ATTITUDE AND KNOWLEDGE OF MOTHERS:
ATTITUDE AND KNOWLEDGE OF MOTHERS WITH THALASSAEMIA MAJOR CHILDREN

ORIGINAL
PROF-1856

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ABSTRACT... **Objective:** The study explores attitudes and knowledge of mothers Thalassaemia Major children. **Design**: Cross sectional study. **Setting**: Thalassaemia center Military Hospital Rawalpindi. **Period:** The study was conducted from the 1st to the 30th August, 2011. **Subjects and Methods**: Fifty mothers were interviewed about the frequency of their child's blood transfusion and thalassaemia related complications, as well as their educational status, the education status of their husband, monthly household income, attitudes and knowledge about premarital screening, antenatal testing, and genetic inheritance of thalassaemia and complications resulting from multiple transfusions. **Results**: Mean age of thalassaemia patients was 9.5±5.5 years and 29(58%) were males and 21(42%) were females. Mean transfusion was 1.6±0.7 per month. Majority (52%) had stunted growth. 58 % of mothers were aware that premarital counseling could prevent disease. 60 % were aware that antenatal testing is available, and 40 % had undergone CVS for antenatal testing. Awareness about inheritance and hazards of blood transfusion was directly linked to maternal and paternal education (P values 0.08 and 0.05 respectively). Majority 30 (60 %) of mothers, irrespective of educational status, agreed that premarital screening should be mandatory. **Conclusion**: To improve patient awareness, instructional videos and audio lectures should be added to pamphlets already in circulation. Additional Facilities for premarital screening and counseling should be made available.

Key words: Thalassaemia , Prenatal testing, Premarital Screening.

INTRODUCTION

Thalassaemia is one of the most common genetic blood disorders in the world. There are approximately 240 million people worldwide who are heterozygous for β-thalassaemia and approximately 200,000 affected homozygzotes are born annually[1]. How ever Thalassaemia major(TM) was only described for the first time in the early 1920s by Detroit pediatrician named Dr. Denton Cooley. a Detroit pediatrician. The signs of the disease included anemia, a large spleen, and characteristic bone deformities. Until recently, the prognosis of homozygotes with thalassaemia major was dismal[2]. From the end of the 1950s transfusions were given to symptomatic patients when their hemoglobin reached extremely low levels. However these transfusions only prolonged life for several years before the patients died of infection or heart failure. The appearance of thalassaemic children in later years is well known: large heads, small noses, protruding zygomatic bones, distended abdomens and thin limbs. Beta-thalassemia major is a transfusion-dependent severe anemia requiring lifelong blood transfusions for the afflicted patients to stay alive. These children have reduced life expectancy, need for regular blood transfusions, daily medication, retardation in growth and sexual maturation. These are great challenges to TM patients and their families[3]. For example there are 3500 patients with severe beta thalassaemia major in Greece, and they consume one-quarter of the country's total blood supply. Iron chelation therapy for them costs 7 million dollars a year[4].

Great lessons can be learned from experiences of other developed countries. The biggest success story is of Cyprus.It was projected that if TM was not controlled, by the year 2000 the cost of treatment of these children would consume the entire health care budget of Cyprus and require all of the blood that the island's population could supply. Following compulsory premarital screening and optional prenatal testing, from 2000 to 2007 there were no TM births[5]. Unlike other countries with the disease, Pakistan has not had a study done on the feasibility of premarital screening. This study was carried out to see how Pakistani women of different socioeconomic backgrounds view mandatory premarital testing and antenatal diagnosis. It was aimed to assess knowledge about the inheritance of the disease, and the complications of multiple transfusions.

SUBJECTS AND METHODS

The study was conducted at Thalassaemia centre located in Department of Paediatrics Military hospital Rawalpindi from the 1st to the 30th August, 2011. In addition 201 patients are registered for transfusion at the center and many others come for treatment. An average of 6-10 transfusions are performed daily in the center. Patients between ages of 2-21 years were included in the study .Patients less than 2 years and those undergoing transfusion for other disorders like hemophilia were excluded. These children were being treated with frequent transfusions and long-term iron chelation therapy. After obtaining permission from the hospital ethical committee, the clinical data was collected by detailed interviews of the mothers with preformed questionnaires. Mothers were questioned about knowledge that premarital counseling could have averted the disease and that antenatal testing could have diagnosed the disease earlier. Their opinion was also sought regarding compulsory premarital testing. Knowledge about inheritance and hazards of transfusion was assessed as nil, fair and good on a scale of three and its correlation with social status and educational status of mother was analyzed. Evidence of complications such as of stunted growth, Hepatitis B and C status and spleenomegaly in affected children was noted.

