



UNDERSTANDING NEUROPATHY

The three common forms of neuropathy found in patients with autoimmune disorders are difficult to diagnose, but they are treatable, and patients can find ways to cope.

By Jennifer Kester

Carolyn Spooner first noticed that she had trouble using her left hand to hit “control, alt, delete” on her computer while at work. But since it was only her middle finger that was numb, and Spooner is right-handed, the business systems analyst didn’t think it was a big deal. “We figured it was a pinched nerve,” she says. A few months later, in January, she started having gesticulations in her left arm. “They weren’t uncomfortable, but odd enough to make me nervous,” she says. It got progressively worse, and by May, her hand was almost nonfunctional. When she put her palm flat on a table, she could move only her pinky.

“I had no idea what the problem was and because it didn’t hurt, it didn’t make sense to me at all,” she says. The doctors tried nerve release surgery and other treatments, but nothing worked. The Candia, N.H., woman was shuffled from specialist to specialist in hopes of learning the cause of her numbness. Then, she stumbled upon a neurologist who thought he knew what the problem was. “He said he’d heard about it, read about it, but never seen it,” Spooner says. His diagnosis: She had multifocal motor neuropathy, a disease of the peripheral nerves, arms and legs.

What Is Neuropathy?

Neuropathy is a degenerative condition of the nervous system or the nerves. People with autoimmune disorders are more prone to peripheral neuropathy, which occurs when tissue in the peripheral nervous system — the communications hub that carries information from the brain and spinal cord to the rest of the body — comes under attack. Three types of peripheral nerves are affected: sensory nerves, which help with sensation; autonomic nerves, which assist internal organ functions; and motor nerves, which work with muscles.

Damage to the peripheral nervous system can interfere with the signals between the brain and the rest of the body. This damage can be caused by more than 100 types of peripheral neuropathy, each with its own set of symptoms and treatments. For people with autoimmune disorders, some common peripheral neuropathies include chronic inflammatory demyelinating polyneuropathy (CIDP), Guillain-Barré syndrome (GBS) and multifocal motor neuropathy (MMN). CIDP and MMN come about from immune system activities rather than from direct damage by infectious organisms, whereas GBS is believed to be brought on by a virus. All three

of these peripheral neuropathies also are demyelinating diseases, which means they destroy myelin, a fatty protein that protects many nerves. When the myelin is damaged, nerve impulses slow or even stop, resulting in neurological deficits.

CIDP has numerous symptoms that include muscle weakness, numbness and pain in the extremities, impaired balance and difficulty walking. The disease usually starts in the legs but can also begin in the arms. It can cause severe weakness and even death if left untreated. CIDP may have patterns of remission and relapse, but most patients will require some form of therapy for life. Although there is no cure for CIDP, it is treatable with corticosteroids. Some patients who don't respond to steroids or suffer from their side effects can use alternative drugs such as mycophenylate. Patients often see dramatic improvement with a therapy regimen with intravenous gammaglobulin (IVIG).

According to GBS/CIDP Foundation International, Guillain-Barré is the most common cause of rapidly acquired paralysis in the United States today, affecting one to two people in every 100,000. GBS has similar symptoms to CIDP, except GBS has a more sudden onset that often requires immediate hospitalization. Most reach the stage of greatest weakness within the first two weeks after symptoms appear, and by the third week, 90 percent of all patients are at their weakest, the National Institute of Neurological Disorders and Stroke says. If GBS doesn't get better or becomes a chronic issue, it is often rediagnosed as CIDP. GBS is treated with plasma exchange and high-dose IVIG. Once a GBS patient recovers, the disorder typically doesn't return.

MMN is a progressive muscle disorder characterized by muscle weakness in the hands. Unlike CIDP and GBS, MMN affects only the nerves' motor fibers — not the sensory fibers — so there's no numbness or pain. MMN's symptoms include muscle wasting, cramping and involuntary contractions or twitching of the leg muscles. The disorder is sometimes mistaken for amyotrophic lateral sclerosis (ALS, or Lou Gehrig's disease), but unlike ALS, it's treatable. Some patients have mild symptoms and require no treatment. Most receive IVIG as the first course of treatment, with 80 percent of patients showing improvement after the infusion, according to the Multifocal Motor Neuropathy Center at Johns Hopkins. An early and accurate diagnosis allows patients to recover quickly.

CIDP is the most common of the conditions, followed by GBS and MMN, according to Dr. Carol Koski, medical director of the GBS/CIDP Foundation International. "In all three of these disorders, there tends to be a male predominance — not quite a 2-to-1 male-to-female ratio," Koski says.

A Difficult Diagnosis

It's often not easy to diagnose peripheral neuropathy, and in most instances, the patient doesn't realize there is a real problem. In Spooner's case, she later realized that she had other MMN symptoms that she ignored at the time. "I was extremely fatigued. I was taking three- or four-hour naps on the weekends," the 54-year-old says. "I had no idea that had anything to do with my hand. I knew I was having trouble with my legs before I could prove it. I would walk with [my] dog, Piper, and it got to be that walking through uneven ground or snow was a problem."

