

Chez nous

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Severe juvenile arthritis: Emersyn triumphs over rare disease

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Hôpital de Montréal
pour enfants
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Montreal Children's
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More than joint pain

Toddler diagnosed with severest form of juvenile arthritis

By Stephanie Tsirgiotis

Five-year-old Emersyn Shragie loves to colour, but two years ago she stopped being able to hold a crayon. Her family first started to notice something was wrong during a family vacation in December 2016. Emersyn would wake up feeling normal, but by mid-afternoon, her whole body was covered in little red welts. "We thought they were hives," recalls her mother,

Natalie Ornstein. "Every morning she would wake up with clear skin, but then 3:00 p.m. would roll around, and she would start having a high fever and the hives would come back."

From bad, to worse

After returning home, Natalie brought Emersyn to her pediatrician, who subsequently referred

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► Top: Shannon, Emersyn, and Natalie

her to an allergist. She was prescribed a high dose of antihistamines, but the hives continued to appear on a daily basis. “We thought maybe she was allergic to something or that they were related to some kind of stress,” says Natalie. Emersyn continued to visit pediatricians and allergists, who all believed she was suffering from a form of chronic hives, but Natalie couldn’t shake the feeling that something was very wrong.

Then in March, Emersyn woke up one morning screaming that she couldn’t walk. She also stopped being able to feed herself, get dressed or walk down the stairs. “She would slide down on her bum like a baby,” says her mom. Her hands and feet began to swell and her skin started to peel. She also stopped colouring.

Diagnosis brings relief

Unsatisfied with their daughter’s diagnosis, Natalie and her husband, Shannon, asked to have more blood work done. “The results were shocking,” she says. “Everything was abnormal.” Natalie immediately brought Emersyn to the Emergency Department (ED) at the Montreal Children’s Hospital (MCH), where the ED physician noticed something in Emersyn’s fingers: arthritis.



► Emersyn gets a new bravery bead from her nurse, Nadia Narducci.

Within the first 10 minutes of meeting Emersyn, Dr. Rosie Scuccimarri, a pediatric rheumatologist at the MCH, knew what was ailing her. The toddler was diagnosed with Systemic-onset Juvenile Idiopathic Arthritis, a rare, and potentially life-threatening form of chronic arthritis. This type of arthritis not only affects a patient’s joints, but can also affect their organs, specifically the lining of the heart, as well as cause

swelling of the liver, spleen and lymph glands. “We were shocked, but relieved at the same time,” says Natalie. “We finally knew how to help her.”

Over thirty of Emersyn’s joints were affected, everything from her hands to her knees and ankles. Her fingers were so swollen that she couldn’t close them to make a fist. She was also severely anemic and her lymph nodes were

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*On the cover: Emersyn Shragie
Cover photo: Owen Egan*

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inflamed. “Because her inflammation was so significant, we were worried about a cytokine-storm which can occur when inflammation becomes uncontrolled,” says Dr. Scuccimarri.

A cytokine-storm is when the body starts overproducing cytokines, a type of small protein produced in response to an infection or an inflammatory disease. Cytokines have the ability to alter the behaviour of other cells, which can lead to very serious complications, like macrophage activation syndrome. “Macrophages are a type of white blood cell and are usually helpful to the immune system, but cytokines can turn them from good to bad,” explains Dr. Scuccimarri. “They start eating up other cells, like platelets, as well as

red and white blood cells. If this is not caught early on, a patient can die rapidly. Thankfully for Emersyn, we were able to stabilize her condition before it got to this point.”

Treating the condition

In order to avoid a cytokine-storm and to decrease her inflammation, Emersyn was prescribed a number of different drugs, including corticosteroids, a type of powerful anti-inflammatory drug. She now takes four pills a day, and receives three injections at home on a weekly basis, a task she shares with her mother. “I was diagnosed with Crohn’s disease in my twenties and I’m currently in remission, but I still need to give myself preventative injections once

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► Dr. Rosie Scuccimarri, Emersyn’s rheumatologist, examines her finger joints.



a week,” says Natalie. “On Saturdays, Emersyn gets her injection first and then she holds my arm while I inject myself. It reassures her to know that she doesn’t have to do this alone.”

