

Surviving Neutropenia

Chemotherapy's potential to kill NB cells means that it also will have effects on the normal cells in your child's body. General background information about these effects and some strategies from NB parents for coping with them can be found in **Getting Through Chemotherapy**. One side effect of chemotherapy that requires special attention is "neutropenia." Additional information about this important aspect of treatment is provided below, as well as some basic tips that have been helpful to some NB parents. (Please use the e-mail link at the end of this section to send comments about coping tips that *you* have found helpful during your child's neutropenia.) The information provided is not in the nature of medical advice - you should always consult with your NB team before considering any suggestions from a parent or any source other than your medical team. Most hospitals will have specific guidelines for caring for a neutropenic child. Your child is in a potentially life-threatening situation during neutropenia, so it is crucial to understand and follow the medical team's guidelines.

What is Neutropenia?

Neutropenia is the medical term for having an abnormally low number of white blood cells called neutrophils. Neutrophils are the 'first responders' to a bacterial invasion, and surround and kill the bacteria. (See **"Understanding the CBC."**)

Neutropenia is dangerous because it makes the body more susceptible to bacterial infections. Children with NB often become neutropenic because neutrophils can be destroyed by chemotherapy and radiation. The children are typically at risk for neutropenia a week or two after completing a chemotherapy treatment. The timing will vary according to the specific chemotherapy and the particular child. But often neutropenia can be anticipated and parents should be on high alert at these times.

When neutropenia is expected, your child's temperature should be taken at least twice a day and whenever the child feels warm (tactile temperature). The temperature may be taken by mouth (orally), by ear (tympanic), or under the arm (works well on sleeping children), but be sure to record how the temperature was taken. Do not take a rectal temperature in a child undergoing treatment for cancer, as this can cause bleeding and infection.

How is the Neutropenia Diagnosis Made?

Neutropenia is diagnosed by a blood test that will determine the patient's "absolute neutrophil count" or ANC. (See **"Understanding the CBC."**) The severity of neutropenia generally depends on the ANC:

- mild neutropenia exists when the ANC falls between 1000 and 1500;
- moderate neutropenia exists when the ANC falls between 500 and 1000; and
- severe neutropenia exists when the ANC falls below 500.

When the child's neutrophils are at the lowest point, the child is said to have reached the "nadir" of neutropenia.

What are the Symptoms of Neutropenia?

At the onset of neutropenia, *the child may have no symptoms*. When a child is neutropenic and develops a fever of up to 100.4 degrees F, it is called “febrile” neutropenia. **This is potentially a life-threatening emergency.** Your child’s immune system does not have the white blood cells to fight off infection, especially with the added risk of a central venous line, and you need to contact your child’s oncology team immediately. Make sure you have your oncology team’s 24-hour contact information with you at all times. Bacteria can grow rapidly in the blood (sepsis) and can be rapidly fatal.

Some symptoms that can coincide with neutropenia include any type of infection, cough, sore throat, shaking and chills, mouth ulcers, and diarrhea. (See **“Getting Through Chemotherapy”**)

What are the Signs of Infection?

The body’s normal means of defense against infection is the immune system, comprised of the skin, the lining of the nose, mouth, gastrointestinal tract, and certain blood cells. When a patient has neutropenia, the body’s immune system is compromised, putting the patient at a higher risk of infection. Most infections occur in the lungs, mouth and throat, sinuses, and skin.

Because infection can be extremely dangerous to a neutropenic child, it is important to be aware of the following signs of infection and to notify your child’s oncologist if any of these symptoms appear:

- Tactile temperature
- Chills, shakes, sweating
- Fatigue
- Headache
- Body aches
- Feeling confused, dizzy or weak
- Sore throat
- Cough or shortness of breath
- Stuffy nose
- Burning with urination
- Redness, swelling, or warmth at the site of an injury, surgical wound, or central line
- Swallowing problems
- Mouth sores
- Sinus tenderness
- Pain in the abdomen
- Diarrhea
- Rectal discomfort with bowel movement

Remember, however, that some common signs of infection, such as inflammation (pus, swelling, and redness), may be absent during neutropenia because the neutrophil cells that cause these symptoms are absent.

When is Hospitalization Required?

The criteria for the hospitalization of a child with neutropenia may differ among medical institutions. However, the child with febrile neutropenia will be admitted. When your child is being considered for admission or admitted, expect blood samples to be taken for a complete blood count (CBC) and blood cultures (to determine if an infection is present). Broad-spectrum antibiotics will be administered and your child will have a minimum of once daily ‘counts’ (CBC) drawn. The results of the counts will be evaluated and treated as needed. The ANC will be monitored closely. (See **“Understanding the CBC.”**)

Visiting the Emergency Department

If your child runs a fever or has signs of infection during the evening or weekend when the oncology clinic is closed, a visit to the Emergency Room or Urgent Care will likely be necessary. It is important to call the hospital and ask for the “pediatric hematology/oncology fellow” on-call and if the doctor thinks you should bring in your child, the fellow will alert the ER to expect your arrival. When presenting to the ER, politely inform the triage nurse that your child has a fever and is neutropenic. Specifically ask that your child not wait in the general waiting room (to avoid exposure to other patients who are in the ER for communicable illnesses and because the treatment for febrile neutropenia is urgent).

In order to protect your immune-compromised child in the Emergency Department, have your child wear a mask and make sure any staff member who approaches your child washes their hands. Finally, it is prudent to bring a packed bag from home because such visits almost always result in hospitalization. It is best to bring along your child’s toys and special items for comfort and entertainment, but also to decrease the likelihood of your child contracting an infection from any toys and products that have been used by other children in the ER.

