

MEDICAL DISPATCH | JANUARY 20, 2014 ISSUE

LIVES LESS ORDINARY

Chronically ill children are living longer than ever. How should we care for them?

BY JEROME GROOPMAN

Gwen Lorimier is eleven years old and has been in and out of hospitals all her life. When she was one and a half, she was given a diagnosis of a rare disorder of the mitochondria, the tiny structures within cells that generate most of the body's energy.

Gwen Lorimier has more than a dozen doctors, overseen by a new kind of pediatrician.

PHOTOGRAPH BY ELINOR CARUCCI



Viewed under a microscope, her muscle fibres, rather than being neatly arranged in rows, are frayed and disorganized. In cases like Gwen's, the condition takes an especially hard toll on the liver, the gastrointestinal tract, and other organs and tissues that require large amounts of energy. But when we first met, in the dark-wood surgery library at Boston Children's Hospital, Gwen showed no outward traces of being sick. She has long chestnut hair, a bright face, and horn-rimmed reading glasses, and she was animated and uncannily sharp. When we sat down to talk, she glanced at her parents, Kim and Sue, to let them know that she was in charge of the conversation.

"I have an energy-deficit disorder, which means that I don't produce enough energy in my cells to power my body correctly," she said. "My organs don't work properly, which makes me have all sorts of problems, especially my stomach, intestines, liver, and pancreas. As one of my doctors put it, 'Her organs are trying to kill her from the inside.'"

Gwen has endured numerous life-threatening situations. When she was five, she developed pancreatitis. By seven, her vital organs were failing; at one point, she was placed on a ventilator. Gwen's doctors saw little hope for her and recommended that her parents take steps to make her comfortable before she died. Instead, they took her to Boston Children's for a risky multiple-organ transplant. The operation lasted twelve hours; Gwen received a new liver, stomach, small intestine, pancreas, and spleen. Five months later, she developed a potentially fatal, lymphoma-like condition called post-transplant lymphoproliferative disorder, which can occur in

patients who receive corticosteroids with other medications designed to prevent their bodies from rejecting transplanted organs. A CT scan revealed fifty tumors in Gwen's lungs, stomach, intestines, and kidneys. The surgical team reduced her immunosuppressive drugs and gave her a new medication, a monoclonal antibody.

Gwen's treatment reflects the advances that have been made in pediatric medicine in recent years. Until the nineteen-eighties, children born with cystic fibrosis often died before the age of ten. Today, with the advent of sophisticated drugs, they live well into their thirties and forties. Children with deformed hearts now live fuller lives following complex cardiac surgery. Cancers such as Hodgkin's disease and acute lymphocytic leukemia are typically eradicated by chemotherapy and radiation; if those fail, bone-marrow transplant is an option. Gwen is a new kind of patient: young, incurable yet not hopeless, and extraordinarily complex from a medical perspective. When I asked her to name the specialists who oversee her care, she listed more than a dozen, including experts in neurology, oncology, ophthalmology, gastroenterology, nephrology, cardiology, immunology, and genetics.

During Gwen's hospitalization, her parents recalled, the surgeons saw her briefly in the morning and evening, ordering more tests and medications, and consultants examined her and ordered still more tests. As days went by with no improvement, her specialists argued about how best to proceed. The transplant surgeons wanted to monitor her organs; an oncologist recommended immediate chemotherapy and urged that she be monitored in the cancer ward. Meanwhile, Gwen was in extreme pain. Each move onto a gurney and back onto her bed was excruciating, and none of the pain medications the doctors prescribed helped. Her parents, exhausted and unsure how to proceed, had heard about a group of pediatric specialists at Boston Children's and at the Dana-Farber Cancer Institute called the Pediatric Advanced Care Team, or PACT, which helps families like the Lorimiers to make difficult decisions.

