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LETTER TO THE EDITOR

Hemoglobinopathies in Turkey

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Turkey is settled at a crossroads of the earth's continents, so migrations throughout the world, especially between Asia, Europe and Africa, have affected Turkey in numerous ways, including in the evolution of hemoglobin disorders. In the middle of the 1940s, many thalassemic patients and abnormal hemoglobins began to be diagnosed at Istanbul University, and the pioneering works of Prof. Aksoy in the second half of the last century were of great significance in this field. Investigations into the hemoglobinopathies and thalassemias have been performed in many different areas of Turkey and are continuing, but most of these surveys are restricted to local regions. These studies have pointed out that these disorders are especially cumulated in the central southern part of Turkey.

Thalassemia surveys were pioneered by Aksoy *et al.* [1,2]. Arcasoy *et al.* found the β-thalassemia incidence as 2.1% in a healthy population in 1978 [3]. In accordance with the data given by the Ministry of Health and by Altay [5] (cumulated from local survey results of different areas of Turkey between 2000-2006), the incidence of β-thalassemia and abnormal hemoglobins has been reported as approximately 4.3%, ranging from 0.6% to 13.0% in different surveys [4,6]. The incidence surveys are shown in Table 1.

The surveys investigating incidences and frequencies were done in different populations, some in cord blood samples, some in premarital screening procedures and some in selected areas known to have high incidence of hemoglobinopathies and thalassemia. In screening programs, attention must be given to the population selection. Screening of hospital records or calculation of percentages of cumulated patient populations should not be used for prevalence/incidence surveys or reports to avoid confusion. Premarital screening programs could be useful in the beginning for identifying couples at risk of having affected children.

The outcome of hemoglobin disorders and thalassemia may be predicted by way of molecular, biological and clinical studies and prevention schedules may run normally [6-8]. In addition to the large screening programs, detection of these blood disorders at birth, such as Hb S, may reduce the risk factors and complications. At this point, prenatal diagnosis has prime importance in the prevention and eradication of hemoglobin disorders. In 1982, Prof. Altay started prenatal diagnosis at Hacettepe University, and it was later extended to Boğaziçi, Çukurova, Akdeniz and Ege Universities. To date, about 1700 chorionic villous sampling (CVS) procedures have been performed and about one-fourth of them were diagnosed as homozygous for thalassemia or hemoglobinopathies and the pregnancies interrupted with medical abortion [9-12]. Most (70%) of the prenatal diagnosis procedures have been performed at Çukurova, Akdeniz and Hacettepe Universities. This means that prevention programs must first be aimed at the Çukurova and Antalya regions. Preimplantation genetic diagnosis procedures were also recently started

Table 1. The incidence surveys of α- Thalassemia, β- Thalassemia, Hb S and Hb D in regions of Turkey

Region	*α-Thalassemia* %	*β-Thalassemia* %	*Hb S* %	*Hb D* %
Çukurova region	3.3[1*]	2.3-4.6	3-47	0.3
Southern Turkey	8	2.4-13.1	2.5	-
Eastern Turkey	-	0.7-3.6	0.5	-
Aegean region	-	2.6-5.1	0.5	-
Marmara region	-	3.4-11.7	2.5	-
Central Anatolia	3.6[2**]	2.1	-	-

1 *Column chromatography [15]
2 **DNA analysis [16]

in Turkey. At the end of the 1980s, bone marrow transplantation in thalassemia and hemoglobinopathies began. Currently, many stem cell transplantation centers are working actively in Turkey and thalassemia-free survival after stem cell transplantation was reported as 74.1% by the Turkish BMT Study Group [12].

In Turkey, the biggest challenge, with respect to the physiological, psychological, socio-economical and medical impacts of hemoglobin disorders, is sickle cell anemia, with its associated complications. The patients are cumulated mostly in the Çukurova region [13-15]. HbS is the most frequently seen disorder in hematology follow-up clinics and also in general practice in Turkey. Sickle cell disease in homozygous HbS condition has various clinical outcomes ranging from mild to severe clinical courses, even between children in the same family, and some cases result in very serious complications [6,13,14]. Thalassemia could be managed by regular treatment programs routinely and safely, but in sickle cell anemia the patient can be lost to sudden death.

To address the problems of hemoglobinopathy disorders, the Turkish Ministry of Health's Department of Mother and Child Health has organized a thalassemia prevention program in 33 cities. The Turkish Government has begun to offer premarital screening programs for couples on a voluntary basis in selected areas with high population frequencies of hemoglobin disorders and thalassemia. Individuals of reproductive age are educated regarding the risks associated with consanguineous marriages and are screened for these disorders. Prenatal diagnosis and follow-up programs may also help to prevent the birth of patients with severe clinical courses. The Turkish Government should provide insurance coverage for the prevention programs.

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