The global organisation working to improve the quality of life for people with primary immunodeficiencies.

Living With Primary Immunodeficiencies

A helpful guide for patients and caregivers

By Sara J. LeBien

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PREFACE

This booklet is written by an American mother of an adult son with a primary immune deficiency disorder; common variable immune deficiency and chronic malabsorption. It represents her family's experiences of living with chronic illness over a period of thirty seven years, and her interaction with other parents of children with primary immune deficiency as well as adult patients. She addresses the emotional side effects of this disorder, and includes experiences of other parents of pediatric primary immune deficiency patients and adult patients who kindly offered suggestions from their personal experiences. In addition, there are quotes from other publications that focus on living with chronic illness.

It is our hope that this booklet will help you be better prepared for what you may encounter, and that it offers practical suggestions for coping with typical daily challenges. In addition, it may help you maintain a more stable management of your loved one's health, as well as maintain your family's stability while living with chronic illness. It is encouraging to know that cures are being found for some of these disorders.

We do not have a cure for the disorder, but we have solutions for many of the problems. Some of us have had a difficult journey, and now we want to pass along information and suggestions to make your journey less stressful.

INTRODUCTION

PRIMARY IMMUNE DEFICIENCY or PRIMARY IMMUNODEFICIENCY disorders often go undetected because many doctors are unfamiliar with them and only treat the symptoms rather than diagnose the basic disorders. These disorders manifest themselves in a variety of ways, and the seriousness of primary immune deficiency varies considerably . Many people with primary immune deficiency have recurring infections, but nothing serious enough to require hospitalization, while others do have more serious complications requiring hospitalization. Some have few restrictions in their daily living, while others have many restrictions.

To deal with these recurring illnesses the parents or spouses of an immune deficient person face many challenges - physically, intellectually, and emotionally. It can be physically tiring to take care of a sick child or spouse along with all the other family responsibilities, and it requires questioning and research to understand the disorder. Many articles and publications are available to us through our primary immune deficiency organizations and various web sites. But living with a primary immune deficiency also has emotional effects on both the children and adult persons with this disorder and the other family members.Most of us experience several emotions while coping with PID. If we are prepared for some of the possible negative aspects of chronic illness and how they affect our family we can begin to deal with those problems effectively before they become serious.

SECTION 1: WHAT HAPPENS AFTER THE DIAGNOSIS

Being told that your child, spouse or partner has a primary immunodeficiency disorder is traumatic because it may be incurable and can be life-threatening, and chances are you know nothing about the disorder. Some in the medical sector refer to it as a disease, but the word disease is frightening to many people and needs to be understood. The dictionary says that "Disease is a condition of the body in which there is incorrect function: ailment, complaint, disorder, malady," but unfortunately, disease has an ominous meaning to our friends, teachers, and schoolmates that sometimes causes fear, suspicion and misunderstanding. It is also very painful for parents to know their child has a disease and just as frightening for the child or an adult, so many of us prefer using the word disorder.

There are literally dozens of immunodeficiency disorders. Some are relatively common, while others are relatively rare. Although there are some that affect a single cell or protein of the immune system, others may affect more than one component of the immune system.

Although immunodeficiency disorders may differ from one another in different ways, they all are the result of a defect in one or another function of the normal immune system.

An immunodeficiency disorder may be caused either by an intrinsic (inborn) defect in cells of the immune system or the immune system may be damaged by an environmental factor or agent. In the former case, the immunodeficiency disorder is a primary immunodeficiency disorder. When the damage is caused by an extrinsic or environmental factor, the immunodeficiency disorder is a secondary immunodeficiency disorder. For instance, AIDS is a secondary immunodeficiency caused by the HIV virus. Secondary immunodeficiencies can also be caused by irradiation, chemotherapy, malnutrition, burns and infections. The secondary immunodeficiences are not discussed in this publication.

The primary immunodeficiencies result from defects in T-cells, B-cells, phagocytic cells or the complement system. Most of them are inherited disorders and may run in families, such as X-linked agammaglobulinemia (XLA) or severe combined immunodeficiency (SCID). Other primary immunodeficiencies, such as common variable immunodeficiency (CVID), do not appear to be inherited; in these cases the cause is unknown but genetic factors may play a role in their causation."10

There appear to be several stages and emotions you may experience while living with PID (Primary Immunodeficiency disorder): **shock, relief, inquiry, anger, guilt, fear, acceptance, adjustment, and chronic sorrow**. You may experience some or all of these. **Inquiry** is ongoing, and **anger, guilt, fear** and **sorrow** may come and go, depending on the condition of the primary immune deficiency patient on a given day or week.

When first hearing the diagnosis, people with primary immune deficiency and their families frequently feel **shock**, What is this? What does this mean? What do we do now? This is followed by a feeling of **relief** that the condition actually has a name and treatment. Then there is a period of **inquiry**; asking questions, getting information, and finding other families living with the disorder who are knowledgeable and helpful. Hopefully, your doctor has informed you of your country's primary immunodeficiency organization that you can contact for help. Continually learning more about primary immune deficiency from your doctors, publications and other patients will keep you well informed.

After finally reaching a doctor familiar with primary immune deficiency, being diagnosed, and beginning treatment, **anger** is frequently felt and harbored for a long time toward the former medical care giver, the doctor who kept treating the infections but never suspected anything serious and therefore, never pursued testing - never questioned **why** the person has recurring illnesses. Anger and resentment also at doctors who behave in a condescending manner toward worried parents, accusing them of being "too concerned", "overprotective" or "neurotic". One mother said, "I am not angry that he did not know about primary immune deficiency disorders. I am angry that he was not curious and interested enough to try to find a reason for the recurring illnesses, and instead, simply patted me on the head and told me not to worry so much." Anger also at extended family members and friends who are not understanding and sympathetic for what you and your family are going through, and do not want to hear about it, learn about it, or share your concerns.

Guilt may haunt parents of children who have inherited X-linked or gene recession disorders, or parents who blame themselves for not having been more assertive sooner with their doctors to force a more aggressive investigation and treatment.The persons with primary immune deficiency, too, may feel guilty about requiring more time and care from their families, or for being afraid, or for not always being able to carry out their responsibilities. You may be surprised at what a young age these feelings are felt but not always expressed.

Another emotion, perhaps the most difficult to overcome, is **fear** - fear of more outbreaks of illness, fear of more uncomfortable tests, fear of a lifetime of illness. Even optimism for a future cure does not necessarily replace fear of illness next week, next month, and next year.

