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Demographic and Clinical Aspects In Thalassemic or Hemophilic Patients Referred to Pediatric Hospital in Tabriz City, 2004, Iran

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Abstract: Pediatric Hematology and Oncology (PHO) is an expanding field. Our aims were determining the needs and demands for comprehensive medical care of children suffering from chronic blood diseases, such as thalassemia or hemophilia and the study of demographic and clinical aspects of thalassemia and hemophilia patients referred to pediatric hospital and detection of relation of gender, educational rate and age, date of diagnosis, drugs, splenectomy, despheral utility and socio- economic position of the families. In a descriptive cross- sectional study, a total of 110 thalassemic and 70 hemophilic patients. Referred to pediatric hospital in Tabriz in 2004 was studied. The questionnaire included demographic and clinical aspects. Data were analyzed by SPSS software version 13. In this study, 65% of thalassemic patients were male and 98% were major type. The most of them (31%) had A^+ blood group. Fifty five percent of patients had major thalassemic sister or brother. Fifty three percnet of patients received infusion (IV) of desferal with blood infusion for 8 h, receive 5-10 ampoules each time. In 48% of patients were diagnosed the disease and beginning of infusion from 1-5 years. Eighty five percent of the patients which use infusion pump (S.C) for the injection of desferal for 8-12 h (3-5 day at week), receive less than 5 ampoules each time. All hemophilic patients were male and 82% of them had factor VIII deficiency. Incidence of the O^+ blood group type was 44%. In 38% of these patients, time of diagnosis was between the ages of 1-5 years. According to the results, studied aspects had efficiency on the thalassemic patients conditions ($p<0.05$). In the hemophilic patients, agents as blood factor VIII, blood group, familiar relations between father and mother, mother education level and living place were significant ($p<0.05$). Time of diagnosis ($p=0.88$), history of disease of ($p = 0.80$) and father education level ($p = 0.42$) were not significant. Also, the suitable time of referring to medical centers induce early diagnosis and their functions about therapeutic techniques, increases the longevity of patients.

Key words: Thalassemic, hemophilic, pediatric, desferal, blood factor

INTRODUCTION

In the recent years we have been able expand and optimize the PHO services throughout Iran, in general and in respect to their prevalence and clinical importance, by trained pediatric hematologist-oncologists, pediatric surgeons and improved Para clinical facilities (Abolghasemi *et al*., 2007; Mardawig *et al*., 2004).

Major beta-thalassemic receive blood transfusion and chelating therapy according to the current standards mostly at regional blood banks centers (Model *et al*.,1999) To curb major beta-thalassemia a premarital screening program has been done and abortion has been legitimized if major thalassemia is diagnosed by CVS. Hemophiliacs are supervised and treated as indicated by Iranian Hemophilia Comprehensive Care Centers (IHCCC). Screening for transfusion related complications and transmitted viral diseases (HBV, HCV and HIV) in both cohorts are carried out in regular intervals and necessary management will be then carried out as indicated at respective specialized units (Wolman *et al*., 1990; WHF Statement, 2006).

Hemophilia and thalassemia are inheritance diseases. The best way for prevention is genetic consulting. In the base of statistics of Iran thalassemia association, 2 million carriers and twenty thousand thalassemic patients are well controlled by genetic consulting pre- marriage programs and essential tests (Kao Antonio, 1997; Borugna *et al*., 2000). Hemophilia is a chronic bleeding disorder with reducing Factors rate. The main protocol for patients is prophylaxis and treatment IIIV or XI.

Prophylaxis with coagulation factors is expensive and utilization of this is only in the developed countries. Hemophiliacs are supervised and treated as indicated by Iranian Hemophilia Comprehensive Care Centers (IHCCC). Screening for transfusion related complications and transmitted viral diseases (HBV, HCV and HIV) are carried out in regular intervals and necessary management will be then carried out as indicated at respective specialized units (Evatt and Robillard, 2000; Propper and Cooper, 2001).

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Before the introduction of clotting factor preparations, the mean life expectancy of patients with hemophilia was less than 30 years and patients mostly died of intracranial or other hemorrhages. In patients with severe hemophilia not infected with viruses, mortality is still 40% higher when compared with the general population (FDA, 2006).

All over the world, the increased awareness of the importance of early diagnosis of genetic diseases has given them priority in primary health care.

Our purposes is determining the demographic and clinical aspects of thalassemic or hemophilic patients referred to the pediatric hospital and determining the needs and demands for comprehensive medical care of children suffering from chronic blood diseases and malignancies, such as thalassemia or hemophilia.

MATERIALS AND METHODS

This cross sectional study was done in Tabriz city, located at North-West of I ran for 110 thalassemic and 70 hemophilic patients in 2006 (In the mention of city population and p = 0.05). The age, gender, educational rate, blood group type, relationship, the age of diagnosis, existence of disease in other members of family , the kind of drug consumption, splenectomy, vaccination position and socio-economic position of these subjects were recorded by questionnaire. Values were expressed as prevalence rates or as the mean±1 Standard Deviation (SD). Conventional $\div^2$ and Fisher's exact test were used to analyze qualitative differences. Probability values of <0.05 were considered significant. Statistical analysis was performed with SPSS 13 statistical software.