Gwen's parents insisted that the team be called in. An attending physician and a nurse practitioner met with Gwen's specialists and worked out a plan for treatment: she would remain on the transplant floor and be given a combined regimen of the monoclonal antibody, chemotherapy, and adjusted doses of immunosuppressive drugs. To minimize painful movement, they coordinated the scheduling of tests and treatment. They made sure that Gwen received regular doses of analgesics as they explored the triggers for her pain. And they visited Gwen and her parents several times a day. They were "not like regular doctors," Gwen said. "They make an effort to say, 'Can I sit down and watch ten minutes of this show with you? Can I talk to

you?’ It’s not, Let me just stand over you and prescribe you some pills.” The treatment was a success. That summer, Gwen and her family vacationed on Cape Cod, and she spent a week at camp, where she swam and went horseback riding. For much of her life, Gwen had been fed intravenously. Now she enjoyed shrimp, lobster, ice cream, and red grapes.

The Pediatric Advanced Care Team is among a growing number of hospital programs that aim to bridge the gap in complex, chronic cases like Gwen’s between traditional family pediatricians, who come to know their patients over time and in depth, and high-tech specialists, who address specific ailments. Such pediatric palliative-care programs, as they are called, have three goals: to coordinate care, help families make difficult decisions about treatment options, and ease the child’s pain and suffering. One recent study found that, of the hundred and sixty-two hospitals surveyed, sixty-nine per cent had a pediatric palliative-care program, most of them established during the past decade. Nevertheless, staffing is so short that such programs care for only a small number of families in need. In November, the American Academy of Pediatrics recommended that all large health-care organizations that provide services to children with life-threatening conditions be given the benefit of a diverse team (made up of doctors, nurses, case managers, bereavement specialists, and others) that can address the physical, spiritual, and emotional needs of chronically ill children.

In many respects, the recommendation runs counter to the current direction of health care. Hospitals, doctors, and nurses, under increasing pressure to standardize care, are managed from the top down; treatments are determined by “pathways” and “algorithms.” This helps insurers and, arguably, some patients, but unique cases like Gwen’s aren’t addressed by these systems. “You are on one pathway, whether it’s the urinary-tract pathway or the heart pathway,” Chris Feudtner, an attending physician on the pediatric palliative-care team at the Children’s Hospital of Philadelphia, told me. “I take care of the kids when they fall off the pathway. People say, when I offer a treatment plan, ‘You are making this up.’ And I say, ‘Yes, I make this stuff up. Because we don’t have a path; we only have three kids on the planet who have this condition, or not even that. So I’m willing to stick my neck out and make it up.’ ”

Soon after meeting Gwen, I was invited by Christina Ullrich, an attending physician at Boston Children’s and Dana-Farber, to sit in on PACT’s Monday-morning rounds. She and eleven other PACT team members—including Joanne Wolfe, the program’s founder and director; Janet Duncan, the founding nurse practitioner; and Marsha Joselow, its social worker—met over coffee and bagels in a glass-walled conference room to review the cases of twenty patients, ranging in age from

a month to thirty-three years. (Adult patients are increasingly common at places like Boston Children's, as more patients with complex conditions diagnosed in childhood live into their thirties or beyond.) One baby was in intensive care after surgery for congenital heart disease. A fifteen-year-old girl who was being treated for leukemia had suddenly developed congestive heart failure. Two others had genetic disorders of the mitochondria, like Gwen, and were hospitalized for metabolic problems. One patient had died recently; during a break in the meeting, Joselow circulated a sympathy card for the family.

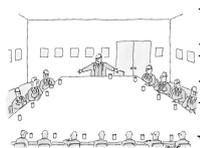
When Wolfe founded PACT, in 1997, it was one of the first pediatric palliative-care programs in the United States. Wolfe, who is fifty-one, was born in Montreal and studied psychology at McGill, then medicine at Harvard. One night during her residency training, at Boston Children's, a boy in the late stages of cancer was dying in the oncology unit; his family had brought him from the Middle East for treatment, but they had unrealistic expectations about his chances and had declined permission for a "Do not resuscitate" order. The ward resident told Wolfe, "We are going to do a slow code"—if he went into cardiac arrest, the staff would only go through the motions of resuscitation. Later that night, one of the boy's older brothers pressed the emergency button; Wolfe arrived at the bedside to see residents administering oxygen by mask (intubation is far more effective), and the brothers yelling. One of them turned to Wolfe, who was seven months pregnant, and cursed her unborn baby.

