



Adolescents with Primary Immunodeficiency Diseases

CHAPTER

20

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Introduction

Adolescents diagnosed with a primary immunodeficiency disease, and their families, face not only the day-to-day challenges of any family, but also the challenges of learning how to manage the effects of that disease while nurturing growth towards adulthood. Today's families have busy lives, with each member of the family dealing with demands of time and energy in their home,

work, education, and social lives. With the variety of primary immunodeficiency diseases, there is a wide range of how adolescents may be impacted by these diseases. Differences in the coping styles of family members and variations in maturity through the adolescent years can all influence how well adolescents and their families will deal with a primary immunodeficiency disease.

Having a Balanced Approach

Family life during the adolescent years has many ups and downs, as the adolescent grows physically and develops the maturity needed for establishing social and family relationships, and an educational path toward an occupation. Through these years, both adolescents and family members experience some ongoing tensions between the adolescent's desire for independence and autonomy and the increasing personal responsibilities that go with that desire. They typically go through a series of steps in this maturing process, commonly having both successes and setbacks in navigating into adulthood.

Each of these ups and downs can be shaped by the adolescent's primary immunodeficiency disease, as it can impact overall health, friendships, family, and career plans. For some, the primary immunodeficiency disease will only minimally affect their personal growth. For others, the disease will substantially affect them in two quite distinctive ways. On one hand, the disease and related health problems can directly challenge adolescent desires for independence and autonomy, as they have to increase their dependence on family members and healthcare providers to meet significant health challenges. At the same time, these health challenges can provide significant opportunities for developing coping skills and maturity beyond their years. Families, then, need to develop flexibility and the support needed to help their adolescents through these challenges.

Families experiencing ongoing health challenges will, at times, tend to struggle with finding a balanced approach to maintaining their family life while addressing these health issues. A lengthy infection, hospitalization, or major change in treatment can be quite disruptive to their lives. Time and energy become so focused on responding to the illness and treatments that other aspects of life become neglected, such as school, work, leisure activity, and social relationships.

While this kind of attention may be necessary at the time of a health crisis, over time, it can lead to isolation from activities and relationships that help the adolescent and family cope in the long run.

There will also be times when the primary immunodeficiency disease and the related health issues may fade into the background for an extended period. A treatment routine becomes successful, symptoms and health complications are absent, and the family experiences an extended period of routine life. Time and energy become invested in pursuing new leisure interests, planning for the future with new interest in school and career, and cultivating relationships. It is at this point that there may be some tendency to ignore subtle health symptoms, neglect basic preventative health care, and be less consistent in maintaining a relationship with healthcare professionals. It is understandable that adolescents would want a break from focusing on the disease, yet ongoing neglect of symptoms or treatment routines can lead to a serious health setback.

Adolescents who best manage their primary immunodeficiency disease are those who find a balanced approach to the disease and to life. An emphasis should be placed on both the disease compromising overall health (the signs, symptoms, and treatments of the primary immunodeficiency disease) and overall health itself (the activities and relationships that promote a healthy lifestyle). However, enough attention must also be placed on addressing the primary immunodeficiency disease without the disease absorbing and defining the adolescent and family life. The family should help adolescents learn the coping skills needed to manage day-to-day issues of the primary immunodeficiency. This gives adolescents a greater sense of control, and helps them develop an identity based on personal strengths and healthy choices rather than on symptoms of disease.

Your Family

Adolescents and their families who cope best with an ongoing health problem typically follow several guidelines during the maturing process. In young adolescents, parents are often more active in taking the lead in learning and setting an example. Later, parents encourage increasing involvement of the adolescent in management of their disease, with parents monitoring the adolescent's increasing responsibility of self care. Finally, as the adolescent moves toward adulthood, parents encourage the adolescent to take main responsibility for managing the disease, with family members as more distant supporters.

Ideas for Successful Family Coping

Learn all you can about primary immunodeficiency disease and help cultivate this learning and curiosity in your adolescent.

Use trusted written and online resources, and resources suggested by your adolescent's physician and healthcare team. Bring your information and questions to physician visits to learn how to best apply the information.

Encourage your adolescent to seek information about successful ways of dealing with their primary immunodeficiency disease.

This learning might be incorporated into their science or health classes. Encourage your adolescent to become an active consumer of health information and have them practice asking questions of their healthcare providers. This will prepare your adolescent to stay informed when he or she leaves home for college or a career.

