Sickle Cell Series

Chapter 1

**Sickle Cell Disease Series: How Atrium Health is advancing care for the Sickle Cell Disease patient population**

***The Burden of Sickle Cell Disease***

Sickle cell disease affects approximately 100,000 patients in the United States, and on average 1 out of 13 African Americans are born with the sickle cell trait, according to the Center for Disease Control and Prevention. The disease is also more prevalent in the Hispanic population. Sickle cell disease is a genetic condition where parents must be carriers and pass it onto their child for it to be inherited.

It is a disease without a cure that affects the red blood cells, causing them to take on a sickle “C” shape. Red blood cells are responsible for moving oxygen throughout the body; yet in sickle cell disease patients, their red blood cells die off early, causing a shortage often leading to anemia. And due to their shape, the red blood cells can cause clots, which can lead to pain, stroke, infection and even organ failure.

Living with the disease is not easy for patients; patients frequently report chronic pain as a side effect of the disease. Frequent doctor visits and prescriptions are often necessary for patients, who tend to be lower-income, which makes paying for medication and healthcare a challenge. Due to the frequent doctor visits, pain episodes and hospitalizations, patients have barriers that affect their ability to lead normal daily lives.

***Providing a unique and robust approach to care***

Patients begin treatment at the onset of diagnosis. Not all treatments work for all patients, which is why Atrium Health works together across departments to meet individual patient needs.

For some sickle cell disease patients, a simple blood transfusion is part of their treatment plan. A blood transfusion decreases anemia. However, every time a patient is given a blood transplant, more iron is put into the body. Too much iron can have adverse effects such as liver disease, heart disease and diabetes. Therefore, the patient has to take more medication to reduce the iron burden.

Atrium Health physicians have been researching and testing a different form of blood transfusion: a red cell exchange transfusion. Red cell exchange transfusion is an alternative treatment that replaces a patient’s red blood cells to help carry oxygen to all parts of the body. Daniel McMahon, MD, pediatric hematologist and director of the pediatric sickle cell disease program at Atrium Health’s Levine Children’s Hospital, says that they have approximately 20 patients receiving exchange transfusions right now. Starting within the last decade, transfusion therapy has taken a bigger role. Levine Children’s Hospital in Charlotte is one of only a few hospitals nationwide offering this alternative treatment to patients.

“I think the transfusion therapy is now so much safer,” said Dr. McMahon. “We’re doing exchange transfusions on a large number of children, which prevent some of the iron overload that occurs with the chronic transfusions. Transfusion therapy is something that we have available to us that really can make a significant difference in people’s therapy.”

Similar to kidney dialysis, only red blood sickle cells are removed and donor blood is added back to the patient. In fact, the same machine is used for both transfusions. The interaction of the hematology, the nephrology and the pediatric departments allow patients to receive cutting-edge treatment like the exchange transfusions.

“The exchange transfusion is a marvelous way of keeping the amount of sickle cells in the blood reduced,” said Dr. McMahon. “It’s quick and easy.”

However, it does come with some complications; patients need a central venous catheter access, which can cause infections and other problems. The therapy also requires more blood and has possible side effects, like nausea and dizziness.

Others forms of treatments, such as stem cell transplants, can be an option for pediatric patients. A stem cell transplant is the only curative form of therapy that requires a stem cell donor, of either peripheral blood or bone marrow, explains Dr. McMahon. The success rate is highest in cases where they have a perfectly-matched donor, which is usually a full sibling who has the same HLA markers but doesn’t have sickle cell disease. In most cases, finding an exact match under these criteria isn’t possible.

Work is being done to find donor matches from alternative sources such as parents or half siblings. Atrium Health and Levine Children’s doctors are participating in national protocols for such sources to look at outcomes and potential long-term complications.

“We have a little more than 500 children with sickle cell disease, and we have about 10-12 children each year undergo stem cell transplant,” said Dr. McMahon. “Most of those are with alternative donors, and we probably do one or two a year with a perfectly-matched donor.”

***Looking to the future***

Atrium Health physicians regularly discover new treatments and research. Ultimately, this leads to a longer life expectancy and better quality of life for sickle cell disease patients.

“Forty years ago, most individuals with sickle cell disease did not live beyond age 18,” said Dr. McMahon. “Now they almost all live into adulthood and their life expectancy is increasing and increasing. One of our biggest goals obviously is to not only have them live longer but to live with good quality of life and be healthy adults.”

*Patient Spotlight:*

*Patient Dejuan Minnigan, a patient of Dr. McMahon and Levine Children’s, has benefited from the exchange transfusions. He had several strokes and needed chronic blood transfusions. Therefore, he was taking the prescription Exjade, a medication in liquid form, for the excess iron. Transitioning Minnigan to exchange transfusions has allowed doctors to keep his sickle cell level even lower than it was before and he was able to stop taking Exjade because his iron levels are no longer too high.*

*Mary Minnigan, Dejuan’s mother, says the new red blood cell exchange therapy basically saved his life.*

*“Now he still gets his exchange, but he doesn't have to take medicine anymore,” says Minnigan. “And he's super healthy, so it's a good process.”*

*Dejuan receives treatment and care from Atrium Health’s comprehensive sickle cell disease clinic, complete with clinical care, a psychotherapist, a social worker, case managers and nurses.*

*“There’s some people in here that work for this hospital that have really gone over and beyond to help my family,” says Minnigan. “Things you wouldn’t believe, like going way past the level of expectations. It’s like a family environment.”*