

# The Link Between PIDD and Skin Cancer

By Jennifer Kester



Research shows that PIDD patients have a 60 percent greater chance of developing cancer, and a handful of specific indications account for the majority of cases.

People who have primary immune deficiency diseases (PIDDs) are susceptible to a number of life-threatening health complications. One that should not get overlooked is skin cancer. According to the Skin Cancer Foundation, skin cancer is the most common form of cancer in the United States, with two million people diagnosed each year. Normally, when abnormal skin cells pop up, the immune system attempts to knock them out. But those with PIDDs have weakened immune systems that aren't

up for the fight, and the cancerous abnormal cells multiply. To make matters worse, the cancer and the chemotherapy wear out the already-taxed immune system even more, making it a vicious cycle.

## **PIDD and Increased Cancer Risk**

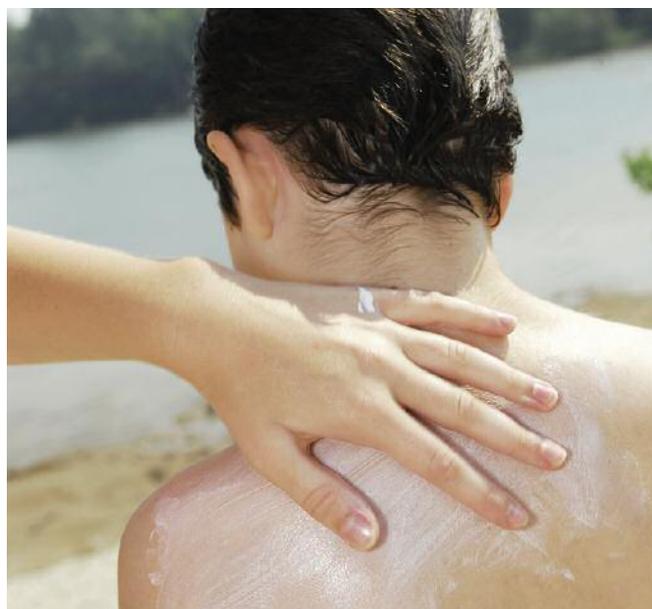
Because the survival rates for those with PIDD have increased in recent years, due in large part to effective immunoglobulin (IG) replacement therapies, researchers

have turned their attention to the higher rates of cancer among PIDD patients. According to a 2008 report in the journal *Anticancer Research*, cancer is the second-leading cause of death, after infection, for children and adults with some form of PIDD. The type of cancer occurring often depends on the patient's type of immunodeficiency, their age, as well as the kind of viral infections contracted. However, the most common type of cancer in PIDD patients is lymphoma, an immune-system-related malignancy. And, according to the Immunodeficiency Cancer Registry (ICR) database at the University of Minnesota, the most common malignancies for those with PIDD are non-Hodgkin's lymphoma (48.6 percent) and Hodgkin's disease (10 percent).

Research from the early 1990s suggests that approximately 25 percent of those with PIDD will develop cancer during their lifetime, a jump from the estimate of 2 percent to 10 percent reported in 1972, when treatment options were far less advanced. Based on estimates from patients in Australia, those with PIDD are estimated to have a 60 percent higher chance of cancer than the general population.

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There are approximately 150 subtypes of PIDD, only a few of which are correlated with cancer formation. According to the ICR, patients with ataxia telangiectasia (A-T) and common variable immunodeficiency (CVID) account for more than half of PIDD-related cancer cases, and one-third of cancer cases occur in patients with Wiskott-Aldrich syndrome (WAS), severe combined immunodeficiency (SCID) and selective IgA deficiency. A number of risk factors make PIDD patients more susceptible to cancer, including an impaired ability to respond to



agents toxic to cells' genetic materials, a decreased ability to remove pathogens, overactive inflammation responses and impaired control over virus-infected cells.

As with other forms of malignancies, CVID and A-T are associated with higher rates of skin cancer. In CVID, sufferers have low levels of antibodies, which make it difficult for them to stave off infections. Those with CVID sometimes contract systemic bacterial infections like meningitis and get frequent skin infections. In addition to skin cancer, CVID patients may also develop breast, prostate, gastrointestinal tract and lymphoid system cancers. Patients with A-T, a systemic disorder that affects the nervous system, have motor skills trouble, which leads to balance problems, as well as slurred speech and dilated blood vessels in their eyes and skin. A-T patients have a 37-fold higher risk of developing cancer than the rest of the population, according to the National Cancer Institute. In addition, they have a 10 percent chance of getting leukemia and lymphoma, and they are at risk for cancers of the stomach, brain, ovary, liver, larynx, parotid gland and breast.

#### **Decreasing Cancer Risk**

To decrease the risk factor of skin cancer, PIDD patients should heed the common-sense advice that applies to everyone: First and foremost, limit sun exposure. Not only does the sun cause skin cancer, it also further weakens the immune system. The Centers for Disease Control and Prevention recommends taking cover in the shade from 10 a.m. to 4 p.m., when the ultraviolet rays are the strongest.

If individuals do go out in the sun during that time, they should protect themselves by wearing a wide-brimmed hat, sunglasses that block the UVA and UVB rays (wrap-around shades are the best at preventing sunlight from seeping in) and loose-fitting clothing made with a tightly woven material. Using sunblock with an SPF of 15 or higher also will help shield exposed skin, and it's important to do so even on cloudy days and during the winter. Slather on the lotion a half-hour before going outside and then reapply every two hours. Of course, it should go without saying that tanning beds should be avoided completely.

## *Although not a lot of research is available on the link between skin cancer and PIDD, one promising avenue is vitamin D.*

However, what will go a long way in preventing skin cancer is to take care of the underlying problem, the immunodeficiency. Keeping the immune system in tip-top shape is a priority, so PIDD patients should be sure to practice good hygiene — such as using a mild soap to keep the skin clean and brushing teeth twice daily — and eat nutritious meals. Patients should also keep up their standard practices to ward off infections, like receiving regular IG treatments (or subcutaneous infusions) and steering clear of large crowds and people who have colds. Another alternative that can thwart a possible chronic infection is the use of long-term antibiotics in low doses, which will require a doctor's prescription.

Therapies offered for PIDD patients often are the same for other persons with the same kind of malignancy. However, shorter time in chemotherapy may be called for, given these patients' already-weakened immune systems. In addition, PIDD patients will need to work even harder to protect themselves from bacteria and other viruses while being treated.

### **The Cancer Research**

Although not a lot of research is available on the link between skin cancer and PIDD, one promising avenue is

vitamin D. A team of researchers found that the natural hormone is connected to immune reactions and could protect against cancer and autoimmune diseases. These researchers examined how the body stops the growth of pathogens and discovered the white blood cells that are integral to the body's ability to fight infections cannot function without adequate levels of vitamin D. However, that doesn't mean PIDD patients should freely bask in the sun just yet; the incidental sun exposure one gets from walking to the car or to the office is probably enough, and if not, there are vitamin D supplements available over the counter and through prescriptions. It is hoped that researchers will uncover more ways vitamin D can help PIDD patients fight cancer more effectively. ■

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