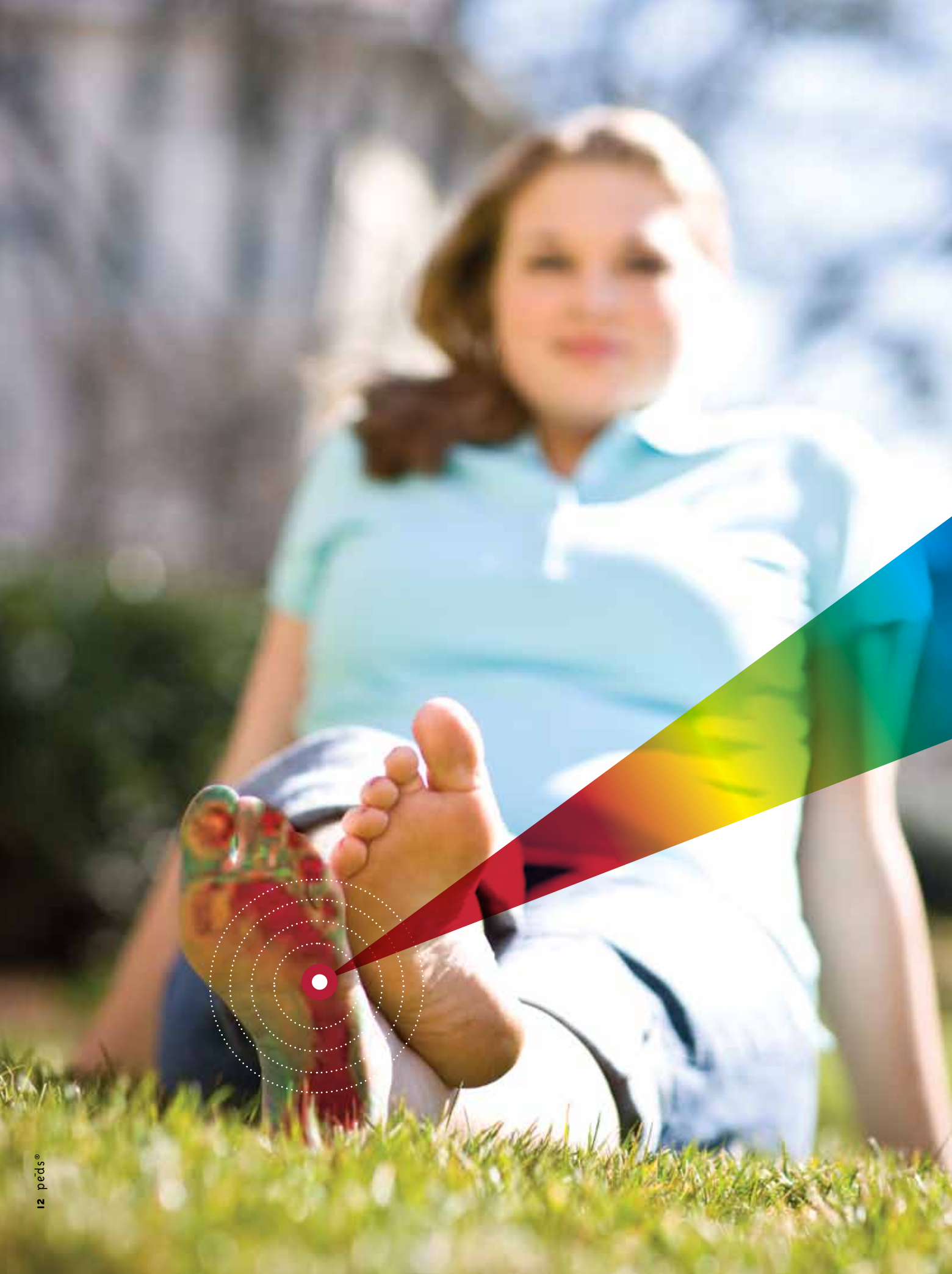

pain

Pain

Providing Accurate Diagnoses and Treatment Goals

When Tiffany Overby was 11 years old, she sprained her left ankle during a game of kickball, it seemed like a simple injury to both her orthopaedist and her mom. But, it was soon apparent that this was no ordinary injury. Although the swelling and discoloration dissipated, the pain did not. In fact, during the next few weeks, the pain became exponentially worse. "It was so bad that when her foot was propped up on a pillow, the air from bodies moving around would result in an escalation of her pain. When she stood up, her foot would turn blue," said her mother, Trish Overby. • Her orthopaedist then X-rayed her foot a second time to rule out a possible missed fracture but nothing appeared concerning. Both her foot and ankle appeared to be normal. But, for Tiffany, life became increasingly difficult for her and her family. "I quit my job to be available 24/7. I was online every day trying to understand what was going on. I knew I needed to find help for my daughter but I did not know where to go," Trish Overby said. "We knew that her pain was real but her peers started to tease her, saying she was just trying to get attention. Even her older brother was skeptical. In disbelief, we watched my daughter go from a totally active young teen who lived to play basketball to a totally inactive shell of her former self." • During the summer, Tiffany and her mom spent hours at the pool trying to desensitize her foot. The water helped to ease the pain—but it was a temporary measure. From that point, Tiffany saw a variety of professionals from pediatricians to orthopaedics to psychiatrists—seven in all. Some were dismissive. "One professional, right in front of Tiffany, said the problem was in her head, not in her foot. He instructed me to take her to a psychiatrist," Overby said. "Others were somewhat sympathetic but admitted that they really did not know what to do."

