

General Care



Chapter 23

The diagnosis of a primary immunodeficiency disease means different things to different people. For most, it represents both an end and a beginning. It is the end of a quest for answers to the questions: Why am I always sick? Why do I have more infections than anyone else does? Why is my child sicker than his brothers, sisters and friends? Sometimes this quest can take a very long time, involve many care providers and lengthy diagnostic testing.

Nevertheless, once the 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 the diagnosis of a primary immunodeficiency disease, but some modifications may be needed. Remember that most people with primary immunodeficiency diseases are able to live full and (relatively) normal lives. Adopting a healthy life style is the key to insuring that this is the case.

General Health Measures

Nutrition

A healthy diet provides nutrients essential for normal growth and development, body repair and maintenance. While good dietary habits are important for everyone, they are especially important for an individual with primary immunodeficiency disease. A lack of adequate nutrition can lead to many illnesses, including infections for which the individual with primary immunodeficiency disease is already at risk. Dietary guidelines for Americans encourage eating a variety of foods, maintaining an ideal body weight, consuming adequate starch and fiber and limiting the intake of fat, cholesterol, sugar, salt and alcohol. (See *Figure 1.*) The primary healthcare provider is an excellent resource for direction and advice regarding a healthy diet.

Special Diets

Unless the individual with primary immunodeficiency disease has another condition, like diabetes, gluten sensitivity or congestive heart failure, there is usually no need for a special diet. However, in times of acute illness, there may need to be some modification of the regular diet. For example, when the patient has an intestinal infection, like a “GI bug,” a diet of clear liquids while the patient is having nausea, vomiting and/or diarrhea may be recommended. The primary healthcare provider will give recommendations, direction and instructions when these modifications are necessary.

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Special Dietary Interventions

In some circumstances, if patients are not able to eat or drink normally, or if they can eat but are unable to absorb nutrients adequately from their stomach and intestines, there are ways to assist them in maintaining adequate nutrition.

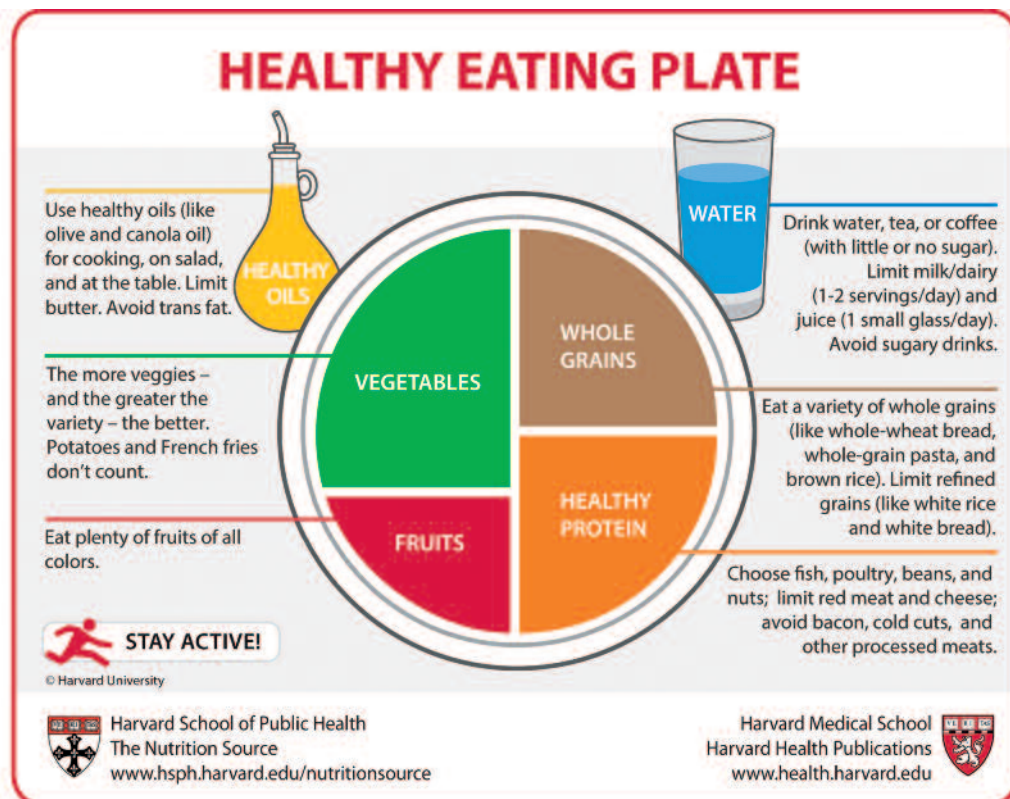
Enteral nutrition, feeding directly into the stomach or intestine with a special tube, may be recommended for patients who are unable to eat enough calories to insure adequate nutrition or drink enough to maintain adequate hydration. This method of feeding may also be suggested for patients who have swallowing difficulties, such as those patients with Ataxia-Telangiectasia. (See chapter titled "Ataxia-Telangiectasia.")

