News
The University of Texas Health Science Center at Dallas
5323 Harry Hines Boulevard Dallas, Texas 75235 (214)688-3404

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CONTACT: Ann Harrell
Office: 214/688-3404
Home: 214/369-2695

**New research in sickle cell treatment helps patients.

DALLAS--George and Carla Sanders lead happy, normal unrestricted lives as young teenagers, their only complaint being more-frequent-than-usual trips to the doctor. Even these wouldn't be so bad if it weren't for the routine bloodwork each time.

George and Carla's older sister Rhonda, 17, is an invalid. After suffering several strokes, Rhonda had to be confined to a nursing home. The brother and sisters are victims of the same disease: sickle cell anemia.

An inherited chronic blood disease that primarily affects people of African descent sickle cell anemia is estimated to affect one in every 600 black Americans. And one in every 10 or 12 are "carriers" of the sickle cell trait that is genetically transmitted. Unfortunately, there is no cure, but the severity of the illness varies widely from patient to patient. Persons who carry the trait are entirely well and have no associated medical problems themselves, but carry the potential for the disease in their genes.

Normal blood cells, says Dr. George Buchanan, associate professor of Pediatrics at The UT Health Science Center at Dallas, are round, almost hollow and doughnut shaped. In persons with sickle cell, these cells have a tendency to twist, or "sickle", into boomerang shapes. Thus, the sickled cells stick together and block the normal flow of blood in the small veins. This blocked blood flow results in pain in such areas of the body as the abdomen, chest, arms and legs, resulting in a condition called sickle cell "crisis." In addition, extensive damage to the internal organs can be caused by the blocked blood flow.

Buchanan, who is director of the sickle cell clinic at Children's Medical Center, the health science center's pediatric teaching hospital, has been involved with both research and patient care since his days as a research fellow at Harvard Medical School in the mid-70s. Under his direction the Children's clinic has expanded so that it is among the largest ten treatment programs in the country, seeing approximately 300 patients on a regular basis. In addition, Buchanan and his associates are on the forefront of research in this area.

The physician reports that the clinic, which is now 7 years old, is now seeing nearly every child with sickle cell disease in North Texas. Most of them see clinic doctors for their total medical care, but others who see private physicians come in to the clinic for treatment of their blood disorder. Because the clinic sees so many patients, it is an ideal place for research into the disease, says Buchanan.

"Sickle cell is a rather poorly understood disease in that very little is known about the clinical manifestations. We know a lot about the molecular and genetic aspects of the disease but very little about how it manifests itself or how to best treat the individual patient," says Buchanan. Generally physicians have far more knowledge of how to treat a disease than the basic scientific facts about the cause on a cellular and molecular level. And no one can predict why patients like Carla and George are able to control their illness with proper medical care while victims like Rhonda are devastated by its manifestations. However, says the researcher, each year brings new gains in treating the sickle cell patient.

Prevention of infection, a major cause of serious illness among sickle cell patients, continues to be a major research focus for Buchanan and his associates. Sepsis, or blood poisoning, causes death in fully 20 percent of the infants with sickle cell disease if special precautions are not taken. Buchanan and his associates have done work that determined the effectiveness of vaccinating children against one of the most prevalent germs causing blood poisoning and pneumonia. In addition, they have been involved in ongoing work on the prophylactic use of penicillin for the child with sickle cell disease. This method of combating infection has been used successfully for some time, but now researchers are looking as just how long the preventive doses should be given.

(over)

"It used to be thought that penicillin should be given to the child until he or she was about five or six. But now there is some concern that the drug should be taken much longer for protection from infection," Buchanan says.

Since blood circulates to all areas of the body, organ-related physical problems can also prove quite severe to the sickle cell patient. Some of these may include enlargement of the spleen or liver, gallstones, kidney problems and, as in the case of Rhonda, strokes.

One of the problems faced by the sickle cell patient who suffers from strokes is iron overload from the blood transfusions that are given as treatment. Too much iron from the transfusions literally begins to poison the patient as the iron from each unit of blood is stored in the body. Now some of the patients are being placed on a small infusion pump much like the insulin pump used in the treatment of diabetes. Medicine flows into the system to release the iron so it can be expelled from the body in the urine. The drug is pumped in through a tube attached to a needle that is placed under the patient's skin at night.

Older children and teenagers using the pump have to be monitored closely because strict compliance is necessary. The device must be left in place every night for a period of about 12 hours.

Pain control in the young person with sickle cell disease is another research interest of the pediatrician's. "Sickle cell pain is not chronic," he says. "Instead, it is severe, acute and intermittant."

Unfortunately, many physicians treating sickle cell patients do not realize the extent of their patients' suffering when they are in crisis. Therefore, Buchanan says he spends a good deal of time educating physicians in the need for liberal intravenous infusions for pain.

"There is neither the problem of chronic use of drugs not the psychosocial situations that would set up these young patients for addiction." he says.

Buchanan and his associates have also been involved in ongoing research they hope will lead to better diagnostic procedures for infection in sickle cell patients. Physicians commonly look for an abnormal number of white cells in the blood as an indication of infection in their patients. Persons with sickle cell disease, however, normally have blood counts with an unusually large number of white cells, so the usual procedure of counting the number of white cells does not apply.

The pediatrician says he believes that newborn screening for sickle cell anemia -- a procedure common for a number of years in most states but new in Texas -- will do much to cut early morbidity and mortality coming from the disease. The sooner parents know that the child has sickle cell, the sooner they can enroll him or her in the program for medical monitoring. In addition, they will receive parent education about the disease, its symptoms and when to seek medical help.

Buchanan's "right arms" in working with parents are sickle cell team members Dr. Robert Sprinkle, assistant professor of Pediatrics, and Susan Smith, pediatric nurse practitioner. Smith's role includes evaluating sick patients with the house staff, evaluating patients at interval check-ups, and providing education for parents. She believes they need to know about the pathophysiology of the disease, its possible complications, genetic patterns, how to manage minor problems at home, and the importance of regular treatment and check-ups.

"Too many people think of patient education as simply giving the patients information to satisfy their curiosity. What we really need to give them is information to help them prevent fatal complications and to handle other problems," says Smith. The nurse/educator has produced educational materials on the disease for both the area program and for the state of Texas. She is also a member of the state advisory commission on newborn screening for sickle cell disease.

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