

Neslihan Erdem¹, Ahmet Çizmeçioğlu², İsmet Aydoğdu³

¹Celal Bayar University Faculty of Medicine, Department of Internal Medicine, Manisa, Turkey

²Karaman State Hospital, Clinic of Internal Medicine, Karaman, Turkey

³Celal Bayar University Faculty of Medicine, Department of Hematology, Manisa, Turkey

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Address for Correspondence: Neslihan ERDEM, M.D.,

Celal Bayar University Faculty of Medicine, Department of Internal Medicine, Manisa, Turkey
Phone: +90 555 729 88 22

E-mail: nesliherdem@gmail.com

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First Observation of Hemoglobin Kansas [β 102(G4)Asn→Thr, AAC>ACC] in the Turkish Population

Türk Toplumunda İlk Hemoglobin Kansas [β 102(G4)Asn→Thr, AAC>ACC] Gözlemi

To the Editor,

Hemoglobin (Hb) Kansas [β 102 (G4) Asn→Tyr, AAC>ACC] is an unstable abnormal hemoglobin with low oxygen affinity and increased dissociation. Hb Kansas has rarely been reported in the literature to date; the first case was defined in the state of Kansas of the United States [1]. The second reported case was a newborn baby with cyanosis from Sarajevo and the third was an elderly patient with polycythemia from Japan [2,3]. There has been no previous report from Turkey [4]. We herein report the first case of Hb Kansas from Turkey, an introduction of clinical significance.

Case: A 28-year-old male patient with cyanosis of the lips and fingertips was admitted to a hospital in the city of Malatya. He had peripheral cyanosis of the hands and feet on physical examination. Blood gas analysis showed low oxygen levels. Complete blood count, blood chemistry, and

cardiac echocardiography results were within normal levels. High-performance liquid chromatography results were as follows; HbA1: 63.6%, HbA2: 32.8%, HbF: 0.2%. Agarose gel electrophoresis was performed to distinguish HbA2, but a band was identified at the level of 39.7% in HbF, G zone, and between HbA1 and HbA2. A blood sample was transferred to our genetic diagnostic center. Following DNA extraction with a commercial kit (Roche, Germany) and amplification of the whole beta globin gene by standard PCR protocols, DNA sequencing (Applied Biosystems, USA) revealed an A to C substitution at nucleotide position 308 (Figure 1). This change was identified as HBB: c.308 A>C, known as Hb Kansas in the HbVar database [5].

Hb Kansas is one of four known hemoglobins with neutral substitutions, along with Hb Köln, Porto Alegre, and Genova [1].

The oxygen equilibrium of Hb Kansas has two unusual characteristics: low affinity for oxygen and low heme-heme interaction. The low oxygen affinity of Hb Kansas should be considered in the differential diagnosis of peripheral cyanosis, especially in the neonatal period and in cyanotic disease and polycythemia in the elderly.

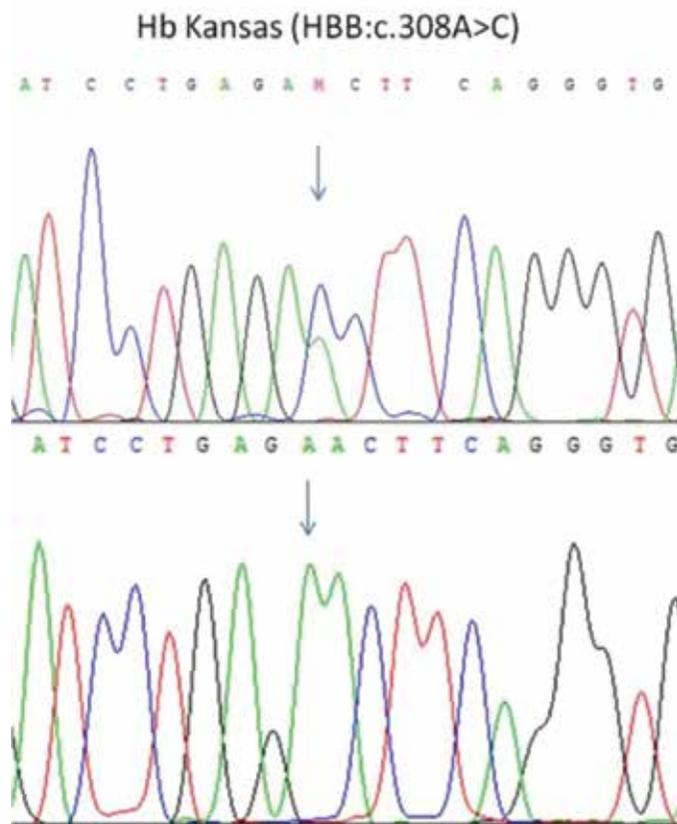


Figure 1. Hemoglobin Kansas in DNA sequencing.

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Keywords: Abnormal hemoglobins, Hb Kansas, Turkish population

Anahtar Sözcükler: Anormal hemoglobinler, Hb Kansas, Türk toplumu

İbrahim Keser¹, Alev Öztaş², Türker Bilgen³, Duran Canatan⁴

¹Akdeniz University Faculty of Medicine, Department of Biology and Genetics, Antalya, Turkey

²Melid Park Private Hospital, Malatya, Turkey

³Research and Application Center for Scientific and Technological Investigations (NABİLTEM) of Namık Kemal University, Tekirdağ, Turkey

⁴Antalya Diagnostic Center of Genetic Diseases, Antalya, Turkey

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Address for Correspondence: Duran CANATAN, M.D.,
Antalya Diagnostic Center of Genetic Diseases, Antalya, Turkey
E-mail: durancanatan@gmail.com

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