Chez nous

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Genetic link helps scientists make new discovery

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Hôpital de Montréal pour enfants
Centre universitaire de santé McGill

Montreal Children's Hospital
McGill University Health Centre

ALSO IN THIS ISSUE:

Seeing double: siblings diagnosed with same rare disease

Genetic link leads scientists to make new immunology discovery

By Stephanie Tsirgiotis and Julie Robert

It all started with a fever. For two months, ten-year-old Thomas Trinh's temperature kept rising and falling, his body ached and his face was swollen. Then in January 2015, his mother, Year Ny Khut, had a feeling that something was very wrong. She brought him to the Emergency department (ED) at the Montreal Children's Hospital (MCH) where a doctor noticed a

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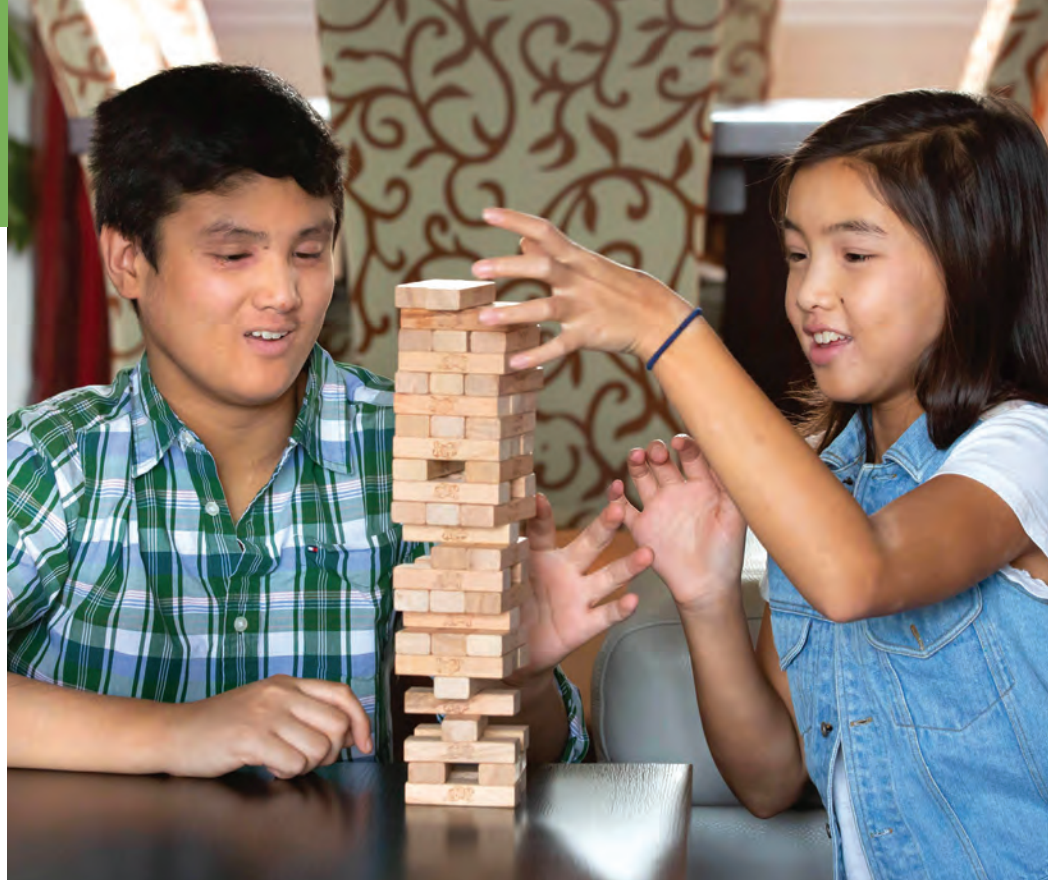
▶ Top: (l. to r.) Brandon, Meagan, King, Thomas and Year Ny.

few small bruises on his back, arms and legs. "The bruises were so small, they were about the size of a small fingernail," recalls King Trinh, Thomas's father. The hospital ran a few tests and confirmed the worst: Thomas was suffering from a type of lymphoma.

