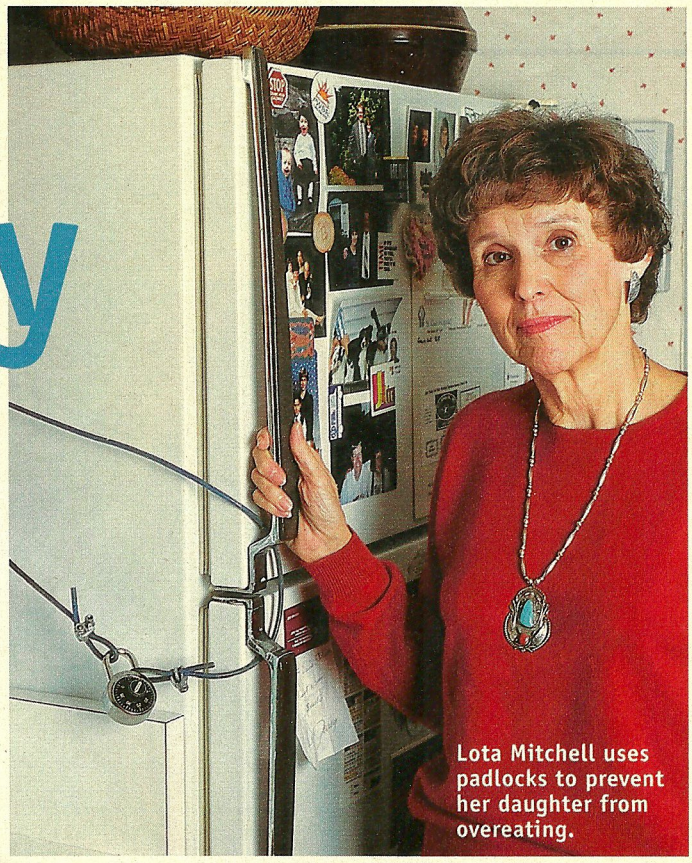
women who **make a difference**

Always Hungry

Telling the world about a tragic disease

By Barbara Klein



Lota Mitchell uses padlocks to prevent her daughter from overeating.

Lota Mitchell was already the mother of two healthy boys when she and her husband, Dave, had their third child in 1969. But Lota's joy over the birth of their daughter was soon overtaken by fear. "Julie was like a limp rag doll," Lota recalls. "She didn't cry and she showed no signs of hunger."

While mother and daughter were still in the hospital, a neurologist was called in for a consultation. "The first words out of his mouth were, 'Your baby is not normal,'" recalls Lota. "That's enough to knock you right out of bed." But just as upsetting, he didn't know what Julie had, or how to treat her.

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Julie struggled through her first years. Developmental milestones, like standing, walking and talking, were late in coming. Says Lota, "The hope was that she'd outgrow the problems, that she'd be normal. The hope went on until kindergarten."

By then, however, Julie was also exhibiting an ever-growing obsession with eating. No food could be left within Julie's reach, says Lota. The dinner table had to be cleared immediately and locks were placed on kitchen cabinet doors. Eventually the Mitchells were forced to turn their Pittsburgh home into a fortress.

For Lota, her husband, and their sons, David and Doug, living with Julie had become a 24-hour-a-day, seven-day-a-week struggle. Worse, the family still didn't know what Julie was suffering from, not until Lota read an article in the local paper that described the eating problems associated with Prader-Willi syndrome (PWS). Lota was shocked by the similarities. She would later learn that PWS is a congenital disorder of the 15th chromosome that not only causes an unrelenting feeling of hunger, but low muscle tone, short stature, incomplete sexual development, cognitive disabilities and behavioral problems.

Still, doctors were unable to offer much hope that Julie would be able to take control of her behavior and appetite.

"Julie had tantrums, and they only got worse as she got older," remembers Lota. "When she was a teenager she would yell and scream and then withdraw for days. It wouldn't matter where she was—at home, school or out in public. There was no way you could talk to her or make peace. You couldn't figure out what the tantrums were about; you could almost never see them coming," she says.

"By the time she was 12," continues Lota, "Julie started looking for food in the neighborhood. If she discovered an open door, she would help herself to whatever was around. I had no idea until my neighbors called to tell me they found Julie in their basement eating all of their ice cream."

These are classic signs of Prader-Willi syndrome, says Jeanne Hanchett, M.D., a pediatrician who has studied Prader-Willi. The earliest indicators are low muscle tone and the inability to cry or take food from a bottle; but at age two or three, a PWS child's appetite becomes insatiable. "It's not a drive for food any of us can understand or appreciate. They never feel full," she explains. "The tantrums are often related to food, but just as often related to nothing at all. Families are overwhelmed by them."

