

THE UNIVERSITY OF TEXAS SOUTHWESTERN MEDICAL SCHOOL AT DALLAS

5323 Harry Hines Boulevard, Dallas, Texas 75235 Contact: Ann Harrell

DALLAS--One peice in the puzzle of finding a cure for sickle cell anemia is being sought by two doctors at The University of Texas Southwestern Medical School.

Drs. Michael R. Waterman and G. Larry Cottam, both assistant professors of biochemistry, have been awarded a \$75,000 contract from the National Heart and Lung Institute of the National Institutes of Health to conduct studies in sickle cell disease.

Sickle cell anemia is an inherited disease which is found predominantly in blacks or people with Negro heritage. It occurs, the scientists said, when an altered type of hemoglobin is present in red blood cells. In sickle cell anemia, the normally doughnut-shaped red blood cells tend to take on a sickle-shaped form when the hemoglobin is deoxygenated. It is from this reaction that the disease takes its name.

Since hemoglobin is the substance in the red blood cells which carries oxygen throughout the body, it is imperative that they move freely, Dr. Waterman explained. The changed, or "sickled," blood cells cannot pass easily through many of the small blood vessels. Thus the twisted cells may pile up, he said, causing blood clumping which blocks the flow of blood to the tissues and inhibits the normal distribution of the oxygen. The reason for clumping of the red cells, according to Dr. Waterman, is that the hemoglobin comes out of solution in the deoxygenated form.

Natural body processes also destroy the sickled cells more rapidly than normal cells, contributing to many other health problems in the victim, the medical expert added.

The studies undertaken by the Southwestern research team are two-pronged. First, they want to understand the chemical nature of the interaction between the altered blood cells, known as deoxygenated hemoglobin S molecules. Secondly, their goal is to study the dynamics of the reaction.

"It is only through an understanding of this aggregration phenomenon of the hemoglobin molecules that the complete nature of the disease can be understood," said Dr. Waterman.

Hopefully, he commented, locating the site of the chemical interaction between the hemoglobin S molecules and identifying the character of the aggregation process will help achieve the ultimate goal in the treatment of the sickle cell disease—a method for reversing the aggregation or clumping process. So far no specific treatment has been identified although recent studies with cyanate show promise, he pointed out.

These changes in the red blood cells, which are identified as sickle cell anemia, may lead to a variety of symptoms, according to current medical knowledge. The most prominent is a periodic attack of acute pain, called a sickle cell crisis, which can last for days. Because altered hemoglobin may be present in varying degrees, some people may not be troubled by the disease themselves but be "carriers" of the trait. These "carriers" may pass the trait on to their children without having the disease themselves.

JULY 26, 1972