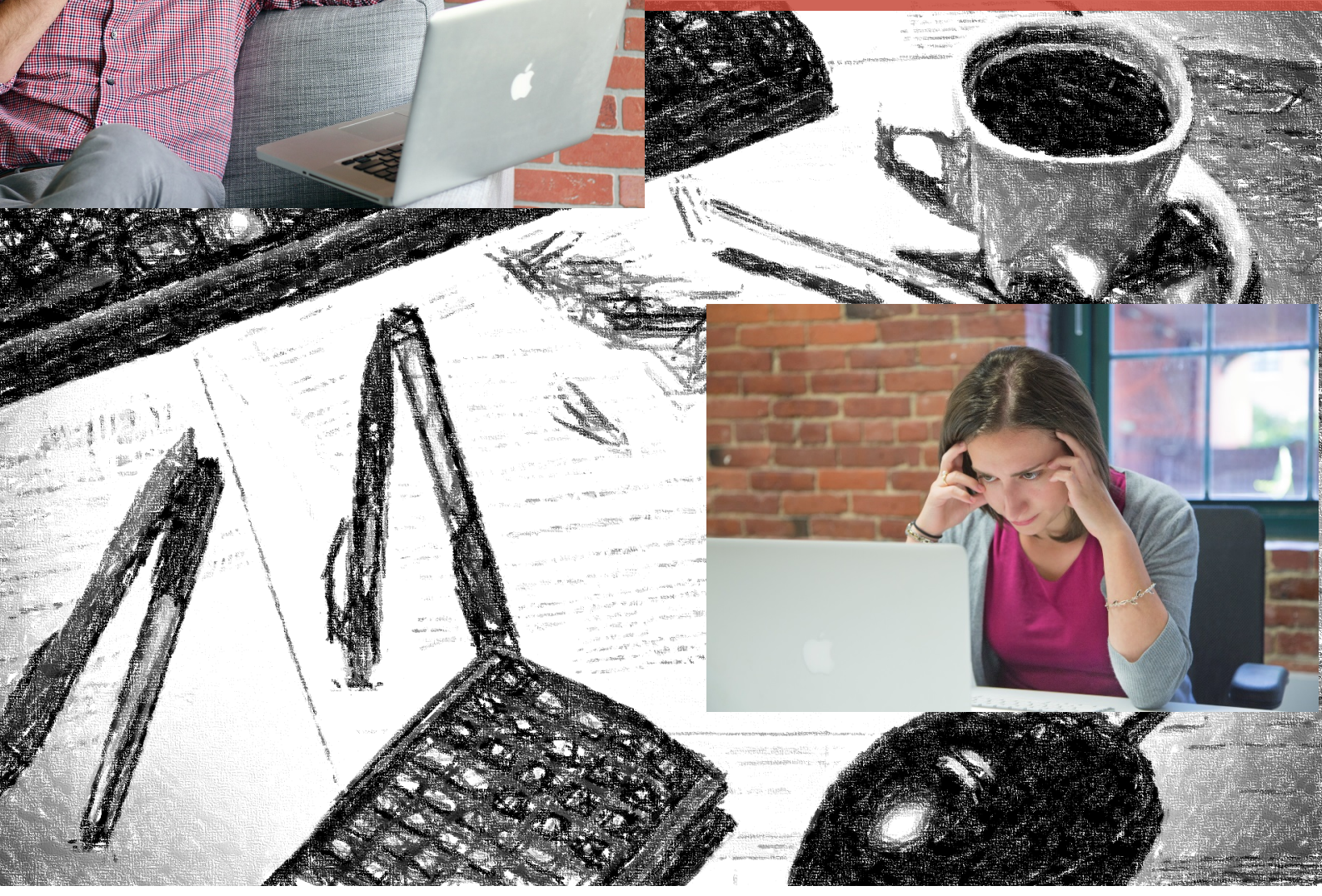
Navigating Insurance

For people with primary immune deficiency, managing insurance may feel like a second full-time job. The guide is designed to provide information about Connecticut insurers and tips to make your journey's navigation easier. .



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Chapter 1

Introduction

Learning that you or a loved one has a diagnosis of a primary immune deficiency is life-changing. We Connecticut Zebras have been there and are proof that you can incorporate your treatment into your everyday routine and live a relatively normal life. In fact, once you've been on your treatment and the infections go away, you may be amazed at how much better you feel!

The Immune Deficiency Foundation has a wealth of information about various immune-deficiency conditions, treatments, insurance, managing school, and employment. The IDF Patient and Family Handbook for Primary Immunodeficiency Diseases, 6th edition, is available for download at no charge. Every family should have a copy. Our goal is not to duplicate what the IDF does, but rather, to provide information specifically for Connecticut.

