

Chez nous

MCH EMPLOYEE NEWSLETTER

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A balancing act: inborn errors of metabolism

—page 2

ALSO IN THIS ISSUE:

ED simulation for better
patient care — Page 6

Celebrating Social Work Month
— Page 9

PFCC Star of the Month,
Eloisa Binder — Page 10

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A day in the life of a...nutritionist!



Chez nous learns about inborn errors of metabolism from the only nutritionist at the MUHC who treats them

By Stephanie Tsirgiotis



► Seven-month-old Florence Corleto was born with maple syrup urine disease, a type of inborn error of metabolism. Her mother, also named Florence (left), always checks new food items with Marie Lefrançois (right) to make sure they're safe for her daughter to eat.

Marie Lefrançois thought about quitting nutrition twice before finding her niche: treating patients with inborn errors of metabolism (IEM). Over the years, her field has grown tremendously and she now treats over 100 pediatric and adult patients a year. The term 'inborn error of metabolism' refers to a series of rare genetic diseases where a child's enzyme activity is malfunctioning and therefore leads to the accumulation of toxic products in their blood, like urea or ammonia. The buildup of these products occurs when certain foods are digested and can lead to serious health problems and developmental issues. And the main treatment is a specialized diet.

THE MISSING ENZYME

In the early 1970s, Quebec started screening for a few types of these diseases at birth, including the most common one, phenylketonuria (PKU). "We can now identify four of these diseases from a heel prick soon after birth, but we're working on increasing that number," says Marie. The common thread between all these diseases is that in every case there is a missing or dysfunctional enzyme. For PKU patients, this missing enzyme can lead to the harmful buildup of phenylalanine in the blood, which left untreated, can lead to irreversible brain damage. "Without proper treatment, adults can be left with the mental capacity of a six-month-old," she says.

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Editor: Stephanie Tsirgiotis
Contributors: Maureen McCarthy
Sandra Sciangula
Graphic design: Vincenzo Comm Design inc.
Photography: Owen Egan
Dr. Lily Nguyen
Sandra Sciangula
Stephanie Tsirgiotis
French translation: Joanne Lavallée

To submit story ideas or texts to *Chez nous*, contact the Public Relations and Communications office at ext. 24307 or send an email to mchpr@muhc.mcgill.ca.

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If one of these diseases is detected at birth, the child is immediately put on a specialized diet. Working with these patients, Marie has to either eliminate a type of food completely or reduce it in order to avoid the accumulation of toxic waste in the body. "Certain foods lead to the production of certain toxic products, which is why we need to treat these patients quickly," she says. "I see newborn patients weekly for the first six months and continue to meet with them regularly into adulthood." Depending on the condition, Marie will either prescribe a tailor-made formula or will give instructions to the mother on how much breastmilk the baby can drink.

Glycogen storage disease type 1 is another type of inborn error of metabolism and is caused by the buildup of glycogen, a storage form of glucose in the body's liver and muscle cells. In order to reduce glycogen in the body, the patient has to eliminate dairy products (galactose), fruits (fructose), table sugar (sucrose) and sorbitol from their diet. "They also need to wake up in the middle of the night to eat corn starch; otherwise their blood sugar will dip dangerously low and it could even be fatal," explains Marie. "So cornstarch, a simple cooking ingredient, becomes a medication."

A BALANCING ACT

Prescribing the perfect balance of nutrients is also challenging, because every patient is completely unique. "Not only do I have to make sure the patient reduces the amount of toxic waste in their system, but I also have to make sure that they continue to grow and develop properly," she says. If the condition is



► Marie is the only nutritionist at the MUHC who treats patients with inborn errors of metabolism. *continued >>*

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On the cover: Seven-month-old Florence Corleto and her mother.

Cover photo: Owen Egan

A day in the life... (cont'd)

diagnosed early on and the diet is strictly followed, patients can go on to live normal lives, but the buildup of waste can happen quickly if the diet is not respected or when the patient gets sick from a simple cold or gastroenteritis. In some cases, the buildup is so dangerous that a child needs to undergo dialysis in order to remove the toxic products rapidly from their body before starting the diet.

