

Adolescents Living with Primary Immunodeficiency Diseases



Chapter 30

Adolescents diagnosed with a primary immunodeficiency disease and their families face not only the day-to-day challenges of any family, but they also face learning how to manage the effects of a rare and chronic disease while nurturing growth towards adulthood. Adolescence is a time of great transition cognitively, developmentally and emotionally. When adolescents are diagnosed, they face some unique difficulties, and it is important to help them manage the impact of a primary immunodeficiency disease while striving to achieve the developmental tasks of a teenager.

Although typical adolescent trials may be more stressful and confusing for those living with primary immunodeficiency disease, you and your adolescent can work together to overcome challenges, enjoy this time and prepare for the transition into adulthood.

Normalizing Your Adolescent's Life

As adolescents grow, they develop the maturity needed to establish and maintain family and social relationships as well as continuing an educational path toward an occupation. They typically go through a series of steps in this maturing process, commonly having both successes and setbacks in navigating toward adulthood. School and social time with friends are often the focus as adolescents begin to explore their independence and separate from prior parental attachments. This can be difficult for both parents and adolescents. A balance must be achieved between maintaining an optimum level of health and being able to actively participate in desired activities. In addition, primary immunodeficiency diseases manifest differently in each patient; therefore families must make choices that best suit the adolescent's physical and mental health, as well as abilities.

You can help your adolescent through this time by teaching them coping skills needed to manage day-to-day issues associated with the primary immunodeficiency

disease while helping them live the normal life of a teenager.

Begin a dialogue with your adolescent, so that they can become a part of the decision making that impacts their life. Lead off any discussion by asking about their feelings, views, and experiences. This approach helps to establish a respectful discussion in both directions, and there will be times when you learn that their viewpoint and concerns may be very close to the concerns that you may have.

Help your adolescent maintain a balanced life. Teens who best manage their disease are those who find a balanced approach to the disease and to life. It is understandable that adolescents would often want a break from focusing on the disease, yet neglect of symptoms or treatment routines can lead to a serious health setback. An emphasis should be placed on both managing the disease (the signs, symptoms and treatments) and maintaining overall

(Normalizing Your Adolescent's Life continued)

health itself (the activities and relationships that promote a healthy lifestyle). Contact the Immune Deficiency Foundation (IDF) about getting your child involved with the IDF Teen Program to help them learn about living with the disease and to connect with others living with primary immunodeficiency: www.primaryimmune.org or 800-296-4433.

Coordinate with your adolescent's school. Living with a primary immunodeficiency disease may disrupt schooling, and as previously mentioned, school is an integral part of an adolescent's life. You should meet with teachers, counselors, nurses and administration to explain your teen's primary immunodeficiency disease and the potential impact on school, such as frequent absences, fatigue and illness. Work with them to develop a plan to help your adolescent keep up with schoolwork when they cannot attend school. A good resource to help you coordinate with their school is the *IDF School Guide*, which can be ordered or downloaded at www.primaryimmune.org.

Encourage your adolescent to explore their talents and interests, and help them make appropriate modifications when necessary. Set realistic expectations based upon their individual capabilities and medical needs, but focus on all that they can do. Encourage participation in athletics, music, dance and whatever peaks their interest. Having fun outside of family, school and medical appointments will build confidence and help them cope with periods of illness.

Allow your adolescent to participate in school and social activities to help them feel that they are living a valuable life with purpose and enjoyment. Remember that school and social events are central to teenagers, and missing out because of a primary immunodeficiency disease can be very difficult for them. Acknowledge such disappointments while balancing their health. Your adolescent should participate in events whenever possible. There is always a chance that patients can become ill since germs exist everywhere, but preventing your teen

from participating in group events can create feelings of anxiety and depression. Making simple modifications, such as using hand sanitizer, avoid sharing beverages, and staying away from actively coughing individuals, can allow them to participate.

There are certain restrictions that some patients with primary immunodeficiency diseases must follow. For example, those with thrombocytopenia should avoid contact sports. Football or soccer is risky for patients with Chronic Granulomatous Disease (CGD) because of exposure to dust or grass with potential pneumonia. These should be explored with your healthcare provider.