A very rare disease

Lymphoma is a form of cancer that affects the body's lymphocytes, an infection-fighting group of cells found in the immune system. The disease causes lymphocytes to change and grow out of control; but Thomas was showing signs of an even rarer condition. "It took the team about a month to diagnose him and figure out the appropriate treatment," says Year Ny. "His doctors said they had only seen two of these cases in their entire career." Thomas was diagnosed with subcutaneous panniculitis-like T-cell lymphoma.

"It was a complete shock to us," says Year Ny. "Nobody in our family had ever had cancer. Could we cure him?" By this point, Thomas's face was so swollen he could barely open his eyes, but his chemotherapy treatments were not working. The MCH oncology team suggested a bone marrow transplant. The entire family was tested and Thomas's older brother, Brandon, ended up being a perfect match. The transplant was a success and Thomas slowly started to feel like himself again. "We were so happy to be able to put all this behind us," says Year Ny. Sadly, that feeling was short-lived.



▶ Thomas and Meagan Trinh were both diagnosed with subcutaneous panniculitis-like T-cell lymphoma.

History repeating itself

A year after Thomas was diagnosed, his younger sister, Meagan, started to complain about a swollen eye. "One of her eyes was red and puffy, but we didn't think it was anything serious. We brought her to the ophthalmologist for some eye drops," says her father. Then a month later, Meagan's temperature started to fluctuate and she noticed something peculiar on her leg – a small bruise. "My heart sunk when she showed it to me," recalls Year Ny. "It looked exactly the same as the ones Thomas had. I couldn't believe it." The family insisted doctors perform a biopsy on Meagan, and shockingly, they

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On the cover: Thomas and Meagan Trinh

Cover photo: Owen Egan

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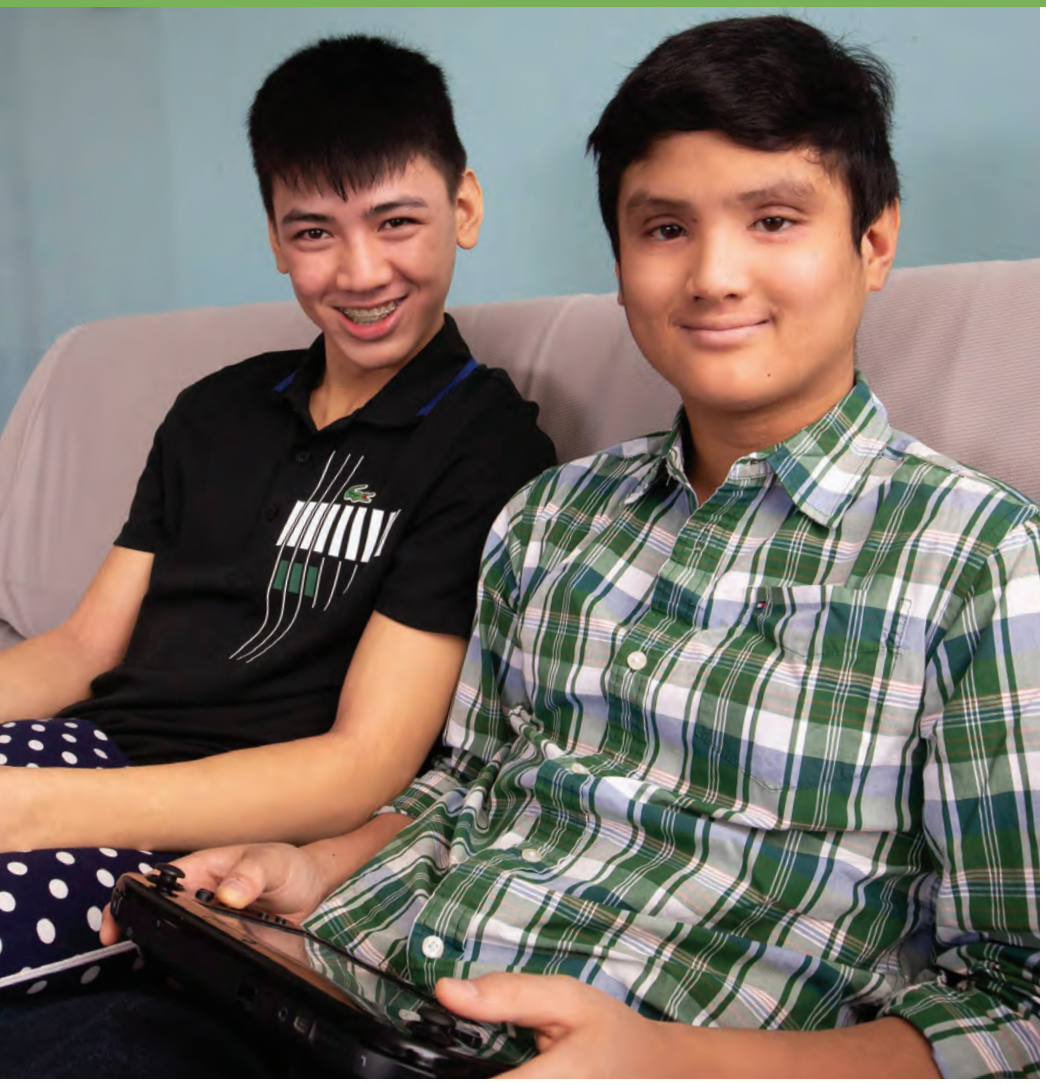

