



CHAPTER

19

Infants and Children with Primary Immunodeficiency Diseases

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Introduction

If your child has received the diagnosis of a primary immunodeficiency disorder, it is important to understand the role you will play in the child's future. It would be nice to think you could adjust to this new role slowly, however, unless there is a family history, you are presented with a number of new challenges with little preparation. One of your first challenges is to minimize the impact of chronic illness upon the child's life without compromising their care.

Once the diagnosis of a primary immunodeficiency has been made, children must learn how to live with this diagnosis on a daily basis. Although having a primary immunodeficiency disease is a chronic disorder, the symptoms and their impact on the child will vary considerably. The diagnosis of a primary immunodeficiency does not mean that the child will be sick every day. Most diagnosed children will continue to play, go to school and socialize normally.

Understanding the diagnosis, using preventive measures, and communicating with medical professionals will help you, your child, and your family live with this chronic health condition.

The family as a whole is affected by any chronic illness and should be encouraged to participate in decision-making events that affect the family unit. Family stresses on top of those encountered while managing chronic illnesses can be minimized if recognized early and addressed immediately. Communication with all parties is essential.

Dealing with a chronic illness in an infant or child can be very emotional. Fear of the unknown can be one of the most prevalent emotions, but can be easily controlled with education. If you want your child to grow up and be able to handle their own care, lead by example. Don't feel ashamed to seek help from medical professionals or others in your situation, and look for credible information sources.

Coordinating Your Child's Care

When your child is diagnosed with a primary immunodeficiency disease, you become part of your child's health management team and his or her primary advocate. Your role in monitoring your child's symptoms and 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 physician 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 diary is an invaluable tool to document events affecting your child's medical care.

Recommendations for items to be kept in the diary include:

- A brief history leading to the diagnosis that can be written by the parent or a physician
- Copies of laboratory evaluations confirming the diagnosis
- A current list of physicians caring for the child with up-to-date addresses and phone numbers

- A chronology of important events, specifically noting types of treatment and therapy, changes in therapy and subsequent responses to that therapy
- A current up to date list of the child's medications
- Allergies to medications
- An immunization record or lack of immunization
- Current insurance information
- Explanation of benefits records can be kept in the diary or separately, but should be periodically reviewed for accuracy

Insurance concerns that arise are more easily resolved through the use of the diary. The diary may be especially useful if the child should need to see a new physician, especially in an emergency. This form of accurate information shortens the lengthy, often repeated history-taking sessions by new physicians, allowing for 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 diary can be extremely helpful for those times when the child is in the care of caregivers other than parents.

Coordinating Your Child's Care continued

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

- Have a list of questions prepared in writing. Doctors cannot spend as much time as they would like with each patient, so be ready with your list of questions.
- Remember to take notes. When possible, take another family member or friend along on the visit. It is always wise to have more than one person familiar with the patient's medical routine. This will allow you time to visit with the doctor individually, if necessary, as well.

Designate a special tote bag just for these medical visits. The tote should contain:

- A couple of toys or age-appropriate activities. It may not be wise to share toys at the doctor's office; you don't want to go home with more germs.
- Favorite books or a new book can help your child stay occupied and calm during long waiting periods.

- A notebook for taking notes
- A contact list with names and phone numbers of family, friends, and school personnel

Sometimes you and your child will go for tests immediately after the visit or the visit could be extended for other reasons. Be prepared for a change in plans or long office visits. For instance, you may need to make other arrangements for your other children.

Encourage the medical professional to communicate directly to your child when possible. Although your child may be young, it is always appropriate for him or her to build a relationship with involved healthcare providers.

Ask for written instructions concerning medicines and treatments. This will help avoid mistakes by all parties, as well as give you written instructions to be placed in your medical diary.

Normalizing Your Child's Life

When a child has a chronic health condition, everyone in the family is affected. Parents may be tempted to be overprotective, which is a very natural response as it reflects the concern of keeping the child as healthy as possible. It is also common for parents to want to compensate for the additional challenges their child with a primary immunodeficiency faces.

Such challenges may include:

- coping with symptoms that may be uncomfortable or hinder regular activities
- daily treatments or medicines
- trips to the physician's office
- uncomfortable procedures

It may be a natural inclination to compensate for challenges by loosening rules and expectations or by providing rewards. However, the loosening of rules, or provision of extra rewards, may result in some undesired consequences. For instance, children may recognize when parents change what

is expected of them and worry about why this has changed or what the change means. Some children may even wonder if it means their illness is getting worse. Changes in expectations, or expectations that are different from their siblings, may also serve to confirm the child's concerns about being different and he or she may perceive that difference negatively. In addition, children may expect this special treatment to continue even when parents or other caregivers begin to transition to more typical behavioral expectations, creating a potential cause of friction in the family. Finally, brothers and sisters are also likely to sense a difference in behavioral expectations and may become jealous and/or resentful of the attention and rewards the child with primary immunodeficiency receives.

