



## Family of Lewisville boy with failing sight looks for answers amid uncertainty



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What do you see when you look through your eyes? Is it simply what's before you — a landscape of objects, a palette of textures?

Or is it something deeper — like visions of what could be or memories of what once was?

In the case of Zach Thibodeaux, it's what you can, while you can.

Last month, in the offices of Dallas' Retina Foundation of the Southwest, what 9-year-old Zach saw was this: a series of flickering lights, viewed in near-total darkness through the cavity of a hollow sphere straight out of Jules Verne. An electrode gripped his numbed eyeball, capturing his electrical responses.

Nearly two years ago, the fourth-grader at Mary Immaculate Catholic School in Farmers Branch was diagnosed with cone-rod dystrophy, a genetic disease that — despite some promising gains — remains without a cure.

His return visit to the medical facility that had finally diagnosed his condition marked a small milestone. While his parents didn't expect much positive news, what they did hope for was something tangible to hold on to after those years of difficulty and uncertainty.

As Zach's sight has rapidly diminished, they've groped for answers. At the same time, they've nurtured their science-obsessed son's curiosity and boyish *joie de vivre*, all while preparing him for the prospect of blindness and his growing academic challenges.

See Zach now compared with two years ago, and you'll see a boy who's excelled at Braille, mastered the abacus and learned to get around with a cane.

Assistive technologies have helped him with reading and communication, and in January, he earned a companion dog to help prepare him for the responsibility of having a guide dog someday.

Heading into the Retina Foundation exam with his mother, Johanna Uek of Lewisville, Zach was chatty and fidgety, having just returned from family trips to Cape Cod and California.

With the help of an airline discount, Uek had also taken him on a quick jaunt to Niagara Falls — part of an ongoing mission to provide iconic images he'll no longer have a chance to see once his sight is gone.

After a blood draw, foundation researcher and clinical trials coordinator Kirsten Locke gave Zach an eye-chart test that stretched his patience; from the other side of the small room, his left eye covered with a patch, he couldn't read any letter smaller than 6 inches high. He saw an N as an A, a B as an F; a V drew a long pause.

"Can you give me your best guess?" Locke asked.

"I just see a bunch of sparkles," Zach answered.

Finally it was time for Zach's electroretinogram, or ERG, in which he would gaze into the hollow sphere called a ganzfeld stimulator. A few eye drops would numb his eye for the first electrode.

She fastened a second one to his head.

"What's that for?" he asked.

"It's so you're grounded. But not because you've done anything wrong," Locke said.

Zach smiled. And with a patch over one eye, the electrode gripping the other and wires trailing from his head, he seemed a cyborg, the kind of experiment he would probably be excited to be working on.

"I can tell why it would hurt if it wasn't numb," he said.

Said his mother: "You're a brave little trouper."

### **Genetic condition**

The Retina Foundation sees four to five new patients weekly, all with genetic conditions for which few to no treatments exist. The most common is retinitis pigmentosa, which eliminates sight at the periphery and moves in.

The next most common is cone-rod dystrophy, whose roots lie in ABCA4, the same gene that causes Stargardt disease, another degenerative eye condition.

Zach's problems began to surface several years ago. He had trouble seeing at night. Bright light was tricky, too. At school, his math scores faltered as he misinterpreted numbers on the board or missed seeing them altogether.

Two doctors told Uek the same thing: His eyes look fine; he's young; he's stressed; he's just getting used to his glasses. But then he started running into things. His face was practically on the table as he did his homework. He wouldn't make direct eye contact.

Four ophthalmologists and a neurologist later, Zach came to the foundation, where a similar ERG unveiled the chaos within his seemingly normal eyes. "He sat so still," Uek recalled. "He was so determined to find out what was going on."

Deep within, the system had malfunctioned. Typically, waste produced by our electrical reactions to light is absorbed by the eye. In Zach's case, the process had broken down, and the toxic buildup was killing the rods and cones of his retinas.

Typical of the disease, the damage has ravaged the center of his vision and moved outward. The sight remaining at his periphery helps him navigate and stay oriented. On the surface, though, his eyes look normal.

"He gets stressed out," Uek said. "It's really hard, because when people see him, he does so much they don't think he's blind, and the things he can't do, they don't get it."

"We went to get him a passport, and the lady said, 'Look straight ahead,' and he said, 'I can't.' I had to tell her he was legally blind."

### **Fighting on all fronts**

Here is what his parents see: a growing boy who for years seemed like any other, a boy full of love and life and potential now threatened by a force no fault of his own.

In response, they've tried all they can to help him fight what seems a losing battle. Prayer circles early on, then medical care and research.

Last fall, Uek and Zach saw optometrist Stephanie Fleming of Dallas Services' Low Vision Clinic, who noted promising studies involving stem-cell treatments or artificial retinas in places around the globe, some of which Uek had already heard about.

She and Zach are all over the latest research, seeking anything that could help his vision or slow or even reverse his deterioration — medical developments, supplements such as vitamin A or acai — even kale.

"Oh, boy," Fleming said when she heard that. "'Thanks, mom.' The boy can eat kale all day long, and it won't help his vision that much. I believe in french fries — there's some benefits there. It makes the psyche *much* happier."

So yes, french fries and ice cream are part of Zach's regimen, too. But medical advances are still out of reach, untested. Fleming encouraged Zach to be patient.

"You don't have to be the first person out of the gate to try it," she said. "You're young. ... There's some promising work with artificial retinas. Stem-cell research is even more promising."

Last month, at the Retina Foundation, Uek told CEO and chief scientist David Birch about advances she'd heard about in China. Maybe they should take him there?

"He's going to be better off waiting until sites open up in this country," Birch said.

One British firm has made exciting progress in gene-replacement therapy, and it shouldn't be long before trials are available in the U.S., he said. As with televisions, everything improves every few years.

He agreed stem-cell research, including trials being conducted at UCLA, looks most promising at the moment.

"We can put stem cells into the retina right now, but they still don't make the connection to the brain," Birch said. "But within a few years ..."

The day's examinations showed Zach's optic nerve and the surrounding blood vessels continuing to deteriorate as his photoreceptors — which capture light and convert it into electrical signals sent to the brain — are being killed off.

Later, Uek said: "I knew it'd be bad news. But it gave us a reference to go with. You know, we don't have forever."

As they left the foundation offices, Zach put on the oversized sunglasses they'd given him to protect his eyes from the sun. "OK, LL Cool J," Uek said as the elevator doors opened.

Through his eyes, Zach still saw a world of imagination that only a child's eyes can create.

"Watch, Mom," he said as they stepped inside. "This is how you walk like a squid."

And he spread his arms and legs wide and danced side to side, wiggling, as Uek shook her head, smiling.

### About the series

**Nine-year-old Zach Thibodeaux** is going blind, the result of a condition called cone-rod dystrophy, a degenerative disease for which there is no cure. In "Zach's Journey," staff writer Marc Ramirez and our photographers are chronicling the Lewisville boy's passage into darkness.

**READ** Zach's full story in our e-book Zach's Journey, available for all e-readers at [dallasnews.com/ebooks](http://dallasnews.com/ebooks).

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