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Broken Heart Syndrome

Investigating Takotsubo cardiomyopathy

BROBSON LUTZ M.D.



POLLY BECKER ILLUSTRATION

To see what current tort issues fuel the local legal community, I click on *The Louisiana Record's* website a couple of times a month. *The Record* isn't only a laundry list of all filed lawsuits, it also provides informative synopses of the more unusual, creative or otherwise notable court filings written in language that the non-legal-minds can understand.

Last October I saw a posting where a woman alleged that a Jefferson Parish hospital and some physicians missed an abnormal reading reported in a CT scan. Her husband had a scan prior to hernia surgery that showed a suspicious spot in his liver. Seven months after his hernia surgery, plagued with diarrhea and weight loss, doctors diagnosed an advanced, metastatic cancer that may have been related to that earlier abnormal liver scan. The patient died shortly thereafter, and the lawsuit alleged a delay in diagnosis and a loss of a better outcome.

"Failure to timely diagnose" malpractice lawsuits are common. While troublesome, lapses often make no difference in outcome, but sometimes delays do lead to missed opportunities for earlier effective therapies. But in this case, there was more. After her husband died, doctors diagnosed the wife with a heart problem "most commonly seen in post-menopausal women who have suffered the

sudden loss of a loved one such as a spouse of many years.”

The lawsuit demanded compensation both for her husband’s missed diagnosis and also for her heart condition caused by his death. Now, this was getting interesting. As is often the case, multiple physicians were named, including the emergency room physician, the hospitalist, the surgeon and the man’s primary care physician, who is a “concierge physician.” He has limited his practice to a small number of patients who pay a monthly fee supposedly for more personalized care, while also taking care of nursing home patients.

The lawsuit, which has been allotted to Judge Kern Reese’s court, identified the wife’s heart condition as Takotsubo cardiomyopathy. The name rang a bell, but I suspected the plaintiff had misspelled the condition. I remembered Takayasu’s disease, also called the pulseless disease. It was named after the Japanese physician who first described it, but it had nothing to do with stress.

So then I searched for “takotsubo” on the Internet as it was spelled in the lawsuit. The result was about 220,000 hits. The first hit was a description of the condition in Wikipedia, the second hit was a Mayo Clinic maintained webpage titled “the broken heart.”

My interest heightened, I emailed Dr. Edward St. Martin, the royal duke of common sense cardiology, for his take on all this. My usual emails to Dr. St. Martin go unacknowledged, or I receive a one- or two-word reply at most. I knew I had hit a hot button when I got back three whole paragraphs.

“I have seen about a dozen cases over the years and remember vividly three women who had this before the condition was first described,” he wrote. “The cardinal feature is the sudden ‘in the moment’ stress. ... The EKG and the striking appearance of the left ventricle by echocardiogram and angiography are striking and unmistakable. If I felt like making a PowerPoint, I could probably give a lecture on it tonight,” wrote St. Martin, who later answered some questions for me which I’ve edited below for space and clarity concerns.

Lutz: Takotsubo cardiomyopathy: real or an Internet hoax?

St. Martin: Real.

What are the other names for Takotsubo cardiomyopathy?

Takotsubo cardiomyopathy was the term originally used in the Japanese literature in the 1990s. The other two names are stress cardiomyopathy and broken heart syndrome. Cardiomyopathy refers to any heart muscle disease. This disease was first described in the ‘90s. The recent generation ... named it broken heart syndrome, but the scientific literature is definitive. All the cardiologists I know refer to it as “Takotsubo.”

How is Takotsubo pronounced?

Just say “tock – oh – sue – bow” with emphasis on “sue.” The “bow” is as in “bow and arrow” not as in “bow to the King.”

Who was this Takotsubo whose name goes with the condition?

A pot, not a person. The weakened left muscular chamber of the heart becomes stretched and distended. It takes on the nearly spherical shape of an octopus pot (Tako-tsubo in Japanese), a rounded globular piece of pottery that’s nearly spherical with a wide flared mouth resembling a fat pottery vase.

Why did it take doctors so long to discover Takotsubo cardiomyopathy as a unique heart problem?

