

## Mary Ingram-Schatz “...*The bad news is that you are not a crackpot.*”



We just completed *June is Myasthenia Gravis Month*. Who knew? Myasthenia is a fairly rare disease; about 20 people in 100,000 are diagnosed. By comparison, one out of every four deaths in the United States is attributed to heart disease. So, I didn't expect we had our own awareness month, but it seems like a good time to write about what the disease is and how it has affected my life.

As you know, myasthenia gravis is an autoimmune disease. When the autoimmune system is working properly, it attacks foreign cells like viruses and bacteria in the body. For people with myasthenia gravis things get weird. When your brain wants to use muscles, it sends messages through the nerves, but nerve cells don't connect directly to muscles. There is a little space called the neuromuscular junction. A chemical called acetylcholine guides the message across the gap. In people with myasthenia, the immune system starts attacking these junctions like soldiers blowing up a bridge. But the body repairs bridges pretty quickly, so you don't lose the muscles permanently, muscle strength and control comes and goes. Symptoms tend to get worse when the immune system is stirred up. So when you are sick, stressed, or tired, you lose muscle control more quickly.

As a patient, this process is completely baffling. I have a somewhat atypical presentation of the disease. For me, I lose muscle control in my hands early on. So, what would happen is that I would be tired, stressed, or sick and suddenly start dropping things. One minute I'm writing, the next minute I can't hold a pencil. I went bowling with my kids and dropped the ball four times. They thought this was hilarious. The odd thing was it wasn't always in the same hand, wasn't every day, and would generally get better with rest. Would you go to your doctor to tell her that? I thought I was losing my mind.

As the disease progressed the symptoms got worse and more frequent. I started getting double vision. My eyelids started to droop. I even lost facial control. Pictures of me during this time look strange because my smile is off. I remember one day, it was soup day at the school where I worked. Teachers brought in crockpots filled with homemade soup to share at lunch. There was this crazy music teacher who was well-known for his epic clam chowder. Of course, I had some; it was very rich and delicious. After school, I went to exercise. I was in a total exercise rut at the time. Every day I did 45 minutes on an elliptical trainer set to 9. That day, I arrived, set the machine to 9 and could not make it move. Using all my strength, I could NOT do it. I kept moving the settings down. Finally, at 3 I could push the pedals. But after 20 minutes I felt like I was going to die. I was out of breath, achy, my vision was doubling, eyelids drooping and I was completely miserable. This, when I had done 45 minutes at 9, three or four times a week, for ages. Being a stubborn sort, I tried again the next day and it was no problem. I blamed the soup. What the hell did he put in that stuff anyway?

One day, just before I was diagnosed, I went skiing with my family. Skiing is a terrible idea if your muscle control is flickering on and off, but what did I know. I damn near killed myself on

the bunny slope. I spent the afternoon sipping cocoa with droopy eyelids and muttering about my age. The problem was not my age. Meanwhile, at home, I found myself deciding what to wear based on my hand muscle control. Some days I felt totally normal. Some days fastening a necklace was impossible. I decided that even if I was losing my mind, I had to see a doctor. The drugs for mental illness are actually really effective. If I was having some sort of breakdown I should address it.

My primary care physician tested my thyroid. I have Graves' disease, which has been in remission for years, but it causes double vision, so he and I both thought it might be back. My thyroid was fine. He said, "I know if you are describing these symptoms, then they are real, so I am going to refer you to a neurologist." Everything in his tone suggested that he thought I was nuts.

My genius of a husband diagnosed me first. He is an oncologist, but from some dark, dusty part of his memory, he pulled out a lecture he had heard in medical school about myasthenia. He had a vague memory of it because he recalled that Aristotle Onassis died from myasthenia and the symptoms were really odd. He read up and thought that was perhaps what was going on.

When I had an appointment with a neurologist he asked a bunch of questions then said, "Well, the bad news is that you are not a crackpot." That's bad news? I had been putting my lack of crackpot-ness in the wrong category for a long time. He referred me to a different neurologist who specialized in neuromuscular conditions. A referral to a specialty neurologist is an "oh crap" moment. No one wants to require that level of medical expertise. She ran a bunch of vaguely unpleasant tests where they zap your muscles with little electrical pulses and see how quickly they weaken. It is super exciting for the neurologist, because the disease is rare and they get to show all their medical students how it works. My muscles failed just like the textbooks said they would, how exciting! She had her medical students re-interview me about my symptoms every time I came in so that they could all have a turn. It is the most useful thing I'll ever do to support modern medical training.

