

## The girl

*in the mirror* | Surgery dismantled the face that Brenna Johnston was born with. Weeks locked in a titanium brace may give her the face of her future – and a chance at a normal life.

# Suspended in a cage



JOHN LOK / THE SEATTLE TIMES

**BRENNA JOHNSTON SPENDS THE SUMMER** *before third grade caged inside a titanium halo. A genetic defect kept her facial bones and skull from growing, threatening her life as the rest of her grew. Five weeks after radical reconstructive surgery at Seattle's Children's Hospital, the device has cranked Brenna's face forward almost an inch. The swelling and halo obscure her new profile, but her dad, Erin, mourns the face he first loved: "With that little smile and those big eyes ... it was just Brenna."*

## THE SECOND OF 3 PARTS

BY JULIA SOMMERFELD  
*Seattle times staff reporter*

If Brenna Johnston had her wish right now, she'd be shooting down that slide. She'd swing the swing so high it would bounce at the top. She'd have the run of the playground.

And if she had a second wish, she'd be invisible. Actually, just less visible. "Like maybe if they could see me, but not really stare at me all the time," she says.

It's recess at summer art camp at Veneta Elementary School in Brenna's hometown of Veneta, Ore., a few minutes from Eugene. But for 8-year-old Brenna, the playground is off-limits this summer. She sits on the railroad-tie curb, nodding toward the monkey bars. "If I went on those," she says, "I'd be history."

And those older girls by the climbing pole are, if not exactly staring, then certainly noticing.

She turns her back to them and scoops a handful of pebbles.

She curls her knees to her chest and props up the metal cage that encircles her face.

A month has passed since surgeons at Seattle's Children's Hospital & Regional Medical Center peeled back the skin of Brenna's face and sawed through her underdeveloped facial bones. They severed Brenna's upper jaw, cheekbones and eye orbits, then wired them to a purple titanium halo called a Rigid External Distraction system, or RED.

Bit by bit, Brenna's bones have been cranked forward by the device, correcting the bowl-shaped profile of Crouzon Syndrome.

A CT scan shows hazy threads of bone knitting the new face back to the skull. But it's too early to tell what Brenna will look like. The device must stay in place another month to stabilize these fragile, developing bones. A bump, a jostle, a wayward ball could collapse her budding cheekbones, upper jaw and eye sockets – and send her back to surgery.

"If I could lock her in a box this summer, I would," says Dr. Richard Hopper, Brenna's craniofacial surgeon.

Now, Brenna's mom, Robyn Johnston, gathers Brenna and her younger brother from art camp. Six-year-old Caden bursts into a sprint toward the parking lot. Brenna gives chase.

"Hey, hey," Robyn shouts after her. "Stop! No running!"

"But Caden gets to run," Brenna whines. "That's no fair!"

"You're right, it's not fair. But we don't want to do this all over again, do we?"

Brenna slows to a shuffle and slouches into a full-body pout.

Lots of things aren't fair this summer. It's not fair that she can't make wind chimes next week at camp because that's on a Monday and Monday is when she flies to Seattle to see "Dr. Hoppy." Not fair that she can't swim, ride her bike, turn cartwheels or slide down slides – even if she's super careful. That she can't ride horses, play soccer, play with any kind of ball – except for whiffle ball with her 3-year-old brother Perry, but not if he's swinging a bat around. Can't hop up the stairs, hop down the stairs, tilt back in her chair or twirl around the living room.

## Waking up to the RED

A month earlier, in the intensive-care unit at Children's Hospital, Brenna couldn't eat, move, see or talk. Her face was so swollen and top-heavy with the RED that she couldn't lift her head. Her eyes were puffed shut into shiny plums. Her jaws ached from being wrenched open.

Tears ran down her face, past the pins sticking through her

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bruised cheeks, under the metal scaffolding, and soaked the neckline of her Bugs Bunny pajamas.

“What’s the matter?” a nurse asked. “Do you hurt? Do you need more medicine?”

“No,” Brenna whined.

“What’s wrong?”

“Bored.”

