

MEDICINE

"We thank God for what she is, even if others say she's ugly. I tell her that she's the most beautiful girl in the world." Galby Maldonato

Cynthia is ready for surgery

PART ONE: An 11-year-old Montreal girl has a rare disorder that has left her with a too-small face and bulging, weeping eyes. Today, she's scheduled for an operation that will be the first of its kind in Quebec

CHARLIE FIDELMAN GAZETTE HEALTH REPORTER



PHOTOS: PIERRE OBENDRAUF THE GAZETTE
Cynthia, with her mother, Ondina Yolanda Duarte, and father, Galby Maldonato. The girl has Crouzon syndrome, a rare genetic disease.



Cynthia holds a photo album with her baby pictures.



The room Cynthia shares with her sister is pink, her favourite colour.



Cynthia and her best friend, Karianne Lavertue, eat lunch at school.



Dr. Mirko Gilardino demonstrates on a plastic skull the tightening technique Cynthia's parents will have to perform after her surgery.

On a recent evening, Galby Maldonato and his 11-year-old daughter sit side-by-side on the couch, looking through her baby photo album. They pause at the most memorable moments – like her first trip to the hospital.

Latin music taped during a Sunday church service fills the St. Michel duplex with joyous sounds.

Suddenly, her father asks her softly in Spanish: "Cynthia, are you sure you want to go through with this?"

Cynthia has a rare genetic disease known as Crouzon syndrome, which causes premature fusion of the bones in a baby's head. As a result, her face is too small and her eyes bulge from their sockets and weep under her glasses. Her bottom lip and lower jaw protrude, making her profile look like a crescent. She also has trouble breathing. Because of her mismatched mouth, she lisps in fluent Spanish and French.

Cynthia, who has had several operations already, has wanted cosmetic surgery ever since she was 8, when doctors first mentioned it could be an option.

The long, painful and complicated operation is to take place today – provided it isn't bumped by an emergency or cancelled because of a lack of beds in intensive care. The craniofacial procedure will be the first of its kind preformed in Quebec.

Surgeons are to shave her head, peel her face from her skull, remove her forehead to reveal the brain, then cut through the bones around the eyes and nose. A temporary metal brace resembling a halo will be attached from ear to ear and then doctors will put the pieces of Cynthia's face back in place.

Over the next few weeks, the halo will help stretch her collapsed mid-face forward.

Her face won't be perfect. She'll need surgery again in a few years to complete the realignment of her jaw. But it will improve her appearance drastically.

Cynthia's resolve doesn't waver at her father's question. She isn't nervous or scared. She straightens her pink top and nods: "Yes."

"I want my eyes to look normal. I want my teeth to come together. I want to look like my brother and sister."

Maldonato tickles his daughter. She flails at him with hands unable to close into fists while he covers in mock fear.

"To me, you are the most beautiful girl in the world," he says, covering her face with kisses.

An image capturing the family's first trip to the Montreal Children's Hospital is Maldonato's favourite in the album. It was taken at a bus stop, everyone smiling in their Sunday best. Cynthia, barely a month old, is a pink bundle in her mother's arms.

That's the day the Maldonatos learned she had Crouzon syndrome, a disorder seen in about one per 25,000 newborns. Treatment is a bone-lengthening procedure for limbs invented by Russian orthopedic surgeon Gavril A. Ilizarov, and then adapted for the face.

"It's just that we were so hopeful ...," Maldonato says, looking down at the smiling faces in the photo as his own eyes fill with tears.

Maldonato, a mechanic, and his wife, Ondina Yolanda Duarte, came to Montreal from a small village in Honduras in 1994 with their first-born, Stacy, now 15. Kevin, 14, was born a year later. Both are healthy.

But a scan during the couple's third pregnancy showed something wrong with how the baby's head was growing. It was shaped like a strawberry. Maldonato says that as parents, they blamed themselves, wondering

whether they had done something wrong to cause the malformation. Doctors said the baby would die or be handicapped. They strongly recommended an abortion.