Acceptance comes when parentscan say "Our child has this disorder, we must live with it, and this is what we must do" or if an adult, "I must accept this disorder and make a life for myself in spite of it." It means accepting the fact that there will be recurring infections and you will have to deal with them. **Adjustment** means change - changing some elements of your lifestyle to provide constant health management. This may require changing doctors, changing the usual diet, avoiding sick friends or classmates, and being more disciplined about good hygiene habits. It also will mean making some sacrifices such as changing plans and missing events you had expected to attend. When there are recurring illnesses the family routine is interrupted, but acceptance and adjustment can become a willing routine. Refer to the sections, "A SUMMARY OF **DO's**.

It may take months or even years for you to progress through some of these emotional periods and some of them will reappear during stressful times, but you **can** deal with them so that you and your family can make the necessary adjustments to cope with the everyday challenges. Then you will understand and accept the disorder and adapt to what needs to be done for your loved one with primary immunodeficiency. This will result in better management of the disorder. You will quickly learn the signs of the onset of an illness, will respond to it swiftly, and will also learn that "this too will pass." Better days will follow. However, even after acceptance and making the necessary adjustments, a mother of a child with an immunodeficiency suggests that "chronic illness can breed **chronic sorrow** that may decrease during the well periods but increase during the sick periods." However, you must not allow that sorrow to dominate your personality and your lifestyle. That is not healthy for you, your primary immunodeficient child (or primary immunodeficient partner) or the rest of your family.

This diagnosis does not mean it is the end of the world. There will be periods of better health too when your family will engage in travel, holidays, sports, all the normal things families do, and you will have a greater appreciation of those good times. There are many others living with the same primary immunodeficiency disorders who can be supportive and informative. You can meet them through your national organisation linked to IPOPI (International Patient Organisation for Primary Immunodeficiences) countries from around the world. (Refer to the listing of organisations plus the E-mail addresses and Web sites.)

Through an enquiring approach, your understanding will be increased and you will experience better control of the condition. This is empowerment which leads to an improved quality of life. Also bear in mind that the primary immunodeficienies are at the cutting edge of medical science and new possibilities for treatment are being developed through bone marrow transplantation and gene therapy.

SECTION 2: WHAT TO EXPECT AS YOUR CHILD GROWS OLDER
INFANCY THROUGH ADULTHOOD

Infant and preschool years

This is a very difficult period because your young child is helpless and cannot communicate with you verbally. It is painful to see him/her in the hospital, so your emotions are overloaded. If your child is sick at home you get little sleep, your family's usual routine is altered, siblings might have to make sacrifices, and the constant vigil of the child's temperature and medication is stressful. The questions become second nature - Is he coming down with an infection? Does he look better today? Worse? Should I call the doctor? Should I do this? Should I do that? At first it is frightening , but in time you will calmly take it all in stride as you develop a routine of recognizing signs and symptoms of illness and responding correctly.

Early school years

It is important that you have a good relationship with the school your child attends at all levels, and that your child's teachers are aware of primary immune deficiency so that there is no confusion with AIDS. Your child may want to take the children's book, OUR IMMUNE SYSTEM to school for the teacher to read to the class. A young boy took this book to school with his new title, THIS BOOK IS ABOUT ME, printed over the original, and took great satisfaction in seeing that his teacher and schoolmates understood primary immunodeficiences and his infusions. You may need to sit down with the teacher and explain in detail about your child's disorder, particular signs of an oncoming illness, treatment, and any special needs. If the teacher is not sensitive to your child's situation, you may wish to speak to the Principal or Head Master and request a different teacher. While your child is home sick it may help him or her to look through family picture albums showing happy events and healthier moments in their lives.

Adolescence and Teen years

At this time there may arise some subtle problems for your child. Many primary immunodeficient children and young adults are active participants in sports, but for others less physically demanding activities may need to be encouraged. Some boys with primary immunodeficiency tend to be physically smaller. In an environment where his physical strength and sports ability may be highly regarded by his peers, he may feel left out, embarrassed, experience low self esteem and even depression.

Keep a good line of communication open between you and your son so that you know what he is experiencing . You may be very surprised at his mature concerns. This is the time to encourage and support his interests in less physically demanding activities. Perhaps instead of being on a team he could pursue being a sports writer. Photography and computers offer all manner of hobbies or later professional pursuits.

A young man with CVID (Common Variable Immunodeficiency) and chronic malabsorption commented in retrospect, "I think all kids want to fit in. I wanted to fit in, to feel a sense of belonging. Illness can really isolate you if you can't spend time with your friends and miss out on important social occasions. I got depressed from feeling inferior, taking medications, being examined by doctors, feeling sick, and waiting to get well. After years of doctors and nurses touching me and sticking me with needles, not to mention all the other uncomfortable and degrading tests, I kind of disconnected from my body emotionally. I not only felt inferior physically, I also felt frustrated and depressed that I needed to consent to it all to get better.

" The depression is lifted after getting well, but if you get sick again, the depression can come back even worse. It is the repeated illnesses and long drawn out illnesses that really depressed me. When you have a history of illnesses and know that another infection or problem will mean doctor visits, side effects from drugs, maybe a hospital stay, weeks or months of feeling sick, needles, and missing out on opportunities, the flood of all those feelings you've had before come rushing in. And even worse, your sense of hope gets beaten down. You get pessimistic about life which is a very disempowering attitude." Help your child find a way to fit in, to feel that sense of belonging. Make extra effort to support and share your child's interests or talents. Seek organized groups sharing his interests. A chess club? A photography club? Or start your own.

Young people at this age are interested in their appearance and social life. A steroid-induced puffy face or missing parties and school functions because of illness is depressing, and it also hurts the parents seeing their child endure this kind of pain. You and your child together will need to find alternatives to those missed events. Create your own celebrations to include their friends when better health permits. Of course this will not entirely replace what was lost, but it may soften the disappointments.

One mother offers her experience: " Girls like pretty things such as earrings in pierced ears, chemicals called makeup, and fashionable shampoos, but pierced ears allow infection, and make-up and shampoos may cause skin problems. In addition, females, by nature, are often more expressive of their emotions and this is often not acceptable in a male dominated health sector, so young girls learn at a very early age not to express the emotional impact of it all. Not wanting to act in an attention-seeking manner or be labeled as 'bad' or 'not coping', she might actually be at risk of being ignored. Being too placid and undemanding could result in doctors and parents not really hearing and therefore, not understanding what the child is experiencing physically and emotionally. She may instead show her emotions at home, late at night when nobody else except her parents are there to hear and console. Also, a girl may prefer a female doctor, or want her mother in the examining room with her during checkups and infusions." In most hospitals this is normal policy for all children, but unfortunately, some doctors still have not realized how beneficial this is emotionally to both the child and the parent, and still refuse their presence.

