Sickle Cell Anemia unmasked

By P. Ann Tatum

The days of the great masquerader are over. Sickle cell anemia has been unmasked.

First recognized in the United States in 1910, Sickle Cell Anemia (SCA) confounded and confused medical men for decades. Little money was earmarked for its research and study because of scant interest the disease stirred in medical circles.

There were a few early pioneers interested in the cure and study of this chronic disease. One of those pioneers is Dr. Roland Scott, a prominent educator and pediatrician at Howard University.

"I've been interested in Sickle Cell Anemia since my days as a student studying pediatrics in Chicago," Dr. Scott said. This was during the late 1930s when Dr. Scott was associated with Chicago's Cook, Provident and Municipal Hospital.

It was there he first saw young SCA sufferers.

"When I was studying pediatrics, I saw a number of children with sickle cell. It was frightening to see these little children with pain crisises," Dr Scott said.

It also was during these early days when the disease was a mystery to many physicians who often misdiagnosed the condition of their patients.

After several more years of specialized study in Chicago and, later in Kansas City, Dr. where he had graduated. At Howard, he and the young doctors and students under his charge continued their interest in fighting SCA.

Because it was still known as "black" disease, research funds were sorely lacking. But thanks to the generosity of groups such as Alpa Kappa Alpha and black chapters of the Elks and Mason, Dr. Scott said some money was available.

SCA was first recognized in Chicago in 1910 by J.B. Herrick, who attended Rush Medical College. Dr. Scott said for a time SCA was referred as Herrick's disease. It was Herrick who noticed the odd shaped red blood cells in the blood of a West Indian student living in Chicago.

An important breakthrough in SCA came 39 years later when in 1949 Dr. Louis Pauling and his coworkers demonstrated that SCA results from the abnormal hemoglobin within the red blood cells of its victims.

SCA is "Like many other diseases in that it exists in mild forms, (can be) moderately severe and very severe," Dr. Scott said.

"The severe cases don't tend to live as long as the not-so-severe cases," he said. It is not uncommon for SCA sufferers with less severe



A simple blood test is a beginning of the process to identify Sickle Cell Anemia.

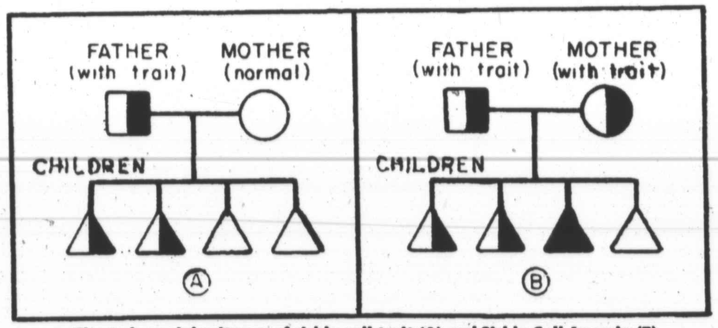
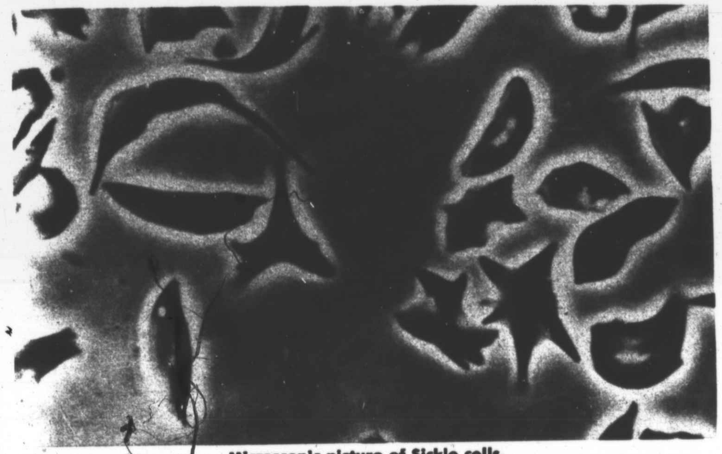


Chart shows inheritance of sickle cell trait (A) and Sickle Cell Anemia (B)



Microscopic picture of Sickle cells.

