

A Child, a Bizarre Tumor, and a Perilous Operation

By Keridwen Cornelius | The New York Times | Published: May 13, 2008



Grace Webster with her therapist Stacey Neill-Wiseman, right, and her father, Perry, left.
The convulsive seizures are a thing of the past.

PHOENIX — Three-year-old Grace Webster perches on the operating table, tiny and cold, covered only by a diaper and her sandy-blond Raggedy Ann hair. Her blue eyes gaze warily at the monster-size machines sprouting tube tentacles that encircle her — machines that will guide surgeons four inches into her brain.

Grace had her first menstrual period at 14 months old. Her body is racked more than 10 times a day with seizures, some of them bizarrely mimicking laughter or rage.

The source of her suffering is a hypothalamic hamartoma, or H.H., a tumor on the hypothalamus that strikes only a few thousand people in the world. And while the tumor is not malignant, until five years ago it was considered incurable, even when baffled doctors could diagnose it. Surgery was risky and largely ineffective. Medication seldom helped. Many children were institutionalized.

Now, thanks to an innovative surgical procedure, scores of these children have been cured at two centers that specialize in the disease. One is in Melbourne, Australia; the other is the Barrow Neurological Institute here in Phoenix.

It is 8 a.m. on April 20, 2007, and on the operating table at Barrow a nurse and a neuroanesthesiologist are trying to coax the anesthesia mask onto Grace's mouth. She is having none of it. Her face turns bright pink and crinkles into a wail. One arm clutches her plush puppy; the other stretches toward the door as she calls for her mother, Erica.

Finally the mask is in place, and Grace's belly billows rhythmically as the anesthesiologist pumps the vapors into her body. The nurses insert tubes into her mouth, arm, toes, back and urethra. (Now it is Grace who is sprouting tentacles.)

She is wheeled into the adjoining room to undergo a specialized M.R.I. It produces a high-definition image integrated with a tracking system that will guide the surgeons to the tumor. The doctors will take images of Grace's brain throughout the surgery, pinpointing the location of structures. The scans will also reveal the exact size of the tumor.

"It's a big, nasty one," says the anesthesiologist, Dr. Steven Shedd. At 3.23 centimeters across, it is about the size of a Ping-Pong ball. It bulges out from the hypothalamus and fills much of the neighboring cavity.

The hypothalamus is one of the brain's jacks of all trades. It maintains the body's status quo, regulating temperature, blood pressure, fluid balance and digestion. It also controls appetite, aggression and embarrassment, accounting for the strange cluster of hamartoma symptoms.

When Grace had her first menstrual period, her mother rushed her to the pediatrician. Tests revealed that Grace's reproductive organs had reached the developmental stage of a 12-year-old. She was given a diagnosis of hypothalamic hamartoma four days later. Soon, the Websters were told, Grace would experience "gelastic," or laughing, seizures.

The seizures begin with a wide, crooked smile, like a string attached to her eye and the corner of her mouth has been tugged slightly. "That's not her smile," said Erica Webster, a business analyst, showing a home video of her daughter. Grace's stare is unfocused, her giggle jerky and her breathing convulsed. Still, to the untrained eye, it looks like an adorable quirk, not the fingerprint of a massive tumor.

Between the ages of 4 and 10 — or earlier in severe cases like Grace's — the character of the seizures morphs from Jekyll to Hyde. Fits of laughter become bursts of room-demolishing rage and intense, paralyzing fits.

"It's like turning on a light switch and having somebody take over your child's body," Mrs. Webster said of the rage seizures. The first time, Grace was coloring contentedly, when — flick — she was screaming and hitting wildly. When her parents blocked her from smacking the furniture, she turned on herself, scratching her face and biting her arms.

Eventually, cognitive development goes from steady to sluggish to stagnant. The children often exhibit autism and social maladjustment. The personality that shone between seizures dims and disappears.

Faced with this dismal future, many parents like Erica and Perry Webster risk surgery to give their child a normal life. Mr. Webster, a civil designer, said they were reassured that "other H.H. kids have undergone the procedure and thrived." But it will be a long day in the waiting room.

Back in the operating room, the mood is upbeat. Rock music is playing. The surgeon, Dr. Harold L. Rekate, a grandfatherly presence with kind eyes, is shaving Grace's hair. There are nine doctors and nurses in the room, sheathing Grace with a linen closet's worth of sterile blankets. By the time they're finished, she is a mound of blue with nothing but a patch of scalp peeking through.

The first neurosurgeon to tackle the tumor is Dr. Scott D. Wait, youngish and TV-medical-drama handsome. "This is such a big tumor," he says, "that for cases like these we bring in multiple surgeons." Today, there will be three.