But girls are not the only ones intimidated into silence. This is frequently true for young boys as well. The attitudes of the physicians and nurses are partially responsible for the emotional comfort of the patients and their parents, and people working with pediatric patients are normally caring individuals. However, some doctors' offices become so crowded and busy that the doctors and nurses lose their compassion and patience. If you or your child are not comfortable with your physician, and you have options, change physicians.(See the section on developing good relations with your doctor.)

Usually our children are very sick the first time they see the inside of a hospital, but if your child's first hospital experience is for a non-emergency purpose, and if your child is fearful of the "unknown", you may want to visit the hospital prior to the appointment day. One mother took her son before his scheduled surgery to see the pediatric ward; where the nurses' station was located, and what the children's rooms looked like. The friendly nurses and the colorful Disney characters painted on the walls satisfied his curiosity and eased his concern.

Teenagers do not want to be different, and they may not want their peers to know they have a health problem. During high school your teenagers may begin to withdraw from participating in school activities because of frequent illnesses which requires canceling activities. These frequent disappointments may cause them to stop trying. They may begin spending time with more solitary activities such as music, reading, photography and computers. These are all good interests that will enrich their lives, but try to encourage your children to maintain contact with others, even if it is only a few friends who understand their health problem and its restrictions. If you participate in their school activities, most likely they will maintain some level of participation.

Make sure the school staff is also aware of your child's problem to insure their sensitivity to his/her needs. An understanding school staff can make a difference. A student with primary immunodeficiency and malabsorption who needed closer access to a toilet was given permission to use the teachers' restroom nearer his classrooms, and when this student was home-bound for several weeks his teachers cooperated with his private tutor to help him keep up with his studies. Such cooperation and sensitivity from the school Principal and teachers provided a continuity and connection to the school.

If parents talk too much about the financial problems and other frustrations, the child with immunodeficiency could grow up feeling guilty of being a burden to the family. Depression could become a serious consequence. Early signs of this should be taken seriously and professional help sought.

While attending university, children may go through a period of denial of their disorder and even refuse infusions. Explain your objections and concerns calmly, ask questions and demand thoughtful consideration on their part, and then let them make the decision. They may get sick before consenting to resume a normal schedule of infusions. You must allow your older teenager to make his or her own decisions, with your guidance, by this time. You are no longer in charge after years of being totally in charge. When entering the university, your child may want to contact the university health center and go in to meet the doctor. Have your immunologist send your child's history so that the staff will be familiar with your child before an illness sends him/her there.

Holistic medicine is of great interest at this age, and if your child becomes dissatisfied with the usual medical treatment, he may try alternative means such as homeopathy, meditation, acupuncture, acupressure, etc. Do not be alarmed. Let your child try what gives him hope, but so long as it is discussed with his doctor. Your child will find out if it really helps or not. It might help psychologically for a while, if not physically. Keep in mind that this is part of your child's need to be in charge, and this is healthy.

Many young people with primary immune deficiency have a normal active social life, but for a few, recurring illness creates a lack of trust in one's body. What will my health situation be next week? Next month? Should I try to plan anything? Will I feel well enough to go? Will that event interfere with my infusion? It becomes easier to withdraw. Just as some parents may stop trying to plan, in case their child will be sick, the immune deficient person may stop trying to plan and slip into a solitary lifestyle. From their child's early years, parents must focus on nurturing positive thinking and encourage their child to take some risks. While accepting the possibility of illness disrupting a planned activity, primary immunodeficient persons should still plan. They should not go through life allowing possible illness to prevent them from taking risks that lead to many successful experiences and joyful events. Go ahead with plans for a backpacking or ski trip. Just because they missed out on one doesn't mean they will miss the next opportunity.

Encourage your teenager to join a support group. Sometimes this is resisted because during the well times he/she doesn't want to be reminded of this disorder. One young man said, "I just want to get on with my life and not think about it." But developing such a relationship could prevent a feeling of isolation and loneliness resulting from recurring illnesses. Even if this contact is through E-mail it would be a connection to others who share the same challenges and can provide empathy and friendship.

The Children's Hopes and Dreams Foundation, Inc. in the U.S. serves children with chronic illnesses with its Pen-Pal Program for ages 5-18. They match children by age, gender, and within a broad catagory of illnessess to help them develop friendships and support with someone who understands and relates to them and their illness. Hospitals and healthcare professionals can provide the necessary applications. IPOPI member organizations around the world could even create their own pen-pal programs matching children with primary imune deficiency. Older children may prefer Email or the IPOPI Web site's Chat Room if they have access to a computer.

Young adulthood

Soon your young adult will be planning a career. In the U.S. this is when medical insurance coverage can become a problem. No longer covered by the parent's policy, the young adult may have group insurance where he is employed, or must obtain his own private insurance. However, the Immune Deficiency Foundation in the U.S. and pharmaceutical companies that produce gammaglobulin have insurance specialists, 'Reimbursement Specialists', who advise and help in these matters. In countries with nationalized medical care the cost of treatment is not as much of a problem.

Other concerns for the future are the demands of a full-time job, maintaining personal relationships, marriage, and whether to take the risk of having immune deficient children. (Before a pregnancy, a couple may want to discuss this with a genetic counselor to help them evaluate their risks.)

These are all very serious considerations requiring thoughtful discussion and encouragement, but always remember that **it is quite possible to live a reasonably good life within the restrictions and responsibilities of this disorder** - there is a man with primary immunodeficiency in the U.S. who not only is married and has children, but is a doctor - an immunologist!

SECTION 3: ADULT ONSET

"Many adults who were born with apparently normal immune systems go on to develop a primary immunodeficiency late in adolescence or even in adulthood. Just as with pediatric patients, the positive side of having a diagnosis is that the uncertainty is over and you can now learn about your disorder, achieve a better state of health as a result of treatment, and make the necessary adjustments to your lifestyle. It is important to distinguish between the adult onset primary immunodeficiencies and AIDS. Because some primary immunodeficiency disorders have their onset in adulthood they are sometimes called "acquired" immunodeficiencies even though they are not caused by the virus that causes AIDS.

The most common immunodeficiencies to affect adults include: the antibody deficiency diseases (Selective IgA deficiency, Common Variable Immunodeficiency, Immunoglobulin G subclass deficiency, and X-linked Agammaglobulinemia) and less commonly disorders of the phagocytic cells (Chronic Granulomatous Disease) and disorders of the complement system.

In most cases, the well informed patient, working with attentive medical staff should be able to pursue a career and live a full life. The newly diagnosed adult must face questions and problems which have already been faced by children who have grown up with these disorders. Feelings of self-pity and fear are quite normal. It is important to realize that you are still the same person, but that you have to come to terms with your diagnosis and treatment and live with them as you go about your life.