Every time Marie sees a patient, she runs a series of blood tests to analyze the child's nutritional status and then compares the results to the previous visit to make sure there are no spikes or drops in the patient's amino acid levels. "If I notice a huge variation it either means that I haven't prescribed enough of something and I need to adjust their diet, or the child is not following the diet," says Marie. "In a lot of cases, you can't visually tell if a child is not following my recommendations. All the answers are in the blood and the diet needs to be followed for life."



► Marie sees newborn patients weekly for the first six months and continues to meet with them regularly into adulthood.

Once a child moves onto solid foods, Marie prescribes specialized food products, like low-protein pasta. These products are not easily accessible and need to be ordered through *Le programme alimentaire Québécois pour le traitement de maladies métaboliques héréditaires*, which is run out of the Montreal Children's Hospital. "We run the program for

the entire province," she says. In total, there are five centres in Quebec which treat these types of patients, but only 10 to 12 nutritionists with this specialized training, and Marie is the only one at the McGill University Health Centre.

THE RIGHT PLACE

One of these patients is seven-month-old Florence Corleto, affectionately nicknamed "Florencita" by the Genetics department. Her mother, also named Florence, was traveling back and forth between El Salvador and Montreal for training in the retail industry, when she was stopped at the airport because airline staff believed she was too pregnant to fly home at 34 weeks. "I felt fine, but they wanted me to get a doctor's note to clear me for travel," she says. After meeting with a doctor, they confirmed that her daughter was breeched and she needed to have a c-section. "My plan was to deliver and go back home immediately, but it didn't work out that way," recalls Florence.

At six days old, Florencita was barely eating and she was sleeping 21 hours a day. Florence brought her to the Emergency department at the Children's where numerous tests were done, including a lumbar puncture to rule out meningitis. "At first they thought she was dehydrated because she was sleeping so much, but she didn't become active even after they rehydrated her," she says. The team also noticed extremely high leucine levels in her body.

The Genetics department was called in and they diagnosed Florencita with maple syrup urine disease. The disease is categorized by the toxic buildup of an amino acid, known as leucine, in the bloodstream. "In those cases this buildup of leucine ends up causing the urine to smell like maple syrup, hence the name," explains Marie. If the disease is caught late, children are left with many serious health issues, including severe brain damage; in some cases, it can cause death.

Florencita was immediately put on hemodialysis to flush the leucine from her body and she was admitted to the Pediatric Intensive Care Unit. "Her levels were so high they were

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worried she was going to go into a coma or die,” says her mother. Doctors and nurses surveyed her closely and monitored her kidneys, amino acid levels and development daily. Marie also prescribed her a special formula, plus additional supplements to cover all her nutritional needs. As she grows, Florencita will continue to be limited in terms of what she can eat. She won’t be able to consume high-quality protein foods or dairy products, because her body can’t process or digest protein. “She will have to become a strict vegan, but we’ll still have to keep track of all the leucine she consumes, which is common in foods like beans and corn,” says Florence.

On top of dealing with her daughter’s illness, Florence is also dealing with another horrible ordeal: she can’t go home. “There are no specialists in El Salvador who deal with this disease. There are also no labs that can run her bloodwork and her special formula is not available in my country,” she says. “I am completely alone here in Canada, but if we go back home right now, my daughter could die.” Marie is now looking into other healthcare options closer to their home, in either Costa Rica or Mexico City.



► Lina Moisan, a nurse in Medical Genetics, measures Florence at every visit.

