

A Risk for Sudden Death in Epilepsy That Often Goes Unmentioned

By Gina Kolata

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Shena Pearson nearly froze in her seat, terrified, as she stared at a power-point slide. She was at her first meeting of an epilepsy foundation, seeking help for her 12-year-old son Trysten, when a neurologist flashed the slide about something called Sudep.

It stands for sudden unexpected death in epilepsy. Her son's neurologist had never mentioned it.

"Oh dear God, my child is at risk, seriously at risk," Ms. Pearson thought to herself.

Sudden death in epilepsy is a little-known and seldom-mentioned phenomenon, but now, after a push by advocates, the federal government has begun a concerted program to understand it. Yet a question remains: When, if ever, should patients be warned?

In a way, the extreme reticence of many neurologists to mention sudden unexpected death to epilepsy patients harks back to the days when doctors and families often did not tell people they had cancer — too terrifying. But today, patients learn not just about cancer but about many other potentially fatal conditions, like an inoperable brain aneurysm that could burst at any time and kill a person. So the quiet about the epilepsy death risk appears to be an anomaly.

Sudep's name pretty much explains what it is: Someone with epilepsy — unprovoked seizures, which are electrical surges in the brain — dies, and there is no apparent cause. Often a person with epilepsy goes to bed and is found in the morning, unresponsive. In some cases, there is indirect evidence of a seizure, like urine on the sheets, bloodshot eyes or a severely bitten tongue, leading to the suggestion that preventing seizures as much as possible with medications could lower patients' risks. But so much about the syndrome remains unknown.

Neurologists say sudden unexpected death in epilepsy is second to stroke as a cause of years of life lost because of a neurological disorder. Sudep kills an estimated 2,600 people a year in the United States — some neurologists say the real figure is almost certainly higher — or one in 1,000 people with epilepsy. For people whose seizures are not controlled with the medication, the fatality rate is one in 150.

Some three million Americans and 50 million people worldwide have epilepsy. About a third of Americans with epilepsy have uncontrolled seizures, said Dr. Daniel Friedman, an epilepsy researcher at New York University. That means about a million Americans could be at high risk of sudden death.

Ms. Pearson's son was having at least 24 seizures a year despite anti-seizure medication. She could not bear to tell him about the sudden death risk. But he found out anyway three months later. He was meeting with an epilepsy support group meeting near their home in Galveston County in Texas and overheard people discussing it.

Ms. Pearson and her son were not alone in finding out about sudden death in epilepsy by accident. Despite the urging of professional organizations like the American Epilepsy Society and leading researchers to give patients the full picture, neurologists shy from a discussion of this phenomenon. The problem is that, at least for now, risk estimates are uncertain, and there are no proven ways to prevent it.

A national study of neurologists found that very few always told people with epilepsy about sudden death. That prompted Dr. William Gaillard, the director of the epilepsy program at the Children's National Health System, to survey the children's neurologists in his program. Most said they usually did not mention it to families.

"Many of my colleagues, myself included, are paternalistic creatures," Dr. Gaillard said in an interview. "Many don't talk about it because it is a low risk, and there is nothing you can do about it. They've made that decision for their patients."

But Dr. Gaillard and others say families have a right to know. In addition, said Dr. Orrin Devinsky, the director of the epilepsy center at NYU Langone Medical Center, knowing about it could provide an impetus for patients to work with doctors to get their seizures under control, as death occurs just after a seizure.

Dr. Walter Koroshetz, the director of the National Institute of Neurological Disorders and Stroke, faced the disclosure question in his own family. In 1990, his father, who developed epilepsy late in life, walked to his refrigerator one day to get something to eat. He had a seizure, fell and died. Five years earlier, an uncle on his father's side had died as a result of Sudep after developing late-life epilepsy. But Dr. Koroshetz, who as a neurologist knew about sudden death in epilepsy, had not mentioned it to his father.

“I did not think it would help him,” Dr. Koroshetz said.

Three years later, another of his father’s brothers developed epilepsy. This time, Dr. Koroshetz said, he had a long discussion about the risk of death with that uncle. He is still alive and doing well.

Dr. Koroshetz says there is not much adults can do to protect themselves except to take anti-seizure medications religiously, because the sudden death risk is associated mainly with uncontrolled seizures.

Yet sudden death is not a newly recognized epilepsy complication; one of the first descriptions was written by George Washington, whose stepdaughter Patsy Custis may have died from it on June 19, 1773, at age 17.

“She rose from Dinner about four o’clock in better health and spirits than she appeared to have been in for some time,” Washington wrote, “soon after which she was seized with one of her usual Fits, and expired in it, in less than two minutes, without uttering a word, a groan, or scarce a sigh. This sudden and unexpected blow, I scarce need add has almost reduced my poor Wife to the lowest ebb of Misery.”

But many neurologists say they did not learn about Sudep in medical school.

“Even among neurologists, it wasn’t well known or publicized or talked about until very recently,” said Dr. Samden Lhatoo, a professor of neurology at Case Western Reserve University School of Medicine.

Dr. Devinsky, 59, said, “People I trained with at great medical institutions never discussed it.”

It was not until an advocacy group, Citizens United for Research in Epilepsy, or CURE, contacted Dr. Koroshetz and asked what is being done about sudden death in epilepsy, that the federal government took action. Now, the National Institute of Neurological Disorders and Stroke has started a major research program. Its investigators say that, so far, it appears that the electrical storm in the brain that occurs during a tonic-clonic seizure — in which a person loses consciousness and convulses — can make the heart stop or brain activity cease in some people.

One research goal is to learn the true incidence of the phenomenon and to study brains from patients who died from it. It has not been easy. Often, especially in older people, the medical examiner will attribute death to a heart problem, even when it is known that the person had epilepsy and there was no evidence the victim had heart disease, Dr. Devinsky said.

John Popovich lost his son John Paul to sudden death in epilepsy. A 19-year-old freshman at the University of Virginia, he had come home to celebrate the holidays last year. His father found him lying face down on his bed one morning, dead. He had had only three seizures in his entire life.

The death certificate did not mention sudden death in epilepsy, and Mr. Popovich and his family, who live in Northern Virginia, had never heard of Sudep until his brother told him about it several weeks after his son’s funeral. Mr. Popovich could not understand doctors’ reluctance to bring it up.

“As a parent who lost a child, I can say that the medical community is not doing anyone a favor,” he said.

The toxicology report after his son’s death indicated that he had not taken his seizure medication for at least a day — the drug was not in his system. If he had known about the risk of sudden death, he would never have missed a dose, his father said.