News
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***Research, treatment and patient education aid 220 sickle cell patients each year at CMC clinic.

DALLAS--Willie Wiggins, 18, entered Cedar Valley Community College in August. Like the other 2,400 freshmen, he is hoping that education will give him an extra push to a bright future. Just five years ago the young man would never have thought he would be well enough to go to college. And as for a future, sometimes he didn't think he would have one.

Wiggins has sickle cell anemia, a chronic blood disease that causes the normal, almost-round red blood cells to twist, or sickle, into boomerang-like shapes. These abnormal sickle cells have a tendency to stick together and clog the normal flow of blood in the small vessels. This blocked blood flow results in pain in such areas of the body as the arms, legs, abdomen and chest. This condition is called a sickle cell crisis. A crisis can also damage internal organs.

Too often parents of children with sickle cell anemia believe that life is hopeless for the child who has this disease, says Susan Smith, pediatric nurse practitioner in Children's Medical Center's sickle cell clinic, a program associated with The University of Texas Health Science Center at Dallas. Compounding the problem is the fact that these misguided parents pass on this belief to their own sons and daughters.

Smith, who works with patient education in the clinic as well as being involved with medical care, is dedicated to changing this attitude.

"Many of these people have heard dire stories from the past that foster the belief that few people survive past age 20. They mistakenly think that someone with sickle cell who is in horrible pain could die any time. They also think that there's nothing medically that can be done. Neither is correct," says Smith.

Fortunately for Wiggins, who was diagnosed as a sickle cell patient when he was only a few months old, his parents heard of the work of Dr. George Buchanan and his associates at Children's. There the young man received an evaluation, regular check-ups and medical care when he was having a sickle cell crisis. Also he and his parents were taught about the disease and how to cope with it.

"Susan and the doctors explained why I got sickle cell," says Wiggins. "And they also have taught me how to avoid getting really sick. You can call them any time or go by and see them, and they'll help you."

The clinic, now in its fifth year, sees about 220 patients a year, a figure that includes most of the children and teen-agers with sickle cell disease in the Dallas area.

"One thing that helps is that we're picking up these cases earlier. We do a lot of professional education in the form of making presentations to public health nurses, public school nurses, private pediatricians and pediatric house staff," says Buchanan.

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The program is one of the largest 20 treatment efforts in the country involved in research as well as patient care. It is the addition of the research component to the Dallas program that the pediatric hematologist-oncologist believes has added so much to its success. Among the Dallas research projects that have contributed to the understanding of treatment for sickle cell patients are the following:

Prevention of infection. A major cause of serious illness among sickle cell patients, infection is being fought in the Dallas program by giving prophylactic doses of penicillin to patients with the disease. Major causes of death in these patients are spinal meningitis and blood poisoning. Fortunately, the germ that causes most of these problems is susceptible to penicillin. Although it has been thought for a long time that giving penicillin to these patients would cause them to develop a resistance to the drug and set up the possibility for infection from other organisms, Buchanan says that did not happen with his research group. When cultured for pneumococcus, a control group (children who did not have sickle cell) had far more germs present in the back of the throat and nose than did the sickle cell patients. The result of this study, which was carried out with Drs. Jane Siegel and David Anglin in the Department of Pediatrics, will soon be formally presented at a national meeting.

Controlling pain during crisis. Sickle cell disease can cause pain that ranges from slight pain in the leg or joints to excruciating pain requiring massive doses of narcotics. While many physicians are leery of giving these massive doses for fear of addiction, Buchanan and his associates have found that in these painful crises, the usual doses of pain medication don't work well, and a continuous intravenous infusion of high dosages of such medicines as Demerol and morphine are necessary. They are, however, given only in the emergency room and in the hospital. Giving these drugs under close medical supervision and for short durations keeps addiction from being a problem.

"This is definitely the right thing to do," says the physician. "But they're given just during the period of crisis--never at home. Nothing stronger than codeine is given outside the hospital."

Knowing your patients. It just doesn't work for a patient with sickle cell to go from hospital to hospital for his or her care. The health team needs to be familiar with the case in order to understand what's going on in each individual circumstance. They need to be familiar with how much pain the patient has in crisis and how his or her metabolism functions in order to choose the correct drugs, a problem Buchanan and biochemist Larry Cottam will soon be investigating. All the CMC patients come in on a regular basis to the clinic where they get to know members of the health team, and, in addition, records are kept in the adjoining emergency room at Parkland Memorial Hospital in case the patient has to come in when the CMC emergency room is closed. Patients are also encouraged to call members of the health team when they need to.

Patient education. Both Smith and Buchanan believe that proper patient (and family) education is an important part of treatment. In a study of compliance with treatment, Buchanan concluded that the better the patients and their families understood the disease and the treatment, the better the compliance. With that in mind, educational meetings for parents and other interested persons are held every few months in evening sessions. In that way the patients and families can learn even more than in individual sessions at clinic and have plenty of opportunity to ask questions.

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Smith believes that parents need individualized education to help them care for a child with sickle cell. Many of her parents, she says, don't know how to read a thermometer so they can't tell how seriously ill the child is. They may be too embarrassed to tell her so. The solution? She begins by saying, "Many people aren't too good at reading thermometers" in a very non-threatening way. Then she puts them through practice runs.

Smith even has parents feeling the children's spleens to see if they are swollen, a sure symptom of a serious crisis in sickle cell disease.

"It's not that difficult," she says. "I tell them I have confidence that they can learn to do it. And they do, too."

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