But Cynthia's parents refused. They saw her as a blessing. It would be hard, they knew, but they would take it one day at a time. And she would know that she is loved.

By age 2, Cynthia wore hearing aids and leg braces. As she learned to walk, her parents spread blankets on the floor to soften her falls.

"We thank God for what she is, even if others say she's ugly," Maldonato says. "I tell her that she's the most beautiful girl in the world – that I love her most."

Duarte pipes in, looking at Cynthia: "No, I love you more."



"I want my eyes to look normal. I want my teeth to come together. I want to look like my brother and sister."

Cynthia's album has pictures of her at 4½ months, then again at 10 months, her head shaved for the neurosurgery she had to undergo to give her brain space to grow.

Cynthia, who is no stranger to hospitals, packed her purple knapsack last week with snacks and diversions – cartoons, drawing materials, her father's iPod and her mom's cake squares, plenty of tissue to blot her eyes – in preparation for meetings with various Montreal Children's Hospital officials about the coming surgery. For the occasion, she wore black pumps and a short jacket.

At the hospital, the Maldonatos sit in room C738 with three families, one in each corner, each living a personal pre-surgery drama behind individual curtains.

Cynthia's mother will of course be sleeping by her side in the same hospital room, clinical nurse Hélène Rainville tells Cynthia kindly.

"She will be with you all the time. But not in ICU. She can be with you all day, but not at night, because you will have a nurse here just for you."

As for going back home, that's weeks ahead, and "only when you can do everything that you are doing today," she says.

"Can I bring something?" Cynthia asks.

"What do you like to do? Books, music, DVDs – bring any favourite activities in your luggage," Rainville says.

One last bit of advice: Enjoy being at home this weekend, and make sure to ask for your favourite dish Sunday night.

Cynthia skips to her mother's side and whispers, "lasagna."

Cynthia loves to eat, her mother confides. "And she loves to cook. Cynthia is the one that helps me most in the kitchen," Duarte says.

Sunday's meal will be the last solid food for Cynthia for many days.

"Any other questions?" Cynthia shakes her head.

"Then it's time for the worst part," Rainville says, and gets a needle to draw six units of blood.

She pokes unsuccessfully for a vein in one thin arm, then in the next. Change of plan, Rainville announces. "I see a beautiful vein here. Are you right-handed or left?"

"Right," Cynthia says with a delicate wave of the right. She doesn't flinch as the needle goes into her left

hand. She fixes her gaze on the syringe. It fills with red.

Neurosurgeon Jose Montes pokes his head behind the curtain: "Good day," he says in Spanish.

Montes, who performed Cynthia's earlier surgeries, will be part of the team in Monday's procedure. "¿Alguna pregunta?" he asks her. Any questions?

Cynthia's treasured pictures in her album include one of her as a smiling baby tucked by her sleeping father – the family's typical Saturday morning routine in bed.

Another is more recent – Cynthia in a gorgeous pink taffeta dress at her sister's coming-of-age 15th birthday celebration, the ultra important Quinceañera, or Latin version of a Sweet 16.

The girls share a room painted in bubble gum pink and sleep together in the lower bunk bed. Except for the stuffed dogs and bears, it is decked in pink things, including a Barbie bedspread and princess figurines from Stacy's party.

"It's my favourite colour," says Cynthia, who sneaks her older sister's clothes and yearns for her waist-length hair.

"Every morning, she asks me if it's grown a bit," says Stacy, who brushes Cynthia's dark shoulder-length curls into a high ponytail.

"And she goes through a bottle of gel every two days," Cynthia says.

"Oh, that's not true," Stacy shrieks. Cynthia shows off one of Stacy's figurines that she helped make: "I got sparkles glued all over my fingers," she says.

Then, Cynthia confides that she's eager for the surgery because she wants to look good for her own 15th birthday celebration.

The family's last stop at the hospital is at the 11th floor at the plastic surgeon's office.

