The Wright Family’s Journey to a Cure

When Sharee Edmonds learned that three of her children had sickle cell disease, she prayed for a miracle that would cure them. That miracle came when her daughter Unity, now 17, was born. In an incredibly rare occurrence, her bone marrow was a perfect match for all three of her siblings. Now with seven siblings in total, Kortne Wright, 26, Alequis Wright, 24, and Cachet Wright, 21, are the only three that had the disease while six carry the trait.
Sickle cell disease is a genetic disease that affects red blood cells in the body. Each year about 2,000 children are born with sickle cell disease in the United States, the majority of which are African American. When both parents have the sickle cell trait, there’s a 25 percent chance that their child will have sickle cell disease. Sickle cell disease can affect many different organ systems in the body. Important organs like the bones, brain, heart, and kidneys, which need a constant blood supply, can be damaged by sickle cells that do not move through the body as easily as normal cells. For the Wright siblings, it was very routine to have what they call “visits,” severe bone pain that was unpredictable in both duration and severity.

“Although they were diagnosed at birth, I didn’t start to notice symptoms and they were ‘invisibles,’” their mother Shari says. “Tears were the most common signs until they got older and started experiencing severe pain. It was very difficult as a mother to watch your children go through this. You learned to avoid certain triggers like extreme temperatures and flu season, but it was hard with three young children.”

In 1999, Alequis, who was 7 at the time, suffered a stroke and needed a bone marrow transplant to save her life. Unity was just seven months old, but the doctors caring for Alequis told her to see if she was an HLA (Human Leukocyte Antigen) match, which she was. HLA typing is used to match patients and donors for a bone marrow transplant by creating a DNA fingerprint. HLA are proteins found on most cells in the body, and the matching process means those to recognize which cells belong in the body and which do not. The more similar they are, the less chance there is rejection. Since half of HLA markers are inherited from the mother and half from the father, each full sibling has a 1 in 4 chance of being a match.

A few years before transplant, Cachet developed an autoimmune disease that left her paralyzed. Despite all the challenges with sickle cell disease, all three siblings say they had a normal childhood and they had learned to live with their disease. When it interfered with school, they worked with teachers or complete home schooling to catch up. They avoided certain triggers and managed their pain when it came. “After the transplant their lives have changed dramatically. They are learning to adapt to normal life without having to constantly worry about sudden pain, cold, or triggers,” Shari said. Both Cachet and Alequis will be attending Gateway Community College this fall. Cachet plans to be a nurse and may return to Yale New Haven Hospital, but this time not as a patient.

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Alequis’ bone marrow transplant was performed at Yale New Haven Hospital and the family decided not to immediately schedule another transplant for another child, but to wait and see how Alequis recovered. In early 2015, when the time came for Kortne and Cachet’s transplant, they met with Dr. Michael Kent, Assistant Professor of Pediatrics and a member of the Pediatric Transplant Program at Yale. He worked to find a regimen that would be well tolerated. “Once a patient is over the age of seventeen, transplant becomes more toxic for them,” explained Dr. Kent. “Our first task was to determine whether or not they would benefit from transplant, and then find a regimen that would be well tolerated.” Kortne and Cachet both received a reduced intensity regimen with less chemotherapy given in advance of their transplant to reduce toxicity, while still ensuring an effective transplant. Kortne and Cachet received their transplants in the summer of 2016 and are more than 6 months post-transplant.

Dr. Shah, Assistant Professor of Pediatrics, joined the Pediatric Transplant Program as Director. Dr. Shah came from a very high volume transplant program and brought with her the expertise and experience needed to treat patients like Kortne and Cachet and offer them a cure. Dr. Shah commented that up until the point of transplant, Kortne and Cachet’s disease was being managed by frequent blood transfusions and pain medication, and when those didn’t work, hospitalization was necessary. “They are both still on graft-versus-host disease (GVHD) prevention medication,” explained Dr. Shah, “but will eventually be weaned off of it completely. Unity’s cells have slowly stabilized in Kortne and Cachet’s bodies and they are not experiencing any further sickle cell disease related complications.” For patients that receive a transplant from a sibling, the cure rate is 80-90 percent. What makes the Wright family’s case so unique is that three siblings were diagnosed with the disease, and one sibling was a donor match for all three. Dr. Shah commented that she has never seen a case like this before.

The Pediatric Transplant Program at Yale is the only one in the State of Connecticut, and as Director, Dr. Shah’s goal is to grow the program regionally. Dr. Shah explained that the Transplant Program, as part of the Pediatric Hematology/Oncology Program at Smilow, performs various types of transplants for both malignant and non-malignant disorders of childhood. Dr. Shah comments, “Transplant is the only curative therapy for patients with sickle cell disease and the earlier they can get it, the better the success rate.”

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