

Chapter 43

Adolescents Living with Primary Immunodeficiency Diseases

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Adolescence is a time of great transition physically, cognitively, socially, and emotionally. Adolescents diagnosed with a primary immunodeficiency disease (PI) and their families face not only the day-to-day challenges of any family, but they also must learn to manage the effects of a rare and chronic disease while nurturing growth towards adulthood. Regardless of when an adolescent was diagnosed, they face some unique challenges, and it is important to help them manage the impact of PI as they strive to achieve the developmental steps or stages of a teenager.

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

Normalizing Your Adolescent's Life

During this life stage, adolescents develop the skills needed to establish and maintain family and social relationships. Education continues as they begin to make decisions about their future life goals. They typically go through a series of steps in this maturation process, commonly having both successes and setbacks in navigating the path 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, PIs manifest differently in each individual. Therefore, families must make choices that best suit their adolescent's physical and mental health, as well as abilities.

You can help your adolescent through this time by teaching coping skills to manage day-to-day issues associated with PI while helping them live a normal life.

Begin a dialogue with them, so that they can become a part of their health-related 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.

Helping your adolescent maintain a balanced life.

Teens who best manage their diagnosis are those who find a balanced approach to PI and to life. It is understandable that adolescents would often want a break from focusing on PI, yet neglect of symptoms or treatment routines can lead to serious health setbacks. An emphasis should be placed on both managing their condition (the signs, symptoms and treatments) and maintaining overall health itself (the activities and relationships that promote a healthy lifestyle). The Immune Deficiency Foundation (IDF) Teen Program offers opportunities for teens to help them learn about living with PI and to connect with others. Contact the IDF at www.primaryimmune.org to learn more.

Coordinating with their school.

Living with a PI may disrupt schooling, and as previously mentioned, school is an integral part

of an adolescent's life. You and your adolescent should meet with teachers, counselors, nurses and administration to explain the PI and the potential impact on school, such as frequent absences, fatigue and illness. Work with these professionals to develop a plan to help them keep up with schoolwork when they are absent. 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.

Encouraging a healthy social life.

Encourage your adolescent to explore their talents and interests, and help them to set realistic expectations based upon their individual capabilities and medical needs, and focus on their strengths. Encourage participation in athletics, music, dance or 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 adolescents to participate in school and social activities whenever possible to help them understand 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 PI can be very difficult for them. Acknowledge such disappointments while balancing their health. There is always a chance that they can become ill since germs exist everywhere, but preventing them from participating in group events can create feelings of anxiety and depression. Making simple modifications, such as using hand sanitizer, avoiding shared beverages, and staying away from actively coughing individuals, can allow them to participate.

There are some restrictions that people with certain PIs must follow. For example, those with thrombocytopenia should avoid contact sports. Football or soccer is risky for those with Chronic Granulomatous Disease (CGD) because of exposure to dust or grass. Restrictions should be determined with the immunology team.

Teenagers already struggle with identity issues and confidence, and feeling different can further complicate this matter. Although they might not want to share information about their diagnosis with peers, it is important to develop strategies to help them educate peers and to explain their condition, including the appropriate terms for diagnosis and treatment. Then, they are prepared if they decide to share this information. Because PIs are rare, adolescents may not know others their age 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.

Assure them that they can disclose as much or as little information as they want. They may not feel comfortable telling everything to everyone. They can choose to confide in some and to provide minimal information to others. This is completely their decision. Make sure that you do not publically share more information about their PI than your adolescent does. Respect their wishes when it comes to this.

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 speaking to your son or daughter about how to talk about their PI 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 PI, that person is not the right person for them. It is important to let them know their value and that people with PI date, marry, have children and lead full lives.

Maintaining Family Life with Adolescents Living with PI

Parenting issues in adolescence can be intensified by chronic conditions. Families may struggle to find a balanced approach to maintaining their family life and addressing the health issues. Time, activities and family decisions may require daily modifications. In addition, adolescents with PI often feel guilty that they are burdening their parents with the additional pressures of their illness. Be sure to let them know all the wonderful things that they add to your life, so that you can let them know how valued they are.

