



CHAPTER

17

## General Care

*This chapter provides information pertinent to the general care of the primary immunodeficient patient in the home, at school, at work, and at play. General measures, both those that maximize the body's resistance to infection and those that reduce the risk of acquiring an infection from the environment are included. In addition, specific illnesses are discussed in terms of their characteristic symptoms and supportive therapies.*

## General Health Measures

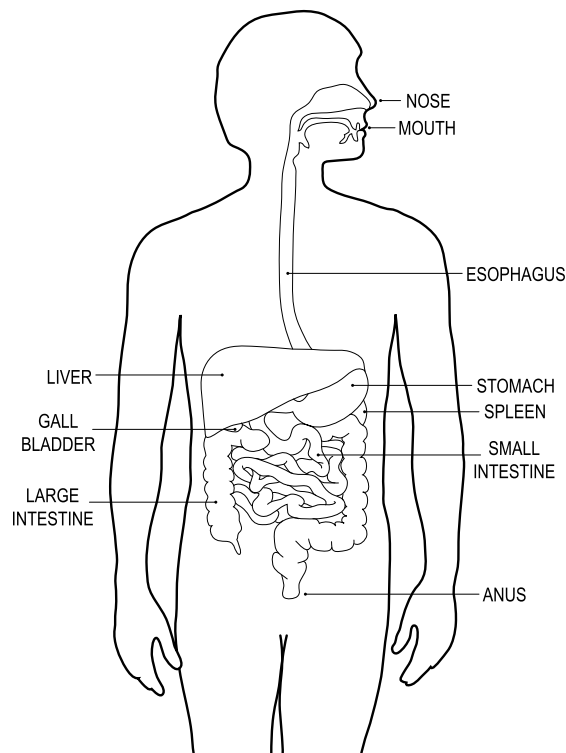
### Nutrition

An adequate diet provides nutrients essential for normal growth and development, body repair and maintenance. While good dietary habits are important for everyone, they are extremely important for the primary immunodeficient individual. Children, in particular, need a balanced diet to grow and develop normally. Dietary guidelines for Americans encourage eating a variety of foods, maintaining an ideal body weight, consuming adequate starch and fiber and limiting the intake of fat, cholesterol, sugar, salt and alcohol.

There are a number of “fad” diets and other diet recommendations that claim to boost immune resistance or help fight disease. It is very important to consider any unusual diet very carefully and to partner with your physician in this decision. The only truly proven dietary measure for increasing immunity is to have a balanced diet with adequate nutrition.

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### CHAPTER 17; FIGURE 1 Digestive System



### Special Medical Diets

In times of infection, illness, or food intolerance, the normal diet may need to be modified. Talk to your doctor to learn when these diets are indicated.

#### Clear Liquid Diet

A clear liquid diet may be used when there is a severe intolerance to food during infection or illness, or when nausea, vomiting and diarrhea are present. Since it is nutritiously inadequate, a clear liquid diet is usually only used for one to two days. The main purpose of a clear liquid diet is to replace lost fluids. Suggested fluids include: electrolyte replacement solutions such as Pedialyte™ and Gatorade™, fat-free broth, strained vegetable broth, strained citrus juices, plain Jell-O™, and fruit ices.

#### Full Liquid Diet

A full liquid diet may be ordered when there is difficulty in chewing or swallowing solid foods, as in pharyngitis, or when advancing from clear liquids. When properly planned, this diet can be nutritious and used for extended periods of time. A full liquid diet includes all foods that are liquid at room and body temperatures. Eggs (soft-cooked), strained meats, fruits and vegetables, ice cream, milkshakes and creamed soups are examples of food which can be added to the diet.

#### Soft Diet

A soft or bland diet is the transitional step between a liquid and a regular diet. Suggested foods include those that are easily chewed, swallowed and digested. Foods to be avoided include those that have high fiber content, are rich and highly flavored or are fried and greasy.

In some circumstances, if patients are not able to eat or drink normally, or if they can eat but are unable to absorb nutrients adequately from their stomach and intestines, there are procedures to assist them in maintaining adequate nutrition.

