Double Jeopardy

ANN-MARGARET HEDGES

During a routine checkup to evaluate Aaron Jackson's sickle cell disease, Nurse Practitioner Amy Cone discovered that the baby also suffered from retinoblastoma, a malignant tumor of the retina. St. Jude oncologist Carlos Rodriguez-Galindo, MD, credits Cone's early detection with saving Aaron's eye and possibly his life.

Imagine finding out that your newborn son has an inherited blood disorder. Still reeling from that diagnosis, you discover that he also has cancer. St. Jude helps one family cope.

By Elizabeth Jane Walker

To watch Aaron Jackson at play is to catch a glimpse of the man he will become. Aaron punches the keys of a computer game with the focus of a top executive typing a proposal. Arranging puzzle pieces, he furrows his brow with the concentration of an artist crafting a masterpiece. And the incandescent smile he bestows on his doctor and nurse practitioner foreshadow the gratitude of a man who understands just how fortunate he is.

At the seasoned age of 2, Aaron has already endured more hardship than many adults. His trials began with the prick of a needle only minutes after birth. That blood test revealed that he had the most severe form of sickle cell disease, an inherited blood disorder of the red blood cells.

His mom, Racheal, was shocked at the news. "His dad and I never knew that we had the sickle cell trait," she says.

Rocky beginning

Sickle cell disease arises from a mutation in the gene for hemoglobin. Aaron inherited one copy of the

mutated gene from his mom and another from his dad, giving him a one-in-four chance of developing the disorder.

Sickle cell disease causes red blood cells—normally round and soft—to elongate into hard, boomerang-shaped rods that hook together, clogging blood vessels. The disorder may cause severe pain, organ damage, strokes, seizures and even death.

When he was less than a week old, Aaron was admitted to the ICU in his local hospital. For 21 days he battled meningitis, a life-threatening infection that causes inflammation of the membranes covering the brain and spinal cord.

"I was terrified," Racheal admits. She soon learned that sickle cell anemia makes her son more susceptible to meningitis and other infections.

Aaron's doctors referred him to St. Jude Children's Research Hospital where Jane Hankins, MD, of St. Jude Hematology explained that Aaron would need twice-daily doses of penicillin until age 5 to protect him against further infections.

Double whammy

Racheal spent the next few months grappling with her child's serious medical condition.

"My other kids had been completely healthy," she says. "This was the first time that I'd ever had to deal with these kinds of health issues."

She took Aaron to St. Jude for regular checkups. Peering into Aaron's eye with an ophthalmoscope during one exam, Nurse Practitioner Amy Cone noticed something odd.

"Have you ever looked at photos where some of the people have red eyes?" she asks. "What you're seeing is the light reflex. When I looked in Aaron's eye, I didn't see that nice, bright red color. Instead, I saw a solid white area."

Cone knew that the ominous white reflex could indicate retinoblastoma, a malignant tumor of the retina. She asked Carlos Rodriguez-Galindo, MD, of St. Jude Oncology to examine Aaron. At first glance, the anomaly was almost imperceptible. "In spite of the fact that I knew Amy had seen a white reflex, it was not easy to find," Rodriguez-Galindo says.

Clinicians at most other sickle cell clinics may not conduct eye exams as part of routine sickle cell checkups.

"Typically, a pediatrician would just focus on the standard problems that accompany sickle cell disease: issues with the spleen, liver, lungs, bones or development," Rodriguez-Galindo says. "Doing a careful light reflex exam is uncommon."

Further testing revealed that Aaron did, indeed, have retinoblastoma, a cancer that strikes only 10 to 14 children out of a million.

In retrospect, Racheal recalled a few instances in which she had noticed a fleeting abnormality in her baby's eye.

"For a second, it would look like a lazy eye that would straighten out," she told Cone. "If we were in a dim light, I could see something like a white cloud in his eye. But I never dreamed it was cancer."

Rodriguez-Galindo says Cone's thorough examination definitely saved Aaron's eye and possibly his life.

"At least 90 percent of kids who have retinoblastoma in one eye must have that eye removed," he says. "The 5 to 10 percent of patients whose eyes can be saved are the ones who are noticed very early. Aaron's was a rare case of unilateral retinoblastoma that was diagnosed early."

Teamwork in action

St. Jude clinicians designed a treatment plan specifically for Aaron. Within a few days, he was receiving his first round of chemotherapy.