Two common methods of providing enteral nutrition are with the use of a nasogastric (NG) tube or a gastrostomy tube (G-tube). A nasogastric tube involves placement of

a small, flexible plastic tube through the nose, down into the esophagus and then to the stomach. A gastrostomy tube involves the surgical placement of a feeding tube through the skin of the abdomen directly into the stomach. It is also possible to place a tube directly into the duodenum or jejunum, which are the upper two sections of the small intestine, bypassing the stomach. A prescribed amount of liquid feeding is administered through the tube continuously or at regular intervals. Various commercial formulations are available to provide balanced amounts of calories, fats, proteins and carbohydrates, as well as other necessary minerals and vitamins.

Total parenteral nutrition (TPN) and hyperalimentation are the terms for nutrition administered intravenously. Solutions containing all essential nutrients, fluids and calories are delivered directly into the blood stream, bypassing the stomach and intestines. TPN is used to

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maintain the nutritional status of an individual who is very ill, malnourished or who is not able to absorb nutrients from their digestive tracts. The TPN solution usually contains protein, carbohydrates, electrolytes, vitamins, water and essential trace minerals. Fats may be supplied in a separate solution. Various types of intravenous catheters are used to administer these solutions. Nutrition via TPN is usually a short-term solution to meet the patient's immediate nutritional needs.

Nutritional Supplements

There are thousands of nutritional supplements available on the open market. These include vitamins, herbal supplements, botanicals, probiotics and naturopathic products. In some cultures, use of herbal supplements is especially common. Many of these products are aggressively marketed and make claims to improve health by “boosting the immune system.” These products are not considered “drugs” by the United States Food and Drug Administration (FDA) so are not FDA regulated. The claims of improving health or strengthening the immune system are not based on scientific data, and virtually anything can be claimed or put into these products.

There is no scientific evidence that any product will boost the immune system or make it stronger. Extreme caution should be used when considering taking any of these products. Some of these supplements can be harmful or interact adversely with prescription medicines the individual is already taking. The healthcare provider's opinion should always be sought before taking any of these products. Sometimes the provider will recommend vitamins, electrolyte supplementation or probiotics for certain patients but remember that supplements are no substitute for a healthy, balanced diet.

Hygiene

General principles of good hygiene are essential for patients with primary immunodeficiency diseases and their families. This includes regular bathing or showering and the use of soap. For some patients, the use of special germ-killing soaps may be prescribed. Regular hand washing should become routine—before and after meals, after using the bathroom, after blowing the nose, coughing—any time there is a concern that excess germs have gotten onto one's hands. It is essential to remember that to be truly effective, hands must be washed vigorously with soap and water for at least 15 seconds, which is generally longer than most people think. It usually takes 15 seconds to sing “Happy Birthday” once or the “Alphabet Song” twice.

When hands are not visually dirty, alcohol-based hand sanitizers can be an effective alternative. These have the advantage of being able to neutralize germs, are portable and can be applied rapidly. The regular use of hand gels has been shown to reduce the occurrence of colds and other viral infections. Individually wrapped and disposable hand wipes are another alternative to soap and water and are excellent for school lunches and for outings.

Some individuals with a primary immunodeficiency are prone to tooth decay and to infections that stem from having decayed teeth. Regular visits to the dentist, proper brushing and flossing should be a key part of the regular health regimen.

A common sense approach to infection prevention is generally the best policy to follow. Individuals with a primary immunodeficiency should avoid exposure to people who have signs of an obvious infection, like people who are coughing, have a fever or have vomiting and/or diarrhea. During periods of influenza outbreaks, it might be wise to avoid crowded areas such as shopping centers and movie theatres. Many patients with primary immunodeficiency disease people have questions about flying or other travel. When in doubt, ask the immunologist or primary healthcare provider for advice.

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Day Care

Families with young children who have a primary immunodeficiency may need to use day care just like everyone else. Unfortunately, children in day care are exposed to many infections that are easily transmitted. While most of these infections are not serious, of course they reduce quality of life and impact parents' education, work and stress. Exposure to infections tends to be greater in large institutional day care settings. Depending on the degree of immune compromise and the effectiveness of therapy, parents may want to consider options with smaller numbers of children or in home day care, if these are available.

