RELATED				
10	Thalassemia major presents with anaemia.	84 (96.5)	0 (0)	1 (1.2)
11	Thalassemia be identified by a blood test	56 (65.9)	25 (29.4)	4 (4.7)
12	If both parents are carriers, they can have thalassemia child	60 (70.6)	18 (21.2)	7 (8.2)
13	Thalassemia is preventable	32 (37.6)	36 (42.4)	17 (20)
14	Thalassemia is of different types	60 (70.6)	13 (15.3)	12 (14.1)
Group mean		60 (70.1)	17.2 (19.6)	7.8 (11.1)
TREATMENT RELATED				
15	Patient can't survive if left untreated	74 (87.1)	10 (11.7)	1 (1.2)
16	Fever, anaemia, diarrhoea, vomiting indicate worsening condition	71 (83.5)	11 (12.9)	3 (3.6)
17	Blood transfusion is a treatment modality	56 (65.9)	25 (29.4)	4 (4.7)
18	Patent can lead normal lives with treatment	54 (63.5)	22 (25.9)	9 (10.6)
19	Thalassemia can only be treated with medications	15 (17.6)	63 (74.1)	7 (8.3)

20	Thalassemia minor person leads a healthy life	53 (62.4)	25 (29.4)	7 (8.2)
21	Thalassemia major is not completely treated with blood transfusions	31 (36.5)	53 (62.4)	1 (1.2)
22	Chelation is one of treatment modalities	14 (16.5)	58 (68.2)	13 (15.3)
23	More chances to develop (transfusion related reaction, kidney failure and stroke)	70 (82.4)	5 (5.9)	10 (11.8)
24	Thalassemia not treatable with surgery	49 (57.6)	25 (29.4)	11 (13)
25	Specific type of food is not a treatment modality	60 (70.6)	15 (17.6)	10 (11.8)
26	Bed rest is not a treatment modality	72 (84.7)	10 (11.8)	3 (3.5)
27	Thalassemia major is curable	42 (49.4)	33 (38.8)	10 (11.8)
28	Incorrect nutrition is not an important risk factor	55 (64.7)	27 (31.8)	3 (3.5)
29	Treatment can't change Thalassemia minor to normal	17 (20.0)	56 (65.9)	12 (14.1)
30	Thalassemia is not sexually transmitted	27 (31.8)	32 (37.6)	26 (30.6)
31	Thalassemia minor does not have a higher risk of infection and/or illnesses	13 (15.3)	58 (68.2)	14 (16.5)
32	Problems in Thalassemia major are due to iron overload and low blood transfusion	72 (84.7)	2 (2.4)	11 (12.9)
Group mean		46.7 (54.9)	29.6 (34.8)	8.7 (10.3)

Table-2: A comparison of responses from Kotli and Muzaffarabad

	Correct response from Muzaffarabad (43 respondents)		**Correct response from Kotli (42 respondents)**		
Assessment No	**Frequency**	**Percentage**	**Frequency**	**Percentage**	***p***
INHERITANCE RELATED					
1	26	60.5	28	66.7	0.3
2	14	32.6	28	66.7	0.002
3	30	69.8	25	59.5	0.22
4	20	46.5	33	78.6	0.002
5	17	40.4	8	18	0.045
6	27	62.8	31	73.8	0.19
7	26	18.60	29	21.43	0.27
8	35	81.4	39	92.8	0.10
9	38	88.4	35	83.3	0.36
Group Mean		55.67		62.31	
PREVENTION RELATED					
10	43	100	41	97.9	0.49
11	24	55.8	32	72.6	0.05
12	29	67.4	31	73.8	0.34
13	21	48.8	11	26.2	0.002
14	29	67.4	31	73.8	0.34
Group Mean		67.88		68.86	
TREATMENT RELATED					
15	35	81.4	39	92.8	0.10
16	31	72.1	40	95.2	0.00
17	24	55.8	32	72.6	0.05
18	26	60.5	28	66.7	0.3
19	7	16.2	8	19	0.4
20	26	60.5	27	64.3	0.44
21	19	44.2	12	28.6	0.10
22	8	18.6	6	14.3	0.4
23	31	72.1	39	92.9	0.01
24	26	60.5	23	54.8	0.37
25	29	67.4	31	73.8	0.34
26	37	86	35	83.3	0.48
27	14	32.6	28	66.7	0.002
28	23	53.4	32	76.2	0.05
29	8	18.6	9	21.4	0.47
30	9	20.9	18	42.8	0.02
31	2	4.6	11	26.2	0.006
32	37	86	35	83.3	0.48
Group Mean		50.63		59.72	
Overall Acceptable level of knowledge	**18**	**41.9**	**32**	**76.2**	**0.01**

DISCUSSION

Present study aimed to assess the effectiveness of thalassemia centre in increasing the knowledge of parents of thalassemia patients. We evaluated those aspects of knowledge which can help in prevention of birth of thalassemic children and also can affect the medical compliance and social support of the affected children. We compared parent knowledge across Kotli and Muzaffarabad health facilities. Kotli has well established thalassemia centre where parents frequently

attend disease awareness sessions organized by health care professionals. Though in Muzaffarabad patients get medical support but health education facilities regarding thalassemia are not well organized.