Aaron underwent seven courses of chemotherapy and three laser treatments to eradicate the cancer. Initially, the dual diagnosis was a concern to clinicians.

"Chemotherapy can put a child at risk for infection and require transfusions. Having sickle cell disease gives you those same problems," says Amber Yates, MD, Oncology fellow. "But Aaron was extremely fortunate. During all of his treatments, he had no infection and only had to undergo one blood transfusion, which is impressive."

PETER BARTA

Occupational Therapist Jessica Sweeney evaluates Aaron's progress during a Rehabilitation Services appointment. During the session, Sweeney offers his mom, Racheal (pictured at left), creative tips to help Aaron avoid treatment-related developmental delays.

Throughout the process, St. Jude clinicians allayed Racheal's fears and taught her how to assist in her son's care, which instilled a sense of empowerment.

"I couldn't have found a more caring hospital for Aaron, because they were so compassionate," she says. "They also taught me a lot. Parents at St. Jude receive a kind of medical education."

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Although Aaron has finished his cancer treatment, he returns to St. Jude every eight weeks for follow-up care. Clinicians in the sickle cell and solid tumor clinics coordinate appointments to minimize the amount of time that Aaron must spend at the hospital. "It's highly unusual for one patient to have two different diseases and to be seen by two separate clinics at St. Jude," Yates says.

Looking ahead

In the hospital's Rehabilitation Services department, Aaron carefully inserts large plastic coins into a piggy bank. Occupational Therapist Jessica Sweeney watches Aaron closely, offering encouragement and checking for any problems he may encounter.

"Because Aaron has some visual limitations related to his cancer, he's at risk for some developmental delays," Sweeney explains. "We monitor his progress and educate his mom about ways to facilitate his development at home. Right now we want to make sure that this little guy can play and just be a kid."

Occupational therapy is just one of many appointments Aaron keeps on a regular basis. During exams under anesthesia, physicians look through the pupil of Aaron's eye into the actual tumor bed. Any new tumors that arise can be removed immediately with a laser. Racheal breathes a sigh of relief after each exam, when she learns that Aaron's eye shows no further evidence of cancer.

"The main goal of retinoblastoma treatment is to save Aaron's life. The secondary goal is to try to save his vision," says Mary Ellen Hoehn, MD, of St. Jude Ophthalmology.

In the St. Jude Eye Clinic, Hoehn shows Aaron a card whose face is divided into halves—one containing wide, black stripes and the other printed with a bland, gray background. "I watch him to see which way he looks," explains Hoehn, who checks Aaron's vision every few months. "If children see the stripes, they will look at the stripes. If they just look away, you know that they're not seeing them."

As the test progresses, Hoehn uses cards with thinner stripes. "This test gives us a rough correlation of his level of acuity," she explains. Aaron currently has about 75 percent usage of the affected eye.

Tiny tornado

Aaron runs through the house scattering toys in his wake. Decidedly independent, the toddler insists on dressing himself and frequently asserts his desires by repeating his favorite mantra: "No!" Aaron's curiosity, mercurial temper and boundless energy both delight and exhaust his family.

Aaron has endured only two pain crises thus far as a result of his sickle cell anemia. If the duration and severity of his pain crises increase significantly, St. Jude clinicians will determine whether he is a candidate for hydroxyurea, a drug that causes the body to produce healthier blood cells. For the past decade, St. Jude researchers have been evaluating the effectiveness of hydroxyurea in children. The hospital is heading a national trial to determine the drug's ability to limit organ damage in infants.

"If the results of current St. Jude research confirm the benefit of hydroxyurea, then all infants with sickle cell anemia may be given the drug in the future," Hankins says.

With his blood disorder currently under control and his cancer in remission, Aaron spends his day in nearly constant motion. He rides toy trucks, tosses balls with abandon and begs his mom to go outside and play. It's tough work being a 2-year-old, but this phase won't last long. Before he knows it, Aaron will be an adult—perhaps pursuing a career, perhaps raising a family. But one thing's for certain: He will be living a life made possible by St. Jude. ●

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Aaron Jackson plays with the stethoscope of Amber Yates, MD, Oncology fellow. Although Aaron has finished his cancer treatment, he returns to St. Jude every other month for follow-up care in the sickle cell and solid tumor clinics.