"It was a nightmare," she told me. The staff had failed to prepare the family. Ultimately, the attending physician transferred the boy to the I.C.U., where he died four days later. On another occasion, Wolfe was responsible for handling the pain medications of a boy with a disorder in which soft tissues effectively turn to bone. "He was in extreme pain," Wolfe said. "I was a graduate of Harvard Medical School, yet I had no idea how to relieve his suffering."

As she pursued pediatric oncology, Wolfe began to study how to help sick children and their families. One attending physician told her that she was wasting her career. Another said that she'd never be supported in her research. But Wolfe found allies in Stephen Sallan, the chief of staff at Dana-Farber, and Samuel Lux, the chief of hematology and oncology at Boston Children's, who shared her view that the care of young patients with serious illnesses could be improved; the Hasbro Children's Fund provided a three-year grant to help build the infrastructure of a new program. There was scant precedent for PACT's approach to pediatrics. "It was nonexistent," Wolfe said. "There wasn't a field."

In 2000, Wolfe published her first studies on the care of children with terminal cancer, in the *New England Journal of Medicine* and the *Journal of the American Medical Association*. Wolfe and her colleagues interviewed a hundred and three parents; eighty-nine per cent said that their children had suffered “a lot” or “a great deal” in the last month of their lives, and nearly a quarter were described as “often being afraid.” Of the children treated specifically for pain, just one in four was treated successfully, and only one in six was successfully treated for shortness of breath. Physicians were far less likely than parents to register and respond to a child’s distress.

*“Before we came
along, gentlemen,
all this was
nothing but
farmland.”*



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In recent years, new regulations have restricted the number of consecutive hours that residents are permitted to work. The aim was to create safer working conditions and lower the number of medical errors, but this has also made it hard for these doctors to provide continuous care. Primary-care pediatricians may

delegate in-patient care to a rotating cast of ward physicians, called “hospitalists,” who change shifts as frequently as every two weeks. When a crisis arises, there may be no doctor with sufficient knowledge of a patient like Gwen to coordinate her care or to help the family set realistic goals. Wolfe sought to create a team that could better organize care in this dynamic environment. In PACT, as in other pediatric palliative-care programs, the doctors have typically trained as specialists. Ullrich received her training in hematology, oncology, and bone-marrow transplantation. Such training enables physicians to advise with an authority equal to that of other specialists.

Many enter the field after having experiences like Wolfe’s. Sarah Friebert, the director of pediatric palliative care at Akron Children’s Hospital, in Ohio, served her residency, in the nineteen-nineties, at the Children’s Hospital of Philadelphia. “It’s a wonderful hospital, and we saved many lives, but I just was overwhelmed by the emotional, spiritual, and even physical suffering that I saw going on around me,” she told me. Marsha Joselow was a social worker in newborn medicine at Boston Children’s while her husband was being treated for lymphoma at Dana-Farber; he died at home, after a tormented few months. She encountered the PACT team when they invited her to attend a presentation in which, using her husband’s case as a prime example, they discussed how families should be better prepared to care for patients at home. Ullrich had been a junior resident at Boston Children’s in 2002, in the bone-marrow-transplant ward, where, despite the staff’s efforts, she felt that the needs of the children and

their families weren't being met. In 2005, she began her fellowship in pediatric palliative care, becoming one of the first doctors in the country to be formally trained in the discipline.