Adolescents and their families who best cope with an ongoing health problem typically follow a pattern during the maturation process. In early adolescence, parents are more involved in overseeing and modeling positive healthcare practices. Later, parents should encourage increased involvement and independence. Finally, as they move toward adulthood, parents should encourage them to take main responsibility for managing the disease, with family members as more distant supporters.

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 the brother or sister who has a PI. Acknowledging the impact of a chronic illness upon siblings not diagnosed with PI is important. Praising siblings for their patience and acknowledging the challenges of having a family member with PI can help decrease resentment and validate the siblings' experience. Encourage sibling input on family decision-making, so they feel that their suggestions are also important.

When two or more children in the family have PI, 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 disorder. It is important to individualize every child's needs and not to generalize experiences.

Preparing for Tough Questions from Adolescents Living with PI

Common questions or issues that you may hear from your son or daughter:

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

It will vary in each individual 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 PI?

This may be related to the question about being treated differently. It also involves learning relationship skills of trust building and sharing. They 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 sharing. Encourage them to take advantage of the IDF Teen Program, which includes weekend events exclusively for teens with PI, to learn more about their disease and to connect with others.

How do I handle my PI at school?

When asked this, you may want to determine if they mean the social aspect of school and/or the academic side. They may 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 include them in setting 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 children were younger can be changed or modified to meet the needs of older children. 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 PI. Schedule an appointment between your adolescent and their immunology team 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 PI 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 diagnosis, 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!

"Why me?" is a question often asked by those with a chronic condition. This is a very tough question, and is often asked by parents as well. This may be a question about their particular diagnosis 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." Remind your child that there are many who are also living with PI, and that it is not a punishment, but

something they just have to learn to manage and live with. Creating a positive mindset will help them to better take control of their condition and life.

Offering Resources and Professional Assistance

Many adolescents with PI often feel misunderstood. As much as family and friends may attempt to empathize the impact of these diseases, only individuals with PI can really understand what this disease feels like. Encourage them to connect with peers with PI through the IDF Teen Program. Having others to relate to is invaluable in providing encouragement. Not only can it foster supportive friendships, but involvement with others with PI can also help them feel that they are neither alone nor different from others. IDF Teen Escape weekends, which are held 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 PI. IDF holds biennial National Conferences and Regional Conferences, during which there are programs for teens. Teens may not feel they need to attend such programs, but those who do attend have said they've had extremely positive experiences. For more information about programs and resources for teens, contact IDF: www.primaryimmune.org. Almost everyone is connected on social media today and there are support groups for every possible subject available almost instantly. Make sure that they do not confuse any posted public information as a substitute for professional advice. Anyone can write anything on social media.

Seek professional assistance if they display 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 PI

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 assistance. Having a place to vent about how difficult this challenge can be is important and receiving support from friends, family and partners is necessary. Just as it is imperative that adolescents are connected to the PI community, parents should be too. Having a connection to another parent who understands your experience can be helpful. Parents of individuals with PI may understand your experience more than friends and family who have not had this experience and may find it difficult to relate to the daily pressures of dealing with PI. Taking care of your emotional well-being can make it easier for you to manage caring for any adolescent with PI. IDF offers a variety of resources and programs to connect you with other parents on the IDF website: www.primaryimmune.org.

Coordinating Your Adolescent's Healthcare

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

Adolescents are more likely to make positive choices if they feel they have some say in the decision-making process. Find providers who are willing to work with you and them, and allow for private time between the children and providers. Your adolescents 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 heard and develop a good relationship with healthcare providers.

Planning appointments around their school and social activities allows them to maintain social relationships, which are 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 adolescents and healthcare providers, develop a personalized list of successful approaches to managing your children's health:

- What health and wellness habits have been most successful in keeping your adolescents 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 teen's PI, 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 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, they can develop lifetime habits of positive coping skills for health challenges.

Being Your Adolescent's Healthcare Advocate

As your children's advocate, you work to make sure their needs are being met by the healthcare team. With younger children, the parent is the chief 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 children mature 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 everyone builds positive relationships with the healthcare providers.