Data was analyzed using SPSS 19.0 for Windows. Chi-square test of significance was used to see significance of association between knowledge and educational status and socio-economic status. P-value $\leqslant 0.05$ were considered significant.

RESULTS

Mean age of thalassaemia patients was 9.5±5.5 years and 29(58%) were males and 21(42%) were females. Mean transfusion was 1.6±0.7 per month. Majority (52%) had stunted growth. Majority of mothers were illiterate 35(70%) as shown in Table-III. 58 % of mothers were aware that premarital testing could prevent disease (Table-III). 60 % were aware that antenatal testing is available and 40 % had undergone CVS for antenatal testing(Table-III). Awareness about inheritance and hazards of blood transfusion was directly linked to maternal and paternal education (P values 0.08 and 0.05 respectively). Majority (60 %) of mothers, irrespective of educational status, agreed that premarital counseling should be mandatory, and the same percentage felt that antenatal testing was a reasonable option.

DISCUSSION

TM is associated with life-long transfusion-dependent anemia, short stature, facial abnormalities, delayed or

Table-I. Socio-demographic features of children with Thalassaemia Major n=50

Mean age (years)	**9.5±5.5**	**Minimum age = 3**	**Maximum age = 21**	
Sex	Male (n=29)	Female (n=21)		
	58%	42%		
Socioeconomic status	<10,000 (n=13) 44%	11-19000 (n=16) 32%	20-30000 (n=21) 42%	
Chelating agents	Oral (n=22) 44%	Parenteral (n=28) 56%	-	
Transfusion / month	1.6±0.7	Minimum = 1	Maximum = 4	
Complications	Student growth 26 (52%)	Hepatitis B/C 3(6%)	Spleenomegaly 8 (16%)	Combination of 2 or 3 13 (26%)

Table-II. Maternal and paternal educational status

	Illiterate	**Matric or less**	**FA or more**	**Total**
Mother	35 (70%)	10 (20%)	5 (10%)	100%
Father	17 (34%)	15 (30%)	18 (36%)	100%

Table-III. Maternal knowledge and attitude n=50			
	Yes	**No**	**Total (%)**
Premarital screening	29 (58%)	21 (42%)	100
Should it be mandatory	43 (60%)	20 (40%)	100
Prenatal testing	30 (60%)	20 (40%)	100
Personal experience with CVS	20 (40%)	30 (60%)	100
Inheritance	Nil s 10 (20%)	Fair 17 (34%)	Reasonable 23 (46%)
Knowledge about Hazards of transfusion	Nil 12 (24%)	Fair 23 (46%)	Reasonable 15 (30%)

Table-IV. Association of Knowledge with educational status .n=50					
Variables	**Illiterate**	**Matric or less**	**FA or more**	**P-value**	**Significance**
Premarital screening can reduce Thal	17	07	05	0.06	Marginal significant
Mandatory premarital screening should be offered	29	09	05	0.84	n.s.
Prenatal testing	16	10	04	0.005	Very highly significant
Personal experience of CVS	11	07	02	0.09	Marginal significant
Inheritance of thalassaemia	12	07	04	0.08	Marginal significant
Hazards of transfusion	07	04	04	0.05	Significant
**Chorionic Villus Sampling*					

Table-V. Knowledge about Thalassaemia major (intra-class comparison)					
Variables	**Low 7000**	**Lower middle 10000-19000**	**20-30,000 Middle**	**P-value**	**Significance**
Premarital counseling can reduce Thal	08	11	10	0.41	n.s.
Mandatory premarital counseling should be offered	12	13	18	0.64	n.s.
Prenatal testing	07	12	11	0.33	n.s.
Personal experience of CVS	05	07	08	0.93	n.s.
Inheritance of Thal	04	08	11	0.58	n.s.
Hazards of transfusion	05	04	06	0.47	n.s.
n.s. = Not significant		*CVS = Chorionic Villus Sampling*	*Thal = Thalassaemia*		

absent puberty, and attendant stigmas and psycho-social problems . Individuals with beta thalassaemia trait may be entirely asymptomatic, unaware of their diagnosis, and marry other individuals with the same trait, and there by perpetuate the disorder[4].

The couple that decides to undergo antenatal testing has an opportunity to choose whether to terminate or continue the pregnancy. For example, in Cyprus, Sardinia, Italy, and Greece[5] when the couple has been informed that the fetus is affected with beta thalassemia major, over 97 percent of the couples terminated the pregnancy to try again. The best way to reduce the burden of the disease is by compulsory premarital screening. This will markedly reduce the genetic burden in the next decades to come[6].

