News
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*******UT Southwestern sickel cell anemia clinic doing research into the disease.

DALLAS--Little Angie fell backward on the nursery school floor. The yellow plastic duck she clutched in her right hand toppled over on its beak. Angie's dark eyes seemed frozen with fear. But she didn't speak. And she didn't move.

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The bedside clock read "3."

Ten-year-old Tanya shook her mother's shoulders, first gently, then with more insistence. "Mamma, Mamma, wake up," she said. "Please wake up. My heart's hurting!"

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Both little girls are the daughters of Karlean Allen. And both, no matter how different their symptoms, are ill with the same disease.

Sickle cell anemia.

An inherited chronic blood disease that primarily affects black people, sickle cell anemia is estimated to affect one in every 400 black Americans, like Angie and Tanya. And one in every 10 or 12 are "carriers" of the sickle cell trait which is genetically transmitted. Unfortunately, there is no cure for the disease. However, persons who carry the trait are entirely well and have no associated medical problems themselves.

In persons with sickle cell anemia, the red blood cells instead of being shaped like round, almost hollow doughnuts, have a tendency to twist, or "sickle," into boomerang shapes. These 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, a condition called sickle cell "crisis." It may also cause extensive damage to internal organs.

Dr. George R. Buchanan, pediatrician at The University of Texas Health Science Center at Dallas, has been interested in sickle cell anemia since his days as a research fellow at Harvard Medical School. Later, when he joined the faculty at that institution, he continued treating patients with sickle cell disease.

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Now an assistant professor of pediatrics at the Dallas medical school, Buchanan is working with sickle cell patient care and research at Children's Medical Center. With the addition of grants from the Zale and Hoblitzelle foundations, the sickle cell clinic has expanded so that it is among the largest 20 treatment programs in the country, seeing approximately 125 patients on a regular basis. Thirteen of the children in the Dallas program have had strokes, like little Angie. And seven of these youngsters, including this little girl, are treated by blood transfusions and seen for checkups every few days to every few weeks.

The physician reports that the clinic 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 only for treatment of this disorder. Since the clinic has been open for 2½ years and has such a large patient load, it is one of the few programs where research into this disease is being carried out.

"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 problems associated with the disease but very little about the clinical manifestations and treatment," says Buchanan. Generally, physicians have far more knowledge of how to treat a disease than the basic scientific facts about cause at the cellular and molecular level.

Prevention of infection, a major cause of serious illness among sickle cell patients, is being studied. Sepsis or blood poisoning is the cause of death for fully 20 percent of the infants with sickle cell disease. Buchanan and his associates are looking at the effectiveness of vaccinating children against one of the most prevalent germs causing blood poisoning and pneumonia. A study on this project will be published in the January, 1980 Journal of Pediatrics.

The usual pediatric inoculations are also given.

A second study is looking at ways of diagnosing 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 anemia, however, normally have blood counts with an unusually large number of white cells because of their disease. The researchers are trying to develop a way of determining infection from the blood count of a sickle cell patient, as well as looking for other diagnostic aids.

Another project is looking at whether there is a disproportional amount of blood clotting, as well as sickling, in the sickle cell patient. There have been some indications that this is so, but Buchanan says that so far his findings do not indicate that this is the case. The final answer to this question will affect the treatment of sickle cell disease.

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

One of the problems faced by children who suffer strokes from sickle cell disease is iron overload from the blood transfusions. Too much iron from the transfusions literally begins to poison the patient as the iron overload is stored in the body. Now some of the patients are being placed on an experimental pump that sends medicine into the system to break the iron loose so the body can expel it. The drug is pumped in through a tube attached to a needle which is placed under the patient's skin at night. All the children on transfusions are carefully monitored for their iron levels. Angie is one who may need the pump soon.

Another area to be looked at is the question of whether having genetic counseling available to persons with sickle cell anemia or who carry the sickle cell trait affects their decision to have children. Since the percentages of passing on the disease or the trait are high, it is thought that counseling is very important. But no studies have been done to determine if passing on this information in a professional setting has made an impact on the decision to have children or to limit the number of children in a family.

Susan Smith is a pediatric nurse practitioner who has recently joined the sickle cell clinic's medical team. She will be very much involved in looking at this question. Smith explains that if both parents have sickle cell traits, their chances for having a child with sickle cell anemia are one in four. The same percentage applies to having a child with normal hemoglobin. However, they are statistically likely to have two chances in four for having children who are "carriers" or have the sickle cell trait.

If one parent has sickle cell anemia and the other is a carrier of the sickle cell trait, the chances are two in four of having children with sickle cell anemia. They are also two in four of having children with the sickle cell trait.

Couples with one partner with normal hemoglobin and one who is a carrier have a two in four chance of having children with normal hemoglobin. On the other hand, the chance for having children who are carriers is also two in four.

Smith's role in the clinic includes an increasing amount of parent education, including an understanding of what the disease is, its genetic patterns, the way sickle cell affects each individual child and the importance of regular treatment and check-ups.

"The thing that makes our jobs easier," Smith says, "is that these parents are really motivated to bring their children to the clinic. Their children are really important to them."

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