Notice all you can about your adolescent's personal body pattern and help cultivate personal awareness and communication skills in your adolescent.

What are the personal signs of a new infection, or successful response to medication and treatment? When energy is low, what are the personal signals of the body that help your adolescent know if it is time to push through the weariness to build their strength, or a time to take a needed rest to refresh and heal? This gives adolescents a greater sense of being in charge and helps make the shift in managing their health from parent awareness and reminding to adolescent awareness and self-reminding. Vacations with extended family, stays at summer camp, and trips with school

or youth organizations give the opportunity for the adolescent to begin to practice greater responsibility in being aware of their body's needs. Use these times to prepare your adolescents for their future when they will live more independently, pursuing college or career goals.

In conversation with your adolescent and healthcare providers, develop a personalized list of successful approaches to managing your adolescent's health.

What health and wellness habits have been most successful in keeping your adolescent happy? What routines for diet, rest, and leisure have been the most refreshing? What activities have promoted the most success with physical fitness? What medications and treatments have been most reliable in managing the symptoms of the adolescent's disease? Having a personalized understanding of your adolescent's primary immunodeficiency, medications and treatment, and strategies for health and wellness will help encourage good habits.

Parents who model good health and wellness habits in their own lives will provide a positive example for their adolescent to follow. Along with modeling, make sure that your adolescent has a full understanding of specific health concerns and treatments, and how preventative care and an emphasis on wellness can help. Reinforce their efforts in taking responsibility for their health, and emphasize how this is important sign of maturity. With opportunity for responsibility and encouragement, your adolescent can develop lifetime habits of positive coping skills for their health challenges.

Create and maintain supportive relationships for your adolescent with other family members, friends, and members of the community in school and such organizations such as scouts, music groups, sports teams, and spiritually based groups.

Sometimes, having a primary immunodeficiency can make an individual feel very different than other people and isolated from their peers. Depending on the particular primary immunodeficiency, and its impact, some adolescents may do well playing on a sports team with very high physical demands, stringent practice requirements, and lengthy traveling games. Others may do better with a sport with varying physical demands, some flexibility with participation, and leaders who are responsive

Your Family continued

to the skills and needs of each person on the team. With younger adolescents, parents may play a greater role in helping them discover these activities and interests, and negotiate their participation. As adolescents grow older, parents can put more emphasis on them exploring their interests, making contact with others, and developing their place in groups and on teams.

Feeling like they belong and are successful may be very challenging when an adolescent has a primary immunodeficiency disease. Not everyone will be understanding or helpful in supporting your adolescent in finding his or her strengths and social supports. Be prepared to help them deal with the ups and downs of their successes and disappointments. While most relationships in adolescents may not carry through a lifetime, the skills and experiences in relationships during adolescence will shape how that adolescent relates to others as an adult.

Share your experience of having a family member with a primary immunodeficiency disease with others—at the right time and with the right people.

This may be limited to immediate family and friends, or it may include sharing with other families in the same situation through a support network. For some adolescents, it may include broader and more public sharing, such as giving a primary immunodeficiency disease presentation in a health class, doing a science fair or project in competition. Sharing can break the social isolation, improve supportive relationships, and give the adolescent a way to show their strengths and successes in dealing with a condition that can be very challenging. However, not everyone in your adolescent's life will have a positive reaction. As with almost any kind of difference that can be noticed between people, the differences caused by a primary immunodeficiency disease can sometimes make the adolescent a target for teasing and isolation. Guide your adolescent in regards to the appropriate people, times and places to relay personal situations.

Your Adolescent

The adolescent years have many dramatic changes: bodies that grow from child to adult, responsibilities that shift from the role of the parents to the role of maturing adolescent, childhood friendships that transform into young adult relationships and schoolwork that takes on new meaning as the adolescent moves towards college and career. Each of these changes can be impacted by the adolescent's primary immunodeficiency disease. Parents, healthcare providers, and other concerned adults in the adolescent's life need to help cultivate open and supportive communication about these issues. These adolescents may still have a lot to learn about growing up, but they also deserve to be respectfully heard as they share their thoughts and feelings that come from the wealth of their experience dealing with day-to-day issues. One of the best ways to show this respect is to lead off any discussion by asking about their feelings, views, and experiences first. This approach helps to establish a respectful discussion in both directions, and there will be times when you learn

that the adolescent's viewpoint and concerns, while said a little differently, may be very close to the concerns that you, as a concerned adult, may be having.