Recommended information to record and keep readily available in a record, either digital or print:

- 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 adolescents 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 all current medications they're taking
- 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

While you are going to begin this record, it is important to encourage them to gradually take over this responsibility.

How you can advocate for your adolescents:

- Ask questions about the diagnosis, treatment and plan. If you do not understand, ask again.
- Inquire about what can be done to improve their 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 they receive Ig therapy, make note of how it is going and/or any side effects.
- Build positive relationships with their providers, teachers and therapists. Know whom 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

Planning for Life after High School

Having this disease should not impede your adolescent from pursuing post-secondary education and/or living independently, but it might influence some decisions in terms of obtaining healthcare and living conditions. Some 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. Have your college-bound student contact student support services at their colleges of choice to discuss the diagnosis and specific needs, as well as possible resources and accommodations. Connecting with the health service is important, especially if they are receiving immunoglobulin replacement therapy. Some colleges and universities will not allow intravenous Ig infusions on campus. While subcutaneous therapy may be allowed, shipping of drugs and supplies may be problematic, especially if all mail and deliveries come to a centralized location.

When researching new providers and facilities, you need to understand your adolescent's insurance benefits and what providers and facilities are covered. 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 and resources in that area. Your current providers may be helpful in making recommendations. Many immunologists are associated with major universities so your adolescent might want to consider these types of schools. If your children 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. Your adolescent should establish care, if possible, before moving day to give the healthcare provider time to get to know them and have time to request any required pre-authorizations for treatment from the insurance plan. It is also key to make sure that the health insurance is accepted as in-network even when out of state in order to maintain their care.

If they receive Ig therapy and are relocating and/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. If they receive 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 they turn 18, they are legally considered an adult. At that point, parents need written permission from their children to access healthcare records and to speak with the healthcare team and health insurance company. 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 them build confidence because they have some control over decision-making and it 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 immunology team 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 adolescents become independent with current healthcare providers and to gradually take on more and more responsibility for themselves and their care.

For more information about how to help transition care to your adolescent, use the IDF Transition Guide, which can be downloaded or ordered on www.primaryimmune.org.

On the following pages are checklists to be completed by your adolescents to help them prepare for the transition to adulthood, and can also be found in the IDF Transition Guide.

Transition Skills Checklist

Ages 12-14

| Yes | No | Almost | General Information |
|-----|----|--------|--|
| | | | 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. |
| | | | My parents and I carry a medical summary, such as the in case of emergency (ICE) report |
| Yes | No | Almost | 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. |
| Yes | No | Almost | 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. |

Transition Skills Checklist

Ages 12-14 (cont.)

| Yes | No | Almost | Medical Appointments |
|-----|----|--------|--|
| | | | 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. |
| Yes | No | Almost | 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. |
| Yes | No | Almost | 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 (cont.)

| Yes | No | Almost | General Information |
|-----|----|--------|---|
| | | | My parents and I keep a personal health record. |
| | | | I carry a medical summary. |
| | | | 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. |

Transition Skills Checklist

Ages 15-17 (cont.)

| Yes | No | Almost | Medications and Treatment |
|-----|----|--------|--|
| | | | 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. |
| Yes | No | Almost | 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. |
| Yes | No | Almost | 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). |
| Yes | No | Almost | Healthcare Transition |
| | | | I know if my health care providers will stop treating me at a certain age. |

Transition Skills Checklist

Ages 15-17 (cont.)

| Yes | No | Almost | Healthcare Transition |
|-----|----|--------|--|
| | | | 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 (cont.)

| Yes | No | Almost | General Information |
|-----|----|--------|--|
| | | | I keep a personal health record. |
| | | | I carry a medical summary. |
| Yes | No | Almost | Medications and Treatment |
| | | | I understand and/or arrange payment for my medications, equipment and treatments. |
| Yes | No | Almost | 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. |

Transition Skills Checklist

Ages 18 and Up (cont.)

| Yes | No | Almost | Medical Appointments |
|-----|----|--------|--|
| | | | I locate and share healthcare information with my providers and in making decisions about my care. |
| | | | I sign medical consent forms. |
| Yes | No | Almost | 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). |
| Yes | No | Almost | 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. |