

A rare brain disorder robs children of language and leaves doctors grasping for answers.

BY LIZA LENTINI PHOTOGRAPHY BY ERIKA LARSEN

When Cody Cawood was 3 years old, he knew all his colors and proudly boasted that he could count to 20. His grandmother, Mary Beth Staab—"Beth" to her friends—a bright woman with more energy than a Kansas tornado, would hop the few short blocks to her grandson's house to help him practice his numbers and his letters every single day. Cody delighted in a mischievous game of reciting all the way up to 19 only to announce 20 as "20-teen," whereupon he would burst into uproarious laughter, even after the joke had been repeated for weeks. Beth would then pretend to protest, Cody would laugh some more, and they would ritualistically start all over from the very beginning. ■ Born two and a half months premature, and with cerebral palsy, Cody overcame his challenges, according to Beth, "with a little bit of his mom and dad's stubbornness." At Kids' Cove, a local school for children with disabilities, Cody was curious, loved to talk, and made fast friends. "He had an infectious personality," Beth recalls. ■ It was shortly after Cody's fourth birthday, in October 2000, that Sara, his mother, found Cody lying in a pool of his own vomit. First his mother suspected that it was the flu. Without real reason for alarm, Sara simply catered to Cody, as any mother would her sick child. But a week later,

when the episode repeated itself, Sara noticed a distinct and troublesome characteristic in his behavior. It was understandable that a sick child would be lethargic, but he also had a glassy, distant look in his eyes, and he was not speaking, just staring and dazed. Sara urged her son to try to go to school, but the only response he could offer was his lazy, hollow gaze falling upon hers. When she called Cody's new pediatrician and relayed the problems, she was told not to worry. After all, children with cerebral palsy were prone to stomach difficulties, the doctor said. Cody would be fine. ■ This clearly wasn't the case. Over the next two weeks, Sara and Beth began to notice a change in Cody's speech. Sentences were now replaced with a single word, a distinct regression

boy, interrupted



for a chatty 4-year-old. Instead of his usual “I want a Coke,” he would just say “drink”; “want to go outside and play” was replaced with “bye,” and “I want to go to sleep” was now one single word: “nightie.”

The third week of Cody’s strange illness brought them more mornings of sickness, sweating, and lethargy. Suddenly now, there was an added feature, frightening and horrific. When asked what was wrong, Cody would turn and bang his head against a wall. If asked a question, he would stop, cry, and start beating himself with force. By December of that same year, Cody didn’t utter a single word. His only modes of expression were violence, confusion, and tears.

Auspiciously, Beth worked for a pediatric neurologist, Samira el-Zind, who performed an EEG, an MRI, and a series of other tests on Cody. The MRI showed no changes; the EEG showed the same abnormal electrical impulses in the left occipital region that had been there ever since he’d suffered a stroke in utero. The doctor did notice some abnormal right and left central spike wave activity, and in addition to Cody’s prescribed anticonvulsant medication Depakote, she put him on Tegretol. But nothing seemed to work. Cody’s behavior worsened quickly. The hitting became more rageful, and now no one could get close to Cody without being punched or even bitten. That January and February saw a desperate course of trial and error as Cody visited a round of doctors and specialists who prescribed a number of different medications, none of which brought Cody closer to his old self. Schooling was now becoming a serious obstacle, as teachers didn’t know how to handle him, and he had to wear a helmet at all times to protect himself from his own furious, beating fists. By March, Cody seemed to have no memory of language and meaning and would only occasionally parrot a random word he’d heard in the conversations around him. His family took advantage of their quiet time with Cody and would try to ease him into the same conversations he’d had just six months earlier: “What’s your favorite color? Do you want to count to 20?” But

Cody seemed incapable of ever responding verbally. A look of confusion would fall over his sad face, followed by the inevitable hitting, screaming, and biting.

There was no such thing as an ordinary day for those who cared for Cody. Once when Sara was driving him to school, he beat himself so badly that blood ran down his face. Sara pulled the car over, got into the backseat, and held his hands to try to calm him down. Meanwhile, some onlookers were watching, suspicious of her actions, as though she’d harmed her own child. Within minutes, a police officer arrived. “My son hits himself,” she told the officer. But he didn’t believe her and ordered Sara to let go of Cody. When she did so, Cody exploded in a fit of fury. Even