An adult with a primary immune deficiency disease has all of the medical problems that a child would have, and yet by the definition of adulthood, is supposed to be responsible for his or her life, career, financial planning, and the future of his or her children. Obviously, this can bring various degrees of stress into a family. It is important to discuss with your physician these aspects of an illness, just as much as one would discuss a physical problem. Sometimes just expressing your fears can have a therapeutic effect.

There are a variety of ways to help keep your frustrations and anxieties to a minimum. You may simply require some time to discuss these feelings with your partner or spouse, understanding friend or health care professional. Many people are helped by meeting with others in a support group setting. Your country's primary immunodeficiency organization can give you information about a support group. Learning as much as possible about an illness is one very specific way to guard against confusion about the illness itself. You will want to make it very clear to your friends and family that it is not AIDS.

One of your main challenges is in maintaining your school or work attendance. You will need to fit your treatment into your schedule. Receiving your infusions of gammaglobulin from Home Care providers may be more helpful, or learning to administer your own infusions at any time of day or night is especially convenient for an adult student or working person. Also, career choices that provide flexible hours or a home-based work environment make life much easier for patients with frequent illnesses".

If school or work is stressful, it would be helpful to learn skills for dealing with stress. You might try meditation, visualization, yoga, or Tai Chi, and the library and bookstores offer many books regarding relaxation skills and stress management.

Laughing can divert your attention, cheer you up and help you put things in perspective. Research even shows that it can be good for you physically. It causes the release of powerful natural pain relievers in your brain...it can help get more oxygen to your blood...11 Look for opportunities for laughter - comedy movies and plays, the antics of animal pets, expressing your sense of humor, and making sure you are in the company of cheerful people as often as possible. Seek activities that are fun. Do not forget the benefits of exercise. If your energy level is low, mild exercise can be invigorating. One university student with primary immunodefiency felt very fatigued until a long bicycle ride rejuvenated him! Remember also the benefits of good nutrition.

Malabsorption, however, which sometimes accompanies primary immunodeficiency, creates other problems. People who have recurring intestinal problems and cannot eat solid food lose social contacts because they have a low energy level and are uncomfortable participating in events focused on eating. Much in our society is based on eating - holiday gatherings, picnics, dining out for dinner, etc. How can a young man invite a young woman out for dinner? How can a young woman with the problem accept? How much fun is it to sit with family or friends at a table laden with food and listen to them comment about how delicious everything is? The result is that people with malabsorption may withdraw from social interaction and gradually fall into a solitary life. This can become a lonely solitary life, especially for a teenager or young adult, unless that person makes great effort to participate in other forms of social interaction.

You should actively participate in your own treatment, but this is nearly impossible during an illness, so you need an advocate or care giver. This could be your spouse or partner, another family member or friend, someone to help communicate with your doctors, provide care, and give you emotional support by boosting your morale. When this illness passes you will once again be strong enough to assert your authority and be more in charge.

"You may experience a feeling of loss - loss of friends and experiences you can no longer keep up with, loss of trust in your body, and loss of peace of mind because you must focus on your health condition and management".12 One young man who experienced this said, "If you find yourself in a ditch, you get out of it." He 'got out of it' through discipline and perseverance, working hard at taking responsibility for improving his health through nutrition supplements, various forms of exercise, meditation, and the pursuit of his special interests and talents when he felt better. A man in Germany with hypogammaglobulin anomaly and malabsorption expresses his feelings about his illness this way - "Because of all the demands on me from this illness, I have to make an effort to remain in good spirits. Those who are rarely spoiled by life, treasure their moments so much more even if those moments are numerous. As long as I have my eyesight I can take great pleasure in the beauty of nature and of art. Moreover, the opportunities I have to hear music bring enormous joy. And also, in spring and summer, I delight in the ensemble sounds of the birds. All of these things have provided great support to me and have given me the will to live and the necessary energy for self discipline."

You must rely on your normal coping methods and devise new ones to confront the changes that have occurred in your life, and reevaluate your lifestyle to meet the challenges you face now. Try to learn to take life as it comes, not in anger but with patience. Primary immunodeficiency is part of your life, but not all of it.

"A true handicap is what we don't do with what we have, not what we can't do with what we don't have." 13

SECTION 4: IMPORTANT ELEMENTS FOR SUCCESSFUL COPING

In addressing the emotional aspects of living with the Primary Immunodeficiency disorders, in my opinion, there are five very important elements in successful coping for the patients, parents and families.

1. Be knowledgeable about the disorder.
2. Maintain a positive attitude.
3. Accept limitations and changes in your lifestyle.
4. Have a support system.
5. Some people find having a healthy spiritual life is helpful.

1. BEING KNOWLEDGEABLE

Learn about your particular immunodeficiency disorder so that you can communicate intelligently with your doctor and nurse, your own family, and others involved with your family. Read, ask questions, **learn**. Here are suggestions from Dr. Ted Wymslo, Director of the Miami Valley Hospital Family Health for Caring magazine:

"TIMES HAVE CHANGED: IT'S OK to ASK YOUR DOCTOR QUESTIONS"

How to get the information you want from your doctor:

Write down questions before you go into the office.

Be assertive (that's different from aggressive) about raising your concerns. If the conversation gets sidetracked, bring it back to your concerns.

Try to be specific in your descriptions of the symptoms or problems.

If the doctor is called out of the room while you are talking, help him/her to remember where you were in the conversation.

Do not be embarrassed to ask for an explanation of a medical term. Ask the doctor to explain it in simple terms until you are satisfied that you understand the explanation. Ask the doctor to draw a diagram or sketch that will make the idea clearer.

If you want more information, ask the doctor about pamphlets or books on the subject. Write or call local branches of national organizations related to your illness for literature.

Join a support group. Besides encouraging each other, care givers can help teach each other practical solutions to common problems.

Be considerate of the doctor's next patient. When you make the appointment, let the receptionist know that you may need extra time for discussion.

Ask, "What symptoms or signs of improvement should I watch for?What are the side effects of this drug? How does this disorder usually progress?

Bring along a friend or family member to help you listen and take notes for you.

Repeat anything vital that's been communicated. Make certain that you understand and have written down what it is that you are to do when you leave - especially having to do with drug dosage or dietary or activity restrictions.

Remember that there are some questions that your doctor simply cannot answer. There are unknowns.

Remember that you have options. You are entitled to a second opinion. If you find your doctor to be consistently uncommunicative and unresponsive to your questions, it may be time to seek another physician."2

HELPFUL PUBLICATIONS:

Several national organizations within IPOPI offer excellent publications explaining the primary immune deficiency disorders and treatment.

