

STUDY OF SEX , AGE AND BLOOD GROUPS (ABO, Rh) DISTRIBUTIONS IN THALASSEMIA PATIENTS IN IRAN

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Abstract

Thalassemias , because of climatic , geographic and ecological conditions , are the most common among the genetically endemic diseases in Iran, specially in provinces adjacent to the Caspian sea and the Persian gulf. There are over 14,000 cases of thalassemia major reported in Iran.

Data , collected by the Iranian Ministry of Health , and analysed at the Department of Human Genetics , School of Public Health , Tehran University of Medical Sciences , showed relative distribution of 3194 patients referring for the iron chelating drug , disferal , in 24 provinces in Iran.

3304 cases were studied for sex and age groups. Higher percentages and sex ratios were observed in each age group and further clarified as the age increased.

3386 cases were considered for ABO and Rh blood groups. Significant high incidence of group O (41.228%) was followed by groups A (29.090%) and B (23.213%) , and group AB with the lowest (6.467%). A significant low incidence of Rh negative was also observed (6.852%).

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Introduction

Thalassemias are a heterogeneous group of heritable hypochromic anemias of different degrees of severity. Thalassemia major, also known as Cooley's anemia, becomes progressively symptomatic during the second 6 months of life. Cells are characterised by absent or deficient synthesis of one or more globin chains. The hematopoietic system responds by producing tremendous numbers of new red blood cells, but with little hemoglobin and fragile membranes, becoming ruptured upon passing through the tissues (2,5).

Iron accumulation resulting from increased gastrointestinal absorption stimulated by the anemia, blood transfusion and decreased utilisation for hemoglobin synthesis. Excess iron deposition, in addition to change of skin colour and bones, specially in head and face region, causes damage to the heart, pancreas and other tissues (5).

Thalassemias due to climatic, geographic and ecological conditions, are the most common among the monogenic (autosomal recessive) endemic diseases in Iran and the region. At the present time there are over 14,000 cases of thalassemia major in Iran, which is probably the highest in the world, and the number is growing. Consanguinous marriages are probably have increasing effects in this autosomal recessive disease. The highest frequency is in provinces adjacent to the Caspian sea and the Persian gulf. Relative high frequency is also observed in the central/south of the country (Fars province), considered the continuation of the Persian gulf area.

Problems caused by thalassemia are shortening of life expectancy, expenditure for treatment, inefficiency of life, psycho-social burden for the patients, their families and the society (1).

The annual national requirement for optimal treatment for existing patients are US\$ 60 million for iron chelating drug Disferal, in addition to infusion pumps, disposables and 300,000 units of blood, which are estimated to be rising by 10% or more each year (4). Therefore, there is a strong need for education, prevention, control and prenatal diagnosis, to decrease the incidence.

The present study is to find the numbers and percentages of disferal receiving patients in each province in Iran, distribution in different sex and age group and association with ABO and Rh blood groups.

Materials and methods

The crude data of this study was collected by the Office of Genetic Disease, Department of Prevention of Diseases, the Iranian Ministry of Health, Treatment and Medical Education, from the public health centers, such as PHC networks, etc., at the urban, sub-urban and rural levels.

The information was then categorized by provinces, age, sex, ABO and Rh blood groups and analysed at the Department of Human Genetics, School of Public Health, Tehran University of Medical Sciences.

Results and discussion

Table 1 shows the numbers and percentages 3194 disferal receiving patients by relative distribution in 24 provinces. This distribution is necessarily in agreement with the distribution in each province of Iran.

Table 2 shows the distribution of 3304 cases in different sex and age groups. Higher percentages and sex ratios are observed in males in each age group and it becomes further clarified as the age increases. It needs further investigation to determine the causes.

Table 3 shows the ABO and Rh blood group distribution in 3386 cases, receiving blood transfusion, according to the information obtained by the Iranian Thalassemia Association, at the time of this investigation, as compared to 291,857 control individuals (3). Statistical analyses show that the highest incidences are in group O (41.228%) followed by groups A (29.090%) and B (23.213%) and group AB has the lowest incidence (6.467%), in a highly significant level. A significant high incidence of Rh negative was also observed (41.228%).

Table 1 - Provincial distribution of 3194 disferal receiving thalassemia cases in Iran

Province	n	%	population of provicne
Tehran	432	13.525	9,982,309
Markazi	31	0.970	1,182,611
Gilan	231	7.232	2,204,047
Mazandaran	327	10.237	3,793,149
E. Azarbaijan	13	0.407	4,420,343
W. Azarbaijan	40	1.252	2,284,208
Kermanshab	98	3.068	1,622,159
Khuzestan	328	10.269	3,175,852
Fars	519	16.249	3,543,828
Kerman	352	11.020	1,862,542
Khorasan	40	1.252	6,013,200
Esfahan	93	2.911	3,682,444
Sistan & Baluchestan	203	6.355	1,455,102
Kordestan	11	0.344	1,233,480
Hamadan	36	1.127	1,651,320
Charmahal & Bakhtiati	15	0.469	747,297
Lorestan	41	1.283	1,501,778
Ilam	68	2.128	440,693
Kohkiluyeh & Boyr-Ahmad	83	2.598	496,739
Bushehr	170	5.322	694,252
Zanjan	6	0.187	1,776,133
Semnan	12	0.375	458,125
Yazd	32	1.002	691,119
Hormozgan	13	0.407	924,433
Total	3194	100%	55,837,163

Table 2 - Age groups distribution and sex ratios of 3304 thalassemia cases in Iran

Age group	Female		Male		Total		M/F
	n	%	n	%	n	%	
0-4	74	2.239	104	3.147	178	5.387	140
5-9	599	18.129	760	23.002	1359	41.131	127
10-14	485	14.679	617	18.674	1102	33.353	127
15-19	197	5.962	227	6.870	424	12.832	115
20-24	48	1.452	77	2.330	125	3.783	160
25-29	26	0.786	30	0.907	56	1.694	115
30-34	13	0.393	15	0.453	28	0.847	115
35-39	7	0.211	8	0.242	15	0.453	114
40-44	1	0.030	5	0.151	6	0.181	500
45-49	1	0.030	3	0.090	4	0.121	300
over 50	2	0.060	5	0.151	7	0.211	250
Total	1453	43.971	1851	56.017	3304	100%	

Table 3- ABO and Rh blood group distribution of 3618 thalassemia cases in comparison with control individuals in Iran

Blood groups	N	A		B		AB		O		Rh-		X ²	P
		n	%	n	%	n	%	n	%	n	%		
Thal	3386	985	29.090	786	23.213	219	6.467	1396	41.228	232	6.852	87.86	0.000
Control	291,857	93,803	32.14	69,228	23.72	23,874	7.79	104,777	36.35	30,295	10.38		

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Thalassemia Control in Iran

The Ministry of Health , Treatment and Medical Education has established an active prevention program for beta thalassemia minor , starting 1996. According to this program , couples who have planned to get married will be mandatory screened throughout the country. The carrier couples , prior to signing the marriage contract will be referred to the counselling centers to be informed about the risk , promote family planning and , if possible , try to separate such candidates.

This program will also take the affected persons in to consideration for treatment and their family members will be tested and carriers referred to the counselling centers.

The carrier couples who do not have affected child / children will also be included in this plan.

It is hoped that this program will raise the society's level of information and knowledge , with the aim of preventing the appearance of new cases of thalassemia major in Iran.