Emersyn also visits a physiotherapist and occupational therapist to help with her mobility. “We do a lot of exercises at home through play, like picking up the little pieces in Light Brite. Her fingers are the worst affected by the arthritis, so we try and work on that,” says her mom. Because her disease is rare, Emersyn is also part of a Canadian study group that is currently looking into its long-term prognosis. “We only see two or three of these cases a year at our hospital, and we don’t know what the underlying cause of it is. We believe its multi-factorial, but we’re not sure if it’s genetic or triggered by an infection,” says Dr. Scuccimari. “Our goal is to put these types of diseases into remission by treating them aggressively.”

Since being diagnosed, Emersyn has come a long way. Only six of her joints are now affected and she started colouring again. “I like to colour Cinderella,” she says. In September, she began kindergarten and started sleeping in her own bed, a big accomplishment which warranted a new bravery bead. “Emersyn has over 75 bravery beads and each one represents a different procedure or treatment she had to endure,” explains Natalie. “She is very proud of them and we are so proud of her.” ●

► Emersyn proudly stands next to her bravery beads.





A day in the life of... an ethicist

By Stephanie Tsirgiotis

As a respiratory therapist, Lori Seller was often confronted with ethical issues. "I worked primarily in the intensive care units, which tend to be very stressful, emotional environments," she says. "And I started to question things; not about the care we were delivering, but about why we did or said the things we did." This curiosity led to Lori returning to school to complete an undergraduate degree in philosophy, followed by a Master's degree in Bioethics at

McGill University, thus propelling her down a whole new career path: ethics.

Ethicists bring to the table special training in moral theories and critical analysis, as well as knowledge of well-established principles of medical ethics, local health law, patients' rights, clinicians' professional obligations and hospital policy. "It is actually quite rare that an ethical dilemma for which I am consulted is

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► Top: Lori Seller

the first time anyone has confronted that issue. Our knowledge of these different considerations and our ability to recognize and navigate their interplay is part of the ethicist's expertise," she says.

Lori explains that ethics goes beyond just following simple rules like "respect confidentiality". "The approach we use here is casuistic which means case-based reasoning. It involves thinking about how we usually do things, and how and why particular aspects of a given case might ethically justify the need to do things differently." In general, Lori is called upon to help name the ethical problem, provide an ethics analysis and recommend a principled resolution.

Lori's work is divided into three categories: clinical ethics, research ethics and teaching. She is part of the McGill University Health Centre's (MUHC) Centre for Applied Ethics and is the only ethics counselor assigned to the Montreal Children's Hospital. As an ethicist, her primary goal is to try and balance competing interests and values. "I wear different hats. I'm part analyst, interventionist and mediator," says Lori. "It's important to remember that we all have our own 'moral compass'. Sometimes our different values don't align and sometimes we weigh the importance of things differently. That can lead to conflicts."

Clinical ethics

About once a month, Lori will be asked to consult on a clinical ethical dilemma, most often in the intensive care units or hematology-oncology unit. "These areas deal with a lot of stress, as well as end-of-life decisions," she says. "I help bridge the gap between families



► Dr. Patricia Fontela meets with Lori to discuss a pediatric research study.

and the clinical team when competing values are in play." In these situations, Lori will sit with the care team to gain insight, and learn more about the dilemma in question.

A dilemma either signifies two competing goods, both of which you would like to achieve, or two competing harms, both of which you would like to avoid. An example might be a case in which everyone feels there is a choice to be made between prolonging life versus prioritizing comfort. "Sometimes what it takes to achieve one goes hand in hand with undermining the other; achieving both perfectly is just not possible, although both are things we strive for in medicine," she says. In these cases, both families and clinicians can feel quite a bit of moral distress. Everyone feels like they are not doing everything they should.

Lori's main goal when meeting with families is to better understand their point of view and ensure there is at least a shared understanding of some of the facts. She also uses this opportunity to explore the family's understanding of the medical team's perspective and

point out any discrepancies or assumptions that she uncovers so they can be discussed. "As a neutral third party, my intervention is usually well received by the family. I make it clear that my role is not to pressure them into agreeing with the clinical team," she explains. "I'm called in to unblock an impasse and to help move everyone forward. At the end of the day, I make recommendations to the team, but nothing is binding. Clinicians and the patient's family are responsible for all final decisions."