Living with Primary Antibody Deficiency

What are Primary Immune Deficiencies?

Travelling with Primary Immune Deficiencies

The History of Immunoglobulin Therapy

Information for IgA Deficient Patients

About...Subcutaneous Immunoglobulin Therapy

About...Intravenous Immunoglobulin Therapy: Information for Patients
Available from:
PiA (Primary Immunodeficiency Association of the U.K.)
Alliance House
12 Caxton Street
London SW1H OQS
United Kingdom Telephone: +44 207 976 7640 +44 207 976 7640
FAX: +44 207 976 7641

PATIENT AND FAMILY HANDBOOK
For the Primary Immune Deficiency Disease
Our Immune System (a children's book)
Available from:
Immune Deficiency Foundation
245W. Chesapeake Avenue Suite 206
Towson, MD 21204 USA Telephone: 410 321-6647 410 321-6647
and 1- 800-296-4433 1- 800-296-4433

CONSENSUS DOCUMENT FOR THE DIAGNOSIS AND MANAGEMENT OF PATIENTS WITH PRIMARY ANTIBODY DEFICIENCIES
Available from:

The Royal College of Pathologists
The Royal College of Physician
The Primary Immunodeficiency Association (PiA)

TEN WARNING SIGNS
Available from:
The Jeffrey Modell Foundation Inc.
43 West 47th Street New York, N.Y. 10036 U.S.A.

UPDATE
Available from:
International Patient Organization for Primary Immunodeficiencies (IPOPI)
Alliance House
12 Caxton Street
London SW1H OQS Telephone +44 207 976 7640 +44 207 976 7640
United Kingdom FAX: +44 207 976 7641

2. MAINTAINING A POSITIVE ATTITUDE

Lemons or Lemonade?

A positive attitude will turn lemons into lemonade, and to get and maintain a more positive attitude we need to give ourselves opportunities to hear the good things too and not be consumed by the sorrow. We can learn from the positive experiences of others. If you are the person with primary immune deficiency you must try to do everything in your power to be as healthy as possible. This means staying on schedule with your treatment, following a good routine of getting necessary medical attention when symptoms first occur - don't put it off. It may mean being more aware of good nutrition and hygiene - beware of germs! Beware of people with germs!

If you are the care giver, you must try to keep yourself healthy physically and mentally. Trying to be perfect and all things to all people is exhausting and not conducive to optimism. Make time for yourself. If you feel good about what you are doing for yourself, you will have a more positive attitude.

One of the biggest problems for care givers is fatigue, and this damages your spirit of optimism. "The widespread belief that not getting enough sleep increases susceptibility to infections and possible other illnesses appears to be confirmed by recent studies.

Sleep stimulates the production of various cytokines and other protective components of the immune system, and in laboratory animals, sleep deprivation is associated with depressed immune system function. In humans, healthy slow-wave sleep is associated with a marked increase in certain immune system activities, and a decrease in those components associated with fatigue and drowsiness.

Stress is a major cause of insomnia and can also depress immune system function. Since lack of sleep itself is stressful, this can set up a vicious cycle. Chronic sleep deprivation is common and insidious, since it may take months for symptoms to appear".5

Parents of young primary immune deficiency patients commonly suffer from loss of sleep and find it difficult to remain emotionally strong when suffering from fatigue. It is normally the mother who is home all day with the sick child and many times, siblings, caring for them along with household responsibilities, trips to the doctor, grocery, etc. Then at night she may not get enough sleep due to a coughing, feverish child. The husbands normally are away at work all day but provide some relief during the evening hours. When a husband realized how exhausted his wife was, he hired a sitter for their children and took his wife to the mountains for a weekend alone. The young mother spent the entire week-end sleeping. She returned home refreshed physically and emotionally.

On the Sunday morning of Mother's Day, a father took their early rising children to a park at 5:30 A.M. to allow his exhausted wife to sleep. What a wonderful Mother's Day gift!

Since neither parent has the opportunity to make up the sleep loss during the day, it is important for them to find a way to take turns with the caregiving at night during an illness, to allow the other to get needed sleep. In situations where there are more than one family member with primary immunodeficiency this is especially stressful. Being a care giver for a sick child is a **joint effort**. A single parent has a special need for additional help, especially if that parent needs to maintain a job outside the home. In the U.S., the Family and Medical Leave Act provides parents with a (limited) paid ' sick leave' to care for sick children. Recurring or lengthy illnesses, however, may make it necessary for a regular care giver to help the single working parent.

Besides finding ways to catch up on sleep, there are other things you can do to care for yourself. Delegate household jobs so that everyone is helping, pay attention to your diet to make sure you are getting proper nutrition, make time for physical activity such as jogging, bicycling, or walking, and do something pleasurable for yourself - read, write or sketch in your personal journal, plant a flower bed, etc. Ladies, take a leisurely bath by candlelight, invite someone for tea in the afternoon, watch a humorous TV movie, etc. And for men, it might be good for you to attend a sports event with men friends or pursue a hobby. It will not be easy to find the time or to feel guiltless taking the time for yourself, but you need it and you deserve it.

The mother of two sons with agammaglobulinemia writes in Caring magazine:

"How to Develop Optimism"

Identify when you are engaged in negative self talk.

Ask someone to tell you when you speak negatively.

Use positive, empowering phrases several times daily. (Tell yourself you CAN do this job or handle this infusion, you CAN ask for help, you CAN say wha you need.) Try to be around positive people.

Don't put yourself down when you make a mistake

Don't give up. Change takes time!

Though it may not be possible to control the situations in our lives, we can control what we say to ourselves about them. If we tend to focus only on the negative, it will stamp out the positive. As a parent, my attitude is being copied by my children. My children are learning patterns of coping and dealing with life based upon what I do and say every day around them. Parents have a remarkable opportunity to mold a child's approach to life, and a parent's optimistic attitude will be modeled by a child, giving him or her a wonderful asset."3 However, avoid unrealistic, insincere or forced positive remarks.

Charles Swindoll writes about **attitude**,"The longer I live, the more I realize the impact of attitude on life. Attitude, to me, is more important than facts. It is more important than the past, than education, than money, than circumstances, than failures, than successes, than what other people think or say or do. It is more important than appearance, giftedness, or skill. It will make or break a company...a church...a home. The remarkable thing is we have a choice every day regarding the attitude we will embrace for that day. We cannot change our past...we cannot change the inevitable. The only thing we can do is play on the one string we have, and that is our attitude...I am convinced that life is 10% what happens to me and 90% how I react to it. And so it is with you...we are in charge of our attitudes."