Courtesy Eric Ward / CC-BY-SA

When I was diagnosed, I had heard of adults with immune deficiency disease, but that was it. I've been a nurse and taught for more years than I am willing to admit. So, don't feel bad that this is all a new world for you. One of the biggest challenges is managing insurance coverage. Treatment is expensive and requires special approval over and above typical prescriptions. Different companies require different information. In this guide, I have compiled the requirements for the insurance companies in Connecticut. Remember that even if you live the state, you may have out-of-state insurance that will not be listed here. However, the general information is similar among all the companies.

I'm Pat Carroll, a former trauma and ER nurse. I was diagnosed in September 2011 with specific antibody deficiency. I did not have the familiar story of having infections throughout my life. I had a very bad bout with Lyme disease in 1991 and developed symptoms of an autoimmune disease in 2005 (that we figured out after the fact.) Then, in 2010, I had H1N1 influenza, followed by a staph pneumonia. I took antibiotics, felt better, and a few weeks later, I was sick again. This cycle happened over and over, and my lung function slowly deteriorated. I had a CT scan in June 2011, which showed permanent damage in my right lung. That's when my lung specialist thought I might have an immune deficiency. He ordered a blood test to measure antibodies in my blood, then gave me a pneumonia vaccine. Eight weeks later, he re-tested my blood, and I only made 3 of the 13 antibodies I should have — the definition of specific antibody deficiency. I started treatment with IVIG the following month.

You can (and should) be an expert on your health conditions.

Insurance companies require lab results and clinical information so that they can confirm the diagnosis. Once you begin treatment, you can never repeat that original lab work. That is why it is so important for you to have your own copy of every test result. I changed insurance companies seven years after my original diagnosis and had a new immunologist. If I did not have my own copies of the lab work, I probably would have missed treatment while I waited for the paperwork required for the new company to approve my therapy**. I cannot emphasize this point too much: you must have documentation of your *original* workup**.

A quick note: As you read, I use the word "you," assuming you are the one with the immune deficiency. But you can substitute "your family member" if that is more appropriate for your situation.

Chapter 2

Your Immune System

The immune system is one of the most complex systems in the body. Researchers are learning more about how it works and immune deficiencies every day. The immune system's job is to protect from infection. Immune deficiencies affect the *humoral immune response*, which refers to proteins in the body fluid that have a special role in this protection system.

Lymphocytes, a type of white blood cell, can be B cells (made in the bone marrow) or T cells. B cells make antibodies, also called immunoglobulins. There are five classes of immunoglobulins, IgG (75-80%), IgM (5-10%), IgA (10-15%), IgE (0.002%) and IgD (0.2%)[1] .

IgM is found in blood and lymph fluid and is the first antibody made in response to an infection.

IgA antibodies are in mucous membranes that line the nose, breathing passages, eyes, ears, digestive tract, and vagina. They protect body surfaces that are exposed to outside foreign substances. IgA is also in saliva, tears, and blood.

IgG is in all body fluids. It is the smallest but most common antibody and is essential to fight bacterial and viral infections. IgG can further be measured in subclasses: IgG1 (60-65%), IgG2 (20-25%), IgG3 (5-10%) and IgG4 (3%)[2] . The normal immune response includes a mix of all four subclasses along with the other immunoglobulins. People with subclass deficiencies may be completely healthy (in which case they have probably not had the test), or they may have infections, particularly when an IgG subclass deficiency exists with other antibody deficiencies.

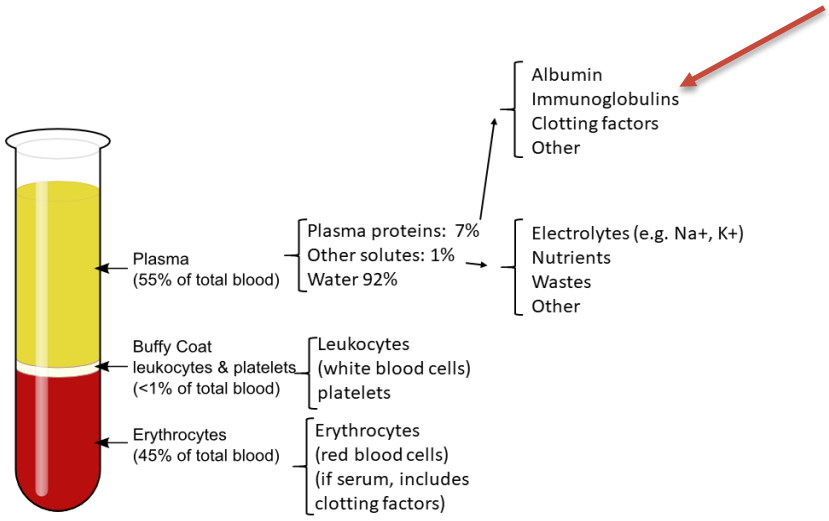


Figure 1 Courtesy Alan Sved

Antigens trigger the immune response -- fighting infection or causing allergies

CD molecules on the cell surface further classify T cells; they describe the cell's actions. Those most associated with immune deficiencies are CD3 & CD19. Some T cells can help B cells develop antibodies to bacteria, and other T cells kill viruses.

