

Immune Deficiency Foundation

Patient & Family Handbook

For Primary Immunodeficiency Diseases

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Chapter 45

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6th Edition

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110 West Road, Suite 300
Towson, MD 21204
800.296.4433

www.primaryimmune.org
idf@primaryimmune.org

Chapter 45

Adults Living with Primary Immunodeficiency Diseases

Carla Duff, CPNP, MSN, CCRP, IgCN, University of South Florida, Tampa, Florida, USA

Many adults with primary immunodeficiency diseases (PI) live full lives. Those who are well-informed and working with an attentive healthcare team can often pursue a career and live an active, productive life.

Introduction

Although the first PI were identified in children, the medical community soon realized that adults can have a PI as well. Advances in medicine, as well as earlier diagnosis and treatment of the childhood immunodeficiency diseases, have allowed many born with a PI to grow into adulthood. In other cases, many children born with apparently normal immune systems go on to develop a PI later in adolescence or adulthood. Unfortunately, the Immune Deficiency Foundation (IDF) survey research has shown that adults with an undiagnosed PI will, on average, experience symptoms of their immunodeficiency for more than a decade before a diagnosis is made.

No matter how old you were when you were diagnosed, it is important to learn about your condition and to choose healthcare providers with whom you can work comfortably. In addition, you should consider the psychosocial aspects of living as an adult with PI.

Normalizing Your Life

PI affects people in different ways, but, like everyone else, individuals with a PI need to feel a sense of accomplishment and purpose and contribute to the world around them. To best manage your life and your health, you need to educate yourself about your disease, build a collaborative relationship with your healthcare providers, and take care of yourself physically and emotionally.

Accept your new diagnosis. Some recently diagnosed individuals may experience a combination of relief, fear, and denial upon diagnosis. In such cases, it can be a relief for you to finally have a firm diagnosis and an identified

treatment plan. At the same time, it can be frightening to have confirmation of a documented illness that is of a chronic nature. This is especially the case for individuals who already may be struggling with one or more conditions which may interfere with their level of functioning and quality of life. You can work towards accepting your diagnosis by creating a support system with family, friends, and healthcare providers to help you effectively manage the impact of a primary immunodeficiency on your life.

Educate yourself about your health issues.

You will be better equipped to manage health issues successfully if you understand them and the potential impact they can have on your life. This is true of your PI as well as any other health issues you may have. Almost no one is likely to care about your health and well-being as much as you do. The diagnosis of any illness, particularly a chronic illness, can challenge your sense of independence and control over life. Educating yourself not only provides you with information about how to care for yourself and gives you the confidence to make decisions about your treatment but it can also help to restore and reaffirm a sense of independence and control. Self-education is a continual process and IDF provides a wealth of information for you. Ongoing research frequently provides new information about these diseases and their treatment, so it is important to review existing information, to register for IDF communications and mailings at www.primaryimmune.org, and to continue asking questions of your healthcare providers. Ensure that while educating yourself about your disease that you use reputable sources like www.primaryimmune.org or www.nlm.nih.gov. Always consider the source of the information that you find; if it sounds too good to be true, it probably is.

Choose a quality healthcare team. It is essential to have a healthcare provider who understands your health problems. Seek out an immunologist who specializes in PI and make sure that you feel comfortable with that person. They should welcome and encourage your questions and input. Individuals who are involved in their own healthcare decisions tend to do better than those who are not as involved, so it is in your best interest to find healthcare providers who consider you a partner in the treatment process. Although many healthcare providers are pressed for time, most of them appreciate if you are curious, willing to learn about your health issues and treatments, and want to collaborate in your care. IDF can help you locate a specialist in your area, go to www.primaryimmune.org.

Build strong social relationships. It is particularly important to build and maintain strong relationships, both inside and outside the family, and to remain connected socially. Schedule quality family time. Meet friends for lunch or coffee. Volunteer your time for a worthy cause. Engaging in activities outside of managing your health will ultimately benefit your health. It is also important to learn to ask for and accept help from the people in your life. Family members and friends often want to help and contribute to your sense of well-being. They can be a valuable resource for you.

