

The girl in the mirror

Our face is our identity, the shape of our personality, the frame of our soul. It's how the world sees us and how we see ourselves. But to little Brenna Johnston, her face is her biggest threat.

THE FIRST OF 3 PARTS

BY JULIA SOMMERFELD
Seattle times staff reporter

The date has been circled on Robyn Johnston's desk calendar for months: June 22.

Tomorrow.

But she's known this day would come for eight long years, since soon after Brenna was born. She knows that after tomorrow, after the surgery, she'll never see her daughter's face like this again.

To Robyn, tonight already feels like a memory.

And to Brenna?

"Tonight is girls' night!" she squeals, bouncing on the couch at the Ronald McDonald House, a few blocks from Seattle's Children's Hospital & Regional Medical Center. Dad and her two little brothers are back home in Veneta, Ore., six hours away. So tonight it's just her and Mom, with popcorn for dinner and her favorite video, "Spirit: Stallion of the Cimarron."

She skirts talk of surgery. "Horses are the best, except maybe unicorns, but that's pretty much a horse with a thing on its head."

Brenna watches the cartoon.

Robyn watches Brenna.

Her eyes trace her daughter's profile, shaped like a quarter moon. Brenna's gray-blue eyes bulge beyond their bony orbits. Her nose, cheeks and upper lip look deflated. Her lower jaw juts far forward, giving her a smile like a friendly snowplow.

Her teeth don't line up to chew; she mashes the popcorn between her tongue and the roof of her mouth, her lips puckering like an old lady with her dentures out. Her nose is too constricted to draw air; her raspy mouth breathing competes with the movie soundtrack.

The sunken face is textbook Crouzon Syndrome. And "trademark Brenna," Robyn says.

A glitch in Brenna's genes stunted the growth of her skull and facial bones. Repeated skull surgeries have saved her life.

Tomorrow's radical face surgery will help her live it.

In a procedure as intimate as it is invasive, surgeons will peel off Brenna's face, then wrench her underdeveloped facial bones from her skull. They will bolt those bones to a device that, over two months, will pull them forward — like braces



JOHN LOK / THE SEATTLE TIMES

EIGHT-YEAR-OLD BRENNA JOHNSTON was born with a rare condition called Crouzon Syndrome. A random genetic defect stunted the growth of her skull and face, limiting her ability to breathe, eat and see. She has had more than 30 surgeries to protect her brain and save her life. In June of this year, surgeons at Seattle's Children's Hospital & Regional Medical Center prepared to give her a new face — and a chance at a normal life.

< Continued from previous page

realigning teeth. If all goes well, her nasal passage will expand. Her eye orbits and cheekbones will cradle and protect her eyes. Her top teeth will meet her bottom teeth.

And, by summer's end, she'll have a new profile.

Now Brenna catches Mom staring at her and flashes an impatient look. Staring is rude. So Robyn scoots closer to Brenna and turns her attention back to Spirit.

But first she commits to the deepest part of her memory the face her daughter was born with.

Operating Room 14



Dr. Richard Hopper specializes in complex face and skull surgeries.

Just after 9 a.m., in Operating Room 14 at Children's Hospital, the surgeon works his long, gloved fingers through Brenna's hair, gently braiding her blond bob into tiny pigtails.

It's a good thing Brenna's under anesthesia; she hates having her hair done. But if there were anyone in the world she'd allow to play with her hair, it's Dr. Richard Hopper — "Dr. Hoppy," to Brenna — her craniofacial surgeon and first schoolgirl crush.

The braiding secures stray hairs out of Hopper's way, clearing a path for the scalpel, and gives his fingers a warm-up.

Brenna's shallow face peeks like a vacant mask from its veil of sterile blue cloths; anesthesia has emptied it of expression. Her eyeballs are shielded with blue plastic lenses in case an instrument slips during surgery.

Hopper parts the braids to uncover a groove along the top of Brenna's scalp, a hairless white scar that runs ear-to-ear, a few finger-widths behind her bangs.

The first surgical cut is usually an elegant one, with the scalp separating cleanly under Hopper's blade. But this path has been traveled too many times before, by too many other surgeons; the scarred tissue is stubborn and bleeds heavily. Hopper tugs through the fibrous route slowly; there are potholes of missing skull — and unprotected brain — just beneath his knife. His assisting surgeon, Dr. Anna Kuang, soaks rag after rag with Brenna's blood.