Antibodies, from IgG immunoglobulin, are Y-shaped proteins that recognize antigens as foreign invaders and bind to them. Antibodies travel throughout the body in blood and lymph. Each antibody binds to just one antigen type, forming an antigen-antibody complex, or an immune complex (see next page). It's like a lock and a key — there is only one match. When this system is working well, the antibody stops the antigen from doing more damage, such as causing infection.

However, the system can get out of control when antibodies mistake normal substances for antigens and bind with them. This concept of attacking normal tissue is called autoimmunity (auto meaning self). In an autoimmune response, the body's antibodies and other elements of the immune system, such as T-cells and B-cells, attack normal tissues. There are more than 100 autoimmune conditions.

Monoclonal antibodies, such as the drug rituximab, act against certain B cells in the immune system. Eliminating these cells can reduce symptoms of diseases such as rheumatoid arthritis.

Figure 3 Courtesy EpicTop10.com on Flickr

An **antigen** is a foreign substance that triggers an immune response in the body, particularly the production of **antibodies.**

In certain people, the body recognizes allergens, such as pollen as antigens, producing an outpouring of IgE. Those antibodies travel to tissues that produce histamine and other substances that cause common allergy symptoms.

Figure 2 Courtesy Biology for AP Courses Open Stax Apr 10, 2020

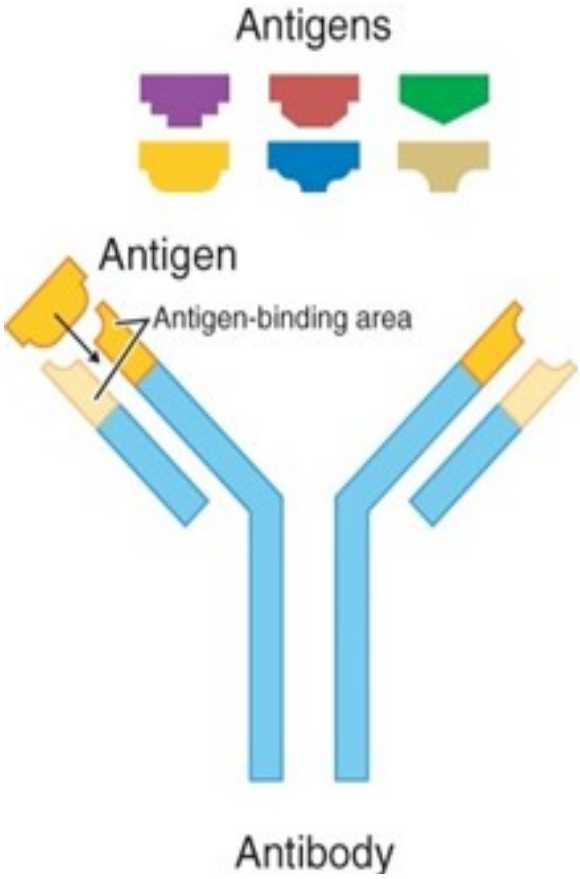


Figure 4 Courtesy Libre Texts Introductory Biology (CK-12)

About 20-25% of people with CVID also have an autoimmune disease[3], but these diseases are not just limited to CVID. Experts now recognize a range of immune dysregulation that can include primary immune deficiency, autoimmune disease, or a combination of these two types of immune system abnormalities.

Do you have a primary immune deficiency AND an autoimmune disease? You're not alone

Chapter 3

Types of Primary Immune Deficiency Diseases[4]

According to the Immune Deficiency Foundation, more than 400 different primary immune (PI) deficiencies have been identified.

Severe Combined Immune Deficiency & X-lined Agammaglobulinemia

These are considered among the most severe conditions because babies are born without T-cell or B-cell function. There are at least 13 different genetic defects that can cause SCID. As of December 2018, all 50 states screen newborns for SCID and other immune deficiencies. Genetic or molecular testing confirms the diagnosis.

DiGeorge Syndrome

This condition is a primary immune deficiency caused by the abnormal formation of certain tissues during fetal development. 90% of patients have the same genetic defect. T-cell abnormalities are the most common. Many patients have other disorders, such as heart defects and feeding difficulties. Some children may be diagnosed at birth, while health providers may not diagnose others until they have frequent infections when they are a little older.

