

Chapter 42

Infants and Children Living with PI

Kristin Epland, FNP-C, Midwest Immunology Clinic, Plymouth, Minnesota, USA

When a child is diagnosed with a primary immunodeficiency disease (PI), each member of the family is affected. The family must come to terms with the illness and be prepared to adjust schedules and priorities. With proper anticipation and realistic expectations, quality of life can be normal.

Helping a Child Understand

Children's understanding of PI depends on where they are in terms of their cognitive development. They adjust differently to illness and family life at each developmental stage. Here is information about how children cope at different ages and how a parent/caregiver can help a child better understand their diagnosis.

Infants and Toddlers (ages 0 to 2) are just beginning to develop trust and security, and they usually do not have an understanding of their diagnosis. They may experience challenges to their development of trust and security when they experience pain, restriction of motion, and separation from parents. A parent/caregiver can help by staying with the child for medical procedures and hospitalizations as well as holding, comforting, and interacting with the child as much as possible. Bringing a favorite stuffed animal, pacifier or blanket along to treatments may be helpful as well.

Preschool Children (ages 3 to 4) are ready to begin to be independent and eager to make choices. They may understand what it means to get sick, but they may not understand why and how. Being in the hospital or adjusting to medication schedules can sometimes take away their freedom and choices. Children may try to challenge limits set by parents as a way to exert some control. A parent/caregiver can help by being firm and consistent with things the child does not have a choice over (such as taking medications, going to the doctor, etc.). However, when possible, let them make some decisions, like what medication to take first, what chair to sit in when getting blood drawn, and what color bandage to use or which site to use for their treatment. Rewarding a child for making positive choices about their health is also important. Do not make promises to the child that cannot be kept. For instance, do

not promise a child that it will only be one poke or that there will not be any blood draws at any certain office visit. Broken promises can damage trust in this age range.

Early School Age Children (ages 5 to 10) are developing a stronger sense of control over their environment. They may have a greater understanding about their disease, but these reasons may not be entirely logical. Children in this developmental stage may believe they caused their illness by thinking bad thoughts, acting out, or by not following rules at home. At this stage, children are also beginning to notice that they may seem different from their peers. When these emotions are not dealt with, they can lead to sadness, anger, anxiety, or withdrawal. Parents can have a big influence on how their child responds to feeling different. Remind them that they are not alone. Engage in an ongoing dialogue about what they are experiencing and encourage them to share their feelings. Parents can help by making sure their child knows that their diagnosis is not their fault and that they did nothing to cause it. It may also be beneficial to allow children to participate in the management of their care. For example, allowing the child to communicate with their doctor or help keep track of their medication schedule can go a long way in helping the child develop a stronger sense of control.

Parents can help children cope with their disease and treatments by encouraging them to practice on a doll or stuffed teddy bear with a toy doctor kit. Letting the child take the doll or bear's temperature or blood pressure, listening to its breathing, or even practice painful procedures (such as shots, blood draws, infusions, etc.) can help relieve anxiety the child may be feeling. Parents/caregivers should participate in this play, but it is important for the child to take the lead.

Older School Age Children (age 11+) want to be more independent from their parents. Relationships with friends and social activities are exceptionally important to children of this age. Children may feel frustrated, angry, and left out if they are forced to miss activities due to illness or restriction. Children of this age may also start to struggle with not wanting to take their medicine, especially if they are feeling well. They may not feel as though they need it any longer. While they are better able to understand their diagnosis and its treatment, they should not be expected to react as adults do. Parents/caregivers can help by explaining to the child how important medications can be in the management of PI, even when they begin to feel better. With the approval of the child's healthcare provider, the child should participate in school or other activities whenever possible. Be sure to include the child in discussions with their medical team when possible. Encourage the child to prepare for a medical visit by writing down questions to ask. This will help children feel included and give them a greater sense of control over the situation. Listening to the child is always essential, and it may be helpful to encourage the child to express emotions through play, art, drawing, music or reading.

Normalizing a Child's Life

Parents of a child living with PI may be faced with challenges, difficulties, and decisions that other parents will never have to face. This may be overwhelming; however, there are ways to support the child and help the family cope.

Medical

Explain the diagnosis to the child in ways he or she can understand. One of the most important things parents can do for children with PI is to provide accurate age-appropriate information and encourage children to ask questions. Children who lack information about their diagnosis tend to make up information that is often inaccurate and scarier than the actual circumstances. It is important to let children know that the diagnosis is not their fault and that it is not a punishment. Make sure the child knows that the parents/caregivers are there to answer any questions they may have. Having open and honest communication with the child helps build trust and a sense of security, and it helps the child cope better. It is important to let the child know that the parents/caregivers trust the healthcare team so the child, in turn, can learn to trust providers.