▶ Thomas (r.) received a bone marrow transplant from his older brother, Brandon (l.).

received the same diagnosis: subcutaneous panniculitis-like T-cell lymphoma.

"How was it possible that this rare disease could affect two people in the same family, in the same year?" asks King. "Why was this happening to us and more importantly, why was it happening to them?" Pediatric hemato-oncologists, Drs. Sharon Abish and David Mitchell, began to suspect that the condition could be genetic. They contacted their colleague, Dr. Nada Jabado, a hemato-oncologist at the Children's and researcher at the Research Institute of the McGill University Health Centre. It was soon discovered that Year Ny and King both carried a mutation in a gene, called HAVCR2. As carriers they had no symptoms. However, they had both passed the mutation down to Thomas and Meagan, but not to their son Brandon.

New immunology discovery

Researchers confirmed that the mutation was preventing a protein, called TIM-3, from working properly. TIM-3 is an important protein because it helps regulate the immune system and stops it from going into overdrive. When the protein is suppressed or inactive, the immune system becomes completely "unleashed" and T-cells are uncontrollably over-activated, resulting in this rare form of lymphoma.

"How was it possible that this rare disease could affect two people in the same family, in the same year?"

When Dr. Jabado began discussing this case with colleagues in Australia and France, they realized that they too had similar cases of patients with the same mutation, most of whom seemed to be of East Asian or Polynesian descent. They also identified another mutation on the same gene in patients of European origin. In all, 17 pediatric and adult cases were identified and the findings were published in Nature Genetics, a scientific publication.

Most lymphoma cases are treated like cancers, when in reality they may often be intensified responses of the immune system. "We're now looking into using TIM-3 as a target to trigger enhanced immune responses in patients with cancer," says Dr. Jabado. "We hope this