## Special Dietary Procedures

### Enteral Nutrition

Enteral nutrition is used when an individual with a normally functioning gastrointestinal system is unable, or refuses, to eat sufficient liquids to maintain hydration and sufficient foods to meet energy needs. Two common methods of providing enteral nutrition are with the use of a nasogastric tube or a gastrostomy tube. A nasogastric tube involves placement of a small flexible plastic tube through the nose, into the esophagus and then to the stomach. A prescribed amount of liquid feeding (containing essential nutrients) is administered through the tube continuously or at regular intervals. A gastrostomy tube involves the placement of a feeding tube into the stomach or small intestine.

### Total Parenteral Nutrition

Total parenteral nutrition (TPN) (or intravenous hyperalimentation) is a term used to describe methods of delivering all essential nutrients, fluids and calories directly into the blood stream. Total parenteral nutrition is used to maintain the nutritional status of an individual who is very ill, malnourished, or whose gastrointestinal function is inadequate. The TPN solution usually contains protein, carbohydrates, electrolytes, vitamins, water and trace minerals. Fats may be supplied in a separate solution. Various types of intravenous catheters are used to administer the solutions through a large vein. Nutrients are introduced directly into the blood stream, bypassing the stomach and intestinal tract.

## Nutritional Supplements

It is important to consider the large number of nutritional supplements that are available. These include a wide variety of vitamins, meal replacements, herbal remedies, botanicals, probiotics and naturopathic medicines. Many of these are marketed with claims to improve various aspects of your health. Since these items are not considered “drugs” by the United States Food and Drug Administration, the claims made by the companies that produce these items do not need to

be based upon scientific data as medicines must. For this reason, extreme caution is recommended when adding these types of treatments to your regimen. Importantly, some of these supplements can even be harmful or interact directly with prescription medicines you are taking. The use of certain supplements, however, is supported by scientific evidence under specific circumstances. You should feel comfortable in discussing the use of these treatments with your physician.

## Hygiene

General principles of good hygiene are essential for patients with immunodeficiency and their families. Hand washing before meals, after outings, and after using the toilet should become routine. It is essential to remember that to be truly effective, hands must be washed vigorously with soap and water for at least 15 seconds (try timing this sometime). When hands are not visually dirty, alcohol-based hand gels are an effective alternative. These have the advantage of being able to neutralize germs, are portable and can be applied rapidly. The regular use of hand gels has been shown to reduce the occurrence of colds in healthy people, and there is no reason to believe that this would not apply to immunodeficient patients as well. Individually wrapped and disposable hand

wipes are excellent for school lunches and for outings. For younger children, periodic washing of toys may be beneficial. Cuts and scrapes should be cleansed, and a first-aid cream applied.

Some individuals with a primary immunodeficiency are prone to tooth decay. Regular visits to the dentist and proper brushing and flossing are to be encouraged.

Individuals with a primary immunodeficiency should avoid exposure to people who are ill with an infection. During periods of influenza outbreaks, crowded areas such as shopping centers and movie theaters should be avoided by patients with severe combined immunodeficiency disease and patients who already have developed significant lung damage.

## Exercise

Participation in age appropriate physical activities should be encouraged. Hobbies and sports promote physical fitness and provide an excellent outlet for energy and stress. Swimming, biking, running and walking promote lung function, muscle development, strength and endurance. In general, people who are physically fit and participate in regular exercise get sick less than people who do not exercise. In some studies, people who exercise regularly were even found

to have stronger immune systems. Although this has not been directly tested in primary immunodeficiencies, exercise can be a fun, rewarding and useful part of your routine.

Some patients with a primary immunodeficiency may have problems controlling bleeding. In these cases, the types of exercises in which the patients can safely participate should be discussed with their physician.

