

SOUTHWESTERN NEWS

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UT SOUTHWESTERN AND U.T. DALLAS WIN NIH GRANT TO ESTABLISH FIRST SICKLE CELL CENTER IN SOUTHWEST AT CHILDREN'S MEDICAL CENTER

DALLAS – Sept. 16, 2002 – A multi-million-dollar five-year federal grant will enable The University of Texas Southwestern Medical Center at Dallas and The University of Texas at Dallas to establish the first National Institutes of Health sickle cell center in the Southwest. The principal clinical site will be at Children's Medical Center of Dallas.

When funded next April, the grant is expected to total almost \$8 million. The comprehensive sickle cell center, led by Dr. George R. Buchanan of UT Southwestern, is one of 10 chosen to form the first national clinical trials network for the disease.

In addition to the two Dallas components of The University of Texas System, the University of Oklahoma Health Sciences Center in Oklahoma City and Tulsa, UT Medical Branch in Galveston, Scott and White Health Science Center in Temple, and pediatric clinics in Paris and Tyler will collaborate in clinical studies and research on this inherited red blood cell disease, which strikes one in 500 African-Americans and about 70,000 people in the United States.

People with sickle cell disease have a genetic error in their hemoglobin, a component of red blood cells. Instead of being soft and round, the red blood cells of a sickle cell patient are inflexible and sickle-shaped, causing blockages in the blood vessels and preventing body tissues from receiving oxygen.

"When this happens," said Buchanan, professor of pediatrics at UT Southwestern, "the patient may suffer severe pain in the bones similar to the pain associated with a heart attack resulting from blocked coronary vessels. This painful complication often extends to the lungs, where it induces acute chest syndrome, a condition very similar to pneumonia. In addition, damage to many other organs, including the brain, spleen, and kidneys, may be disabling or even life-threatening.

"We are very pleased to receive this grant from the National Heart, Lung and Blood Institute of the NIH. These funds will support major new initiatives in treatment with the aim of eventually curing sickle cell disease."

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U.S. Rep. Eddie Bernice Johnson, D-Dallas, said federal funding is essential to programs such as the sickle cell center. "I will continue to champion this cause in Congress. Our investment in medical research will no doubt lead to breakthroughs in the fight against this disease. The joint efforts of these institutions will have a positive impact on the communities involved and on those personally affected by sickle cell disease."

UT Southwestern will administer the grant and receive the majority of the funds. UTD will receive the second-largest amount, about \$1.1 million in research funding over the 5-year period. The balance will be divided among the other institutions.

"The outstanding track record of the sickle cell program at UT Southwestern and Children's over the last 20 years helped us obtain this highly competitive grant," said Buchanan, who is also the medical director of the Center for Cancer and Blood Disorders at Children's. "In addition, the research of UTD's Dr. Steven Goodman helped complete the package necessary to win designation as one of the few NIH centers focusing on the illness."

Goodman, who came to UTD from the University of South Alabama last year, where he was director of their Comprehensive Sickle Cell Center, said the combination of his basic research with the research and clinical care initiatives at UT Southwestern makes "the perfect marriage." Dr. Goodman is also an adjunct professor of Cell Biology at UT Southwestern.

Goodman, director of UTD's newly established Sickle Cell Disease Research Center and head of Molecular and Cell Biology, and Dr. J. Victor Garcia-Martinez, associate professor of internal medicine and microbiology at UT Southwestern, will lead the basic science research effort.

Goodman will study "Dysregulation of the Sickle Cell Membrane Skeleton." Dr. Goodman discovered the molecular basis of the irreversibly sickled cell. Garcia-Martinez will investigate "Pre-Clinical Evaluation of Gene Therapy for Sickle Cell Disease." Garcia-Martinez is internationally known for his expertise in developing gene therapy models in HIV infection and hemoglobinopathies.

"A long-term goal of our center's laboratory research is to understand how altered red blood cell membranes lead to the formation of rigid sickle cells and how best to implement gene therapy that will ultimately cure the disease," Buchanan said. "There have been many advances in treatment during the past two decades that improved the lives of these patients, but some of

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the therapeutic approaches may fail or have harmful side effects.”

Buchanan, who oversees the sickle cell disease program at Children’s, said 600 patients are treated for the disease each year by UT Southwestern faculty physicians. Approximately 2,000 babies are born with the disease each year in the U.S., and better management of the disease now enables patients to live into middle age, according to the NIH.

Among the center’s activities will be clinical programs for adults with sickle cell disease, serving patients at Parkland Memorial Hospital, Zale Lipshy University Hospital and St. Paul University Hospital. Dr. Cynthia Rutherford, professor of internal medicine at UT Southwestern, will direct the adult program with the assistance of a full-time nurse coordinator.

A patient service core will provide education and genetic counseling in conjunction with community outreach and will be staffed by a newly hired patient service coordinator. A full-time genetics counselor, funded as part of the grant, will be added to the staff at the Sickle Cell Disease Association of America, Dallas Chapter, to work with Children’s and UT Southwestern physicians.

In addition, a Scholars Program will foster the career development of young physicians and other health-care professionals who conduct research on the illness.

By congressional mandate, the NHLBI funds 10 Comprehensive Sickle Cell Centers every five years.

Few rigorously performed clinical trials have been conducted in sickle cell disease, and the establishment of a network of leading sickle cell researchers offers a tremendous opportunity, Buchanan said.

Clinical projects supported by the grant will include studies directed by Buchanan; Dr. Zora Rogers, associate professor of pediatrics at UT Southwestern and associate medical director of the pediatric sickle cell disease program; and Dr. Charles T. Quinn, assistant professor of pediatrics at UT Southwestern. They will conduct research on:

- “Outcome of a Newborn Sickle Cell Cohort”;
- “Epidemiology, Treatment and Secondary Prevention of Priapism”;
- “Dexamethasone for the Acute Chest Syndrome of Sickle Cell Disease”; and
- “Ketorolac vs. Ibuprofen for the Painful Crisis of Sickle Cell Disease.”

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