Finally, Peter L. Meehan, M.D., an orthopaedic surgeon at Children's Healthcare of Atlanta and Clinical Associate Professor of Orthopaedic Surgery at Emory University School of Medicine, contacted Claudia Venable, M.D., a pediatric anesthesiologist at Children's Healthcare of Atlanta and Director of the Children's Center for Pain Relief.

When seeing Dr. Meehan, Tiffany and her mother first learned that her painful foot probably was due to a condition known as complex regional pain syndrome (CRPS), formerly known as reflex sympathetic dystrophy (RSD).

"Dr. Venable called us that day to talk to Tiffany about being admitted to Children's," Overby said. "Her first words were, 'I think I can help you.' After months of pain, that was a defining moment in my daughter's life. She made an incredible connection with Dr. Venable. In Tiffany's eyes, that stood for hope."

Although Trish Overby was frustrated about the length of time it took to get a diagnosis and begin treatment, she also understood that this condition can be a puzzling disease. Children who have this ailment may not develop pain until weeks after a minor, or what even seems to be a trivial, injury. The pain may have a waxing and waning quality; the affected limb may have color changes, ranging from pink and warm to a dusky blue. The pain usually is completely out of proportion to the injury and patients describe unusual sensations, such as pins and needles, or shooting pains, like zingers or shocks. Usually in a distal extremity or, more commonly, the lower limbs in children, this issue commences with a burning pain that is extremely sensitive to touch. In fact, light touch can cause distinct pain. Children usually pull away as the examiner comes toward them and instinctively wince when they are touched. Many children with this condition have stopped wearing socks and shoes. The development of allodynia (pain caused by stimuli that are not normally painful) and hyperalgesia (increased sensitivity to pain) are distinctive features of this condition, and they are synonymous with the diagnosis of CRPS.

Unfortunately, children who experience this escalating pain with every touch look for ways to avoid putting weight on their foot. Some children hop to their first visit with their physician. Disuse of the limb makes matters worse. The extremity may appear swollen and shiny over time. The color changes are noticeable and the child readily describes how his foot turned blue. Over and over again, parents and children will comment on the fact that well-meaning professionals who are not familiar with this condition are quick to minimize the symptoms and advise the patient to get over it. So, as the pain continues, there comes an escalation in anxiety and sadness as the child becomes a disruption in both the family setting and at school.

The diagnosis of CRPS is based on the follow criteria: the presence of an initiating noxious event or cause of immobilization; continuous pain, allodynia or hyperalgesia in which the pain is disproportionate to any known inciting event; evidence at some time of edema, changes in blood flow or abnormal sudomotor activity in the region of pain; and a diagnosis excluded by the existence of other conditions that would otherwise account for the degree of pain and dysfunction. After the diagnosis is made, the first effort to manage the pain is to send the patient to an experienced physical therapist. The important treatment for CRPS, no matter when it is diagnosed, is utilization of the extremity. Children in an increased amount of pain sometimes find that this is an unpleasant challenge. Usually, the patient requires some pharmacologic treatment. The typical treatment begins with nonsteroidal anti-inflammatory agents, such as Motrin or Advil. Treatment progresses to tricyclic antidepressants, such as amitriptyline and nortriptyline, starting in lower doses and given at night. Anticonvulsants, such as gabapentin and pregabalin also are the next mainstay of treating this unusual, neuropathic pain. During the course of an escalating painful crisis, opioids may be used. While they might be tried, they are generally ineffective for the pain of CRPS.

Tiffany's pain crisis continued to wax and wane. Because of her pain, there were days when she could not tolerate going to physical therapy. There were many tearful times and Tiffany continued to feel helpless and distraught. Her mother was so concerned that she hid all the sharp objects in the house from Tiffany. There was a great deal of tension in the family. After her pain became increasingly intolerable and physical therapy could no longer be done, she was admitted to Children's for the placement of an epidural catheter to manage her pain. With the catheter in place, she was given local anesthetic and opioid and experienced immediate relief. Tiffany was proud to show off her ability to walk around the hospital. Her parents, brother and sister rejoiced to see Tiffany finally out of pain. The catheter was left in for several days while Tiffany was able to fully participate in her physical therapy routine. Thanks to many skilled and motivating physical therapists, Tiffany began to regain some physical strength. While in the hospital, she talked to a pediatric psychiatrist who helped her better understand the complex connection between the emotional and the physical pain she had experienced. In retrospect, Tiffany and her family felt that this was essential in getting everyone back on a more even keel. Dr. Venable remembers the day she left the hospital: "she was all but dancing."