Help your adolescent develop strategies to educate peers and to explain their condition, including the appropriate terms for diagnosis and treatment. Teenagers already struggle with identity issues and confidence, and feeling different can further complicate this matter. Because primary immunodeficiency diseases are rare, your adolescent may not know many other children with the same illness. They must develop strategies to cope with questions and misconceptions they may come across. If they are able to clarify peer questioning and talk about their experiences, peers will be less likely to gossip about the condition.

Start conversations about dating not only to encourage positive decisions but also to help you get to know your teenagers as they mature. Dating is a hallmark of adolescence, and talking to your adolescent about how to talk about their primary immunodeficiency disease and safety concerns is important. Although rejection is a normal part of dating and people reject others for all sorts of reasons, reiterate that if someone rejects them because of primary immunodeficiency disease, that person is not the right person for them. It is important to let your adolescent know their value and that people with primary immunodeficiency diseases date, marry, have children and lead full lives.

Family Life with Adolescents Living with Primary Immunodeficiency Diseases

Adolescents and their families who best cope with an ongoing health problem typically follow a pattern during the maturing process. In early adolescence, parents are more involved in directing learning and serving as role models. Later, parents encourage increased involvement of the adolescent in management of their disease, with parents monitoring their increasing responsibility of self-care. Finally, as the adolescent moves toward adulthood, parents encourage them to take main responsibility for managing the disease, with family members as more distant supporters.

Families may struggle to find a balanced approach to maintaining their family life and addressing the health issues of their adolescent. Time, activities and family decisions may require daily modifications. In addition adolescents with primary immunodeficiency diseases often feel guilty that they are burdening their parents with the additional pressures of their illness. Be sure to let your adolescent know all the wonderful things that they add to your life to let them know how valued they are.

Increased attention to medical care and modifications to the family routine may cause strain amongst siblings. Siblings can feel jealous of the attention given to their brother or sister with a primary immunodeficiency disease. Acknowledging the impact of the disease upon the non-patient sibling is important. Praising the sibling for their patience and acknowledging the challenges of having a sibling with a chronic illness can help decrease resentment and validate the sibling's experience. Encourage sibling input on family decision-making, so that the non-patient sibling feels that their needs are just as important as the patient's.

When two children in the family have primary immunodeficiency diseases, there can be a greater connection and shared understanding gleaned from the common experience. There may be less resentment between siblings than if one child did not have the disease. However, the emotional impact can vary based on individual personalities, coping skills and different degrees or manifestations of the disease. It is important to individualize each child's needs and not to generalize their experiences.

Preparing for Tough Questions from Adolescents Living with Primary Immunodeficiency Diseases

Common questions that you may hear from your adolescent:

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 how much they want to blend in with the crowd. Helping them find their own distinctive qualities and talents will help build confidence.

What do I tell my friends about primary immunodeficiency diseases?

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. Conversely, they can be hurt by less mature peers who use personal information as a way to bully or tease. Help them make wise choices in their friendships and personal

(Preparing for Tough Questions continued)

sharing. Encourage them to take advantage of the IDF Teen Program and IDF Common Ground to learn more about their disease and to connect with other teens.

How do I handle this at school?

When your adolescent asks this, it might be more about the friendship aspect of school. They 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 you to help establish positive relationships with school personnel and realistic expectations for balancing health and school performance. Consult the *IDF School Guide*, which can be ordered or downloaded at www.primaryimmune.org.

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

As adolescents acquire new levels of responsibility, there will be times that they will want to do things differently. Begin by listening to their concerns. It is possible that a treatment or management regimen made when the child was younger can be changed or modified to meet the needs of the older child. 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 their current responsibilities throughout their life, not only with healthcare but also with home responsibilities, schoolwork and recreational activities. Having a greater sense of control in other areas often helps balance the sense of lacking control that can come with living with a primary immunodeficiency disease. Schedule an appointment with your adolescent's immunologist to discuss why taking medications and continuing treatment is important.

Am I going to be dealing with this disease forever?

Younger adolescents may ask this when they realize that primary immunodeficiency disease will not be like other health problems they 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 earlier learning experiences to these new challenges, and suggest talking with their healthcare providers.

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 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. Assure your adolescent that this is not their "fault." Reach out to the people and resources that you currently seek out to find meaning in life.