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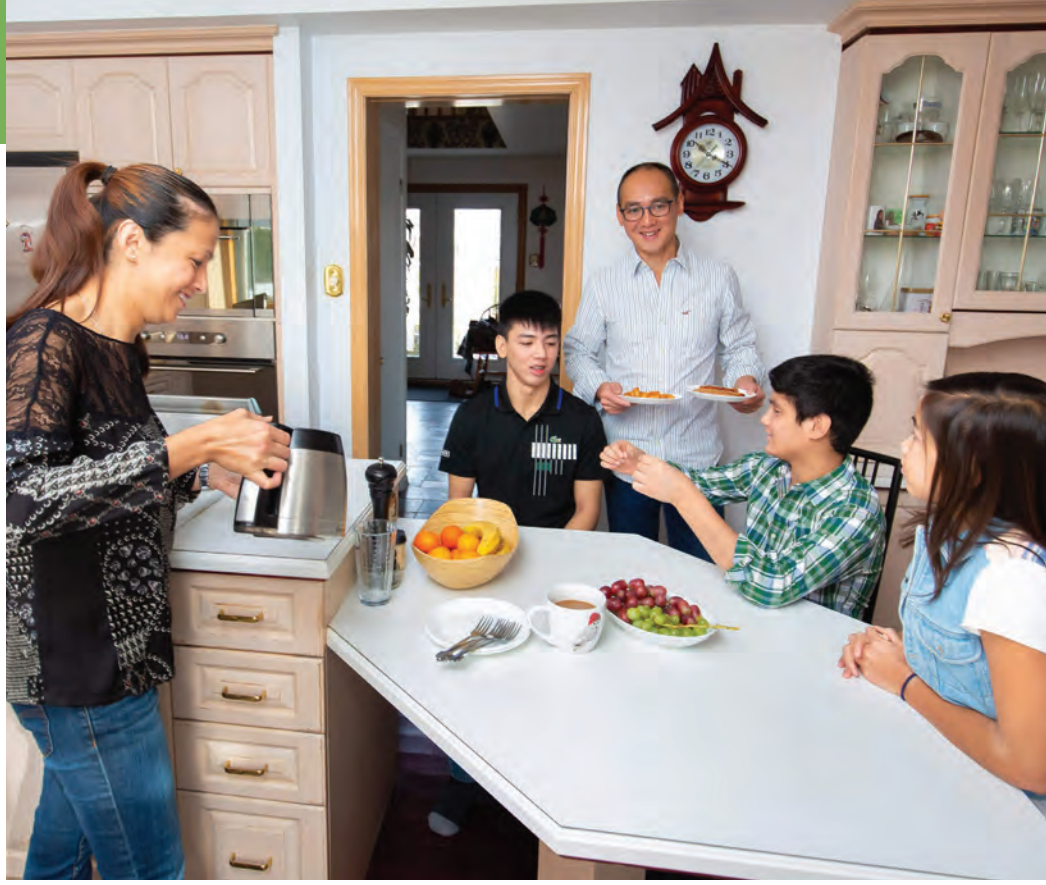
will provide much better results and fewer side effects than cytotoxic chemotherapy."

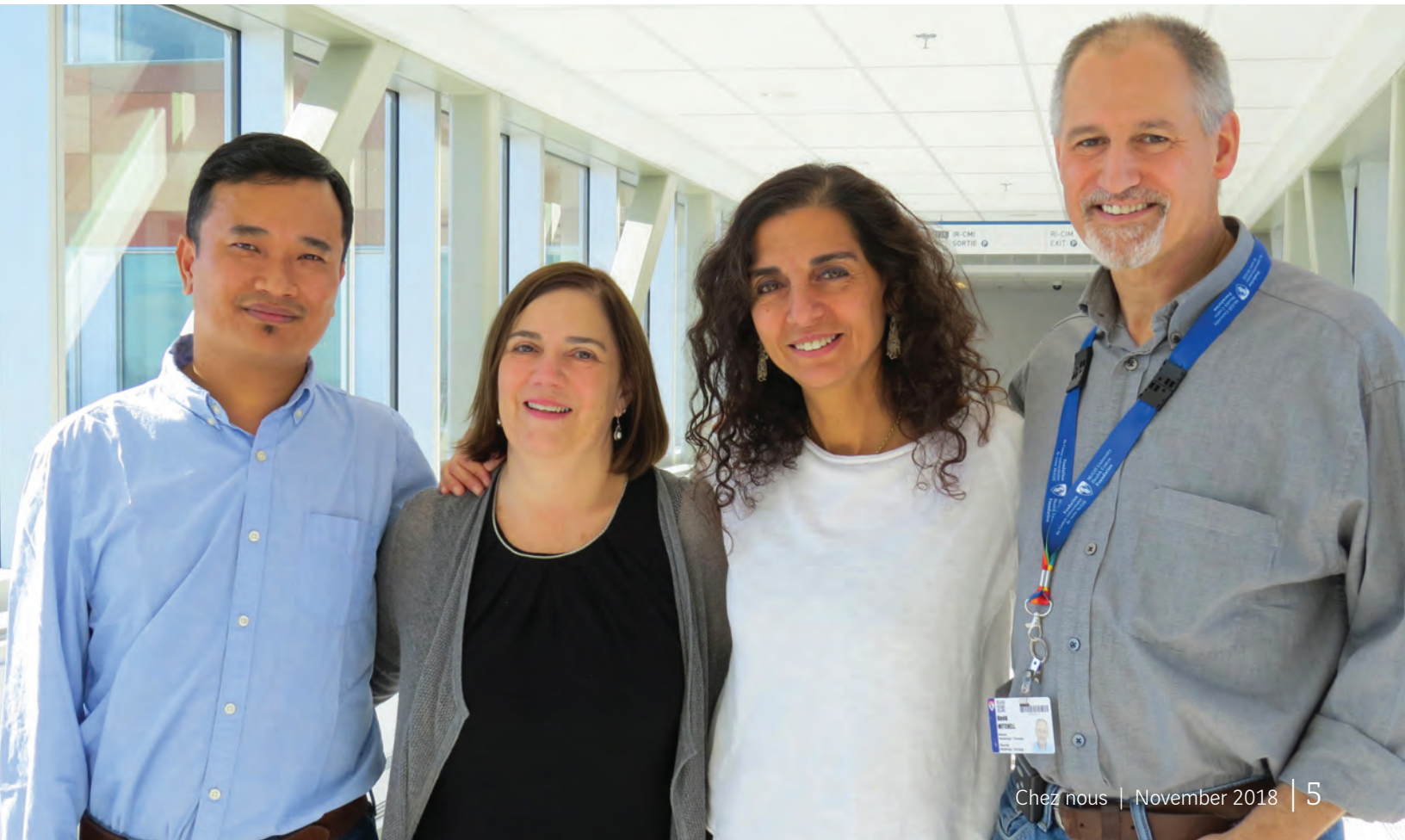
Researchers are now trying to see if patients with autoimmune diseases such as lupus—a disease where the immune system turns against the body itself—may have some TIM-3 dysfunction. This could potentially lead to more effective treatments for various types of cancers, infectious diseases such as HIV, or perhaps malaria, as well as multiple sclerosis.

