Living with Scleroderma: Eating, sitting, bending, healing can be hard

Alvin Crowl lives with scleroderma (which sometimes means he can't enjoy his GTO as much), and he and wife Angie are organizing a cruise night to help raise awareness of the autoimmune disorder. THE NEWS JOURNAL/ROBERT CRAIG

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HELP THE CAUSE

Two upcoming events are being held to benefit the Delaware Valley chapter of the Scleroderma Foundation.

The first is "Stepping Out to Cure Scleroderma," a 5K and fun walk scheduled for 9 a.m. April 15 at Bellevue State Park. "Cruise for a Cure," a car show with raffles, concessions and other activities, will be 11 a.m. to 3 p.m. that day on the nearby grounds of Mount Pleasant High School. Spectators are free, but admission is $20 for those bringing a car for display.

For more information about the car show, contact Alvin and Angie Crowl at 425-5054 or anglcrowl@aol.com. To register for the race and walk, visit www.runtheday.com.

Alvin Crowl doesn't get frustrated when people ask him the reason for his tan in the middle of the winter. For Crowl, it's an opportunity to explain the many ways his autoimmune disease affects his body.

Crowl, 62, of Wilmington, has scleroderma, which causes his skin to harden and take on a darker hue. His fingers are so sensitive to cold -- and sometimes develop ulcers on the tips -- that he wears gloves almost year-round. It's too hard for him to bend over, so he uses a piece of plastic to help put his socks on. He wears shoes without tie-up laces.

But those are just the symptoms people can see. Crowl also suffers from digestive problems related to the condition. Last year, he was hospitalized with heart failure and internal bleeding related to his scleroderma. He received eight infusions of iron.

"He doesn't do much in the winter," said his wife, Angie, who is helping to organize a fund raiser next month to raise money and awareness of the disease. "Even when the weather is better, he's still affected. On a cold, rainy day in the spring, it's the same as if it was 20 degrees out for him."

Tough diagnosis

Named from the Greek words meaning "hard skin," scleroderma can be a frustrating disease for patients and physicians alike. It's caused by an excessive accumulation of scar tissue in different parts of the body, from the skin to small blood vessels. For reasons not completely understood, the mechanisms that usually control the accumulation of this tissue go awry, said Dr. Sergio A. Jimenez, director of the Scleroderma Center at Thomas Jefferson University Hospital in Philadelphia.

"So it keeps piling up. Just like strong, tough scar tissues can't be removed, it causes the skin to get thicker and affect the mobility of joints," said Jimenez, whose center draws people from across the globe who have already been diagnosed or who need confirmation of the disease. "It can also go to the internal organs, lungs, where it can affect the transfer of oxygen, so they can't breathe well. Many other organs are affected, resulting in renal failure, renal crisis, heart failure and problems in the intestinal tract."

That range of symptoms can make it hard for people to get the care they need. It can take five to 10 years for someone to be diagnosed with scleroderma, often because the symptoms aren't seen as being related, s aid Christine Gaydos, executive director of the Delaware Valley chapter of the Scleroderma Foundation. About 300,000 people are estimated to have the disease in the U.S. Women are disproportionately affected, about six to eight times more often diagnosed than men.

"I think in the beginning it can mimic other disorders, like rheumatoid arthritis or just pain in the joints," Jimenez said. "It takes a person who is aware of the disorder to actually

make the diagnosis."

How it began

Crowl first experienced skin symptoms years ago, when he was working on airplane engines for the Delaware Air National Guard. He thought the color change in his hands -- from pink to white and blue in the winter -- was the result of working with jet fuel and other chemicals. But in 1997, he found out the discoloration was the result of Raynaud's phenomenon, which occurs in about 90 percent of scleroderma patients.

Two years later, he found out the bigger issue was scleroderma. As his skin has thickened, he has lost some mobility, particularly with bending over. He also has had to deal with hard-to-heal wounds and ulcers on his body, including one that kept him from sitting for more than half a year. That was difficult, his wife said, because it meant the car enthusiast couldn't spend hours behind the wheel of his 1972 GTO.

"But even with everything that comes up, he still keeps a good attitude," she said. "You'd never know what was going on."

Because scleroderma is considered a spectrum disease, patients can experience a range of severity depending on what parts of the body are affected. Less-severe forms include hardened skin on hands and forearms, but more crippling cases can impact kidney, lung and heart function. Scarring also can damage the esophagus, causing problems with swallowing and acid reflux. In the intestines, it can cause bloating, gas and constipation that last for days. The only part of the body not affected by scleroderma is the brain.

"I have my days when my stomach doesn't feel right, so I won't eat. When I really have it bad, I can't eat for a week or two weeks -- I just throw everything up," said Crowl, who lost 30 pounds during an episode last year.

Jimenez said although there is no cure for scleroderma, there has been progress in treating its complications, improving the quality of life for patients. In some cases, patients have had symptoms go into remission or their progression halted.

In the past, patients died from kidneyrelated problems caused by scleroderma. But better treatment control, including dialysis, has reduced the number of deaths. These days, patients with scleroderma face the most risk from lungrelated complications, such as pulmonary hypertension, which occurs when the vessels in the lungs tighten, reducing the circulation of blood.

In addition to their symptoms, patients also have to deal with the challenges of living with a rare condition, Gaydos said, noting t hat scleroderma is considered by the National Institutes of Health to be a rare disease. That means educating people.

"People say, 'I've heard of it, but I really don't know a whole lot about it,'" Gaydos said.

WHAT IS SCLERODERMA?

Scleroderma is an autoimmune disease that affects connective tissues in the body. It causes changes in the skin, blood vessels, muscles and internal organs.

Hardening of the skin is one of the most visible signs of the disease, but this thickening of collagen also can happen to internal organs, including the lungs, heart and kidneys. It also can affect the intestinal tract, making it difficult to eat, swallow or get ride of waste in the body. About 300,000 people in the United States are estimated to have scleroderma. Women are six to eight times more likely to develop the disease than men.

Symptoms may include fingers or toes that turn blue or white in response to hot or cold temperatures (a condition known as Raynaud's phenomenon), joint pain and ulcers on the fingertips or toes, shrinking muscles and injured tendons. Patients frequently lose movement in their joints, especially the hands. The long-term prognosis for scleroderma patients depends on the severity of symptoms.

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SOURCES: Scleroderma Foundation, PubMed Health