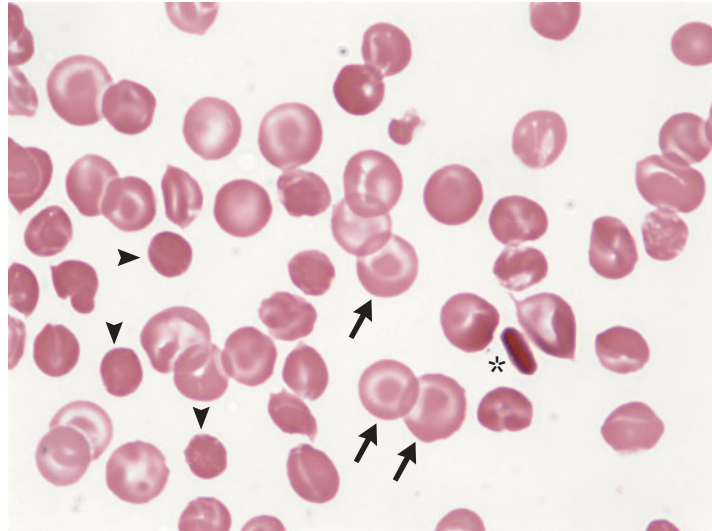


Homozygous Hemoglobin C Disease



Rick M. Fairhurst, M.D., Ph.D.
National Institute of Allergy
and Infectious Diseases
Bethesda, MD 20892

James F. Casella, M.D.
Johns Hopkins University
School of Medicine
Baltimore, MD 21205

A GHANAIAN CHILD WAS FOUND ON ROUTINE SCREENING TO HAVE A hemoglobin level of 9.0 g per deciliter, a hematocrit of 24.3 percent, a mean corpuscular volume of $53.8 \mu\text{m}^3$, a red-cell distribution width of 28.8 percent, and an uncorrected reticulocyte count of 1.6 percent. There were no signs of splenomegaly. Testing of the father, who was asymptomatic, revealed only hemoglobin C on electrophoresis and a complete blood count similar to that of the child. A blood smear showed a constellation of findings that were characteristic of homozygous hemoglobin C disease: target cells (arrows), microspherocytes (arrowheads), rod-shaped cells containing hemoglobin C crystals (asterisk), anisocytosis, and poikilocytosis. The child is now healthy and asymptomatic.

Homozygous hemoglobin C disease is generally a benign condition associated with mild hemolytic anemia and splenomegaly. In terms of geographic distribution, the hemoglobin C allele is found at the highest frequencies in West Africa, where it has been associated with protection against malaria.

Copyright © 2004 Massachusetts Medical Society.