

Chem 412 Seminar

2. April, 2001

Sherri Strop

Sickle-Cell Anemia: When Good Red Blood Cells Go Bad

Sickle-cell anemia is a genetic disease that is caused by a single amino acid substitution in the B-chain of hemoglobin. This mutation causes red blood cells to have a sickled shape. It is an autosomal recessive disease that effects approximately 1/500 African Americans. When there is low levels of oxygen present deoxyhemoglobin forms, and sickle cell hemoglobin molecules come out of solution and form aggregates that cause the sickle shape of the cells. The sickle cells often get trapped in small blood vessels. There are many symptoms of sickle-cell anemia. They include: painful episodes, hand-foot syndrome, acute chest syndrome, formation of gallstones, leg ulcers, strokes, anemia, renal failure, and hearing loss. There is currently no cure for this disease. Treatments include blood transfusions, hydroxyurea, and bone marrow transplantations. There have been new, exciting developments in treatment possibilities in the past two years. In this seminar, I will discuss the cause of the disease, why it is found in the African population, the symptoms, diagnosis, treatment, and advances in treatment possibilities that have occurred in the past two years.

References:

- 1) Bloom, Mirian. *Sickle Cell Disease*. Jackson: University Press of Mississippi, 1995.
- 2) Blouin, Marie-Jose, et al. Genetic correction of sickle cell disease: Insights using transgenic mouse models. *Science* 6:177-182, 2000.
- 3) Christensen, Damaris. NO News: Nitric oxide may help treat sickle cell anemia. *Science News* 157:2000.
- 4) Hebbel, Robert P. Blockade of Adhesion of Sickle Cells to Endothelium by Monoclonal Antibodies. *The New England Journal of Medicine* 342:1910-1911, 2000.
- 5) Karlsson, Stefan. The first steps on the gene therapy pathway to anti-sickling success. *Nature Medicine* 6:139-140, 2000.
- 6) Kaul, Dhananjaya, et al. Impaired nitric oxide-mediated vasodilation in transgenic sickle mouse. *American Journal of Physiology* 278:H1799-H1806, 2000.
- 7) Sickle Cell Guideline Panel. *Sickle Cell Disease: Screening, Diagnosis, Management, and Counseling in Newborns and Infants. Clinical Practice Guideline No. 6, A HCPR Pub. No. 93-0562*. Rockville MD: Agency for Health Care Policy and Research, Public Health Service, U.S. Department of Health and Human Services. April 1993.
- 8) Toppet, Michele. Antisickling activity of sodium cromoglicate in sickle-cell disease. *Lancet* 356:309, 2000.