"Our family has been through a lot over the last few years, but we're glad these findings will help so many other people," says King. "This all happened for a reason." ●



▶ Above: The Trinh family has been through a lot over the last few years, but believe this all happened for a reason.

▶ Below: (l. to r.) Dr. Tenzin Gayden, Dr. Sharon Abish, Dr. Nada Jabado and Dr. David Mitchell joined forces and expertise to solve a medical mystery that originated at the Montreal Children's Hospital, leading to an important international discovery.



A day in the life of a...

food service attendant

By Stephanie Tsirgiotis

Denise Bourgon is hard to keep up with. She walks fast, works fast and thinks fast. "I'm always thinking about what I need to do next," she says. Denise has been working in Food Services at the McGill University Health Centre for the past 24 years. She started off at the Montreal Chest Institute and was transferred to the Montreal Children's Hospital in 2008 where she began working in the formula room.

Every detail counts

The formula room is not easy to find. It's tucked away inside the kitchen on S2. Small and tidy, the space has everything Denise needs to

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▶ Top: Denise Bourgon

> **We don't have the right to make a mistake because we're dealing with children and their health.**

make specialized liquid formulas for newborn patients at the MCH, or for older pediatric patients with restrictive dietary needs. Denise starts off every morning reviewing her long list of daily orders. Each formula is tailored to the child's nutritional needs and is accompanied by a recipe prescribed by an MCH nutritionist.

"I make enough formula to last 24 hours, and on average, we prepare 15 to 25 special formulas a day," she says. "It takes a lot of concentration and attention to get every detail just right. I always tell my colleagues that we don't have the right to make a mistake because we're dealing with children and their health."

▶ Unit coordinator Bernard Riel thanks Denise for her quick turnaround. She delivers formula to the milk room on each floor.

Each specialized formula varies in complexity; some are liquid concentrate infant formulas diluted in sterile water, while others are in powdered form or ready-to-feed. "Some of the formulas I make are a combination of nine ingredients. Each recipe must be followed precisely in order to have the exact right amount of nutrients for every patient," she says. "And for mothers who are breastfeeding, I prepare special concentrates they can mix in their breast milk." Denise also prepares milkshakes or calorie-rich shake mixes, called Scan dishakes, for children who need to gain or maintain their weight.

