

## **The ups and downs of Erek's world**

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**By Aaron Derfel**

*Children's hospital is home to 10-year-old boy born with a severe form of rare genetic disorder*

*Therapeutic clown Dr. L'Air de Rien (Olivier-Hugues Terreault) nuzzles 10-year-old Erek Cokay, who has myotubular myopathy, at the Children's Hospital.*

*Credit: Gordon Beck, The Gazette*



Erek Cokay sat in a wheelchair next to his bed on a sunny spring morning, eagerly waiting to be taken to school. It was 7 o'clock, and already Erek had been dressed in some of his favourite clothes, including a purple Raptors basketball sweatshirt.

His face was freshly washed, his cheeks turning pink as the sun rose beyond the window of his bedroom. His thick eyelashes fluttered from behind his glasses.

A compact ventilator machine was helping 10-year-old Erek breathe. The machine makes a whooshing sound 24 hours a day. It was hooked up to the back of the wheelchair.

An oxygen tube ran from the machine to a hole - a tracheostomy - that a doctor had cut in his throat when Erek was a baby.

Erek stared at the stuffed animals lining a shelf in his room. On one of the walls hung a team portrait of the Canadiens.

The room is on the sixth floor of the Montreal Children's Hospital, Erek's home since he was one. He's one of four children living in the acute-care hospital because there is no other place - no other home - where there are respiratory therapists, doctors and nurses who can take care of their complex medical needs round-the-clock.

"Good morning, Erek," said Sharlene Olscamp, an aide with the Victoria Order of Nurses, as she entered his room.

His eyelashes fluttered, and his lips trembled. That's his way of saying "hello" or "yes," because he finds it hard to speak.

For almost two years, Olscamp has been accompanying Erek to the Mackay Centre, monitoring him during his lessons there, and returning to the Children's with him after class is over. The hospital staff call her a shadow. Olscamp prefers to call herself Erek's second mother.

Ereĸ's mother, Lucy Cokay, was off work that morning, fighting a cold. She was planning to visit her son in the afternoon.

"I'm going to have to suction you before we go to school," Olscamp told Ereĸ.

She removed the tube from his throat and, within a few seconds, the ventilator machine sounded an alarm.

Beep! Beep! Beep!

"It's OK, Ereĸ," she said soothingly.

She slipped on a pair of latex gloves, grabbed a thin, sterile instrument and stuck it in Ereĸ's trachea. She swivelled it deftly inside, suctioning all the saliva and mucus that had accumulated in his windpipe.

Ereĸ's face turned red. Olscamp placed the tube back in his trachea, and squeezed an air bag to ventilate him a little before she reconnected him to the machine.

Ereĸ was born with a severe form of a rare genetic disorder called myotubular myopathy. It causes muscle weakness and wasting.

Some children with myopathy can walk. But in Ereĸ's case, the myopathy has slowly robbed him of what meagre muscle co-ordination he once enjoyed.

Three years ago, he could sing lines from nursery rhymes, but that all changed after a bout of pneumonia. Today, he can barely utter a single word, partly because the ventilation tube obstructs his vocal cords.

He used to wield some strength in his arms.

Today, he strains to raise his right arm, while the left is limp. Hospital attendants have to dress and undress him, bathe him, lift him out of bed and strap him in his wheelchair.

It was time to go to school. They had to be quick or they would miss the bus. Olscamp walked ahead of Ereĸ.

"Come on, Ereĸ. Let's go."

Ereĸ, until that moment, trapped and frozen in his myopathy, sprung into action. With his right hand, he grasped a joystick on a tray before him. The electric motorized wheelchair rolled forward.

Ereĸ eyed the hospital corridor, calculating the angles he needed to manoeuvre the wheelchair, and he drove it gracefully with the greatest precision. It moved in short spurts like a cat, the animal Ereĸ adores.

He paused at the nurses' station.

"You have fun at school today," said one of the veteran nurses, Corinne Soden, smiling at him.

Olscamp and Ereĸ reached the elevators, but a maintenance crew had shut down three of the four for repairs. Olscamp shook her head in frustration, fearing they might miss the bus. Five minutes later, an elevator light came on.

