

My Story: Living with Narcolepsy

A 34-year-old ED nurse discusses how her life has changed after developing narcolepsy with severe cataplexy.

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Editor's Note

Tricia Higgins is her own clinical trial with a cohort of 1. Tricia has been diagnosed with narcolepsy with severe cataplexy. This dual diagnosis is characterized by the inability to maintain voluntary wakefulness, abnormal REM, and sudden paralysis or weakness in muscle tone without loss of consciousness. If left uncontrolled, her symptoms make it nearly impossible for her to live normally. Her ability to live a higher quality of life is made possible by a cocktail of medications that were all developed for purposes other than narcolepsy. Patients and physicians facing uncommon or atypical disorders like Tricia's must consider a complicated series of decisions. Most therapeutic regimens and pharmaceutical developments are based on treating the underlying mechanism of a disease, a pattern that is growing more pronounced following tremendous advances in basic science and translational research. But what happens when that paradigm fails? How do things change for both patient and physician when determining an accurate diagnosis—let alone treatment—can take years? What happens when it is not possible to address the mechanism of a disease, when the clinical focus must be shifted to treating symptoms? The ethical and professional challenges to the treating physician are very real, but so too are the thoughts and reactions of the patient. In her own words, this is Tricia Higgins' story.

When I was 34 years old my world started caving in. I was a mother of 3, wife to a wonderful man completing his third degree, homemaker, emergency room nurse, and good friend to anyone in need. I was a volunteer in my children's classrooms and a CCD teacher at our local Catholic Church. Yet suddenly I was the one in need, a role I hated from the beginning and still hate today.

It all began while I was giving instructions to a mom who had brought in her sick child to the Emergency Room. Suddenly, I found myself on the floor looking up at the ceiling lights. I hadn't felt weak, or sick, but I couldn't get up. Slowly the feeling passed and I was back to work trying to forget the event. A day or so later while at work I once again slumped to the ground. After my third fall I decided to make an appointment with my primary care doctor. I explained to him that I always know where I am when I fall but I just can't move. My only consistent symptom is exhaustion; other than a stiff neck or a bump on my head, I could usually go on with my routine. On my way out of his office, I fell again—exactly the same fall, no warning, no aura; I just remember coming to and looking up at the lights.

When I was admitted to the hospital for the first time, the medical work-up began. I had the usual tests of EKG, carotid Dopplers, echocardiogram, routine blood work, all of which were essentially negative. I did fall while on a cardiac monitor but no arrhythmia was present. Eventually, the cardiologist and neurologist agreed to send me home.

I remember falling a total of 17 times over the course of 2 months before I was convinced that I needed a more drastic treatment. I was started on Tegretol, an anticonvulsant, but my falls continued, so my dosage was gradually increased. I experienced lots of nausea and vomiting when I started Tegretol, but eventually I stopped falling. I was thrilled. I didn't like the diagnosis of complex partial seizures, but the prescribed medication stopped the falling. I agreed to stay on the Tegretol for a 1-year trial, but by the time the year was up, I was ready to quit because I hated the side effects. I

was slowly taken off the Tegretol, but then I started falling again. When I was placed back on the medication, the side effects returned, this time with an additional elevation of my liver enzymes. Finally I decided the treatment was worse than falling. It was at this point that the trial and error of using different combinations of medications began and this gave a whole new meaning to the words "medical practice."

After the Tegretol experience, it was suggested that I seek professional help from a psychiatrist. There was consistent underlying depression that seemed to resurface after each subsequent fall. I missed work occasionally, but I continued to work part-time. I didn't know it then, but my neurologist thought that the falling was somehow under my control. After a trial of Depakote and Neurontin, I had a severe outbreak of hives and had to be taken off both medications. The falls continued as did the side effects from other medications, now including tinnitus, extreme photosensitivity, and headaches. While at a routine appointment with my neurologist, I had a seizure in the CT department. I was re-admitted to the hospital and referred to a neurosurgeon and another psychiatrist. I was restarted on Tegretol, and the nausea and vomiting started all over again. I remember the neurologist coming into my room while I was on the bathroom floor heaving into the toilet too weak to get back into bed. He just stared at me and asked what was wrong. After talking with the psychiatrist, I was discharged and sent home.

Here was my reality: falling at home was no big deal. Mostly I would sleep until I felt more alert. Falling at work proved to be more of a problem. I no longer transported patients by myself. I could no longer hold a baby alone in my arms. I also made sure that I stood with my back directly up against a wall.

Sometimes I would go for weeks without falling, but then they would become more frequent. I was never able to predict their onset. Medications would be added, all of them causing more undesirable side effects. I was ordered to stop doing the late night shifts at the hospital, to see if a regular routine was what I needed to keep from falling. I continued this way for 5 years, seeing a neurologist intermittently while the falls continued.

I was eventually referred for further testing to Brigham and Women's Hospital in Boston. After I was monitored over a period of 7 days, the attending neurologist arrived with a psychiatrist and explained that the falls I was having were not electrical seizures. They said it was a psychiatric disorder—non-electrical seizures and somatization—that caused me to fall. They explained that all I had to do was follow up with a psychiatrist, discover what was triggering my emotions, and then I would no longer have problems falling. They told me that my condition was treatable as long as I was willing to believe that I could get better. When asked if I could come off the anti-convulsant medications they hesitated. I said if these events were non-electrical in nature then I want to stop all medications. The doctors discouraged this since they knew that the falls were clearly less frequent when I was on medication. After they left, I couldn't believe that I was doing this to myself. I cried and cried. Alone in my room with the lights off and no one to talk to, I wanted to end it all. Just thinking that I was doing this to myself was unforgivable. I wanted to die. I was the root of the problem.