3. ACCEPTING LIMITATIONS AND CHANGES IN YOUR LIFESTYLE

Part of the adjustment means making some changes and some sacrifices. The family routine may be upset occasionally. Siblings may need to miss an anticipated activity because the parents need to care for the sick child. The working parents may need to rearrange their work schedule or work related activities in order to be care givers during an illness. Social life may be disrupted. But these inconveniences can be worked out successfully based on family unity, love, and understanding. The disappointments can be balanced with other enjoyable activities at another time. Adults with primary immunodeficiency may need to adapt to a different work environment that provides flexibility in work hours and a healthier work place. In this era of high tech, many people are working at home with computers. Young adult patients preparing for life's work must be realistic about their health and may want to consider professions that offer flexibility and a healthy environment.

4. HAVING A SUPPORT SYSTEM - HOW FAMILY AND FRIENDS CAN HELP

Grandparents and other relatives as well as friends and neighbors should understand the immune deficiency disorder your child or spouse has and what that means to your family. However, it is not unusual for some relatives to avoid discussing it, preferring to remain detached from your family's situation, and some relatives may never reach a level of understanding and emotional support you need and desire. But you should not allow your hurt, disappointment and anger with them to build up to the point that it damages the family unity, and becomes another stress in your life. Accept your relatives as they are, forgive them for not being willing or able to cope with your problem, and turn to your friends instead. Their interest and loving support are therapeutic, and their willingness to help in practical ways provides both physical assistance and emotional strength when you need it.

It helps to have a group of willing friends to call upon for help so that one person is not called too often. And don't feel guilty that you aren't doing anything in return. You will no doubt have an opportunity later to help them in some way, if it will make you feel better, but remember, a good friend does not require repayment.

HOW SUPPORT GROUPS, E-MAIL AND THE WEB CAN HELP

Each country that is a member of IPOPI (International Patient Organization for Primary Immunodeficiencies) has its own immune deficiency organization that provides literature, newsletters, and occasional local meetings. These meetings for patients and their families offer an excellent opportunity to learn from guest speakers and to get acquainted with each other.

Communicating with others in similar situations can be very helpful in many ways. Just knowing there are other people experiencing your same challenges can be comforting, and talking with them in person or through E-mail can be helpful in practical ways as well as emotionally. IPOPI members gain strength from each other in sharing their personal experiences and their knowledge about medical treatments, hospitals, physicians, and information about current research.

Reported gains from self-help groups:

* Acceptance of me as an O.K. Person
* The feeling that I'm not alone
* Knowing that I'm not lazy, crazy, or neurotic
* Validation that it's not "all in my head"
* New friendships, networking
* Reinforcement for efforts toward positive action
* Help with goal setting
* Improved communication skills
* Help in becoming better informed about my disease

There are more than 10,000 health-related web sites alone and thousands more on-line support communities. For Web surfers and E mail readers, a good place to start is the IPOPI web site where you will find excellent information just for primary immunodeficiencies, a guest book, a Chat room, and a Forum for interactive communication with others going through the same thing you are experiencing.

Places to go for medical information:

American Association of Immunologists:
http://glamdring.ucsd.edu/others/aai

Health World (provides free access to Med Line):
http://healthy.net

Immune Deficiency Foundation:
http://www.immunedeficiency.org

International Patient Organization for Primary
Immunodeficiencies (IPOPI)
http://www.IPOPI.org

the Jeffrey Modell Foundation)
http://academic.mssm.edu/peds/modell/guidelin.html E-mail address:
Modell@AOL.COM

Journal of Immunology:
http://ji.journals.at-home.com/JI

Medaccess: http://www.medacess.com

Medinfo:http://www.medinfo.org

National Institute of Allergies and Infectious Diseases
http://www.niaid.nih.gov/publications

The National Organization for Rare Disorders (NORD)
http://www.nord-rdb.co
Email address: orphan@nord-RDB.com
(provides rare disease information through the Rare Disease Database on the World Wide Web)

OncoLink: http://www.oncolink.upenn.edu/

Yahoo's Health section
http://www.yahoo.com/Health

5. KEEPING A HEALTHY SPIRITUAL LIFE

Having a sick child or a sick spouse or partner, or being the immune deficient patient, is a heavy burden, too heavy to carry by yourself. Bernie S. Siegel, MD, retired general and pediatric surgeon in the U.S. who writes about humanizing medical care, said he thinks life "is like an enormous gift that is handed to you that may be hard to carry, and you may need some help carrying it."7

Don't forget your spiritual needs. Whatever your religious faith, reach out to connect with a higher power for renewed strength, guidance and peace.

In Living With Chronic Illness, author Cheri Regester, who has personal experience living with a chronic congenital disease, says "...illness and troubling questions it raises can and do challenge, reform, and even enrich religious belief, and that this renewed faith, in turn, aids endurance." She continues to say," I find that my faith is a very heali thing for me, because I know it's always there, it's a constant and it's not going to go away".9

An adult with primary immunodeficiency recommends finding a spiritual path you connect with, and believes that the power of spirit and mind should not be underestimated.

SECTION 5: A SUMMARY OF DO NOT'S WE SHOULD BE AWARE OF

Do not allow negative energy to dominate you. This can wear you out physically and emotionally, so that eventually it can even change your personality into a tired, withdrawn, sorrowful, angry, or complaining martyr. It is easy to go through periods of feeling very sorry for the member of your family who has an immunodeficiency, or for yourself, but this can isolate your friends and even your spouse or partner. Those with primary immunodeficiency often suffer from fatigue also, particularly when they have a chronic or low grade infection. At this time chemicals are released into the blood that cause a variety of symptoms including weariness, loss of appetite, weakness, moodiness, and even depression. The state of mind may need to be treated also just as the physical illness needs to be treated. Discuss this with your doctor. Do not ignore depression.

Do not allow caregiving for the sick to so dominate your family that you neglect the needs of your other children or spouse. Jealous siblings or jealous immune deficient children could become a secondary problem to deal with. It is only normal that the sick child may feel left out of many activities the other children are enjoying, or the siblings might feel neglected when your focus is on the sick child. Do not let resentment become a permanent attitude. All family members have to sacrifice sometimes. There simply are things you will occasionally have to give up - a family vacation, planned excursion, evening out, school field-trip, camp, birthday party, etc. Sickness comes at inopportune times and you will need to adjust to this and find ways to compensate for those disappointments. Do fun things on the spur of the moment on the good days and allow less important things to wait.Don't stop planning and doing, but be realistic, of course. Try to do as much as possible inspite of your circumstances. A father of two sons took turns taking each one alone on an excursion of the child's choice. Each son received the full attention of their father during an interesting and fun outing, and the sibling of the child with the disorder accepted disappointments at other times due to his brother's illnesses without resentment.