Leaning on his desk, Dr. Mirko Gilardino holds a T-shaped brace that he will affix to Cynthia's head during the surgery. It looks like a metal halo with screws, something from a science-fiction movie.

Cynthia turns it over in her hands. It will work similarly to braces on teeth to slowly straighten her face, one millimetre a day, he explains.

Once Cynthia is out of the ICU, her parents will turn screws in the brace – one millimetre a day – with a half turn in the morning and a half at night.

The "active" period, when the bones will move, is to last three weeks, but Cynthia will have to wear the brace for at least three months, Gilardino says after illustrating bone movement on a plastic and plaster model of a skull.

He then turns to Cynthia. "Do you want me to shave your whole head?"

Her "No" is barely audible. The shake of the head is unmistakable.

"I didn't think so. I'll take as little as possible," he promises gently. Cynthia wipes her tearing eyes. She's hoping the surgery will fix her respiratory and eye problems. She says she's tired of strangers asking her why she is crying.

The day is almost over. Cynthia looks expectantly at her mother.

"She loves to go shopping," Duarte explains.

Maldonato jokingly chides Cynthia for taking advantage of the situation, and then covers her face with kisses.

"You are the most beautiful girl in the world."

She wiggles away: "He always says that."

The Gazette will follow Cynthia's journey. Health Reporter Charlie Fidelman will report on the surgery, and also chronicle Cynthia's recovery.

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CYNTHIA'S STORY: PART 2

WITH SAWS, HAMMERS and other tools that sound like they belong to a carpenter, surgeons work to change a brave girl's life

'Okay, you can close her up'

CHARLIE FIDELMAN GAZETTE HEALTH REPORTER

On the eve of a big operation, Montreal plastic surgeon Mirko Gilardino likes to relax while playing Chopin's Nocturnes on the piano.

Before a major procedure, neurosurgeon Jose Montes visualizes the cuts he will make into the skull. At breakfast that day, he skips coffee.

The day before the cosmetic surgery that will change her life, Cynthia Maldonado, 11, sings at a special church service in front of 300 people who came to support her. For supper, she eats a hamburger.

Cynthia has a rare genetic disorder known as Crouzon syndrome, which is characterized by a premature fusion of the head bones of a developing child. It affects about one in 25,000 newborns.

The condition has left her with a face that looks too small, as if it has collapsed inward. She also has problems breathing, a protruding lower jaw and bulging, weeping eyes.

Cynthia, who lives in St. Michel, has waited nearly four years for the corrective surgery that will make her look more like her brother and sister.

On Monday, Cynthia finally got her wish for a new face, thanks to Gilardino, Montes, and their medical team at the Montreal Children's Hospital, who performed a complicated craniofacial procedure that was the first of its kind in Quebec.

"She's very brave," Cynthia's mother, Ondina Yolanda Duarte, said that morning while waiting in 10th-floor surgery antechamber at the Children's. At that point, the family was praying for a free bed in intensive care because, without a bed, the surgery would have been cancelled.

When Gilardino appeared in scrubs and a big smile at 8:45 a.m., they knew their prayers had been answered.

As nurses led Cynthia away, she gave her mother a hug: "Don't cry, Mama."

Gilardino, an only child, loved taking his toys apart. He always wanted to be a surgeon. The 34-year-old is now director of craniofacial surgery at the Children's, a McGill University Health Centre teaching hospital.

To greet patients, he wears tailored suits. For surgery, he's barefoot in Birkenstock clogs and wears green scrubs over a silver medallion necklace etched with his mother's initials. For his polished good looks, he is known as the Children's own George Clooney.

Montes, who is originally from Mexico, is the Children's chief neurosurgeon. He has a special relationship with Cynthia's parents, who immigrated from a small village in Honduras in 1994. He's known them since he operated on Cynthia when she was 4½ months old, then again when she was 10 months, to give her brain space to grow.

Montes always speaks to the Maldonados in Spanish, and they consider him a friend.

