UT SOUTHWESTERN MEDICAL CENTER NEWS

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Risk for stroke, death not higher for sickle cell children with early complications

DALLAS – Jan. 29, 2007 – Children with sickle cell disease who experienced major complications such as pain and lung disease early in life are at no greater risk for stroke or death during later childhood, new research from UT Southwestern Medical Center shows.

In addition, sickle cell children who have pain episodes ("crises") or dactylitis, a type of painful swelling of the hands and feet, as infants or toddlers are at no greater risk of having those symptoms recur in later childhood. The study's results, however, showed that children hospitalized for chest problems early on are more likely to see those problems recur up to adulthood.

The study – following more than 200 children with sickle cell disease from birth through teenage years – appears in the January issue of *Blood*, the scientific journal of the American Society of Hematology.

The findings are an important step in trying to identify predictors that reveal how the mysterious disease will progress as children age, said Dr. Charles Quinn, assistant professor of pediatrics at UT Southwestern and the study's lead author.

"Everybody who has sickle cell disease is affected differently by the disease. Some seem to have a lot of problems with pain and lung disease and some have very few problems and may have a normal life span," said Dr. Quinn. "We don't really understand why everyone with the same disease can be so different."

The myriad medical issues make it difficult when counseling parents of babies with sickle cell disease about what they can expect, he said. "We can't give them very much in the way of specifics, exactly what this child will likely go through or what to expect from the disease in the future," said Dr. Quinn, a pediatric hematology specialist at Children's Medical Center Dallas.

People with sickle cell disease have a genetic error in their hemoglobin. The disease turns the usually soft, round red blood cell that carries oxygen through the body into an inflexible, sickle-shaped cell that causes blockages in blood vessels and prevents body tissues from receiving oxygen. It is estimated that at least 70,000 Americans have the disease.

UT Southwestern researchers at Children's and at the National Institutes of Health-funded Southwestern Comprehensive Sickle Cell Center launched the study to try to determine whether

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problems from the disease in the first three years of life offered any indication of later problems.

They initially looked at whether some of the more common problems associated with sickle cell disease – pain events, dactylitis and acute chest syndrome – predicted early death or stroke. Researchers found that none of those factors result in higher risk.

But they did find that acute chest syndrome – damaged lung tissue marked by fever, chest pain and difficulty breathing – did correlate with recurrent episodes throughout the remainder of their childhood.

That may indicate a need for closer follow-up for those children and perhaps justify more aggressive treatment strategies.

Children hospitalized for acute chest syndrome and early painful events in the first three years also were at slightly higher risk for later painful episodes.

"Some doctors would think that if they have early pain, they are destined to have frequent pain later in life, but that's not necessarily the case," Dr. Quinn said.

The swelling condition dactylitis did not indicate any greater likelihood of pain episodes or lung disease up to adulthood, the UT Southwestern researchers found. "That finding in particular is at odds with other studies that showed that early dactylitis does predict later adverse outcomes," Dr. Quinn said.

Researchers reviewed cases of 264 children who are part of the Dallas Newborn Cohort, a unique patient pool started in 1983 when newborn screening for sickle cell disease was launched by the state. Researchers have been able to follow children with the disease to track how sickle cell patients fare. Earlier findings showed that children with sickle cell disease are living longer, dying less often from their disease and contracting fewer fatal infections than ever before.

Dr. Quinn said this latest step of identifying potential clinical signs is an important one for predicting the future for sickle-cell patients.

"This finding is something that could potentially be applied anywhere, whether you have a high-tech lab or you practice in a small clinic in a rural community," Dr. Quinn said. "We're always looking for something to help predict the future. Lots of investigators have looked at many laboratory markers with mixed results. What we did was to look at very simple, clinical manifestations of the disease that are easily seen by parents and by doctors and that didn't require any special laboratory or equipment to make this sort of prediction."

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Also involved in the study were Dr. Zora Rogers, associate professor of pediatrics; Dr. George Buchanan, professor of pediatrics and director of the Southwestern Comprehensive Sickle Cell Center and the Barrett Family Center for Pediatric Oncology; and Elizabeth Shull, a registered nurse who collected the data.

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UT Southwestern Medical Center, one of the premier medical centers in the nation, integrates pioneering biomedical research with exceptional clinical care and education. Its more than 1,400 full-time faculty members – including four active Nobel Prize winners, more than any other medical school in the world – are responsible for groundbreaking medical advances and are committed to translating science-driven research quickly to new clinical treatments. UT Southwestern physicians provide medical care in 40 specialties to nearly 89,000 hospitalized patients and oversee 2.1 million outpatient visits a year.

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