Parents of thalassemia major were poor in knowledge in many areas. Among overall study group, lowest response was about the knowledge that thalassemia minor does not have high risk of infection or illness 15.3% CR (correct response). This reflects that the difference between thalassemia minor and thalassemia major is not well appreciated. Other deficient area under observation was about the chelation therapy (16.5% CR). Lack of awareness, regarding chelation therapy, can result in excessive iron deposition which can damage organs e.g. liver, heart and endocrine system and compromise the quality of life and reduce the life expectancy.

Comprehension of the fact that only medicines are going to treat the child (17.6% CR), results in less than optimum blood transfusion and substandard management. Misconception that thalassemia carrier can be changed to normal (20% CR) can motivate thalassemia carriers to approach the quacks in the hope of getting normal. With the misconception that thalassemia is an allergic reaction (29.4% CR), parents may put child unnecessarily on antihistamine as medicines are easily accessible in our society. The misconception that thalassemia is sexually transmitted (31.8% CR) can generate sex related issues in the couples at risk. A conviction that thalassemia is completely treated by blood transfusion (36.5% CR) restricts the parents from other options of treatment, i.e., bone marrow transplantation.

We compared correct responses with a study done in thalassemia centre in Karachi[21]. Participants in our study have better knowledge about the risk of iron overload (72 *vs* 9.9%), non-transmission by sex (31 *vs* 14.5%) that rest not being a treatment modality (74.7 *vs* 36.6%). Thalassemia carriers lead a normal life (62.4% *vs* 48.8%). Our participant are relatively less aware in the following areas: Thalassemia can be identified by a blood test (65.5% *vs* 93.6% CR) thalassemia is preventable (37.6% *vs* 63.44% CR) only medical treatment can treat thalassemia (17.6% *vs* 73.3% CR) and role of chelation in treatment (16.5% *vs* 59.3% CR).

Reason of far higher knowledge score in some areas of our study may not be due to better efforts to educate the parents in our population. It is to note that study by Maheen *et al*[21] was conducted about 15 years ago. During these years knowledge explosion and means of communication of knowledge have tremendously improved and possibly that is reflected in our study.

Gaps in knowledge have been noted in both health facilities. Parents in Muzaffarabad are relatively knowledge deficient than Kotli. Comparison of findings between Muzaffarabad and Kotli has revealed interesting facts. Particular areas include: role of genes in the disease pathology (32.6 *vs* 66.7% CR). This means that concept of origin of disease is not clear in parents. Parents with an understanding that thalassemia is not curable (32.6% *vs* 66.7% CR) may not assert adequately. If the parents believe that siblings do not have a chance to have thalassemia (46.5% *vs* 78.6% CR), recognition, management and prevention of thalassemia becomes difficult.

Specific nutritional elements are not required for the patients of thalassemia (53.4 *vs* 76.2% CR). Wrong information may divert the parents' efforts to arrange good food at the expense of appropriate management. An understanding that thalassemia can be transmitted through sex (20.9 *vs* 42.8% CR) can generate sex related problems among couples. One with the thought that thalassemia carriers don't lead a normal life (4.6 *vs* 26.2% CR) can generate unwanted anxiety among carriers. Blood transfusions, are important part of management (55.8 *vs* 72.6% CR) lack of this information may produce serious flaw in the management process. During the process of blood transfusion, reactions can occur which may be serious. Lack of this knowledge may be detrimental. Not having the information that thalassemia can be diagnosed by a blood test delays the diagnosis. If parents don't know that disease can be prevented, they will never agree for pre-marital counselling, anti-natal testing, and other preventive strategies. The high knowledge score in parents of thalassemia at Kotli were because of the presence of an established thalassemia centre.

Comparison of average correct responses for shared questions between our study and other studies[25] showed Italians (66.7%) Kotli (55.9%) Muzaffarabad (44.9%) Americans (30.8%) and Italian Americans (21%). Italy has implemented many programs to control thalassemia so their population is better educated and same holds true for Kotli. More exposure and more organized efforts result in better parent knowledge.

Widespread literature, education programs and systematic screening of β-thalassemia collectively increase the knowledge about the disorder and decrease the incidence of β-thalassemia. Screening program and prenatal diagnosis in Greece, Cyprus, Italy and Sardinia[26] lead to a decrease in birth rates of thalassemia affected children.

The survey questionnaire which was prepared on the basis of common problems faced during daily practices also motivated the parents to acquire more knowledge about various aspects of Thalassemia.

The outcome of our study shows potential intervention areas. Deficiencies in knowledge about occurrence of thalassemia, treatment modalities and misapprehensions about thalassemia minor and thalassemia major are prominent in our area of study.

Present findings are not only relevant to two health facilities in Azad Kashmir but are also applicable to other thalassemia centres providing care to the Thalassemia children in the region.

Models that have been successful in other places[27,28], e.g., Italy can be used to devise such programs in Azad Kashmir. Results of the present study warrant a need to do so. The best would be to implement an organized program.

CONCLUSION

Establishment of thalassemia centre improves the knowledge of parents of children suffering from thalassemia. There is a need to establish thalassemia centres at all major hospitals where dedicated staff must cater for their needs. General/high risk targeted awareness raising programs which should be culturally appropriate and linguistically adjusted will play a very important role in thalassemia prevention and treatment.

STUDY LIMITATIONS

Samples were not chosen randomly so there is a potential of bias. The answers to the questions were obtained from parents of thalassemia children so are not representative of knowledge of general population.

CONFLICT OF INTEREST

The authors have no conflict of interest to be declared.

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