

Your Body's Best Defense

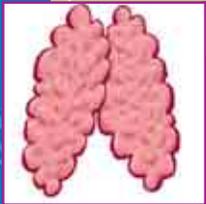


Every day, your body fights off infections, germs, bacteria and parasites. It is destroying these invaders, called *pathogens*, with the help of its number one defense: *the immune system*.

The immune system's job is to keep your body healthy. This requires many different parts of the body to work together against pathogens. Here are the main components of your immune system:



1 Tonsils - Located in the back of your throat, the tonsils protect the entrance to your respiratory and digestive systems by destroying bacteria with the help of white blood cells.



2 Thymus Gland - Located underneath the middle of your breastbone and above your heart, the thymus gland stores white blood cells until they are mature, and then provides them with specific jobs.



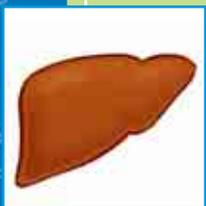
3 Lymph Nodes - Small, bean-shaped nodules on the lymphatic vessels, lymph nodes are located primarily in your armpits and groin regions. They filter pathogens out of the lymphatic system.



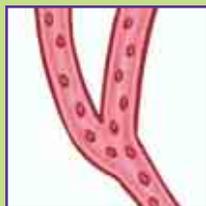
4 Bone Marrow - Located inside your body's bones, it produces red and white blood cells. *Red blood cells* carry oxygen to other cells and body parts, and remove carbon dioxide. *White blood cells* look for and destroy pathogens. The different types of white blood cells are phagocytes, B-cells and T-cells.



5 Spleen - The largest lymphoid organ in the lymphatic system, the spleen is located to the left of your stomach. The spleen removes pathogens from the blood as it passes through.



6 Liver - Your body's largest internal organ, the liver contains white blood cells. These cells destroy bacteria in the blood as it passes through the liver. It also processes nutrients found in the blood and produces bile used in digestion.



7 Blood - Red and white cells travel throughout your body's blood vessels. While white blood cells defend against pathogens, your red blood cells nourish your body.



Some people are born with immune systems that don't work as well as others. This might be caused by a genetic disorder called **Primary Immunodeficiency (PI)**, which might cause a person to be sick a little more often.

For more information, visit the Jeffrey Modell Foundation's Web site at www.info4pi.org or call 1-866-info-4-pi.

Now that you know how your immune system works, do your part to help it by keeping your body healthy with proper nutrition and exercise.



Jeffrey Modell Foundation

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WHAT IS PRIMARY IMMUNODEFICIENCY (PI)?

Dear Nurses,

You devote your life to helping others and do what you can to make sure your students are healthy and happy. When students are not feeling well, you might be the first person who treats them.

If there are students who are in your office regularly, ask yourself the following questions:

? Do the students you see frequently exhibit recurring illnesses without explanation?

? Are you familiar with Primary Immunodeficiency (PI)?

? Do you think your students' families are aware of PI?

PI is a genetic defect in the immune system that does not allow the body to fight off germs and viruses as quickly as a person with a healthy immune system. To date, there are more than 100 identified forms of PI that greatly range in severity. PI is characterized by chronic, recurring, persistent and/or debilitating illnesses that in severe cases, can be fatal.

PI affects close to 10 million people around the world, including more than 500,000 in the U.S. But how many people know about it? Not enough.

You provide information to parents about their children's health, and in turn, parents trust you as a reliable health source.

To help you provide parents with information on health issues facing children today, the Jeffrey Modell Foundation (JMF) and Lifetime Learning Systems have collaborated to bring you this free, educational program: **What is Primary Immunodeficiency (PI)?**

This program is designed to inform school nurses, educators and parents about PI – what it is and how it is detected – in order to raise awareness of PI so that it does not continue to go undiagnosed in children.

Included with this program are:

- ◆ four reproducible informational panels, including:
 - this letter to nurses
 - a letter to parents/guardians
 - an informational page about PI
 - a student activity page
- ◆ a poster of the immune system to hang in your office as a teaching tool for students (the poster is located on the reverse side of these information panels).

Please photocopy and distribute the information about PI to families and educators (in school mailings, with report cards, in teachers' mailboxes, at open house, during conferences or when parents come in to pick up their sick children, etc.). Please feel free to photocopy the activity page to distribute to students while they are waiting in your office, or send it home with the parents' information for students to do with their families. If you suspect the presence of a PI disorder, urge the family to visit the JMF Web site at www.info4pi.org.