The Gazette was there as these two doctors led the surgery that aims to fulfill Cynthia's biggest wish. The team spent hours peeling her face from her skull, breaking the bones underneath, then attaching a metal brace to pull the little girl's collapsed face forward. Here is the story from the operating room.

10 a.m. Intubated and draped in a flannel sheet in OR 2, Cynthia has been under anesthesia for an hour. Her head is cradled on a doughnut pillow.

After several attempts to insert a thin catheter into an artery on Cynthia's right wrist, anesthesiologist Natalie Buu tries her left arm. She probes, straightens her shoulders and probes again. The catheter line will monitor blood pressure and oxygen levels during the surgery.

"Okay I got it," she says, relieved.

"Can we move the table just two inches?" Gilardino asks as nurses perform a pre-op inventory: blades and drill bits in trays, blood units in the fridge, and the right patient on the table.

Computer screens mounted on the wall show scans of Cynthia's head.

Watch out for the soft spots, Gilardino cautions his residents as they inject Cynthia's head with droplets of epinephrine that will



PHOTOS: PIERRE OBENDRAUF THE GAZETTE

Operation is a go, Dr. Mirko Gilardino indicates to 11-year-old Cynthia Maldonado as her mother, Yolanda Duarte, looks on. Cynthia had been waiting for nearly four years for surgery for Crouzon syndrome, a rare genetic disorder. On Monday, Cynthia finally got her wish.



Gilardino checks the halo secured to Cynthia's face during her operation at the Montreal Children's Hospital on Monday.

help reduce bleeding.

Gilardino asks for the shears. He clips Cynthia's dark, curly hair below the crown at the back of the head — over a scar of an earlier procedure — and clears a thin strip, fulfilling a promise he made to her to shave as little as possible. He braids the rest of her hair into strands; one strand dangles off the gurney.

A nurse mops the floor. Wearing magnifying glasses similar to a jeweller's, Gilardino washes to the elbows, dons a gown over the scrubs and slips each hand into two gloves.

11 a.m. Gilardino's knife penetrates as he makes the first cut. He carves along a blue line drawn on Cynthia's right ear to the left where the scalp was shaved.

Cynthia's head splits. Blood drips from the braid to the floor.

Gilardino separates the scalp from the bone, and the skull emerges from the crown to the temple and then the forehead toward the eyes.

Gilardino reaches the eye sockets and scrapes gently: "You don't want to rip the whole muscle. It will make her (Cynthia's) life easier," he tells residents Oren Tessler, Ronnie Borsuk and Hassan Alajmi.

11:35 a.m. Exposed to the top of the nose, the skull looks like a lunar landscape seeping blood. It's time for the neurosurgeon to take over.

"I have 15 minutes to get something to eat," Gilardino says, and slips out of the OR.

11:45 a.m. Montes bores holes at each temple with a manual drill. He makes a few burrows then takes a lightweight, nitrogen-powered surgical saw to connect the holes in the skull like someone connecting the dots.

It whines and smokes, throwing fine bone powder where it pierces the head. The air smells burnt.

Abruptly, the high-pitched whine stops. "I lost power," Montes says, surprised. "Hold on, Dr. Montes, we need to lower the pressure," replies a surgical nurse.

"It was working great," he says.

Once power is restored, Montes traces the same trajectory he made when Cynthia was a baby. He then moves into new territory, cutting the forehead bones right above the eyes. He frees a piece of parchment-thin bone of skull from the uppermost layer of matter that separates the brain from the cranium. He wraps her crown in gauze.

11:55 a.m. Cynthia's brain is visible. It moves and pulsates with every heartbeat.

Montes is done. He's going to another OR to remove a brain tumour from another patient.

"Call Dr. Gilardino if there's anything," he says. "I'm downstairs."

12:10 p.m. Gilardino is back, wearing a fresh gown and gloves.

"How are we doing? No issues?" Gilardino asks Buu.

Nurses count the discarded blood-soaked gauze strips to make sure none is left in the patient.

