Rare Stevens-Johnson syndrome can be unpredictable, deadly

April 7 2010, By Avi Selk

For Cody Strickland, it began with poison ivy. In his 21 years, the outdoorsy electrician was no stranger to the rash, which caught him for the last time in early February, at a bonfire party on the shores of Lake Pat Cleburne.

It was no big deal -- he took care of it the next day with a few doses of medicine at the local hospital.

A couple of days later came the blisters, the spreading redness. Then the rush by ambulance to Parkland Memorial Hospital. Then the coma.

Strickland spent his last three weeks in the hospital's burn unit, sheets of skin sliding off his body.

By March 3, his father was staring at a death certificate and the name of a disease he had not known existed a month earlier.

"I just can't explain it," says James Strickland. "I don't know how I'm dealing with it. He got Stevens-Johnson syndrome. That's what it says on the death certificate."

Then he explains that poison ivy didn't kill his son -- the medicine did.

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UNPREDICTABLE

Stevens-Johnson syndrome is the bomb in the pill bottle: an unpredictable drug reaction that makes your skin and internal-organ lining fall off. It can be triggered by almost any drug, preceded by the slightest malady -- a cold, a fingernail infection, a headache.

Estimated to affect just a few people per million a year, the disease is so rare that a spokeswoman for the Centers for Disease Control and Prevention had to look it up on the Internet when queried. The Stevens-Johnson Syndrome Foundation in Colorado says it hears of at least three new cases in Texas each month.

But if the disease hardly ever strikes, it can strike at any time. And while doctors think that certain people may be genetically predisposed, they cannot predict who.

"There's no way to avoid it. No way to know who can be at risk," says Bernard Cohen, head of pediatric dermatology at Johns Hopkins Hospital in Maryland. (The disease is most common in children.)

"Someone could take Aleve for six years and the next time, for some reason, their body is sensitized."

Most people survive the disease. Strickland's case was especially severe -- his father says he lost the skin across 95 percent of his body. The doctors who treated him at Parkland would not comment.

Of the survivors, Cohen says, many make quick recoveries.

Cecilia Garcia is not among them. For her, it began with a cold.

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'I WANT TO DIE'

Two years ago, the Frisco, Texas, mother and McDonald's worker treated her cold with her husband's prescription Bactrim -- a sulfa-based medication that has a relatively strong association with Stevens-Johnson syndrome.

Garcia had completely forgotten taking the medicine two weeks later, when a doctor told her the red spots spreading across her body were caused by a drug allergy.

The doctor prescribed her more Bactrim.

Julian Garcia still isn't sure if Bactrim was the trigger. All he knows is he came home from his mail route two days later and couldn't recognize his wife's face.

"She just blew up," he says.

Garcia was rushed to the hospital, where the last thing she remembers before slipping into an induced coma is screaming, "Quiero morime" -- "I want to die" -- as she watched two doctors and a nurse scrub away her bubbling skin with wet towels.

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LIMITED TREATMENT

Your skin is made up of three layers. From top to bottom: the epidermis, the dermis and fat. When you get a rash or a sunburn, the damage occurs within the epidermis, which itself is composed of many layers.

When you get Stevens-Johnson syndrome, your epidermis blisters off completely, as do mucous membranes that cover your eyes, the inside of your mouth, your body cavities -- wherever your organs are exposed to air.

Without the protection of the top layers, "bad stuff gets in and good stuff gets out," explains Cohen. "Without those areas of the body functioning, you're basically dead meat."

In most cases, all doctors can do is take you off whatever medication you were on, put you in a burn ward and try to keep you alive until your skin regenerates.

That's what they did for Garcia, who awoke from a three-week coma staring at her raw, pink skin through half-destroyed eyes.

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PAINFUL COMPLICATIONS

Two years later, the scars that cover Garcia's body are the least of her complications.

"I was like a baby when it first happened," she says in Spanish. "I couldn't eat, couldn't walk, couldn't do anything."

Her eyelids are so scarred that the lashes grow inward and scratch her corneas. One eye barely functions. Her mangled tear ducts no longer produce tears. She is half-blind -- and what vision she has she pays for in pain.

When Garcia leaves the house -- which she rarely does except for the seemingly endless eye surgeries and doctor appointments -- she wears tinted goggles or oversize sunglasses and wraps her head in a scarf because daylight is excruciating to her.

So while her husband scours the Internet for news of experimental eye treatments, Garcia sits most days in a dark living room, listening to a television that is too bright to look at, often crying with scarce tears.

When the pain becomes unbearable, Garcia says, she sometimes risks a Tylenol.

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AT A GLANCE: STEVENS-JOHNSON SYNDROME

Who discovered it? Pediatricians A.M. Stevens and F.C. Johnson

What causes it? Almost any medication, including over-the-counter drugs

Who can get it? People of all ages, but a large number of victims are children

What are the symptoms?

• Rash, blisters or red splotches on the skin

• Persistent fever

• Blisters in mouth, eyes, ears, nose, genital area

• Swelling of eyelids, red eyes

• Conjunctivitis

• Flu-like symptoms

SOURCE: Stevens-Johnson Syndrome Foundation