Exercise

A healthy life style always includes exercise. Physical activity should be encouraged for all people, immunocompetent or immunodeficient. Not only are these activities good for the body, they are good for the mind, as well. Regular exercise 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 are physically fit and participate in regular exercise are known to get sick less than people who do not exercise. Organized sports may be an excellent outlet for children who are struggling with coping with their illness. Playing on a team with immunocompetent children may help the child with an immunodeficiency feel that he or she is not so different and is "just a regular kid" like everyone else.

Some kinds of exercise may be contraindicated for people with specific immunodeficiencies. For example, a boy with Wiskott-Aldrich Syndrome who is known to have a low platelet count should not engage in contact sports. People with Chronic Granulomatous Disease (CGD) should never swim in the ocean or fresh water. The immunologist can recommend appropriate types of exercise for their patients.

Sleep

Getting an adequate amount of sleep is an essential requirement for good health. Most scientists recommend a consistent number of hours of sleep per night and consistent bed times and waking times, as well. While "sleeping in" on a Saturday may seem like a special treat, it may not be the best thing to do to insure good health. Erratic sleep patterns have been shown to have negative effects on the immune system. Some helpful sleep guidelines include:

- Go to sleep and wake up at roughly the same time each day.
- Avoid late nights.
- Avoid consumption of caffeine (such as caffeinated coffee, sodas or tea) or alcohol in the evening.
- Avoid eating heavy meals in the evening or snacking right before bedtime.
- Minimize potential disturbances during the night.
- Avoid long naps during the day that could interfere with the regular sleep schedule.
- Plan the schedule around a night that will include an age-appropriate amount of sleep.

Adequate amounts of sleep are essential for children. Children age 3 and younger require naps during the day in addition to their nighttime sleep. (See Table 1.)

Age Appropriate Nightly Sleep

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Age	Average Nighttime Sleep Duration (Hours)	Average Daytime Sleep Duration (Hours)
6 mos	11	3 1/2
1 yr	12	2
2 yrs	11 1/2	2
3 yrs	11	2
4 yrs	11	1
6 yrs	11	Ø
8 yrs	10 1/2	Ø
10 yrs	10	Ø
13 yrs	9	Ø
16 yrs and up	8	Ø

(Adapted From Pediatrics. 2003. Vol 111, Page 302-307.)

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Stress

The notion that people get sick more often when they are under increased stress is supported by scientific data. Chronic illness, itself, is known to be a major life stressor. Some studies suggest that stress negatively affects the functioning of the immune system. There are also scientific studies that suggest reducing stress can improve immune function. Many stress reducers are easy to incorporate into one's daily life. These include massage therapy, biofeedback, meditation and hobbies.

The importance of physical activity and adequate sleep in helping to reduce stress has already been discussed.

If you find that you are unable to deal with the stresses in your life, you should absolutely discuss these concerns with your primary care provider. They can assist you or refer you to someone who can help you to minimize and effectively deal with stress. You should never feel that there is nothing that can be done for the stress that you feel is overwhelming and keeps you from living and enjoying your life.

Primary Care

Regularly seeing the primary care provider for health maintenance screening is important for everyone, but even more so for the individual with a primary immunodeficiency. Some types of immunodeficiencies are associated with other illnesses. For example, it is known that some patients with Common Variable Immune Deficiency are at a higher risk for developing autoimmune diseases or leukemias and/or lymphomas than those in the general population. Children especially should have annual physicals. Sometimes failure to grow or develop properly may be the first signal that there is a problem in a child.

As discussed in this Handbook, our immune systems consist of two major categories of defense: the innate immune system and the adaptive immune system. The innate system is the first line of defense with its various components ready-to-go immediately when they encounter a microbe threatening our bodies. This is a critically important defense mechanism, but the innate system is not pre-programmed against every potential threat. It is the adaptive immune system that has the capacity to ramp-up a protective response to new threats. It does this by the generation of specific immune T-cells and B-cells, and the production of antibodies that are specifically designed to combat the new threat. The adaptive immune system takes several days before it reaches full power, but once it is fully activated, it remains on duty for a long time. If the body encounters the same threat again sometime in the future, this system has memory and is therefore able to respond much faster.

Immunization

Perhaps the greatest advances to improve general health over the past two centuries have been the introduction of vaccines that effectively protect individuals from many of the worst microbial threats in our environment. Since primary immunodeficiency diseases interfere with the ability of the body's immune system to respond appropriately, does it make any sense or do any good to give vaccines to patients with primary immunodeficiency diseases? Like so many things in life, it depends.

Vaccines are designed to activate the adaptive immune system to respond to specific microbes that innate immunity alone is not capable of controlling. We are familiar with the usual "childhood vaccines" that have greatly reduced the incidence of serious infectious diseases that in earlier generations sickened or killed

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millions of people. Polio, measles, whooping cough, mumps, Rubella, HPV, chickenpox, HiB, tetanus, meningococcus, diphtheria, rotavirus and influenza are the vaccines most commonly given today, and most communities require that children be up-to-date on their immunizations before being able to attend public school. So, what about people with primary immunodeficiency diseases?