Once Hopper has completed his cut, he slides a tiny spatula under the scalp to separate the skin and sheets of tissue from the skull. Then, as if he were rolling off a tight leather glove, he peels back the mask.

"I've got little bones"

When strangers ask about her face, Brenna usually hides behind her mother. But sometimes she'll answer, describing her condition as she understands it: "I've got little bones."

A look at her skull, exposed now on Hopper's operating table, proves her description apt. The facial bones from her eye sockets to her upper jaw are tiny, thin and compressed — a baby's bones under an 8-year-old's skin.

Doctors knew something was off the moment she was born. She had turned breech at the last minute, and was



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MIRRORS DON'T HOLD MUCH INTEREST for Brenna. Her parents have never made a big deal of her looks, and she's usually too busy — decorating hand puppets with glitter glue or updating her sticker books — to fret about her appearance. Her mother, Robyn, tries to memorize the face her daughter was born with. "We've told her, but I'm not sure Brenna really understands how different she'll look after the surgery."

delivered by emergency C-section. Her skull was strangely elongated, like a football resting on the tee of her neck.

She was whisked away for CT scans and MRIs. She was put on oxygen and a breathing monitor. When she was returned to Robyn's arms a day later, her baby-fast pulse was visible in her bulbous forehead and her breaths were strained and irregular.

"I was scared to death," Robyn says. "We didn't know what it meant. The doctors didn't even know."

Three weeks later, Brenna was diagnosed with Crouzon Syndrome, a genetic skull defect that occurs in fewer than 1 in 25,000 births. Brenna's mutation was random; her two younger brothers were born with normal skulls and faces.

"If you think of the genome as the Encyclopedia Britannica, then her mutation is a single wrong letter in a single word," says Dr. Michael Cunningham, director of the Craniofacial Center at Children's Hospital.

If the mutation had occurred just 11 letters away, she would have dwarfism rather than Crouzon, Cunningham says. But for Brenna, that single genetic typo spelled a constellation of errors in her skull and face.

Normally, an infant's skull is made of plates of bone separated by fibrous tissue called sutures. As the brain develops, the sutures act like expansion joints until growth is complete and the sutures fuse.

But Brenna's coronal sutures — which arc across the top of her head like a headband — fused in the womb. Left untended, her brain would have grown in the direction of least resistance, misshaping her skull, leading to brain damage and possibly death.

And while the rest of Brenna's body grew — lanky limbs and feet so big she trips over them — the middle of her face didn't. As she grew from an infant to a toddler to a little

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girl, her nose, eye sockets and cheeks appeared to sink.

The structural faults radiated through her head. Her ear tubes and tear ducts were malformed. Her nasal passages were too narrow. Her tongue was in the wrong position in her mouth.

The surgeries started when she was 4 months old. There have been more than 30 since.

Some were relatively minor – to insert drainage tubes into her too-small ear canals or breathing stents down her collapsed nose.

Others were major. A shunt was planted in her brain to relieve pressure. She's had her skull reconstructed – enlarging her cranium with titanium plates and patches of her own bone – three times.

This has been Brenna's life. She doesn't remember the first day of kindergarten, but she remembers Mary Ellen, the nurse she had when she was 4. Drop her in any corner of Children's Hospital and she can make her way to the playground.

Her security blanket is a hospital rag named Towely.

The function of a face

Brenna lies under the glaring lights of the operating room, her face folded down like a leather shoe tongue. Her exposed forehead bone is dotted with tiny titanium plates and screws left over from earlier surgeries.

An array of CT scans posted around OR14 show the moonscape of her skull, pocked with strange shadows and rough-looking craters. Her actual cranium is a patchwork of rib grafts, metal plates and synthetic bone paste.

Brenna's hair, thick and cowlicky as a well-loved Barbie's, usually camouflages these bumps and valleys. But the scalp beneath feels rough and dimpled, like a lava stone.

Hopper and Kuang pry off 20 plates that have outlasted their usefulness and risk poking through the skin. They

leave others where bone has healed over them. Brenna will have a smooth forehead now.

Hopper peers through microscope lenses attached to his glasses. He confirms what he sees on the scans behind him: The anatomy of Brenna's face is all wrong.

"There are a huge number of functions of the face you forget about unless they don't work well," Hopper says.

And while it is inevitable to focus instead on the form of the face, the two are inextricable and equivalent.

A straight, protruding nose signals an unobstructed airway. Aligned teeth can bite and tear. Deep-set eyes are shielded from scratches, and brows protect them from falling debris. Pronounced cheekbones do more than lend an aristocratic appeal – they're bumpers for the eyes and a major support for the entire facial structure.

