



Ashley Holley

SICKLE CELL

“It’s not just about the cure—it’s about improving the quality of life for patients until there is a cure.”

I'm an adult living with sickle cell disease, and I've spent the majority of my professional career studying it, first as a research assistant in a biochemistry lab and now both as a clinical research coordinator and hematology oncology medical practice administrator.

I was diagnosed shortly after birth, and it was always clear to me that I was different. As the daughter of two diligent parents, I was prepared for anything. Every morning, I would sit down to breakfast with penicillin, folic acid, and vitamin C. I hated those pills, but I knew that I would be very sick without them. My parents convinced the school principal to set aside one hour during the day for me to use her office to rest as well as enforce mandatory water breaks for me during recess.

I made it through my first pain episode when I was about eight years old. The pain was excruciating and the speed of onset was terrifying. I recovered quickly and moved on with my life, still without a full understanding of the magnitude of my illness. When I was 14 years old, all of that changed. I went to the emergency room one evening with a fever and pain. After a second round of x-rays, they noted lower lobe infiltrate. The doctor told me I was suffering from acute chest syndrome, something like pneumonia, but not really.

Over the next couple of days, my condition worsened. My ACS progressed and eventually my left lung collapsed. My hemoglobin levels dropped as a result of aplastic anemia induced by what was thought to be parvovirus at the time. I also developed an infarct in my spleen which caused my spleen to collect my circulating blood.

I almost lost my life during those two weeks, which shifted the way that I viewed and coped with my disease. Every day, I was tortured with the uncertainty of what was turning into a very life-threatening and unpredictable disease.

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- Sickle cell disease is an inherited blood disorder that affects red blood cells.
- When sickle-shaped cells block small blood vessels, less blood can reach that part of the body. Tissue that does not receive a normal blood flow eventually becomes damaged.
- The sickle cells also block the flow of blood through vessels resulting in lung tissue damage (acute chest syndrome), pain episodes (arms, legs, chest and abdomen), stroke and priapism (painful prolonged erection).
- Health maintenance for patients with sickle cell disease starts with early diagnosis, preferably in the newborn period and includes penicillin prophylaxis, vaccination against pneumococcus bacteria and folic acid supplementation.

Source: Sickle Cell Disease Association of America — www.sicklecelldisease.org

As I grew older, my disease impacted every part of my life. I developed avascular necrosis of my left hip and it quickly advanced to stage four. It relegated me to crutches and a wheelchair for two years, and I had to discontinue sports and dancing. I'm still waiting for it to give out so that I can replace it.

In college, my roommate and I studied for many mid-terms and finals in my hospital room when I was lucid enough. I managed to finish in five years, despite withdrawing for a quarter for health reasons. I've had about six additional run-ins with acute chest syndrome, and there's growing concern about whether I'll develop pulmonary hypertension or a chronic lung disease.

My echocardiogram seems to indicate that I'm at risk, but no definitive diagnosis has been made and no plans or options for treatment have been presented.

Drs. Clarice Reid, Marilyn Gaston, and Roland Scott changed the medical landscape, and greatly improved sickle cell patients' survival with their work on prophylactic penicillin, newborn screening, and the establishment of comprehensive sickle cell centers and a national sickle cell disease organization. Unfortunately, advocacy has waned and clinical advances are slowing. Medicine has managed to keep us alive, but our quality of life is compromised by the accumulation of co-morbid conditions such as pulmonary hypertension, avascular necrosis, and other degenerative organ diseases.

I'm hopeful we can finish the work that has been started. It's not just about the cure—it's about improving the quality of life for patients until there is a cure.

Ashley Holley was a patient speaker at the ATS 2012 International Conference in San Francisco, Calif.