If you have students who you see regularly, who are continuously afflicted with chronic ailments and take longer to recuperate from repeated illnesses, please do not rule out PI as a possibility for the child's recurring health problems. If parents know about PI and are educated about it, they can take action. Once informed, they can speak to their child's pediatrician about PI and will be able to rule it out as a possibility, or get help for an affected child.

Thank you for taking the time to learn about PI and educating others about it. For more information, please visit the Web site for the Jeffrey Modell Foundation at www.info4pi.org.

Sincerely,
Jeffrey Modell Foundation & Lifetime Learning Systems



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Please complete and return the enclosed survey to help us with future programs.

If you or a loved one has recently been diagnosed with PI, please contact the Jeffrey Modell Foundation at info@jmfworld.org for a free patient starter kit.

This publication was supported by grant funds from the U.S. Department of Health and Human Services' Centers for Disease Control and Prevention (CDC) and the generosity of Baxter Bioscience.



WHAT IS PRIMARY IMMUNODEFICIENCY (PI)?

Dear Parents/Guardians,

What you want most for your loved ones is for them to be happy and healthy. As a parent, you do what you can to make sure your child is feeling well, and you take care of your child when he/she is ill.

When it comes to your child's health, the more knowledge you have, the more you can help your child. That is why the Jeffrey Modell Foundation (JMF), in conjunction with Lifetime Learning Systems, developed **What is Primary Immunodeficiency (PI)?**, a program designed to inform parents about Primary Immunodeficiency (PI), including what it is and how to detect it.

PI is a genetic defect in the immune system. This genetic flaw does not allow the body to fight off germs and viruses as quickly or as regularly as a person with a healthy immune system. So far, there are more than 100 identified forms of PI that vary in severity. PI is characterized by chronic, recurring, persistent and/or debilitating illnesses that, in severe cases, can be fatal. One in every 500 Americans is diagnosed with PI; however, doctors estimate that PI goes undiagnosed in an additional one in every 500 Americans.

Here are some additional facts about PI:

- ◆ PI symptoms sometimes appear as a common infection (often of the sinuses, ears or lungs), a gastrointestinal problem or inflammation of the joints.
- ◆ Infections can be chronic, coming back often and with more severity, requiring more time for recovery.
- ◆ Males and females of all ages are at risk of having PI, since it is a genetic flaw in the immune system.
- ◆ Early diagnosis and treatment are necessary to prevent recurring infections from inflicting permanent damage.
- ◆ PI should not be confused with AIDS. PI is genetic, meaning the immune system's flaw is built into a person's DNA. AIDS is acquired by becoming infected with the HIV virus.

If your child exhibits any of these symptoms on a frequent basis, contact your pediatrician and ask him/her to rule out PI as a possibility. PI can be diagnosed with a blood test. Diagnosis and treatment begin with awareness.

Visit the JMF Web site at www.info4pi.org for more information about PI. You will find that it is an interesting and valuable resource for information.

Sincerely,
Jeffrey Modell Foundation & Lifetime Learning Systems



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10 Signs of PI

Primary Immunodeficiency (PI) causes children and adults to have infections that come back frequently, or are unusually hard to cure. If you or someone you know is affected by two or more of the following warning signs, speak to a physician about the possible presence of an underlying Primary Immunodeficiency.

- 1) Eight (8) or more new ear infections within one (1) year.
- 2) Two (2) or more serious sinus infections within one (1) year.
- 3) Two (2) or more months on antibiotics with little effect.
- 4) Two (2) or more bouts of pneumonia within one (1) year.
- 5) Failure of an infant to gain weight and/or grow normally.
- 6) Recurrent deep skin or organ abscesses.
- 7) Persistent thrush in mouth or elsewhere on skin, after age one (1).
- 8) Need for intravenous antibiotics to clear infections.
- 9) Two (2) or more deep-seated infections.
- 10) A family history of Primary Immunodeficiency.



If you or a loved one has recently been diagnosed with PI, please contact the Jeffrey Modell Foundation at info@jmfworld.org for a free patient starter kit.



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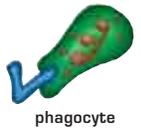
Primary Immunodeficiency (PI) causes children and adults to have infections that come back frequently, or are unusually hard to cure. In America alone, more than 500,000 people suffer from one of the 100 known PI diseases.



macrophage



lymphocyte



phagocyte

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How is PI diagnosed?