Resources and Professional Assistance

Many adolescents with primary immunodeficiency diseases often feel misunderstood. As much as family and friends may attempt to understand the impact of these diseases, only patients can really understand what this disease feels like. Encourage your adolescent to connect with peers with primary immunodeficiency diseases through the IDF Teen Program. Having other adolescents to relate to is invaluable in providing support. Not only can it foster supportive friendships, but their involvement with others with primary immunodeficiency diseases can also help them feel that they are neither alone nor different from others. IDF Common Ground is a social networking site designed specifically for adolescents, where they can connect with other young people who understand what it is like to live with a primary immunodeficiency disease: www.idfcommonground.org. IDF Teen Escape weekends, which are held a few times each year in various cities throughout the country, are designed to help teens develop coping skills, promote and nurture friendships and provide educational guidance for those living with primary immunodeficiency disease. IDF holds

biennial National Conferences and Retreats, during which there are programs for teens. For more information about programs and resources for teens, contact IDF: www.primaryimmune.org or 800-296-4433. Teens may not feel they need to attend such programs, but those who do attend have extremely positive experiences.

Seek professional assistance if your adolescent displays symptoms of depression or anxiety. They can feel isolated as well as overwhelmed by the impact of feeling ill, limitations in productivity and awareness of financial burdens. It is important for you to recognize the signs and symptoms of depression and contact a mental health professional. Symptoms of depression include:

- Shifts in overall mood and outlook
- Changes in eating and sleeping patterns
- Negative self-talk
- Increased isolation
- Irritability, anger
- Hopelessness, tearfulness

Coping for Parents with Adolescents Living with Primary Immunodeficiency Diseases

Parents of children with chronic illness not only worry about their children's physical and emotional care, but they also carry the extra burden of managing financial and insurance issues. This can be incredibly stressful. Of course children require support, but parents also need to receive some extra, outside support. Having a place to vent about how difficult this challenge can be is important and receiving support from friends, family and partners is necessary. But just as it is imperative that your adolescent is connected to the primary immunodeficiency disease community, you should be

too. Having the connection to another parent who understands your experience can be emotionally healing. Parents of patients may understand your experience more than friends and family who may find it difficult to relate to the daily pressures of primary immunodeficiency diseases. IDF offers a variety of resources and programs to connect you with other parents. Taking care of your emotional well-being can make it easier for you to manage caring for your adolescent.

Coordinating Your Adolescent's Healthcare

Managing the healthcare of your adolescent calls for a high level of communication and teamwork among healthcare providers, family members and the adolescent. Knowing what local resources are available and creating positive relationships with healthcare providers, teachers, and others involved with your teen increases the chance that your child's healthcare needs can be effectively met.

Adolescents are more likely to make positive choices in the future if they feel they have some say in the decision-making process now. Find providers who are willing to work with you and your adolescent, and allow for private time between the child and providers. They may want to share concerns about issues that they do not want to share with you. Some level of privacy is appropriate and necessary. Keep in mind that they will be more likely to share information and therefore receive better care if they feel listened to and develop a good relationship with healthcare providers.

Planning appointments around your adolescent's school and social activities allows them to maintain social relationships, which is key to emotional well-being. Try to schedule their doctors' appointments, infusions and/or blood draws on the same day or a day and time that does not interfere with school or social activities.

In conversations with your adolescent and healthcare providers, develop a personalized list of successful approaches to managing their 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 their disease?

Having a personalized understanding of your teens's primary immunodeficiency disease, medications and treatment, and strategies for health and wellness will help encourage good habits. In addition, parents who model good health and wellness habits in their own lives will provide positive examples for their adolescents to follow. Along with modeling, make sure that they have a full understanding of specific health concerns and treatments, and how preventative care and an emphasis on wellness can help. Reinforce and praise efforts to take responsibility for their health, and emphasize how this is an important sign of maturity. With appropriate support, your adolescent can develop lifetime habits of positive coping skills for health challenges.

Being Your Adolescent's Healthcare Advocate

As your child's patient advocate, you work to make sure the needs of your teen are being met by the healthcare team. With younger children, the parent is the chief patient advocate. As a caregiver, the parent is in the position to tell the healthcare providers what happens every day and supply the healthcare provider with critical information. As the child matures during adolescence, they must begin to learn to advocate for themselves. To be effective advocates, it is important that you both learn as much as possible about the disease, treatment options and available resources, and that you build positive relationships with healthcare providers.