12:15 p.m. "Okay, let's do it," Gilardino says before taking a hammer, chisel and saw to Cynthia's face. He tells his three residents that their No. 1 job while he is breaking the bones is to protect Cynthia's brain.

Four sets of arms hover where Cynthia's brain meets the roof of her eyes. Tessler has the mallet, Alajmi has suction and Borsuk holds a metal depressor to move the brain aside just a few millimetres

from the skull.

The sharp surgical pick is in Gilardino's hand. He positions the sharp end and nods to Oren to start tapping the top of the pick with the mallet.

Tap tap. Tap tap. Tap tap. "Keep going," Gilardino tells Oren.

Tap tap. "Yeah." Tap Tap. "Yeah." The suction machine gurgles over the hiss of the respirator. Blood drips down Cynthia's braid. It splatters Gilardino's Birkenstocks.

12:45 p.m. "We're going to be bleeding here Dr. Buu."

"We're good," replies Buu, who is monitoring Cynthia's vitals.

Gilardino fractures the bones first around Cynthia's eyes, then around her nose. He drives a pick into each cheek above the jaw.

Tap tap. Tap tap. Tap tap. He then tests her face to see if it broke. Each of Gilardino's hands is wrapped around a set of pliers, or surgical forceps. He directs both into Cynthia's face. Pincers enter into each nostril and through the mouth.

Gilardino rocks the pliers back and forth, side to side. "Careful not to break the palate," he says.

But Cynthia's face doesn't give.

12:55 p.m. Tap tap. Tap tap. Tap tap. This time, it is Gilardino on the hammer. It's still not breaking.

"Hold her head down for me," Gilardino says, and leans his weight on the pliers.

Craack!

12:59 p.m. "Okay. We're going to put her face back on."

1:05 p.m. Gilardino puts in temporary sutures by each temple: "The face has a tendency to fall."

1:50 p.m. Gilardino finishes attaching tiny metal plates, two above the nose and two below, which will anchor the halo-shaped apparatus that will stretch Cynthia's face forward. He steps into a pool of blood.

He puts the skull back in place, pulls the scalp over the skull like he's fitting a wig and turns it over to his residents: "Okay, you can close her up."

Oren and Ronnie take up needle and suture thread. There's a lot to sew.

Gilardino sits for the first time.

2:45 p.m. Gilardino asks for the halo-shaped device. He tightens it over Cynthia's temples and then screws the nose bolts in.

The key, Gilardino says, is to not create any space in the sinus cavity for bacteria to grow. That's why the broken bones are put back in exactly the same spot. The halo will realign the mid-section of her face like braces straightening teeth.

3:05 p.m. There are enough discarded bloody gauzes to fill nine bags. Cynthia has lost a litre of blood.

Cynthia's parents have been waiting on the 7th floor since 9 a.m. They held hands and prayed. No one ate.

At 4 p.m. Gilardino sits down on a couch opposite them and tells them that everything went very well, better than expected.

Their tears fall after Gilardino leaves the room.

The next night, Cynthia is able to see with both eyes and move her hands and feet. Dressed in a charcoal suit, pale blue shirt and silver tie, Gilardino pops into the ICU to test her reflexes.

"Comment ça va, ma belle? Hello, Beautiful. Can you squeeze your fingers?" Cynthia obliges. Gilardino's hand curls into the universal OK sign. Perfect.

Cynthia should go home Monday. Her recovery time will be considerable. She will wear the halo for three months.

Gazette Health Reporter Charlie Fidelman will be checking in on Cynthia's recovery. Our third instalment will tell readers about her life with a new face.

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UNDERSTANDING CYNTHIA'S SURGERY

Check out our interactive graphic explaining each stage of Cynthia's remarkable surgery. Only at montrealgazette.com



SCENES FROM CYNTHIA'S LIFE, AND THE O.R.

Get to know Cynthia, her family and friends in a video by The Gazette's Pierre Obendrauf. We also take you inside the operating room for her surgery. Warning: graphic content. montrealgazette.com