Anyone who has lived with CRPS can tell you there are good days and bad days. Tiffany was pain-free for several months but continued to relapse. Flare-up after flare-up would present and these painful episodes often would coincide with an event at school: someone would accidentally step on Tiffany's foot or she would play too hard or experience pain around exam time. She consistently turned to Children's for relief. While a lumbar sympathetic block was given and physical therapy treatment was increased, it seemed the epidural catheter gave the most consistent relief for getting her pain level down to zero. On one occasion, the possibility of a Medtronic implanted pain pump was mentioned. The family became very enthusiastic knowing that there might be a longer lasting solution to her chronic pain. David Wrubel, M.D., a pediatric neurosurgeon at Pediatric Neurosurgery Associates at Children's Healthcare of Atlanta and Medical Director of the Children's Neuro Spine Care for Kids program, was called to implant the intrathecal pump. The hockey puck-size device with a chamber for medication was placed in Tiffany. This intelligent device drips micro-amounts

Complex Regional Pain Syndrome (CRPS)

(From Raj, Practical Management of Pain, Chapter Chronic Pain Management in Children)

- The pain generally follows mild trauma and is disproportionate in intensity to the inciting event
- The pain persists indefinitely if untreated
- The pain is asymmetrical without dermatomal distribution
- Early on there is vasodilation with increased temperature, hyperhydrosis and edema
- The skin can also be cool, red, clammy—vasomotor
- Later, there may be a vasospastic quality with persistent coldness of the affected area
- The intensity of the pain my lead to depression and anxiety, particularly if undiagnosed

Definitions:

- Type 1—no distinct nerve injury
- Type 2—involves a nerve injury
- Allodynia—pain is produced by innocuous stimuli such as stroking, which elicits excruciating pain (very characteristic of neuropathic pain)
- Hyperalgesia—a patient has a decreased threshold to pain—usually along a glove or stocking distribution
- Neuropathic pain—pain that is unusual with qualities described as burning, shooting, pins and needles
- Nociceptive pain—pain that is usual such as incisional pain or pain from a broken bone

of medication to Tiffany's cerebrospinal fluid. Luckily, this became Tiffany's salvation as it stopped her pain. There were no more pain crises during the next nine months. She started ninth grade and became the manager of the basketball team. She was closely involved with the sport she had hoped to play. "To my surprise, part of the manager's job was working out with the basketball team. Knowing Tiffany, I had never imagined that such an activity would be possible for her. It was exciting to see her at her best," said Dr. Venable.

Maurice Sholas, M.D., Ph.D., Medical Director, Children's at Scottish Rite Rehabilitation Services, and his partners have formed a working relationship with the Children's Center for Pain Relief. "Every surgeon, physician and nurse at Children's is skilled at managing pain from surgery to life-threatening illnesses like cancer. Our role is to go beyond the ordinary. Challenges arise when a child does not have that normal recovery period or when the child does not recover in a timely fashion and is in severe pain. In addressing these issues in children, we work together. "We coordinate all of the modalities," Dr. Sholas said. "We pivot from pain to function."

Although the Children's Center for Pain Relief is not a new department, the leadership is. Dr. Venable became the director of the Children's Center for Pain Relief six months ago and Dr. Sholas heads up the inpatient and outpatient rehabilitation team. Together, they have developed an approach that includes the Children's Center for Pain Relief, Neuro Spine Care for Kids program, as well as the Children's Psychological Services and Rehabilitation Services. Being involved in a variety of cases, whether the child has CRPS or some other painful condition, such as sickle cell disease, chronic back pain, fibromyalgia or post traumatic pain, these professionals strive to formulate a unified message and treatment plan for the staff, family and child.

Currently there are 28 inpatient beds at Children's at Scottish Rite and 15 to 20 outpatient beds dedicated to rehabilitation services. At Children's, there are three main campuses, as well as seven outpatient therapy centers and eight sports medicine centers. "We are ready, willing and able partners," Dr. Sholas said.

"Chronic pain is very frustrating for both the parent and the child but just having a name for the disease is a huge relief and allows us to prescribe a full range of treatments," he said. "It is the parent's instinct to protect a child, however, with a pre-adolescent or adolescent, it is not just the parent's job. The child has to learn to own the problem. If someone else does it for them, the treatment is less likely to be successful. I want to unburden parents and empower the child to do as much as he or she can."

Now 15, Tiffany is doing well. Despite an occasional flare-up, she is involved in school and her sports team. Her journey is far from over, however, instead of working with a team of 11 physicians, she now only sees three on a regular basis. She also attends a weekly physical therapy prescribed by Children's Rehabilitation Services near her metro Atlanta home.

Her parents realized how far she had progressed when she was able to wear high heels to a school dance. In the picture, she deliberately posed with her left foot toward the camera—a clear sign that she was becoming the same carefree girl they feared they had lost forever.

Claudia Venable, M.D., is a pediatric anesthesiologist at Children's Healthcare of Atlanta and Director of the Children's Center for Pain Relief.

Maurice Sholas, M.D., Ph.D, is Medical Director of the Children's Healthcare of Atlanta Rehabilitation Services at Children's at Scottish Rite.

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