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PREVALENCES OF COAGULOPATHIES IN DIFFERENT PROVINCES OF IRAN

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Abstract

Prevalences of coagulopathies (n = 6010) including factor VIII deficient (hemophilia A) individuals (n = 3205) were investigated and analysed in 24 provinces of Iran.

For all coagulopathies, the prevalence (per 100,000) was the highest with 24.45 in Semnan (north east of Iran), followed by Hamadan (west) with 17.56. The lowest was observed in Kordestan (west) as 0.88, followed by Kohkiluyeh & Boyr-Ahmad (center) and Esfahan (center) as 1.20 and 1.32, respectively.

For hemophilia A, the highest was observed also in Semnan as 14.19, followed by Hamadan as 9.81. The lowest was observed in Hormozgan (south east) as 0.17, followed by Kohkiluyeh & Boyr-Ahmad as 0.20.

The analyses showed that a higher prevalence existed above a hypothetical geographical central line, from west to east of Iran ; a lower prevalence was observed below the line.

More extensive epidemiological investigations are needed to establish and explain the geographical gradient of the coagulopathies in Iran.

Introduction

Coagulopathies, as single gene defects, are the second priority, after thalassemia, for maintenance, treatment and prenatal diagnosis in Iran. Hemophilia A, as an X linked genetic disease, being the most common coagulation disorder, with an incidence of about 1-2 in 10,000 males, is casued by mutations in the factor VIII coagulation gene (6). Families have been observed with numbers of affected individuals; one such family had 13 patients, 18 obligate carriers and 14 females as possible carriers of hemophilia B, in four generations, as the

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largest reported pedigree so far in Iran (8).

One form of serious complecations is that some hemophiliacs become refractory to therapy by the given factors, because a circulating inhibitor or antibody forms (3,12,17,21). Also, factor VIII auto-antibody inhibitors are rare but may present massive and life threatening hemorrhages, mainly in older individuals (5,1). Acquired or induced von Willerbrand's disease is rare but some cases have been reported (11).

Obstetric management and precautions must be considered under special attention for carriers of certain coagulopathies. Studies on pregnancy, labour and the puerperium, have shown the risks and the importance of indication for special care.

Prenatal diagnosis should be considered for affected cases (14,15,16) ie. it is now possible to identify a fetus with hemophilia by examining DNA polymorphisms in amniotic fluid fibroblasts.

Patients with bleeding disorders present life threatening problems of upper gastrointestinal tract bleeding; one newly emphasized cause is the infection by *Helicobacter pylori*. It has been found that gastric and duodenal ulcers were diagnosed more often in patients with bleeding disorders (4). Intraarticular bleeding (hemarthrosis) in hemophiliacs starting in early childhood is a well acknowledged problem (23).

Some of the coagulopathies require certain therapeutic procedures such as coagulation factors or blood transfusions that may cause serious complecations. Contaminations with hepatitis viruses (9,11,19,20) have been proven to be serious problems. Mortality and morbidity of transfused patients have been increased by introduction of human immunodeficiency virus (HIV) (10,18,22,24).

These issues present the necessity to determine the prevalence of the disorders and provide the grounds for fundemental search in the geographical and ethnological differences in the Iranian population.

This study was performed to find the numbers and prevalences of coagulopathies as a whole, and factor VIII (hemophilia A), as the main type, in different provinces of Iran.

Materials and methods

This study was performed on patients with different types of coagulopathies who have been registered in the Haemophilia Center of Iran. Data were taken from the records available in this center. Exact final diagnoses were confirmed by clinical as well as laboratory findings (25). Total numbers of patients with coagulopathies (n = 6010) including factor VIII deficiencies (n = 3205) were considered for analyses.

Results and discussion

Table 1 shows the numbers and prevalences of 6010 individuals affected with different types of coagulopathies, and 3205 factor VIII (hemophilia A) patients in particular, as they consist of the highest proportion of the group, in 24 provinces of Iran.

Among the coagulopathies, factor VIII deficiency had the highest percentage (53.33%) followed by factor IX deficiency (11.21%), von Willebrand's disease (9.15%), platelet deficiency (7.42%); all other types of coagulation disorders, consisted of 18.72%. Compared to the investigation in 1987 (7), a decrease of von Willebrand disease would suggest better awareness of the population about the risks of consanguinous marriages.

In a similar study in the United States (n = 2743), an estimation shows a national population of 13,320 cases of hemophilia A, and 3,640 cases of hemophilia B (25).

As it is shown in table 1, the prevalence (per 100,000) was the highest with 24.45 in Semnan (north east of Iran), followed by Hamadan (west) with 17.56, and the lowest was observed in Kordestan (west) as 0.88, followed by Kohkiluyeh & Boyr-Ahmad (center) and Esfahan (center) as 1.20 and 1.32, respectively.

Factor VIII deficiency was also observed to be the highest in Semnan with prevalence of 14.19, followed by Hamadan as 9.81, and the lowest in Hormozgan (south east) as 0.17, followed by Kohkiluyeh & Boyr-Ahmad as 0.10.

It is obvious that there is almost a total geographical conformity in these two groups because a large proportion (53.32%) of all coagulopathies is the factor VIII deficiency.

The analyses of the prevalences of both groups showed that the incidence presents almost a hypothetical geographical central line from west to east of Iran, the highest in the north of the line, and the lowest below the line.

More extensive epidemiological investigations are necessary to establish and explain the geographical gradient of the coagulopathies in Iran.

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Table 1- Provincial distribution and prevalence (per 100,000) of 6010 individuals with different types of coagulopathies, including 3205 factor VIII patients, in Iran.

Province	All Coagulopathies		Factor VIII		Population* of province
	N	Prevalence	N	Prevalence	
Tehran	1489	14.91	805	8.06	9,982,309
Markazi	173	1.46	99	8.37	1,182,611
Gilan	262	11.88	140	6.35	2,204,047
Mazandaran	366	3.64	164	4.32	3,793,149
E.Azarbaijan	489	11.06	252	5.70	4,420,343
W.Azarbaijan	142	6.21	75	3.28	2,284,208
Kermanshah	223	13.74	138	8.50	1,622,159
Khuzestan	362	11.40	197	6.20	3,175,852
Fars	160	4.51	93	2.62	3,543,828
Kerman	166	8.91	81	4.35	1,862,542
Khorasan	535	8.89	278	4.62	6,013,200
Esfahan	487	1.32	234	6.35	3,682,444
Sistan & Baluchestan	66	4.53	29	1.99	1,455,102
Kordestan	102	0.82	67	0.54	1,233,480
Hamedan	290	17.56	162	9.81	1,651,320
Charmahal & Bakhtiari	21	2.81	13	1.74	747,297
Lorestan	132	8.79	82	5.46	1,501,778
Ilam	22	4.97	2	0.45	440,693
Kohkiluyeh & Boyr-Ahmad	6	1.20	1	0.20	496,739
Bushehr	23	3.31	11	1.58	694,252
Zanjan	164	9.23	107	6.02	1,776,133
Semnan	112	24.45	65	14.19	458,125
Yazd	102	14.76	58	8.39	691,119
Hormozgan	43	4.65	16	0.17	924,433
Total	6010		3205		55,837,163

* The populations of provinces were taken from the 1975 census (13).

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