I was happy to help, because she gave me drugs that changed my life. First, she gave me pyridostigmine. It doesn't really manage the disease, but it makes the symptoms better. It just boosts up those junctions that are under attack. I LOVED it. Doctors always ask if you are feeling fatigue and frankly I never really know what to say. I mean, I'm a working mother. I was teaching kindergarten at the time and those little munchkins get all their energy by sucking it right out of the adults around them. Plus, my own kids are busy and active. Am I tired? Well, duh. But, you know how you have an overhead light in your kitchen with three or four bulbs in it? And one day, the last bulb goes out and you replace all four. Suddenly, the room is crazy bright and you realize that it had actually been kind of dim in there for a long time, but it happened gradually so you didn't really notice? Taking pyridostigmine was like putting in four new bulbs. I felt awesome. I had forgotten what normal felt like. But pyridostigmine is just a myasthenia band aid.

After that, I took prednisone for over a year. Prednisone is a double-edged sword if ever there was one. All my symptoms went away. It is like a miracle. However, my hair thinned, I gained a few pounds, I didn't sleep well, and I get a little short tempered (not a great quality in a kindergarten teacher). Over the long term, it has caused osteopenia as well. But I figured at least I was only short-tempered temporarily. Lots of people are like that all the time –

right? Once I was stable, I had a surgery called a thymectomy to remove the thalamus gland. At one time this was a pretty debated treatment among neurologists. Some said that it was very useful as treatment, others disagreed. Recently, a paper was published in The New England Journal of Medicine, Randomized Trial of Thymectomy in Myasthenia Gravis, giving strong support to thymectomy as a treatment. To learn more you can go to <http://www.myasthenia.org/Research/Latestnews.aspx>. The thalamus gland isn't shaped like a jellybean like I imagine glands to be. It is more like someone dropped a bowl of Jello and there are little gooey pieces scattered inside your chest. Essentially, they jam a tiny robot vacuum cleaner in through your side, and suck up as many pieces as they can find. To my mind, this is a giant improvement over a previous procedure that involved cracking ribs and snuffling around near a bunch of really important organs by hand. Still, recovery is long. When they told me it would take 6-8 weeks, I assumed that only applied to little old ladies and whiners. But it actually takes 6-8 weeks. I had to walk around with a baseball-sized bag of intravenous painkillers in a fanny pack for a couple of weeks after I got home. Stylish! My myasthenia is still part of my life, but perhaps it isn't as bad as it would have been.

Anyway, after that life got back to normal. The medications and surgery made the symptoms manageable for me. It took a while, but the muscle weakness and double-vision stopped. I got my energy back. I felt normal. I went off medications for a while. Then we moved to a new state.

Moving is stressful, emotionally and physically, but I thought I was fine. I was exercising, enjoying my new home. I had a new neurologist, who is a leading expert in myasthenia. He was recommended by my previous doctor who told me, "I met him on a web chat for people who are into weird diseases." That kind of makes them sound like fetishists, but it was an excellent recommendation all the same.

I made an appointment right away although I felt fine. I wasn't interested in being on medication again. However, in one of our early office visits, something in one of the tests made him nervous and he sent me for a breathing test. I was embarrassed about taking the technician's time. They ask all these questions, "Can you walk up a flight of stairs without getting winded?" of course. "Can you walk a mile?" Sure. I was swimming every day and felt fine. What the heck was I even doing there? Then, I failed the breathing test. One of the scariest parts of myasthenia is that it can start to affect the muscles that control breathing. Even scarier is that my breathing was reduced to 30% of my expected capacity and I didn't even know it. Shouldn't I have felt that? But, it was dangerous. A case of pneumonia would have sent me to the ICU. So, I went back on prednisone and other drugs as well.

Today, I still take medications and see the doctor regularly, but the disease is well managed and not controlling my life. I'm still exhausted some days, but it is hard to say if that is the disease or just regular exhaustion. I occasionally get little flickers of disease if I'm sick, but nothing like before.

The disease name myasthenia gravis is Latin for "extreme weakness." Frankly, getting a disease called "extreme weakness" just pisses me off. But these days, I've got my muscles back, at least for the moment. I worry that I may not be as capable and active as I am now forever. It is a good reminder to live for today, take the vacations, go on the hike, play with the

kids. Continued good health isn't a guarantee for any of us, so enjoy every day, take care of yourself, and have fun right now!

I write a blog about health, exercise, and nutrition as it relates to autoimmune disease (<http://autoimmune.blog>)

*Mary*