VITAL SIGNS

BY ANNA REISMAN

A woman with mild congestive heart failure takes a drastic turn for the worse. Why is her heart suddenly flagging?

WHEN 43-YEAR-OLD BARBARA HARRIS FOUND HERSELF PANTING AS she climbed the steps to her front door, she knew something was wrong. She was overweight and had high blood pressure, but she'd never been sick like this. In the hospital, she was shocked when her doctors told her she was suffering from mild congestive heart failure. Because her high blood pressure had gone untreated for years, her heart muscle had been damaged and was now unable to pump enough blood for her body's needs. Blood returning to the heart was backing up, resulting in fluid buildup in her lungs and making her short of breath.

She also had a mild heart murmur, probably a remnant of a childhood bout with rheumatic fever. An echocardiogram (an ultrasound of the heart) confirmed that her mitral valve was slightly leaky, allowing blood back into the atrium and forcing her heart to work harder to pump it out. Harris was treated for two days and went home with medication to lower her blood pressure and decrease the buildup of fluid.

Two weeks later, on a cold February evening, she was back in the hospital.

"It was OK at first when I went home," she told us from her hospital bed. I was working that month on the inpatient wards as an attending physician, along with a resident and a medical student. Harris adjusted the prongs of the oxygen tubing in her nostrils. "But it started again pretty quickly. I didn't even want to walk anymore; it got too hard. I'm out of breath. And my ankles are killing me." I pressed one of her ankles, leaving a little dent in the warm flesh, and asked her to rotate her feet. Wincing, she moved them a tiny bit. Ankle swelling could be a sign of heart failure, but mild cases do not usually cause pain. It was probably a red herring.

Once a patient has congestive heart failure, treatment is a delicate balancing act that includes nuanced adjustment of medication, a low-salt diet, and frequent weight checks. In some people this is easy; in others, missed medications or too much salt can cause the symptoms of uncontrolled congestive heart failure to return. But Harris had been avoiding salty food, and she hadn't missed any pills. She was so compliant, in fact, that her blood pressure and leaky heart valve probably could not explain the relapse at hand.

It was time to rethink. We sat in the doctors' station and talked about what might have triggered her symptoms. A heart attack was a possibility; we would get better information from a stress test. Alcohol could do it, although Harris told us she didn't drink. So could a condition affecting the heart valves (she did have the heart murmur, but it was mild) or bacterial endocarditis, a potentially life-threatening infection of a valve.

Over the next two days, we kept her busy. We cultured her blood for bacteria, which, if present, would increase the likelihood of bacterial endocarditis. We also sent her for various heart tests. But none of this told us anything new. Her blood was clear of infection and her heart was pumping well enough to rule out a heart attack. What had knocked her heart out of control?

ON HER THIRD DAY IN THE HOSPITAL, there was a seismic shift in Harris's condition. Her nurse beckoned to us from down the hall, where we were examining another patient. When we came into her room, she was sweating and massaging her knees. The nurse said her temperature was 102 degrees.

"Look at this," Harris panted, pulling her sheet aside so we could see her legs. "My ankles are fine now, but I'm hurting in my wrists—I can barely move my hands. And my knees, look how puffy!"

Dan, the resident, listened to her heart, his brow furrowed in concentration. "Her heart sounds different," he said. "It's weird. She has a bunch of new murmurs." Before, the lub-dub of her heart had been clear; now, all I could hear were loud whooshing sounds.

I was stunned. Dan and I were thinking the same thing: It *had* to be bacterial endocarditis. In endocarditis, small masses of bacteria form on the surface of a heart valve, and these little infected clumps—septic emboli—can slip off into the bloodstream. Depending on where they settle, they can cause major problems: abscesses, strokes, kidney disease, blood clots, and, by interfering with a heart valve's normal function, congestive heart failure.

I asked the student to draw some more blood to culture again for bacteria and to order another echocardiogram, which would show whether bacteria had built up on a valve.

