

Health

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Girl with rare disease doesn't know pain

PATTERSON, Georgia (AP) -- Ashlyn Blocker's parents and kindergarten teachers all describe her the same way: fearless. So they nervously watch her plunge full-tilt into a childhood deprived of natural alarms.

In the school cafeteria, teachers put ice in 5-year-old Ashlyn's chili. If her lunch is scalding hot, she'll gulp it down anyway.

On the playground, a teacher's aide watches Ashlyn from within 15 feet, keeping her off the jungle gym and giving chase when she runs. If she takes a hard fall, Ashlyn won't cry.

Ashlyn is among a tiny number of people in the world known to have congenital insensitivity to pain with anhidrosis, or CIPA -- a rare genetic disorder that makes her unable to feel pain.

"Some people would say that's a good thing. But no, it's not," says Tara Blocker, Ashlyn's mother. "Pain's there for a reason. It lets your body know something's wrong and it needs to be fixed. I'd give anything for her to feel pain."

The untreatable disease also makes Ashlyn incapable of sensing extreme temperatures -- hot or cold -- disabling her body's ability to cool itself by sweating. Otherwise, her senses are normal.

Ashlyn can feel the texture of nickels and dimes she sorts into piles on her bedroom floor, the heft of the pink backpack she totes to school and the embrace of a hug. She feels hunger cravings for her favorite after-school snack, pickles and strawberry milk.

That's because the genetic mutation that causes CIPA only disrupts the development of the small nerve fibers that carry sensations of pain, heat and cold to the brain.

"There are all kinds of different nerve cells that help us feel different sensations," says Dr. Felicia Axelrod, a professor of pediatrics and neurology at New York University School of Medicine. "You can have one sense removed, just like you can lose your hearing but still smell things."

Number afflicted unknown

Specialists such as Axelrod don't know how many people suffer from CIPA. As director of a treatment center that specializes in CIPA and related disorders,

Axelrod has 35 patients with the disease on file. Only 17 of them are from the United States. Japan has the world's only association for CIPA patients. It has 67 members.

In Patterson, a rural town of 800 people in southeast Georgia, John and Tara Blocker had no idea the disorder existed before they took Ashlyn to the doctor for a bloodshot, swollen left eye when she was 8 months old.

The doctor put drops in Ashlyn's eye to stain any particles that might be irritating it. The infant smiled and bounced in her mother's lap while the dye revealed a massive scratch across her cornea.

"They put the dye in her eye and I remember the look of puzzlement on all their faces," Ashlyn's mother says. "She was not phased by it by any means."

Tests by a geneticist led to Ashlyn's diagnosis. To have the disorder, Ashlyn had to inherit two copies of the mutated gene -- one from each parent.

Ashlyn's father, a telephone technician, and mother, who holds a degree in physical education, were largely on their own in learning to cope with their daughter's strange indifference to injury.

Many things they couldn't anticipate. Ashlyn's baby teeth posed big problems. She would chew her lips bloody in her sleep, bite through her tongue while eating, and once even stuck a finger in her mouth and stripped flesh from it.

Family photos reveal a series of these self-inflicted injuries. One picture shows Ashlyn in her Christmas dress, hair neatly coifed, with a swollen lip, missing teeth, puffy eye and athletic tape wrapped around her hands to protect them. She smiles like a little boxer who won a prize bout.

Her first serious injury came at age 3, when she laid her hand on a hot pressure washer in the back yard. Ashlyn's mother found her staring at her red, blistered palm.

"That was a real reality check for me. At that point I realized we're not going to be able to stop all the bad stuff," Tara Blocker says. "She needs a normal life, with limitations."

So when Ashlyn goes to her kindergarten class at Patterson Elementary School, she gets daily check-ups with school nurse Beth Cloud after recess. Cloud and Ashlyn's mother discussed having her wear a helmet on the playground, but decided it would look too odd.

And when teacher's aide Sue Price puts ice in Ashlyn's chili at lunch, her dozen classmates get ice in theirs too.

Infections with no outward symptoms also concern them. They heard of a case where a child with CIPA had appendicitis that went untreated until her appendix burst.

"It's a lot to take in. It opens your eyes to things you wouldn't normally think about," says Tara Blocker. "If she sees blood, she knows to stop. There's only so much you can tell a 5-year-old."