

Adults Living with Primary Immunodeficiency Diseases



Chapter 32

Many adults with primary immunodeficiency diseases live full lives. The well-informed patient working with an attentive healthcare team can often pursue a career and live an active, productive life.

Introduction to Adults Living with Primary Immunodeficiency Diseases

Although the first primary immunodeficiency diseases were identified in children, there has been a growing awareness that adults also can have a primary immunodeficiency disease. Advances in medicine as well as earlier diagnosis and treatment of the childhood immunodeficiency diseases have allowed many patients born with a primary immunodeficiency disease to grow into adulthood. In other cases, many children born with apparently normal immune systems go on to develop a primary immunodeficiency disease later in adolescence or adulthood. Unfortunately, the Immune Deficiency Foundation (IDF) survey research has shown that adults with an undiagnosed primary immunodeficiency disease

will, on average, experience symptoms of their immunodeficiency for more than nine years before a diagnosis is made.

No matter how old you were when you were diagnosed, it is important for you 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 primary immunodeficiency diseases.

Normalizing Your Life

Primary immunodeficiency diseases affect people in different ways, but, like everyone else, individuals with primary immunodeficiency diseases 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 that interfere with their level of

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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 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 primary immunodeficiency diseases 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.

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 primary immunodeficiency diseases and make sure that you feel comfortable with that person. They should welcome and encourage your questions and input. Patients 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 patients who are curious, willing to learn about their health issues and treatments, and want to collaborate in their care. IDF can help you locate a specialist in your area, go to www.primaryimmune.org or call 800-296-4433.

Take advantage of resources. 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.

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 primary immunodeficiency diseases, often feel isolated and that they are struggling 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 another patient 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 Primary Immunodeficiency Diseases

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 be no longer 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 patients 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.

If you and your partner consider growing your family, it is important to understand the genetic implications of primary immunodeficiency diseases. Your immunologist or a genetic counselor can address these questions and concerns. (*See chapter titled “Inheritance.”*)

Managing Stress

Not everyone with the same disease is affected in the same way. It is typical for patients 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 patients feel and the ways they cope vary greatly from patient to patient. 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.

(Managing Stress continued)

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 and Health Insurance for Adults Living with Primary Immunodeficiency Diseases

Adult patients, in choosing a job or career, 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.

Patients with primary immunodeficiency diseases work in all kinds of jobs. For many patients, 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.

(Employment and Health Insurance for Adults Living with Primary Immunodeficiency Diseases continued)

Health insurance is a concern that all people with a primary immunodeficiency diseases 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 chapter titled “Health Insurance.”*)

Coordinating Your Healthcare and Being Your Own Healthcare Advocate

It is essential for you learn how to coordinate your healthcare and become your own healthcare advocate by establishing a relationship with your healthcare providers. You have to learn to understand each other. Communication is the key, and the secret to effective communication is in your grasp.

Communication is how we share information. 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. Communication does not just use words; it also uses tone of voice, body language, emotion and touch. Noise interferes with communication. We live in a noisy world. “Noise” can come from our environment, our culture, our psyche, and our choice of words and how any of those affect how our words are understood.

To improve your care, it is important to pay careful attention to the communication with your healthcare team and the noise affecting it. Your healthcare team includes anyone who helps you get the care that you need. The members of your healthcare team 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. When you first meet your provider, remember it may be the start of a long-term relationship, so smile, introduce yourself, shake hands, make eye contact and pay attention. 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. Do your best to keep the environmental noise down, such as silencing 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.

(Coordinating Your Healthcare and Being Your Own Healthcare Advocate continued)

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 the IDF eHealthRecord, an online personal health record (www.iedehelathrecord.org). 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 receive immunoglobulin (Ig) replacement therapy
- An immunization record or lack of immunization
- Current insurance information
- Explanation of benefits records can be kept in the journal or separately but should be periodically reviewed for accuracy

Prepare your questions. If you have been searching the Internet for information about your condition or treatment, do not bring in a stack of printouts. Instead, bring a list of the most important questions that you have. Do not expect that you are going to find a miracle cure for a chronic disease. Search 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.

Feel comfortable asking questions. 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. Remember, communication is a two-way process, and it is neither effective nor complete until the message is understood.

At your initial visit, ask questions like:

- What is the best way to get a message to you?
- 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, right after you say your hello; it will help them focus on what you need. Ask questions, but be concise. Ask your questions early, not at the end of your visit. Give the provider a chance to think and carefully consider your questions.

Forget your stereotypes about your providers. With their white coats and degrees on the wall, they seem like authority figures. 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. They are your partners in your healthcare.

(Coordinating Your Healthcare and Being Your Own Healthcare Advocate continued)

Our attitudes or inhibitions can hamper communication by creating psychological noise. 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?

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.

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. How you can advocate for yourself:

- 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 your 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 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. Identify and silence as much of the noise around your communication as you can. Apply the aforementioned principles. 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.