## Sleep

Sleep is an essential requirement for good health. An appropriate amount of sleep each night that is consistent from day-to-day is highly recommended. Although there have not been specific studies of sleep habits in patients with primary immunodeficiency, erratic sleep has been shown to have negative effects on the immune system in other types of patients. For this reason, viewing sleep as a part of your therapeutic program is very important. Some helpful guidelines include: 1) trying to go to sleep and wake up at roughly the same time each day; 2) trying to avoid late nights; 3) avoiding consumption of caffeine (such as caffeinated coffee, sodas or tea) in the evening; 4) trying to minimize potential disturbances during the night; 5) avoiding long naps during the day that could interfere with your regular sleep schedule; and 6) planning your schedule around a night that will include an age-appropriate amount of sleep. Children aged three years and below also require naps during the day that should be considered an essential part of their sleep schedule (see table).

### Age Appropriate Nightly Sleep

(adapted from *Pediatrics*. 2003. vol 111, page 302-307)

Age	Average nighttime sleep duration (hours)	Average daytime sleep duration (hours)
6mos	11	3 1/2
1yr	12	2
2yr	11 1/2	2
3yr	11	2
4yr	11	1
6yr	11	0
8yr	10 1/2	0
10yr	10	0
13yr	9	0
16yr+	8	0

## Stress

The common notion that people get sick more often when they are under increased stress is supported by scientific data. Some studies also show that stress negatively affects the immune system. There are also scientific studies that show reducing stress can improve immune function. Since some of these measures can be low-risk, they may be worth considering. These include massage therapy, biofeedback, meditation and

hobbies. These interventions have advantages that may or may not apply to particular patients. You should partner with your physician or healthcare team in considering or choosing specific interventions (some may actually be covered by your medical insurance). However, regular exercise and sleep are perhaps the most important stress-reducing measures and should be taken seriously.

## General Care During Specific Illness

This section provides basic information about some of the illnesses patients may experience that may not require hospitalization. Medical terms often used in association with these illnesses are defined and their characteristic symptoms are described. General supportive measures designed to provide relief of symptoms and prevention of complications are also provided.

These illnesses are grouped according to the body “system” involved. These systems include the visual (eyes), the auditory (ears), the respiratory (nose, throat, lungs) and the gastrointestinal (stomach, intestines). It is important to stress the need for physician communication and supervision for an individual with a primary immunodeficiency during any illness. The frequency of even “minor” illnesses should be reported because they can influence the preventive therapy the physician feels is necessary (i.e. immunoglobulin, antibiotics).

The goals of medical treatment and supportive care of any primary immunodeficient individual are to reduce the frequency of infections, prevent complications and prevent an acute infection from becoming chronic.

The patient, family and physician must work together as a unit if these goals are to be accomplished.

### Visual System

#### Conjunctivitis

Conjunctivitis (pink eye) is an inflammation or infection of the lining of the eyelid and of the membrane covering the outer layer of the eyeball (conjunctiva). It can be caused by bacteria, viruses or chemical irritants such as smoke or soap. Conjunctivitis may occur by itself, or in association with other illnesses, such as the common cold. The symptoms commonly associated with conjunctivitis are redness and swelling of the eyelids, tearing, and discharge of pus. These symptoms are usually accompanied by itching, burning, and discomfort from light. In the morning, it is not unusual to find the eyelids “stuck” together from the discharge that has dried during the night. These secretions are best loosened by placing a wash cloth soaked in warm water on each eye. After a few minutes, gently clean each eye, working from the inner corner to the outer

corner of the eye. Meticulous hand washing is necessary for anyone coming in contact with the eye discharge in order to prevent the spread of the infection. It may be necessary to be seen by a physician to determine the type of conjunctivitis involved, and the type of treatment required.

