



The Connection Between Pulmonary Disease and Immune-Mediated Illness

By Jennifer Kester

Breathing problems due to pulmonary disease are often a cause of immune and autoimmune diseases, and can lead to serious complications if not diagnosed and treated early.

Since she was a child, Melissa Hauser frequently got sick. She suffered from repeated sinus infections and respiratory infections, and she came down with pneumonia at least once a year. Then, in 2005, she contracted pneumonia and she never healed. After checking out of the hospital, she landed right back there, and when she got off antibiotics, she had to go right back on them. In one month, she was forced to go on a hospital respirator three times. The problem: None of her doctors could



pinpoint the reason for her chronic health issues.

That changed when Hauser started college and she switched to a pulmonologist, a lung specialist, closer to her school, rather than continuing to see the one near her home in Vernon, Conn. When she met her new doctor, she told him she “felt compromised,” the 27-year-old recounts about that meeting. “I told him my symptoms, [and] then he did a blood test.” After years and countless tests that came back inconclusive, his simple blood test revealed that her hunch was right: She was diagnosed with hypogammaglobulinemia, which for Hauser, who had low immunoglobulin (IG) levels, was classified as a common variable immune deficiency (CVID).

What Is Pulmonary Disease?

According to a 2010 report from the American Society of Hematology, one in 25,000 to 50,000 people have CVID. Since it is a disorder that damages the immune system, patients can't fight

off infections, making them prone to recurrent sinus and lung problems. Pulmonary disease is one of the more serious CVID complications, and it is one of the leading causes of death for those with CVID. Also known as chronic obstructive pulmonary disease (COPD), pulmonary disease causes an airflow blockage that makes it hard to breathe. Emphysema, chronic bronchitis and, in some cases, asthma all are types of pulmonary diseases.

Although her diagnosis wasn't optimal, Hauser was relieved. “You know your body when something isn't right,” she says. But, until then, all the doctors she saw didn't believe her when she tried to explain something was really wrong. “I was told: ‘You don't know what you're talking about,’ ‘No, that's all in your head’ and ‘You are making it up.’” Even now, many doctors don't really understand. “If you're not a classic textbook case, [or] if you don't fit,” says Hauser, “the doctors automatically think ‘It must be in your head; you must be making it up.’”

PIDD and Pulmonary Complications

Like Hauser, some patients with an undiagnosed primary immune deficiency disease (PIDD) initially visit a pulmonologist to seek treatment for what they believe are breathing problems, only to learn they have PIDD. Pulmonologists are more likely to be aware of the possibility that a patient with recurrent respiratory problems could have immunodeficiencies, says Dr. Les Szekely, a pulmonary specialist at Doylestown Hospital, which is located about an hour north of Philadelphia. “We check for it a lot because it's sort of on our mind,” he says.

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“Once you've seen it and you've diagnosed it, treated it and had success, it sticks in the back of your mind.” Szekely says that immunodeficiencies are rare — he has only six patients who are prescribed IVIG — but he suspects that there are many more out there. “There are a lot of people who just aren't diagnosed,” he explains.

Even after a diagnosis, some PIDD patients continue to see a pulmonologist because of frequent pulmonary infections, as well as complications associated with the lungs. These PIDD patients are those who suffer from chronic bronchitis; asthma; granulomas (small areas of chronically inflamed tissue, such as from an infection), particularly chronic granulomatous disease, an immunodeficiency disorder resulting from an inability of phagocytes, like white blood cells, to kill harmful microbes; and bronchiectasis, in which the airways widen and become flabby and scarred.

Autoimmune Diseases and Pulmonary Complications

Some patients with autoimmune diseases who are treated with IG also have pulmonary complications, which require them to seek out a specialist. Those who have chronic inflammatory demyelinating polyneuropathy (CIDP) experience decreased muscle tone and activity, which

increases the risk of lung infections. People with Guillain-Barré syndrome (GBS) can suffer serious breathing difficulties with the onset of the disease. And, in more severe GBS cases, a ventilator may be necessary. In cases of myositis, one of the complications that can arise is refractory interstitial lung disease, a chronic condition that is unresponsive to treatment, causing swelling and scarring of the lung, affecting patients' ability to breathe.

For some autoimmune diseases, a pulmonologist is the primary specialist that a patient sees. One example is Wegener's granulomatosis, which causes blood vessels and other tissues to become inflamed, thus limiting blood flow to the organs and destroying normal tissue. Because one of the main organs affected in Wegener's granulomatosis is the respiratory tract, it's necessary for patients to see a pulmonologist. Another condition that requires a pulmonologist's care is sarcoidosis, which is the growth of clumps of inflammatory cells in different areas of the body — the lungs being one of the most common places.