“Every day I think about how blessed we are,” says Florence. “My daughter wouldn’t be alive today without everything the hospital team and this province has done for us. How ironic that my daughter was diagnosed with this disease in the capital of maple syrup?” ■

Students get a glimpse of their future

MCH organizes a career exploration day for high school students



On March 13, 46 high school students interested in pursuing a career in healthcare had the opportunity to tour the Montreal Children’s Hospital (MCH) and Research Institute of the MUHC. The grade 10 and 11 students from the English Montreal School Board (EMSB) met with Jessica St-Onge, a nurse in the MCH Emergency department, Vicky Fortin, a specialised technologist in Medical Imaging and Dr. Chris Karatzios,

an infectious diseases specialist and pediatrician on the inpatient wards. They also toured Dr. Pia Wintermark’s lab while she explained her current research into newborn brain and eye damage. ■

Practice makes perfect

Multidisciplinary simulation aims to better patient care

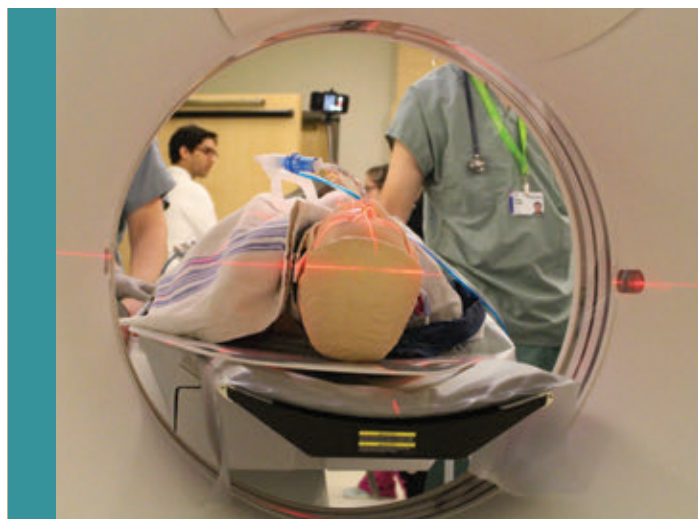


By Sandra Sciangula



► Trauma Team Leader, Dr. Hussein Wissanji, a pediatric surgery fellow, maintains constant communication with team members throughout the entire simulation process.

Last month, over 50 healthcare professionals from the Montreal Children’s Hospital (MCH) were involved in a pediatric emergency medicine simulation involving a “10/10”, the most severe state of trauma seen in the Emergency Department (ED). Of the 50 professionals, about 20 participated in the simulation while 30 others watched via live video stream from the Research Institute Auditorium. The exercise was developed and organized by a team under the direction of Dr. Ilana Bank, Pediatric Emergency Medicine specialist and Director of the MCH Simulation Program. The team practiced on a mannequin, referred to as “the patient”, which represented a 14-year-old boy who was brought to the ED after a scooter accident. An actress was brought on board to play the role of the patient’s mother, and ALSi technology was used to display the patient’s vital signs.



► The “patient”—a mannequin used for simulation exercises—undergoes a CT scan.

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► Members of the team prepare to image the patient in the Interventional Radiology department.



► The patient is transported to the PICU.



► The PICU team takes over from the Trauma team once the patient arrives in the PICU.

Although there was no actual danger, everyone involved treated the simulation very seriously and carried out their roles as if the situation were real.

As soon as the simulation began, the energy in the room rose, the machines beeped, and everyone got to work with intense focus. The group was subdivided into pairs of specialists taking on specific roles and sharing tasks: one respiratory therapist assisted with ventilation while another prepared the intubation equipment; two ED nurses carefully recorded and prepared each medication that was requested, while another nurse helped manage the patient at the bedside. There was a constant hum of activity and communication within groups and among the larger group. Trauma Team Leader (TTL) Dr. Hussein Wissanji, pediatric surgery fellow, managed the group like a conductor leads an orchestra: he commanded moments of quiet and gestured to people for the information he needed to make on-the-spot decisions about the patient's care, announcing the next steps, and maintaining frequent communication with the patient's mother.

THE PATIENT'S ROUTE TO THE PICU

The simulation started in a crash room—also known as a red room—in the ED, where the most severe cases are treated. The team included two emergency room physicians, three ED nurses, two respiratory therapists, one radiation technologist, one interventional radiologist, two anesthesiologists, a social worker, a member of the spiritual care team, a physician and resident from orthopedics, a fellow from the Pediatric Intensive Care Unit (PICU), and the TTL.