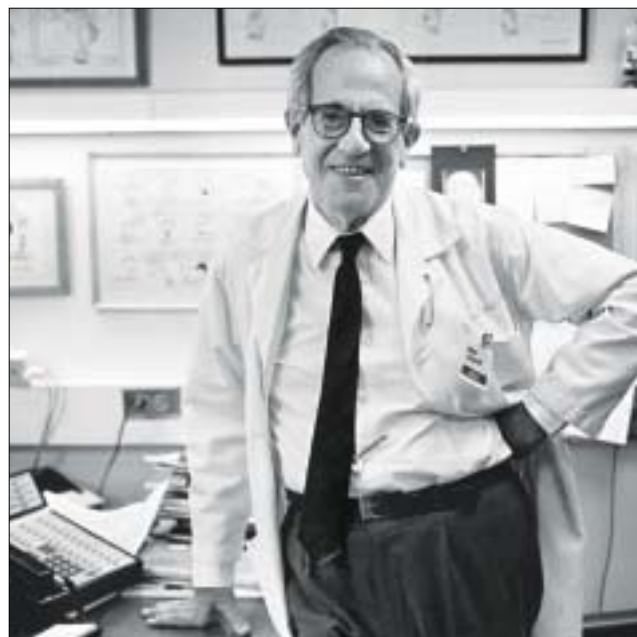
so, the officer wasn’t convinced. Sara urged him to follow her to Kids’ Cove, where the officer interrogated the principal about Cody and his mother and, after being assured of Cody’s condition, told Sara he would “let it go.”

In September of 2000, Sara had just entered nursing school, and the combination of classes and Cody’s illness brought her to the point of exhaustion. There seemed to be no preventing his rages. At one point, Sara and Beth considered handing Cody over to the state of Kansas so that his medical bills would be taken care of, but in the end Sara just couldn’t bring herself to give up her only child.

On the morning of September 11, 2001, Cody bit a nurse’s finger so hard he broke the bone, and he wouldn’t stop screaming and beating himself, even through the early afternoon. Beth arrived at school to pick him up and said he was like a “crazed animal.” Sara and Beth held Cody down and tried to calm him all evening, but by 9:30 that night they were out of options. Beth, still working for the pediatric neurologist Samira el-Zind, called the doctor in hysterics, asking to be seen right away: “Something has to be done for this poor child! He’s going mad right in front of us!” When they met the doctor at Wesley Medical Center, Cody



Cody at age 2½, before the onset of Landau-Kleffner syndrome, and neurologist William Landau, who first helped to identify the disorder.



was immediately put on the sedative Ativan, without any results. Doctors then tried a heavier sedative, Nembutal. There seemed to be no stopping the biting, hitting, and screaming.

The next morning happened to be Sara’s final exam in nursing, and so Beth sent her home just after midnight, as the doctors were administering a heavy dose of Thorazine. The Thorazine, without effect, was followed up by lithium, which also proved futile. At 2:30 a.m. the doctors finally found an effective medication, the tranquilizer Haldol. The doctor was worried that all these medications might make Cody unable to breathe on his own, but by 6:30 Cody was up and fighting, just as before. The hospital called in Mark Romereim, a pediatric psychiatrist, who ordered Cody to be put on Risperdal,

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eventually increasing Cody's dosage to that of an average adult. Kore Liow, an epileptologist, was then called in to consult. He ordered a video EEG at the Epilepsy Center and, after five days of monitoring Cody, diagnosed him with Landau-Kleffner syndrome.

No one in Cody's family had heard of Landau-Kleffner syndrome (LKS). A disorder in children often occurring between the ages of 3 and 7, LKS affects the child's ability to understand and use language. All children with LKS suffer from abnormal electrical brain waves, just like those in Cody's EEG. These children may suffer epileptic seizures at night, hence Cody's awakening sick and dazed. Hypersensitivity and aggression can accompany the disorder.

Before there was LKS, there was William M. Landau, neurologist, and Frank R. Kleffner, speech pathologist, two doctors who observed six children with acquired aphasia and convulsive disorder at the Central Institute for the Deaf. Landau and Kleffner studied the effects of drug and speech treatment on the children, noting in their summary: "In all cases a severe paroxysmal electroencephalographic abnormality, usually diffuse, is observed; electroencephalographic improvement tends to parallel improvement in speech re-education." "Syndrome of Acquired Aphasia With Convulsive Disorder in Children" was published in 1957—50 years ago this month—and caused quite a stir, opening the door for some 160 diagnoses between 1957 and 1990. It was Landau, along with John F. Mantovani, M.D., who published a follow-up study in 1980 of nine patients evaluated 10 to 28 years after the onset of LKS; the first six were from the original Landau-Kleffner study. Landau, who is still practicing at Washington University School of Medicine, wishes more progress had been made over the past five decades. "I'm glad to have survived the past 50 years," he says, wryly, but adds: "I'm very discouraged—we haven't learned much at all. We know nothing about cause yet." In terms of what went wrong, he says: "I think research has not been done adequately. We need multi-institutional organization of research programs that will examine potential causes and potential treatment in a systematic way. I'd like to tease the current population of potential investigators to get off their butts."