Even people with severe asthma who don't have an immune disease also may be primarily treated by a pulmonologist. Research shows that patients with severe refractory asthma often respond to IG treatments.

Testing for Pulmonary Disease

For those with immune-mediated conditions, pulmonologists run a battery of tests to check for pulmonary diseases. The most basic is a blood test, such as what was done in Hauser's case, in which doctors check immunoglobulin levels. A CT scan of the chest gives a better look at the

lungs and thoracic lymph nodes than a simple X-ray. Pulmonary function tests are able to measure how well the lungs work, including how they expand and how much oxygen they can hold. One common test uses a spirometer, an instrument that patients breathe into, which gauges the air entering and leaving their lungs.

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Pulmonologists also look at oxygen saturation levels by measuring the amount of O₂ that is carried by the red blood cells. One method of measuring O₂ uses a pulse oximeter, a device that shines a light on the fingernail bed and calculates the amount of oxygen in the blood. This works because blood cells reflect different levels of light based upon the amount of oxygen they are carrying. Another form of testing doctors use is sputum, or phlegm, cultures. The pulmonologists check the mucus to see what types of organisms are colonized or causing an infection in the chest.

Treating Pulmonary Disease

IG is a common therapy for pulmonary disease, but there also are other therapies. To assist with breathing, asthma inhalers and nebulizers can be used. A pulmonologist also may prescribe corticosteroids to reduce swelling in the breathing tubes. If there is an infection in the lungs, antibiotics are usually recommended by the doctor. And, those who have difficulty getting rid of secretions in the chest are given devices to help break up the mucus and bring it up. Szekely says this is important for PIDD patients because leaving the chest congested with mucus can lead to pneumonia. Last but not least, patients with severe COPD and low oxygen levels may receive supplemental oxygen, which comes in portable devices and does not require a hospital stay.

Hauser's lung function is poor, and she is in constant



need of supplemental oxygen. She uses a concentrator, a machine that takes oxygen out of the air and condenses it for use by the patient. She also has portable oxygen tanks for when she leaves her home.

To further deal with her illness, Hauser is on intravenous immunoglobulin (IVIG). According to a 2006 report from the *Cleveland Clinic Journal of Medicine*, regular IV infusions help reduce the rate of infection and protect pulmonary function. Hauser started with once-a-month treatments, but the benefit wore off after a week, so two years ago, she changed to a weekly low-dose IVIG treatment. Each week, she devotes a full day out of her schedule to receive the treatment: She drives an hour to her doctor to receive the infusion, which takes six hours, and then drives an hour back home. "There are times when I don't want to do this, but you know you need the IVIG to survive," she says. "It's a double-edged sword: You need it and it keeps you healthy, but there are definitely things you have to give up."

One of the things Hauser had to give up was her aspiration to become a nurse, since being around sick patients jeopardizes her health. Then, after graduating from college in 2009, she wanted to get her doctorate in biology to conduct research, but that wasn't possible with her constant illnesses. Her frequent infections and other health issues also made it difficult for her to hold down a job. But, she hasn't given up on pursuing her dream of working in the medical field and is taking online classes to learn medical animation. "I know that there are some people who say, 'I wish I wouldn't have to work,'" she explains. "But I want to work; I pray for the day I can work a 20-hour week."

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Hauser still struggles to find the right combination of medicine to alleviate her symptoms. To help her breathe, she uses DuoNeb, a solution that she inhales every four hours through a nebulizer. She also takes a medication that breaks up mucus. Aside from her pulmonary problems, Hauser suffers from gastrointestinal ailments and joint pain, as well as severe migraines that get so bad she's been forced to go to the emergency room to seek relief.



"I found out that I have a bunch of health issues that doctors don't seem to be able to figure out," she says of her non-lung-related ailments. "Finding a doctor [who] will put them all together is really hard."

Szekely recommends that those who suspect they might have a pulmonary issue should get it checked out. "A thorough evaluation by a pulmonary specialist would be advised — it's a different look, it's a different approach," he says. "If you are having problems, seeing a respiratory specialist won't hurt. If you have pulmonary problems and aren't currently managed adequately or think you have any respiratory issues, seeing a pulmonologist would be a good idea."

Early Diagnosis Is Key

Hauser says the keys to avoiding major pulmonary problems are early diagnosis and proper treatment. "The longer you go without treatment and diagnosis, the more lasting effects [there are]," she says. "When you get respiratory infections that frequently, you get scarring in your lungs. The amount of infections I had caused my lung function to go down."

She tells people to go with their gut feeling if they think they might have a condition like hers. "My biggest piece of advice is trust yourself and advocate for yourself," she says. "A doctor can't tell you how you feel. You know your body best." ■

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