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Practice makes perfect... (cont'd)

Once the patient was stabilized, he was brought from the ED to CT for a scan where the decision was made to transfer the patient to the Interventional Radiology department for management of an arterial bleed, and finally up to the Pediatric Intensive Care Unit, where the PICU team took over his care.



► Dr. Wissanji answers questions from the patient's "mom"—an actress who takes part in the entire simulation process.

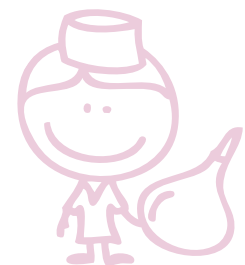
READY FOR THE REAL THING

The ultimate goal of the simulation is to improve patient care. Dr. Bank's exercise is meant to mimic real life as much as possible by involving members of the various disciplines who would normally participate in this type of trauma case. The idea is to practice effective communication and give the professionals a chance to learn about each other's expertise, their role within a department, and their way of working. "This is an opportunity to share the mental model and practice the transfer of care so that it can be as seamless as possible when it comes to a real-life situation," says Dr. Bank. The entire simulation and subsequent debriefing was recorded, annotated and shared with the group, so that areas of success and areas needing improvement are reviewed by everyone who took part.

Dr. Wissanji says one aspect of the simulation he really appreciated was the feedback from the "mom". "This is valuable information that we rarely get in these types of trauma cases," he says. When asked if he would recommend his peers participate in the next simulation, Dr. Wissanji doesn't hesitate. "Practice makes perfect. Hands down, a yes!" ■



► Dr. Ilana Bank leads a debriefing session in the RI Amphitheatre immediately following the simulation.



Shining a light on our social workers

By Sandra Sciangula



Clinical social workers help families cope with the stress of their child's hospitalization, illness or injury. They counsel families, advocate for them, assess patient and family psychosocial needs, provide counseling, assist patients' transition to home, and link families to community and government resources. For Social Work Month, we shine a light on Marie Laberge and Laura Amer, dedicated social workers who find a genuine sense of satisfaction in helping the patients and families they work with.



As a social worker in the outpatient psychiatry clinic at the Montreal Children's Hospital (MCH), Laura Amer's role is a therapeutic one. She works directly with a psychiatrist to evaluate the nature of the patient's symptoms in order to devise a treatment plan. "In psychiatry cases, one of the roles of the social worker is to help patients feel comfortable enough to open up and talk about their pain so we can begin to understand it," says Laura. Children and adolescents are referred to the outpatient psychiatry clinic either from the emergency department or by other healthcare professionals in the community. The MCH outpatient psychiatry clinics treat the most complex and challenging cases. "The majority of our patients are referred to us after they have already undergone evaluations and therapy in the community without success, and when they get to us they can feel hopeless or like they will never get better," says Laura. She adds, "It is profoundly rewarding to see a child who came to us feeling such deep pain, eventually begin to feel better, and even smile."

"Advocating for families is a significant part of social work in the NICU," says Marie Laberge who has been a social worker for over 30 years and the last 20 at the Montreal Children's Hospital. She works primarily in the Neonatal Intensive Care Unit (NICU) where parents are faced with what is often the most difficult and emotionally charged time of their lives. "We intervene at a time of crisis in the family's life," says Marie. Helping parents adapt to what is usually a long stay in the largest unit in the hospital is the crux of Marie's work and that of her two NICU social work colleagues. Services provided include instrumental assistance like lodging, support in applying for special government benefits, referral to community resources as well as providing for the family's emotional needs through supportive counseling and crisis intervention. "We really care for the whole family," says Marie. "Feeling like I can empower parents and be there for them at a critical time is extremely rewarding." ■



Eloisa Binder, PFCC Star of the Month

By Maureen McCarthy



Isabelle and Olivier remember well the feeling of bringing their son Guillaume home after six months at the Children’s in both the neonatal intensive care unit (NICU) and the pediatric intensive care unit (PICU), as well as on the medical floor. “It was the first time in his life that he wasn’t surrounded by machines telling us exactly how he was doing,” says Isabelle. Before leaving the hospital, the family was referred to the Children’s Complex Care Service (CCS), where they met nurse clinician Eloisa Binder. “In the year that followed, Eloisa helped us so many times in so many ways to care for Guillaume,” says Isabelle, who nominated Eloisa for this month’s PFCC Star award.