Recommended information to record and keep readily available in a journal or in IDF ePHR (www.idfephr.org):

- Brief history leading to the diagnosis, written by you or a healthcare provider
- Copies of laboratory evaluations confirming and supporting the diagnosis
- Current list of healthcare providers caring for your adolescent with accurate addresses and phone numbers
- Chronology of important events such as infections and surgeries, specifically noting types of treatment and therapy, changes in therapy and subsequent responses to the treatment, therapy, infection, surgeries and/or hospitalizations
- List of your adolescent's current medications
- Allergies to medications
- Infusion log for those receiving immunoglobulin (Ig) replacement therapy
- Immunization record
- Current insurance information
- Explanation of benefits records can be kept in the journal or separately but should be periodically reviewed for accuracy

How you can advocate for your adolescent:

- Ask questions about the diagnosis, treatment and plan. If you do not understand, ask again.
- Inquire about what can be done to improve your adolescent's health such as diet, physical activity, sleep and social activities.
- Maintain consistent communication with the school.
- Know your insurance policy and communicate to your provider if there are any changes.
- If your adolescent receives Ig therapy, make note of how it is going and/or any side effects.
- Build positive relationships with your adolescent's providers, teachers and therapists. Know who to call and when.
- Ask about and seek out resources for further information at the local, state and national level.
- Connect with IDF for additional resources: www.primaryimmune.org or 800-296-4433.

Plan for Life After High School

Having this disease should not impede your adolescent from pursuing post-secondary education and/or living independently, but it will influence the decision in terms of obtaining healthcare and living conditions. Some patients may choose to live at home and attend a local college or university. Others choose to attend school and/or live on their own, sometimes far from home. Consider what is best for your adolescent. Contact student support services at their colleges of choice to discuss the child's diagnosis and possible resources and accommodations. If your adolescent secures a part-time or full-time job, make sure it suits their interests and abilities.

When researching new providers and facilities, you need to understand your adolescent's insurance benefits and what providers and facilities are covered. Be sure to choose healthcare providers who best suit your adolescent's needs. The location of the potential healthcare providers may influence decisions about where to live or attend school. If they plan to relocate after high school or attend college away from home, you and your adolescent should research immunologists in that area. Your current providers may be helpful in making recommendations. Many immunologists are associated with universities that can support their research, so you could consider those universities and cities. If they move out of state, it is also important to remember that hospitals and clinics may not be able to accept the orders from your current healthcare provider. You and your adolescent

should establish care, if possible, before moving day to give the healthcare provider time to get to know your adolescent and time to request authorization for treatment from the insurance plan.

If your adolescent receives Ig therapy and is relocating or going away to college, it may not always be necessary to change infusion providers. If they are receiving infusions through a specialty pharmacy in a homecare setting, they may be able to continue with the same provider. Be sure to check with their current infusion provider several months before moving. Additionally, if a change in providers is required, their current provider should participate in coordinating the care and transition to the new provider. Also, inquire about their policy for administering intravenous medications in university housing.

If your adolescent receives infusions in a clinic or outpatient hospital setting, it will be important to coordinate the care in advance with as much notice as possible. Additionally, the receiving clinic will likely need to get a new insurance authorization to provide care. Failure to obtain a new authorization could result in denied claims.

Transitioning Responsibility to Your Adolescent

During childhood and adolescence, parents have the responsibility of making all healthcare decisions. Once an adolescent turns 18, they are legally considered an adult. At that point, parents need the patient's written permission to access healthcare records and to speak with the healthcare team. To prepare for this transition, adolescents should begin actively participating in their care early on and understand their diagnosis, treatment and insurance. When appropriate, allow them to make choices that ultimately meet the determined goal. This helps your adolescent build confidence because they have some control over decision-making and prepares them to participate and ultimately take care of themselves in adulthood.

Planning the transition of care should begin in early adolescence. The primary care providers and immunologist should be involved to insure a smooth transition, and the plan for independence should be tried and tested long before the adolescent turns 18 or lives away from home. Testing the transition plan would involve having your adolescent become independent with current healthcare providers.