Wiskott-Aldridge Syndrome

Unlike other PI, boys with WAS also have problems with bleeding due to abnormal platelets. Abnormalities in T-cells and B-cells cause the immune deficiency. Infants can have eczema. Immune problems typically become evident in toddlers and older children. Genetic testing makes a definitive diagnosis.

Ataxia-Telangiectasia

A-T is a syndrome that affects how toddlers walk, the shape of some blood vessels, and causes progressive neurological symptoms, particularly with coordination. About 65% of people with A-T have very low or absent IgA, as well as other immunoglobulin deficiencies and low levels of T-cells and B-cells. Genetic or molecular testing can confirm the diagnosis.

Combined Variable Immune Deficiency

CVID is one of the most commonly diagnosed PI conditions. The name “variable” comes from the differences in this condition from person to person. Some people have low IgG and IgA, others also have low IgM, and some only have T-cell defects. Often, the diagnosis is made in adults in their 30s and 40s, and the frequent infections are recognized in retrospect. The diagnosis is confirmed by testing the response to vaccination, typically the pneumococcal polysaccharide vaccine.

IgG Subclass Deficiency

An IgG subclass deficiency is diagnosed when total IgG levels are normal, but one of the types of IgG is low. According to the Immune Deficiency Foundation, this is a controversial diagnosis, and experts disagree about the significance of a low subclass as a cause for frequent infections. Antibody response to the pneumococcal polysaccharide vaccine is critical for a diagnosis that requires treatment.

Selected Specific Primary Immunodeficiency

Selective IgA deficiency is diagnosed when IgA is undetectable, but there are no other deficiencies in the immune system. Many people have no illness as a result; others may develop a variety of clinical problems.

Selective IgM deficiency is defined as low IgM levels in patients with recurrent infections that can be severe. However, some patients may have no symptoms. Without symptoms, there is no reason to measure immunoglobin levels, so the actual number of people with this condition may be undercounted.

Hypogammaglobulinemia is the name for low IgG levels; agammaglobulinemia is the absence of IgG.

Specific Antibody Deficiency

Specific antibody deficiency can be tricky to diagnose because the levels of immunoglobulins are all normal. However, people have frequent, recurrent infections requiring antibiotics. The problem here is that IgG cannot make specific antibodies to fight off infection. The levels are normal, but the immunoglobulins are not functional. Again, the key to diagnosis is measuring the response to the pneumococcal polysaccharide vaccine.

Chapter 4

Information Required by Insurance Companies

Insurance companies are quite prescriptive in their Clinical Policy Bulletins . They describe the disease or condition, how it is diagnosed (to the company's satisfaction), and the resulting treatments they can provide. In general, the requirements for diagnosis and which treatments are available for which diagnosis are now quite standardized. However, you may run into a situation in which the choice of Ig replacement drugs is limited. This limitation comes from the employer offering the plan (fewer choices saves money) or from the choices the policyholder made when it came time to select the plan each fall. Most people without health challenges simply choose the least expensive plan and move on without another thought. However, those selections can have significant ramifications if you are surprised with a serious illness.

Tests Requested by Insurance Companies

Not all tests are required for each diagnosis – see the reference table in the Appendix

- Genetic or molecular testing to confirm certain diagnoses
- Measurement of IgA, IgM, IgG (and IgG subclasses) levels
- Measurement of T and B cells in total and types of T cells and B cells by a test called flow cytometry (e.g., CD3, CD19)
- Measurement of antibodies before and after vaccination with pneumococcal polysaccharide vaccine to demonstrate impaired antibody response
- Measurement of protective antibody titers after tetanus and diphtheria or *Haemophilus influenzae* type B (HiB) vaccination

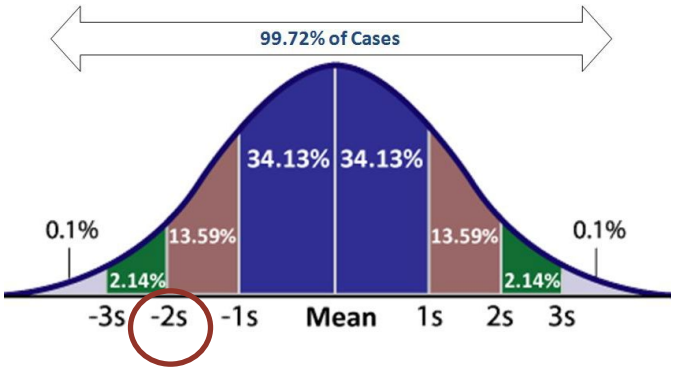


Figure 5. Courtesy MADe/Wikimedia Commons

Some insurance companies specify the results they are looking for; others rely on the laboratory range for normal values. Some state that the result must be "lower than 2 SD below the laboratory mean value for age." (See Figure 5 above. The red circle indicates 2 standard deviations below the mean.)