Do not suffocate your child with too much love and over-protection. Our children with primary immunodeficiency need extra TLC (Tender Loving Care) at times, but then let go and allow your child to experience life. Discuss the necessary restrictions for your child with his doctor, but in most cases, let him go to camp, backpacking, play in sports (even when it means spending the rest of the day lying around the house very tired), and let him be outside in cold weather. Know the health conditions of his playmates, in case he needs to avoid some children at times, but do not overdo it. **Do not let fear of sickness hang a dark cloud over your lives.** Let your child take certain risks and grow up unafraid. This will result in a continued desire to pursue interesting experiences during healthy periods. An American university student with Common Variable Immunodeficiency (CVI) was allowed to go backpacking alone in Australia and New Zealand.This young man's parents trusted his good judgment in taking care of himself. He returned home weeks later very tired but thrilled with his experience and a renewed confidence in his abilities. Through the years of coping with primary immunodeficiency your child will learn what his/her abilities and limitations are, and will adjust to the limitations. As parents we must not forget to emphasize and support his/her abilities, not just dwell on the limitations.

Do not accept a lack of communication. Talk, talk, talk in your family. It is just a matter of interested questioning which leads to good discussion and understanding. It is easy to do this with healthy, active children, but some parents may feel more reticent to hear what their sick child has to say, but that is exactly what you need to keep in touch with. This is especially necessary during the adolescent and teen years when your child may become more private and unwilling to express his fears and frustrations. Do not wait for him to bring it up - ask.

Parents may choose not to talk about the emotional stress in order to protect the primary immunodeficient child from possible painful discussion, and the child may think he is protecting his parents by not expressing his true feelings, all the while missing valuable time and opportunities to share and help one another through the stressful times.

Do not try to be everything to everybody. Somehow we think we can be superhuman. Wrong! We do not have to be a volunteer in school or community organizations, so **do not** let anyone make you feel guilty for not helping. Stand firm. Say NO! Keep yourself calm and in good health so that you can focus your energies on your family and yourself. Put off trying to be an excellent housekeeper or gardener (unless that is your therapy). Make time for yourself. This is easier said than done, but it is important for you to have pleasureable activity and not become consumed with being a caregiver to your child or partner.

Do not let your marital relationship suffer. Being a caregiver to a sick child is time consuming and emotionally and physically draining from loss of sleep and the strain of living with illness and trips to doctors and hospitals. This can be damaging to a marriage, The mother may become too much of a full-time home nurse and neglect the needs of her husband. She must be sensitive to his additional responsibilities and pressures at work. Likewise, the husband must be sensitive to what his wife is going through all day caring for a sick child while he is at work, and should not put additional demands on her. If both parents work there is additional stress. **Both need to be understanding, unselfish, patient, and willing to work out the problems that may occur.** Take time for the two of you to be alone, to simply sit down together at the end of the day without the TV or noise and interruptions of the children. Declare "Mum and Dad's Talk Time- Do not Disturb"

Do not be afraid to ask for help! Parents of immune deficient children or a spouse of an adult with primary immunodeficiency learn to be vigilant, organized, decisive and calm, but this can give the impression they do not need help. There are times when the most experienced and capable caregiver needs help - time to yourself for an hour or a morning, a simple errand, a chance to rest and to catch up on much needed sleep, or someone to listen to you when you are hurting. When someone says, "Call me if you need anything, **call!**

Just remember, you are not alone. There are many who have had or are having the same experiences you are having. As mentioned before, there are patient organizations in each of the IPOPI member nations available to you by telephone, and some by E-Mail and FAX. IPOPI has a web site with a Chat Room and Forum for your interaction. Many of the national organizations hold local meetings with excellent speakers from the medical sector, and they send informative newsletters and print valuable publications. If you do not live near enough to attend their meetings perhaps audio tapes of the talks or discussions could be loaned to you by mail.

Self-help books can be found at the local library and bookstores, and for more personal assistance, hospitals have counselors. Do not be embarrassed, ashamed or afraid to get this professional help.

SECTION 6: A SUMMARY OF DO'S TO KEEP IN MIND

Do learn about your child's particular immunodeficiency disorder, or your own, so that you can communicate intelligently with your doctor and nurse, your own family, and others involved with your family. Read, ask questions, learn. Help your child to understand the disorder and to know that he/she is not alone. Participate in family activities your immune deficiency organization offers so that your child can meet other primary immunodeficient children.

Do communicate well. Meaningful communication is :

1. **unhurried, thoughtful discussion** with your doctor and nurse. Do not let yourself feel hurried out of the office before you are satisfied with answers to your questions.

2. meaningful **talking with** and **listening to** your immune deficient child. Know, as best you can, what is going on in his/her head and heart, not just what the IG levels and temperature are and the date of the next infusion.

3. good talks between parents and other family members. Good communication can keep your child and your entire family on a positive path and a good life in spite of illness.

Do develop a good relationship with your doctor. The management of your child's immunodeficiency treatment will be less stressful if you and your child like the doctor, trust the entire office or hospital environment, and communicate well together. Occasionally through the years both parents may want to make an appointment with the doctor simply to sit down together and discuss your child's (or your) progress, treatment options, and future. If you and your doctor communicate well you will have a greater understanding of the condition which leads to better control. Be an active participant in discussions. If you are not satisfied and have an option, change doctors. Every sick person involved in medical care needs an advocate. You, as a parent or partner of a primary immunodeficient person, are that advocate and will need to be the one to ask questions, take notes, be sure you understand, and probe for better service or answers when it is necessary. As one mother said, " I am totally pro-active in my daughter's care because I am her voice".

Do develop a good relationship with your child's school. It is imperative that the school teachers and principal understand your child's immunodeficiency disorder and the treatment.You do not want an insensitive school environment to be an additional stress for your child. School officials must know the threat of illness and restrictions your child has and any special needs he/ she requires. A twelve year old German boy said, "Thank God I have an understanding teacher who is so careful with me". To eliminate the need for frequent notes explaining school absence due to illness, a parent in New Zealand arranged with the school for a laminated card her child shows the teacher each time she reenters school. The card gives her name, that she has primary immune deficiency and is often in need of medical attention. It is signed by the Head Master. This simplifies and expedites the attendance procedure each time.This same parent stresses the importance of allowing your older child to be part of the school advocacy process instead of a parent always being the one to explain things to the school Principal/Head Master or teacher. This allows your child to have some control in these matters. Arrange for a classmate to furnish class notes or other school information for home study. It follows too, that classmates will be interested and helpful if they understand your child's primary immune deficiency needs.