On the following pages are checklists to be completed by your adolescent to help them prepare for the transition to adulthood.

Transition Skills Checklist

Ages 12-14

General Information	Yes	Almost	No
I can tell someone the name of my primary immunodeficiency.			
I can describe the effect of primary immunodeficiency disease on my body.			
I can share my medical history with a doctor or nurse.			
I can list my medication and food allergies.			
I tell my parents about changes in my health.			
My parents keep a personal health record for me, such as IDF ePHR.			
My parents and I carry a medical summary, such as the in case of emergency (ICE) report from IDF ePHR.			
Medications and Treatment			
I can list the proper names of my medications, the dosage and times they should be taken.			
I can explain why each medication is necessary, the result of not taking it as prescribed and its side effects.			
I take all medications as prescribed and notify a parent when the supply is low.			
I use and take care of medical equipment/supplies and notify a parent if there is a problem or supplies are low.			
I can list medical tests that need to be completed regularly.			
Medical Appointments			
I tell my doctor or nurse about how I am feeling.			
I answer at least one question during a medical appointment.			
I ask at least one question during a medical appointment.			
I spend some time alone with the healthcare provider during a medical appointment.			
I talk with my parents and healthcare providers about the medications and treatments I need.			
I tell the healthcare provider I understand and agree with the medication or treatment prescribed.			
Understanding the Healthcare System			
I know the date and reason for my next medical appointment.			
I know the names of my healthcare providers and how to contact them.			
I know the name of my health insurance and the importance of being insured.			
Healthcare Transition			
I am taking more responsibility for my healthcare.			
I have talked to my parents and healthcare providers about whether I will need to see new providers when I'm an adult.			
I have talked to other teens about their healthcare transition experience.			

Transition Skills Checklist

Ages 15-17 (Transition Skills to be added to the 12–14 Checklist)

General Information	Yes	Almost	No
My parents and I keep a personal health record, such as IDF ePHR.			
I carry a medical summary, such as the ICE report from IDF ePHR.			
Medications and Treatment			
I can explain why each medication is necessary, the result of not taking it as prescribed, its side effects and the management of side effects.			
I can select medication for a minor illness, such as a headache.			
I can refill a prescription.			
I can list medical tests that need to be completed regularly and make sure they are scheduled.			
Medical Appointments			
I answer many questions during a medical appointment.			
I ask many questions during a medical appointment.			
I spend most of the time alone with the healthcare provider during a medical appointment.			
I decide with my parents and healthcare providers about the medications and treatments I need.			
I can contact the appropriate healthcare providers to tell them about changes in my health.			
Understanding the Healthcare System			
I can explain the difference between a specialist and primary care physician.			
I can explain legal rights and responsibilities available to me when I am 18.			
I can explain how my health insurance works (provider network, deductible, co-pays).			
Healthcare Transition			
I know if any of my healthcare providers will only treat me until I am 21.			
I have talked to my parents and healthcare providers about things I should think about if I need to see new providers when I'm an adult.			
I have identified some healthcare providers that will care for me when I'm an adult.			
I have talked to other teens and young adults about their healthcare transition experience.			

Transition Skills Checklist

Ages 18 and Up

General Information	Yes	Almost	No
I keep a personal health record, such as IDF ePHR.			
I carry a medical summary, such as the ICE report from IDF ePHR.			
Medications and Treatment			
I understand and/or arrange payment for my medications, equipment and treatments.			
Medical Appointments			
I check myself in at appointments and provide my insurance card.			
I answer all questions during a medical appointment.			
I ask the questions during a medical appointment.			
I am alone or choose who attends a medical appointment with me.			
I decide with the healthcare provider about the medications and treatments I need.			
I locate and share healthcare information with my providers and in making decisions about my care.			
I sign medical consent forms.			
Understanding the Healthcare System			
I can explain the difference between a specialist and primary care physician.			
I can explain legal rights and responsibilities available to me when I am 18.			
I can explain how my health insurance works (provider network, deductible, co-pays).			
Healthcare Transition			
I have decided which things I should consider when selecting a new healthcare provider.			
If necessary, I have transitioned to a new healthcare provider.			
If necessary, I have shared medical information with a new provider.			