The mean is the average. A standard deviation is a statistical measurement of how spread out values are from the mean. (See Figure 5.) A group of researchers[5] examined results from 330 healthy children age 0-18. The mean IgG was 853 mg/dL, with a standard deviation of 362 mg/dL. So, 2 SD below the laboratory mean would be 853-362-362 = 129 mg/dL. This calculation is an example of the concept; the actual values will depend on the laboratory doing the testing.

If the values are spread out (distributed) equally, only about 2% of values will be 2 SD below the mean.

Other Reports Requested

- Workup for kidney or gastrointestinal disease that could cause low IgG
- Workup that excludes other causes of immune deficiency such as drug-induced, other genetic abnormalities, cancer, or infections such as HIV
- Documentation that the prescription is from or in consultation with an allergist, immunologist, ENT, pulmonary specialist, or infectious disease practitioner who treats people with immune deficiency
- Documentation of frequent and severe bacterial infections requiring antibiotics, particularly ear, sinus, lung, gastrointestinal, or sepsis (a body-wide infection of the blood)
- Documentation of the number of episodes of antibiotics and the total number of days treated
- Documentation that the health provider has maximally treated any underlying conditions such as asthma or allergies to minimize risk for infection, with supporting lab results or imaging

Besides laboratory reports, insurance companies also require a detailed medical history, particularly of infections and antibiotic use. You may need to go back through insurance claims to build a complete history, particularly if more than one health provider has treated you. See the form in the Appendix to help you keep track. Different companies require slightly different information.

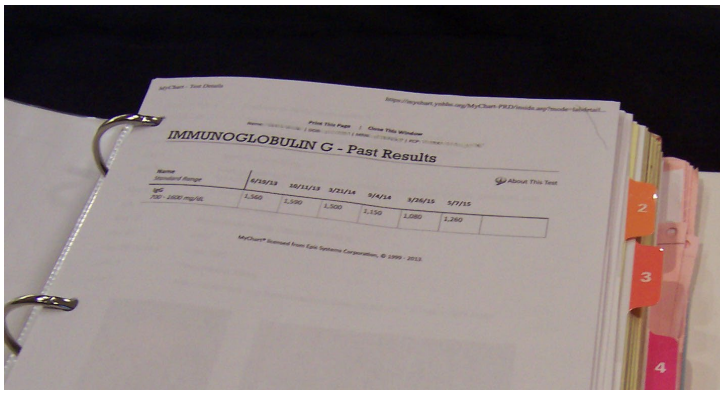
Chapter 5

Organizing Your Data

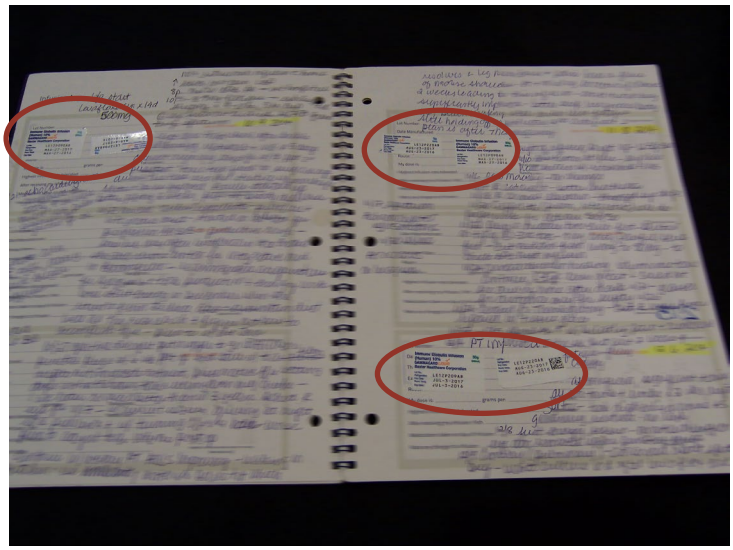
The person who cares most about your health and the documentation of your medical history is YOU. Now that you know you will be managing a primary immune deficiency disease for yourself or a loved one, you need to develop a method of organizing your information. When I started my journey, I bought a large notebook with dividers and set it up this way:

- Section One: Original laboratory results, physician summary of workup and diagnosis
- Section Two: Laboratory results once treatment begins
- Section Two: Keeping track of infections and antibiotics
- Section Four: Infusion notes (copies of nurses' notes for in-home infusion of IVIG, personal notes)
- Section Five: Information from infusion company and insurance approvals

These are photos from my original notebook. The top page is from the laboratory results section. This is a printout of my IgG levels from the patient portal of my electronic medical record.



