BY H. LEE KAGAN

A Twitch of the Heart

When a cardiac nurse collapses during a stress test, her physicians must track down a rare heart condition before it kills her.

I WAS CHECKING ON MY PATIENTS in the cardiac monitoring unit at the hospital where I am on staff, when Denise, a 31-year-old nurse on the unit, stopped me to ask about chest pains she was having.

"I think I need to come see you," she said. Denise had been my patient for several years. "I've been having these pains off and on. It's been more than a month, and they're not going away."

Denise was clenching her fist over her mid-chest—a signal that, despite her relatively young age, she might be experiencing cardiac pain. Patients describing angina, the major symptom of a heart starved for oxygen because of narrowed coronary arteries, often clench their fist against their chest to illustrate what they're feeling. Typical angina is a pressure-like pain felt in the middle of the chest that is brought on by physical exertion. It fades away with rest. The ache may radiate into the neck or jaw or down an arm.

But Denise's chest pain was not typical of angina. Her pains were occurring at random times, unprovoked by anything she could identify. And the discomfort went away spontaneously after several minutes, whether she stopped what she was doing or not. Exercise didn't bother her at all, she said. As I eyed her overweight frame, however—she was an even five feet tall and weighed 150 pounds—it struck me that serious exercise was something she probably thought about more than she actually did.

Most people with atherosclerotic coronary artery disease (CAD) are over 50 years old. But it does occur, rarely, in people as young as 20. Diabetics, patients with kidney failure, and people with inherited metabolic disorders are the likeliest to have premature CAD. But Denise had none of these risk factors. Nor did she have hypertension or a family history of heart disease, two other important risk factors for coronary blockages. In fact, her only risk factor was a half-a-pack-per-day cigarette habit.

The more risk factors a patient has, the greater the odds their chest pain is due to CAD. In Denise's case, I wasn't too concerned. More likely, I guessed, stomach acid refluxing into her esophagus was causing the pain. Esophageal spasm can feel just like angina, and Denise's weight would make her susceptible to acid reflux, which is unrelated to physical exertion. Still, I decided it would be prudent to give her a stress test to definitively rule out CAD.

A Routine Test Turns Scary

A few days later, Denise was walking on the treadmill in my office. She wore an oversize T-shirt, baggy shorts, and sneakers. She said she felt fine. Earlier I had confirmed that her resting vital signs and cardiac exam were normal. Her baseline electrocardiogram, a measure of the heart's electrical activity, was also unremarkable.

During a cardiac stress test, the patient walks through a series of three-minute stages of increasing speed and incline, with each succeeding level more physically demanding. As the heart rate increases, blood pressure, cardiac rhythm, and the electrocardiogram are carefully monitored and the patient is asked to report how she is feeling, particularly if she experiences any chest pain.

Denise sailed through the first level, walking at a rate of about two miles per hour on a 10 percent incline. But when we bumped up to stage 2—still a fairly low degree of exertion for most people at three miles per hour at a 14 percent incline—the easy stroll turned into serious work for my overweight patient. Her smile faded. And then things went south very quickly.

I watched Denise become wobbly-legged on the treadmill, looking as if she might stagger off at any second. Her eyes seemed unfocused. The monitor showed that her heart rate, instead of increasing, had plummeted from its baseline of 76 into the low 40s, a drastic drop that signified she was in distress. I hit the emergency stop button, hopped onto the treadmill, and helped her take the three steps needed to get over and onto the exam table.

By the time I had her lying down, Denise wasn't answering me. When I glanced at the monitor I saw too much flat line and not enough squiggle. She was in a severe bradycardia—slow heart rate, in the low 20s. Her heart was beating much too slowly to generate enough blood pressure to supply oxygen to her brain. She was unconscious and nearly in cardiac arrest.

"Call 911!" I shouted. My nurse, who had been monitoring the test with me, quickly relayed my command to the front desk. She then brought the red crash cart, stocked with medications including drugs to speed up the heart, close to the exam table. Mentally I ran through the cardiac resuscitation training that I had hoped never to have to use.

