

Infants and Children Living with Primary Immunodeficiency Diseases



Chapter 29

When a child is diagnosed with a primary immunodeficiency disease, each member of the family begins an unexpected and challenging journey. The whole family must come to terms with the illness and perhaps make major changes in schedules and priorities. As a result, it is a journey that may have unforeseeable turns but also many joys and rewards—a journey that can be meaningful.

Helping Your Child Understand

Children's understanding of their primary immunodeficiency disease 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 you can help your child better understand:

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

Preschool Children (ages 3-4) 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. You can help by

being firm and consistent with things your 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. Praising your child for making positive choices about their health is also important.

Early School Age Children (ages 5-10) 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, by hitting their sibling or by not following rules at home. At this stage, children are also beginning to notice that they may seem different from their peers. You can help by making sure your child knows that their primary immunodeficiency disease 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

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control. Parents can also 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. You 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

better. They may not feel as though they need it any longer. While they are better able to understand their primary immunodeficiency disease and its treatment, they should not be expected to react as adults do. You can help by explaining to your child how important medications can be in the management of primary immunodeficiency, even when they begin to feel better. With the approval of your child's healthcare provider, your child should participate in school or other activities whenever possible. Be sure to include your child in discussions with their medical team when possible. This will help children feel included and give them a greater sense of control over the situation. Listening to your child is always essential and it may be helpful to encourage your child to express these emotions through play, art, drawing, music or reading. *(See the chapter titled "Adolescents Living with Primary Immunodeficiency Diseases.")*

Normalizing Your Child's Life

Parents of a child living with a primary immunodeficiency disease are often faced with many challenges, difficulties and decisions that other parents will never have to face. This may be overwhelming, however, there are ways to support your child and help your family cope.

Medical

Explain the diagnosis to your child. One of the most important things parents can do for children with primary immunodeficiencies is to provide accurate age-appropriate information and encourage the 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 also important to let children know that the diagnosis is not their fault and that it is not a punishment. Make sure your child knows that you are there to answer any questions they may have. Having open and honest communication with your child helps build trust and a sense of security, and it helps your child cope better.

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

- Infection precautions, including co-sleeping, school, sleep overs, camps and airline travel
- Use of antibiotics if they get sick or following known exposures
- Vaccines for your child and family
- Avoidance (if necessary) of swimming, gardening, playing in the leaves, etc.

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- Precautions about school sports if necessary
- Nutrition
- Dental hygiene
- Medical alert badge
- Dental and medical procedure pre-medications
- Genetic counseling of parents
- Insurance issues

Prepare your child for medical procedures. Children need to know what to expect in their lives. Explain, in an age-appropriate way, the reasons why the procedure is 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 critically important to give consistent and accurate information. For example, do not tell your child that something will not be painful or uncomfortable when you know that 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 your child deal with feelings about the diagnosis.

Try to understand the many emotions that children experience regarding their primary immunodeficiency disease. You can help your child cope with difficult emotions by talking openly about how everyone in the family may be experiencing something similar. Providing routine and predictable times to check in with your child gives them opportunities to talk and to share, and it gives you opportunities to reassure them that their feelings are normal and acceptable. You can ask questions in a way to get your child talking by using open-ended questions. “What kind of questions do you have?” is very different than “Do you have any questions?” You 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 your child

get rid of unhappy feelings. Some examples include using play or art to express feelings.

Give your child some choices. Many children living with primary immunodeficiencies 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 your child for the reactions of others. Children with primary immunodeficiency diseases 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. You can help by teaching your child a simple and short explanation of the diagnosis. Make sure your child is comfortable explaining what is necessary to keep well. It may help for you and your child 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 primary immunodeficiency disorders 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, including family retreat weekends, patient education meetings and a national conference held every other year. You can share and ask questions on IDF's social network, *IDF Friends*, www.idffriends.org. You can ask IDF to connect you with a trained peer support volunteer that

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has experience living with a child who has a primary immunodeficiency disease. Children often benefit from having contact with others who have the same illness. IDF can connect your child with other children living with a primary immunodeficiency disease.

Coordinate with your child's school. Living with a primary immunodeficiency disease may disrupt a child's schooling. It is important for parents to meet with teachers, counselors, nurses and administrators to explain their child's primary immunodeficiency disease

and the potential impact on school (such as frequent absences, fatigue, activity restrictions). You should talk about what parents and other children in the class should be told about your child's primary immunodeficiency disease. A plan should be developed to help your child keep up with schoolwork when they cannot attend school. A good resource to help you coordinate with your child's school is the *IDF School Guide*, available to order or download at www.primaryimmune.org.

Family Life with Infants and Children Living with Primary Immunodeficiency Diseases

A child living with a chronic illness 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 your family in many ways. You may experience increased worry, stress and 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 healing process for you and your family. Parents 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 illness. Financial worries may also increase.

Siblings may experience a wide range of emotions when their brother or sister is living with a primary immunodeficiency disease. 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 children 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 your child lead as normal a life as possible.

To whatever extent possible, you should try to treat your child with a primary immunodeficiency disease just like any other child. At the same time, you need to take into consideration your child's health and any special needs that they may have. This can be quite a balancing act, but it is important for parents to encourage their child's participation in activities that involve other children of the same age.