"The hallmark feature of Landau-Kleffner syndrome is a loss of receptive and expressive speech and language skills," says Sharon Willig, associate director of speech-language pathology for the American Speech-Language-Hearing Association (ASHA). The inability of those affected to understand the spoken word eventually hinders their own language skills, thereby rendering most of these children gradually or suddenly mute. It's for this reason that LKS children are often misidentified as developmentally delayed or possibly hearing impaired.

As with many other epileptic syndromes, LKS children often resume normal brain activity around age 15, when the brain cells are reaching toward maturation, perhaps spurred by hormonal change. Unlike similar conditions, children with LKS are often left with neurological impairments, even after the electrical abnormalities have returned to normal. The neurological effects

of LKS on a child can range anywhere from minimal to severe. For their parents, it's the hardest waiting game they'll ever have to play.

"I don't know if it's mysterious, but it sure is obscure," says Kleffner, who prefers to be called "Frank." Retired since 1991, Kleffner, now 82, moved on from LKS years ago and now finds it "at times a nuisance." Of his famed publication he says with humility, "I really didn't think at the time it was any great accomplishment." He credits Landau as being the foreseer of its impending significance, the real pioneer. "A few years after our paper, we got labeled a *syndrome*," he says, emphasizing the last word of his sentence with slight disdain. "I got a call from Landau one day asking me if I'd heard of Landau-Kleffner syndrome, and I thought he was kidding. Apparently he'd found it in a medical journal!" Kleffner now considers LKS "a badge of eponymic ignorance [the title of a paper by Landau]. We don't know much more now than we did then." To even attempt to research and write an article of this kind just "opens up a can of worms . . . you're only going to come up with loose ends."

Decades of continuous speculation have caused many scientists to refrain from offering opinions on LKS's cause and curability. One doctor who remains undeterred is Josiane LaJoie, a pediatric neurologist specializing in epilepsy surgery and treatment at New York University Medical Center. LaJoie has treated patients with LKS since 1999 and believes these children have an "inherent brain susceptibility. There is activation of abnormal circuits in the brain that prevent the normal function of the affected brain cells. The presence of these abnormal circuits may also inhibit the natural process of removal of unnecessary developing brain cells known as pruning. While we are asleep, the events and the experiences of the day are recorded as memory. The abnormal brain circuitry of LKS patients renders the brain incapable of imprinting memory, further impairing brain function. The areas affected are primarily responsible for speech."

The past 50 years of study and research have brought few concrete answers. "Much has changed since that landmark paper was published in 1957," LaJoie says. "We have subsequently learned that similar EEG findings can be found in a variety of patients with different clinical symptoms. Because of this, the margins of what defines a clear-cut LKS patient have been blurred." Kleffner himself, in his interview for this article, agreed, expressing concern that many LKS children were actually misdiagnosed. "Some use the term LKS to describe any patient with either language regression and/or EEG findings similar to those in LKS," LaJoie continued. "These patients may also have other symptoms not found in the typical patient with LKS. It is well known that children on the autistic spectrum can also undergo a similar regression and have EEG findings similar to those found in LKS patients. There is much debate about whether these patients should be classified as LKS or a variant of LKS. Just as with any medical condition, there is a spectrum of clinical symptoms among LKS. With increased awareness of vast possibilities of presentation of these patients, more patients are properly diagnosed and treated. However, not every patient will follow exact guidelines, making clinical decisions and classifications more difficult."

LaJoie believes that steroids are the best treatment for a patient's language issues. "If unsuccessful," she adds, "other options include



LEFT TO RIGHT: COURTESY BETH STAAB; COURTESY DR. LANDAU.

Think of the sensation of being locked in without any clue how to get out.

intravenous immunoglobulin therapy or possibly surgery. Multiple subpial transections [MSTs] are a type of disconnection procedure that allows the preservation of function of brain cells but prevents the spread of abnormal electrical impulses. MSTs have been found to be helpful in the recovery of language in LKS patients. Although MSTs are as invasive as they are controversial, it is sometimes a child's best chance at a functional recovery."

As for the future, LaJoie believes that magnetoencephalography, a technique that records the magnetic activity in the brain and helps determine the origin of the seizures, may be beneficial. "Well-designed research protocols are needed in order to better evaluate the effectiveness of various therapeutic interventions," she says. "With more research we will be better able to determine which patients should be referred for surgery quickly. I believe that we will be more aggressive in treating these patients and possibly intervening surgically much sooner, thereby optimizing their neurological outcome."

The outcome for LKS kids is decidedly varied, but aggressive speech and language treatment is a course of action that all specialists seem to agree on. The later the onset of LKS, and the more language development they achieve in their early years, the better. Children who have learned to read and write before the onset of aphasia can sometimes continue reading and writing even after their verbal skills seem to have disintegrated. Some children develop communication akin to sign language. "For children who are non-verbal as a result of Landau-Kleffner syndrome," says Sharon Willig of ASHA, "speech-language pathologists can recommend and provide assistance with the use of augmentative or alternative forms of communication, such as sign language, communication boards, or speech-generating devices. These communication systems are used in conjunction with verbal language treatment."