### Auditory System

#### Otitis Media

Otitis Media is an infection of the middle ear and is usually caused by bacteria or viruses. A small tube called the Eustachian tube connects the middle ear with the back of the throat and nose. In the infant and small child, the tube is shorter and straighter than in the adult, providing a ready path for bacteria and viruses to gain entrance into the middle ear. In some infections and allergies, this tube may actually swell and close, preventing drainage from the middle ear. The characteristic symptom associated with otitis media is pain, caused by irritation of the nerve endings in the inflamed ear. A baby or young child may indicate pain by crying, head rolling, or pulling at the infected ear(s). The older child or adult may describe the pain as being sharp and piercing. Restlessness, irritability, fever, nausea, and vomiting may also be present. Pressure in the infected ear drum tends to increase when the individual is in a flat position. This explains why pain is often more severe at night, causing the individual to wake up frequently. As fluid pressure increases within the ear drum, pain becomes more severe and the ear drum may actually rupture. The appearance of pus or bloody drainage in the ear canal is an indication of a possible ear drum rupture. Although pain is usually relieved when the ear drum ruptures, the infection still exists. Whenever an ear infection is suspected, the patient should be seen by a physician. Antibiotic therapy is begun in order to stop the infection and prevent hearing impairment. Decongestants may also be prescribed to shrink mucous membranes, promoting better drainage from the middle ear. A follow-up examination may be performed in approximately ten days to be sure that the infection has cleared and that no residual fluid remains behind the ear drum.

## General Care During Specific Illness continued

### Respiratory System

The following respiratory illnesses will be discussed in terms of definitions and symptoms. Because the general care of the patient during these illnesses is similar, it will be handled as a single discussion at the end of the section.

#### Rhinitis

Rhinitis is a term used to describe an inflammation of the nose. It is usually caused by bacteria, viruses, chemical irritants and/or allergens. Symptoms may include sneezing, difficulty in breathing through the nose, and nasal discharge. The nasal discharge may vary from thin and watery, to thick and yellow or green.

#### Pharyngitis

Pharyngitis is a term used to describe an inflammation of the throat (sore throat). It is usually caused by a bacterial or viral infection. Symptoms include a raw or tickling sensation in the back of the throat and difficulty swallowing. Temperature may be normal or elevated. Sore throats caused by streptococcus (strep throat) can, in rare incidences, cause serious complications such as rheumatic fever. Whenever you or your child complains of a sore throat, your doctor should be contacted.

#### Acute Sinusitis

Sinusitis is a term used to describe an inflammation of one or more of the sinuses (see *Figure 2*). The sinuses are small cavities, lined with mucous membranes, located in the facial bones surrounding the nasal cavities. The purpose of the sinuses is thought to be to decrease the weight of the skull and to give resonance and timbre to the voice. The basic causes of sinusitis are the blockage of normal routes of sinus drainage and the spread of infections from the nasal passages. Headache, aching in the forehead and cheekbones and tenderness over the face in these same areas are characteristic symptoms. In addition, there may be pain in and around the eyes and in the teeth of the upper jaw. The pain and headache associated with sinusitis is typically more pronounced in the morning due to accumulated secretions in the sinuses during sleep. Being in an upright position during the day facilitates sinus drainage providing temporary relief. Depending on the amount of post-nasal drainage, cough, throat irritation, bad breath and decreased appetite may also be present.

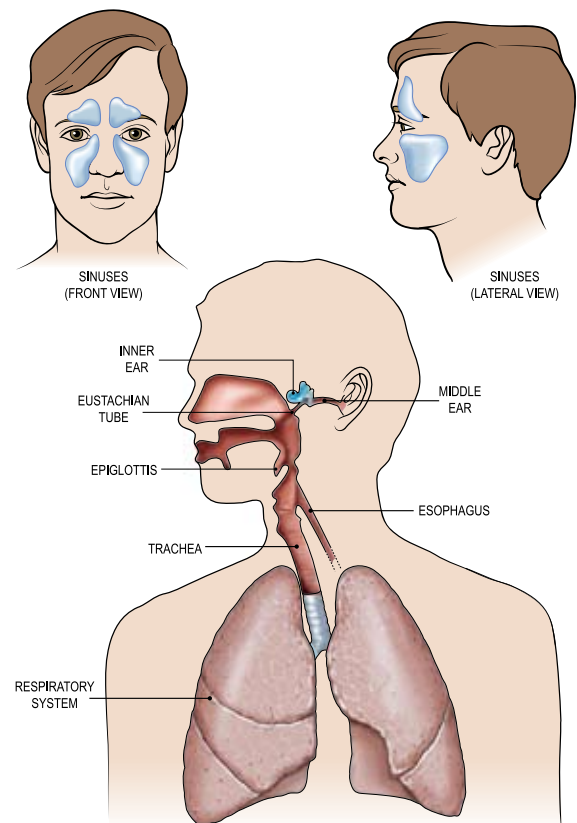
Temperature may be within normal limits or slightly elevated. Repeated or prolonged episodes of acute sinusitis may lead to chronic sinusitis.