At other times, clinical teams will ask Lori to provide some guidance about how to best meet their professional ethical obligations. For example, questions like: what is an appropriate level of disclosure in this context, or am I obligated to offer interventions that exist but that I do not recommend in this circumstance? In these cases, Lori works solely with the clinical team and will not necessarily meet with the family. "I'm often called when there is uncertainty about the 'right' or 'least wrong' thing to do or about what 'should' be done in a particular context. Sometimes I can be offered as a resource to families, a neutral third party who can help them

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to articulate their values and promote shared decision making. Other times, the clinical team just wants to work through the options and discuss their professional obligations.”

Research ethics

As co-chair of the MUHC’s Research Ethics Board (REB), Lori spends the majority of her time reviewing and validating research trial proposals. “Any research involving humans needs to be reviewed and approved by an ethics committee,” she explains. Every year, the MUHC reviews about 800 new research studies, 150 of those in pediatrics. When reviewing these types of research projects, the committee asks two significant questions: is this ethical and is this scientifically valuable?

The board—made up of professionals with wide-ranging expertise in science, ethics, law, as well as a family partner—reviews the study, the language used in the consent form and ensures that regulatory requirements are met. Ultimately, the REB’s mandate is to ensure the rights, well-being and safety of participants. When dealing with children, there are additional protections in place as

“ I’m part analyst, interventionist and mediator... ”

they may have difficulty understanding how research is different from clinical care and weighing the risks and benefits of participation. Studies cannot begin until they are reviewed and approved by the REB.

“These research trials are so important when it comes to advancing medicine,” she says. “I’m continuously amazed by how innovative the MUHC and its Research Institute is. We see a lot of studies and research ideas created by our very own physicians. Clinicians in this institution are always looking for and finding new and better ways to treat their patients. It’s very inspiring to witness. I always joke that we are lucky these people are using their brains for good instead of evil!”

Teaching

Another important part of Lori’s job is education. She teaches ethics to medical students and residents, as well as nurses and allied health professionals. She also gets involved in case debriefings, where she sits down with a clinical team after the fact to review a situation that caused moral or ethical strife. “Sometimes these situations occur because the team has not reached a consensus and ends up sending mixed messages to the family. It’s important to figure out what the real medical options are and to present those in a consistent way when it comes to these difficult decisions,” says Lori. “And I’m happy when I get to play a little part in that.”

Lori is also invited to attend psychosocial rounds in several departments. “This gives the team a chance to recognize potential ethical issues that might come up and to discuss them with me so we can work to prevent the situation from escalating or becoming a full-fledged dilemma. I also think that those who see me as a member of their team are more comfortable reaching out to me to discuss ethical issues when they come up.” ●

► Lori discusses a clinical ethical dilemma with palliative care team, Evelyn Ouellet and Dr. Stephen Liben.





Cure SMA Québec

A Children's mom starts group for support, education and friendship

By Maureen McCarthy

Émile was a little over one year old when he was diagnosed with a rare genetic disease. Spinal muscular atrophy, commonly referred to as SMA, is a condition that affects the motor nerve cells in the spinal cord, severely limiting an individual's ability to walk, eat, and breathe. Émile's mom, Amy, was devastated by the news but it didn't take long for her to seek information and support. "I was really happy to find out about a group called Cure SMA Canada, and through their

organization, I went to a conference in California for professionals and families living with SMA," she says. "I learned so much while I was there and met some amazing people, and it all made me think, 'Why not start a group in Québec?'" And so she did.

Amy approached SMA Canada with her idea, and shortly after, Cure SMA Québec was launched. Within a few days, 80 people had joined. They

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► Top: SMA Québec's membership includes about 100 families whose children live with SMA. Around 20 of the families took part in a four-day summer camp last year outside Montreal.

now have over 500 members representing about 100 families living with SMA. “People were really excited to get this going. Our first meeting was in someone’s apartment, and then it took off from there,” she says.

