19

"Yes, I have Usher Syndrome"

Anatomy is destiny.

—OLD MEDICAL SCHOOL SAYING

More than half the individuals who are deaf and blind in the United States have Usher syndrome. This is about ten thousand people, three percent of those who are born deaf. Fortunately, RP usually descends slowly and spares children. Even at my old school, those students who groped for the walls of hallways and felt for the table under their books usually had enough sight to learn through vision.

Art Roehrig says that I accepted my diagnosis quickly. Acceptance didn't feel quick. Painful, yes. Many layered, yes. Intermittent and reversible, yes. Quick? No way. For a long time, I couldn't even talk about it. There is an ancient story about a Spartan boy who found a mountain lion, hid it under his shirt, and went about his tasks, even as the lion, restless and hungry, clawed at his stomach. That was me and my diagnosis. I helped students in the library, consulted with teachers on appropriate readings for their classes, took care of Jason, and went out most weekends with Lance to visit friends. I laughed and gossiped. And all the while there was this little lion of a diagnosis clawing at me under my shirt.

Rachel Stone, now married to Ray Harris, was teaching at Kendall, the elementary school on the Gallaudet campus, just down the hill from the high school where I worked. Rachel and I had remained friends since our Gallaudet days. We were both part of a group of couples that got together once a month in each other's homes to watched captioned movies, available then through the federal government. Rachel would later write *Let's Learn about Deafness,* a book for teachers of deaf students that included historical information about deaf people and deaf education, and activities for the classroom. She was taking graduate courses in counseling and pregnant with her second child—her first had been a deaf girl.

"I feel great," she said, smiling and touching her stomach.

I was pleased that she had found time to stroll into the library and talk.

"You look good," I smiled.

She had just switched to maternity clothes, and she was as lively and effervescent as ever.

"I hope it's another girl," she said, "and I hope she's deaf."

I laughed. In those days, no one had a clue about a child's sex until he or she was born, and it didn't surprise me one bit that Rachel wanted another deaf baby.

"Did you save the baby clothes?" I asked.

"Oh, yes," she said. "They were hardly worn."

I thought about Jason. He was starting to walk now, a miniature man toddling about the room.

"They grow so fast," I said, shaking my head.

She nodded, still smiling as a student arrived with a book in his hand.

"Just a second," I told Rachel and turned to quickly stamp the boy's book. I reinforced the date that the book was due by pointing to it and meeting the boy's eyes. We always hoped that this would get our books back on shelves before the advent of summer, though it rarely did. The boy nodded and left. I turned back to Rachel.

"Why don't you admit you have Usher syndrome?" she asked suddenly.

"What?!"

Her question took me completely by surprise. I studied her face, looking for a smirk or an expression of fear. I saw only friendly concern. She had used the correct sign for Usher syndrome. She may have studied it in her master's program at Gallaudet, I realized.

"I don't have Usher syndrome!" I said my heart pounding.

Rachel shook her head. "You don't see us," she said.

She pointed at the boy disappearing around the corner, and I realized that he must have tried to get my attention before I saw him.

"We wave for your attention, and you just ignore us," Rachel was positively gentle.

"I'm busy!" I exclaimed, gesturing frantically at the papers before me.

Rachel's face softened even further, but it held a trace of exasperation, too. I tried to think of a way to change the subject and couldn't. Helplessly, my eyes returned to Rachel's.

"Are you afraid that you will lose friends?" she asked.

It was another question I wasn't ready to answer. I looked away. My frustration was fading. So were my defenses. I couldn't stay mad. Being mad took too much unpleasant energy. I glanced back at her.

"Everyone knows!" she was blunt without being unkind. "Everyone knows and no one cares. You are still the same person."

I looked away again. What purpose was served in pretense anyway? Who did I think I was kidding?

"It's best to just say it," said Rachel.

"Yes, okay," I burst out finally. "I have Usher syndrome."

I looked back at the floor after I said it. It felt good and bad at the same time, another kind of box was falling away.

