

Immune Deficiency Foundation

# Patient & Family Handbook

For Primary Immunodeficiency Diseases

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## Chapter 38

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For Primary Immunodeficiency Diseases

*6th Edition*

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110 West Road, Suite 300  
Towson, MD 21204  
800.296.4433

[www.primaryimmune.org](http://www.primaryimmune.org)  
[idf@primaryimmune.org](mailto:idf@primaryimmune.org)

# Chapter 38

## General Care

M. Elizabeth Younger, CRNP, PhD, Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

*The diagnosis of a primary immunodeficiency disease (PI) means different things to different people. For most, it represents both an end and a beginning. It is the end of what may have been a lengthy quest for answers to the questions: Why am I always sick? Why do I have more infections than anyone else I know? Why is my child sicker than his siblings or friends? This quest can involve multiple healthcare providers, a long period of diagnostic testing, and, perhaps, even a misdiagnosis. Nevertheless, once a diagnosis has been made, it represents a beginning—the beginning of a life spent moving forward while dealing with a chronic illness.*

*It is seldom necessary to make major life changes in response to a diagnosis of PI, but some modifications may be needed. The individual's new normal may need to be defined in the context of the condition. It is important to remember that most people with PI are able to live full lives. A diagnosis of PI is merely a part of that life; it should never become the life. Adopting a healthy lifestyle is the key to making sure that this is the case.*

### General Health Measures

#### Hygiene

General principles of good hygiene are essential for everyone, including people with PI and their families. This includes regular bathing or showering, and the use of soap. For some, the use of germ-killing soaps may be recommended. **Regular handwashing should be routine**—before and after eating, after blowing the nose, after coughing—anytime there is a concern that germs have gotten onto one's hands. It is important to remember that to be truly effective, hands must be washed vigorously for at least 15 seconds. This is generally longer than most people think. It usually takes 15 seconds to sing "Happy Birthday."

Individuals should avoid touching the nose or eyes, especially after touching a doorknob or flat surface. Viruses can remain alive on these surfaces for long periods and are easily transmitted from the hands.

When hands are not visibly dirty, alcohol-based hand sanitizers can be an alternative to hand washing. They have the advantage of being portable and easy

to use. These products have been shown to reduce the occurrence of colds and other viral infections. Individually wrapped disposable hand wipes are another great alternative to soap and water.

Tooth brushing and dental care are also key components to maintaining good hygiene. People with some immunodeficiencies are especially prone to gum disease and to infections that come from having decayed teeth. Regular visits to the dentist, as well as brushing and flossing multiple times daily, are essential.

A common sense approach to infection prevention is generally the best policy to follow. In addition to the maintenance of good hygiene, individuals should avoid exposure to people who have signs of illness or an obvious infection. This includes people who are coughing or have a fever. During flu season, it is wise to consider staying away from places where there are large crowds of people like a shopping center or movie theatre. With the recent measles outbreaks, it is also advisable to avoid social gatherings in which there is a group of parents who do not vaccinate their children. Some individuals choose to wear a

mask. It is best to consult the care provider about this. Masks tend to get moist from nasal or oral

secretions quickly. As soon as masks become moist, they are no longer effective. People who wear masks sometimes have a false sense of security and forget about other, more effective, preventative measures like frequent handwashing.

Many people with PI have questions about flying or travel. Depending on the specific type of PI a person has, travel to a particular place might not be advisable. For example, in some parts of the world yellow fever is a serious problem. The vaccine for this disease is a live viral vaccine and contraindicated for some people with antibody deficiencies. Travel to an area where there is yellow fever would not be a good idea for these individuals. Similarly, travel to a part of the world where healthcare services are not readily available might not be a particularly good idea. It is always wise for the individual with a PI to consult the immunology team when considering extensive travel. Consultation with a provider specializing in travel medicine may also be indicated.

## Nutrition

A healthy and balanced diet provides the elements necessary for appropriate growth and development, as well as body repair and maintenance. While good dietary habits are important for everyone, they are especially important for an individual with a PI. A

lack of adequate nutrition can predispose people to developing many illnesses, including infections for which someone with a PI is already susceptible.