Taking the extra step

After lunch, Denise delivers and distributes the prepared formulas to each ward, with most of her clientele making up the neonatal intensive care unit. "I deliver my stock to the milk room on each floor," she says. "Even though I don't have any interaction with the patients, I feel like I know them because I've been preparing their formulas for weeks or months, and sometimes even years."

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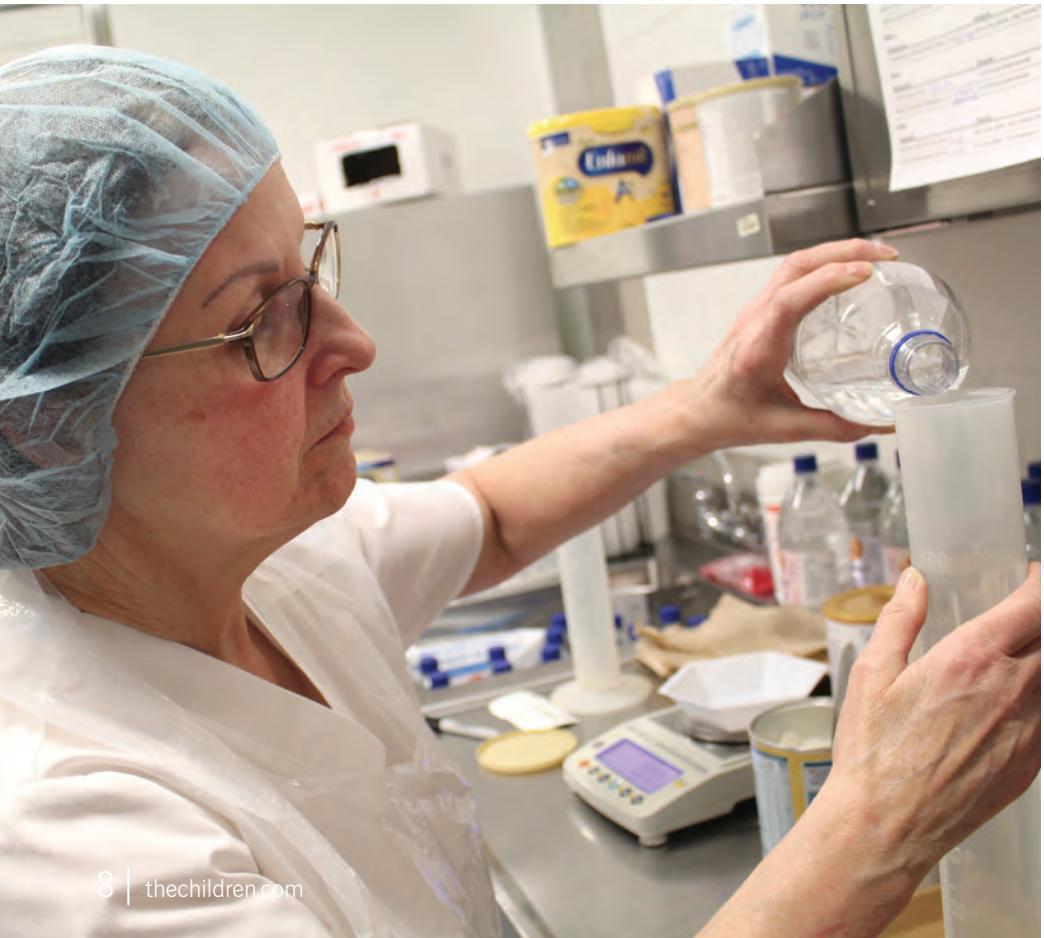
When dropping off the new stock, Denise also takes the time to observe whether or not yesterday's orders were fully consumed. "We want to make sure that we're making the right amount of formula for each patient. If I notice that a patient's formula hasn't been touched or that every last drop has been drunk, I follow up with a nutrition technician to see if we should be decreasing or increasing the amount I make," she says. "I work very closely with that team. They answer my questions and keep me up-to-date on certain situations. We sometimes call each other 20 times a day!"

Using good judgment

Considering the number of new admissions a day, Denise is constantly reprioritizing and juggling her responsibilities. "I get called with a lot of different emergencies, so I've developed good judgment over the years in terms of what I should tackle first," says Denise. Besides preparation and delivery, she is also responsible for reviewing the formula room's inventory list, checking expiry dates and restocking pre-op, the emergency department and the recovery room with ready-to-feed formulas.

