

ADULTS AND PRIMARY IMMUNE DEFICIENCY DISEASES



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INTRODUCTION: Although the first primary immunodeficiency diseases were identified in children, there has been a growing awareness that adults, too, may have a number of primary immune deficiency diseases. Because of the advances in medicine, earlier diagnosis and treatment of the childhood immune deficiency diseases has allowed many patients who were born with primary immune deficiencies to grow into adulthood. In other cases, many adults who were born with apparently normal immune systems go on to develop a primary immune deficiency late in adolescence or even in adulthood.

The most common immune deficiencies 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 phagocytic cells (Chronic Granulomatous Disease) and disorders of the complement system. Details of these specific immune deficiency disorders are covered in other chapters in the Handbook. There are several features of immune deficiency diseases of which a newly diagnosed adult should be aware. 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 following chapter reviews the types of problems that adults with primary immune deficiencies develop, discusses how you and your doctor can coordinate care, and outlines some of the psychosocial aspects of living as an adult with these disorders.

COMMON SYMPTOMS

Recurrent infections are the most common problem that patients with immune deficiencies experience. Typically, patients will have recurrent infections in the sinuses (sinusitis) and in the chest (i.e., bronchitis and pneumonia). Early recognition of illness is important to allow rapid treatment before infections become severe. Some early signs may be as obvious as changes in color or consistency of drainage from the nose or changes in sputum coughed up from the chest, or as subtle as easier fatigability or a short-ened temper.

In addition to recurrent respiratory infections, one of the more common symptoms that antibody deficient patients may experience is diarrhea. The diarrhea may be caused by a variety of infections or even by an overgrowth of the "normal bacteria" that live in the gastrointestinal tract. Either of the above events results in decreased absorption of important nutrients required for normal body function. Giardia is one of the more common protozoal intestinal infections that can cause diarrhea. Patients with compromised immune systems are uniquely susceptible to Giardia that can be treated easily with oral medication. Finally, it is not unusual for immunodeficient adults to have chronically red eyes, a condition known as "conjunctivitis." In many patients, if the immunodeficiency can be treated with gamma globulin, the conjunctivitis often improves although

additional antibiotics are sometimes needed.

Some patients also experience arthritis-like symptoms as well as several other symptoms which are seen in patients with so-called "autoimmune" diseases.

These conditions are covered in the chapters in this handbook which deal with specific disorders (see chapters on Selective IgA Deficiency and on Common Variable Immunodeficiency). It is important that patients be familiar with the common symptoms that accompany their diagnosis, so that appropriate care can be sought.

Most doctors who provide care to patients with immune deficiencies know that these patients may require frequent antibiotics, and that the antibiotics often need to be given earlier in the course of an illness than in people with normal immune systems and for longer periods of time.

GENERAL CARE

It is important for any immune deficient patient to understand as much as he or she can about the workings of the immune system, and when medical care is involved that often means staying healthier longer. There are a few things that patients can do in their everyday lives to help maintain good health. In particular; good nutrition is of great importance. A balanced diet is essential for normal growth, development, body repair and maintenance and especially important in preventing and fighting off disease.

The general principles of good hygiene are also very important. Simple things like washing hands before meals and after using the restroom go a long

way to prevent illness and should become routine habits. Many viruses, including the ones responsible for the common cold, are spread by unwashed hands.

Any cuts or scrapes on the skin should be cleansed completely and any unusual redness or drainage should be reported to a physician so further treatment can be initiated promptly. Dental hygiene and regular dental check-ups are important since some patients are more prone to tooth decay and gum diseases.

Regular exercise helps to maintain optimal function of the body and is also a good means of stress relief for the mind. Specific treatment for the immunodeficiency disease should be coordinated between the patient and the health care team members. Each adult should do everything possible to foster good communication between themselves and their health care providers.

Many times patients can give their physicians very important information since they are the ones who have experienced which treatments have truly been of benefit and which have not. Intravenous gamma globulin and other treatments can be administered at home, saving both money and time and giving the patient more control over their health.

BECOMING AN ADULT PATIENT

Growing up with illness often speeds the development of a mature personality. The ability to understand, and become responsible for, complicated medical care can begin at a very early age with parental help and guidance. Including even the youngest of children in the discussions and decisions regarding treat-

ment fosters a positive attitude toward good health rather than anger at "having to take medicine". This sense of responsibility will serve them well in later life once they are on their own. Having young adults leave home is difficult, compounded by the added needs that often accompany any chronic illness. Knowing that a young adult has been able to gradually take care of his/her own medical problems while living at home eases the fears of moving out.