Dietary guidelines for Americans encourage eating a variety of foods including starch and fiber, achieving and maintaining an ideal body weight, and limiting the intake of fat, cholesterol, sugar, salt and alcohol. (See Figure 38:1). The primary healthcare provider is an excellent resource for direction and advice regarding a healthy diet.

### Special Diets

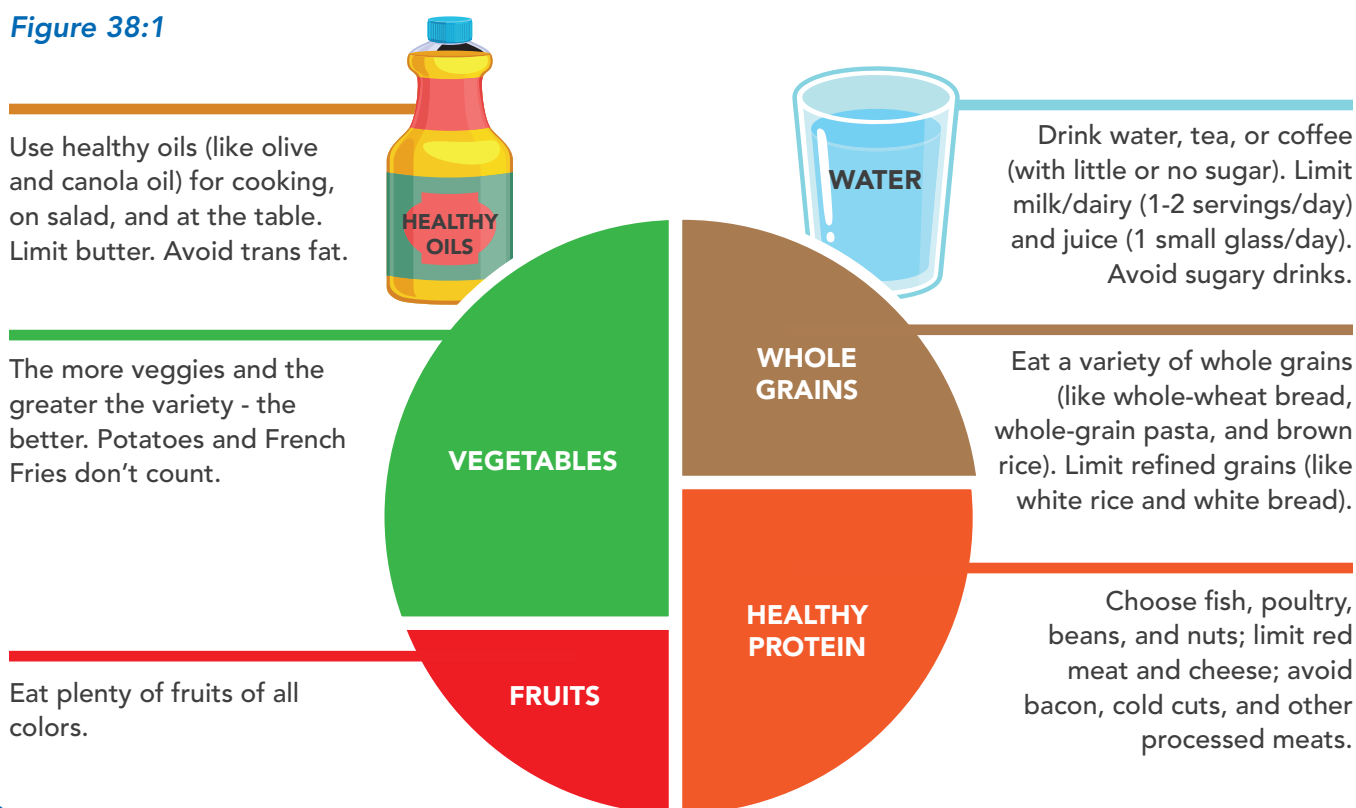
Unless the individual with a PI has another condition like diabetes, food allergies, or gluten sensitivity, special diets are not usually needed. During times of acute illness, though, dietary modification may be necessary. For example, when an individual has a gastrointestinal (GI) "bug" with nausea and vomiting and/or diarrhea, the healthcare provider may recommend a clear liquid diet. The provider will give instruction for the dietary modification.