It is helpful to remember that children need limits and consistent expectations and responses to their behavior. This provides security to children by increasing the predictability of their world. Developing and maintaining expectations, or "family rules" for all children in the family helps

Normalizing Your Child's Life continued

them understand their role in the family and what to expect as well as what is expected of them. If your child with primary immunodeficiency is unable to do his chores, reevaluate the expectations and find something else that he or she can do to contribute to the family. If he or she is able but not willing, or chooses not to follow through on an expectation, the consequences should be clearly stated, age-appropriate and similar to siblings, and carried out. This process of limit-setting and discipline should be the same for all children in the family.

Similar to the pitfalls of relaxing family rules and expectations, providing children with rewards requires some careful consideration. As parents find, because there are so many trips to the physician's office, it becomes clear that they cannot reward the child with every needle stick

or test. Such procedures or treatments may indeed be challenging for your child. Planning and practicing ways of coping can help you and your child better manage difficult events. For those times when a reward is appropriate, provide your child with a few choices that blend into his or her everyday world. For example, on treatment days, allow your child to select a much loved activity or a favorite meal for dinner.

Providing knowledge of your child's condition to their friends and their friends' parents at an early age helps to foster acceptance. They will grow up together knowing the special circumstances that surround them. Your child will know he or she is not so different after all.

Preparing for School or Other Care Outside the Home

In addition to their role with the health care team, parents also act as the link with their child's other caregivers, such as those adults who interact with and supervise their children in childcare or school. The transition from infant/toddler to school-aged child is particularly challenging. Often this is the first occasion the child and parent are separated for an extended length of time. Also, the addition of new care providers can create anxiety for children and parents alike. Conversely, this opportunity to grow intellectually and emotionally should be greeted with enthusiasm as it represents a great milestone in life.

Children are very perceptive and will often share their parents' emotions during this change in life. An optimistic outlook beginning weeks, even months, before the first day away eases the transition to school or care outside the home. Many parents recommend advance preparation to feel more comfortable with any specific concerns related to their child's health needs. Preparation includes a refresher course on your child's particular primary immunodeficiency and his or her current therapy. By reviewing your child's medical diary with school officials and personnel, you will aid in educating them about your child's condition and potentially facilitate the prediction of illness patterns in this new

environment. Timing and early warning signs of illness should be discussed with key personnel (i.e., school nurses, teachers, counselors, and principals). Your child's physician and other health care providers may also be called upon to answer any specific questions. Other items to consider include transportation on normal and sick days, as well as a phone "call down" list in case of illness. Appropriate letters from the physician about physical limitations, if any, medications to be given at school, and immunization recommendations, should be obtained in advance to allow resolution of specific concerns prior to the beginning of school. In addition, plan in advance with your child's teachers for specific needs that may impact school routine. Special arrangements may be necessary for children who need frequent meals or restroom privileges due to intestinal malabsorption, hall passes or scheduled nursing visits for medication administration, and/or assignment of classes to minimize the effects of absences due to regularly scheduled treatments or doctor's visits. Yearly review of these items should allow a safe and smooth transition throughout the school experience.

Some parents have reported two types of misunderstandings that may arise among other people with little knowledge of primary

Preparing for School or Other Care Outside the Home continued

immunodeficiency diseases. One is the perception that parents of children with primary immunodeficiency diseases are overprotective. Often, a child looks healthy to others, but the child's parents are aware that a simple cold can lead to other complications. Due to their keen awareness of their child's history, these parents are often in the physician's office before symptoms are apparent to others. As a parent, you know your child best of all and will often pick up the early signs of potential trouble. Another situation resulting from the misunderstanding of primary immunodeficiency diseases is the fear that a child with a primary immunodeficiency disease will spread illness to others when, in fact, the opposite is true. Families of a child with primary immunodeficiency may fear going to public

places or having their child attend school due to a perceived risk of illness exposure. It should be emphasized that most children with primary immunodeficiency diseases are able to attend school safely. In some very special instances, home schooling, homebound and even dual-enrollment options can be viable alternatives. Your child's physician can help in making this decision, but as a general rule, if your child has no restrictions on being in public spaces (i.e., movies, malls, airplanes), they may safely attend school. It is important to prepare yourself and your child for handling such misunderstandings. Planning what to say in a situation where someone expresses worry that your child will spread illness to others, for instance, can be beneficial and minimize the tension of the situation.

Hospitalizations

Everyone in the family is affected when a child is admitted to the hospital. Parents worry about the well-being of their child in the hospital, who will take care of siblings at home, and about missing work. The child in the hospital is likely to experience stress related to procedures, separation from family and friends, and/or disappointment related to missing out on regular activities such as field trips or other school events. Brothers and sisters may worry about the child in the hospital and about how their own lives will be affected (e.g. who will take care of them while their parents are at the hospital, how their lives will change). Siblings may also feel jealous or resentful of the attention that the hospitalized child receives.