Ullrich, who is forty-one, has blond hair and wears rimless reading glasses. "Perhaps it sounds like reinventing the wheel," she said. "But the PACT approach was what I had hoped medicine would be. As a doctor, you focus on pathology and physiology; you learn pharmacology and how to order drugs. The social and psychological dimensions of care are often not well respected. But I learned that you can't treat a child's pain effectively without understanding her anxiety and her social situation. It's not just a matter of writing a prescription."

After Monday rounds, I went with Ullrich to visit Avery Nault, a seven-year-old boy with Leigh's disease, an inherited metabolic disorder that deforms the bones, causes muscle spasms and lung problems, and makes it difficult for the patient to hear and speak. Avery had been in the hospital for nine days, recovering from surgery to remove a segment of his bowel. His specialists included doctors in neurology, metabolism, orthopedics, and pulmonary medicine.

When we arrived at Avery's room, he was asleep, with an oxygen mask over his nose and mouth. Even as he slept, his arms and legs were bent at the joints—the result of excessive muscle tension, another by-product of the disease. We spoke quietly with his mother, Nicky, who works as an audiologist for the elderly; her husband is an assistant principal and the basketball coach at a high school in Lawrence, thirty miles north of Boston. The Naults have two other children: Janessa, who is thirteen and healthy, and Isaiah, who is eleven and has a potentially fatal mitochondrial disorder.

Avery was given a diagnosis of mitochondrial disorder at birth. At nine months, Nicky Nault recalled, he began to scream and didn't stop for two weeks. "We didn't know what was wrong with him, but we knew he was in some sort of pain, because all his limbs and toes were curling," she told me. "I was exhausted. I couldn't function, I couldn't even care for my other kids." When Avery was evaluated by the pain specialists at Boston Children's, they told Nault that, because Avery couldn't talk about his pain, there was no way to know what the problem was; it could be anything, they said, even a tooth coming in. "Then somebody slipped me a flyer about PACT," she said. When Joselow arrived at Avery's hospital room, along with a PACT doctor, the first thing she did was give Nault a hug. "I just broke down, because it wasn't only about Avery; it was about the family," Nault said. "If Avery is not sleeping, we are not sleeping, we are not functioning. I felt that was the turning point for me—to say, you know, I have somebody there for

us.” She relies on PACT when making hard decisions about Avery’s care. “They are very open and honest with me. I like that. I’m a very straight shooter, and I like the same back.”

Because Avery’s respiratory system doesn’t function properly, he may stop breathing suddenly, aspirate his saliva and choke, or develop pneumonia. In 2010, during an acute crisis, he was placed on a ventilator in intensive care. The Naults considered a tracheostomy for Avery, which would eliminate the need for emergency intubations. But the surgery would have left Avery tethered to a ventilator, and unable to play with his dog and roughhouse with his brother and sister. With PACT’s guidance, the Naults decided that a tracheostomy would preclude the activities that made Avery happiest. Janet Duncan, the nurse practitioner, said, “We bring a little bit of a different perspective, because we sit with families, who teach us about how they make decisions. It’s not that there is a right or wrong; it’s really what is the best decision for your family, for your child.”

Early on, Nault insisted that something be done to relieve Avery’s pain. “I describe Avery as being a used car. You take that car to the shop, and you can’t fix everything, so you have to have priorities about what can be fixed,” Nault told me. PACT takes an empirical, customized, and sometimes controversial approach to controlling pain, which can mean administering powerful drugs at very high doses. For Avery, they tried a number of medications until they found that a potent muscle relaxant called baclofen, given in combination with methadone and Valium, eased the worst of it. When, occasionally, his pain still flares, Nault says that emergency-room residents raise their eyebrows at the high doses of drugs like methadone.

Gwen had a similar experience in 2012, after four of her teeth were extracted. Extractions are typically straightforward, but anesthesia can disrupt Gwen’s metabolism and equilibrium, and prompt extreme nausea and sustained bursts of pain. “Pain takes a lot of my energy, and I don’t have extra energy to give,” Gwen told me. She emerged from anesthesia vomiting and in excruciating pain that could not be controlled in the recovery room. Ullrich recommended a powerful opioid, intravenous Dilaudid, every two hours, a regimen that, she concedes, “was pretty bold. It certainly isn’t what we’d do if we were doing things by the books. But there’s not much about Gwen that has ever been by the books.”