Denise was now unresponsive, with no palpable pulse. Her shorts were darkened where she had urinated. Two involuntary jerks shook her entire

H. Lee Kagan is an internist in Los Angeles. The cases described in Vital Signs are real, but names and certain details have been changed.

body, the result of her brain receiving next to no blood flow. Moving quickly, I interlaced my fingers and put my lower palm onto her breastbone. I braced my arms to begin CPR as my nurse tilted back Denise's head and placed a plastic airway in her mouth to keep the tongue off the windpipe. I stole one more look at the cardiac monitor to check the rhythm. And then I froze.

A Surprising Recovery

Denise's heart rate was speeding up: 30s, 40s, 50s. Regular heart rhythm was climbing its way back up to where it belonged. Whatever had clamped down her heart rate was releasing its grip. I lifted my hands from her chest, reached to her neck, and found a pulse in her carotid artery. She opened her eyes and I watched as awareness slowly returned to her face. "Did you feel the chest pain again?" I asked. She nodded and I told her not to try to sit up, to just relax. My nurse placed oxygen tubing under her nose, and I was nearly done starting an intravenous line when the paramedics walked in.

At the hospital, my cardiology colleague, Dr. Andrew Johnston, took Denise to a lab to perform a test called a coronary angiogram. He threaded a thin plastic tube into her major coronary arteries and squirted dye into each one to make the insides of the vessels visible via X-ray. If he found a blocked artery, he might be able to relieve the obstruction and place a stent to keep the artery open. But the angiogram delivered another surprise.

"The arteries are all normal," Andrew told me over the phone when he was done. "And the heart muscle walls are all moving well, undamaged. There was no heart attack."

Normal coronaries. No injury to the heart muscle. I felt relieved. But what had happened on the treadmill?

Andrew went on to tell me that when he dripped a small amount of acetylcholine (a potent neurotransmitter) into the arteries, it induced severe spasm in one of the vessels.

That's when it hit me. "Prinzmetal's," I said. "She's got variant angina." It was the first case I'd seen since finishing my residency 15 years earlier.

In 1959 cardiologist Myron Prinzmetal was the first to identify a variant form of angina, a chest pain caused by a sudden and severe spasm of a coronary artery that obstructs blood flow. The spasm is reversible, can occur at any time, and is often unprovoked. In the safety of a lab, injecting certain substances such as acetylcholine can induce the spasm to confirm the diagnosis.

Studies have shown variant angina to be rare, occurring in approximately 4 out of 100,000 people in the United States. It is believed to be slightly more common in women and generally affects patients younger than those with CAD. We don't know why some people's arteries spasm, but it is probably related to a malfunction of the cells that line the insides of the vessel walls and the nerves that stimulate the smooth muscle surrounding the arteries. Left untreated, Prinzmetal's can increase the risk of cardiac arrest.

Life After the Diagnosis

When I saw Denise at the hospital that evening, she was feeling well. "Congratulations," I told her. "Today is the day you quit smoking." She didn't return my smile. I told her that smoking is one of the few recognized provokers of coronary spasm.

Her chart showed she had been started on a calcium channel blocker, an oral medication that prevents the spasm. As long as she stayed on that pill, her prognosis for a full, normal life was excellent.

