

- Wedding proposal pays tribute to CC
- Patient returns to celebrate 50th anniversary open heart surgery

Focused on transparency, CC expands patient access to medical record notes

On July 1, the NIH Clinical Center expanded how it shares information with its patients.

The Clinical Center already shares some documents from patients' electronic medical records, or Clinical Research Information System (CRIS). This summer, more notes will be sent to the Clinical Center's patient portal for patients to review, including inpatient and outpatient progress notes that log lab results, treatments and medical procedures. Launched in 2013, the patient portal is a secure system that provides patients with access to their medical information.

As with other healthcare organizations already sharing notes, the Clinical Center is moving forward with this initiative to enhance patient engagement. The goal of this shift is to help patients remember critical information regarding care transitions, care plans and important next steps or testing; to help empower patients to be active partners in the research and clinical care processes; and to improve overall quality and safety of care.

Researchers have reported that organizations who share visit notes through patient portals have found it beneficial. Some of those findings show that patients report improved adherence with care plans and medication regimens and physicians report improved patient satisfaction and trust. Additionally, considerable research demonstrates that engaged patients have better outcomes.

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First randomized, controlled study finds ultra-processed diet leads to weight gain

Studies throughout the world have shown associations between processed food and negative outcomes. Eating processed food may make you gain weight. It seems obvious, right? Surprisingly though, most scientific findings around this topic have only relied on self-reporting and are simply associations.

In May, Dr. Kevin D. Hall, a senior investigator with the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) published a study on the first randomized, controlled trial at the NIH Clinical Center (<https://tinyurl.com/y26my5cp>). The study



The metabolic kitchen can create precisely measured ultra-processed and unprocessed meals for study participants. Photo credit: Jennifer Rymaruk

found that people who ate processed food ate more calories and gained more weight than when they consumed a whole food diet with foods that were unprocessed.

DIET *page 4*

First-in-human gene therapy launches for pediatric patient with rare neurogenerative disease



In May, a ten-year-old girl named JoJo received gene therapy for an extremely rare neurogenerative disease called GM1 gangliosidosis. GM1 progressively destroys nerve cells (neurons) in the brain and spinal cord, leading to muscle and movement weakness, difficulty swallowing and a shortened life expectancy. The a first-in-human gene therapy trial was led by Dr. Cynthia Tift with the National Human Genome Research Institute and partners at the University of Massachusetts and Auburn University. The therapy provides a replacement, or normal, copy of the gene *GLB1*.
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The gene is encapsulated in a virus that has shown it targets the central nervous system.

"It's like a viral infection. The viral code dumps the DNA into a cell," Tifft said. "All the guts of the virus (the DNA of the virus) has pretty much been eliminated to make room for the gene you want. The gene we want is sitting in the cell and will begin to make the enzyme that is deficient in the child."

JoJo is one of 35 patients who are a part of an NHGRI natural history study for GM1.

"There was very little known about the disease until about 10 years ago when we started doing a natural history study," said Tifft, principal investigator of the trial. "Based on data that we obtained doing our natural history study in the Clinical Center, the only place we could have ever done it, we basically had enough data to launch a gene therapy trial."

As for most scientific research, Tifft said this has been quite the odyssey. JoJo has been coming here for four years. She used to talk and walk quite well, but now only has the ability to say a few single words and cannot hold herself upright.

When it came time for the 30 minute IV infusion of the gene therapy, a medical advancement a decade in the making, Tifft recalls the moment feeling "almost spiritual."

"It was kind of surreal almost because you think you know what's going to happen and how things are going to work," Tifft said. "While JoJo slept peacefully, 30 of us stood quietly in her dimly lit room listening to the infusion pump and watching her sleep."

While it's still early to tell, at about three weeks post-gene transplant, Tifft said "JoJo stood independently for five minutes. I haven't seen her stand like that for almost two years. That makes me really optimistic."

The care team plans to bring in the second patient mid-August and the third patient mid-November. They will follow each patient for five years, post-gene therapy.

- Molly Freimuth

Patient celebrates 50th anniversary of open heart surgery at Building 10



Former Clinical Center patient Tom Kaminski (center) returned "home" to the NIH in June in celebration of his successful open heart surgery five decades ago in Building 10. Kaminski was born with a tint of blue skin and blue fingernails. Due to a congenital heart defect known as tetralogy of Fallot, it was difficult for blood to flow to his lungs and for his body to get the oxygen he needed. He couldn't run around with friends in the neighborhood. He couldn't ride a bike. His parents decided to take action. In 1969, they brought him to one of the only places at the time doing heart surgery – the NIH Clinical Center.

"It was like somebody had taken two doors and opened them up and said, 'Okay, there's the world that you were not able to be a part of. Go ahead and be part of that world.' Now I could go play. Ride my bike. It changed everything," Kaminski said. Kaminski's wife Lyn joked, while flipping through her husband's complete medical record stacked high in front of her, that the best notes in the entire record was that the "effective correction of this child's cardio-malformation shows that he will derive good benefit". "50 years later, he certainly did," she said.