▶ Above: Besides preparation and delivery, Denise is also responsible for reviewing the formula room's inventory list.



"I find my job very fulfilling because I get to participate in a child's recovery," she says. "We all play a part in helping these patients get better. Doctors and nurses use their brains to help these children, and I use my arms." ●

▶ Left: Denise makes enough formula to last 24 hours, and on average, the team prepares 15 to 25 special formulas a day.

PICU takes on medication safety

A key mandate for the Quality Improvement committee

By Sandra Sciangula

The Pediatric Intensive Care Unit (PICU) at the Montreal Children's Hospital (MCH) formed a Quality Improvement Patient Safety Committee (PQUIPS) made up of a multidisciplinary group of caregivers, including doctors, nurses, parents and pharmacists. Ni Nora Ruo is a pharmacist in the satellite pharmacies of the intensive care units. As part of PQUIPS, Ni Nora heads a sub-committee that examines medication safety.

"Medication errors are the most frequent medical errors in hospitals," explains Ni Nora. In an effort to improve medication safety, she and her committee first mapped out the medicine management process, from the moment a medication is prescribed to the moment it is administered to a patient. The multi-step process was then compared to incident reports from the previous year to identify where the weak points are in the process. "Sometimes, the cause for error is as basic as illegible handwriting," she says.

The committee was then able to prioritize four areas of focus: prescription ordering, review of the administration method of the medication, the independent double check for high alert medications, and a review of the administration plan. Each area of focus is then studied in detail using the Healthcare Failure Mode and Effect Analysis, which is used to identify potential causes leading to medication errors and to prioritize areas for improvement using an algorithm.

To raise awareness on some of the identified issues in an interactive way, the members of the sub-committee organized a simulation where participants from across the hospital could participate by identifying medication errors surrounding a mannequin. The activity was well received with a high percentage of participants finding the exercise educational and with an equally high percentage of participants saying that they plan on changing the way they handle medication.

Another aspect of medication safety that the sub-group is immediately rolling out is a parent survey asking how involved they would like to be in the prescription and administration of medication for their child. The questionnaire—approved by the hospital's Research Ethics Board—will help the committee understand how best to involve parents and how much information is too little versus too much for a parent. "A parent knows their child best, so we believe that a parent's involvement can improve the quality of overall care the child receives," she says.

▶ Top: (l. to r.) Gabrielle Girard, Ni Nora Ruo, Elain Grant, Sarah Shea, Chrysanthi Roussianos. Absent from the photo: Jeannine Julien, Gabrielle Cunningham-Allard, Razan Babakr, Valerie-ann Laforest, Anne-Isabelle Dubé, Irene Gernet, Laurent Methot, Saleem Razack.

MCH pediatric surgeons develop new app for congenital diaphragmatic hernia

By Maureen McCarthy

Congenital diaphragmatic hernia (CDH) is a "hole" in the diaphragm muscle through which the intestines can move into the chest. Advances in neonatal care have improved survival rates in the last 30 years, but survivors can experience significant long-term health issues that affect feeding, growth, and brain development.

Dr. Pramod Puligandla, Pediatric Surgeon and Pediatric Intensivist at the Montreal Children's Hospital (MCH), was the project lead of the Canadian Congenital Diaphragmatic Hernia Collaborative that recently developed a guideline for standardized CDH management. The guideline was published in the Canadian Medical Association Journal (CMAJ) in January 2018.

Building on what was achieved with the CDH Guideline, Dr. Puligandla and Dr. Kathryn LaRusso, Research Fellow in the Department of Pediatric Surgery at the MCH, worked with Montreal-based Stradigi AI to develop an app that would provide healthcare professionals with a comprehensive tool to assist management decisions related to CDH care.

The main features of the app include rapid access to recommendations and evidence summaries for all phases of CDH care from prenatal diagnosis to hospital discharge and follow-up; links to medical calculators for key patient data; a daily ICU rounding checklist and flowsheet; a guideline compliance checklist for quality improvement; and access to the full guideline document with expanded discussions and a complete set of references. The app is also an excellent resource for teaching and education during patient care rounds.

