Frequently Asked Questions about Primary Immunodeficiency Diseases

What are primary immunodeficiency diseases (PI)?
Primary immunodeficiency diseases (PI) are a group of more than 250 rare, chronic disorders in which part of the body’s immune system is missing or functions improperly. While not contagious, these diseases are caused by hereditary or genetic defects. Some types affect a single part of the immune system; others may affect one or more components of the system.

Who does PI affect?
According to the Immune Deficiency Foundation (IDF), there are approximately 250,000 people diagnosed with PI in the U.S. Thousands more go undetected. Although some disorders present at birth or in early childhood, the disorders can affect anyone, regardless of age or gender.

How is PI diagnosed?
Medical and family history, physical exam, blood and immunoglobulin level tests and vaccines to test the immune response may be included in the diagnosis process. IDF estimates that the average length of time between onset of symptoms and diagnosis is between nine and 15 years. Fifty percent of those patients are 18+ years of age when diagnosed.

What are the symptoms of PI?
You should be suspicious if you have an infection that is:

- **Severe** – requires hospitalization or intravenous antibiotics
- **Persistent** – won’t completely clear up or clears slowly
- **Unusual** – caused by an uncommon organism
- **Recurrent** – keeps coming back
- **Runs in the Family** – others in your family have a similar susceptibility to infection

If any of these describe your infection, ask your physician to check for the possibility of a PI.

People with PI are more susceptible to infections and health problems that lead to serious and debilitating diseases. It is critical to get an early diagnosis and proper medical care.

How is PI treated?
Immunoglobulin (Ig) replacement therapy is the primary treatment for PI. In a recent IDF survey, it is estimated that 70 percent of those diagnosed with PI reported that they are being treated with immunoglobulin.

Why does IDF say THINK ZEBRA?
The PI community often identifies with zebras. This is based on an old saying. In medical school, many doctors learn the saying, “when you hear hoof beats, think horses, not zebras” and are taught to focus on the likeliest possibilities when making a diagnosis, not the unusual ones. However, sometimes physicians need to look for a zebra. Patients with PI are the zebras of the medical world. So IDF says THINK ZEBRA! Patients and family members can be seen wearing zebra striped clothes and using accessories adorned with black and white stripes. Children identify with zebras and it has been a way for children with PI to describe their disease to others—they are like zebras.

How can I find out more?
For more information, contact IDF at 800-296-4433 or visit www.primaryimmune.org