For other suggestions refer to the PiA publication, "Educating a Child with Primary Immunodeficiency: A Guide for Schools"

Do make sure everyone knows that primary immunodeficiency disorder is NOT AIDS.
There should be no confusion about this which could lead to fear and inaccurate information being passed around in the school **Make sure they understand that primary immunodeficiency is not contagious or transferable, and is not caused by the HIV virus**

Do work out a plan for medical emergencies. Arrange in advance for a friend or neighbor to come quickly if you suddenly need to leave your other children to go to the hospital or doctor, or if you need transportation or something else done for you during this stressful time of sickness. Having someone available is comforting and saves valuable time. Having a willing group of friends is even better so that no one person is called upon too often.

Do know the location of a twenty-four hour chemist (pharmacy) so that you won't lose time hunting for one in the middle of the night during an emergency or on a weekend when many may be closed.

Do have important medical information handy. Have on hand your insurance card or whatever forms your particular medical system requires, your doctor's telephone number, your chemist's/pharmacy's telephone number, etc. Keep this information with you at all times so that you don't waste precious time looking for it when it is suddenly needed.

Do keep a medical journal. Take this each time to the doctor's office or hospital to record important information or instructions. Date it, because later on, maybe even years later, you may not remember something that could be significant. Or at the time of illness, when you are emotionally involved - maybe even frightened - you may not recall afterward what the doctor said or what he/she did. Write it down at the time. Later it may be quicker for you to find a piece of information in your journal than for the doctor to look through a sizable case history. The PiA in the UK has a booklet designed for this purpose.

Do keep your own personal journal. "Therapists report that writing in journals during stressful times can help avoid depression and also has physical benefits. Studies at Southern Methodist University in Dallas, Texas in the U.S. suggest that people who write about upsetting events visit their doctors about half as often as those who write only about trivial events".15 One mother gave her son his own journal and special pen for writing or illustrating his thoughts. Another suggestion is to write letters expressing your frustrations, but not mail them. This is a private, safe place to express your anger and frustration.

Do try to alleviate your child's great fear of the needle. Adults also have needle fear and some hospitals offer help from their needle counselor who talks with the patients prior to infusion, helping them with methods of relaxation and giving them the opportunity to talk with others who have infusions. For a young child, it is important that the physician and nurse show compassion and understanding of the fear. A local anaesthesia, lignocaine cream, is sometimes used an hour before infusion. Be assertive about getting this kind of helpful atmosphere. No child should be forceably held down while a needle is inserted, nor be punished for resisting. At some hospitals a Play Specialist teaches young children how to put an IV drip into a doll. Some families prefer Home Care service which comes to your home with the gammaglobulin and gives the infusion. In the UK, 'Action for Sick Children', London, published a book, 'NEEDLES', and it is available at the PiA office (the Primary Immunodeficieny Association),

Do have pets, if possible. One young adult with primary immunodeficiency said, "I remember we always had animals; cats and dogs, and I think they really helped a lot. Dogs can be very empathetic and are used in many hospital settings to cheer people up. The dogs we had over the years definitely brought joy into my life."

Do take precautions while traveling. Plan ahead. Ask your doctor for names and telephone numbers of doctors in the country or city where you are going. Be sure to keep all medications with you - not in luggage that might be lost or temporarily misplaced. Take along extra prescriptions, and call your airline in advance to learn about national and international regulations regarding carrying medications. **Remember - people with primary immunodeficiences must not have vaccinations of live serums.** If your trip will extend into the next normal infusion day, ask your doctor if a larger dosage of gammaglobulin would be appropriate prior to leaving on your journey. Keep your own doctor's telephone number with you in case you need to call while traveling, and have something in writing, preferably by your doctor, stating your child's (or your) diagnosis and treatment. (Actually, this advise pertains to any member of the family who is on medication.)

For more help refer to the IPOPI publication, *"International Listings of Treatment Facilities Travel Guide and National Member Organizations"*.

You Mustn't Quit

You mustn't quit when things go wrong as they sometimes will.

When the road you're trudging seems all uphill,

When the funds are low and debts are high

And you want to smile, but you have to cry.

When care is pressing you down a bit,

Rest, if you must, but never quit.

Life is queer with its twists and turns,

As every one of us sometimes learns.

And many a failure turns about,

When he might have won if he'd stuck it out.

Stick to your task though the pace seems slow,

You may succeed with one more blow.

Success is failure turned inside out,

The silver tint of the clouds of doubt.

And you can never tell how close you are,

It may be near when it seems afar.

So stick to the fight when you're hardest hit,

It's when things seem the worst that you mustn't quit.

From DEAFinitely speaking, reprinted from Louisiana I.D.E.A.S with permission of Immune Deficiency Foundation Louisiana

SECTION 7: WHAT IS IPOPI?

International Patient Organization for Primary Immunodeficiencies

IPOPI is an international organization that was created in order to achieve worldwide improvement in the care and treatment of people with primary immunodeficiencies.

For more information: www.ipopi.org

SECTION 8: REFERENCES

1. Sorrow That Lives On, from Kai Tiaki: Nursing New Zealand, April 1995 by Jill Hollingum, RcpN, New Zealand.

2. Reprinted from the magazine, Belonging, by Caremark Therapeutic Services, U.S.A.

3. The Value of Optimism, Part Two, by Julie Down, R.N.,C.,M.S., reprinted from Belonging immune therapy, Caremark Therapeutic Services, U.S.A.

4. Louisiana I.D.E.A.S , Fall 1995, Immune Deficiency Foundation Louisiana Chapter, U.S.A.

5. Lousiana I.D.E.A.S , Spring 1996, Immune Deficiency Foundation Louisiana Chapter, U.S.A.

6. Living Creatively With Chronic Illness, by Eugenie G. Wheeler and Joyce Dace- Lombard. U.S.A.

7. How to Live Between Office Visits, by Bernie Siegel, MD, U.S.A.

8. Ministry to Persons With Chronic Illnesses, by John T. Vanderzee, U.S.A.

9. Living with Chronic Illness, Days of Patience and Passion, by Cheri Regester, U.S.A.

10. PATIENT AND FAMILY HANDBOOK, Immune Deficiency Foundation, U.S.A.

11. Living Creatively with Chronic Illness, by Eugenie G. Wheeler and Joyce Dace- Lombard, U.S.A.

12. Living With Chronic Illness, Days of Patience and Passion, by Cheri Regester, U.S.A.

13. Guideposts, October 1995, by Christopher H. Brewer, Bethesda, MD , U.S.A. Reprinted with permission of Immune Deficiency Foundation Louisiana Chapter.

14. When You're Ill or Incapacitated: 12 Things to Remember in Times of Sickness, Injury, or Disability, by James E. Miller. U.S.A.

15. The Primary Source, the Immune Deficiency Foundation Northern California Chapt