#### Croup

Croup is a general term used to describe an infection in children which causes narrowing of the air passages leading to the lungs such as the larynx, trachea and bronchi. Croup can be caused by viruses or bacteria. The child's temperature may be normal or slightly elevated. The onset of croup may be sudden or occur gradually. In some instances, the onset occurs at night and the child may awaken with a tight "barking" cough. Breathing is difficult due to the narrowing of the trachea (windpipe). Croup can be a frightening experience for both the parents and child. Unfortunately, the child's anxiety may increase the severity of the symptoms. It is important for the parents to remain as calm and as reassuring as possible.

### CHAPTER 17; FIGURE 2

## Common Sites of Infection



## General Care During Specific Illness continued

Always notify your physician when you suspect your child has croup. Your physician may recommend that the child be seen immediately.

### Acute Coryza (Common Cold)

Acute coryza (common cold) is an acute inflammation of the upper respiratory tract (nose and throat or nasopharynx). Early symptoms include a dry tickling sensation in the throat, followed by sneezing, coughing and increased amounts of nasal discharge. There may also be symptoms of fatigue, chills, fever and general aches and discomfort.

### Influenza (Flu)

Influenza (flu) is a term used to describe a highly contagious respiratory infection which is caused by three closely related viruses. Influenza may occur sporadically or in epidemics. Usually epidemics occur every two to four years and develop rapidly because of the short incubation period. The incubation period includes the time a person is exposed to an infecting agent to the time symptoms of the illness appear. Symptoms of the flu include sudden onset of high fever; chills, headache, weakness, fatigue, rhinitis, and muscular soreness. Vomiting and diarrhea may also be present with one type of influenza.

### Acute Bronchitis

Acute Bronchitis is an inflammation of the bronchi (the major branches off the trachea or windpipe). It often accompanies or follows an upper respiratory tract infection, such as the common cold. Symptoms include fever and cough. At the onset, the cough is dry, but gradually becomes productive (producing mucus).

### Pneumonia

Pneumonia is an acute infection of the lungs and can be caused by bacteria, viruses, and fungi. Symptoms include chills, high fever, cough, and chest pain associated with breathing and coughing. In some cases nausea, vomiting, and diarrhea may also occur. Patients who develop pneumonia must be treated by a physician since permanent lung damage may develop if it is not treated aggressively.

### General Care of the Individual with Respiratory Illness

The treatment of respiratory infections is directed toward the relief of symptoms and the prevention of complications. Your doctor may prescribe

a medication to relieve fever and general body aches. Antibiotics may be prescribed to control infections of bacterial origin and/or to prevent complications. Expectorants may be prescribed to liquefy (water down) mucus secretions. Decongestants to shrink swollen mucous membranes may also be ordered. Fluids should be encouraged, and drinking a variety of beverages is important. Beverages served with crushed ice can be soothing to a sore throat. Warm beverages, such as tea, may promote nasal drainage and relieve chest tightness.

During the acute phase of any illness, there may be an initial loss of appetite. You or your child should not be forced to eat, and large meals should not be offered. It is often better to offer small frequent feedings of liquid and soft foods. Once the appetite returns, a high-caloric, high protein diet, to replace the proteins lost during the acute phase of the illness, should be offered (see section in this chapter titled *Nutrition*).