The bottom photo is of my first infusion log book from the company that made the drug I infused. The red circles highlight the stickers from the bottles of the Ig replacement drug. All bottles have peel-off stickers for the patient. It's important to keep these in case there is a recall or you have a reaction to the drug and need to notify the manufacturer. Yellow highlights the date of the infusion. The handwritten notes summarize my health – any imaging, visits, symptoms or med changes. Note the three-hole punch so it fit in my notebook.



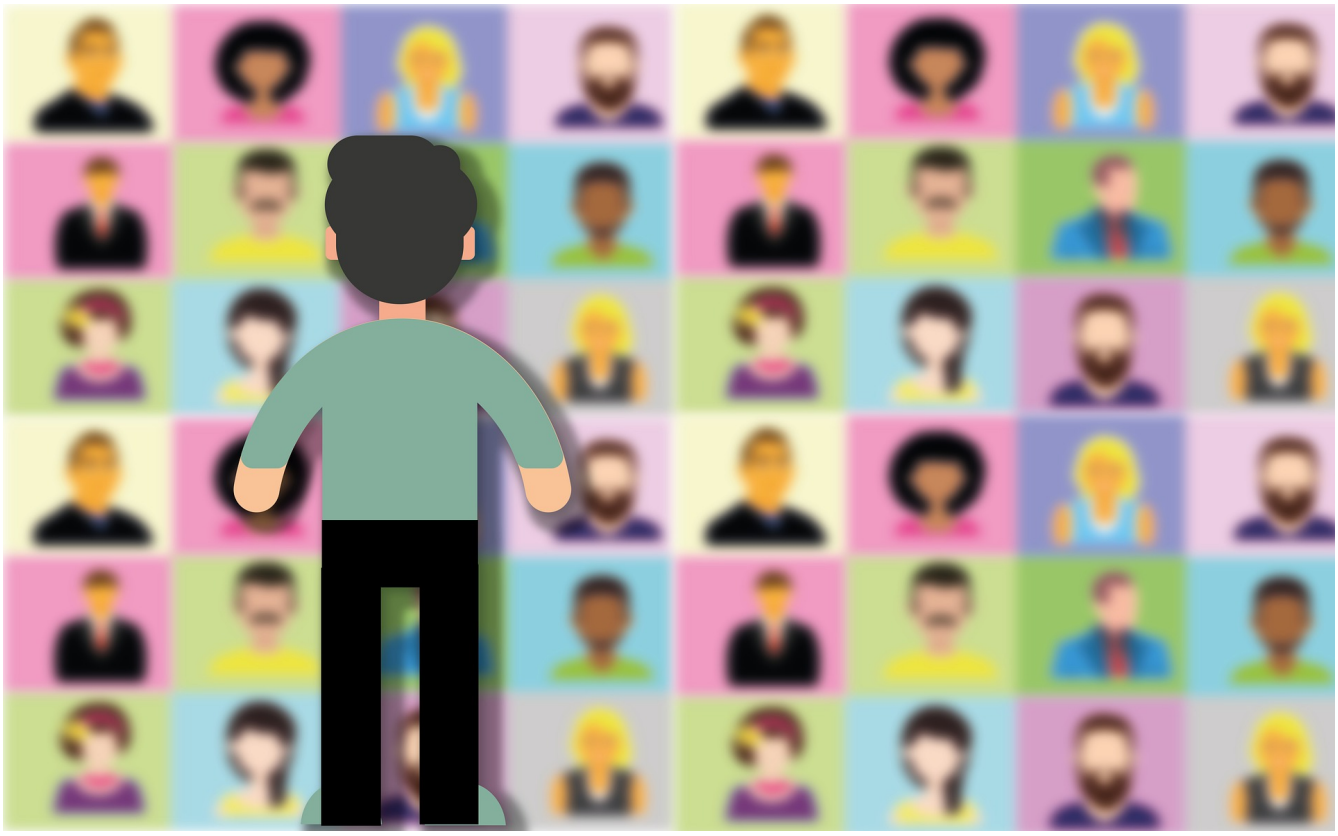
A notebook, a computer, or a password-protected phone will all work to keep track of your information ***if you use it***

If you are managing your condition with Ig infusions, check with the drug company to see what resources are available to help you track your infusions and your health status. Many companies have a "welcome kit" and infusion logs and apps.

You're not alone on this journey.

