If you have questions, or any specific topics you would like to be reviewed, please call the Children's Hospital Hematology Department at (205) 939-9285, your local sickle cell foundation, or ask the sickle cell doctor or nurse practitioner in clinic.

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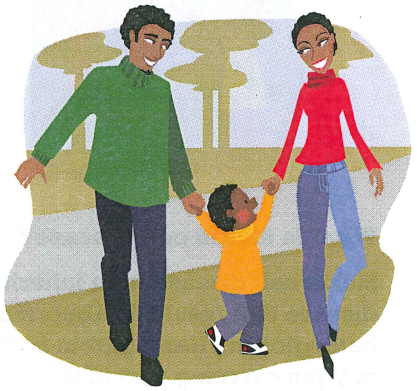
UAB DIVISION OF PEDIATRIC HEMATOLOGY AND ONCOLOGY

So you have a sickle cell disorder



YOU HAVE SICKLE CELL DISEASE. WHAT DOES THAT MEAN?

What does it mean to have sickle cell disease? Endless days in the hospital, lots of doctor visits, constant pain, and no fun! Not necessarily.

There are many different types of sickle cell disease and people with the same type can have very different experiences. This handout discusses hemoglobin S and how it affects the severity of the different types of disease. Now please read on and learn more about sickle cell disease.

What is sickle cell disease?

Sickle cell disease is an inherited disease that results in abnormal hemoglobin. Hemoglobin is the oxygen carrying part of the red blood cell (RBC). It is what makes the blood look red. Remember, there are many different types of sickle cell disease and people with the same type can have very different experiences.

How do you know if you have sickle cell disease?

If you were born in or after 1988, you have probably known since the day you were born that you have sickle cell disease due to a program called newborn screening. Newborn screening requires every child born in Alabama to be tested for sickle cell disease. Persons born before 1988 probably discovered that they had sickle cell disease through pain crisis or an illness.

How do you know what type of sickle cell disease you have?

Because sickle cell disease affects hemoglobin in the red blood cells, a test called a hemoglobin profile is done on a blood sample. A hemoglobin profile shows what kind of hemoglobin and the amount of each you have.

There are many different types of hemoglobin (Hgb), but the most common types are the following:

- **Hgb A:** Normal hemoglobin or Adult hemoglobin.
- **Hgb S:** Sickle hemoglobin is a unique hemoglobin associated with sickle cell disease. When Hgb S loses oxygen, it attaches to other Hgb S molecules. Over a short period of time, more and more Hgb S molecules come together and form rigid rod-like structures. These structures distort the red blood cell into a sickle shape. Because of the rigidness of the cell, it is unable to fit through small blood vessels. These vessels become clogged and red blood cells are not able to get oxygen where it is needed. This causes a pain crisis.
- **Hgb F:** Fetal hemoglobin is the hemoglobin you have when you are born. Between ages 6 months and 2 years, most people's bodies slow down in the production of fetal hemoglobin. People with Hgb AA or Hgb AS types maintain a Hgb F level of 1-2%. People with different types of sickle cell disease maintain Hgb F levels between 2 and 30% depending on the type.
- **Hgb C:** Hemoglobin C is another type of hemoglobin. It does not sickle, but when it is found in combination with Hgb S, it forms one of the sickle cell disorders.

Other types of hemoglobin include Hgb D, E, and G. Different combinations of hemoglobin determine the type of sickle cell disease.