Gwen’s condition seems to have stabilized; she has not had a major hospitalization in nearly a year. (When I met her, at Boston Children’s, she had just had her monthly checkup.) But Avery’s visits to the hospital had become more frequent. Last year, he was able to attend kindergarten at a mainstream school. “He graduated, had the school play at the end of the

year, and had a big birthday party, and the whole class came,” Nault said. “I thought that was an accomplishment, that Avery went to school and got to see his peers.” But there was a cost: during the course of the year, Avery was hospitalized twelve times. “This year, I had to step back, and say, ‘You know, twelve admissions in one year—that’s not our goal,’ ” Nault said. “I feel O.K. about it. I gave him that one year in his life with so much interaction.”

Palliative care is often confused with hospice care, which focusses on the last few weeks or months of life. Feudtner, Wolfe, and their colleagues published a study in 2011 that found that most children in pediatric palliative-care programs are still alive a year after their initial consultation. Nonetheless, Wolfe told me, when PACT was launched the medical staff at Boston Children’s frequently referred to it as a death-and-dying group. She said, “We would walk onto a floor and people would make comments like ‘The Grim Reapers are here.’ ” Ullrich told me, “There’s this concern that if we get involved we’ll cause families to give up hope.” Medical specialists, who are inclined to be territorial, pose another challenge. “There does tend to be mistrust, and we need to be very careful in terms of how we present ourselves,” she said.

Caring for children with chronic illnesses is expensive, and pediatric palliative care is an additional financial burden. But the early data on such programs suggest that they are beneficial and cost-effective. In 2012, researchers at U.C.L.A. found that participating in California’s public pediatric palliative-care program reduced the number of days that a child spent in the hospital by a third, saving more than sixteen hundred dollars per child per month. The program has been extended through 2017.

Still, paying for such programs is a challenge. The care that Ullrich and others provide is time-intensive and often falls outside a hospital’s normal billing structure. A surgeon might bill a thousand dollars an hour for performing a tracheostomy, while lengthy conversations with a family about whether a tracheostomy is in a child’s best interest are reimbursed at a fraction of that amount. Pediatric palliative care “deserves to be valued as any other specialty, available to every child who needs it,” Sarah Friebert, of Akron Children’s Hospital, told me. But hospitals make money through admissions, and programs that emphasize the importance of home visits from social workers, chaplains, and others not typically considered part of the medical establishment don’t fit easily into the system. “As long as we live in a fee-for-service world, what we do isn’t going to be very popular,” Friebert said.

Meanwhile, Friebert has been lobbying Congress and the Department of Health and Human Services to support legislation that would provide funding for training in palliative care, including for pediatric patients. A section of the Affordable Care Act, called the Concurrent Care for Children requirement, ostensibly stipulates that Medicaid pay for palliative care for children who are eligible for hospice care. “But it doesn’t work at the state level, because various states have not successfully implemented it,” Friebert said, mainly because they are overwhelmed by higher priorities. Such care generally requires a physician to determine that a child is within the last six months of her life, should her condition follow its normal course—a complicated assessment to make in pediatrics. The Naults initially found that their insurance would cover Avery only if he went to a local hospital, which was unable to meet his medical needs. PACT negotiated on their behalf, so that he could be cared for at Boston Children’s.

“Ask them to be patient—baked pheasant carnivalé isn’t something we just throw together.”