Pictured above, Dr. Laura Olivieri, a National Heart, Lung, and Blood Institute (NHLBI) research grantee and cardiologist at Children's National Hospital, Dr. Douglas Rosing, head of the NHLBI Cardiac Consultation Service, Kaminski's wife Lyn, Tom Kaminski holding teddy bear that was with him in the operating room and an astronaut toy his mother gave him after surgery, Kaminski's son J.T., Dr. Gail Pearson, associate director of NHLBI's Division of Cardiovascular Sciences and Dr. Richard Childs, clinical director of NHLBI's Division of Intramural Research. View more online: www.cc.nih.gov/about/news/newsletter.html

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Overall, sharing notes with patients can lead to improved communication, collaborative decision-making and enhanced patient engagement.

"For care providers, this will encourage us to think more deeply about the information we provide," said Dr. James Gilman, CC CEO. "The discipline needed to make notes more understandable can lead to greater insight to the best care options, as well as flag erroneous information and potential risk of harm."

Members from OpenNotes (www.opennotes.org), an organization promoting and studying the effects of

sharing more information with patients, visited the CC June 18 (<https://go.usa.gov/xyq5G>). They presented a session open to all, including patients. Topics covered included guidelines for writing notes, hearing a patient's perspective and further details regarding the CC's implementation. There was also a breakout session with behavioral health and pediatric providers.

Finalized progress, visit and other notes shared through the implementation of OpenNotes are now available in the Patient Portal.

- Donovan Kuehn

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Lost in the laundry: a patient's stuffed animal lamb goes missing

Pants? Check. Shirts? Check. Stuffed animal Lamb? Check. As 19-year-old Kayden Reinke filled his suitcase and set forth from Ohio to the NIH Clinical Center his beloved "Lammie" was packed and ready to accompany him as he faced the summit of his medical journey.

The 34-year-old lamb was passed down from Reinke's mother to him. Lammie is "at a point in her life where her original material isn't there," said Reinke's grandmother, Rosalind Janezic. The past few years Lammie hasn't been coming to all of Reinke's medical appointments but, "we decided this is important. Lammie is going to come along. She has literally got him through so many medical emergencies."

Last October, Reinke had a bone marrow transplant in hopes to treat his immune deficiency disorder. But just days after the transplant, the key member of his care team filled with fluff had vanished.

"I come out [of the shower] and she's nowhere to be found," Reinke recalls. "I checked every little area in the room. I checked underneath the bed. I checked behind the bed...gone! I thought she had disappeared."

"We were just so worried that Lammie was never going to be found," Janezic said. "This is the second time she's been missing in action. Kayden had a procedure done [elsewhere] when he was really little, and she was missing in the hospital. The first time she was found. This time, [we thought], there's a possibility we may not. I think we both cried at night thinking we might never see this special Lammie again."

Reinke's nurses on 3NE quickly called the one person who could continue the search – Chauncey Buford, the Manager of Laundry Services for the 200 bed hospital.

"When you throw stuff in a whole bunch of linens its chances of being found are really slim," said Buford, who has been working at the NIH for 30 years. He estimates only about 30% of items can be found and recovered. The Clinical Center sends out roughly 1,200 pounds of laundry a day to be cleaned.

The nurse explained the situation of the young man, the family significance of Lammie, and that Reinke was just in the early and fragile stages of recovering from his transplant.

"Right away I said to myself, you know what, this is one that I'm going to have to put my hands, feet, everything in [to continue the search]," Buford said.



Kayden Reinke, at the age of 2, holds Lammie.

That's just what he did. Taking infection control precautions, Buford dug through upwards of 30 bags of soiled laundry. But, no luck.

"I called over to the [laundry contractor] plant and explained to them 'I know I've called over for blankets and all kinds of stuff. But this one is a hot one. We have a young patient over here and that lamb has been in the family [for a while]. I gave them the whole nine yards so they became a part of [the search].'"

Then, fifteen miles away from the Clinical Center at the laundry plant, and nearly 72-hours since she went missing, the laundry contractor located Lammie.

"It felt much longer," Kayden said jokingly as he thought Lammie was lost closer to a week.

"One of the best things that could have happened while we were there, after his bone marrow transplant, was the return of Lammie," Janezic said. "We are so thankful to everyone involved to try to locate Lammie. We so appreciated it. Lammie is part of our family."

"I was just doing what I'm hired to do," Buford said humbly. "It's more than a job to me. It was a time for the Clinical Center to shine. That's what I'm always thinking when I'm in this office down here doing laundry. Which seems not to be a very important part at all, but I think no matter where we are and what we're doing we're connected to this symbol-

ism. If one person does it for the team, the whole team shines. It was strictly done to continue what NIH is doing every day and that's just laying the red carpet out for patients."

While the reunion between the family heirloom and patient was filled with hugs and tears – it was also filled with water. Lammie immediately was given a "water park in the dark" rinse in the washing machine at The Children's Inn at NIH after her long adventure, Janezic added.

