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REVIEW ARTICLE

A review of abnormal hemoglobins in Turkey

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ABSTRACT

In this review, abnormal hemoglobins published in Turkish population during the last four years are presented. Further, analysis of the 88 abnormal hemoglobins is geven.

Key Words: Abnormal hemoglobin

ÖZET

Türk popülasyonunda yeni belirlenen anormal hemoglobinler

Türk popülasyonunda yapılan çalışmalarda çok sayıda yeni anormal hemoglobinler tanımlanmıştır. Bu gözden geçirme makalesinde son dört yılda yayınlanan anormal hemoglobinlerle, moleküler analizi yapılan 88 olgunun dökümü verilmektedir.

Anahtar Sözcükler: Anormal hemoglobin

An extensive review by Altay concerning the "Abnormal Hemoglobins in Turkey" appeared in the journal four years ago[1]. Since then, several other variants have been reported in both international and national journals. The aim of this mini-review was to compile the newly published abnormal hemoglobins in the Turkish population since Altay's paper[2-11] (Table 1)[12,22].

For the last five years, 89 variants other than Hb S, each belonging to one family, were referred to our laboratory for further molecular analysis. DNA of these samples was isolated with the standard phenol-chloroform extraction method. Non-radioactive fluorescence dye-based DNA sequencing of beta and alpha globin genes was performed using BECKMAN Coulter CEQ8000 genetic analysis system as described previously[5]. Our data concerning the variants referred for molecular analysis is given in Table 2.

Altay pointed out that the exact number of subjects having abnormal hemoglobins in Turkey is not known due to the absence of a national registry system for these conditions[3]. This aim can be achieved under the auspices of the Turkish Hematology Association.

Table 1. Abnormal hemoglobin variants in the Turkish population published since 2002

Name	Structure	Reference
a. Variants of the alpha chain (single base changes)		
Hb Setif	[a94 (G1) Asp-Tyr]	2
Hb Q-Iran	[a 75 (EF4) Asp-His]	18
Hb Hasharon	(a47 Asp → His)	11
Hb Bronovo	[a103 (His → Leu]	19
b. Variants of the beta chain (single base changes)		
Hb C	(B6 Glu-Lys)	14
Hb E Saskatoon	(B22 Glu-Lys)	14
Hb Volga	[beta 27 (B9) Ala-Asp]	3
Hb Siirt	[beta 27 (B9) Ala-Gly]	4
Hb Hamadan	[B 56 (D7) Gly-Arg]	5
Hb Pyrgos	[B 83 (EF7) Gly-Asp]	6
Hb D Punjab	[B121 Glu-Gln]	14
Hb Beograd	[B121 Glu-Val]	7
Hb G-Coushatta	[B22 (B4) Glu-Ala]	8,14,20
Hb J-Iran	[B77 (EF1) His-Asp]	8,16,17,21
Hb Tyne	(B5 Pro-Ser) and Hb S (B6 Glu-Val)	9
Hb G-Copenhagen	(B47 Asp-Asn)	11
Hb D-Iran	(B22 Glu-Gln)	11
c. Variants of the delta chain (single base changes)		
Hb A2 Yialousa	(D82 C-T Ala28Ser)	15
d. Hybrid Hbs		
Hb Lepore Boston		10,11
e. Abnormal hemoglobin variants that have been reported in compound heterozygote state with thalassemia or sickle cell		
Hb Hamadan	[B 56 (D7) GLY-ARG]-beta thalassemia	5
Hb D Punjab	(B121 Glu-Gln)/Hb S	13
Hb G-Coushatta	[B22 (B4) Glu-Ala]/Beta thalassemia	20
f. Homozygosity of Hb variants		
Hb C	(B6 Glu-Lys)	12
Hb D Punjab	(B121 Glu-Gln)	22
Hb Hamadan	[B 56 (D7) GLY-ARG]	5
Hb Q-Iran	[a75 (EF4) Asp to His]	18

Table 2. Our data concerning the variants other than Hb S referred for molecular analysis, each belonging to one family

Variant	n
Hb D Punjab (B121 Glu-Gln)	35
Hb O Arab (B121 Glu-Lys)	23
Hb E Saskatoon (B22 Glu-Lys)	5
Hb Lepore	4
Hb C (B6 Glu-Lys)	4
Hb E (B26 Glu-Lys)	3
Hb J Iran (B77 (EF1) His-Asp)	3
Homozygous Hb D Punjab (B121 Glu-Gln)	2
Hb D Punjab (B121 Glu- Gln)/Hb S compound heterozygous	2
Hb Hamadan (B 56 (D7) GLY-ARG)	2
Hb Beograd (B121 Glu-Val)	2
Hb Pyrgos [B 83 (EF7) Gly-Asp]	1
Homozygous Hb Q Iran (a75 (EF4) Asp to His	1
Unknown	2

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