The face Brenna was born with betrayed her on all these counts.

There's a blockage instead of an airway behind her nose. Asleep, she relies on an oxygen tank to keep her breathing. Limited oxygen and fitful sleep sap her energy and could delay mental and physical growth. Left unchecked, her sleep apnea could cause irreversible heart damage.

Doll-sized cheekbones and shallow sockets leave her eyeballs exposed to the slightest brush.

"She plays soccer," Hopper says. "Can you imagine what a ball could do to an eye, bulging out like that?"

Her eyelids must be coated in thick ointment at night because they don't close all the way. The constant exposure could eventually scar her corneas. She already needs glasses to see the chalkboard.

An adult's finger can fit into the gap made by her underbite. She mashes soft food between her tongue and the roof of her mouth. She grinds crunchy food with the few molars that meet. Tough or sticky food – she doesn't even try.

As a plastic surgeon, Hopper sees the precise reasons, measured in millimeters, why Brenna's bones will never fit the conventional definition of beauty.

As a pediatric surgeon, he sees how Brenna's face makes her suffer.

Hopper lifts a small reciprocating saw from the sterile blue cloth and aims to fix all that.

Normal comes at a cost

Brenna's face grows on you quickly. Especially those wide-set blue eyes. Like a floppy-eared puppy, there's something absurdly adorable about her out-of-proportion features that grabs your heart and reels you in.

"Her cuteness is so different from anyone else," says Brenna's father, Erin Johnston. "We just fell in love with it."

But to a stranger's first glance, she just doesn't look right. That's yet another function of the face – the social function. The ability to make a first impression, a subtle emotional expression and, when it's time, an attraction.

"When you look at a face you do a T-shaped analysis – you go from the eyes down the nose to the lips and back



JOHN LOK / THE SEATTLE TIMES

TO FIX THE FUNCTIONS of Brenna's face, Dr. Richard Hopper will drastically alter its form. He will peel back her skin, saw through her cheekbones, eye orbits, nose and upper jaw, and wrench the bones forward. He warns her mom: "It's going to be a big shock for you. Her eyes will stay the same, but everything else will look different."

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up again,” Hopper says. “But if you notice one little thing wrong, you go to it automatically and stick on it.”

Brenna has been called “Alien Face” by a kid at McDonald’s, been pointed at on the playground and asked more times than she can count: “What’s wrong with you?” People who don’t know better assume she’s mentally retarded or speak to her like she’s deaf.

Now, to fix the functions of Brenna’s face, Hopper must also drastically alter its form. Not only will Brenna be able to breathe and chew, she will look more “normal.”

“In so many ways this surgery is her chance for a much more normal life,” Erin Johnston says.

But on the eve of surgery, Robyn Johnston is wistful. For all her daughter would gain, part of her would be sacrificed.

Hopper feels this, too: “There’s a bit of bittersweet satisfaction. I can make the child function better and be more accepted by society, but you lose something you won’t get back – a certain uniqueness.”

Deconstructing a face

At the level of bone, the face is an architectural marvel. Beams, ledges, arches and columns form and suspend the structures that protect the underside of the brain and allow us to breathe, see, chew and speak.

To give Brenna a face that functions properly, Hopper has to tear down and rebuild the frame.

His concentration sharpens as he prepares to sever Brenna’s facial bones from the rest of her skull. The OR goes silent. This is the riskiest part of the surgery. There’s a chance of blindness, stroke, even death.

Hopper works the serrated saw clean through the miniature cheekbones, which look like porcelain flying buttresses. He saws through the rims of the eye sockets and across the top of the nose, like he’s drawing opera glasses. An earthy smell – thick and organic – trails his blade.

He takes up a chisel and turns the edge into a blind corner around the backside of the facial bones, where concealed pillars fasten the facial plane to the base of the skull. By feel, he tap-tap-taps a path through the blocks of bone until the steady resistance gives way.

“It’s like working with marble, predicting which way it will crack, but even an expert can think it will break one way and it goes another,” Hopper says.

As he operates, the 37-year-old surgeon hears the voices of his teachers and colleagues coaching him on. His mentor from New York University, Dr. Joseph McCarthy, cautions: “Don’t force it.” The senior craniofacial surgeon at Children’s, Dr. Joseph Gruss, chides in his South African accent: “Remember, there’s a brain under there.”