Premarital screening for genetic diseases is superior to neonatal screening since the former is primary prevention while the latter is secondary or tertiary prevention[7]. Premarital screening can potentially reduce the burden of inherited hemoglobin diseases by reducing the number of high-risk marriages. This was proven to be beneficial in nearby countries with similar endemicity levels of hemoglobinopathies[8,9]. Because of the burden on the healthcare system and effect on the quality of life in patients with β-thalassemia, premarital genetic screening was mandated in Saudi Arabia in 2004[10].

Mandating the premarital screening for inherited hemoglobinopathies was long awaited in Saudi Arabia and was preceded by religious and scientific debate[11]. In this highly conservative population, premarital screening in Saudi Arabia is the main preventive tool. In our study despite diverse socioeconomic and ethnic population majority were in favour of compulsory premarital testing.

The situation in Iran has also recently changed. Genetic counseling has generally had little or no impact on the choice of spouse. Separating and finding another spouse was the only option in Iran before prenatal testing was generally available and abortion of an affected fetus was permitted by the government[12]. On the other hand, once pregnancy has occurred, the couple will almost invariably seek prenatal diagnosis[13].This can be achieved before 13 weeks using chorionic villus biopsy.

In our study women even those with no formal education had personal experience of undergoing invasive testing in the form of CVS for antenatal testing universal antenatal screening for thalassaemia should be implemented in Pakistan. Mean corpuscular volume and haemoglobin should be used for screening [14]. As mentioned earlier, Premarital screening has also been found to be an acceptable even in conservative Saudia Arabia. Carrier screening at an early age (in school) and premarital screening programs aimed at identifying individuals before marriages may be more effective and culturally acceptable among our communities compared to prenatal diagnosis carried out in the antenatal clinic. Implementation of mandatory national premarital screening program, and screening young and unmarried women for detection of carriers have dramatically reduced the incidence of infants born with major thalassaemia in several countries worldwide [13]. For example, premarital screening to identify carrier couples and subsequently provision of screening in Iran has resulted in a 70% reduction in the annual birth rate of affected infants and a large amount of medical expenses [10]. Similarly, an earlier study in Montreal showed that high-schools students have high level of interest in thalassaemia screening, and participated at a rate of nearly 80%[16].

Components of a successful eradication program include education of public and physicians as well as community leaders. .Control of thalassemia requires treatment of the individual patient as well as a community-based educational effort to increase awareness of this problem In general, termination of pregnancy is not a consideration among many Asians because of a complex web of conservative values[15]. Additionally, a number of studies worldwide showed that attitudes toward prenatal diagnosis were related to religious convictions. Muslim couples, for instance, have been reported to refuse prenatal diagnosis on religious grounds. However in our study, the great majority were willing for invasive prenatal testing. .This trend was also shown in a Malaysian population survey. Only slightly more than one-third of the study participants supported selective termination of affected fetuses and the same was true for premarital screening[16].

Premarital screening is possible in Pakistan and majority of the participants in our study found it acceptable. Differences between this study and the Malaysian study[16] may be because the Malaysian study targeted the general population where as we targeted families with TM children. Families with TM children are more likely to be familiar with the consequences of the disease, and the burden its treatment poses for families and society.

Many illiterate and uneducated women in the current study, regardless of socioeconomic class, were unaware of basic aspects of the disease. A study from Karachi assessed awareness amongst TM families and found that knowledge concerning premarital screening was present in15%, prenatal testing in 5% and personal experience of CVS in 5%[17]. In our study these values were 58%, 30% and 20% respectively. The reason for better knowledge amongst mother was higher educational status of fathers in our study. Another reason for higher uptake of antenatal diagnosis was that it is offered free of cost to all armed forces families. Facility of bone marrow transplant was also available to some of the families free of cost by armed forces Bone Marrow Transplant Center.

A study examining parent knowledge about genetic transmission before and after educational videos proved such videos can be effective tools in educating the population[18]. Therefore, such videos and audio lectures should be produced in local languages to complement fliers and written materials already available.

CONCLUSIONS

To improve patient awareness among uneducated patients, instructional videos and audio lectures should be added to pamphlets already in circulation. Additional Facilities for premarital screening and counseling should be made available.

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Article received on: 03/10/2011 Accepted for Publication: 20/10/2011 Received after proof reading: 02/12/2011

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Article Citation:
Shukr F, Ali S, Yasmeen A. Attitude and knowledge of mothers with thalassaemia major children. Professional Med J Dec 2011;18(4): 703-708.

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AN IDEA THAT IS NOT DANGEROUS IS UNWORTHY OF BEING CALLED AN IDEA AT ALL.

OSCAR WILDE