Following are some relatively simple strategies that may help minimize the stressful effect of hospitalizations on the child and family:

For the hospitalized child, bring favorite items and activities such as stuffed animals, a special blanket, books, videos, toys or games from home. These all help make the hospital environment more familiar and comfortable. If the hospitalization will be longer than a few days, ask if pictures and get well cards can be taped to the walls. Become familiar with what procedures will be conducted and when you

may accompany your child. When possible, prepare your child for procedures or events by helping him or her know what to expect (different parts of a procedure, what it may feel like, sound like, look like) and by planning how to cope or get through it (utilize Child Life specialists, nurses, and procedure technicians). Maintain regular limits and routines.

For siblings, maintain routines as much as possible. Try to keep siblings at home if possible, rather than sending them to stay elsewhere; consider bringing alternative caregivers to stay with them in the home if necessary. Communicate honestly and openly about the situation and provide updates as needed. Support the continued connection between children at home and the hospitalized child through phone calls, notes or cards, and visits, if possible (check with your child's nurses about visiting policies). Maintain regular limits.

For parents, utilize the support and resources available in your community and at the hospital. Continue to take care of yourself, try to eat nourishing meals and get enough sleep. Be sure to take short breaks to get outside or at least out of the hospital room. This may help you gain energy and perspective at times.

A Child's Understanding of Illness

Very young children can sound like experts regarding their illness when they repeat the words and explanations they have heard adults use. However, the ability to repeat such statements does not indicate that children truly understand the meaning of the words they have just used. Asking a child, "What does that mean to you?" can help you evaluate his or her individual level of understanding. As children continue to grow and develop, they will need to revisit questions related to their primary immunodeficiency disease (e.g. "What is this illness?"; "How come I have it?"; "How did I get it?"; "What's the medicine for and why do I need it?"). Sometimes changes in your child's behavior can be a clue to initiate these conversations.

Preschool Child

The child may perceive treatment, procedures, or hospitalization as punishment because of their immature understanding. What these children need to know is that they did not cause the illness and that the treatments are not punishment—instead it is the best way the doctors and nurses know for helping them stay well or get better. If it's a particularly challenging treatment or procedure, it may help to say, "we wish there was an easier way, but this is the best way."

School-age Child

The child begins to develop an understanding of the interior body and an understanding of illness. This age-group benefits from employing their natural curiosity to facilitate understanding about the body systems and their specific symptoms and treatments. Books and/or videos, such as children's anatomy books (resources listed in the Resources section) and even science "experiments" can encourage more advanced discussion and understanding.

Learning from Your Children

Children are resilient. However, there may be times when you are not sure how well your child is coping. You may have noticed some changes in your child's behavior that occur more commonly than the occasional tough day that all children have.

Watch for continuing patterns of:

- eating or sleep disturbances
- changes in school performance
- an increase or appearance of fears
- changes in social behavior
- regression in developmental milestones
- withdrawing from others

These actions may alert you that your child needs some extra support. Often, talking and giving your child the opportunity to share concerns with you and together planning ways to cope in the future is all that's needed.

Other times, children and parents may benefit from additional support from extended family and friends, and caring adults in the child's school or community (such as guidance counselors, religious youth group leaders, and mental health providers).

How to Ask Questions That Get Children Talking

Open-ended Questions

“What kind of questions do you have?” is very different than “Do you have any questions?”

“What do you think will happen?”

“What do you think is the best (or worst) thing that could happen?”

“What are you wondering about?”

When You’re Concerned About A Specific Behavior

“I’ve noticed that you’re not eating much lately, and that’s not like you. I think there’s something on your mind.”

“Lately, you’ve been getting angry about things that don’t usually bother you. Why do you think that is?”

Multiple Choice

“I’ve read (or heard) that lots of kids whose brother or sister is in the hospital worry that...” then offer several likely possibilities (such as, it could happen to them, they won’t be able to do things their friends are doing like the school trip). Ask “What has this been like for you?”

Playing and Learning from Your Children

Pretend Play/Dramatic Play

By using dolls, animals, action figures, even cars and trucks, children play out their experiences. Adults can learn what is on children’s minds by watching and by participating. Play with cars can become play about the mommy car, the daddy car, the baby car, the big brother car. To learn the most from your children, guide the play gently—perhaps setting the characters (“You be the mommy and daddy dolls, and I’ll be the baby and sister dolls”) and setting the scene (“The mommy and baby dolls are at the hospital. What do you think is happening there?”). Using questions such as “What does he say?” or “What is she thinking/feeling now?” can further extend the play. Usually children take over the play and begin to direct all of the characters. If you sense that your child is reluctant or wants to do different play, give him or her the freedom and control to move on.

Drawing

Children often use drawing and other forms of art for emotional expression. Encouraging children to talk about their drawings or artwork can be eye-opening for adults. Open-ended questions such as, “What is happening in this picture?; What is this person thinking or feeling?” or “Tell me the story of this picture.” can help you learn about your child’s inner world.

Learn from your children by observing their behaviors and playing and talking with them. These connections will help you identify and change potentially problematic stress reactions before they interfere with your child’s normal activities.