When they arrived at the hospital lobby at 7:30, there was no bus outside. Olscamp huffed dejectedly. They waited for a half hour before she marched over to the reception desk to phone the Mackay Centre.

They ended up missing the bus by a couple of minutes. Ereĸ watched hospital workers shuffle inside through a revolving door. He had missed the bus in the past, and each time he was forced to return to his room.

Tears trickled down his cheeks.

"I need to get Ereĸ to school!" she pleaded over the phone.

Olscamp, a hefty woman who has the manner of a drill sergeant except for the tenderness she reserves for Ereĸ, finally persuaded somebody to send over a bus.

"The handicapped always have to fight," she said as they boarded the yellow bus that Tuesday morning.

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Shortly before Ereĸ turned 1, doctors in London, Ont. - where he was born, prematurely - suggested to his mother that they wean him off the respirator. Without it, everyone knew Ereĸ would surely die.

"They wanted me to let him go," Lucy Cokay recounted. "I couldn't do it. I couldn't let him go. I just couldn't."

Cokay had already endured the death of her firstborn, Antonio, who probably had myopathy as well. Antonio died of internal hemorrhaging at the age of 16 months.

She had also been widowed. Her husband, Sami Cokay, died of lymphoma when Ereĸ was only three months old.

"I just couldn't handle another loss," she explained in a soft, quavering voice.

Muscle biopsies confirmed Ereğ's myopathy. There was no family history of the disease, but a genetic test - which was unavailable at the time - would later reveal that the mother carried the genetic abnormality on the X chromosome.

That means that only boys can be affected. Cokay gave birth before Ereğ to a healthy girl, Yasmin, now 12 years old.

Cokay decided to move with her children to Montreal to stay with her mother. She tried looking after Ereğ at home, but the task soon proved insurmountable. She was often up all night, tending to his needs while raising Yasmin at the same time.

In 1994, Ereğ was permanently admitted to the Children's. Dr. Stephen Liben was put in charge of him, administering antibiotics whenever Ereğ sweated with pneumonia in the ICU.

There is no cure or treatment for myopathy. The best doctors can do is treat symptoms like stomach acid or give him "artificial tears" at night because his eyes tend to stay open.

"Being handicapped doesn't equal suffering necessarily," Liben said. "Being very handicapped, you can enjoy the pillow on the back of your head, and the feeling of sheets and the feeling of sunshine. With enough support and help, you make the most of it."

Yet it's a struggle for Ereğ's mother to communicate with him. He keeps a red binder with pages of pictograms which he can point at to indicate whether he likes something, or he's cold, among many other everyday things.

"Sometimes he does say 'yes' or 'no', and you're still shaking your head, wondering whether what he said is the right answer," she confided.

Myopathy usually doesn't hinder intelligence. However, studies have shown that institutionalized children do develop more slowly.

Ereğ's cognitive skills were tested and he scored low. The tests, though, were biased in favour of children who could answer questions by moving their hands - something Ereğ found difficult to do.

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At the Mackay Centre, Olscamp changed Ereğ's diapers before class. Lying down, he gazed at a couple of photos of kittens that were taped to the wall, his dark brown eyes flitting from one to the other, the fingers on his right hand caressing the paper.

The Mackay Centre, founded in 1870 as a school for the deaf, has expanded its mission to teaching more than 160 children with a wide range of physical disabilities. It has won praise for a program of "reverse integration," allowing 28 children who are not disabled to attend class.

Ereĸ shares his Grade 2 augmentative communication class with five other children, mostly those with cerebral palsy. Ereĸ is the most disabled, and that's why he requires Olscamp's assistance.

"I'm glad you were able to show up today," said Ereĸ's teacher, Sarah Knitter, as he wheeled himself into the bright classroom.

"Ereĸ, why don't you come up here and do some writing?"

Ereĸ approached the front of a "smart board," a large computer screen. A boy was sitting in a wheelchair next to him. Knitter handed Ereĸ a specialized mouse.

The screen displayed several categories of words: subjects, verbs and objects. By moving the cursor on the screen with the mouse, a pupil can click on the various words to write a sentence.

