

THE MYSTERIOUS *and* CASE *A* OF MOTHER'S HUGS VANISHING DELAYED BONES

By Sandy Van and Kevin McClanahan

Cooping in her mother's lap, Sky Meadow Harms is the center of the universe. And when it's time to hit the floor crawling, she is picked up, hugged and turned loose by dad, Ryon, for whom the birth of his daughter and the life of his wife are nothing less than miracles.

Laurie Harms, 31, suffers from a disease so rare that fewer than

200 cases have been recorded worldwide. She endured nearly two decades of pain and uncertainty before reaching Cedars-Sinai Medical Center and the care of Frank L. Acosta, Jr., MD. Four other neurosurgeons had already given up on her care.

Fortunately for Laurie, Ryon and Sky, Dr. Acosta tends not to look back at what has been, but to imagine and explore what may be. The director of Spine Deformity in the Department of Neurosurgery, Dr. Acosta trained in complex and reconstructive spine surgery, and studies such intricacies as tissue engineering to repopulate worn out spinal disks with



healthy cells. But Laurie's case was unlike anything he had ever seen.

"Her condition was quickly going downhill," recalls Dr. Acosta. "Her head had actually collapsed over her shoulder because of bone loss in her neck. We could not send her home like this."

Laurie was 13 when she began to experience progressively debilitating pain in and around her left ear. Doctors dismissed the symptoms as ear infections, sinus problems or muscle spasms, but after years of emergency room visits, a CT scan revealed disintegration of her neck bones and an undetermined tumor mass.

The pain, bone loss, and questions kept coming, and at age 22, Laurie was told she had what amounted to a broken neck. She needed bone fusion surgery right away.

Single, and facing major surgery and an uncertain future, a shaken Laurie thought of Ryon. They had worked together at a small café three years earlier, but their lives went in different directions. Though she had rarely seen him since they parted ways, she picked up the phone and punched in his number.

"I'd always loved Ryon, and he was the first person I wanted to call," explains Laurie. "He knew from my voice that it wasn't good news."

Ryon appeared on her doorstep the next day, leaving his apartment and job behind, and he was at her side at a major Los Angeles-area hospital where surgeons fused the bones in her neck.

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By the time they were married in 2003 the rate of her bone loss had slowed. Except for periodic emergency room visits and the chronic pain she suffered, they lived a fairly normal life for a while.

They also got a better understanding of the enemy they faced when an oral surgeon finally identified the mysterious disease—a decade after the original onset of Laurie's pain. Gorham-Stout syndrome, also known as vanishing bone disease, is characterized by the body's inability to normally

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regenerate bone. It also is associated with the formation of painful, fluid-filled tumors, a condition related to lymphangiomatosis. Bone deterioration can start, stop and start again and travel to adjacent structures, resulting in profound disability and even death.

"When I read the symptoms, they fit me to a T, and I realized what I had experienced for so long suddenly made sense," Laurie remembers. "It was a relief to know but, at the same time, I realized it was going to be a struggle in the years ahead."

Laurie defied the two-year survival predictions but experienced a major setback and a new round of treatments in 2008. Eventually, her oncologist arranged for her to try a new type of chemotherapy, but at the last minute she opted out. A week later she discovered she was pregnant. "Ecstatic but terrified" at the prospect of this surprise and potentially high-risk pregnancy, Laurie and Ryon chose to proceed despite the odds. Sky Meadow was born premature but healthy at 4 pounds, 8 ounces on September 28, 2009.

The new year brought another series of medical crises. Laurie's spine collapsed and she was rushed to an emergency room. When Ryon walked into his wife's hospital room, he saw

her head resting on her shoulder. "As soon as I saw her I knew that her neck was broken," he recalls. The bones had literally crumpled under the weight of her skull.

She lay helpless in a hospital bed—neck broken, arms limp and useless, and voice muted by tubes that kept her alive. Pressure on nerves in her neck and shoulder triggered intense pain and paralysis in her arms. Normally petite, at little more than 100 pounds on a 5-foot-2-inch frame, Laurie had dropped to 85 pounds. Holding her baby was out of the question, and her doctors gave up hope.

In desperation, an aunt called her neurologist, Hart Cohen, MD, at Cedars-Sinai, who put her in touch with Dr. Acosta. By that afternoon, family members were rushing X-rays to his office for review.

"Dr. Acosta called us the next morning—a Saturday—to tell us he thought he could help Laurie. It was the most amazing feeling because all of a sudden we had hope again," Ryon recalls. "Other doctors told us they couldn't do anything, that nobody could help her. Without even knowing us, Dr. Acosta agreed to see us."