Connect with others like you. Individuals living with chronic illnesses, especially unusual or rare disorders such as PI, often feel isolated and alone. Contact with other individuals who live with these diseases is a way to both gather knowledge and acquire an important sense of connection with others who share your experience. IDF can put you in touch with others living with PI through its peer support program and can provide information about regular educational meeting opportunities that occur at various regional and national locations.

Maintain a positive attitude. If there are activities in which you can no longer engage, focus instead on what you can do. Consider the gifts and abilities you have and use them to contribute to the world and people around you.

Family Life for Adults Living with PI

Maintaining strong and healthy family relationships can be a special challenge when you have a chronic illness, but these relationships are vital to your

health. Consider your family as your team. You and your family must work together to remain strong and caring and communication is the key. Your family members should share their thoughts and feelings with each other on a regular basis. One of the most effective ways to do this is to share a daily meal together. This is a great opportunity to share your experiences, plan family activities and outings, and reminisce about good times spent together. In addition, everyone must make a contribution to the team. Everyone needs to feel a sense of accomplishment and to feel good about themselves and their contributions. In some cases, because of your health, you may no longer be able to work or complete other tasks for which you have been previously responsible within the family. You need to discuss the changes in your roles or responsibilities and work with your family members.

It is very common for those with PI to take out their frustrations on family members when they are feeling overwhelmed, angry, or stressed. Remind yourself that you may be feeling upset about your situation and are not necessarily angry with other family members. At times, it is important to share that thought with your family. Consider what other family members need or want as well. Usually you will find that it is exactly what we all want: love, understanding, and appreciation.

Having Children

If you and your partner consider growing your family, it is important to understand the genetic implications of PI. Some PI are genetic, meaning they are passed down from parent to child. Your health care provider or a genetic counselor can address these questions and concerns. (See Inheritance Chapter.)

Managing Stress

Not everyone with the same disease is affected in the same way. It is typical to experience increased stress as they face unexpected illness, hospitalizations, and missed work. They may simply be unable to manage their usual responsibilities and may require the help of others while they recuperate. Some can become absorbed in their own problems and feel angry, hopeless, or depressed. The amount of stress some feel and the way they cope vary greatly in each individual. Recognizing and managing this psychological stress can be challenging, but it is important to identify stress and how it affects your physical and emotional health, as well as to develop effective ways of coping.

The best ways to address and manage stress differ from person to person and sometimes it takes time to understand your limits. Keep in mind a variety of activities that help you manage stress. Remember that you may not be as efficient when you are stressed or overwhelmed with fatigue, so it is of no benefit to push yourself at those times. Make time for rest and relaxation. Take a nap, learn how to meditate or use deep breathing or other relaxation exercises. Make time to read for pleasure or enjoy music. Exercise is also an excellent way to relieve stress, whether you walk, ride a bike or engage in a more strenuous workout. Know the kinds of stress-reducing activities that are helpful to you and best suited to your lifestyle and physical abilities.

Many individuals benefit from speaking with a mental health professional, such as a psychiatrist, a clinical psychologist, a social worker, or a pastoral counselor. If you are wondering how you will know when it is time to seek help, consider the following suggestions:

- When your feelings and/or your behavior regularly interfere with your ability to function on the job, at home or as a member of your family.
- If you are trying to move forward but feel stuck or if you feel uncomfortable to the point that you feel a need to do something as soon as possible.
- When your family members become overwhelmed, unable to manage or struggle to manage everyday stress or when relationships seem to be falling apart.

