

# PRIMARY IMMUNE DEFICIENCY

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## Symptoms or Behaviors

Primary immune deficiency does not have unique symptoms. It is often mistaken for other illnesses. The infections can be chronic.

- Fever/chills
- Swollen glands
- Fatigue
- Pain
- Nasal congestion or drainage
- Headache
- Cough
- Muscle aches
- Rash
- Inflamed joints
- Nausea/vomiting/diarrhea

**Become familiar with symptoms specific to the student.**



## About the Disorder

A primary immune deficiency results when one or more essential parts of the immune system are missing or not working properly at birth because of a genetic defect. The immune system protects health in two important ways. First, it recognizes “foreign invaders” like bacteria, viruses and other infectious organisms. Once the invaders are recognized, the immune system reacts by sending special cells and proteins to eliminate the infection. This is called the immune response. The immune system isn’t located in just one part of the body. The thymus, liver, bone marrow, tonsils, lymph nodes, spleen, and blood cells all have a role in the immune system. These organs help make, store, and transport the cells and proteins that fight foreign invaders. The major cells and proteins of the immune system are B-lymphocytes (B-cells), T-lymphocytes (T-cells), phagocytes (eater cells) and special proteins. Because the immune system is composed of many different parts, there are many types of primary immune deficiency disorders with more than eighty currently identified. A primary immune deficiency disorder can be specific to one cell type or generalized to several components of the immune system. Some PI disorders have few symptoms, others are severe and may be life threatening. PI deficiency disorders can affect males and females of all ages and races. Frequency of occurrence varies from one in five hundred persons (IgA deficiency caused by faulty B cells) to one in a million (SCID or Severe Combined Immunodeficiency caused by abnormalities in both B and T cells).

Primary immune deficiency diseases do not have unique symptoms of their own. Warning signs that a primary immune deficiency may be present include frequent ear and sinus infections, pneumonias, meningitis and abscesses. The individual may be on antibiotics for two or more months with little effect.

**Diagnosis** is made by detailed medical histories, physical examinations, blood tests and vaccines to test the immune system.

**Treatment** includes aspirin or ibuprofen for fever and body aches. Decongestants are given to shrink swollen membranes in the nose, sinuses and throat. Expectorants thin mucous secretions and postural drainage helps to clear secretions from the lungs. Antibiotics are given to control or prevent infections and may be administered intravenously. In some life-threatening primary immune deficiency disorders, bone marrow transplantation may be done. For many people with antibody deficiencies, Immunoglobulin (IVIG) is administered intramuscularly or intravenously. IVIG infusions take two to four hours and are administered every two to four weeks. Prevention of infections by washing hands, eating a healthy diet and staying away from people with colds and other infections is essential.

A chronic illness can be difficult for the whole family. The student can have long periods of normal health and suddenly become very ill. Waiting for the next attack can result in great anxiety. Lack of public awareness about primary immune deficiency can isolate a family.

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## Educational Implications

The student with a primary immune deficiency disorder is highly susceptible to illness. They often have a high number of absences due to illness, hospitalizations and specialized treatments resulting in missed lessons. Infectious outbreaks such as chicken pox may further exclude the student from attending school resulting in more missed classes.

Side effects from medication can impact concentration, energy level and comfort, and subsequently academic performance (prior to infusions, they are weak and tired; after infusions, they may experience headaches, fatigue, backaches, chills, nausea, vomiting, hives and fever, lasting 2-3 days). Due to chronic illnesses, students are at risk for developing anxiety or depression. A plan to address the needs of the specific primary immune deficiency disorder needs to be developed and shared with staff. Staff may need help in understanding that primary immune deficiency is not HIV.

## Instructional Strategies and Classroom Accommodations

- Provide additional time to complete assignments
- Reduce number or length of assignments
- Provide tutoring/additional instruction to understand work missed while ill or out of the classroom
- Home based instruction may be needed (prevent illness when school has outbreak of communicable diseases)
- Provide a place to rest if fatigue is present
- Reduce physical distance student needs to travel during the school day
- An emergency healthcare plan needs to be developed addressing procedures if child is injured (scrapes, cuts), becomes ill or is exposed to children with infectious diseases (chicken pox, for example)
- Inform parents of communicable diseases to avoid unnecessary exposure
- Support good social relationships and self-esteem; observe for signs of anxiety or depression
- Provide private changing areas during PE for students with medication tubes who are uncomfortable changing in front of peers
- Teach and reinforce importance of covering nose and mouth when coughing and good hand washing techniques to classmates
- Should not have close contact with a person who recently received the flu mist vaccine, as it is a live virus.

## Resources

### **Immune Deficiency Foundation**

25 West Chesapeake Ave.  
Suite 206, Towson, MD 21204  
1-800-296-4433

[idf@primaryimmune.org](mailto:idf@primaryimmune.org)

*Booklet: Our Immune System  
(available from IDF)*

### **National Institute of Child Health Human Development**

Public Information and  
Communications Branch  
31 Center Drive, Room 2432  
Bethesda, MD 20892-2425  
301-496-4133

<http://www.nih.gov/nichd>

*When the Body's Defenses are  
Missing: Primary  
Immunodeficiency*

### **The Jeffrey Modell Foundation**

(Nonprofit research foundation  
devoted to PID)

1-800-Jeff-844

[info@jmfworld.com](mailto:info@jmfworld.com)

### **Howard Hughes Medical Institutes**, Office of

Communications, Arousing the Fury  
of the Immune System

[www.hhmi.org](http://www.hhmi.org)