First, it is important to recognize that many types of primary immunodeficiency diseases are fully capable of making a normal response to vaccines. Those with innate system defects like CGD and other phagocytic cell defects, individuals with complement deficiency, and even some with significant adaptive system deficiencies can produce antibodies to many vaccines and thus benefit from immunization. However, there are many others with primary immunodeficiency diseases that will be unable to develop protective immunity following vaccination, and in some cases the vaccine itself may represent a threat to the recipient.

Since vaccines for chicken pox, measles, mumps, smallpox, rubella, rotavirus, BCG, yellow fever, oral polio and the influenza nasal spray are live attenuated vaccines, individuals with primary immunodeficiency diseases could potentially contract infections if they receive these immunizations. In practice, infants with Severe Combined Immune Deficiency (SCID) are at greatest risk and it is the general recommendation that others with defects in adaptive immunity also avoid receiving any live agent vaccines. (*See chapter titled "Severe Combined Immune Deficiency and Combined Immune Deficiency."*) Since some of these live vaccine viruses (oral polio, rotavirus) can be found in the some body fluids and stools for up to two weeks following vaccination, it may be necessary to limit contact between any recently immunized individual and infants with SCID until the period of viral shedding has passed. For children and adults with primary immunodeficiency diseases who are receiving immunoglobulin (Ig) replacement treatment, the infused antibodies should give them adequate protection from any secondary spread of vaccine virus.

The usefulness of vaccination during treatment with intravenous or subcutaneous Ig therapy is not fully understood, in part due to complexity of the range of underlying immune defects treated with Ig therapy. Assessing patients' antibody responses to vaccine is confounded by antibody in infused Ig. Vaccines can also stimulate T-cell responses, which may have antibody-independent (such as cellular immune) protective effects, but those are harder to measure and even less well understood with respect to their role in protection independent of immune globulins.

Some patients with milder forms of immunodeficiency (such as selective IgA deficiency, mild hypogammaglobulinemia, partial DiGeorge syndrome) can receive live virus vaccines at the discretion of their doctor.

Purified protein, polysaccharide or non-viable whole-agent vaccines pose no infectious risk to patients. However, for most vaccines, the patient's antibody response is likely to be inferior to what is provided by Ig therapy. New vaccine agents may be exceptions to this general rule. For example, antibody to new strains of influenza may not be found yet in therapeutic Ig, and consideration should be given to administering such a vaccine to patients receiving Ig treatment. Although there may be theoretical benefit to inducing T-cell responses in patients on Ig, clinical benefit is unproven, and this practice may not be cost-effective, particularly for expensive vaccines such as HPV.

For families with a member who has a primary immunodeficiency disease, we recommend that all members of the family group, including the patient, should keep their immunizations up-to-date. This is particularly important for readily communicable diseases like influenza with many different strains circulating that change from year to year. Why do we recommend that everyone be immunized to influenza? First, some patients with a primary immunodeficiency may respond and benefit directly from the influenza vaccine. Even if they do not, there is little down side to receiving the killed vaccine. Family members who are able to respond to a vaccine will be protected. Even if the patient with a

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primary immunodeficiency disease does not respond to the immunization, they will benefit from having everyone else in the home protected from infection and thus not susceptible to bringing the virus home with them. This is particularly important if there are other school-aged

children in the home. We want to create a “protective cocoon” of immunized persons surrounding patients with primary immunodeficiency diseases so that they have less chance of being exposed to a potentially serious infection like influenza.

General Care during Times of Acute Illness

Even after a diagnosis of primary immunodeficiency disease has been made and appropriate treatment initiated, people with primary immunodeficiency diseases are still going to get sick. Hopefully these illnesses will be reduced in number and intensity, but it is unrealistic to think that because one is being treated for one’s primary immunodeficiency disease that they will not be sick any more. When acutely ill, people with primary immunodeficiency diseases should:

- Seek medical advice. Do not ignore symptoms such as fever or a productive cough and think they will “just go away.”

- Never self treat. Taking leftover antibiotics or those prescribed for another family member is not a good idea.
- Follow the healthcare provider’s advice. If 14 days of antibiotics are prescribed, take 14 days. Do not stop after a week because you feel better. If there is a recommendation to stay home from work or school for several days, then stay home.

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

Summary of General Care

The diagnosis of a primary immunodeficiency disease is a life-changing event, but it can be viewed in a positive rather than a negative way. The diagnosis and initiation of 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 recommendations and advice of the healthcare team for treatment of the primary immunodeficiency diseases can maximize the potential for a full and normal life.