When I lifted my eyes back to Rachel, I felt suddenly like crying. She hugged me.

"I've had it a long time. A long time," I told her.

She nodded. She looked like she might cry, too.

"It must be hard," she said.

I nodded. "I remember at my old school we would all go running for the dorm and I would always trip on the step. No one else

tripped. Just me." I paused and laughed a little in my tears. "It never dawned on me that everyone one else could see the damn thing!"

And that was that.

They say news travels through the deaf community faster than through the hearing community. This may be true. Once any of us finds out information of any sort, we feel obliged to immediately inform our friends. After I told Rachel, I felt that everyone knew, and that helped me to be able to talk about it more easily. Somehow that made me feel better.

From the beginning, Lance and I had wanted two children, but the dramatic loss of vision during my first pregnancy scared me. My mother-in-law came with me to the doctor's again. Again, I watched her hands and lips as the doctor spoke. The doctor said that no one knew why pregnant women reported accelerated vision loss and he agreed that a second pregnancy was dangerous; it might destroy more of my remaining vision.

"Do you think the child would have Usher syndrome?" I asked. It was the most important question of all.

He and my mother-in-law talked for a while before she turned to me.

"There's little chance that the child would have Usher syndrome," she said. *"Lance's deafness was a total surprise, and nobody in our family has ever been blind."*

"Our children would probably be safe?" I asked.

She nodded.

I glanced from my mother-in-law to the doctor. His face was impassive.

When I was pregnant, doctors couldn't test unborn children for genetic problems other than Down syndrome. A few years later, we could test for a number of the really outrageous recessive genetic mischief-makers—cystic fibrosis, sickle-cell anemia, Huntington's chorea, Tay-Sachs disease, some forms of breast cancer, and Usher syndrome—but most people didn't. Even when it was available,

testing was no easy matter. Chromosomal material put through its paces to check for Huntington's chorea, for example, would yield no information about Tay-Sachs or Usher. Each test had to be performed separately, each cost a bundle, and most were not covered by insurance. No wonder testing was rarely even considered until one was in terror for oneself or one's children, and then it was almost always too late.

Still, our dream was for two children. We both felt Jason should have a brother or sister, just as Lance and I did.

"It's up to you," Lance told me.

That's all it took. As had happened before, I became pregnant the moment I considered pregnancy.

Lance suggested that if the child were a girl, we name her Rebecca, after his grandmother.

"Such a hard word to say!" I told him. Our lips went through the shapes of the sounds, three of them in percussive succession.

"I don't like it," I admitted. With Jason, we had chosen a name that was similar to his grandfather's without being exactly like it.

"How about Rachel?" I asked finally.

"You want to name her after Rachel Stone?" he asked.

"Partly," I said. "But partly I really like the sound of the name."

"It's a good Jewish name," Lance said.

Rachel Sara Fischer, six pounds, twelve ounces, was born nine months later. Like her brother, she was perfect. Her birth was easy, too.

"I'm a baby machine," I told my husband.

He shook his head. I knew what he was thinking. My visual field had swooped inward again with Rachel's birth; unremitting black closed in another pronounced notch.

"Well . . . at least from the nose down," I tried to laugh.

Like most deaf people, vision more than anything else defined my world. It was through vision that I had language and learning, vision that was sharpened by use and the need to survive. I didn't mind being deaf. Although deafness took away hearing, it gave me community—and that community was based on sight. No matter how I tried to look at it, blindness was terrifying. It was right up there with death.

Rachel was only a baby when I drove to an evening appointment and bumped over an unseen median strip. It had been painted the same color as the road, and I didn't see it even after I drove over it. Frightened, afraid of hurting other people as much as myself, I stopped driving at night altogether.

It is amazing how we can compensate for such loss. Even now, able to see only one-sixth of the range of normal human sight—under the best conditions—I sometimes forget how limited my eyesight is. I am surprised when I bump into things. I forget that everyone else doesn't have to look down just to be able to see the floor.