Julienne Dallara wasn't hit with an onslaught of symptoms when she was diagnosed 13 years ago. "For a couple of weeks, I felt strange," she says. "I knew something was wrong, but I couldn't tell what. One morning, I couldn't urinate. So I went to my obstetrician, because that was the only doctor I had," says Dallara, who was a new mom at the time. Her obstetrician referred her to a neurologist, who then sent her to the hospital. "The next morning, I woke up in the hospital paralyzed from the waist down," she says.

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Dallara was diagnosed with transverse myelitis, a neurological disorder caused by inflammation across both sides of one segment of the spinal cord. Three years later, she also was diagnosed with CIDP. "They tried all kinds of stuff when I was in the hospital, but nothing seemed to change; I couldn't feel anything from the waist down." After 10 days in the hospital, she was transferred to a rehab facility so she could learn to live in a wheelchair. "They thought I would make a relatively full recovery because most people with myelitis do," she says. She went home with a wheelchair and walker and was working on getting better. Then, two months later, she had a relapse and became completely paralyzed from the waist down and couldn't stand up for four or five months, until she received chemo. "That got me back on my feet again," she says.

Treating Peripheral Neuropathy

Dallara's immune system is hyperactive and it attacks her body, including the myelin of her spinal cord. To stay healthy, doctors have to keep depressing her immune system. "They don't know what's causing it, or why," the Los Angeles resident says. "So for the last 13 years, I've been trying to keep the symptoms under control." To do this, she gets an infusion of IVIG every three weeks. "The way it makes sense to me is that in order to keep from being paralyzed, I can either destroy all the bad guys or I can flood my body with so many good guys that they outnumber the bad guys," she says. "By doing chemotherapy, we are killing the bad guys, but it's hard on me as well. What IG does is flood my body with good guys. It does that for a while and I can keep on going." She also takes medication to manage the pain. The first couple of years, she didn't have painful neuropathy, just paralysis, but over the years, the burning, buzzing and banding feeling — "like someone's wrapping Saran wrap really tightly around my leg," she says — has gotten worse.

Multifocal motor neuropathy is a progressive muscle disorder characterized by muscle weakness in the hands.

Spooner also uses IG to alleviate her symptoms. "If I don't have the IG treatment every four weeks, I immediately notice an increase in fatigue and start to lose ground in my hand," she says. "The IG has taken care of the hand problem, but I have more trouble now with my legs." To help her walk, she takes muscle relaxants regularly to control muscle spasms.

In addition, both Spooner and Dallara look for nonmedical ways to cope with their neuropathies. "I have found that the way for me to deal with it is to keep so busy that I don't notice it as much," Dallara says. "If I start to pay attention to it, it gets worse. I have noticed that when I get stressed, it gets worse." However, she also has to be careful to not overdo it. "The job I'm in is very demanding, and the hours are long," says the 49-year-old, who sells

handicapped vehicles. "It's not unusual to work a 12-hour day. It helps keep my attention off the pain, but it makes the pain worse when I stop. It's a balancing act." Spooner tries to work around her condition by planning her days carefully. "I work full time and I have to not plan anything in the evenings," she says. "If I'm out one night and I go to bed early the next night, I really need two or three nights to make up for the first night." Spooner also exercises regularly, focusing on building her core to help with her walking, and tries to eat well, cutting out diet soda and limiting sugar.

Both women picked up hobbies that don't require mobility or a lot of physical strength. Spooner has gotten into genealogy and knitting. It was particularly hard for Dallara to stop being as physically active, since prior to her paralysis she was an actor, singer and dancer. But she found creative outlets in oil painting and jewelry making.

Coping with Neuropathy

For those newly diagnosed with a neuropathy, Spooner advises them to find out what they can about their disease. But, she warns that they shouldn't be disappointed when the answers are, "Everybody's different" or "We don't really know." "You have to recognize the reality of the situation, but you also have to believe that there are going to be good days and that there are things you can do," she says. "A positive attitude makes a big difference." She also gets her own copies of tests and records and keeps it all in a three-ringed binder, which she brings to appointments so that she's always prepared. Another thing that Spooner finds helpful is listening to meditation tracks on her iPod to help her relax. "They re-energize me," she says. She also takes power naps during the day, sometimes ducking into an empty conference room at work to catch some z's. "It gives me the boost I need to get through the day," she says.

Dallara maintains that it's important to keep trying to get better. "In my line of work, I see people with many different disabilities, and I'm seeing more and more people diagnosed with neuropathy each year," she says. "More and more people are coming into my office saying, 'I'm in a wheelchair because I have neuropathy,' or 'I'm on a scooter because I have neuropathy.' It's not an excuse to stop living. Keep pushing for more and more that you can do." ■

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