

FIRST OBSERVATION OF HAEMOGLOBIN C IN BAHRAIN

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We present the first case of Haemoglobin-C in Bahrain and discuss the significance of this abnormal haemoglobin in the presence of high incidence of sickle haemoglobin in Bahrain. Bahrain Med Bull 1996;18(2):

Haemoglobin C (Hb-C) was the second abnormal haemoglobin to be detected, not long after the description of haemoglobin S (Hb-S)¹. It is found in 17 to 28% of West Africans with the same prevalence as that of Hb-S^{2,3}. But unlike Hb-S disease the selective factors that account for this high prevalence are unknown at present. The prevalence of Hb-S is around 10% in the Bahrainis, but until now no instance of Hb-C has been documented⁴. We describe here one such family.

THE CASE

A 26 year old pregnant Bahraini lady was on regular antenatal examination at Salmaniya Medical Centre, Bahrain, for her fifth pregnancy. She had four previous full term normal deliveries. These children were between 4 to 9 years of age. At the time of examination at 36 weeks of pregnancy her haemoglobin was 9.2 g/dl, haematocrit 0.27 L/L, red cell count $3.4 \times 10^{12}/L$, MCV 79 fl, MCH 27 pg, MCHC 34 g/dl, reticulocyte count 2.5%, total white cell count $10.9 \times 10^9/L$ and platelets $188 \times 10^9/L$. Her peripheral blood film showed microcytic hypochromic red cells with some target cells. Haemoglobin electrophoresis at pH 8.6 showed two major bands at position of A2 and A (Fig 1). Haemoglobin electrophoresis in acid agar gel at pH 6.0 showed two major bands one at A and another at C (Fig 2). Confirmation of Hb-C was done by the 'Iso Lab-Hemocard' monoclonal antibody test for Hb-C. Quantitation done by HPLC showed Hb-C 32.2%, Hb-A2 2.8%, Hb-F 0.5% and Hb-A 64.5%, thus making her a case of Hb-C trait. She was also found to be deficient in iron and

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folic acid. On enquiring she was found to be of Egyptian origin married to a Bahraini man. Investigation of children revealed one of the four children, a boy was also Hb-C trait like his mother.

DISCUSSION

In Hb-C, the glutamic acid in the sixth position from the N terminal of the beta globin chain has been replaced by lysine⁵. On haemoglobin electrophoresis at alkaline pH, Hb-C moves in the same position as Hb-A2, Hb-E and Hb-O Arab. However Hb-C is readily distinguishable from other haemoglobins by acid agar gel electrophoresis.

Hb-C trait is inherited as an autosomal dominant trait. Although clearly a black African characteristic, this abnormal haemoglobin is reported in various

racial groups including Egyptians⁶. In the homozygous form the condition is clinically mildly symptomatic such as low haemoglobin (8-10 g/dl) and splenomegaly. In the heterozygous form it is almost always asymptomatic. However like other traits of haemoglobinopathies low haemoglobin levels can be present during pregnancy⁷. In Bahrainis who have high prevalence of Hb-S, the gene for Hb-C can interact with that for Hb-S to create Hb S-C disease which clinically manifests as sickle cell disease.

CONCLUSION

This is the first case of Haemoglobin C in a Bahraini family. The abnormal haemoglobin was acquired from Egypt through a mixed marriage between Egyptian mother and Bahraini father. Though clinically asymptomatic, haemoglobin C can manifest in a severe form when associated with sickle haemoglobin, thus forming part of the spectrum of sickle cell disorders.

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