Grace Webster, who is now 4, had surgery last year to remove a large tumor from her hypothalamus that had been causing severe seizures. Dr. Wait slices into the scalp and peels back the skin to expose the skull. Before he drills, he touches the bone with a wand, a skewerlike navigational device that takes pictures of the brain. The squeal of the drill drowns out the music, which has taken a turn toward the elevatorish.

It's 11:22. Dr. Wait maneuvers into a surgical chair and presses his face into a giant microscope. The magnified image pops up on a TV screen on the wall. A scalpel and tiny scissors move into view, gently slicing and clipping away the brain's outer membranes. Underneath, purple veins rope and red arteries spider web around the flesh-colored convolutions of the brain.

11:55. The music is silenced. Dr. Wait delves slowly into the cleft between the two hemispheres, slicing through the corpus callosum — the bridge connecting them — into a membrane called the septum pallidum.

Surgically, this is a road less traveled. In the past, surgeons took the shorter path from the underside of the brain, with little success. Many parents were told there was no hope.

Two of those parents were Lisa and Jon Soeby of Phoenix, whose 4-year-old son, CJ, had as many as 300 seizures a day. The Soebys discovered that an Australian neurosurgeon, Dr. Jeffrey Rosenfeld, had pioneered a different surgical route: he approached the hamartoma from the top, diving between the hemispheres so they separated like two halves of a book. At the book's binding lay the tumor.

Since the doctors at Barrow would not perform the novel procedure, the Soebys flew to Melbourne. It was a success, and in 2003, Dr. Rosenfeld traveled to CJ's hometown to teach his "transcallosal" approach to the neurosurgeons here. They now specialize in it, receiving hamartoma patients from around the world. Grace Webster, from Anderson, CA., is patient No. 101.

12:20. Dr. Wait is navigating Grace's third ventricle, a tiny basin of cerebrospinal fluid. There in the fluid convulses a pink, fatlike mass: the tumor. Dr. Wait and Dr. Rekate consult the M.R.I. scans on the monitor. Green lines converge in a bull's-eye on the hamartoma. Dr. Wait digs into the tumor, which bleeds; the suction tip wails and vacuums the blood.

"Let me play for a little while," says Dr. Rekate, switching seats with Dr. Wait. He wields an ultrasonic surgical aspirator, an instrument that vibrates at 23,000 revolutions a second, simultaneously emulsifying, irrigating and sucking up the tumor.

1:26 p.m. A man strides in wearing a surgical cap with a piano-key pattern: Dr. Robert Spetzler, the director of the Barrow institute. Dr. Rekate surrenders the chair. Dr. Spetzler, sinewy and intense, probes at the tumor with the aspirator. Twenty minutes creep by. Dr. Rekate gasps. "Is that the basilar artery?"

It is the equivalent of a land mine, the main blood supplier to the back of the brain. As Dr. Spetzler swipes the aspirator around the vessel, he murmurs something like "I feel very uncomfortable here."

Puddles of bloody fluid flood the cavity, but only from minuscule arteries. The topography of the heart monitor and vitals signs remains reassuringly mountainous.

Dr. Spetzler attacks the seemingly unending tumor from all sides. The tissues begin to look deceptively similar. "This is where I trust what the wand is seeing," Dr. Rekate says, "because it's not so obvious inside."

2:17. Dr. Spetzler rises. "I don't think there's any more," he says. He exits unceremoniously, with a "Thank you" from Dr. Rekate. When the door shuts, Dr. Wait shakes his head. "Holy cow," he says, about Dr. Spetzler's decision to run the aspirator over the basilar artery. "I don't think I'll ever do that."

Dr. Wait begins to close Grace up and prepare her for another M.R.I. Dr. Rekate leaves. The rock music returns.

At 5 p.m., the results are revealed. "Wow, we got all of that thing out," Dr. Wait says. But he adds, "I can't guarantee she'll never have another seizure."

Once removed, a hypothalamic hamartoma will not grow back. But Grace's fate remains to be seen. At Barrow, 90 percent of hamartoma patients emerge from transcallosal surgery with at least 90 percent fewer seizures, Dr. Rekate says, and 60 percent of those are seizure-free. Ten percent are not helped at all. Two patients have died.

A year after the operation, Grace still suffers from a form of diabetes and low thyroid function, but these conditions are improving. Thanks to physical and speech therapy, she is catching up in her development, growing more active and sociable.

She must continue to get Lupron injections every 28 days until she is 11, to stave off the precocious puberty. But most important, she no longer experiences the seizures that convulsed her with seeming rage or laughter.

Now when Grace laughs, it is not the tumor, but the lyrical giggle of a sassy, seizure-free 4-year-old.