The first step in seeking help is to contact your insurance company to review your mental health coverage and benefits. You will want to know any in- and out-of-network deductibles and co-pays and if there are any restrictions on the type of professionals you can see. Your insurance company can usually provide you with a list of mental health professionals in your area who are participating providers with your plan. Another way to identify a potential therapist is to get a recommendation from someone you trust, like a family member, friend, your healthcare provider, or clergy. In addition, most state psychological associations or state social work associations have referral services to help you identify a suitable professional.

Employment with PI

When choosing a job or career, adults with PI must think in terms of ones that are suitable for

their condition. Depending on the nature of your condition, you may or may not be limited physically. However, there may be complications that need to be considered. Factors like time and stress and how they affect your condition and treatment cannot be ignored. You may need to limit your exposure to large numbers of other individuals who may transmit infections.

In seeking employment, be aware that there are laws against discriminating against an applicant based on a chronic health condition. However, that does not mean that the laws are easy to enforce. You may want to familiarize yourself with these laws.

Those living with PI work in all kinds of jobs. For many, the health insurance coverage associated with employment is the most problematic. Small employers, for instance, may not be able to cover you, so choosing an employer who can provide adequate health insurance may be important while considering careers. New Health Insurance Portability and Accountability Act of 1996 (HIPAA) legislation has improved the ability to transfer insurance coverage from job to job once you are insured. The Family Medical Leave Act (FMLA) also ensures continued employment in the face of prolonged work absences due to illness. Disability in this population is not common but can happen. You need to be prepared should this occur. (See Employment Chapter.)

Relocation

Adults with a PI may find it necessary to relocate to another city or state for education or employment. It is vital that you notify your healthcare provider of any plans to relocate as soon as possible to ensure that there are no disruptions in your medical care or treatment. Notifying your healthcare provider as soon as possible ensures that there will be time to address the following:

- Establishing care with new healthcare providers. Your current provider may be able to recommend a new provider or you can contact the IDF for a provider in your area. Often providers have long wait list for new patients, so it is important to make an appointment for as soon as possible after you relocate.
- Signing a release of medical records. In order for your current provider to send your medical records to your new provider you must sign a release of medical information form.

- Needing to continue therapy. Once you relocate, your current provider may no longer be able to order and/or refill your medications. If you are receiving Ig replacement therapy, new authorizations may need to be obtained and care transferred to a new center. In order to prevent a lapse in your treatment, this authorization will need to be obtained as soon as possible after you relocate. Discuss with your healthcare provider how you will continue to receive treatment prior to your relocation.

Most healthcare providers are familiar with patients relocating and will be able to work with you to ensure your transition is as smooth as possible.

Health Insurance for Adults Living with PI

Health insurance is a concern that all people with a PI must face. Decisions regarding school or employment may be affected by insurance coverage. This issue cannot be taken lightly by anyone with a pre-existing condition. If you allow your insurance to lapse or do not look into the options that exist before coverage terminates, your ability to qualify for insurance may be seriously jeopardized. It is important for an engaged or married couple to face the issue of health insurance realistically and understand its importance in career decisions.

It is also essential that you understand how the Affordable Care Act (ACA) of 2010, also known as healthcare reform, affects you. The law puts in place strong consumer protections and provides new coverage options. (See Healthcare Chapter.)

Coordinating Your Healthcare and Being Your Own Healthcare Advocate

It is essential for you learn how to coordinate your healthcare, become your own healthcare advocate, and establish a relationship with your healthcare providers. Communication is key.

Effective communication is essential in all relationships. It needs a sender, a message, and a receiver. It is a two-way process and is not complete until the receiver understands the message.

To improve your care, it is important to pay careful attention to the communication with your healthcare team. Your healthcare team includes anyone who

helps you get the care that you need and can include doctors, nurses, ancillary therapists, case managers, and social workers. Support personnel and insurance providers may also be key people.