It took eight years and more than 30 operations to get to this point — surgeries that propped open her airway, spared her hearing and expanded her skull to make room for her growing brain.

It will take another two months — months of discomfort and boredom — to construct a face that works.

Brenna was barely out of surgery when Robyn began to work the RED, twisting the four screws to slowly tighten the wires, a millimeter a day. When she first felt the resistance increase beneath the screwdriver, she recoiled, realizing she was actually cranking her daughter’s face forward.

“It doesn’t hurt as bad as it sounds,” Hopper assured her. “It’s more of a tolerable ache, like getting braces tightened.”

By the end of summer, the RED should tug Brenna’s face forward about 2 centimeters — almost an inch. The extra space should allow her to get adequate oxygen to her brain. It should protect her eyeballs from scratches. It should let her bite into a hamburger and crunch popcorn.

It should give her a chance at a normal life.

### “Just Brenna”

When Brenna is released from the hospital 11 days after surgery, her face is somewhere between before and after.

The transformation is so gradual that Robyn — used to adjusting the RED each day, to the feedings, the ointments, the medicines — barely notices her daughter’s profile growing.

But when Erin Johnston sees his daughter for the first time since surgery, he is stunned into tears. Even through the swelling, Brenna’s face is unfamiliar. She has cheekbones now.

“I wasn’t ready,” Erin says. “I didn’t know it would happen so fast.

“I loved that face for eight years. Whatever other people thought, I thought it was cute. With that little smile and those big eyes ... it was just Brenna.”

### A random error

Robyn and Erin Johnston met as students at the University of Oregon. Erin, a chemistry student with a sarcastic



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**A BUMP OR FALL COULD SHATTER** Brenna’s developing facial bones, so she must spend summer on the sidelines. The playground at Veneta Elementary School, where she attends art camp, is off-limits. “If I could lock her in a box this summer, I would,” says her surgeon, Dr. Richard Hopper.

streak, had signed up for an aerobics class to meet women. He couldn’t believe his luck when he saw the instructor. Robyn Peters had long, wavy blond hair, a take-charge manner and a rowdy laugh. She was studying for her master’s degree in exercise science and needed help with her chemistry homework.

They spent much of their courtship outdoors — camping, backpacking, scuba diving. They envisioned a future with two children and a house on the water. They were both 29, and had been married a year, when Brenna was born.

The pregnancy was uneventful until a last-minute C-section. Brenna was born with a warped skull, struggling to breathe.

Three weeks later, doctors diagnosed her with Crouzon Syndrome.

“We loved her to death but were really shocked,” Robyn says. “We were healthy. We did all the right things.”

A random error in a single gene stunted the growth of Brenna’s skull and facial bones. Her brain didn’t have room to grow; doctors warned that she might be mentally retarded. Her depressed nasal passages stifled her breathing; as a newborn, Brenna would work herself into a sweat as she strained for air.

She was 4 months old when she had her first surgery, to insert stents, like cocktail straws, from her nostrils to the back of her throat to help her breathe.

She was 6 months old when doctors at Oregon Health & Science University in Portland made the first incision across the top of her scalp. In a six-hour operation, they removed sections of the top of her fused skull and forehead to relieve pressure and give her brain room to grow. They expanded

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the vault with titanium plates and slices of bone from other parts of her skull.

The swelling afterward was so extreme that Robyn documented its progression with snapshots every 12 hours. By hour 60, Brenna's head was the size of a pumpkin and her ears were folded over. "She truly looked like she was going to explode," Robyn says.

But after the swelling subsided, Brenna's new forehead was crooked. It pushed her left eye socket lower than the right; one eye began to wander. And parts of her newly expanded skull were too thin to protect her brain.

### The long road to Seattle

After the surgery failed, the Johnstons were referred to Seattle's Children's Hospital, where a team of doctors specializes in complicated craniofacial cases: kids from throughout the Northwest with prematurely fused skulls, cleft palates and rare disorders. It took countless letters and calls to convince their insurance company to cover it.

They've since made the six-hour drive north to Seattle as many as 50 times.

