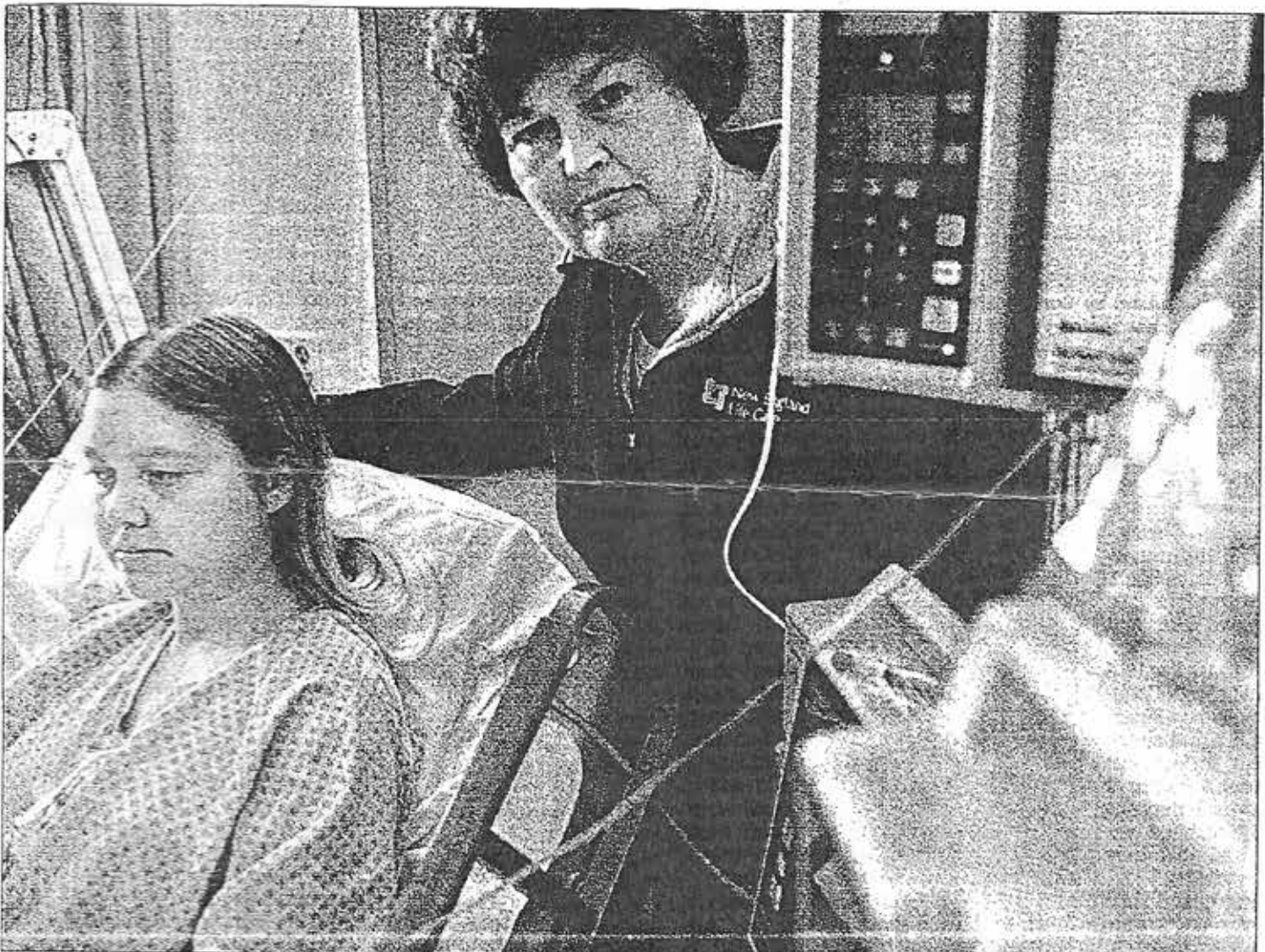


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"My body rejects my stomach, and there's nothing I can do," says 21-year-old Joy Weiss (left) from her room at Eastern Maine Medical Center. With her is her mother, Barbara Weiss. Joy has Crohn's disease, an inflammation of the gastrointestinal tract. While Crohn's is treatable in many people, Joy hasn't improved with medicine or surgery. "She doesn't have a life, basically," her mother says. (Photo by Kristen Schmid)

An effort to stem Crohn's

Cell transplant offers hope to Mainer with intestinal disease

By Misty Edgecomb
Of the NEWS Staff

BANGOR — Joy Weiss hasn't eaten in six days. The 21-year-old Mariaville woman is a patient at Eastern Maine Medical Center for the umpteenth time since she was diagnosed with Crohn's disease as a child, lying in a hospital bed with a morphine epidural in her spine and intravenous lines feeding nutrients directly into her bloodstream.