A wayward crack at this point could work its way into the brain cavity and spill the cerebrospinal fluid that cushions the brain. A fissure could splinter vertically and split the face up the middle.

Hopper places the chisel at the root of Brenna’s nose, between her eyes. Her brain is directly below his edge. He



BRENNA IS A FLURRY OF NERVOUS ENERGY the night before surgery. She bounds off steps, bounces on couches and leaps over sidewalk cracks. This kind of play will be off-limits after tomorrow. She will spend her summer with her face caged in a titanium brace that will slowly pull her bones forward. Any bump or fall could send her back to the operating room.

presses through the nose and pries the attachments from the skull.

After about an hour of sawing and chiseling, the facial framework – from the eye orbits down to the upper jaw – is freed.

The surgeons tug the block of facial bone upward with forceps, stretching the muscles beneath to their limits.

A final, radical procedure

The traditional procedure for children with Crouzon Syndrome is to sever the facial bones and wrench them forward, as Hopper has just done. The gaps are then filled with bone from rib grafts.

This is the surgery Brenna’s parents have been bracing for since she was an infant.

But the severity of Brenna’s case requires her face to advance at least two centimeters – twice as far as the limits of traditional surgery – to unblock her airway and protect her eyes. That would mean at least two of these intensely invasive surgeries. And after all her previous surgeries, she has no rib bones to spare.

Hopper, who came to Children’s three years ago from New York University, introduced a more extreme procedure called midface distraction.

After splitting off the facial bones, Hopper will bolt and wire Brenna’s skull to an external device – an elaborate titanium halo – to slowly draw the face forward and stimulate the body to grow its own bone.

Some surgeons say the procedure is too torturous, the outcomes too unpredictable. Hopper, the only surgeon in the Northwest who performs this operation, believes the results are worth it. Brenna is his 14th patient.

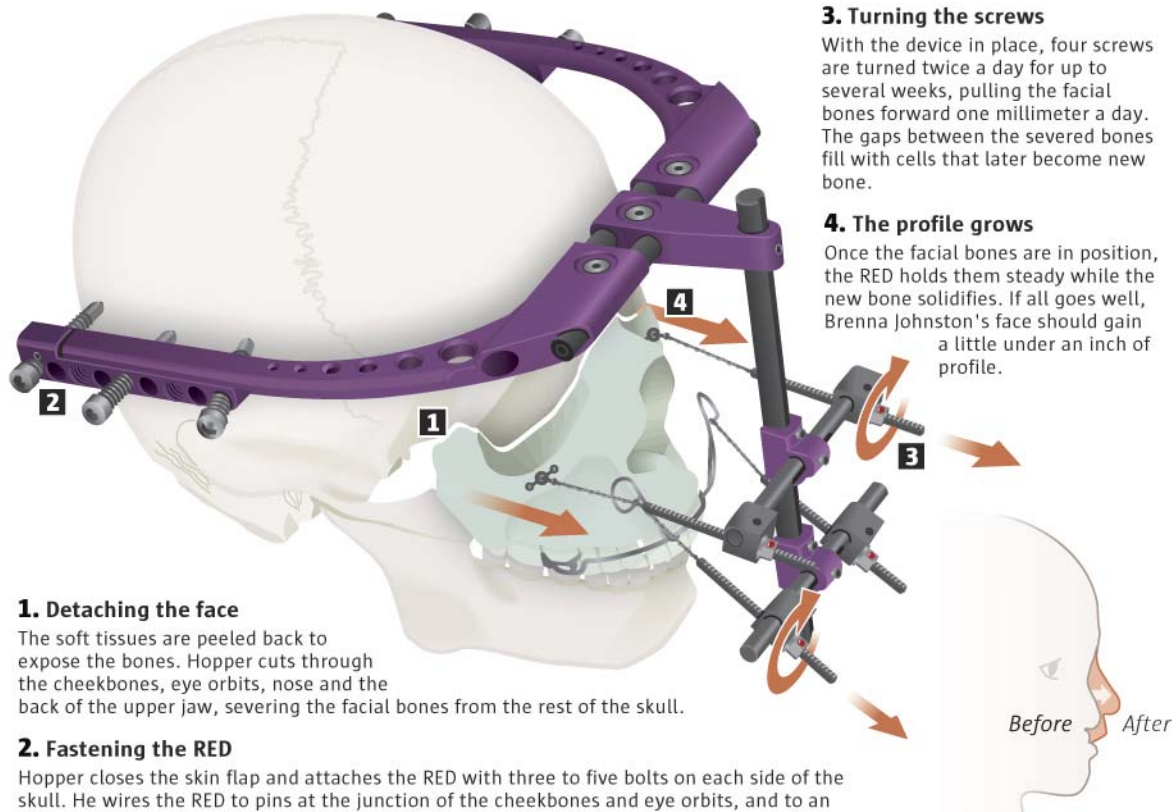
The device he will use is called a Rigid External Distraction system, or RED. Once he bolts it to Brenna’s head, she