It can be hard to adopt a new habit. Typically for a habit to become routine, you need a trigger to get you started, the behavior that you want to make part of your life, and a reward. But like starting to exercise, managing a complex illness isn't easy. There will be days when you just don't have time to add one more thing. There will be days you don't feel well. There will be days you just don't want to deal with having an immune deficiency. But there is always tomorrow to start over.

And, the Immune Deficiency Foundation sponsors Get Connected Groups so you can meet others who are also dealing with these challenges. Join IDF (it's free!) and if you go to the CTZebras.com site, you'll see the meeting schedule and link to register.



Chapter 6

Summing it Up

Our goal is to provide state-specific resources for people with primary immune deficiency disease and those who love them and may manage their care. The Immune Deficiency Foundation is the leading organization providing support and advocating for us. Not only do they have written materials, videos, and Webinars, but they also have experts available by phone to help members with individual challenges.

The most important person in your care is YOU

Parents of children who have had repeated infections and adults who have been sick most of their lives have often learned to accept what health providers tell them without speaking up. If you know something is wrong and a health provider is disregarding your symptoms, then you need a new provider who will listen to you and discuss your concerns. It takes the average person seven years to finally get a diagnosis of primary immune deficiency, so it takes persistence.

For all of your health care interactions, once you find a provider with whom you are comfortable and you have your workup, ask questions so you understand what tests you have had, the results and what they mean, and the diagnosis the provider makes. Then you're a true partner in your care.

Appendix

These tools are provided to help people with primary immune deficiency disease and those who care for them. They do not constitute medical advice or specific recommendations for any individual situation.

1. Initial insurance worksheet

This form can be filled out on a computer or printed and filled in by hand. It covers the basic information that applies to almost everyone with a primary immune deficiency or having a workup. This can be put in the front of your notebook or log for quick reference.

2. Infection and antibiotic worksheet

The initial diagnosis for many primary immune deficiency diseases requires documentation of frequent infections that require antibiotics. This worksheet can be used retroactively, in conjunction with pharmacy claims, to document previous infections or it can be used to keep track of infections and antibiotics as they occur.

3. Insurance requirements

This table was compiled from insurance company documents called “Clinical Policy Bulletins” in the fall of 2020. This form is not to replace official documents, but to give patients and caregivers an overview of the documentation required by Connecticut insurance companies before they approve Ig replacement therapy.

This E-book contains *general* health information that cannot be applied safely to any *individual* situation. Medical knowledge and practice can change rapidly. Therefore, do not consider this E-book as a substitute for professional medical advice.

Initial Insurance Worksheet

Date of diagnosis	MM		DD		YY	
Patient name / Age						
Name of physician ordering Ig therapy	Physician Name					
Office telephone	Telephone					
Diagnosis and code						
Have you requested records for yourself?	Yes	☐	No	☐		
Name of drug, dose, through the vein (IV) or under the skin (SC)	Drug, dose, IV/SC					
Name of infusion company	Infusion Company					
Infusion company contact person	Contact Name					
Infusion company telephone	Phone Number					
Laboratory: IgG, IgG subclass, IgA, IgM, T cells, B cells	Yes	☐	No	☐	Requested Date	
Results of response to pneumonia vaccine	Yes	☐	No	☐	Requested Date	
Results of genetic or molecular testing	Yes	☐	No	☐	Requested Date	
Diary of infections & antibiotics past 12 months	Yes	☐	No	☐		
Results of workup of other condition that could be associated with infections	Yes	☐	No	☐	Requested Date	
Results of workup of other cause(s) of immune deficiency	Yes	☐	No	☐	Requested Date	
Insurance company and Policy Number						
Customer service telephone						

Additional Notes

Infection and Antibiotic Worksheet

Name _____

Page # _____

Date of onset	Symptoms/Diagnosis	Antibiotic name, dose, dates	Hospital / ER
9/23/2020	Sinus pressure, headache, green secretions, T 100.8 max x 3 day before antibiotics	amoxicillin/clavulanate 2000mg every 12 hr x 14 days	No