Reinke, nearly 9 months post-transplant, says "things are going as good as they can be" with his health. He will come back for his one-year appointment in Fall 2019 – but this time, Lammie will stay safe at home.

- Molly Freimuth

Patient proposal pays tribute to the place that gave her a 'second chance at life'



On May 9, after completing her outpatient appointment, patient Marissa Amuso walked out to the common area in Building 10 to find her boyfriend Branden Bolivar on one knee asking for her hand in marriage. Amuso has been participating in clinical trials at the Clinical Center since 2000 and is currently participating in a trial for a haploidentical cord blood stem cell transplant. "Getting proposed [to] at the Clinical Center was very meaningful to me, not only to be surrounded by the best doctor, physician assistants and nurses (my transplant team), but the Clinical Center itself," Amuso said. "I have been coming here for years, and without this place, I would not still be here. NIH has become a second home to me, and my doctors and nurses feel like part of my family." Nuptials are expected in mid-2020.

The small study, led by Hall, admitted 20 healthy volunteers for one continuous month. In random order, for two weeks on each diet, healthy volunteers ate either meals made up of ultra-processed foods or meals of minimally processed foods.

Foods were considered “ultra-processed” if they had ingredients predominantly found in industrial food manufacturing, such as hydrogenated oils, high-fructose corn syrup, flavoring agents and emulsifiers. They included foods such as potato chips, sugar-sweetened drinks, processed meats and French fries.

For the unprocessed foods, the team used the NOVA classification. This system categorizes foods based on the extent and purpose of food processing, rather than in terms of nutrients. It included minimally processed food, especially plant foods like vegetables, fruits, nuts, whole grains, legumes, milk, eggs, fish and meat. For example, an ultra-processed breakfast might consist of a bagel with cream cheese and turkey bacon, while the unprocessed breakfast was oatmeal with bananas, walnuts and skim milk.

Previous studies published across the country were observational — looking at large groups of people that showed associations between diets high in processed foods and health problems. But because none of these studies randomly assigned people to eat specific foods and then measured the results, scientists couldn’t say for sure whether the processed foods were the culprit, or whether there were other causes.

“Though we examined a small group, results from this tightly controlled experiment showed a clear and consistent difference between the two diets,” said Hall. “This is the first study to demonstrate causality — that ultra-processed foods cause people to eat too many calories and gain weight.”

The NIDDK researchers worked closely with the Metabolic Clinical Research Unit, which is especially equipped to study issues involving diet and metabolism. Dietitians in the Metabolic Kitchen prepared controlled meals and matched nutritional components

in the processed and non-processed diets. Each diet contained the exact same quantity of calories, sugars, fiber, fat, salt and carbohydrates. The foods needed to be commercially available and listed in the U.S Food and Drug Administration nutrition database to provide accurate information about the nutritional content. Participants could eat as much or as little as they wanted.

This study was considerably shorter than most studies. It took less than a year to complete thanks to the amount of staff support in the Nutrition Department and Metabolic Unit. In addition, the only parameters for inclusion were age range and a certain Body

Mass Index. An even number of women and men were enrolled.

day, ate at a faster rate and gained weight. On average, participants gained 0.9 kilograms, or 2 pounds while they were on the ultra-processed diet and lost an equivalent amount on the unprocessed diet.

This is the first study that shows causality: ultra-processed food causes people to consume calories and gain weight. If you remove those ultra-process foods and give them the same calories, they report liking the food just as much, yet they lose weight, and they eat fewer calories.

Justin Butner, one of the study’s volunteers, said that the experience has caused him to think of processed food differently.

“Ultra-processed foods are so calorie-dense that feeling full meant I’d over-eaten.” Butner said. “Some days I would get through my meal in a few minutes without really noticing I was eating. It wasn’t satisfying.” In contrast, he found that unprocessed foods tend to be more filling and the process of eating more enjoyable.

While the study reinforces the benefits of unprocessed foods, researchers



Dr. Kevin Hall, center, and Dr. Stephanie Chung, right, talk with a healthy volunteer in the Metabolic Unit at the NIH Clinical Center. Photo credit: Jennifer Rymaruk

note that ultra-processed foods can be difficult to restrict. “We have to be mindful that it takes more time and more money to prepare less-processed foods,” Hall said.

“Just telling people to eat healthier may not be effective for some people without improved access to healthy foods.” However, the take-away is, as much as you can, try to avoid ultra-processed food and choose more whole foods. You can still like your food and it will be easier to lose weight.

One of the remaining questions is what is causing the increased calorie intake? It was not the components: sugars, salt, carbohydrates and fat.

“We need to figure out what specific aspect of the ultra-processed foods affected people’s eating behavior and led them to gain weight,” Hall said. “The next step is to design similar studies with a reformulated ultra-processed diet to see if the changes can make the effect on calorie intake and body weight disappear.”

The basic question for the researchers was - do people eat more on a processed diet than on an unprocessed diet? The answer was “yes.” On the ultra-processed diet, people ate about 500 calories more per

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- Debbie Accame