I followed up with a psychiatrist and explained the physicians' findings in Boston. I told him that the reason I was falling was some underlying emotion, and, once I discovered what I was trying to suppress, I would be cured. The psychiatrist did not agree with this conclusion. I told him that he had to agree with the conclusion or I would never get better; I begged him to make me better. He explained that he still disagreed with the other physicians' conclusions and thought that no one had come up with a correct diagnosis yet.

I was slowly able to compose myself and go back to work, but secretly I felt like a fool. I felt like a fake and a liar, but I kept most of these feelings to myself. The falls continued, but as long as they were not at work, I figured my job was not in jeopardy—and then I fell carrying a portable monitor up a flight of stairs on my way to a code in the ICU. My co-workers said they needed to count on me, but they never knew when I was going to fall. I felt totally defeated. It had been my decision to keep working, and now I no longer had that option. I started teaching courses at the hospital, but then I began to fall in the classroom.

At this point my life was spiraling out of control. Each fall would send me into a deeper depression until I was eventually placed on antidepressants. I was also started on medication for unexplained paresthesias and peripheral neuropathies. I began having trouble with sleep paralysis and terrible nightmares. I felt tired all of the time.

By 1999 I had been falling for 7 years, and my primary care doctor asked if I was getting enough sleep at night. I

looked at him as though he had lost his mind; until recently I had worked shifts in the ER; I had 3 active boys, and my husband was in law school at night. I told him every mom I know is tired. Then he said he wanted to enroll me in an overnight study for sleep disorders. I was adamantly against it. Every test I had taken had been normal; I was convinced this one would be normal too and would be another waste of time and money. I said I could not handle being tested again only to be told the results were negative. After much coaxing he scheduled the test in a hospital where I knew no one. I cried going to the hospital and begged my husband not to leave me. The sleep lab was in a prominent psychiatric hospital, and I was afraid that my husband was going to abandon me because, by this time, I too was convinced that I was losing my mind.

Within 2 days of the test, my doctor reported that at last I had a diagnosis that explained my symptoms. He said that I had narcolepsy with cataplexy. Once I started reading about narcolepsy, I knew that this was it; this was what I had been dealing with. Excessive daytime sleepiness, vivid dreams, automatic behavior—I could have written the book. I was able to accept the diagnosis of narcolepsy and the constant modifications needed for my treatment. I now wear a mask to bed to help with sleep apnea, and I think the quality of my sleep has improved. The use of Ritalin has improved my alertness during the day. Taking Prozac and Nortriptyline for the parasthesias has also helped the cataplexy. It has taken many trials of various combinations of medications to help decrease my symptoms, but with my current regimen, including Xyrem, my cataplexy is better than it has ever been.

As I contemplate just how my life has been impacted as a result of narcolepsy, the first thing that comes to mind is how everyone thinks that it's comical. They laugh and then realize when they are laughing alone that I am serious about this disorder.

This illness has cost me my job, my friends, my self-confidence, my dignity. Work, the place we all go to make some money, pay the bills, and keep our children in school, that place where we feel like we make a difference in people's lives—for me that place was in the hospital as a nurse. I would gladly accept the challenging patients. I used to feel fulfilled even when I was fatigued at the end of a shift. My coworkers could always count on me for overtime or extra shifts. I would cover for someone who needed to be home with his or her kids due to illness. I was always there when something extra was needed, and I rarely took a break. Now I am no longer employed; I have been on disability for 4 years, forced out (or so it felt) because I had no control over my cataplexy and subsequent falls.

For me, cataplexy presents itself with total loss of muscle control. I will just fall to the ground, without premonition, without aura. I am always aware that I am on the ground, and I can hear what is going on. I am unable to move or speak for a period of seconds to minutes. It is the loss of control that bothers me so much. It is the unpredictability. And then it is the explanations to others when I am able to respond and speak. I am used to it, but it is hard to convince those around me who aren't not to panic.

Along with excessive daytime sleepiness, this is what plagues my days. But don't think that it is just my days that are affected. At night I often find myself screaming at a hypnogogic hallucination that I swear is really happening and invading my nighttime sleep. Many nights my husband has to wake me and assure me that there are no creatures coming to get me. Sometimes just before drifting off to sleep, or just before waking up, I will experience sleep paralysis. You want to move but you can't. You want to call out but you cannot speak the words. I used to think I was dying, and was afraid to fall asleep.

The excessive daytime sleepiness is difficult to describe. It is so painful to stay awake sometimes that my body actually hurts. It is struggling to do the housework, make the meals, stay ahead of the piles of laundry. It is missing appointments, being late to meetings, forgetting chores. It is being unable to drive and relying on others to give you and your children rides to events. It means lots of bike rides to the store when you forget something.

And it is the isolation. You only go out when you have to. Even walking brings its own set of risks. I am constantly worried that a fall will result and that the depression will resurface. It is easier to be reclusive because then no one knows what is going on with me. Hibernation and cataplexy in my own home can stay a secret. Just when I think it would be easier on everyone else if I were no longer here to burden my family, a ray of hope, a sliver of sunshine gives me inspiration to move forward.

Tricia Higgins is a native of New Jersey who currently makes her home in Morristown. She obtained her BSN from Catholic University in Washington, DC, and worked professionally as an emergency room nurse.

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