PubMed ID: 10791557 (by Ashley-Koch A, Yang Q, Olney RS, published in *Am J Epidemiol*.)
Title: *Sickle hemoglobin (HbS) allele and* ***sickle cell disease****: a HuGE review.*
Abstract: **Sickle cell disease** is caused by a variant of the **beta-globin** gene called sickle hemoglobin (Hb S). … either two copies of Hb S or one copy of Hb S plus another **beta-globin** variant (such as Hb C) are required for disease expression. … individuals with **sickle cell disease** exhibit significant morbidity and mortality. Symptoms include chronic anemia, acute chest syndrome, stroke, … Disease expression is variable and is modified by several factors, the most influential being genotype. Other factors include **beta-globin** cluster haplotypes, alpha-globin gene number, and fetal hemoglobin expression. … newborn screening, better medical care, parent education, and penicillin prophylaxis have successfully reduced morbidity and mortality due to Hb S.