Lota, who was studying toward a master's degree in social work, started to write a paper about Prader-Willi in 1979. "I put together some practical advice ▶▶▶▶▶▶▶

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"It's not a drive for food any of us can **understand.**"

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about locking away food and clearing the table," she says.

A year later Lota attended a conference sponsored by the Prader-Willi Syndrome Association (USA). "The interesting thing was no one knew much of anything," she recalls. "The people who were professionals were busy asking questions of the parents, and the parents were asking questions of the professionals."

She so impressed the association that she was elected to the board. Lota's paper, which was revised to incorporate information from the conference, was published in the spring of 1981 and became a widely used resource for the organization. Lota now also had a new challenge—to get a medical center to offer help to PWS sufferers and their families.

In 1981 the Children's Institute in Pittsburgh (formerly known as the Rehabilitation Institute) generously created a four-week summer program for PWS kids. "I had called and told them that if you give me the services, I'll give you the people," Lota says.

That was just the beginning. Today, the institute offers a year-round comprehensive inpatient program for children and adults with PWS.

"Lota was instrumental in getting the PWS program started," says Dr. Hanchett, director of the institute's outpatient PWS clinic. "She was not only concerned about her own daughter, but all persons with the syndrome. She showed us the need."

Ken Smith, administrative manager of the inpatient program, underscores the institute's early role in trying to help PWS sufferers. "Thirty years ago families were left to fend for themselves," he recalls. But even now, "With no formalized dietary intervention, individuals with PWS are destined to get into trouble, with death being a very real possibility. It's not uncommon," says Smith, who is also chairman of the board of directors for the association, "for someone with the syndrome to gain 10 pounds over a weekend. Their muscles are weaker. They gain weight at an incredibly fast rate."

Children with PWS now use a growth hormone to increase their muscle mass and height, but ultimately, Dr. Hanchett says, "You can't overcome the syndrome, not with medication, surgery or behavior modification. Right now, we can't change or repair the chromosome."

Offering 24-hour-a-day supervision, the PWS program has a core staff of 15—including a psychiatrist, pediatrician, psychologist, nurses, dietitians, and occupational, physical and speech therapists. "We're able to deal with the medical complications associated with obesity, like diabetes and congestive heart failure, as well as the behavioral problems and the special dietary restrictions," Smith explains. "Our program is the only one of its kind in the world."

In 1995 at the age of 11, Whit Park of Oklahoma City, Oklahoma, desperately needed treatment for Prader-Willi syndrome. "He was in crisis," says his mother, Penny. "He weighed 235 pounds and his behavior was erratic. I believe he would not have lived."

Whit stayed at the institute for more than five months. "When he came home he weighed 140 pounds. He understood the diet and he learned to try to keep a grasp on his behavior," his mother says. "The program is probably the best in the country. It's incredible. They've taken the time, energy and dollars to truly find out about the syndrome."

The institute can accommodate only up to 16 patients at a time, who stay an average of 30 to 60 days. Since its founding, about 500 people from around the world, some as young as 4, have gone to the institute for help.

By the early '80s, Lota's life had settled into a more predictable routine. She had earned her degree in social work and had a position as an employee assistance program counselor. Julie was doing well. Then tragically, Lota's older son, David, died in a car accident. He was only 23. "Nothing good will ever come from my son's death," says Lota solemnly. "It will be with me until the day I die." But now, she says, she no longer mourns Julie's illness. "When Julie was born I grieved very much that she wasn't normal, and the grieving went on and on until David was killed. Now I can see the blessings Julie has brought into my life."

Lota appreciates the blessings Julie has brought into her life.

Even in tragedy, Lota has never stopped helping those with PWS. "Lota has been one of the most significant people within our organization," says executive director Janalee Heinemann, whose stepson, Matt, has the disease. "She was instrumental in getting me and many others involved. She's a great taskmaster."

For the past five years, Julie, 31, has been living with other PWS adults in a group home near Scranton, Pennsylvania and taking on jobs at a nearby sheltered workshop. Lota and Julie talk by phone at least once a week and, "We visit about a half dozen times throughout the year." Every now and again Julie attends get-togethers with the entire Mitchell family, including Doug, his wife and their four-year-old daughter.

"But if I'm not careful," says Lota about Julie's trips home, "if I don't keep the refrigerator locked, we're right back where we started. Prader-Willi syndrome doesn't go away. And it will never go away until there's some kind of medication that stops the drive for food."

Lota is proud of her daughter's accomplishments. "Julie isn't a complainer. She has seldom expressed how she really feels about having the syndrome. I know she certainly doesn't like it. Who would? But she has a social life. She has a work life. She's content." ■

For more information, visit the Children's Institute Web site at www.amazingkids.org., or www.pwsausa.org.