Continued on next page >

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Building a new profile

Dr. Richard Hopper uses a device called the Rigid External Distraction system, or RED, to align the underdeveloped facial bones of children with severe Crouzon Syndrome. The device is attached during a lengthy surgery and must stay on for about two months. Here's how it works:



1. Detaching the face

The soft tissues are peeled back to expose the bones. Hopper cuts through the cheekbones, eye orbits, nose and the back of the upper jaw, severing the facial bones from the rest of the skull.

2. Fastening the RED

Hopper closes the skin flap and attaches the RED with three to five bolts on each side of the skull. He wires the RED to pins at the junction of the cheekbones and eye orbits, and to an acrylic mouthpiece bonded to the upper teeth.

3. Turning the screws

With the device in place, four screws are turned twice a day for up to several weeks, pulling the facial bones forward one millimeter a day. The gaps between the severed bones fill with cells that later become new bone.

4. The profile grows

Once the facial bones are in position, the RED holds them steady while the new bone solidifies. If all goes well, Brenna Johnston's face should gain a little under an inch of profile.

Source: Dr. Richard Hopper

KRISS CHAUMONT / THE SEATTLE TIMES

will have to live caged inside it for two months.

Robyn's first reaction to the RED was visceral: No way am I letting you put that thing on my kid.

But when Hopper said it could take care of Brenna's face all at once – that it could be her last major surgery – Robyn agreed.

Building the cage

Now Hopper's team pulls the flap of facial tissue up, smooths it over Brenna's skull and sews it back in place. Her severed facial bones are left floating beneath.

Hopper secures the titanium halo. He twists five sharp-tipped screws through Brenna's scalp on each side, in rows above her ears.

He suspends a vertical post and two horizontal beams down the center of her face. This graphite ladder forms the scaffolding he'll use to construct her new face.

If Hopper played the roles of carpenter and marble cutter during the first half of the operation, now he's an engineer, calculating proper vectors to suspend her bones.



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THE SURGERY IS OVER, but the swelling – and suffering – have just begun. With the titanium halo bolted to her skull and the scaffold wired to her cheeks and upper jaw, Brenna can't see, talk or lift her head for days. Robyn will turn screws on the front of the device twice a day to crank Brenna's severed facial bones forward a half-millimeter at a time.

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He threads wires from plates on her cheekbones to the upper beam of the scaffold. Pins stab through the fragile skin near the corners of her eyes.

He bonds an acrylic retainer to her upper teeth so the jaw can be wired to the lower beam.

Twice a day for the next few weeks, Robyn will turn four screws on the front of the scaffold to crank Brenna's facial bones forward, a half-millimeter at a time.

The constant tension will signal the body to fill in the gaps between the severed bones, a process doctors have harnessed but don't fully understand.

Once Brenna's airway opens, her orbits cradle her eyes and her teeth align, the brace will suspend her face until the new bone solidifies.

When Brenna starts third grade, she'll have a new face.

The long summer ahead

After nine hours in surgery, Hopper peels off his mask, gown and gloves. The hard part is over – for him.

Brenna is rolled out of the OR, trussed into the metal brace that encircles her face like a medieval cage.

If it works, she'll be able to breathe, chew and play soccer without fear of blindness.

If something goes wrong, if Brenna stumbles in the next few weeks and bumps the disjointed bones out of alignment, if she gets an infection, if her bones don't set the way Hopper predicts, she could be back in surgery within a year, starting over.

"For Brenna, this is just the beginning," Hopper says. "Some people call this a two-month-long surgery."

That's a long time when you're 8 and it's summer break.



TWO WEEKS AFTER SURGERY, Robyn and Brenna leave Seattle for their home in Veneta, Ore. They've made the six-hour drive to Children's Hospital dozens of times over the years, but this summer they qualified for free transport from Angel Flight, a nonprofit group that shuttles patients to medical care. They'll fly back to Seattle each Monday through the summer so Dr. Richard Hopper can check on Brenna's progress.

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