Here are some common questions that may be helpful in starting a conversation with your adolescent:

So, am I sick or am I well?

Have a discussion about the balance needed to cope with a primary immunodeficiency disease. Your adolescent's healthcare providers may be helpful in advising on how to focus energies on both managing the disease and living life more fully. Help your adolescent identify enjoyable interests and activities that may be less impacted by days when the adolescent is not feeling well. Music, arts, crafts, and other creative activities can be enjoyed alone and with groups. These creative outlets also can provide needed distraction and relaxation when an adolescent is experiencing health challenges.

Your Adolescent continued

I hate being treated differently! Why can't I be just like everybody else?

Adolescents will vary in how much they wish to express their uniqueness or blend in with the crowd. Helping your adolescent find their own unique qualities and talents will help build confidence and reduce the likelihood that they see their uniqueness mainly as being the person in their group that has an uncommon disease.

What do I tell my friends about primary immunodeficiency disease?

This may be related to the question about being treated differently. It also involves learning relationship skills of trust building and sharing. Your adolescent can benefit from a trusted peer who can understand and offer personal support when a primary immunodeficiency can disrupt a regular routine. Your adolescent can also be hurt by less mature peers who use personal information as a way to bully or tease. Help your adolescent make wise choices in their friendships and personal sharing.

How do I handle this at school?

When your adolescent asks this, it might be more about the friendship aspect of school. Your adolescent may also be asking about how to deal with teachers, coaches, assignments, and team requirements. While a long-term goal is self-responsibility, some school issues may require parents to be more active in helping establish positive relationships with school personnel and in establishing realistic expectations for balancing health and school performance.

Why do I have to go see my physician/take my medications/continue my treatments?

As adolescents learn new levels of responsibility, there will be times that they will want to do things differently. Begin by hearing out the adolescent's concerns and responding with the details of treatment decisions previously made, realizing that some of these decisions and routines may have been originally made when the adolescent was much younger and not involved in the information gathering or decision making process. Some of the questions about care may relate to a healthy

need to have a greater sense of control over their life. This may be a good time to review the adolescent's current responsibilities throughout their life, not only with their healthcare, but also with their home responsibilities, schoolwork, and leisure activities. Having a greater sense of control in other areas often helps balance the sense of lacking control that can come with some of the symptoms of a primary immunodeficiency disease.

Am I going to be dealing with this disease forever?

Younger adolescents may ask this when they realize that their primary immunodeficiency will not be like other health problems they may have experienced, like a sprained ankle or broken bone, which has healed and is now forgotten. This may be about that balance of addressing the illness and health aspects of their disease, and realizing how health and wellness habits will help them. Older adolescents may ask this when they are thinking about their future—career plans, college plans, or developing relationships. Discuss how they can apply their earlier learning experience to these new challenges of young adulthood, and suggest talking with their physician or other healthcare professionals.

Why do I have to have this disease? It's not fair!

This is a very tough question. It is one often asked by parents as well as their adolescents. This may be a question about their particular disease and how the immune system works. Often, though, this question is looking beyond scientific answers and looking more toward personal beliefs and values about life. Families need to reach out to the people and resources they have for finding meaning in life.

Your Adolescent's Healthcare Providers

Some adolescents may experience few medical problems that are related to their primary immunodeficiency disease while others may have very complex medical concerns that involve a number of physicians and other healthcare providers. Maintaining good communication with, and among, these providers is important to effectively manage a primary immunodeficiency disease. Clear and accurate information about both the history and current status of your adolescent's health condition is needed by all who will be providing care. Healthcare providers need to know exactly what your adolescent is experiencing and how each of the providers is contributing to your adolescent's care.

To maintain clear communication and good teamwork with healthcare providers, keep a diary or notebook outlining your adolescent's symptoms and treatments. Prior to a visit or phone call with your healthcare provider, use these notes to summarize your adolescent's current condition and plan any specific questions that you may have. Ask all healthcare providers to provide you with copies of all major treatment summaries, laboratory and diagnostic test results, and correspondence. When requested by healthcare providers, help make sure that they are also copied with reports from other providers. Keep these organized in your adolescent's healthcare

notebook as a reference for new providers. Including adolescents in helping with this communication process will help them be ready for their adult role of managing their own healthcare.