But today, Weiss is grateful for the gravity of her illness. Because of her struggle, she has been selected as the first person in the world to receive a stem cell transplant to treat — and perhaps cure — Crohn's disease. "I still can't believe that of all the people in the United States, I'm going," Weiss said. "I'm sick with Crohn's, but other people are dying. I could last another couple of years if I had to." Crohn's, like lupus and multiple sclerosis, is an autoimmune disorder, a particu-

larly nasty family of genetic diseases in which the sufferer's immune system is programmed to attack the body itself. Crohn's disease tells the body's white blood cells to rally against the digestive tract, which irritates the tissues, causing pain and impeding the body's ability to absorb nutrients from food. "My body rejects my stomach and there's nothing I can do," Weiss said. Weiss' illness began with stomach pains that her pediatrician first diagnosed

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Crohn's

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as food allergies or some kind of bacterial infection. She tried countless antibiotics, but nothing helped, so her doctor finally diagnosed Crohn's.

More than 50,000 Americans have the disease, according to a national research foundation. Some can control the disease simply by limiting their diet. So Weiss' pediatrician began removing foods that seemed to irritate her digestive tract.

"They said no dairy, no fried foods, no seeds or nuts, no salads and nothing with citric acid — so that's basically everything," Weiss said. "Sometimes I just would eat anyway."

But even with weeks of a soft, bland diet, Weiss often experienced severe pain that had to be dulled with morphine. Her doctor prescribed Prednisone, a powerful drug that reduced the inflammation, but with nasty side effects.

The drug is a "two-edged sword," said Barbara Weiss, Joy's mother, who is a registered nurse. While Prednisone is effective as an anti-inflammatory, it weakens tissues throughout the body, she said.

As a little girl, Joy Weiss ruptured her spleen, which had been weakened by the drug, in a minor fall from her horse.

As a teen, she had a port implanted in her chest so medications could be delivered directly to her superior vena cava (a large vein at the top of the heart) because her veins were so fragile that they collapsed whenever medication began to flow through them. The surgery caused one of her delicate lungs to collapse.

At 21, Weiss suffers from osteoporosis, but her body cannot absorb the calcium supplements prescribed to slow its progression. She is chronically anemic because she cannot absorb vitamins and minerals from her food.

Despite the medication and sur-

geries to remove badly damaged sections of her colon and intestine, until Weiss was 19, she could not absorb enough sustenance from her food to exceed a weight of 80 pounds.

"We've been in and out of the hospital so many times, it's a blur," said Barbara Weiss.

Each flare-up of Joy's digestive irritation results in weight loss that weakens her body and makes her digestive tract even more susceptible to attack. Even minor exertion, such as going out to a movie, can spark the pain.

"If I can do it, I'll go out with my friends, but I know I'll spend the next couple of days in bed," Weiss said.

"She can't have a life," added the elder Weiss.

Weiss frequently would have to begin an intravenous course of painkillers or nutritional supplements — not the easiest thing to accomplish in the school cafeteria or at a teen-age slumber party.

Sometimes she would endure the pain rather than start a morphine feed at school. "I'd just leave it in my backpack," Weiss said. "It sounds awful, I know, but sometimes I just didn't want to be bothered."

At her best, Weiss was able to teach horseback riding at a local stable. During one particularly difficult year, she recalls having attended only six more days of school than she missed.

She tried countless treatments, including Remicade, which has been touted as a "wonder drug" for Crohn's patients, said Weiss.

"You're supposed to get, like, a six-month remission — I got nothing," she said.

Despite the challenges, Weiss managed to graduate from Ellsworth High School with her lifelong friends in 1998. Friends, family and faith carry her through the tough times, she said.

"If I didn't have God to pray to, to help me out with this, I wouldn't last a day," she said.

But continuous pain in recent weeks led Weiss' doctors to suggest a colostomy, an operation that

would open her colon and force her to use an external waste bag for the rest of her life.

"Physically, I needed it, but mentally and emotionally I just wasn't ready," Weiss said.

Seeking any alternative, Weiss' doctor, Bangor-based pediatric surgeon Thomas McGill, went online and read about an experimental stem cell transplant proposed by world-renowned research physician Dr. Richard Burt at Northwestern University.

McGill was unavailable for comment Friday, but the Weiss family

"I have a little mare waiting, and she's going to need a lot of work."

— Joy Weiss

described his call proposing the treatment.

"Joy just about jumped through the phone," Barbara said.

Weiss traveled to Chicago last month to meet Burt and to undergo a series of tests, which indicated that she possessed the unique combination of severe symptoms and physical stamina that makes her a perfect candidate for the world's first autologous (meaning that the cells come from her own body) stem cell transplant for Crohn's.

If successful, the treatment could be a remedy for the incurable disease.

