

A Day in the Life of **PIDD Patients**

No two PIDD patients are the same. All are affected differently by their disease, and their responses to IG therapy vary widely.

By Annaben Kazemi



Three patients, one disease, yet none functions the same as the other — despite all being treated with immune globulin (IG) therapy. This can be said for the thousands of individuals affected by primary immunodeficiency disease (PIDD). Some are highly functioning and living normal lives with only an occasional infection; others are marginally functioning due to frequent infections; and yet others struggle with so many complications that they are rarely able to function at all. Why does PIDD present so differently in patients?

The truth is that no two PIDD patients are the same. There are more than 180 types of immunodeficiency, each with a wide spectrum of symptoms affecting many systems of the body. Physicians diagnose PIDD based on poor or absent response to immunization, serum concentration of IgG (IgA, IgE, IgG and IgM) and the number of infections that are classified as recurring, persistent, debilitating and chronic.

IG is the standard therapy with the goal of allowing PIDD patients to lead full lives. But because the disease is so complex and varied, IG therapy varies in its ability to achieve adequate IgG trough levels to allow patients to fight off infection. For instance, one person may become severely ill with an IgG trough level of 700, whereas another might become that sick with a level of 600. This makes it a challenging task for immunologists to prescribe the optimal dosage of IG therapy to ensure an adequate IgG trough level for each patient. Complicating the dosage issue are the side effects and the infusion method — intravenous (IV) versus subcutaneous (SC), the former of which results in a greater variance in the IgG level during the infusion cycle.

In essence, there are no typical PIDD patients, and PIDD patients seem to have no typical days. So much depends on how they are feeling, what their complications are, how their bodies are responding to therapy, as well as where they are in the infusion cycle. This is illustrated by the stories of three PIDD patients whose lives are as varied as the disease itself.

Robert's Story

Robert, a busy 42-year-old executive and avid baseball fan from New York, was diagnosed as an adult with PIDD, but he doesn't think of himself as sick. Robert wakes up early each morning and gets in a 30-minute walk with his golden retriever before starting his busy day. He tries to maintain a healthy lifestyle by eating only organic foods, taking supplements and exercising daily. He power naps

most afternoons and is religious about getting enough sleep. He is able to go to work every day, help coach his son's baseball team and live "pretty normally."

Before Robert was diagnosed with a PIDD, he saw countless doctors for most of his adult life for recurring allergies, ear infections and chronic sinus infections. At one point, Robert's sinuses were so bad that he was told to have ear, nose and throat surgery. Within a month of his surgery, Robert had three more sinus infections. His allergist tested for an immune deficiency, and he was diagnosed with common variable immune deficiency (CVID). A year after being diagnosed, Robert began treatment with IVIG therapy, which he now receives at an infusion clinic every four weeks.

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Robert hasn't had a serious infection since starting IVIG, but he still checks in with his immunologist twice a year. When the occasional infection does occur, it is resolved with a prolonged course of antibiotics, but it doesn't impede his lifestyle as it used to. He states that the time he spends in the clinic (four to six hours a month) to get IVIG has completely revolutionized his world. "Sometimes having this disease is really horrible, and it can get you down," explains Robert. "The risk of infection is real; subsequently, a healthy or unhealthy amount of fear has accompanied my disease. But I've learned life goes on and it's mostly pretty good."

Now that he has experienced a prolonged period of wellness, Robert admits he is not as much of a germaphobe as he was when first diagnosed with CVID. He used to open every public door with a paper towel in his hand. He says he realizes that it could be much worse. "Much more serious infections like pneumonia can occur in CVID, but I've been rather lucky in that department," he says. "Those antibodies appear to have shown up for work."

He coaches his son's baseball team and is either out on the field or in the batting cage most nights of the week.

"I love baseball. I've either been playing the game or coaching just about my whole life. It's what I love to do," Robert emphasizes. "I hope that other people with immune disorders will see that I am an adult living vibrantly with the disease and use me as an example that it is possible to enjoy an active lifestyle despite being diagnosed with CVID."

Sarah's Story

Sarah, a 19-year-old college student who lives in the Pacific Northwest, had been sick her entire childhood and spent her adolescence filled with tests and doctors' appointments. After a serious hospitalization with pneumonia, Sarah was finally diagnosed with hypogammaglobulinemia when she was 14 years old, after it was determined that the pneumonia and flu vaccines had no effect on her immune system. Upon diagnosis, Sarah began treatment with IVIG therapy every three weeks. But because Sarah suffered severe migraines after IVIG therapy that kept her in bed for 24 hours at a time, affecting both her studies and lifestyle, she was transitioned to a twice-weekly regimen of SCIG.

On a typical day, Sarah wakes up and studies for a while, checks in with her online friends and updates her infusion log with how she is feeling. Sarah chose to go to a college close to where her parents live so that she can continue to live at home rather than in a dorm, where illness can spread quickly among even healthy students. Her mom is her biggest supporter and ally, and she checks in with her regularly. But her mom encourages Sarah to take ownership of her illness.