Under the first category were the following names: Lea, Leigh, Leroy and Lucy. Knitter instructed Ereĸ to write a sentence.

His right hand took hold of the joystick on the mouse. Slowly, methodically, he slid the cursor across the screen in a perfectly horizontal line above the first category. He paused. Again, slowly, he pushed the cursor down in a perfectly vertical line over the list of names.

He stopped when he reached Lucy - his mother's name. Slowly, he edged the cursor into the very centre of the name and clicked.

He repeated this precise pattern for the other categories of words. This was his sentence:

"Lucy lays on the light."

"Very good!" Knitter said. "We'll be able to print that later."

The students gathered in a semicircle, and sang songs. When Ereĸ's turn came, he pressed on the button of a voice box that sang for him.

During snack time, the teachers distributed slices of cantaloupe, grapes and cheese crackers. None of the food went to Ereĸ because he can't swallow.

When he was an infant, doctors installed a gastrostomy tube in his stomach. Four times a day, a bag filled with milky nutrients drips down a line, feeding him.

His mother does let him taste things, though. She likes to dip a finger in her coffee and touch the tip of his tongue. EreK, she said, is a "coffee fiend."

As EreK watched the children eat and laugh, one of the teachers informed Olscamp in front of him that he couldn't attend a field trip the next day. EreK's mother couldn't take time off from her job as a lab technician at St. Mary's Hospital to accompany him.

For field trips, EreK must be escorted by two people. He would have to stay in the hospital the next day, missing the outing to the petting zoo.

Tears streamed down his cheeks. No one seemed to notice but Olscamp. She rubbed them away, and whispered in his ear that everything would be okay.

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After his morning lessons, EreK returned to the hospital. When he wheeled himself into his room, his mother and grandmother, Rita Zinno, were already there waiting for him.

"Hi, EreK!" said his mother, a youthful-looking woman of 46. She flashed a big smile, bending down to kiss him on the cheek.

EreK's face brightened. For the first time that day, a faint smile appeared on his face.

Olscamp turned on EreK's computer in the corner of the room before leaving. He played a game on his Mac, clicking on letters that matched each other. Again, he directed the cursor to the very centre of each letter.

"Wow!" his mother said. "Good job. You did it. Just like your father - a perfectionist."

He continued playing until his head flopped forward. He was growing fatigued, and needed to rest in bed.

During the afternoon, two therapeutic clowns - Dr. Giggle and Dr. L'Air de Rien - stomped into his room, making a ruckus.

"EreK, give me a kiss," Dr. L'Air de Rien asked, craning his face toward the boy's mouth.

EreK's lips touched the clown's red nose.

"Ahhh!" Dr. L'Air de Rien exclaimed joyously.

EreK was being paid a lot of visits that afternoon. Pascal Comeau, the music therapist, dropped by to play Zip-A-Dee-Doo-Dah and John Lennon's Imagine.

As Comeau strummed the strings of a guitar, Cokay talked with a social worker about Ereğ's health.

"He's had a great year so far," the mother said, noting Ereğ hadn't come down with pneumonia.

Like most children with myopathy, Ereğ is at heightened risk of respiratory infections. To protect him from catching other children's germs, the hospital assigned him a private room.

Comeau handed Ereğ a music stick and let him slide it across the keys of a glockenspiel. Next, Ereğ clanged within a metal triangle, but with each instrument, his right arm grew weaker.

Cokay watched her son. She, too, was tired because she hadn't slept well the night before. She rubbed her eyes and nose.

"I think you've had a long day," Comeau told Ereğ, letting him rest. His eyelids started drooping.

Cokay readjusted his legs under a blanket to make him more comfortable. When the ventilator sounded a false alarm, she scrambled to the machine to check it.

Soden, the nurse, strolled into the room to suction him, and could tell right away that Cokay's cold had gotten worse.

"You need to go home and have a cup of tea," Soden advised her.

It was 4:30. The grandmother leaned over to tuck the blanket around Ereğ.

Mother and grandmother then turned to leave.

"Bye, Ereğ," she said, waving at him. "See you Friday. Oh well. Let's go home, Ma."

From behind his glasses, Ereğ's eyelashes fluttered.

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