

Hudson boy's cause of death: Broken heart

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HUDSON, N.H. -- Zachary **Tompkins** ' sudden and mysterious death at home in his sleep March 8 left his parents, two brothers, relatives and friends brokenhearted.

After months of uncertainty, Necole **Tompkins** said an "intuitive" medical examiner discovered through a genetic test that Zach had Long QT Syndrome, or LQTS, an inherited electrophysiological disorder that can stop one's heart without warning.

In other words, Zach's heart was broken, too.

There was never a single symptom to let even the most attentive mother know that Zach, an unfailingly healthy and happy 11-year-old honors student and athlete, had a potentially fatal heart condition -- a "silent killer" -- hidden in his chest.

"Zach was never even sick," Necole said.

She was devastated to learn that she had passed the LQTS gene to two of her sons.

"Knowing that I carry this gene, and that I gave it to both Zachary and Nicholas, I'm wrestling with my emotions, ranging from guilt to loneliness to sadness," Necole said. "As a mother, I'm supposed to protect them, not give them something that's going to hurt them."

Genetic test results showed Zach's younger brother, Nicholas, 8, and Necole both have LQTS. The genetic tests, conducted at one of only two labs in the country equipped to test DNA samples for LQTS, showed Zach's dad, Mike, and oldest brother, Christopher, don't have the disorder.

"Not only do we continue to grieve for the loss of Zach, we now grieve with the new knowledge that Nicholas and I have LQTS and that our lives have changed forever," Necole said. "However, knowledge is power, and doctors have prescribed medications for Nicholas and me, and we're confident those medications will work."

Now that the mystery of how Zach died is solved, Necole has vowed to solve another mystery involving LQTS on a national scale: Why is there no standardized genetic testing for LQTS among pregnant women in the United States, especially considering the estimates from medical experts who say 1 in 4,000 Americans possesses the defective LQTS gene?

"I'm just so baffled why none of us know more about this," she said. "There's got to be some kind of hang-up."

The relative newness of the genetic test for LQTS may be largely to blame for the lack of widespread understanding or testing for the syndrome, suggested Theresa McNeely, vice president and spokesperson for Newton-based Clinical Data Inc., which owns PGX Health, the first laboratory to begin offering LQTS gene-testing beginning in 2004. There is also the

substantial cost of \$900 per test, McNeely said.

Most insurance companies fully reimburse clients for LQTS genetic testing if they post an erratic electrocardiogram reading, faint inexplicably, or exhibit other warning signs of an abnormal heart condition, McNeely said.

But the same insurance companies, including Cigna, have a similar LQTS clause in their standard-coverage policies that reads: "CIGNA does not cover genetic screening for LQTS in the general population, because such screening is considered not medically necessary or of unproven benefit."

The insurance companies may be forced to rethink their policies for LQTS among the general population as the numbers of needlessly grieving and outraged parents climb, Necole said. Thousands are dying every year from LQTS needlessly, she believes.

"I'm ready to go to the White House right now," said Necole, who is also a nurse. "LQTS is a dominant gene. You have a 50 percent chance of passing it to your child. Cystic fibrosis, which (OB/GYNs) test for now, is a recessive gene, which both your parents have to have in order for your child to get it. So why aren't we more educated about LQTS? What's the hang-up?"

Wholeheartedly, McNeely agrees. Quoting from Clinical Data Inc.'s most recent in-house findings, McNeely said, "Greater awareness of, and improved testing for LQTS is revealing the disease's true prevalence: It is now estimated that 1 in 2,500 people has LQTS."

"As people get more educated, hopefully ahead of tragic events such as what happened to the **Tompkins** family, we anticipate there will become more routine testing for the Long QT gene and other inherited genetic conditions," McNeely said.

Despite struggling this month with sending off to school two children instead of three, Necole renewed her pledge to continue open communication with the Hudson community about Zachary's passing, and to "never give up" (to quote the title of Zach's favorite book by former New England Patriot Tedy Bruschi) in making life-saving noise about the silent killer that took her son's life.

"I've got a big mouth on a small girl and I'm going to use it," Necole said. "People have asked me, are you sure you want to keep talking about this, how Zach passed? But if I keep quiet and don't talk about it, then I'm doing a disservice to Zachary and Nicholas. On top of that, Nicholas will think it's something horrible or shameful, instead of something manageable. He didn't do anything wrong."

Necole said her husband's "unconditional love and support" and their "everlasting love for our three boys" has kept the family going. She also credits the outpouring of love and support from the Hudson community for keeping her from inactivity or depression.

Several fundraisers that have been held to date to build a Hudson sports stadium in Zach's name -- including a golf tournament and the "Zach Attack 5K" -- have been tremendous successes, generating more than \$100,000.

"We all got a special gift from Zachary," Necole said. "Our community -- our Hudson family -- is standing by us in our mission to build a stadium in Zachary's honor. And we will continue until we complete our goal."

The next Zach's Stadium fundraiser, "Rock the Stadium," is Friday, from 7:30 p.m. to 2 a.m., at The Brewery in Lowell, with live music featuring the on-stage debut of Christopher **Tompkins** on guitar, playing a tribute to his late brother. For tickets, contact Danyelle at 978-479-3003, or danyelle@wildflowerphotography.org.