"If it will work for me — I've tried everything — then it will work for everyone else," Weiss said.

The treatment sounds simple. I will kill Weiss' defective immunity; then build her a new immune system using stem cells from her own bone marrow.

"If a person's immune system has become deranged, the concept is that you go in and destroy the entire system," Burt said.

But a person cannot survive

without immunity to disease, so doctors regrow the system using blood stem cells from the patient's bone marrow — primitive cells that have the potential to grow into any type of cell, including the white blood cells, or lymphocytes, that make up the backbone of the body's immune system.

"A stem cell doesn't know what it wants to be when it grows up," explained Patricia Manning, a registered nurse who coordinates stem cell treatments at EMMC.

Stem cell transplants have been used successfully for decades to rebuild cancer patients' natural immunity after intense radiation or chemotherapy treatments.

For a dozen years, Burt has been performing stem cell transplants to build new immune systems for other autoimmune diseases such as lupus, multiple sclerosis and rheumatoid arthritis with promising results. Logic suggests that Crohn's patients would experience the same miraculous cure.

Doctors conservatively estimate that the cost of such a transplant at \$150,000. This does not include the cost of transportation and lodging the mother and daughter will need for their monthlong Chicago stay.

Weiss' insurance carrier, Anthem Blue Cross, had denied coverage for the procedure, on the basis that stem cell transplantation had not yet been proven effective for Crohn's disease.

"From the information that we have, this procedure is experimental and investigative," said Clark Dumont, Anthem's director of communications for the Northeast. "Generally, clinical trials are not considered eligible for benefits."

But late last week, the family dropped appeals when they learned that Medicaid will cover the transplant's cost.

So Weiss will travel to Chicago in the next month, where she will take a combination of low-dose Cytoxan (a chemotherapy drug) and drugs known as growth factors. These medications cause a large number of stem cells to grow, then move

from her bone marrow into her bloodstream.

Burt's staff will send about 15 liters of Weiss' blood through a machine that removes the white blood cells and stem cells, then returns the blood to her body, through a mechanism that resembles kidney dialysis.

Weiss' stem cells will be frozen and preserved, and she will be treated with massive doses of Cytoxan essentially to kill her natural immune system.

Her preserved stem cells will be re injected into her bloodstream. Weiss again will be given growth factors to encourage the cells to reproduce, and they will develop into the white blood cells that the body lacks, and take up residence in Weiss' bone marrow, where they will provide her with natural immunity.

During the growth period, which will last several weeks, Weiss will have no immunity to disease, and must remain in a sterile room or risk dying from any number of infections.

If the procedure goes as planned, she will return home several weeks later, with a normal immune system that will protect her slowly healing digestive tract. Ideally, Weiss will be able to go off pain medication and will control irritation of any permanent damage with diet.

"Since this has never been done in Crohn's, we can't be absolutely sure what will happen," Burt said.

In the best-case scenario, Weiss' recovery could take months or even years as her embattled organs begin to heal. Burt will track her progress through yearly visits, and regular communication with Dr. McGill in Bangor.

Crohn's is known as a genetic disease, present in Weiss' DNA. So why would her cells behave differently after a stem cell transplant? Because Crohn's cause is far more complicated than the presence of a single gene. The interaction that causes the disease is so complex that, despite decades of research, doctors still do not understand it completely, Burt said.

"There's a genetic susceptibility, but because you have the genes doesn't mean that you necessarily get the disease," he said.

In studies of identical twins, one sibling developing the disease means that the twin will have a one-third chance of showing symptoms. Crohn's is caused by a chance and complex interaction of several factors that doctors don't yet completely understand.

A genetic predisposition is important, but so is gender (as a rule, women are more susceptible to all autoimmune disorders), age and exposure to an infectious agent.

A sophisticated type of white blood cell, called a T-cell, adjusts to attack whatever foreign agent is in its immediate vicinity. Its potential targets run into the billions. In a healthy body, this system can allow a person to build immunity to new diseases, Burt explained.

Basically, Crohn's disease occurs when a person's T-cells accidentally learn to recognize the cells of the stomach, intestines and colon as their enemy because they are exposed to the cells during a period when the T-cell is actively fighting a true intruder, he said.

Even if Joy Weiss responds positively to her stem cell transplant and develops a perfect immune system, she will be at risk to develop Crohn's again, should the correct combination of environmental factors occur.

"If people do get better following stem cell transplants, we still can't tell you exactly why," Burt said.

Weiss tries to restrain her excitement about the possibilities of the stem cell transplant — should the treatment fail, life-altering treatments such as a colostomy could be in her future.

But the 21-year-old survivor's eyes light up when she talks about going to college to study equine science, teaching riding and just climbing back onto her beloved horses as soon as possible.

"I have a little mare waiting, and she's going to need a lot of work," Weiss said. "There's a lot that I want to do."