Become informed about special medical issues affecting the child. These may include:

- Infection precautions, including school, sleepovers, camps, and airline travel
- Use of antibiotics if they get sick or following known exposures
- Vaccines for the child and family
- Avoidance (if necessary) of swimming, gardening, playing in the leaves, etc.
- Precautions about school sports if necessary
- Nutrition
- Dental hygiene
- Genetic counseling of parents
- Insurance issues

Prepare the child for medical procedures. Children need to know what to expect before medical appointments. Explain, in an age-appropriate way, the reasons why procedures are being done, who will be doing it, what equipment will be used, and whether or not it will be painful or uncomfortable. This will give the child an opportunity to ask questions, build trust, and cope in their own ways instead of worrying about the unknown. It is important to give consistent and accurate information. For example, do not tell the child that something will not be painful or uncomfortable when it will be. Child life specialists are available at many hospitals and can help prepare children for hospitalization, surgery, and various medical procedures.

Emotional

Help the child deal with feelings about the diagnosis. Try to understand the many emotions that children experience regarding their PI. Parents/caregivers can help the child cope with difficult emotions by talking openly about how everyone experiences similar emotions at times in their life—isolation, anxiety, sadness—for various reasons. Providing routine and predictable times to check in with a child gives them opportunities to talk and to share, and it gives parents/caregivers opportunities to reassure them that their feelings are normal and acceptable. Parents/caregivers can ask questions in a way to get a child talking by using open-ended questions. "What kind of questions do you have?" is very different than "Do you have any questions?" Parents/caregivers can also ask questions about specific

behavior: “Lately, you have been getting angry about things that do not normally bother you. Why do you think that is?” Finally, provide ways to help the child get rid of unhappy feelings. Some examples include using play or art to express their emotions.

Give a child choices whenever possible. Many children living with PI tend to think they have little control over their lives. Children need opportunities to make choices—to have power over any part of their lives they can control. This can be done by offering the child choices whenever possible (such as what they would like for dinner, what activity they would like to do that day). When appropriate, it can also help to have the child participate in making small decisions about their treatment (such as what arm to get a shot in, what day of the week or month to take their treatment, what site to use to get their infusion, etc.).

Social

Prepare the child for the reactions of others. Children with PI often do not know how or what to tell others about their illness and symptoms, particularly at times when they look healthy on the outside.

Parents/caregivers can help by teaching the child a simple and short explanation of the diagnosis. Make sure they are comfortable explaining what is necessary to stay well. It may help to role-play examples of how to answer questions that others might ask and to handle any teasing that might occur. Be sure to include siblings in these discussions as well, as they often experience similar situations with their peers.

Look for role models. Many children with PI feel different and isolated. Being around others with the same diagnosis can often help them in this regard.

- The Immune Deficiency Foundation (IDF) offers many ways for children and families to interact throughout the year at events held across the country: www.primaryimmune.org/events. Children often benefit from having contact with others who have the same illness.
- Parents/caregivers can also connect online through IDF Friends, www.idffriends.org, or in the IDF Facebook groups, www.facebook.com/ImmuneDeficiencyFoundation.
- Additionally parents/caregivers can request IDF to connect them with a trained peer support volunteer that has experience living with a child who has PI: www.primaryimmune.org/idf-peer-support-program.

School

Living with a PI may disrupt a child’s schooling. It is important for parents/caregivers to meet with teachers, counselors, nurses, and administrators to explain their child’s PI and the potential impact on school (such as frequent absences, fatigue, activity restrictions). Consider what parents and other children in the class should be told about the child’s diagnosis, if anything. One issue that should be discussed with a child’s teacher and school nurse includes a reminder to other parents/caregivers in the classroom to keep children who have infectious symptoms home in order to limit exposures to infections. For instance, ask the teacher to review protocol surrounding fever or vomiting with class parents. A plan should be developed to help the child keep up with schoolwork when they cannot attend school. An excellent free resource to help coordinate with a child’s school is the IDF School Guide, available to order or download at: www.primaryimmune.org.

Family Life

A diagnosis of a chronic illness such as PI affects the entire family system. Research shows that how well a child with a chronic illness copes depends on how the entire family is supported. A family that has healthy coping skills is more likely to follow treatment and care plans, and to be active in seeking support.

Chronic illness can affect a family in many ways. Some parents/caregivers may experience increased worry, stress, problems with sleep or appetite, sadness, anger, a sense of loss, and even a feeling of relief. These conflicting emotions can be difficult to deal with, but they are a normal part of the coping process. Parents/caregivers may have less time for each other and for social activities they once enjoyed. Planning for fun times may be difficult due to the unpredictability of the child’s illnesses. Financial worries may also increase.

Siblings may experience a wide range of emotions when their brother or sister is living with a PI. These emotions often include anger, guilt, embarrassment, sadness, loneliness, fear, and confusion. Siblings may also experience jealousy if they receive less attention. It is important to talk with siblings about their feelings and not to simply dismiss them, thinking they will get over it on their own.