Patricia Prelock, professor and chairwoman of the Department of Communication Sciences at the University of Vermont, conducts treatment research for children with autism spectrum disorders, focusing on supporting their social skill development, a theory of mind, and peer interactions. Her approach is to avoid a prescribed therapy for every child and make things personal. "As opposed to looking at LKS alone, we found it very useful to consider who the child is in the context of their family, school, and community and what the child's strengths and unique challenges are, building on their strengths while accommodating their challenges."

The enigmatic nature of LKS leaves parents scrambling for answers their doctors can't provide. Working to alleviate that problem is Friends of Landau-Kleffner Syndrome (FOLKS), a U.K.-based non-profit organization dedicated to providing information and promoting research studies, which they publish online. Run by a volunteer committee of parents with LKS children, FOLKS aims to fulfill the need for LKS information among parents and medical professionals worldwide, serving about 300 members. Richard Budnyj took over as the secretary of FOLKS in 2003. "The biggest issue," Budnyj says, "is that unlike with many illnesses, our children look 'normal' and therefore are mistaken as children with a behavioral problem." Budnyj should know. His own son's LKS was diagnosed at age 3. He went through four regressions of speech and language, losing every word he knew. To control the boy's severe epileptic seizures, doctors put him into a coma. It seemed to help. Four years later, he's

a happy sports lover with a fantastic sense of humor. "He may still be behind about two years on speech and language, and he may never end up being the brightest child," says Budnyj, and then, gratefully, "but he has gone four years without further regressions. His next few years will be crucial to his development and what the future holds. All we can do is hope he continues to progress." Budnyj believes in accentuating the positive: "I don't know what the future holds for any of the children with LKS, but we can look to examples that give us hope."

Somewhere in the middle of Montana, a 52-year-old mother of two is proudly in the midst of her second career, as a cattle rancher. Her first, motherhood, garnered two accomplished children now in their midtwenties; the elder, a daughter, is now a practicing attorney, and the younger son is still in law school. "I've had a very lucky life," she says in a strong, affable tone. Though she prefers to remain unnamed, LKS researchers would know her best as "patient #7" from Mantovani and Landau's follow-up study. At 5 years old she was a talkative child, but then at 5½, at the onset of LKS, she began to stutter. "It's strange," she says, her voice clear with an enviable resonance, like that of a broadcaster, "this 'brain thing' happened so long ago, and it's still a part of my life." She remembers that time as mere "snatches" of memory and sensations. "It was as though I could hear things—they would come in, and come out garbled," she describes. Performing the EEGs then was a "very pokey" process—harsh tips were pressed into her young scalp. Her treatment took place at the Central Institute for the Deaf, where she attended weekly sessions to learn how to read, write, and talk, a frustrating challenge she remembers well. "I still have the notebooks and the flashcards with pictures of a chair or meatloaf—I had to relearn how to speak," she says. She was treated by Landau until she was about 16, then attended a Missouri state school and married her college sweetheart. "I had a relatively normal life," she says, and then, introspectively, "but I knew I was different. I felt different from the other kids I went to school with."

Truth be told, LKS children are different. They are as mysterious as they are rare, stumping medical science and allowing half a century to race by with few definitive answers. However, there's a lot to be learned from these children and the saintlike disposition required to treat, approach, and care for them. "We all take spoken language for granted," says LaJoie. "We wake up, say good morning to our





Beth Staab, Cody, Cody's mom, Sara Pinkston, and his stepfather, Chris Pinkston.

family, and are able to express our wants, needs, and emotions. Communication allows us to form connections with one another. Imagine that one day you were unable to connect with your loved ones with spoken language. Think of the feeling of being locked in without any clue how to get out. Think of the anxiety and frustration that you would feel. This is what these children go through. Often, these children are dismissed too quickly. However, with a modicum of time and attention, one can see a young child who desperately wants to be heard. The impact of the depth of their impairments not only on the child but on their entire family underlines the need to find a permanent treatment and cure for this condition."

As for Cody, seven years after the onset of LKS, doctors

haven't noticed any significant progress, and as of now, his family is planning to put him in adult handicapped housing when he finishes school at age 21. "I have a picture of Cody when he was a little boy at Kids' Cove climbing up the big chain-link fence that surrounded the playground," Beth says. "Now he doesn't even know what a fence is." It's still too early to determine if Cody will follow in the footsteps of many LKS kids or if his EEG will return to normal around age 15 and, even if it does, what the effects of living with LKS will be. For now, those who love Cody are remaining hopeful and taking each day in stride. "At 11 years old, he still loves for me to rock him on my lap, just like when he was a baby," says his grandmother with a smile. ■