General comfort measures also include rinsing the mouth with plain water at regular intervals. This will relieve the dryness and "bad taste" that often accompanies illness and mouth breathing. A vaporizer is helpful in increasing room humidity. If you use a vaporizer; it must be kept clean, to prevent contamination with molds and bacteria. A petroleum jelly coating can provide relief and protection to irritated lips and nose. Body temperature fluctuations may be associated with periods of perspiration. Bed linens and clothing should be changed as often as necessary, and your child should be protected from drafts and chills. Finally, adequate rest is important. If persistent coughing or post nasal drip interferes with rest, elevation of the head and shoulders with extra pillows during periods of sleep should be attempted.

The individual should be encouraged to cover the mouth and nose when sneezing and coughing. Soiled tissues should be promptly discarded. Frequent hand washing is essential to prevent the spread of the infection. In some cases of bronchitis and pneumonia (depending on the age and level of understanding) coughing and breathing deeply at regular intervals should be encouraged. Coughing protects the lungs by removing mucus and foreign particles from the air passages. Deep breathing promotes full expansion of the lungs, reducing the risk of further complications. In some situations, a physician may order chest postural drainage, chest physiotherapy, or sinus postural drainage.

## General Care During Specific Illness continued

### Gastrointestinal System

#### Diarrhea

Diarrhea is characterized by frequent, loose, watery bowel movements (stools). Diarrhea may be caused by viral, bacterial, or parasitic infections. Certain medications may also cause diarrhea. Diarrhea may be mild to severe in nature. Whether it is mild or severe depends on the frequency of stools, their volume, how loose they are, the presence or absence of fever and how much fluid the child or adult can take and retain by mouth. The significance of diarrhea is related to the amount of body fluids lost and the severity of dehydration which develops. Infants and young children are at a greater risk for dehydration than older children and adults.

*Symptoms of dehydration include:*

- Poor skin turgor (loss of elasticity)
- Dry, parched lips, mouth and tongue
- Thirst
- Decreased urinary output
- In infants, depressed (sunken) fontanelles (soft spots) when the child is lying down
- A sunken appearance to the eyes
- Behavioral changes ranging from increased restlessness to extreme weakness.

The general care of diarrhea focuses on the replacement of lost body fluids and the prevention of dehydration. When diarrhea is mild, changes in the diet and increased fluid intake may compensate for fluid losses. The doctor may suggest giving the infant or young child clear liquids. If clear liquids

are tolerated (no vomiting) and the frequency and volume of stools decrease, you may be instructed to offer diluted formula or milk.

An infant may find comfort in a pacifier. Sucking may help relieve abdominal cramping. Burping is still necessary to expel any swallowed air. The older child and adult may be encouraged to drink fluids such as weak tea, Gatorade™, bouillon and “flattened” soft drinks. If nausea and vomiting are present, offer the older child and adult ice chips and popsicles. Fluids taken too quickly, or in too large of an amount, may precipitate vomiting. If these fluids are tolerated, gradually offer small sips of other fluids. Bland foods, such as rice cereal, yogurt, and low fat cottage cheese can slowly be added to the diet (see section titled *Nutrition*).

General comfort measures include coating the rectal area with a petroleum jelly preparation. This will help protect the skin and reduce irritation from frequent diarrheal stools. Soiled diapers and clothing should be changed immediately. The older child and adult may be encouraged to rinse his mouth with water regularly. This helps to relieve mouth dryness and “bad taste” associated with illness and is especially important after vomiting. In infectious diarrhea, several measures are used to reduce the chances of spreading the illness to other family members. Frequent hand washing is essential for everyone. It may be easier for the infected person to use disposable cups, dishes, and utensils. Soiled diapers, clothing and linens should be kept separate and washed separately from other family laundry. Bathrooms should be cleaned with a disinfectant solution as often as necessary.

### Use of the Internet

As many patients and their families have access to the internet, it is very important to carefully consider the source of any information available. Although there are many valuable resources on the internet, including nearly all of the scientific data that helps guide expert care of primary immunodeficient patients, there are also just as many personal opinions and unproven testimonials.

Talk to your physician and healthcare team partners about finding good sources of information on the internet and in interpreting other information you may have found.