RESULTS AND DISCUSSION

In this study, 65% of thalassemic patients were male and 98% were major type. The age of patients was 12±2.11 years. The most of them (31%) had A blood group. The most of parents had relationship (58%).

Fifty five percent of patients had major thalassemic sister or brother. Forty two percent of fathers had elementary educational level and 44% of mothers were illiterate. Fifty three percent of patients received infusion (IV) of desferal with blood infusion for 8 h (one time at week); receive 5-10 ampoules each time. In 48% of patients were diagnosed the disease and beginning of infusion from 1-5 years. Eighty five percent of the patients which use infusion pump (S.C) for the injection of desferal for 8-12 h (3-5 day at week), receive less than 5 ampoules each time. All of the patients received the blood each of 21 days (Table 1 and 2). All hemophilic patients were male and 82% of them had factor VIII deficiency. The age of hemophilia patients were 3±0.28 years. Incidence

Table 1: Demographical aspect of thalassemic children

Characterizations	Number	(%)	p-value
Genus			
Male	72	65	0.022
Female	38	35	
Blood group			
O+	30	27	0.0001
O-	6	5.5	
AB+	8	7	
AB-	2	2	
A+	34	31	
A-	2	2	
B+	22	20	
B-	6	5.5	
Relation of parents			
Near	64	58	0.001
Far	26	23	
Non	20	19	
Levels of social and economical of family			
Low	90	82	0.001
Middle	18	16	
High	2	2	

Table 2: Clinical aspects of thalassemic children

Characterizations	Number	(%)	p-value
Age of disease diagnosis			
Birth	2	2	0.01
>3 month	6	5	
3-6 month	24	22	
1-5 year	28	25	
>5 year	42	38	
>10 year	4	4	
	4	4	
Appearance of disease in family			
One sister or one brother major	26	54	0.016
Two sister or two brother major	4	8	
One sister or one brother minor	16	33	
Two sister or two brother minor	2	4	
Age of beginning of desferal injection			
>6 month	8	8	0.001
6-12 month	8	8	
1-5 year	48	48	
5-10 year	32	32	
>10 year	14	14	
Splenectomy			
Yes	20	18	0.001
No	90	82	
Vaccination			
Yes	90	82	0.001
No	20	18	

Table 3: Demographical aspects of hemophilic children

Characterizations	Number	(%)	p-value
Blood group			
O+	24	44	0.027
O-	5	9	
AB+	13	9	
AB-	-	-	
A+	13	24	
A-	4	7	
B+	4	7	
B-	-	-	
Relation of parents			
Near	10	18	0.001
Far	10	18	
Non	35	64	
Levels of social and economical of family			
Low	45	81	0.001
Middle	8	15	
High	2	4	

Table 4: Clinical aspects of hemophilic children

Characterizations	Number	(%)	p-value
Age of disease diagnosis			
3-6 month	17	31	0.886
6-12 year	12	22	
1-5 year	18	33	
>5 year	8	14	
Appearance of disease in family			
Yes	30	55	0.016
No	25	45	
Splenectomy			
VIII	45	82	0.001
IX	10	18	

of the O^+ blood group type was 44%. In 31% of cases, fathers had elementary education. Fifty three percent of mothers were illiterate and 55% of patients' parents were not relatives. In 38% of these patients, time of diagnosis was between the ages of 1-5 years (Table 3 and 4).

All over the world, the increased awareness of the importance of early diagnosis of genetic diseases has given them priority in primary health care (Issaragrisil, 2001). The objectives of the study, were showing the position of thalassemic or hemophilic patients due to treatmental programs and enhancing the knowledge of parents about diseases. In the mention of results, studied aspects had efficiency on the thalassemic patients health conditions ($p<0.05$). In the hemophilic patients, factors as blood factor VIII, blood group, familiar relations between parents, mother education level and living place were significant ($p<0.05$). Other factors such as time of diagnosis ($p = 0.88$), history of disease of ($p = 0.80$) and father education level ($p = 0.42$) were not significant.

Results were well corresponded with some reports: Yazdi *et al.* (1998) showed 64% of thalassemic patients were male and Sadati *et al.* (2004) reported 60% of patients were male and 40% were female.

The most of ills had relative parents (58%) and was similar to Warwick and Modi (1997). Blood group detection is very important to Blood bank programs for providing packed cell and blood factors for patients. Fifty four percent of patients had thalassemic sister or brother and it is important for the new cases prevention programs (Sher *et al.*, 1995).

CONCLUSION

Therefore, educational programs for patients increase awareness about diseases topics and prevent the new cases birth of thalassemic in the future. Also, the suitable time of referring to medical centers induce early diagnosis and their functions about therapeutic techniques, increases the longevity of patients.

In the mention of results, we can prevent enhancing of disease with educational programs for relatives and pre marriage tests. Also, time of referring to the clinic for diagnosing of diseases has importance for prognosis.

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