A FEW FACTS ABOUT SICKLE CELL ANEMIA

— SCA is an inherited disease in which there is a defect in the hemoglobin (coloring matter of red corpuscles of the blood).

— Its victims are predominately black. The trait, sickled red blood cells, have been found in one of every 12 Black Americans. The active disease is found in about one of every 600 blacks and in one of every 1,200 American whites.

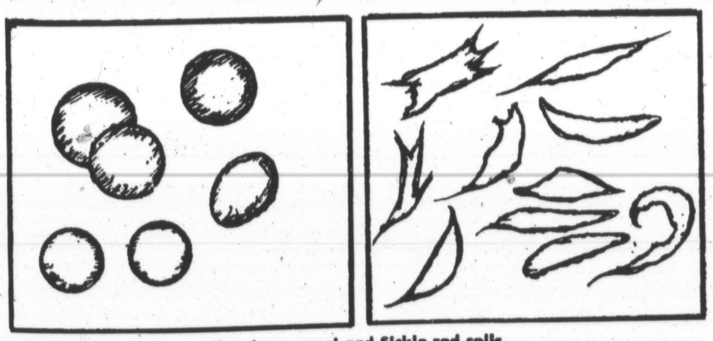
— Estimates vary, but approximately 50,000 persons in the U.S. are sufferers.

— SCA and its variant forms have been described in people and their descendants from Southern India, South and Central America, Greece, Italy, Syria, the Caribbean Islands and other countries.

— SCA occurs when two people, who both carry the gene for the sickling trait (which causes no symptoms) mate. If the child inherits the gene from each parent, the presence of a double dose in the child will result in sickle cell anemia.

— The trait is a condition where a person has inherited a single dose of the sickling gene from one parent. The trait cannot turn into Sickle Cell Anmia and these people normally are healthy and require no treatment. They can, however, pass the sickling gene to their children.

(This information was provided by the Center for Sickle Cell Disease, Howard University, College of Medicine, Washington, D.C. 20059.)



Showing normal and Sickle red cells.

forms of the disease to live well into their 40s, 50s and 60s.

"People are living longer with sickle cell anemia because of better care, antibiotics... people also consult their doctors more, they ask more questions forcing their doctors to know more about the disease, Dr. Scott said.

Medical care obtained through Medicare and Medicade benefits also result in better care for the poorer sufferers.

Of course the passing of the Sickle Cell Anemia Prevention Act in 1971-72 greatly boosted interest in the disease.

According to the text of hearings in Washington, D.C., Sen. Ted Kennedy, of Massachusetts, testified the act would accomplish two things:

"First it will authorize the appropriation of more money for the battle of sickle cell anemia... Second, it will serve to catalyze the enormous potential for all our health and public education resources to help solve problems of those who are affected."

The act freed $25 million for research, voluntary screening counseling and public education during the early 1970s. Also established were 10 centers devoted to SCA study and research.

Dr. Scott said a cure for SCA is coming. Much advancement has occurred in its treatment and research. He mentioned a bone marrow transplant which recently took place. The verdict is still out, but "it looks like the patient is cured, but it's too early to tell,' he said.

The outlook is encouraging, but Dr. Scott lends some sobering thoughts.

"It still is a chronic illness, it has its remissions, but people need to be under good medical care they should avoid things that can throw them into a crisis."

Stress, anxiety, alcoholic beverages, and for some, riding in airplanes can trigger a SCA crisis. Strenuous exercise such as a competitive sport also can bring on a crisis, he said.

A crisis, which is an attack of SCA, is characterized by intense pain. That and depression are two of the major problems of the disease. Dr. Scott said depression occurs because SCA interperes with people's lives.

Children fall back in the school work because of frequent hospital stays. "Adults have a hard time manuvering in the job market," Dr. Scott said.

"Some employers will not hire a person with sickle cell anemia...high job absentism," Dr. Scott said. It also is often difficult for a SCA victim to get insurance.

Still, with the new drugs, treatments and procedures becoming available with better medical attention, SCA suffers stand a better chance of living longer, healthier and productive lives. This is because the great pretender, the great masquerader has been unmasked.