Amy points out that the group members support each other in many different ways. “We organize regular outings such as picnics or dinners, and events around SMA Awareness Month in August, and we’ve even organized dinners at the Children’s for those of us whose kids are followed here,” she says. The group’s Facebook page also provides a way for members to organize events, share information and ask questions on new treatments and clinical trial news, and even help each other out in an

► Amy and her son, Émile.



emergency. Amy recalls one time when she borrowed a breathing machine from another family nearby when Émile’s machine malfunctioned. “The opportunity to get out and meet new people is really important,” she says. “Plus, social media networks are really important for groups like ours, and in some cases, our group is helping to break the isolation for families who live with the disease.”

From the National Assembly to summer camp

From her first days as the de facto leader of the group, Amy is now co-president of Cure SMA Québec’s board along with Isabelle Boudreault, whose child is also followed in Complex Care at the Children’s. One of their key accomplishments last year was being invited by Amy’s MNA Dave Turcotte to present a petition to the National Assembly in Québec regarding reimbursement for a new drug which has promising results in treating SMA. They received a standing ovation after their presentation. Amy believes it was real proof of how the parents are pushing things forward and advocating for their children.

Last summer, Amy and several others organized a four-day trip to a summer camp near Montreal. “There were around 20 families who took part with some staying overnight and others making day trips,” says Amy. She explains that in the day-to-day lives of families living with SMA, the siblings are often left out



► Along with many other families, Amy (r.) and her children Elli and Émile attended a demonstration in Ottawa last March to urge the federal government to help fund treatment for SMA.

of the equation, but at summer camp, they’re meeting other children just like them, and they’re included in all the activities. “I think they really understand each other and they start to form friendships too,” says Amy. “It’s fun for them to share with other brothers and sisters who are living the same thing.”

Amy is already planning the next outing which will be a trip to either Valcartier or Jay Peak waterpark. So how do Amy and the other parents keep things going? “Well, we’re all busy parents and we’re all volunteers on this,” she says. “But what it gives us in return is really worth all the work we do.”

Find out more about Cure SMA Québec on the group’s Facebook page. ●



CHEO visits MCH

On October 10, twenty administrators, architects, clinical engineers, project planners and clinical staff from the Children's Hospital of Eastern Ontario (CHEO), visited the Montreal Children's Hospital. CHEO is in the beginning stages of a capital planning process and

their staff were interested in learning from organizations who recently underwent a large infrastructure change. They were particularly interested in lessons learned related to the planning process and overall outcomes. ●

Mock Code Orange

On October 18, the Montreal Children's Hospital (MCH) took part in a city-wide Code Orange simulation. Twenty pediatric mock patients with varying degrees of traumatic injury, including children in need of massive blood transfusions, were transported to the Children's. Healthcare workers from different sectors of the hospital took part, and some of the patients were even transported to the operating room and intensive care unit. "This simulation took over 10 months to organize and was extremely successful because it showed us what we do well and what the gaps are," says Dr. Elene Khalil, MCH emergency physician and medical co-director of emergency

preparedness and director of education at the McGill University Health Centre (MUHC). Dr. Ilana Bank, director of the MCH's Institute of Pediatric Simulation, also reiterates the importance of these exercises. "Simulating these events is the best way to get better and be prepared," she says. ●



The Montreal Children's Hospital Foundation launches its new image!

On September 25, the Children's family got a sneak peak of the new visual identity of the Montreal Children's Hospital Foundation, whose role is to support all of the hospital's departments.

Heal. Love. Bond.

Inspired by the staff's dedication to their young patients, the new logo reflects how they heal sick children, love them and form unbreakable bonds with them and their families. "We interviewed more than 30 staff members, patients and their families, and these messages emerged time and time again. The advertising agency Cossette captured this level of commitment very well when they toured the hospital," says Foundation President, Renée Vézina.

The new logo is composed of three elements—a single bandage, representing the hospital's core mission to heal children, and a heart shape, representing love—joined together they form the figure of a child, representing the unique bond between donors, the hospital, our patients and their families.

Since 1973 the Foundation has raised \$455 million for the Children's. The Foundation is confident that its new image will only add to this success by rallying even more donors to support the Children's, its staff and patients. ●



guérir
heal



aimer
love



s'unir
bond

la fondation
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de montréal
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children's
hospital
foundation



Memorial service

A memorial service is being organized to remember the children who have died recently at the MCH. All staff members are warmly invited to attend this service, which will be held on **Tuesday, November 27, at 2:30 p.m. in the Auditorium of the MUHC RI, (ES1. 1129).**