### Special Dietary Interventions

In some circumstances, if individuals are unable to eat or drink normally or are unable to absorb nutrients from their GI tract, there are ways to assist them in getting and maintaining adequate nutrition.

Enteral nutrition, feeding directly into the stomach or small intestine via a special tube, may be recommended for those individuals who cannot eat

Figure 38:1



enough calories to ensure adequate nutrition or drink enough fluids to avoid becoming dehydrated. This method of feeding may be suggested for those who have swallowing difficulties, such as those individuals with Ataxia-Telangiectasia. It may also be a short-term solution for those who develop mucositis (inflammation of the mucus lining of the GI tract) during hematopoietic stem cell transplantation, also known as bone marrow transplantation.

Two common methods of enteral feeding are with a nasogastric (NG) tube or a gastrostomy tube (GT). An NG tube is a small, flexible plastic tube that is inserted through the nose and threaded down the esophagus into the stomach. In infants, these tubes can be passed through the mouth instead of the nose. A GT is a feeding tube that is surgically implanted directly into the stomach through the abdominal wall. These types of tubes can also be placed into the duodenum or jejunum, the upper two parts of the small intestine, and bypassing the stomach. Prescribed amounts and types of liquids are administered through the tubes either continuously or at regular intervals. There are many commercial products designed to be given in tube feedings. These products differ in the amounts of calories, nutrients, and the other components that they contain. The healthcare provider will decide which is the best product for an individual to use.

Total parenteral nutrition (TPN) and hyperalimentation are the terms used for nutrition administered intravenously. TPN is used to maintain the nutritional status of an individual who is very ill, malnourished, or unable to absorb nutrients from the GI tract. TPN solutions usually contain protein, carbohydrates, fats, electrolytes, vitamins, water, and essential trace minerals. Various types of intravenous catheters are used for TPN. Nutrition via TPN is usually a short-term solution designed to meet an individual's immediate nutritional needs.

### Nutritional Supplements

There are thousands of nutritional supplements available at supermarkets, specialty stores, pharmacies, and on-line. These include vitamins, minerals, herbal supplements, botanicals, probiotics, and naturopathic products. Many of these products are marketed aggressively and make claims to improve health by "boosting" the immune system. These supplements are not considered to be drugs by the United States Food and Drug Administration (FDA) and so are not FDA regulated. Virtually anything can be put into these products, and manufacturers can put anything they wish to on the labels.

There is no scientific data to support that any of these products improve health or strengthen the immune system. Extreme caution should be used before taking these products. Some of these products can be harmful or interact adversely with the medications an individual is taking. The healthcare provider should always be consulted before an individual takes any of these products. Sometimes the provider will recommend a pro-biotic, vitamin, or mineral supplementation. Some of these products may require a prescription. The important thing to remember is that these products are not a substitute for a healthy, balanced diet.

## Day Care

Families who have children with PI may need to use daycare just like those families with immunocompetent children. Unfortunately, all children in day care are exposed to lots of easily transmitted infections. It is not uncommon for a child to have seven to eight viral infections during the first year of daycare. While most of these infections are not serious and are self-limited, they, nevertheless, have an effect on quality of life as well as parental stress. Exposure to infections tends to be greater in larger daycares. Parents may choose smaller daycares or home day-care settings. They may choose to forego daycare, instead hiring a nanny or au pair. Parents need to consider carefully their options, weighing the benefits of daycare like providing the child with peer socialization, against the potential risks of infection.

## Exercise

A healthy lifestyle always includes exercise. Regular physical activity should be encouraged for all people—including people with normal immune systems and those with PI. Physical activity is good for the body and good for the mind, as well. Regular activity is an excellent stress and anxiety reducer. Activities such as swimming, biking, running, and walking promote lung function, muscle development, strength, and endurance. In general, people who get regular exercise are known to have fewer infections than those who do not exercise. Organized sports may be an excellent outlet for children who are struggling to cope with their illness. By competing on a team with other children, it may help the child with PI to feel that he or she is not so different and just a "regular kid" like everyone else.