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Timothy Quill, the director of the palliative-care program at the University of Rochester Medical Center, is more reserved in his advocacy. It “adds another layer of specialized care for seriously ill patients on top of an already complex, expensive health care environment,” he and a co-author wrote in a recent article for the *New*

England Journal of Medicine. Moreover, many elements of palliative care can be provided either by specialists—cardiologists, neurologists, oncologists, and the like—or by “generalist clinicians,” such as primary-care doctors. And, Quill wrote, bringing in a palliative-care specialist might be seen by a child’s other physicians as an opportunity to back off and provide less attention.

Quill believes that only the most complicated cases should be referred to palliative specialists. Even when a case requires the involvement of a doctor from a program like PACT, he says, the child should generally return to her original doctor for ongoing treatment. “Just as everyone who takes care of sick kids needs to know C.P.R., they should also know how to manage pain, and how to have conversations that involve tough decisions,” Quill told me. For that matter, he said, the entire health-care system should be reformed to “reintroduce the primary-care pediatricians as able to provide basic palliative care—defining their role with gravitas.”

Joanne Wolfe told me that palliative-care experts are working to share their approach with other medical staff. One easy change, she said, would be to improve the kinds of questions that pediatricians ask families upon first meeting them, particularly in the hospital. Typically, a doctor's first questions are narrowly focussed on the child's illness; instead, PACT recommends asking about the child as a person—how he or she contributes to the family, and how the illness has affected them—as a way of beginning to gauge and manage expectations. Questions with seemingly obvious answers—when asked what they hope for, parents almost always reply that they hope for a cure—can open the door a little bit for parents simply to talk. “You are giving permission for people to disclose their worries,” Wolfe said. Harvard Medical School has devoted half a day in the students' four years of study to hospice and palliative medicine. “It's a step in the right direction,” she said.

Unlike most doctors, PACT makes regular house calls. A few months after I met the Naults, I accompanied Ullrich on a routine visit to their home. Avery's older brother, Isaiah, was snuggling with Milo, the family's small dog, on the carpet. The garage and the ground floor had been refashioned into what Ullrich called a “home I.C.U.” Cabinets were filled with antiseptics, syringes, gloves, suction apparatus, tubing, and medicine bottles. The refrigerator held dosages of immunoglobulin (for Avery's low antibody levels), insulin, and an array of nutritional products. Avery, in his wheelchair, turned toward me and smiled. His mother positioned a hearing device in his ear and secured it with a Nike headband.

In the hospital, Avery had been sedated, his face covered by an oxygen mask. This is how residents, specialists, hospitalists, and other consultants often see such patients—uncommunicative, immobile, and in pain. There is no sense of what life is like for them at home. I was reminded of older patients with incurable cancer, whom I see in clinic. They describe their grandchildren, tell stories and jokes, discuss books they have recently read or films they've seen. But when they require hospitalization and are prostrate in bed, on oxygen, with intravenous lines providing fluids and nutrition, the staff sometimes sharply question me: “Why are we keeping him alive?”

Ullrich hugged Nault, then said hello to Avery, a handsome boy with black hair and a toothy grin. She checked a catheter above his collarbone, through which he receives nutrition and a muscle relaxant. Then she used her stethoscope to check for signs of pneumonia. Examining his abdomen, she noted that the scar from his recent operation had healed well.

We went into the den, where there was a Lucite box holding a basketball signed by his father's team at Central Catholic High School. On top of the box were two state-championship rings. "We have the players meet the boys," Nault said. "They're like another family. When Avery is in the hospital, they bring meals to the house." Photographs of the team with Coach Nault were displayed on a shelf. In one photo, Isaiah and Avery, smiling broadly, were seated in front of the squad.

Soon Lisa Kynvi, an older woman with short hair, arrived for a music-therapy appointment. She had a guitar slung over her shoulder, and she opened a bag that contained a tambourine and a maraca. She said hello to Avery, then placed belled cuffs over his arms. As she began to strum the guitar and sing, Avery moved his arms along to the music. "Avery has a lot there, but he can't speak," Nault said.