Transitioning from late adolescence into young adulthood may involve changing healthcare providers. This may occur as they move away from home to attend college or begin a career. For others, the transition occurs as older adolescents are shifted from pediatric care providers to adult care providers. Discuss this with current physician and healthcare providers for the best way to manage these care transitions.

There may be times that are particularly stressful in family life. This may be from significant changes in family life, such as a change in parent employment, financial changes, a move, divorce or death. Or, the stress may be more directly related to coping with a primary immunodeficiency disease. Whatever the source of these high levels of stress, it can impact any healthcare problem, so do not hesitate to address the sources of family stress. Your adolescent's physician should be kept updated on these matters, so that general guidance can be offered or a referral can be made to a family counseling professional for more specific help.

Your Adolescent's School

Some adolescents with a primary immunodeficiency disease will not experience any difficulties at school due to their particular condition. Other adolescents, with more substantial health concerns will need significant assistance in the educational setting. For adolescents with a number of concerns, you will be wise to make advance contact with school personnel to discuss your adolescent's healthcare condition and how it may impact school. School personnel should be included in advance of problems with adequate information. They can often develop plans that will work to address concerns of reducing exposure to infection, coordinating schoolwork during absences, and providing certain modifications of schoolwork when needed.

Federal and state educational regulations include requirements and guidelines for how schools should respond to a student's health and its impact on their learning. Parents, school personnel, and the adolescent's physicians and other healthcare providers can coordinate specific plans through mandated programs such as the Section 504 Plan or Individualized Education

Plan (IEP). Additional information about these regulations and guidelines for modifying your adolescent's educational setting are available through your local school district, Web sites of your state education department, student advocacy Web sites and in the Immune Deficiency Foundation's publication, *A Guide for School Personnel*.

Include your adolescent as much as possible in these planning meetings and discussions with school personnel. Just as adolescents need to develop skills in managing healthcare decisions, they need to develop skills in managing education decisions that lay the foundation for their future with college or career plans. For your older adolescent with college plans, be aware that there are also federal guidelines for higher education institutions. Many student service departments of colleges include resources for health, career, counseling, and other campus-based services that can help. When your adolescent begins to explore college choices, it may be useful to include visits with these services when touring campus or examining literature or Web sites.

Adolescent Insurance Concerns

Most young adults in this country are covered under a group insurance plan offered by one of their parent's employers. Many of these employer sponsored plans end dependent coverage when the dependent child turns 19, if not attending college, or to age 23 or 25, if enrolled in school full-time. When dependent children are no longer eligible for group insurance under a parent's plan, they are entitled for up to 36 months of COBRA coverage (if the employer has 20 or more employees). Some plans continue to cover totally disabled dependents as "adult disabled children" beyond the usual end date for dependent children. To know how dependents are covered under the employer sponsored health plan, the parents should ask their human resources department for

a copy of the Summary Plan Description (SPD). A SPD is a requirement of the law (ERISA) that sets the terms of each employee benefit plan in a written plan document.

Medicaid, usually available through the state's Children's Health Insurance Plan (CHIP), is another form of insurance that usually ends when the dependent child turns 19. Medicaid for disabled Supplemental Security Income (SSI) can end if the young adult is no longer designated as disabled or if income or assets exceed the allowable limits. Some states offer incentive programs to Disabled Medicaid individuals who work and make over a certain amount of money.

Adolescent Insurance Concerns continued

If an adolescent's insurance is about to end, it is important to understand what is available well in advance. Here are some important questions to research to provide a better picture of what insurance options are available:

- How long am I entitled to insurance under my parent's plan?
- Will COBRA be available to me after I graduate? If yes, how will the premiums be paid?
- Will my Medicaid end when I turn 19?
- If I get a job, how long is the waiting period before benefits begin?
- Does my state have a high risk pool? What are the premiums?
- Does my state Medicaid plan offer incentives to work despite my disability?

Coordinated Support

As you, your family members, and your adolescent with a primary immunodeficiency disease handle the challenges that may accompany that disease, remember to keep your support system close at hand. Particularly stressful times, when support is most needed, may also be the times when it is the most difficult to maintain those connections with family, friends, and the professionals that are in your support network. Stay current through your adolescent's healthcare providers, your reading, and networking. Keep your circle of support informed and involved. Show your adolescent how to both be realistic about the challenges as well as encouraged by the choices in life.