Brenna's baby bones were too fragile to operate on again right away; her forehead was paper thin in spots. The surgeons at Children's decided to wait until she was 4 to redo the dramatic skull surgery. In the meantime, medical problems stacked up at every stage of Brenna's development. She needed tubes to help her ears drain properly and a series of nasal stents to help her breathe. Her tonsils and adenoids were removed to help clear her airway. Her blocked tear ducts were surgically probed. Teeth were extracted to relieve her painfully overstuffed tiny jaw.

Between surgeries, she saw speech therapists, nutritionists and eye doctors. She underwent hearing tests and sleep

studies. Developmental assessments showed she wasn't mentally retarded.

Robyn quit her job managing a fitness center to manage Brenna's medical care.

Erin worked long hours as a chemist, but the pay was good and the health benefits would prove essential: The cost of Brenna's medical care over her lifetime runs in the hundreds of thousands; the RED procedure this summer alone cost \$114,000.

Still the co-pays, deductibles, uncovered expenses, like orthodontia, and even the gas for the frequent trips to Seattle, added up. Robyn sold Mary Kay cosmetics to make some extra income.

And Robyn and Erin returned to those dreams of home and family they had as newlyweds.

"After we learned she's not going to die, it became about what can be done to live as normal a life as possible," Robyn says.

Genetic counselors assured them they wouldn't pass Crouzon Syndrome on to future kids.

Caden was born when Brenna was 2. "We knew we wanted her to have a little brother so she'd have an ally," Robyn says.

He has the kind of devilish grin that seems to come with redheaded boys. Sometimes he defends his sister, like when she gets a timeout and he insists, "Mommmmm, she didn't DO anything." Other times he aggravates her, like when she wants watch a movie and he refuses to get off the Game Cube. Sometimes, he plays the victim, like when Brenna mimics everything he says until he blows up.

Perry was born three years after Caden. He has his sister's bright blond hair, and is his mom's replica and shadow. By age 3, he has learned to play with Caden when he's feeling wild. When he's quiet, he does arts and crafts with big sister "Bwenna."

But the boys' perfectly formed skulls and plump toddler cheeks made Brenna's oddities all the more apparent. As an infant, her buggy eyes and wagging tongue seemed adorable. But as the rest of her grew, the middle of her face sunk into a basin. Her profile skipped the convexities of youth – the chubby roundness, apple cheeks and pouty lips – and went straight to the concavity of old age.

To Erin and Robyn, it was never about Brenna's looks.

"We just didn't go there," Robyn says. "When you're talking about your kid not being able to get enough oxygen to her developing brain or having her vision or hearing deteriorate, something like her face looking different doesn't seem very important."

### The summer Brenna was 4

This is what Brenna remembers about turning 4: "I had my birthday and then was in the hospital for a really long time. Like maybe for as long as summer is."

The pressure inside her brain vault was spiking and she was getting headaches. A brain scan revealed that screws



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**AFTER BRENNNA WAS BORN** with Crouzon Syndrome, a geneticist assured her parents that the genetic defect was random. So Erin and Robyn Johnston chose to have as normal a family life as possible. That meant two younger brothers for Brenna, and typical sibling rivalries. Caden, 6, is the reigning champion at "Super Mario Bros.," but when Brenna pulls an upset, she tends to rub it in.

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from the first skull surgery, when she was 6 months old, had worked their way into her brain.

In a 14-hour operation, neurosurgeon Dr. Richard Ellenbogen extracted the screws from her frontal cortex and patched the membrane around her brain. Craniofacial surgeon Dr. Joseph Gruss split parts of her skull to expand her cranium and reconstruct a new forehead.

"It was a beautiful forehead," Robyn recalls. "While it lasted."

But cerebrospinal fluid began to leak from Brenna's nose — the first sign of infection. Doctors placed a drain in her skull and put her on IV antibiotics. Nurses flushed more antibiotics under her scalp. The surgeons went back in to scrub the grafted bone.

Brenna didn't wake up for a week. But once she did, she took a clinical interest in her situation. Her nurses referred to the cerebrospinal fluid as "brain juice." The tube coming out of her skull was a "brain drain."