Some kinds of exercise may be contraindicated for people with specific types of PI. People with

low platelet counts should not engage in contact sports. People with Chronic Granulomatous Disease (CGD) should never swim in fresh water. Likewise any individual with PI should be cautious about swimming in lakes or streams because of the increased susceptibility to a protozoa called Giardia.

The immunology healthcare team can recommend appropriate types of exercise if there is a specific concern. Nevertheless, everyone should get their steps in!

## Sleep

Getting enough sleep is critical for everyone, not just those who have a chronic illness. Adequate rest is an essential requirement for good health. Most research studies support getting a consistent number of hours of sleep each night as well as having consistent bed times and waking times. While sleeping in on a Saturday morning may be a special treat, it may not be the best thing to do to insure good health. Erratic or inconsistent sleep patterns have been shown to have negative effects on the immune system.

Some helpful sleep guidelines include:

- Going to sleep and getting up at roughly the same time everyday.

- Avoid late nights.
- Avoid consuming caffeine, such as coffee, tea, cola, in the evening.
- Avoid eating heavy meals in the evening or snacking right before bedtime.
- Minimize potential disturbances during the night (Don't go to sleep with the television in the bedroom on). Overall, limit screen time on any device before bed.
- Avoid long naps in the daytime that might interfere with the regular sleep schedule.

Adequate amounts of sleep are also important for children. They usually require more sleep than adults do. Children age 3 and younger require daytime naps in addition to their nighttime sleep.

## Stress

The notion that people get sick more often when they are under stress is supported by scientific data. Chronic illness alone is considered a major life stressor. Some studies suggest that stress has a negative impact on immune function and that reducing stress can actually improve

# Age Appropriate Sleep

### Table 38:1

Age	Average Sleep Requirement (in hours)
Newborn (0 to 2 months)	12 to 18*
Infant (3 to 11 months)	14 to 15*
Toddler (1 to 3 years)	12 to 14*
Pre-schooler (3 to 5 years)	11 to 13*
School-age (5 to 10 years)	10 to 11
Teen (10 to 17 years)	8.5 to 9.25
Adults (26 to 64)	7 to 9
Seniors (65+)	7 to 8

\*Includes nap

Adapted from the National Sleep Foundation Guidelines ([www.sleepfoundation.org](http://www.sleepfoundation.org))

immune function. Many stress reducers are easy to incorporate into one's daily life. These include massage therapy, biofeedback, meditation, and hobbies. Physical activity, even a walk around the block after lunch and adequate sleep, including a power nap can also help with stress reduction.

If you find that you are unable to deal with the stressors in your life, you should **absolutely** discuss these concerns with your healthcare providers. They can assist you or refer you to someone who can help you to deal effectively with your issues. You should **never** feel that nothing can help you to deal with the problems that overwhelm you and keep you from living and enjoying your life.

## Primary Care

Seeing a primary care provider regularly for health maintenance screening is important for everyone, but even more so for the person with PI. Many types of PI are associated with other illnesses. For example, it is known that people with Common Variable Immune Deficiency (CVID) have a higher incidence of autoimmune disease. The primary care provider who knows the individual well and sees them regularly may be the first person to recognize a symptom of one of these autoimmune conditions. Similarly, the first sign of a problem in a child may be seen by the primary care provider in the regularly assessed parameters that may indicate failure to grow or develop properly.

## Immunization

Perhaps the greatest advances made to improve general health over the past 225 years have been the development of vaccines to protect individuals from some of the worst diseases in our environment. Many people with PI, especially those with antibody disorders, have questions about vaccines.

People with many kinds of PI are fully capable of making protective levels of antibody to vaccines. Vaccines are very important for these people. They include people with complement deficiencies, phagocytic disorders like CGD, and even people with some antibody disorders like those with Selective IgA Deficiency. However, there are others with PI who are unable to develop protective immunity following vaccination. In some of these cases, the vaccine itself may pose a threat to the recipient.