Ways to help you communicate with your healthcare providers so you can be heard and understood:

- Treat each healthcare appointment as if it is an important meeting. Remember, communication is more than just words. You have probably waited a while for the appointment, and it will not last as long as you might like, so make the most of your face-to-face time. Silence your cell phone. Make your visit personal, minimize distractions; do not bring the whole family or kids into the exam room. Remember, it is an important meeting, and you do not need any interruptions.
- Be prepared. Plan ahead and do your homework. Get any necessary insurance authorizations ahead of time. This will help you to keep organizational noise down.
- Bring your medical information to your visit. You can keep a journal, create a folder, make computer documents, or use an online personal health record. **However, you choose to document your healthcare, make sure to include:**
 - » A brief history leading to the diagnosis, written by you or your healthcare provider
 - » Copies of laboratory evaluations confirming the diagnosis
 - » A current list of providers caring for you along with their accurate addresses and phone numbers
 - » The chronology of important events, specifically noting types of treatment and therapy, changes in therapy and subsequent responses to that therapy, surgeries, and/or hospitalizations
 - » A list of your current medications
 - » Allergies to medications
 - » Infusion log if you are currently receiving immunoglobulin (Ig) replacement therapy
 - » An immunization record or lack of immunizations
 - » Current insurance information

- » Prepare and feel comfortable asking your questions. Ask questions, but be concise. Ask your questions early, not at the end of your visit so that the provider has a chance to carefully answer your questions.
- » Bring a list of the most important questions that you have.
- » Never be afraid to ask a question because you think it might be seen as dumb or because you feel that the provider knows best.
- » If you do not understand the meaning of the words that your provider is using, do not be afraid to say so.


- **At your initial visit, ask questions like:**

- » Whom should I talk to in the office when I need to get a message to you?
- » What should I do when I get sick after hours or on the weekend?
- » Which hospital do you admit your patients to?
- » May I contact you by e-mail if I have a question?
- Be sure to take notes electronically or bring a notebook and pen along.
- Express your concerns in your own words. Use words with which you are comfortable. Tell the provider what the reason for your visit is, it will help them focus on what you need.
- Forget your stereotypes about your providers. Remember, healthcare providers are people, just like you, with a job to do. Their job is to help you find a way to stay as healthy as possible. It is important to find providers with whom you can be yourself around. They are your partners in your healthcare.
- Be honest. Do not be afraid to talk about what goes on in your bathroom or your bedroom. If you smoke, drink, take illicit drugs, use herbs or see alternative care providers, say so. Remember, your healthcare information is private and law protects that privacy. Whatever you do, do not be afraid to tell the truth. Your relationships with your healthcare providers are intimate ones. It is their job to help you, not to judge you. If you did not follow their advice, did not adhere to

the treatment plan, or did not buy the medicine because your insurance did not cover it and you could not afford it, tell them. How else will they know if what they thought should work was effective?

- Advocate for yourself. No one knows how much your disease affects your life better than you do. No one understands the changes you have to make every day to deal with your treatment as you do. To live your life to the fullest, you need to be your own healthcare advocate.
- Ask questions of your providers about your diagnosis, treatment, and plan. If you do not understand, ask again.
- Inquire about what can be done to improve your health. Consider such things as diet, physical activity, sleep, and social activities.
- In terms of school or work, maintain consistent communication with your school and/or your employer in the event that you miss days and understand their policies and procedures.
- Know your insurance policy and let your provider know if there are any changes—especially if those changes mean you have to change providers or your therapy and medications will no longer be covered.
- If you receive Ig therapy, make note of how it is going and/or any side effects. Keep an infusion log, including date, time, product name, and product lot number.
- Build positive relationships with your providers. Know whom to call and when.
- Ask about resources for further information at the local, state and national level.
- Connect with IDF for additional resources: www.primaryimmune.org or 800-296-4433.

Remember, communication is how we all relate to each other. Think about the things that you need to stay healthy. Think about how you can best communicate those needs. By doing this you will have some of the tools you need to successfully coordinate your healthcare and be your own advocate. No matter what your diagnosis is, this is your life. Make the most of it.



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