*CT Insurance Requirements**

	Aetna	Anthem	Cigna	Connecticare	Harvard Pilgrim	Oxford	United Healthcare
SCID or X-linked agammaglobulinemia							
genetic or molecular test confirming diagnosis	X		X		X		
pretreatment IgG <200mg/dL	X		X		X		
absent or very low CD3 T cells <300/mcl or maternal T cells	X				X		
pre-treatment IgG below lower limit for age or ≥2 SD below age-adj mean		X					
no evidence of renal or GI causes of hypogammaglobulinemia		X					
prescribed by or in consultation with allergist, immunologist, ENT, pulmonologist or infectious disease who treats PI				X			
frequent and severe infections				X			
Wiskott-Aldrich, DiGeorge, ataxia-telangiectasia or other non-SCID							
genetic or molecular testing confirming diagnosis	X				X		
history of recurrent bacterial infection (pneumonia, ear, sinus, sepsis, GI)	X				X		
impaired antibody response to pneumococcal polysaccharide vaccine	X				X		
pre-treatment IgG below lower limit for age or ≥2 SD below age-adj mean		X					
no evidence of renal or GI causes of hypogammaglobulinemia		X					
prescribed by or in consultation with allergist, immunologist, ENT, pulmonologist or infectious disease who treats PI				X			
frequent and severe infections				X			
Combined Variable Immune Deficiency (CVID)							
age 4 years or older	X				X		
other cause of immune deficiency excluded: drug-induced, genetic, infections such as HIV, cancer	X				X		
pretreatment IgG <500mg/dL or ≥2 SD below age-adj mean	X				X		

*CT Insurance Requirements**

	Aetna	Anthem	Cigna	Connecticare	Harvard Pilgrim	Oxford	United Healthcare
history of recurrent bacterial infections (sinus, lung)	X	X	X		X		
recurrent infections require multiple courses of prolonged antibiotics			X				
impaired antibody response to pneumococcal polysaccharide vaccine	X	X		X	X		
pretreatment IgG <lower limit of age-adjusted laboratory reference or ≥2 SD below age-adj mean		X					
no evidence of renal or GI causes of hypogammaglobulinemia		X					
pretreatment IgG below the lower limits of laboratory normal on at least two occasions more than 3 weeks apart			X	X			
lack of protective antibody titers (tetanus and diphtheria or HiB) measured 3-4 weeks after immunization			X				
Inadequate response to pneumococcal polysaccharide vaccine Age < 6 years <50% of serotype are protective (≥1.3mcg/mL) ; Age 6 or older, <70% of serotypes protective			X				
evidence that underlying conditions such as asthma or allergic rhinitis are adequately treated w supporting imaging or lab results, as indicated			X	X			
age 2 or older				X			
IgA or IgM serum levels below normal measured on at least two occasions more than 3 weeks apart				X			
IgG Subclass Deficiency							
recurrent bacterial infections (sinus, pulmonary) requiring antibiotics	X	X	X		X		
evidence that underlying conditions such as asthma or allergic rhinitis are adequately treated w supporting imaging or lab results, as indicated			X				
impaired antibody response to pneumococcal polysaccharide vaccine	X	X	X		X		
*IgG1, IgG2, or IgG3 ≥2 SD below age-adj mean assessed on at least 2 occasions**	X	X	X		X		
normal IgG, IgM, normal/low IgA	X				X		
Selected Specific Primary Immunodeficiency							
recurrent bacterial infections (sinus, pulmonary) requiring antibiotics	X		X		X		

*CT Insurance Requirements**

	Aetna	Anthem	Cigna	Connecticare	Harvard Pilgrim	Oxford	United Healthcare
impaired antibody response to pneumococcal polysaccharide vaccine	X		X		X		
hypogammaglobulinemia: pretreatment IgG <500mg/dL or ≥2 SD below age-adj mean OR	X				X		
selective IgA deficiency: IgA <7mg/dL w normal IgG and IgM OR	X				X		
selective IgM deficiency: IgM <30mg/dL w normal IgG and IgA OR	X				X		
agammaglobulinemia IgG <200mg/dL			X				
Specific Antibody Deficiency							
recurrent bacterial infections (sinus, pulmonary) requiring antibiotics	X		X		X		
evidence that underlying conditions such as asthma or allergic rhinitis are adequately treated w supporting imaging or lab results, as indicated			X				
impaired antibody response to pneumococcal polysaccharide vaccine	X		X		X		
normal IgG, IgA and IgM	X		X		X		
normal response to protein antigens (diphtheria and tetanus toxoid)			X				

*These requirements were compiled in the fall of 2020. This document is informational only. Do not make any medical decisions or take any actions without consulting a licensed health care provider familiar with your case. Insurance companies change criteria, rules, and formularies regularly. Check with your insurance company for the latest information based on your diagnosis and prescribed therapy.

References

1. University of Michigan Health
2. IGLiving Magazine June-July 2010 p16-21
3. IDF Patient & Family Handbook for Primary Immunodeficiency Diseases FIFTH EDITION. Copyright 2013 by Immune Deficiency Foundation, USA
4. Turk J Med Sci. 2019; 49(2): 497–505

This E-book contains general health information that cannot be applied safely to any individual situation. Medical knowledge and practice can change rapidly. Therefore, do not consider this handbook as a substitute for professional medical advice.