Families can benefit from strategies that help them to relieve stress, share responsibilities, gain support, and explore emotional worries. Approaches include:

- **Help the child lead as normal a life as possible.** To whatever extent possible, try to treat the child with a PI just like any other child. At the same time, parents/caregivers need to take into consideration the child's health and any special needs that they may have. This can be a balancing act, but it is important for parents/caregivers to encourage their child's participation in activities that involve other children of the same age.
- **Maintain family routines.** As much as possible, maintain regular family routines (such as wake-up times, mealtimes, bedtimes, regular activities, chores, discipline, etc.) as this can help offset some of the disruption experienced due to living with a PI. Children typically do better when their daily routines are predictable and consistent. Of course, this is not always possible, but every effort should be made to maintain structure and schedules for family members. Continue to enforce the usual discipline in the household. Setting and enforcing limits that are fair to all family members will decrease resentment in siblings.
- **Help the other children cope.** A child living with a PI may demand a lot of parental attention. Siblings may feel jealous, angry, lonely, and worry about their sibling and about their parents/caregivers. They also might worry that they might get the disease. Explain the disease to siblings. Try to get them to ask questions and to express their concerns. Parents need to keep lines of communication open with all of their children. It often helps children feel like an important member of the family if they can have a part in caring for their sibling in some way. One way to help siblings is by focusing on fun family activities when the child with a PI is healthy. It can be beneficial for parents to spend individual quality time with each child, letting each of them know how much they are loved, valued, and appreciated.
- **Make having fun together as a family a priority.** Living with a child's PI may cause the whole family to be under increased stress. Getting support from each other may be harder during times of stress, but it is also even more important. Spend time together that is not focused on the disease and make it a priority to carve out time for whole family activities. It is equally as important to have special alone time just for parents/caregivers and even for one-on-one parent-child dates.

Coordinating The Child's Healthcare

When a child is diagnosed with a PI, parent/caregivers become part of the child's healthcare team and their main advocate. The role in monitoring the child's symptoms, responses to treatments, and communicating the observations and concerns is vital to the medical team's assessment and treatment of the child. In many cases, more than one provider will be involved in caring for the child; therefore, coordinating communication and keeping comprehensive and accurate records of the child's medical course is essential. Many parents/caregivers suggest that a journal is an invaluable tool to document events affecting the child's medical care. There are also online personal health records that can be helpful.

Recommended information to record:

- Brief history leading to the diagnosis, written by a parent/caregiver or the child's healthcare provider.
- Copies of laboratory evaluations confirming the diagnosis.
- Current list of providers caring for the child with accurate addresses and phone numbers.
- Chronology of important events, specifically noting types of treatment and therapy, changes in therapy and subsequent responses to that therapy, surgeries and/or hospitalizations.
- List of the child's current medications.
- Allergies to medications.
- Immunization record.
- Current insurance information.
- Explanations of benefits (from the insurance company) can be kept in the journal or separately but should be periodically reviewed for accuracy.

Insurance concerns that arise are more easily resolved through accurate record keeping. A journal or online health record will be useful if the child should need to see a new provider, especially in an emergency. This form of accurate record keeping shortens the lengthy, often repeated history-taking sessions by new providers, allowing more time to focus on the immediate issues at hand. It is wise for more than one person in the family to be aware of the child's medical routine. A well-documented

medical record maintained by parents/caregivers can be extremely helpful for those times when others care for the child.

In addition to bringing a journal to each medical visit, additional suggestions when visiting a medical professional include:

- **Prepare questions:** Have a list of questions prepared in writing.
- **Take notes:** Document the visit by writing details about the visit. When possible, take another family member or friend along. It is always wise to have more than one person familiar with the child's medical routine.
- **Plan ahead:** Be prepared for a change in plans or long office visits. Sometimes parents/caregivers and the child will go for tests immediately after the visit or the visit could be extended for other reasons. If this is the case, make arrangements for other children.
- **Communicate directly with the child:** Encourage the healthcare professional to communicate directly with the child when possible. Although the child may be young, it is always appropriate for them to build a relationship with their healthcare providers.
- **Ask for written instructions:** Request written instructions concerning medicines and treatments. This helps avoid mistakes by all parties, and provides written instructions to be placed in the journal or scanned and saved into an online personal health record.
- **Prepare a tote bag:** Designate a special tote bag just for these medical visits and include the following items:
 - » Toys and/or activities: It may not be wise to share toys at the doctor's office to reduce exposure to germs. You can also prepare age-appropriate activities to engage them.
 - » Books: Take along favorite books or a new book to help the child stay occupied and calm during long waiting periods.
 - » Game device or smart phone: These are also useful for distraction and to alleviate boredom.
 - » Notebook: Parents/caregivers or another family member can take notes.

