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G6PD Deficiency in Turkish Cypriots

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Abstract: 1108 Turkish Cypriot men and 318 male labourers from mainland Turkey were screened for G6PD deficiency and haemoglobinopathy traits. The results revealed a 6.7% G6PD deficiency rate in the Turkish Cypriot men and a 1.6% prevalance rate in the Turkish men. The mean haemoglobin level of the G6PD deficient males was approximately 1g/dl lower than that of the non-deficient males.

Key Words: G6PD deficency, thalassaemia.

Introduction

The geographical distribution of G6PD deficency correlates closely with that of the haemoglobinopathies. There is good experimental evidence from in vitro studies of the mechanism of protection against malaria. Parasites exhibit impaired growth in G6PD-deficient cells both in hemizygous males and heterozygous females (1, 2).

The main disadvantages of G6PD deficiency in hemizyous males are an increased risk of severe neonatal jaundice, due to slightly increased haemolysis, and favism, which has been common in eastern Mediterranean people for a long time (3-5). Favism is a term used to describe acute haemolytic anemia after the consumption broad beans (Vicia faba) - a very common food in Cyprus.

A few previous studies of Cypriots have identified G6PD deficiency in 3 to 12% of those tested (6-9). These figures are surprisingly low compared to the 16% beta thalassaemia carrier rate (10) since the proportion of a population heterozygous for beta haemoglobinopathy traits is usually similar to the percentage of males hemizyogus for G6PD deficency (11-13). Because of this surprising relationship and uncertainty over the real figures, this study of the prevalence of G6PD deficency in Turkish Cypriots was performed.

Materials and Methods

1108 Turkish Cypriot men who came to the thalassaemia laboratory for premarital thalassaemia-carrier screening were also screened for G6PD deficency. As premarital heterozygote testing is mandatory in Northern Cyprus, they represented a random sample of the population. In addition, 318 male labourers from mainland Turkey, applying for work permits in Northern Cyprus and so obliged to undergo HIV tests, were also screened for G6PD deficiency and haemoglobinopathy traits. Haemoglobinopathy traits were diagnosed using standard methods (14-16). G6PD deficiency was diagnosed by electrophoresis of lysed whole blood samples on cellulose acetate at pH 8.6, and incubation for twenty minutes at 37°C with G6PD reagent (7.6 mM glucose-6-phospate; 1.6 mM NADP; 2.8 mM MTT; 0.3 mM phenazine methosulphate-Helena Laboratories, Texas), which forms a purple complex withthe enzyme band (17).

Results

The result are summarized in Tables 1 and 2. Seventy-five of the 1108 Turkish Cypriot males (6.7%) were G6PD deficient, and 1033 were normal (having electrophoretic mobility which was the same as the normal Mediterranean B variant). The G6PD-deficient males were told that they have a genetic enzyme deficiency which can cause haemolysis when they take certain drugs and a list of drugs to avoid was provided. It was explained that the condition is passed from mother to son.

The mean haemoglobin level of G6PD-deficient males was approximately lg/dL lower than that of the non-deficient males, though the red cell indices did not differ

Males	Number	G6PD Deficient	β-thal trait	HbS trait
T.Cypriot	1108	75* (6.7%)	176*(15.9%)	-
Turkish	318	5**(1.6%)	8**(2.5%)	3(0.9%)

* 26subjects had both β-thalassaemia trait and G6PD deficiency
** One subject had both β-thalassaemia and G6PD deficiency

Table 1. G6PD status and haemoglobinopathies in Turkish Cypriot males and Turkish labourers.

G6PD status	β-thalassaemia status							
	Normal				β-thalassaemia trait			
	No.	Hb(gr)	P	SD	No.	Hb/gr)	P	SD
Normal	883	14.8	<0.001	±1.25	150	13.5	>0.1	±1.04
Deficient	49	13.7		±1.20	26	13.5		±1.20

Table 2. G6PD deficiency and β-thalassaemia traits in male Turkish Cypriots in relation to their haemoglobin levels

between the two groups. Twenty-six (35%) of the Cypriot G6PD deficient males also had a beta thalassaemia trait. 1.6% of the males from Turkey were G6PD deficient, 2.5% had a β-thalassaemia trait and 0.9% had a Hb S trait. One had both G6PD deficiency and a β-thalassaemia trait.

Discussion

This study confirms the findings of previous studies done in Cyprus, showing a prevalence of G6PD deficiency in males of 6-7%. The reduced haemoglobin levels of the G6PD-deficient males studies is consistent with the fact that in G6PD-deficient males the red-cell lifespan is a little shorter and haemolysis is slightly greater than in normal individuals (11).

The thalassaemia-trait prevelance rate found in this study was 15.9%. The finding that the frequency of males hemizygous for G6PD deficiency is less than half that of heterozygotes for β-haemoglobinopathies was unexpected, because the prevelance of the two conditions is similar in many populations. In both cases, the prevalence represents a balance between selective advantage in protecting against falciparum malaria, and childhood mortality due to the disorder. The homozygous haemoglobinopathies are almost uniformly fatal, while G6PD deficiency can lead to death from neonatal jaundice or acute haemolitic crisis: the frequency with which these occur being strongly influenced by environmental factors. The fact that the prevalance of male hemizygotes for G6PD deficiency corresponds fairly closely to the prevalence of heterozygotes for beta haemoglobin disorders in many populations suggests that the two conditions have rather similar selective advantages and disadvantages (18).

However, Cyprus is an exception and the reason is not clear. It is conceivable that environmental factors have increased the selective disadvantage of G6PD deficiency in Cyprus, where fava beans have been a very common food for a long time. At this stage it is impossible to obtain objective data, but favism is well-known in Cyprus and was often fotal in the past due to lack of medical facilities. In recent times, there has been an effort to reduce the frequency of favism by advising mothers not to feed fresh fava beans to their children. Nevertheless, during the spring when broad beans are readily available, some children are still admitted to hospital with haemolytic crises.

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