"The goal of the CDH Guideline was to improve outcomes for children with congenital diaphragmatic hernia by standardizing care across Canada," says Dr. Puligandla. "Managing CDH requires the expertise of a variety of specialists as well as primary care physicians, and the app provides easy access to tools and resources useful to all clinicians involved in the care of children with CDH."

Support for the app development was provided by a Medical Staff Services Association Innovation Grant at the MCH.

The app is free to download and is available on the Apple App Store. Enter "CDH" in the Search function. The app will soon be available for Android phones. ●

▶ Top: (l. to r.) Dr. Kathryn LaRusso and Dr. Pramod Puligandla.

Ceinture Olivier

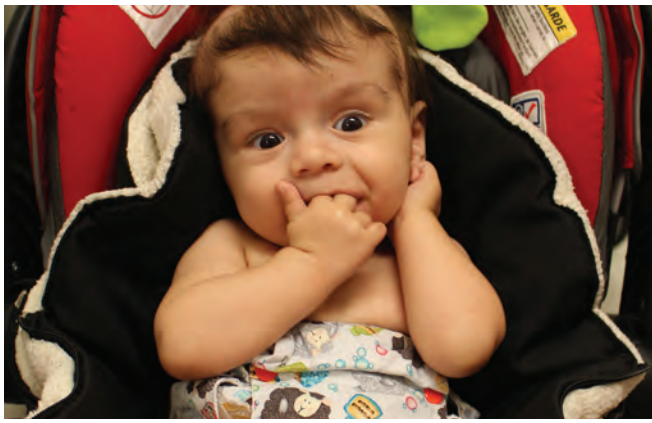
A family shares their creative solution to help other patients

By Sandra Sciangula

While undergoing chemotherapy for a brain tumour at the Montreal Children's Hospital (MCH), six-month-old Olivier had a gastrostomy tube (G-tube) inserted in his abdomen to facilitate his feeding. To reduce the risk of the G-tube being dislodged, Olivier's mom, Liliane Laroque, reached out to her aunt Johanne Long for help. A technician in industrial engineering by profession and owner of THL Costume, a company that creates outfits for dancers and figure skaters, Johanne was up for the challenge. She modeled her design on a G-tube belt that is only available in the United States and France. The morning after her niece's call, Johanne had a prototype ready.

Dr. Hussein Wissanji, pediatric surgery fellow at the Children's, and one of the surgeons who inserted Olivier's G-tube, was impressed with Liliane and Johanne's innovative solution. The two women delivered 36 belts to Dr. Wissanji's office for distribution to other patients who will have a G-tube inserted. "This is a heartwarming initiative," says Dr. Wissanji. "It's amazing that a family facing such adversity can come up with an ingenious solution and want to so generously help other patients."

The now aptly named *Ceinture Olivier* has been modified and designed to fit infants of all sizes and there is even a model that is appropriate for bath-time. With the help of a company that has donated the material and a group of volunteers who sew, Johanne has made over 100 belts that she will distribute to each of the four pediatric hospitals in Quebec.



A new Welcome Guide
for parents and families

The new MCH Welcome Guide for parents and families is now available. The Guide, which was developed with input from staff and parents, contains information about the hospital and its services, and aims to provide parents with a better understanding of what to expect during their child's stay at the Children's. The Guide has key information on topics such as parking, food and retail services, and visiting guidelines, and includes information on the various members of a child's care team and the support services available to families.

Distribution of the new Welcome Guide will begin in the coming weeks, and parents and families will receive a copy at the Admitting office.

Complete the *Chez nous* survey
for a chance to win!

The Children's Public Relations and Communications office would like your feedback on *Chez nous*! Our short survey takes only a couple of minutes to complete and your answers will help ensure that *Chez nous* remains a valuable way to communicate important news and information about the MCH. You'll find the survey online at **surveymonkey.com/r/Chez_nous_EN_2018**.

If you include your name, email and phone number at the end of the survey, you'll have a chance to win a **$50 Amazon gift certificate**. Keep in mind you can also choose to remain anonymous if you prefer.

Deadline to complete the survey is **December 6, 2018**.