Ullrich and Nault reviewed their treatment options. "We regularly reset goals, and identify what the family hopes for," Ullrich had told me earlier. Now she talked with Nault about the possibility of surgery to treat Avery's worsening scoliosis, which will make it increasingly difficult for his chest to expand so that he can breathe normally. They had discussed the option several times. But the surgery was risky and would likely require follow-up procedures. Nault told Ullrich that they would decline. "I don't want to make any decision based on my sadness," she said. "It has to be about Avery."

Chronically ill children tend to be curious about the meaning of their plight and fearful of what might happen after death. "Even young children ask 'Why?'" Ullrich said. "Their inner world is filled with these questions, and you go where they take you, with honesty." The program provides younger children with books, such as "The Tenth Good Thing About Barney," by Judith Viorst, the story of a cat who dies and joins the family garden as the nurturer of new leaves and flowers. Through an initiative funded by the National Institutes of Health, older children, who are more likely to be stoic, can use a special computer program to express their feelings about their condition. Ullrich told me about a small boy who was dying of neuroblastoma. "His mother made it very clear to him that she would see him again in Heaven someday," Ullrich said. "But he was worried about how he would find her. So they made a plan to meet in the front left corner of Heaven."

A hundred and seventy or so children die each year at Boston Children's, and pediatric palliative care manages most of the cases. In the spring, the hospital holds a memorial service for children who have died in the past two years. Ullrich told me that this was helpful to the staff as well as to

families. PACT also holds individual bereavement meetings and stays in touch with families for two years after a death. “One family came back a couple of weeks ago, and couldn’t get their minds off thinking about everything that they thought they did wrong,” Marsha Joselow, the team’s social worker, told me. “We were able to step back with them and show the steps that led to that decision.” Typically, Joselow said, a parent “regrets that, if only they had called the doctor a week before, they would have known the diagnosis or complication and the child could have lived longer.” Or they feel that “we made this horrible decision to have a tracheostomy for our child. We thought it was best, but in reality we feel that we jeopardized their quality of life even if the procedure lengthened that life a bit.” Janet Duncan, the nurse practitioner, added, “We are often the ones to help these families remember that they went through a very thoughtful, careful process of what would help their child live as well as possible, and they did the very best they could with the information they had.”

Ullrich and Nault had moved on to discuss paperwork, including a new medical-order form for patients with advanced illnesses. The form is attached to the patient’s chart, to make difficult decisions easier in the event of a crisis in the hospital or at home; among the items listed were C.P.R., ventilation, intubation, dialysis, and such measures as artificial nutrition and hydration. Nault looked at it for a few long moments. “We haven’t gotten specific about cardiac medication,” she said finally. “But, obviously, if his heart stops, then it’s time to let Avery go. For Avery. He has fought such a hard fight; he will let me know when he is ready to go.”

Pediatric medicine has extended the lives of children like Gwen and Avery, but such patients are more fragile than most. Palliative-care programs like PACT help, in part, by narrowing the gaps in care where catastrophic failures can occur. The parents of both Gwen and Avery described instances in which PACT intervened to keep their children from being discharged from the hospital either prematurely or without proper medical oversight. “Avery wouldn’t be alive if it weren’t for PACT,” Nault told me more than once.

But programs like PACT do more than simply extend a child’s life; they work to insure that the extension is meaningful to the child and her family. Gwen told me proudly that, with her parents’ help, she has started a Web site, chronicallycool.com. It offers blog posts about her experiences, and a section with resources for parents and siblings, and she plans to add stories from other children with chronic illnesses. The site includes games with medical themes. “There is a heart-transplant one, and I found a game where you have to get all the mucus out of the lungs with suction.”

I asked Gwen what PACT does for her. “It’s like when you’re filling in concrete,” she said. “The transplanterers are the people who put the layer down, then PACT are the people who go after and fill the holes, so the whole thing doesn’t start to crumble. But if it does start to crumble they’re the people who actually go with the hard hats and fix it.” ♦



Jerome
Groopman, a
staff writer
since 1998,
writes

primarily about medicine and
biology.