The CCS sees children with complex medical needs and Eloisa is one of nine clinical nurse specialists who manage and

coordinate care. The CCS has a number of specialized programs including the bronchopulmonary dysplasia (BPD) clinic, which is one of Eloisa’s areas. “When Guillaume first went home, he was on oxygen so we followed him regularly in the BPD clinic,” says Eloisa. In CCS, parents are encouraged to be in contact as much as they need. “I make sure they know that nothing is too minor to ask about. I keep in touch with them a lot between appointments.” In addition to BPD, Eloisa also follows post-cardiac transplant patients, and multiplex (multiple complex need) children.

Understanding the journey that families take from the NICU to going home is something Eloisa knows well. Before she

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joined the CCS team a decade ago, she worked for 15 years in the Children’s NICU; while there she took the extracorporeal membrane oxygenation (ECMO) pump specialist course and was one of the nurses on the NICU ECMO team when it first started.

Practicing patient and family-centered care for Eloisa means treating a family the way she’d want her own family to be treated. A self-described “people” person, she feels that with her many years in pediatrics, she has an understanding of what parents are going through. “Parents understand and know their child so it’s important that they have their say in any discussion or decision, and feel part of a team,” she says. “Sometimes they can be intimidated about talking to someone on their child’s care team, so it’s important to make sure they feel comfortable expressing their thoughts or to have someone to advocate for them.” Eloisa points out that it’s not something she does

consciously, it’s just the way she approaches her role as nurse. “If there’s something better that could be done for a child, or some other way to help the family, then you do your best to make it happen.”

Isabelle agrees that Eloisa always went the extra mile. Guillaume was born very premature and in the year after he went home he had significant problems with his breathing and with his feeding. At one point, he had to be fed with a nasogastric tube. “From our first appointment in Complex Care, Eloisa was with us,” says Isabelle. “She was a huge help to us, a great listener, compassionate. She really supported us as we transitioned to looking after Guillaume at home, and was a constant support whenever we had doubts or worries.” Isabelle also says that the communication and the trust they developed with Eloisa was very important to them “We felt like a partner in Guillaume’s care, and that meant everything to us.” ■



Our PFCC Stars are featured on the Children’s Facebook page and Instagram. Here’s what people had to say about February’s PFCC Star, Judy Edes.

Alison Jung

Judy is amazing. She so helped our family when our little boy was in the hospital for a month. She was encouraging, supporting and always radiating a smile that perked us up!

Mikokato

She’s amazing! So well deserved! Happy for you Judy!

Pamela Audette

Congratulations Judy, keep up the fantastic work you do!

Mélanie Mireault

Wow, she truly deserves it! 😊

Celebrating Child Life Month

The Montreal Children's Hospital wouldn't be the same without our Child Life department. Thank you for everything you do for our patients and families. You help make their stay enjoyable, educational and welcoming.

Here's what some patients had to say about our Child Life team:



"We have taken part in the Parental Presence at Induction (PPI) program with Sabrina and Nathalie, and now, I cannot imagine my son going into the OR without me. We truly appreciate the members of the Child Life team."

—Édith Lacroix, Félix-Antoine's mom

"Julie is so attentive. She knows what each child in the Medical Day Hospital likes. She hooked me up with the wifi password so that I can listen to music and watch YouTube videos while I am here. She goes out of her way and makes sure that there are age appropriate movies for me, like war movies or Batman."

—Julien, 15



"Anna used a doll to help explain that I was going to lose my hair, but that I was still beautiful. And then she gave me the doll to take home! I also love playing with Afifah when I come to the day centre. We do lots of arts and crafts together. I love drawing flowers, because they remind me of spring."

—Mia, 4

"I'm on dialysis three days a week for five hours at a time, so Doreen organizes a lot of fun activities for me. I especially like Wednesdays because we get to cook! I love baking and eating the cupcakes we make. My favourite are the vanilla ones with icing and candies."

—Halil, 13