PI can be diagnosed with a blood test. A blood test will show the genetic makeup of the immune system and whether or not any parts are missing or not working correctly. An immune specialist, called an immunologist, might also choose to test your child's immune system with a series of vaccines to see how your child's body responds to the shots. A physical exam and an evaluation of family medical history may also be part of the path to diagnosis.

How is PI treated?

With more than 100 different types of PI, there are also different types of treatments. The earlier the detection of PI, the better. Some recurring infections can be treated with antibiotics to help prevent long-term damage and improve the patient's quality of life.

Another alternative for treatment is antibody replacement therapy, also known as IVIG therapy. It works by replacing the antibodies (proteins that help white blood cells fight and destroy germs) that the body isn't able to make on its own.

In other cases, bone marrow transplants, enzyme replacement or gene therapy may be appropriate.

What is the Jeffrey Modell Foundation?

The Jeffrey Modell Foundation (JMF) is a non-profit organization established in Jeffrey's memory by his parents, Vicki and Fred Modell. Jeffrey struggled with a form of PI for years, before succumbing to pneumonia in 1986 at the age of 15. The Foundation is dedicated to early and precise diagnosis, meaningful treatments and ultimately, cures for Primary Immunodeficiencies. Today, JMF focuses its efforts in research, physician education, patient support and public awareness.

The Jeffrey Modell Foundation believes that with treatment intervention, patients with PI will be able to gain or maintain control of their lives by:

- participating in work, school, family and social activities;
- decreasing the number and severity of infections;
- having few, if any, side effects from medications and other treatments; and
- feeling good about themselves and their treatment program.

Visit the JMF Web site at www.info4pi.org for more information about PI.

Your Body's Best Defense

Every day, your body fights off infections, germs, bacteria and parasites. It is destroying invaders, called *pathogens*, with the help of its number one defense: *the immune system*. The immune system's job is to keep your body healthy.

Finish the activities below to learn more about your body's immune system. The answers are at the bottom of the page.

The Truth About Your Immune System

Test your knowledge about your immune system by determining if each statement is true or false. Check off your answer.

- 1) Blood cells are produced inside your bones in the bone marrow.
 True False
- 2) Germs and viruses trigger your red blood cells into defensive mode.
 True False
- 3) Some types of bacteria live inside your body and help protect it from germs.
 True False
- 4) Your thymus, the gland where some white blood cells go to become T-cells, shrinks in size as you get older.
 True False
- 5) White blood cells sometimes attack healthy cells within your body by mistake.
 True False

Finding Out About the Immune System

Match the words below with their definitions.

- | | |
|------------------------|-------------------|
| 1. Immune system _____ | 4. Antibody _____ |
| 2. Fever _____ | 5. Pathogen _____ |
| 3. Tonsils _____ | |
- A) any foreign invader found in your body, such as a germ, virus or bacteria
 - B) located in the back of your mouth near your throat, they protect the entrance to your respiratory and digestive systems by destroying bacteria
 - C) the cells and organs that work together to keep your body healthy and fight off germs, bacteria, viruses and infections
 - D) a protein that white blood cells use to mark virus cells so that other white blood cells will know to attack the virus
 - E) a body temperature higher than 98.6 degrees Fahrenheit – sometimes when a virus is detected, your white blood cells will tell your brain to raise your body's temperature, making it too hot for the virus to multiply

For more games and activities, visit www.info4pi.org and click on NPI.

Immune System Order

The words below are scrambled. Put the letters in the correct order to find out the parts of the immune system.

pelsen

htmsuy

slintos

ylhpm oedsn

beon rarwom

Think About This!
How many words can you make using the letters in the phrase "immune system"?

Answers:
The Truth About Your Immune System:
 1. True. Bone marrow is the substance within the core of your bones where red and white blood cells are made.
 2. False. Red blood cells carry oxygen throughout your body and bacteria trigger white germs, viruses and bacteria trigger white blood cells into action.
 3. True. There is harmless bacteria that lives within your stomach and skin that helps to stop harmful bacteria from reproducing.
 4. True. Your thymus is large at birth, about the size of a lemon, but as you get older, it decreases to the size of two grapes joined together.
 5. True. Sometimes white blood cells attack healthy cells within your body, destroying an autoimmune disease.
Finding Out About the Immune System:
 1. C, 2. E, 3. B, 4. D, 5. A
Immune System Order:
 spleen, thymus, tonsils, lymph nodes, bone marrow