People with CVID, Severe Combined Immunodeficiency (SCID), significant T cell deficiencies or agammaglobulinemia should never

receive live vaccines. These include the vaccines for chicken pox (varicella), measles (rubeola), German measles (rubella), mumps, smallpox, yellow fever, oral polio, oral Salmonella typhi vaccine, or rotavirus. These vaccines could actually cause the disease they are supposed to prevent in the people with these conditions. Infants with SCID are at the greatest risk for this problem. As some of these live viruses are shed in body fluids and stool for up to two weeks following vaccination, it might be necessary to limit contact between anyone recently immunized with these vaccines and an infant with SCID. For those people receiving immunoglobulin (Ig) replacement therapy, the infused antibodies should give them adequate protection against any shed virus. It's best to talk to an individual's immunology providers if there are any vaccine questions.

The usefulness of vaccination when one is receiving Ig replacement therapy is not fully understood, in part to the complexity of the range of underlying immune defects in those being treated with Ig replacement therapy. Assessing these peoples' antibody responses to vaccines is confounded by the antibody present in the Ig. However, it is possible that these individuals' T cells will be stimulated by inactivated vaccines, and therefore there is potential for some benefit. Thus, the viral influenza vaccine is generally recommended for individuals with CVID, and certain other individuals with antibody deficiency, such as IgG Subclass Deficiency, Selective IgA Deficiency and Selective Antibody Deficiency.

Some individuals with milder forms of PI affecting humoral immunity such as those with Selective IgA Deficiency, mild hypogammaglobulinemia, and partial DiGeorge Syndrome can safely receive all vaccines. Again, the immunology providers are the best people to advise regarding whether an individual should or shouldn't get a particular vaccine.

Purified protein, polysaccharide, or non-viable whole-agent vaccines pose no infectious risk to individuals who have problems making antibodies and are receiving Ig replacement therapy. However, for most vaccines the antibody response is not likely to be protective and these individuals rely on the antibodies they receive in their Ig for protection. For this reason, many immunologists do not recommend routine vaccination for individuals on Ig replacement therapy as they believe there is little benefit to be gained. New vaccine agents are the exception to this rule. For example, it is well known that the influenza strain changes from year to year. In a given year, there may not be a protective level of influenza

antibody in the Ig pool. Antibody to a new influenza strain will not be present in Ig replacement therapy for approximately two years, which is roughly the time between plasma collection to production of a lot of Ig. Many immunologists recommend an annual influenza vaccine for their individuals on Ig replacement therapy for this reason.

The Immune Deficiency Foundation (IDF) recommends that families of people with PI receive all recommended immunizations. The only exception is oral polio vaccine, which can be transmitted. If a close contact develops a rash after varicella vaccination, isolation of the person with PI is recommended with administration of zoster immunoglobulin. This is especially important in the case of highly communicable diseases like influenza, which change from year to year. Even if people with PI do not benefit from receiving the vaccine directly, they will certainly benefit from having a cocoon of protection from their immunized family members. (See Immunizations Chapter.)

## General Care during Times of Acute Illness

Even after a diagnosis of a PI has been made and appropriate treatment has been initiated, people with PI are still going to get sick. It is the hope that these illnesses are reduced in intensity and number, but it is unrealistic to think that they will be eliminated. When acutely ill, people with PI should always:


- Seek medical advice. Do not ignore symptoms such as a cough or fever and think they will just go away.
- Never self-treat. Taking leftover antibiotics or those prescribed for another family member is never a good idea.
- Follow the healthcare providers' advice. If 14 days of antibiotics are prescribed, take them all. Do not stop after a week because things have improved. If there is a recommendation to stay home from work or school, then stay home.

Undertreating or trying to ignore an acute illness may seem okay in the short run, but it can absolutely have long-term negative consequences.

## Summary

The diagnosis of a PI is a life-changing event. It can be viewed in a positive way. The diagnosis and treatment are the first steps on the road toward wellness and an improved sense of well-being. Adopting a healthy lifestyle and complying with the advice and recommendations of the entire healthcare team can maximize the potential for a normal, full life.

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