- » Contact list: Include a contact list with names and phone numbers of family, friends, and school personnel.
- » Snacks: Bring snacks in case the visit may be extended.

Being a Child's Advocate

A parent/caregiver is their child's best advocate. It is important to communicate concerns and questions to the child's providers. Using a journal or an online personal health record will help remind a parent/caregiver about what to discuss with the child's provider at various visits. Overall, parents/caregivers should follow their intuition. They generally know when something is going on with their child.

How to Advocate for a Child:

- Ask questions about the child's diagnosis, treatment, and plan. Ask again if there is something unclear.
- Inquire about what can be done to improve the child's health, such as diet, physical activity, sleep, and social activities.
- Maintain consistent communication with the school as the child may miss school days.
- Know the child's insurance policy and communicate if there are any changes to the provider.
- If the child receives immunoglobulin (Ig) replacement therapy, make notes of how it is going and/or any side effects.
- Build positive relationships with the child's providers, teachers, and therapists. Know whom to call when.
- Ask about resources for further information at the local, state, and national level.
- Connect with IDF for additional resources: www.primaryimmune.org or 800-296-4433.

Transitioning Responsibility to the Child

As children develop, they begin to form their own thoughts and opinions of their care. When it is appropriate, offer choices to the child. This helps the child build confidence because they have some control over decision-making and will prepare the child to participate and eventually take over their healthcare in adolescence and adulthood. The better prepared the child is, the easier the transition will be.

When to Ask for Help

Having a child with a PI forces the entire family to cope with many changes and stressors. It can cause emotional and behavioral challenges for the child, parents, siblings, and extended network of family and friends. Because of these challenges, family members may be more likely to experience adjustment difficulties as they learn to adapt.

It is important to support the child's emotional and behavioral needs as well as the needs of the entire family. Counseling services can be a valuable part of the child's treatment plan. The most successful families tend to be those who are working together as a team to face the new responsibilities of managing a long-term illness. They build on their family's strengths to cope with the new stress and can help the family grow closer together.

Every situation is unique, but there are similarities in how children and families react to the stress of living with PI. Adjustment difficulties commonly observed in children with chronic health conditions and/or in their parents and siblings include the following:

- Disturbance of mood: feelings of anxiety, fear, sadness, depression, hopelessness, irritation, anger, emptiness, and/or guilt; frequent worrying; disinterest or lack of pleasure in activities formerly of interest.
- Behavior difficulties: mood swings, temper outbursts, aggressive behavior, not cooperating with medical care, changes in activity or energy level, separation anxiety or clinging behavior, regressive behaviors, reenactment of their situation/trauma, and/or acting out by not listening, fighting, or even hitting.
- School issues: academic problems, change in school performance, and/or difficulty with concentration.
- Social issues: isolation from peers, feeling disconnected from people, lack of interest in things they previously enjoyed, and/or fights with friends.
- Self-esteem issues: sense of being different, low self-confidence, and/or negative comments about the way they look or feel.
- Family issues: increased strain in relationships, different perceptions of issues, blame, communication difficulties, fights with siblings, and/or ignoring other family members.

- Parent issues: time-management difficulties, financial worries, marital stress, guilt, self-blame and/or blame of others, grief, and/or discipline problems
- Physical issues: changes in eating, sleep disturbances, stomachaches, headaches, tiredness, and/or over-activity.

Remember that it is a sign of strength to be able to ask for help from counselors and other support professionals. Support can be sought at any time. Parents/caregivers do not need to wait for a crisis. In fact, it is better to arrange for support sooner rather than later. Also, it is normal to experience the need for support at some times and not at others. Adjustment is an ever-changing process.

Addressing the Needs of the Parents/Caregivers

Parents/caregivers should remember to take care of themselves. Addressing personal needs will allow a parent/caregiver to provide better care for the child.

Learn about a child's diagnosis. Being knowledgeable allows parents/caregivers to make informed decisions about their child's care and to know which behaviors and symptoms are normal and which are not. It also helps parents answer questions their child may have about their disease.

Practice self-care. This may seem like a difficult task for many parents/caregivers. Nevertheless, it is vital for parents to take care of themselves. Otherwise, they will not be able to give good care. It is important to get connected with other parents who know what it is like to have a child living with a PI. Allow others to help and be sure to carve out time to do something you enjoy. Find someone to listen to the worries and make it a priority to spend quality time with the partner on a regular basis. Learn to deal positively with the stress by eating right, exercising, keeping a journal, and spending quality time with the children.

Remain hopeful. Coping with PI can be discouraging and scary at times. It is important to stay positive and hopeful. Do not ignore or dwell on worries and negative feelings. Instead, recognize and address them in a positive manner. Trying to find the positive side of things enables parents/caregivers to teach their children a valuable lesson as well as maintaining their own peace of mind.