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Lost in Transition: Sickle Cell Clinic Helps Adults Continue Care

State grant supports expansion of UC San Diego Health's comprehensive sickle cell clinic

Vanessa Hughes remembers cheerfully playing childhood games one minute and running to her parents writhing in pain the next. During those early development years, Hughes was studying her ABCs while also learning how to moderate her playtime to reduce the risk of going into a painful sickle cell disease crisis.

Often it was trial by fire.

"I didn't know what was going on. I just needed it to stop," recalls Hughes, now 24 years old. "There were times when I would have a pain crisis and my dad would carry me to the backyard so I could feel the cool air because I would be running a fever. In those days, all they could do was give me Tylenol so I wouldn't end up in the ER. Still, most often I would."

More than 250 people with sickle cell disease live in San Diego. The number may seem small, but its impact is severe.

Sickle cell disease is a lifelong condition, passed down from a parent's genes, resulting in abnormally shaped red blood cells that are hard, sticky and clump together, which reduces the blood's ability to carry oxygen throughout the body and to vital organs.

"The most common manifestation of this disease is pain, often in the bones, caused by occlusion of small blood vessels by sickled red blood cells. These episodes are termed 'crises' and are triggered by various factors, such as extreme heat or cold, infection or physical exertion. They can go on for days at a time," said Srila Gopal, MBBS, a UC San Diego Health hematologist who leads San Diego's only [adult sickle cell disease clinic](#), supporting child-to-adult transition health services.



Pictured as a child, Vanessa Hughes spent a lot of time in hospitals getting treatment for chronic pain and other complications caused by sickle cell disease.

“By adulthood, many patients are at increased risk for bone necrosis and poor skeletal health, in addition to other organ dysfunction,” said Gopal. “There are a handful of medications to modify the course of the disease or improve quality of life, but access and compliance can be challenging for many individuals living with this disease.”

Stem cell transplantation offers promise for people living with this disease, but it is often not practical because of lack of suitable donors. In reality, children and adults often find themselves in emergency rooms seeking pain medications.

The Centers for Disease Control and Prevention (CDC) estimates the average life expectancy for people with the most severe form of sickle cell is 30 years lower than that of people without it. In California, this gap is even wider, with an estimated lifespan of just 37 years for patients with sickle cell

disease.

Part of the problem is that, as children, people with sickle cell disease usually have access to comprehensive pediatric services that provide them with disease management skills and specialists to help them during and after a crisis. As they transition out of pediatric programs, adults get lost in the maze of health care and their well-being suffers.

“We know that having a way to offer coordinated care changes patients’ lives. And helping patients transition from pediatric to adult care makes a world of difference,” said Gopal. “It reduces emergency room visits and hospitalizations while improving quality of life. Our goal is to make comprehensive health care more accessible to people with sickle cell who have not been part of organized care.”

Governor Gavin Newsom’s 2019 Budget Act created the Networking California for Sickle Cell Care Initiative, directing \$15 million to expand services in several counties throughout the state by creating at least five new centers in priority areas, including the specialized clinic at UC San Diego Health.

According to the CDC, the condition affects roughly 100,000 Americans, primarily persons of African ancestry. One out of every 365 African American babies is born with this condition in the United States. One in 13 African American babies are carriers of the sickle cell trait.

Located at UC San Diego Medical Center in Hillcrest, the comprehensive sickle cell clinic includes nurse case manager support, same-day infusion services, nutrition services, social work, access to clinical trials and coordination with other specialties. The team includes Gopal, a physician assistant, nurse practitioner, nurse case manager and social worker. The California initiative will add a community health worker who will connect the clinic to the community by assisting with setting up appointments and transportation as well as speaking with patients about medication adherence.

“Transitioning from pediatrics to a hospital with all adults was scary at first, but I feel I’m being heard by my doctor,” said Hughes. “My friends with sickle cell who go to other hospitals don’t get care from people who specialize in sickle cell or do research. The care they receive isn’t personalized for them individually. I definitely made the right choice by choosing UC San Diego Health with specialists who also do research.”



Now 24 years old, Vanessa Hughes transitioned from pediatric health care services to San Diego’s only adult sickle cell disease clinic at UC San Diego Health where she receives comprehensive care that is helping her stay out of the hospital.

Working together, the multidisciplinary team that includes hematology, emergency personnel, pain management and a same-day infusion center can treat patients more quickly and effectively to control pain and reduce the risk of complications, said Gopal. Sickle cell disease can cause chronic pain, pulmonary hypertension, chronic kidney disease, multiple organ failure and increased stroke risk.

Patients who experience a health crisis can get help from the only same-day infusion center in San Diego County. Here, patients can receive their first dose of pain medication immediately, as well as oxygen, hydration or other necessary treatments instead of waiting in an emergency room.

Until recently, when Hughes had a pain crisis, the best solution was fluids to rehydrate her body and pain medications. For a time, she would undergo blood transfusions, but the side effects caused problems, such as iron overload, which would lead to additional issues.

At UC San Diego Health, Gopal offers red blood cell exchange apheresis, a nonsurgical therapy that removes the patient's red blood cells and replaces them with donor cells. Hughes began receiving the treatment in 2019.

"I am coming up on one year of not visiting the ER thanks to apheresis and the same-day infusion center," said Hughes. "I don't have to go rushing into the ER every time I feel a pain crisis coming on. I just call my doctor and request an appointment at the infusion center where I receive what I need to try to feel better instead of landing in the ER again."

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