Dr. Acosta asked neurosurgeon J. Patrick Johnson, MD, to assist with the surgical plans and procedures. "I have encountered many complex cases, but I had never seen anything like this," recalls Dr. Acosta. "The whole concept of fusion depends on bone growing across the hardware we place along the spine, so the bone loss of Gorham-Stout syndrome presents a major challenge. You can place any type of screw and rod but if fusion doesn't occur, the procedure is not going to solve the problem."

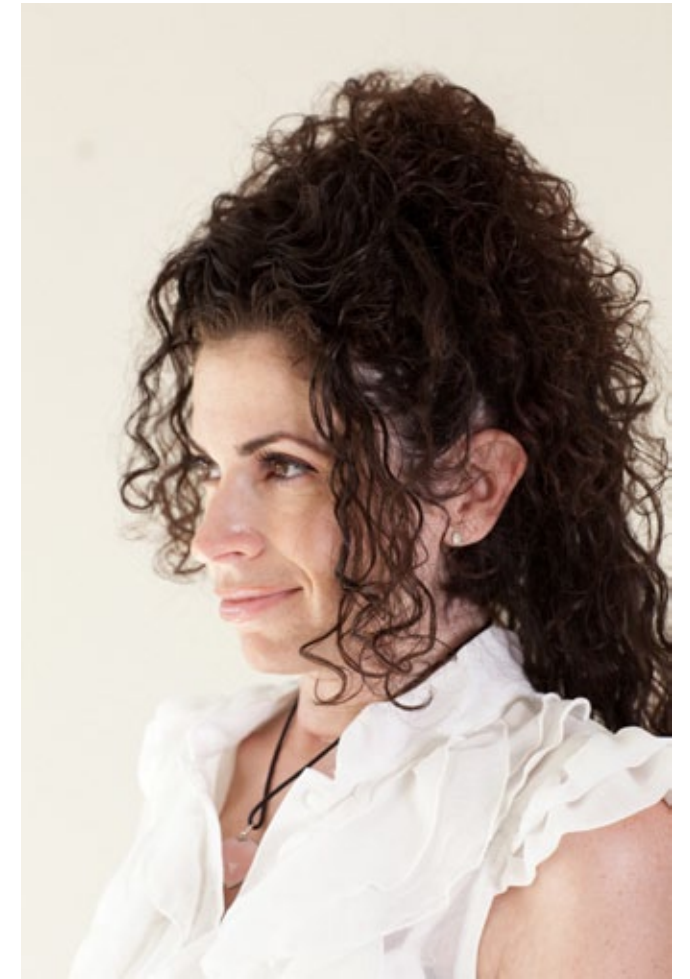
During two complex surgeries, the neurosurgeons repositioned an existing plate and extended the previous fusion with rods along the thoracic spine, anchoring the hardware in solid bone. They also adjusted disk spacing between two vertebrae and placed a protective plate over the hardware. After a week's recovery, Laurie went home with a virtually rebuilt upper spine and 36 staples in her head and back.

"Laurie has done really well since the surgery, regaining a good deal of arm and hand function, and she continues to make progress," says Dr. Acosta, who expects to perform

additional surgery in the coming months. "She isn't out of the woods yet—and the bone loss has to be stopped."

While there is still no cure for Gorham-Stout syndrome, Laurie remains optimistic. She is back up to weighing nearly 95 pounds and full of energy. She has started driving again, and is working hard in physical therapy to increase her upper body strength and mobility. She improves almost weekly and is now able to lift up 20-pound Sky by herself.

"I have this beautiful little treasure and she's all the motivation I need," says Laurie. "I already feel so blessed—every little accomplishment is simply a bonus." 🐾



What's Now, What's Next NEUROSCIENCE AND THE BRAIN

Keith L. Black, MD
Chairman and Professor, Department of Neurosurgery; Director, Maxine Dunitz Neurosurgical Institute; Ruth and Lawrence Harvey Chair in Neuroscience

Most important scientific achievement of the past 5 years:

Our progress with the dendritic cell vaccine for brain cancer is one of our more important scientific achievements in the past five years.

We have shown an increase in two-year survival of patients with Glioblastoma Multiforme (GBM), the most aggressive form of brain cancer, from 8 to 42 percent. We have also correlated

that dendritic cell vaccination and chemotherapy work synergistically to improve treatment.

The next big thing: We are currently working on a project we call the "Brain Window," a non-invasive imaging technique to diagnose early stage Alzheimer's disease thru the eye. Our team of

researchers has shown in animal studies that nerve-cell damaging amyloid plaques appear earlier in the retina than in the brain. Using this knowledge, we are developing a noninvasive optical

imaging technique to potentially detect early stage Alzheimer's disease.