(Family Life Continued)

Maintain family routines. You should, 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 primary immunodeficiency. 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 regular routines and schedules for all family members.

Help your other children cope. A child living with a primary immunodeficiency disease demands a lot of parental attention. It is no wonder that brothers and sisters often feel jealous, angry and lonely and worry about their sibling and sometimes about their parents. They also might worry that they might get the disease. You should explain the disease to your other children. Try to get them to ask questions and to express their concerns. Parents need to keep open lines of communication 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 your child with a primary immunodeficiency disease 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 primary immunodeficiency 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 and even for one-on-one parent-child dates, as mentioned earlier—each parent spending individual time with each child.

Coordinating Your Child's Healthcare

When your child is diagnosed with a primary immunodeficiency disease, you become part of your child's healthcare team and their main advocate. Your role in monitoring your child's symptoms, responses to treatments and communicating your observations and concerns is vital to the medical team's assessment and treatment of your child. In many cases, more than one provider will be involved in caring for your child; therefore, coordinating communication and keeping

comprehensive and accurate records of your child's medical course is essential. Many parents suggest that a journal is an invaluable tool to document events affecting your child's medical care. IDF ePHR, an online personal health record designed for the primary immunodeficiency community, is another tool to help record your child's medical information:
www.idfephr.org.

(Coordinating Your Child’s Healthcare continued)

Recommended information to record:

- Brief history leading to the diagnosis, written by you or your child’s healthcare provider
- Copies of laboratory evaluations confirming the diagnosis
- Current list of providers caring for your 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 your child’s current medications
- Allergies to medications
- Immunization record
- Current insurance information
- Explanation of benefits records 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, and a journal or IDF ePHR will be useful if your 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 issue 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 you can be extremely helpful for those times when others care for your 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 patient’s medical routine.

- **Plan ahead:** Be prepared for a change in plans or long office visits. Sometimes you and your child will go for tests immediately after the visit or the visit could be extended for other reasons. If this is the case, you may need to make arrangements for your other children.
- **Communicate directly with the child:** Encourage the medical professional to communicate directly with your child when possible. Although your 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, as well as give you written instructions to be placed in your journal or scanned and saved into the ePHR.
- **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 because you do not want to go home with more germs. You can also prepare age-appropriate activities to engage them
 - *Books:* Take along favorite books or a new book to help your 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:* You 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 Your Child's Advocate

As a parent, you are your child's best advocate. It is important to communicate with your child's providers the concerns and questions you or your child may have. Using a journal or IDF ePHR will help you remember what to discuss with your child's provider at various visits.

How you can advocate for your child:

- Ask questions about your child's diagnosis, treatment and plan. If you do not understand, ask again.
- Inquire about what can be done to improve your child's health such as diet, physical activity, sleep and social activities.
- Maintain consistent communication with the school as your child may miss school days.
- Know your insurance policy and communicate if there are any changes to your provider.
- If your child receives immunoglobulin (Ig) therapy, make notes of how it is going and/or any side effects.
- Build positive relationships with your 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 Your Child

As children develop, they begin to form their own thoughts and opinions of their care. Again, when it is appropriate, offer choices to your child. This helps your child build confidence because they have some control over decision-making and prepares your child to

participate and eventually take care of themselves 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 primary immunodeficiency disease 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. It is also important to support the needs of the entire family. Counseling services can be a valuable part of your 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 primary immunodeficiency diseases. 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 or fear, sadness and depression, hopelessness, irritation, anger, disinterest or lack of pleasure in activities formerly of interest, emptiness, guilt, and frequent worrying
- **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 acting out by not listening, fighting, or even hitting
- **School:** academic problems, change in school performance, and difficulty with concentration
- **Social issues:** isolation from peers, feeling disconnected from people, lack of interest in things they previously enjoyed, and fights with friends
- **Self-esteem issues:** sense of being different, low self-confidence, and negative self-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 ignoring other family members
- **Parent issues:** time-management difficulties, financial worries, marital stress, guilt, self-blame and/or blame of others, grief, and discipline problems
- **Physical issues:** changes in eating, sleep disturbances, stomachaches or headaches, tiredness, and 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. You 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 Your Needs

Parents should remember to take care of themselves. Addressing your own needs will allow you to provide better care for your child.

Educate yourself about your child's diagnosis. Being knowledgeable allows parents 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.

Take care of yourself. This may seem like a difficult task for many parents. Nevertheless, it is vital for parents to take care of themselves. Otherwise, you 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 primary

immunodeficiency disease. Allow others to help by giving you a break and be sure to carve out time to do something you enjoy. Find someone to listen to your worries and make it a priority to spend quality time with your partner on a regular basis. Learn to deal positively with your stress by eating right, exercising, keeping a journal and spending quality time with your children.

Be hopeful. Coping with a primary immunodeficiency disease can be discouraging and scary. It is incredibly important to stay positive and hopeful. Do not ignore or dwell on your worries or negative feelings. Instead, recognize and address them in a positive manner. If you try to find the positive side of things, you will be teaching your child a valuable lesson as well as maintaining your own peace of mind.