Brenna liked to see what was coming out of her brain. She pouted when doctors replaced the external drain with a permanent shunt that channels her excess brain fluid through a tube that runs into her stomach.

But for all the intervention, the infection proved too strong. A month after the first surgery, doctors reopened the incision across Brenna's head and cut away her smooth and symmetrical new forehead bone. The infection had invaded the bone graft.

Brenna was sent home — 10 weeks and eight surgeries after coming to Children's — with no forehead. Her scalp was sewn over the crater in her skull. It felt spongy to the touch, like a rubber ball; that was the membrane around the brain, protected now only by skin.

When Brenna started preschool, she wore a helmet.

### Summer means surgery

Brenna had grown wobbly and uncoordinated. She took corners a little too wide and stumbled over sidewalk cracks. She seemed dazed a lot and needed more repetition to learn.

"I don't know if it was all the surgeries or the infection or what, but that summer took its toll," Robyn says.

Yet more surgery would be needed. Brenna's skull would have to be opened again, and her forehead rebuilt. The base of her brain was slipping into her spinal column, and a fluid-filled cyst had formed in her spinal cord.

She was 5 the summer pieces of her spinal column were removed to help decompress her brain.

She was 6 the summer she got another forehead — a patchwork of rib grafts, titanium mesh and synthetic bone material.

She was 7 the summer they collapsed the cyst in her spinal cord.

Finally, the summer she was 8, her skull was big and stable enough to address the final and most obvious piece of the Crouzon puzzle: the face.



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**THE SIMPLEST THINGS**, like breathing and eating, have been ordeals for Brenna. As an infant, she needed a special bottle to accommodate her underbite. She mashed small morsels of solid foods between her tongue and the roof of her mouth, and avoided chewy foods. Here, dad Erin maneuvers a bite of hamburger under her titanium halo. Each bite is such an effort that by midsummer, Brenna has lost 5 pounds.

### Feeling the stares

The worst thing about wearing the RED isn't the needle pokes or sleeping with a big metal brace on her face. It's not having to drink milk through a syringe or slurp soft foods past the bulky retainer.

"The thing I really don't like is people staring at me," Brenna says. "At art camp, a girl said, 'Why do you have that stuff on your face?' She said, 'What's wrong her? Why is she like that?'"

"It made me have tears for a little bit."

She was 3 or 4 when she noticed that people were always noticing her.

"Mom says just to smile and say 'hi' and sometimes I do. But sometimes I just ignore them like I don't see them."

It got so it wasn't so bad around home, where most everyone knows — or knows of — Brenna. Around Veneta most people stopped staring long ago.

Until this summer. The RED means she looks different again — and this time the stares are worse. Brenna learned that back at the hospital, soon after the surgery.

She had shuffled out of her room with Robyn, headed for the playroom, when a younger girl turned a corner and came face-to-face with Brenna. She gaped at Brenna's scabby scalp stitches, furious pins and wires sticking out of her cheeks, the RED caging her face. The little girl screamed and ran back in the other direction.

Robyn took a breath, about to recite the standard speech. The one that comes after a rude stare, about how some people haven't been taught any manners and that's their problem.

But Brenna looked up at her silently. Her weary eyes seemed to say: I don't even want to hear it.

Not a word passed between them. They held hands and walked to the playroom.

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### A furry incentive

By mid-July, Brenna can breathe through her nose. Full, deep breaths that fill her lungs with oxygen, that energize and relax her. She can sleep through the night without waking to gasp for air. Caden finally sleeps through the night, too; his sister's raspy moaning used to give him nightmares of ghosts.

Robyn calls and has the oxygen tank picked up. "Her room finally looks like a little girl's room, not a nursing home," she says.

The RED is finally starting to pay off. Maybe this will be the last surgery of Brenna's childhood. And yet ...

"I'm not going to get my hopes up," Robyn says. "Because Brenna doesn't read the textbooks. She's always developing problems the doctors didn't see coming."

But Brenna is counting more immediate rewards:

"I can whistle through my nose. Listen." She issues a quiet hoot through her nostrils.

"And I can wrinkle my nose." She scrunches it up like a bunny.