It should be expected that not everything will continue smoothly once the new young adult leaves home. A large part of becoming an adult requires taking your own chances and learning from your own experiences. As mature adults, immunodeficient patients should be cautious that these new experiences do not become life-threatening events.

Feeling "normal" is an important part of adolescence and early adulthood. If a sense of normalcy is established early by the parents, the patient may feel less compelled to fit a lifetime of experience into the first few years away from home.

Accomplishments and compliance to treatment should be recognized and rewarded, while problems should be identified, discussed in a positive way (in regard to prevention of recurrence) and then dropped with no feelings of guilt or punishment remaining. This creates a good line of communication between patients and their parents.

Peer pressure can, on occasion, push someone into unwanted situations. Remember anyone worthy of your friendship will accept your limitations and respect your self-control. Mature adults will quickly realize that the time and attention required to maintain good health will eventually allow for more normal lives and social activities and the continued freedom of living outside the

parental home.

Another integral part of preparing for adult problems includes the construction of a medical diary that includes a summary of your past medical history, a list of current problems and medications, physician's phone numbers, insurance information, and even the appropriate chapters/diagrams/paragraphs from this Handbook. A concise diary is useful when moving to a new area or even on vacation or weekend trips should immediate care be required.

Many adult patients and their friends have commented on the growing number of people who believe that traditional medicine is unnecessary or bad. Frequently they believe that if you "think right" or "eat naturally" you can do without medicine. Most of these people know little about medicine and less about immune deficiency disorders but they are often very persuasive in their offers of easy solutions. Remember that many of these people are motivated by profit and not your good health. Discuss ANY variation from your prescribed treatment with your doctor before giving it a try.

THE NEWLY DIAGNOSED ADULT PATIENT

Some people who have been recently diagnosed have felt unwell for years without an answer to their illnesses and problems. In some cases, a diagnosis can actually be a relief by finally providing an answer. However, at the same time, the newly diagnosed adult patient must face questions and problems that have already been faced by children who have grown up with these disorders.

Feelings of self-pity and fear are quite normal. Sections in this chapter, as well

as other chapters of this Handbook will address some of these feelings. Above all, 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.

SELF EDUCATION

This is the key to caring for one's own health. The more an individual understands about his or her primary immune deficiency disorder; the more comfortable that person will feel. Just as most parents of children with primary immunodeficiency disorders reach out for as much education as possible, the patients themselves, whether they grew up with the disorder or were recently diagnosed as adults, must ask questions, obtain educational materials and understand the realities of their immune deficiency.

Most importantly, patients should read about their disorder and become informed, getting involved with their own care. This can help produce a feeling of independence and control over their own life. One way to begin this process is to seek out health care professionals who specialize in these disorders.

Physicians who have little interaction with patients with primary immunodeficiency diseases may either overestimate the difficulties or underestimate the need for complete evaluation. It is important to ask as many questions as you can of a specialist. No question about your disorder is too trivial. New methods of investigating and treating these illnesses are being developed each year, and it is in the patient's best interest, regardless of age, to find out what these are. Another

way to gather knowledge is through contact with, or attendance at, a local support group. Talking to other patients and families is often helpful, and any feelings of isolation you may be experiencing can be dispelled.

ADVANCED EDUCATION

Since the goal of adults is to be self-sufficient, the importance of receiving an education is hard to overestimate. It is important for people with a chronic illness or medical condition to obtain a job with good health insurance and a position with enough flexibility to allow appropriate medical attention when necessary. The bottom line is that advanced training and education provide a greater range of choices and flexibility for anyone, and particularly for a person with a preexisting health condition.

For the individual who goes away from home to college or to be on his/her own, the preceding section on becoming an adult patient suggested potential problems. One of these is the tendency of many young adults to "downplay their illness" or fail to disclose their medical history to the school. It is hard for a college or school infirmary to care appropriately for a student who has not informed the school of his/her specific diagnosis. An immune deficient individual may need antibiotics more often, or sooner in an illness, than another student.

For a student with a more serious deficiency, school infirmaries may not be an ideal place to receive care. One way to manage this problem is for the parent or student to find out in advance what physician in the area of the school has had experience in treating immunodeficient patients and to transfer medical records before

school starts. Then, if more complicated problems arise, appropriate arrangements can be made. A referral from your current physician may help.

EMPLOYMENT

Adult patients, in choosing a job or career, must think in terms of ones that are suitable for their condition. Depending on the nature of your condition, you may or may not be limited physically. However, there may be complications that need to be fully considered. Factors like time and stress, and how they interact with your condition and treatment, cannot be ignored.