We were flabbergasted when the echocardiogram results came in that afternoon. It looked like another patient's heart, and a really bad heart at that. The mitral valve no longer leaked a small amount; now a jet of blood regurgitated backward into the left atrium with each beat. And

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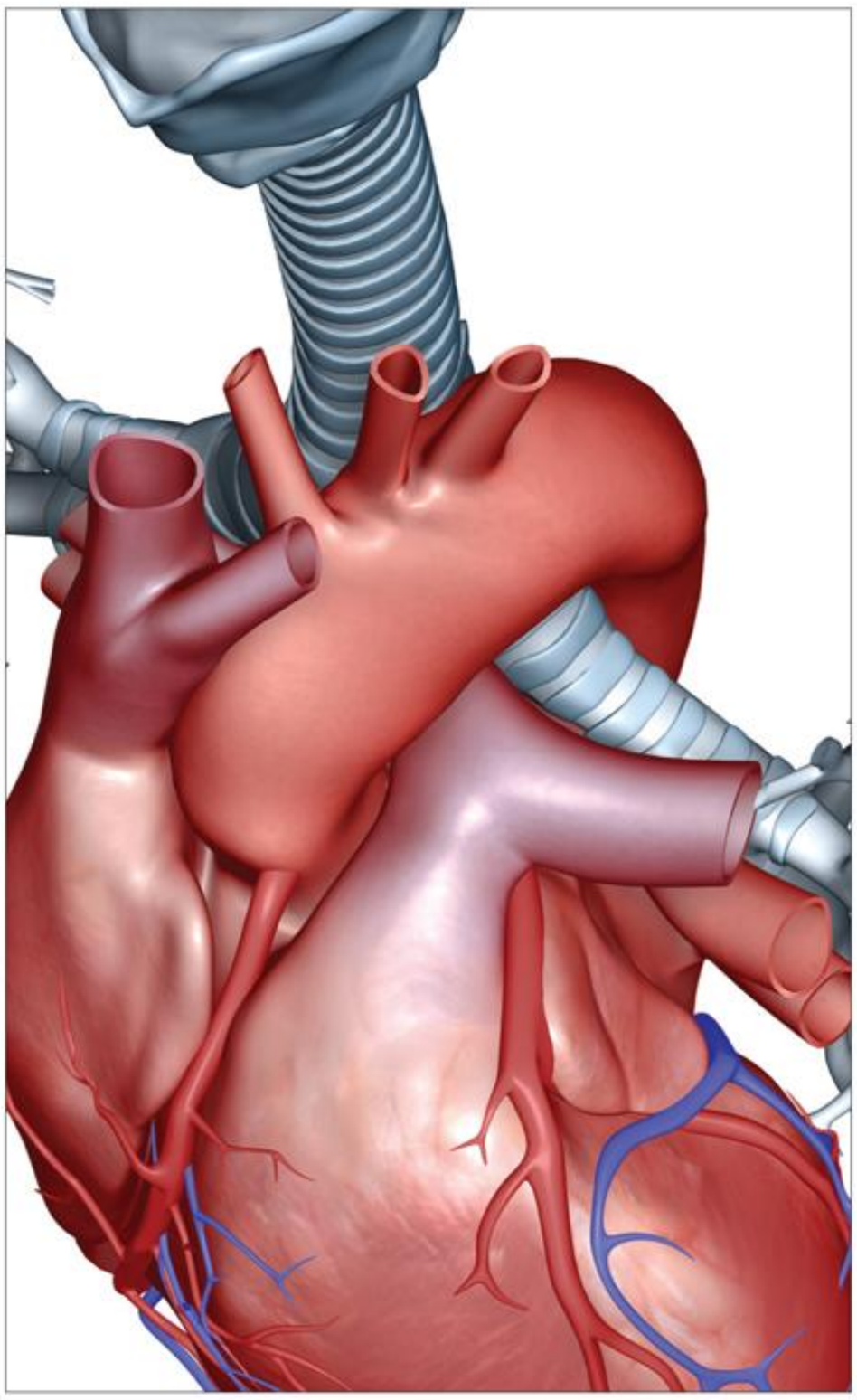
it wasn't only the mitral valve. The aortic valve also spewed blood in the wrong direction. The valves were damaged, but the echocardiogram showed no bacterial clumps. This didn't seem to be bacterial endocarditis. So why were Harris's heart valves worsening before our very eyes?