But the biggest reward: "When I get my RED off, I get a puppy because I've had it so tough and been so brave."

She's been to every pet store in the Eugene area several times, but wants to go again today. Robyn gets her ready, combing Brenna's hair into pigtails behind the RED. She checks each of the screws bolted into her daughter's scalp.

"Are you done yet?" whines Brenna.

"With you? I'll never be done," says Robyn, planting a kiss on her daughter's cheek. Brenna rolls her eyes, impatient to go.

She is willing to face the mall in Eugene, face the strangers, face the stares, if puppies are involved.

"I'd do just about anything for a puppy," she says. "Even wear this RED and not complain too much."

Puppies, she says, don't notice the RED. She cuddles ev-



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**AFTER SEVERAL WEEKS**, the RED has opened Brenna's nasal passages enough that she can breathe through her nose. She's discovered new smells and for the first time, can sleep without an oxygen tank. "Does that mean I can go on sleepovers?" she asks.

ery puppy in the shop, giggling when they lick her face.

Watching her daughter's big new smile through the wires of the RED, Robyn sees an ending to this part of Brenna's story.

And it's a happy one.

### A sudden threat

Six weeks down, two to go.

It's a lazy morning, the first weekend of August. Robyn studies her daughter across the breakfast table. Brenna's flyaway blond hair looks slick and matted. Robyn tries to take a closer look, but Brenna is cranky and pulls away.

That evening, at bath time, Robyn finally gets a good look. The RED's second screw on the left side has formed a pocket, an abscess filled with green ooze. It smells sweet and rotten.

She pages Dr. Hopper, frantic. He tells her to get to Seattle first thing Monday morning, but feels the same dread: MRSA.

A couple weeks earlier, several sores on Brenna's torso and arm, possibly spider bites, were infected with the microbe methicillin-resistant *Staphylococcus aureus*, or MRSA — a scourge of long hospital stays. With her immune system diminished by surgery, Brenna was colonized by the germ.

A flood of antibiotics knocked down the sores — and the fears. Everything would be OK — as long as they kept the MRSA away from the wounds around her scalp, away from the healing bones and the places where the RED attached to Brenna's skull.

### The choice

Dr. Hopper is stunned when he walks into the examina-

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**THE JOHNSTONS TRY TO BUILD A NORMAL LIFE**, but Brenna's condition can be all-consuming. Robyn feels like there's never enough of her to go around. She hired a nanny this summer to help care for the two boys while she traveled to Seattle weekly with Brenna. On this night, Brenna needs her halo cleaned, but Caden wants a bedtime story.

tion room the following Monday. Brenna is a rag doll, too weak to sit up by herself. Her legs are flushed and warm, her eyes vacant. Usually eager to please “Dr. Hoppy,” now she whines and won’t look at him.

He examines the oozing screw site and knows what the cultures will later reveal: MRSA has infested the wound.

He puts Brenna on IV antibiotics and schedules surgery for the next day. The infected pin has to come out.

“That’s not Brenna in there,” he says, leaving the room. “Surgery like this can knock the stuffing out of a child. I’m worried there’ve just been too many blows, physically and psychologically.”

That night, Hopper lies awake. He can’t close the mental file on Brenna.

The infection could make its way to the shunt in the back of her brain, to her central nervous system, to her bloodstream. In this weakened state, she might not survive that.

Hopper weighs the consequences:

He can remove the RED the next day and pull her out of immediate danger. Once the hardware is off, the wounds can heal and the infection will lose its foothold.

But it has been only six weeks. He’s never removed the device so early. Her bones could fall back, pinch her airway and force them to redo the surgery. He’s not sure the broken little girl he saw today could pull through another brain infection – or another summer in a cage.

Hopper must choose: Brenna’s face.

Or her life.



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**ROBYN COMFORTS BRENNNA** as she faces yet one more medical procedure at Children’s Hospital. “I wish I could go through all of this rather than her,” Robyn says. This evening, Brenna whimpers, “I’m scared, I’m scared, I’m scared,” until the tear-choked words form a comforting, numbing chant.