Sarah's SCIG infusions take about one hour. She prefers SCIG to IVIG because she feels it gives her more control and freedom. With her busy college schedule, she likes being able to infuse when it suits her time frame rather than scheduling an appointment. She also likes that it takes less time (one hour at a time as opposed to four to six hours each time in the clinic). However, there also is a downside. "It's kind of a drag to stick yourself or have someone else stick you at multiple sites; it's just not something you look forward to," Sarah explains.

Sarah has to see her immunologist every three months to monitor her levels while on SCIG. She sees her primary care doctor even more often, and she is especially susceptible to infections when she gets run down around the middle and end of each semester. Not being able to take rests when she needs them has taken its toll as well. She was treated for a respiratory infection last fall and ended



up on a nebulizer for several weeks. But it's the constant aches and pains that wear her out. According to Sarah, "Fatigue has been my worst enemy. On bad days, thinking too hard is like competing in an intense sporting event, and there are times I just don't want to get up off the couch."

While Sarah admits that since transitioning to SCIG her energy levels have been much more stable, she is still not as fully functional as she'd like to be. She doesn't go out as much as other co-eds, and she has to constantly turn down their invitations. It's hard when they say things like "But you look so good today" and "I can't even tell you're sick; aren't you better now?" No matter how hard she tries, she can't make her college friends understand how exhausting it is for her or that her disease isn't something that can be cured.

Jenny's Story

Jenny, a "58 and fabulous" grandmother from Nevada, went undiagnosed and misdiagnosed until she was in her early 50s. "I had been told for many years that I had other conditions instead," she said.

For years, Jenny had long periods of normal health and then was suddenly struck by high fevers, pneumonia or sinus and throat infections that she couldn't shake for months at a time despite several rounds of antibiotics. She eventually needed intravenous antibiotics. While her ear, nose and throat doctor was baffled, she continued to struggle through life, dragging herself to work, wondering

why she got sick so easily and so often. According to Jenny, the illnesses would just seem to “hit all at once,” leaving her fatigued and chronically worn out.

Unfortunately, while Jenny fought for a diagnosis, she faced a number of difficulties, among them misunderstandings and discrimination by her employer. Many people thought it was “all in my head,” Jenny says. Eventually, she had to go on disability because the constant exhaustion was causing her to miss so much work. In addition to missed workdays, the doctors’ visits, treatments and hospitalizations all were having a significant financial impact on Jenny’s family. And the insurance companies “just didn’t understand or seem to care.”

Jenny was diagnosed five years ago with acute chronic sinusitis, but it wasn’t until she still didn’t respond to treatment and was undergoing her fourth surgery that the underlying PIDD diagnosis was made. When treated with IVIG, she developed severe muscle pain, headaches and back pain. She was then switched to SCIG therapy, but she still experienced problems and could not tolerate the medication in large doses. Through a lot of trial and error, Jenny and her doctor decided she needed SCIG infusions on a daily basis at a very slow rate, which takes four to five hours.

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Jenny has been receiving daily SCIG infusions for four years now, and she still struggles with fatigue, although it’s not as debilitating as it used to be. Her days are centered around her infusions, and she always takes an afternoon nap. She does try to do a little bit of light housework, read or play “Words With Friends,” and she cooks dinner for her husband. But she misses out on a lot of social and family events because she just can’t deal with

the aftermath of exhaustion and infection. It has been a frustrating journey, and Jenny says “the worry has turned even my healthy periods into times of great anxiety.”

While Jenny doesn’t feel she has a “normal” life because she is isolated and rarely leaves the house, she concedes that it’s better than it was. “I’ve had to struggle with having this disease and all the discouragement that goes along with having it,” she says. “I still can’t do the things others can, but I lean on my faith as a source of support and encouragement.”

An Individual “Normal”

PIDD patients work, play, marry and have families just like people unaffected by disease. While a PIDD shouldn’t keep patients from living a normal life, the degree to which their illness affects their lifestyle does vary widely. As these stories illustrate, every person is affected differently.

A patient’s level of functioning depends upon the type and degree of antibody deficiency and the treatment regimen prescribed. While some patients, like Sarah, start to feel better and are able to begin normal activities after beginning treatment, other patients, like Jenny, continue to be greatly impacted by symptoms and complications even with treatment. Then, there are patients like Robert: His lifestyle is relatively unaffected by his disease; he’s never had to stop working, and his treatments are far less time-consuming than Sarah’s or Jenny’s. It’s important for PIDD patients to understand that it’s normal for different people to experience differing levels of functioning.

Because of the extreme variance in the disease, as well as in the response to treatment, patients need to be aware of their condition and advocate for access to trained specialists who understand their disease and are aware of the most recent developments in managing symptoms. Equally important is for patients to stay informed about obtaining and keeping health insurance coverage and about the laws and regulations that govern insurance. Education and awareness are key components to helping PIDD patients make informed choices so they can live their lives to their fullest potential.

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Editor’s note: The patients in this article are fictitious, developed through a series of composite interviews with identifying details changed to protect the privacy of individuals.