Dan wondered if we were missing a clue in her medical history. "New murmurs, the ankle pain that went to her wrists and knees, congestive heart failure.... Could she be having another bout of rheumatic fever?" he asked.

Because it had happened so many years earlier, we hadn't given much thought to her history of rheumatic fever, an inflammatory disease that can affect the heart, joints, and central nervous system.

"Interesting thought," I said to Dan. "But something would have triggered it. And she said she'd been fine except for the congestive heart failure two weeks ago."

"That's true," Dan said. "But what if she had something minor, like a sore throat, and didn't get treated for it?"



The aorta (pictured here wrapped around the trachea) carries blood out into the body. In some cases of heart failure, though, blood leaks back into the heart instead.

RHEUMATIC FEVER IS A COMPLICATION of something astonishingly basic: an untreated strep throat infection. When a particular type of strep bacterium—the group A beta-hemolytic streptococcus—infects the throat and isn't treated promptly, it can trigger an autoimmune response, causing the body to turn on itself.

One of the proteins on the strep bacterium's surface, the M protein, structurally resembles certain heart proteins. The body's natural response to a bacterial infection is to create antibodies to fight it; in the case of this particular streptococcus, the antibody to the M protein also works against the body's own heart proteins. The result: autoimmune destruction of some heart tissue and the heart valves. Damage to the valves can lead to permanent and often serious rheumatic heart disease.

The prevalence of rheumatic fever and rheumatic heart disease has plummeted in the past century in the United States and other industrialized countries, thanks to antibiotics and improved living conditions. In 1994, the last year the Centers for Disease Control and Prevention tracked the incidence of acute rheumatic fever, there were just 112 cases in the country.

Only a small number of sore throats in adults are caused by the strep bacterium that can, if untreated, trigger rheumatic fever. And most of us don't think about getting a culture every time we have a sore throat, since most are caused by viruses. But in people with a history of acute rheumatic fever, it's a different story. They are prone to significant risk from strep throat, especially if their original bout of rheumatic fever affected their heart.

WE HAD TO GO BACK TO HARRIS FOR that missing piece of the puzzle.

She stared out the window and thought back over the previous few months. At first she couldn't recall being sick, but then she remembered. Back in January, she said, she'd had a sore throat and fever.

"I thought I had the flu," she said. "It lasted a couple days, and then I started to feel better."

That was the clue we were looking for. We sent off one more test that might give us the answer, a blood test for an antibody signifying recent exposure to strep.

The next day the puzzle was solved. Harris had recurrent rheumatic fever.

The original damage her mitral valve sustained as a result of her childhood rheumatic fever wasn't too serious. But the immune response to her strep throat in January had been dramatic, with a new wave of antibodies eventually wreaking havoc on her heart valves. It's a condition I had only read about and have never seen since.

The response to treatment—at least for the joint pains—can be magical. Penicillin and high-dose aspirin erased Harris's discomfort in less than 24 hours. The damage to her valves, unfortunately, was permanent. Although we could control her symptoms with diuretics and other medications, and although antibiotics stopped the strep bacteria's destructive barrage, we could not repair the heart itself.

Because rheumatic fever is such an uncommon disease in the United States, nobody had educated Harris about the importance of promptly evaluating a sore throat. It simply wasn't something that most doctors—including me—thought about anymore.

Our patient was better for now. We were finally treating the infection that was weakening her heart valves. If she hadn't been treated, she probably would have suffered additional valve damage and ended up with more advanced congestive heart failure. She might have needed more medication and, perhaps, surgery to replace the damaged valves.

None of us would ever forget how dangerous an untreated strep throat infection could be, nor how a missing clue could so easily hide in plain sight.