In seeking employment, be aware that there are laws against discriminating against an applicant based on a chronic health condition. However, that does not mean that the laws are easy to enforce. You may want to familiarize yourself with the wording of the laws. For many patients, the health insurance coverage associated with employment is the most problematic. Small employers, for instance, may not be able to cover you. Hence, large corporations and government jobs should be considered in thinking about careers.

HOME CARE

Adult patients must learn to fit their treatment into their school and working lives. For intravenous gamma globulin the use of home health care services permits you to be treated in a home environment. This is particularly useful in avoiding missed time from work. You will want to discuss with your physician whether you are a candidate for home health care, and you will also want to make sure that your insurance will cover the home health care.

In many cases, your physician can provide you with the names of home health care agencies in your area. Many adult patients who need infusions of intravenous gamma globulin can learn to give their own infusions. This can be less expensive and more convenient for the working person. In other cases, a nurse who is employed by a home health care company can deliver all of the home care. Discuss with your physician what options are available to ensure that you receive the treatment prescribed for you.

HEALTH INSURANCE

Health insurance is an issue that all people with a primary immune deficiency disorder must face in decisions they make about school or employment. This cannot be taken lightly by anyone with a preexisting condition. If you allow your insurance to lapse, or do not look into the options that exist before you are no longer covered by your parents' insurance, or change jobs, your ability to afford the treatment you need may be seriously jeopardized. Additional information on health insurance issues can be found later in this Handbook.

DATING, MARRIAGE, AND CHILDREN

Some people may find it difficult to discuss their disorders with their friends, and particularly with girlfriends and boyfriends. It often depends on an individual's own personality as to how much they want to explain, and when they feel comfortable discussing their medical condition. When you do discuss your disorder with a boyfriend or girlfriend, be sure that you make it clear that you have a primary immunodeficiency disease, and that

it is not contagious. Often when a patient becomes seriously involved with another person, it may be helpful to have that person accompany them on a visit to the immunologist to better understand the disorder.

When a couple is considering marriage, it is important for both to understand the genetic implications of the disorder, and whether it could be passed onto children or grandchildren. This is a question about which the immunologist and a genetic counselor can offer guidance. You may wish to refer to the chapter on Inheritance in this Handbook.

It is important for an engaged or married couple to also face the issue of health insurance realistically, and understand its importance in career decisions.

EMOTIONAL STRAINS

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.

For the adult who has recently been diagnosed, there may be feelings of confusion, self pity and, above all, fear. These thoughts and feelings are normal. The positive side of having a diagnosis is that the uncertainty is over and you can be on your way to understanding your illness and the emotional difficulties that may arise from it. It is difficult to have a chronic illness, and to be susceptible to repeated or recurring infections, or other medical ailments. One of the difficulties about primary immunodeficiency disease is the unpredictability of manifestations such as infections. This in itself can place pressures on oneself or family and friends.

In addition, the possibility of unexpected absences from work, last minute changes of one's social activities, or even hospitalizations may cause added strain. 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 airing your fears can have a therapeutic effect.

A problem in families in which the immunodeficiency is genetically determined are feelings of guilt on the part of the parent who has passed the defect on to a child. Again, the best way to deal with these feelings is to discuss them with your spouse, and also your physician and genetic counselor. Remember that this is out of your control, and you also have passed on a number of extremely good qualities.

Children with an immune deficient parent may themselves have a fear of becoming ill when they are older. In most cases the fear is unfounded and can be dispelled with the proper information and testing. 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 a spouse, understanding friend or health care professional.

Many people are helped by meeting with others in a support group setting. For many patients, learning as much as possible about an illness is one very specific way to guard against confusion about the illness itself. As discussed in the section on Self Education, understanding one's own immune deficiency can lead to taking control of one's own life.

SUMMARY

Adults with primary immune deficiency live in the real world. They work, play, marry and have families like other people.

There is no reason why their immune deficiency should alter this. However, they must be aware of the condition and use common sense in recognizing symptoms and treating infections.

These adult patients must make sure that they have access to trained specialists who understand their disorder and are aware of the most recent developments in treatment. They must be more careful than people without a pre-existing condition in obtaining and keeping their

health insurance coverage and in the laws and regulations that govern insurance.

Because of their condition and because of the health insurance risks, some choices of employment may not be optimal. Education and awareness are keys to helping immune deficient adults make life's choices and realize their potential. Always remember that you are not alone and support is only a phone call away.