Cardiac testing advances. Immediate coronary angiography in people presenting with heart attack symptoms and two-dimensional echocardiography were the diagnostic advances that led to the recognition of Takotsubo cardiomyopathy. Early two-dimensional echocardiography looking at the left ventricle demonstrated the octopus trap appearing left ventricle in persons with symptoms of an acute heart attack who had unexpected normal appearing coronary arteries by angiography.

Who is at risk for Takotsubo cardiomyopathy?

Most persons diagnosed with Takotsubo cardiomyopathy are middle-aged or postmenopausal women. Virtually every patient has had a triggering event causing a sudden release of adrenalin. In my experience Takotsubo occurs relatively frequently in women who have heart attack symptoms.

So what triggers Takotsubo cardiomyopathy?

The people we see with the syndrome have a sudden surprising emotional shock. Suddenness, surprise, anger, fear and devastating loss are all frequent precipitating events. The syndrome has been associated with death of a family member, extremely emotional arguments, public speaking, devastating financial loss and surprise birthday parties, among many others. Most often the stress has to do with a family member. Tragic loss or illness, devastating news, marked anger and conflict are frequently present. The triggering event isn't always sudden or unpleasant. There was a case reported at a College of Cardiology meeting where an elderly woman had inherited a huge sum of money and was signing a check for \$1 million to donate to her alma mater at a public event.

So how do these triggers and stresses cause heart problems?

The triggering event causes a sudden release of adrenalin, the "fight or flight" hormone. The sudden outpouring of adrenalin causes arterial spasm including the coronary arteries supplying the heart. Blood pressure and heart rate increase. The body is calling for more work, yet the blood supply to the heart, which has had no time to warm up, is decreased.

In biology, stress is defined as any change in the environment that evokes a response in the organism. Human beings are undoubtedly the king of the hill insofar as psychic stress.

What happens with a classic heart attack?

A classic heart attack is due to a blood clot triggered by arteriosclerosis in the coronary arteries resulting in a total blocking of a coronary artery with resultant deprivation of blood to a region of heart muscle. If the artery isn't opened quickly, the heart muscle deprived of blood dies and is replaced by scar tissue leaving the heart weaker. Various abnormal rhythms may occur in a classic heart attack and some can be lethal.

How does having Takotsubo cardiomyopathy compare to having a heart attack?

These patients can be very sick on admission and sometimes the damage to the left ventricle, the main pumping chamber of the heart, can be sufficient to put the patient into congestive heart failure. However, the outlook for total recovery is very good. The survival rate is much better than with heart attacks, and the likelihood of an ongoing or recurrent problem is close to zero. The affected heart muscle is likely more "stunned" than permanently damaged. Most patients undergo a complete or nearly complete resolution of the problem and return to a normal lifestyle.

Tell me about the cases of Takotsubo cardiomyopathy you have diagnosed.

When the initial descriptions of Takotsubo cardiomyopathy in the English literature were published, I immediately recalled three women who clearly had had this syndrome. The first was a reporter who was robbed at gunpoint in a dark parking lot after she was leaving her place of employment late at night. The next two were women who lived alone, worked long hours, got home at night, opened the door and were each met with a crowd in the house exclaiming "happy birthday." Each of the three underwent immediate coronary angiography revealing normal arteries. I also treated an elderly lady who experienced Takotsubo delivering the eulogy at her sister's funeral and a number of others – about eight to 10 altogether. All of those whom I have followed long term have fully recovered.

Almost all folks claim undue stress. Is it possible to be walking around with stress cardiomyopathy and not even know it?

The stress that would be claimed by most folks is chronic day-in-and-day-out stress related to life in general. The stress that precipitates Takotsubo cardiomyopathy is an acute stress different from that of daily life experience. In view of the usual presentation with acute severe chest pain and other features, such as shortness of breath and the fact that the disease often gets much better within a few days or weeks, it's doubtful that an individual would be walking around with this condition and not even know it.

Once a person has had stress cardiomyopathy will they get it again the next time they are under severe stress?

Given what is known about the types of stress that produce Takotsubo cardiomyopathy, it's very unlikely that it will happen twice in the same individual. I am unaware of any such cases.