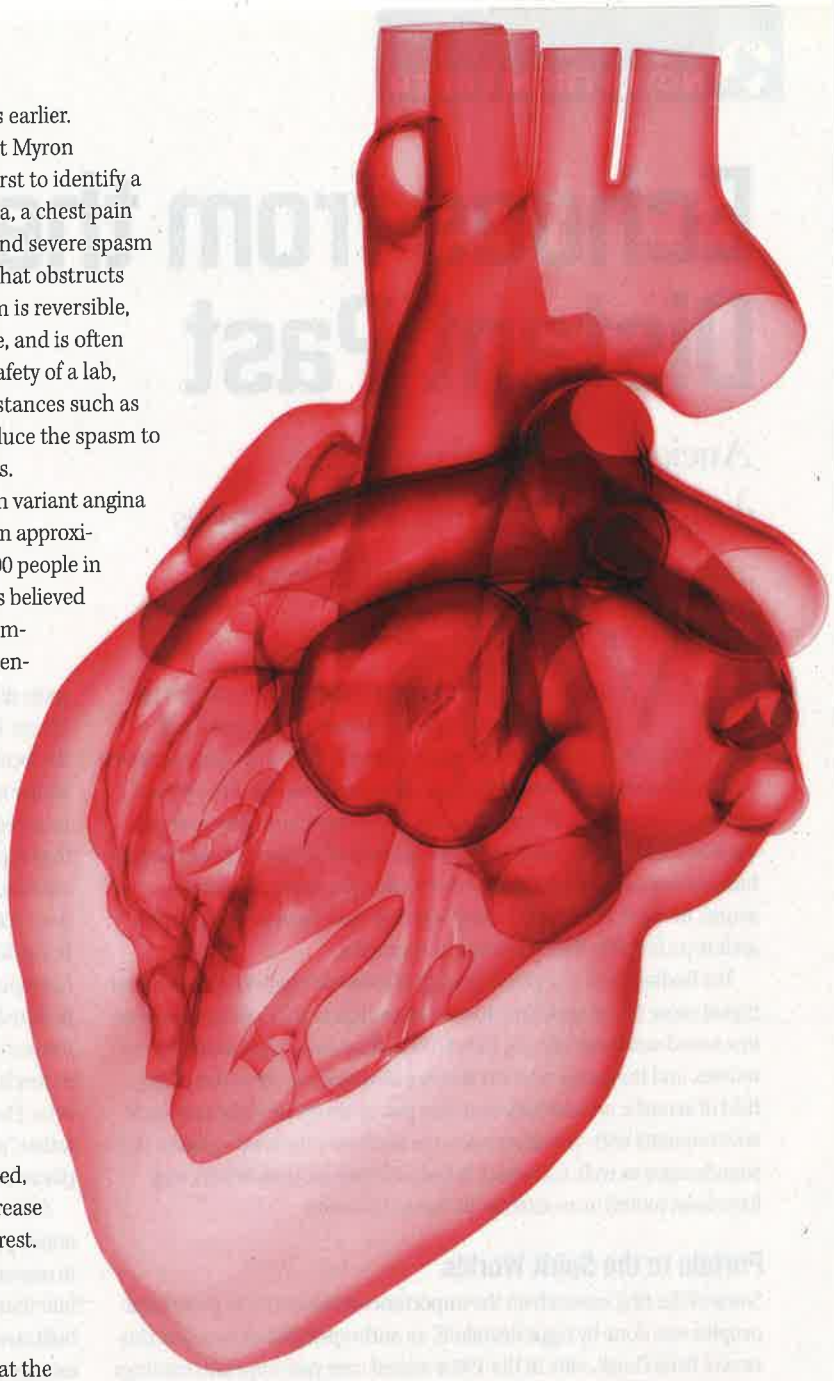
At a follow-up visit in my office I explained to Denise that hyperventilation can also trigger spasm in some people with Prinzmetal's. "When I had you walk on the treadmill, it didn't take long for you to be breathing hard," I told her. "That induced the coronary spasm and slow heart rate. Once you became unconscious, your breathing slowed, the spasm relaxed, flow was restored, and we had you back."

She was happy to report that she had quit smoking. But she also told me that she had tried skipping her pill one day.

"And ... ?" I asked.

"I felt the pain again."

I wagged a finger at her, but didn't say a word. I would save the weight-loss discussion for another day.



A human heart pumps 2,000 gallons of blood per day—50 million gallons over a lifetime.

MEDICALRF.COM/SCIENCE PHOTO LIBRARY