Discharge from the Hospital

There are no set criteria for a child to be released from the hospital following an admission for neutropenia. If no infection was found, the child may be released after a certain period of time without a fever and if evidence exists that the ANC is rising. (Remember, the ANC is typically checked at least daily during a febrile neutropenia admission.) However, if an infection or another complication was discovered, the child may have additional requirements to meet prior to being released.

One of the things most families experience in NB treatment is the feeling of being “trapped” in the inpatient ward of the cancer hospital after being hospitalized for neutropenia -- the child will start feeling better and better and pleading to go home, but his or her ANC will not yet be at the point required for discharge. One of the biggest frustrations during NB treatment is keeping your child engaged and happy during such hospitalizations! This is the time when the resources of the hospital’s child-life program, those gifts of board games, crafts, and books, and if permitted, visits from friends and family members, provide a lifeline to the harried NB parent. Now is the time to reach out to those friends who are eager to help and don’t know what to do. A well-timed visit and a gift of a set of Uno cards can work miracles!

Treatment of Neutropenia

As mentioned above, once your child is admitted to the hospital during neutropenia, cultures will be performed on central venous access lines or ports and peripheral blood, and broad-spectrum antibiotics will be administered. Several antibiotics are given, because upon admission to the hospital it will not yet be clear if your child has a specific infection; by administering broad-spectrum antibiotics, the hope is to start treating any such infection that may exist. Blood cultures will be started immediately, and once any actual infection is identified the appropriate specific medication for the particular infection will be administered.

As a precautionary measure, many children with neuroblastoma take a specific antibiotic, sold under the brand names Bactrim or Septra among others, two to three days each week to prevent “pneumocystis carinii” pneumonia (PCP). (See **“Getting Through Chemotherapy Treatments.”**) Alternatively, the child may receive a periodic IV injection of a specific antibiotic at the hospital. If you are administering such an antibiotic to your child at home and another antibiotic is prescribed

for your child to treat a suspected infection during a neutropenic phase, DO NOT discontinue the Bactrim or Septra unless your child's primary oncologist orders this. The other antibiotics may not protect the patient from PCP.

To stimulate a child's immune system during neutropenia, generally the child will be treated by the administration of a "growth factor" into the subcutaneous tissue. These growth factors are special proteins that naturally occur and stimulate the bone marrow to produce more white blood cells. "Granulocyte-Colony Stimulating Factor" (G-CSF) is a man-made product that stimulates the production of neutrophils and also enhances the activity of mature neutrophils.

G-CSF is usually administered by subcutaneous injection (an injection just under the skin). Cancer centers have varying schedules for these injections, which are commonly done by parents. If you are expected to administer G-CSF, your child's oncology nurse will give you all the necessary information and training. The recommended sites include the abdomen below the navel, upper outer arms, and upper outer thighs. Rotation of the sites is often recommended to prevent scarring and discomfort. The injection is not usually painful, but occasionally the preservative may sting.

Some families choose to have G-CSF injections accomplished through the use of a device called an Insuflon. The Insuflon is a soft plastic catheter that is inserted just under the child's skin. It allows several injections through the same injection port. The theory is that since it stays inserted under the patient's skin, it will alleviate the needle pain associated with injections. This technique works well to diminish the fear associated with daily injections in some children. Other children, however, find that having the Insuflon inserted is more stressful than the shots themselves. Also, much of the pain associated with the G-CSF injection is due to the medicine itself, and the Insuflon does not alleviate this.

If the daily injection of G-CSF is a stressful experience for your child, try experimenting with different techniques in an attempt to make the shot less painful. Some children feel that having EMLA cream or an ice cube placed on the injection site lessens the pain from the injections. Some children prefer having the G-CSF injected literally as fast as possible; others prefer it to be injected very slowly. Other coping tactics include distracting the child with television or books during the injection, and having the child participate as much as possible in the injection process. Some parents put together a toy box with a prize given to the child after each shot. The child's anticipation of the shot can be the worse than the actual experience (allowing the child to create and control the "rituals" to be done before administering the shot helps in some cases, but can also backfire). Occasionally, parents give the shot when the child is asleep. Some children actually prefer this method. (Just remember to hold the child securely at all times during the injection, whether given awake or asleep.)

If the child's central venous line or port is accessed for any reason at the time when G-CSF is being administered, the oncologists at some institutions may permit G-CSF to be given through the line. There is some data that suggests that giving the injection under the skin is more effective, but the child's discomfort with the shot may outweigh this concern. Of course, the risk of infection of the central line should always be weighed against the child's aversion to the injection.

Common side effects of G-CSF are bone, joint, or muscle pain (including muscle cramping) and injection site reactions. Tylenol and antihistamines such as Benadryl or Vistaril will generally alleviate some of these symptoms. Your child's oncology team will discuss any potential side effects and their appropriate treatments.

Neulasta

Neulasta is similar to G-CSF in a long-acting form and boosts white blood cells. NB patients usually use Neulasta less often than G-CSF because it is generally only administered to adults and, therefore, it is often difficult to obtain insurance coverage it. Those patients who have successfully

received Neulasta have been required to order an adult dosage from which they have to prorate their child's correct dosage amount. This may or may not necessitate the administration of the shot by the oncology clinic. The benefit of Neulasta is that it is given in a single shot 24 to 48 hours after the cessation of chemotherapy. This is obviously a significant quality of life issue for many children.

Avoidance Tactics and Safety Precautions while Your Child is Neutropenic

There is nothing that a neuroblastoma patient can do to avoid neutropenia. The child and caregivers can, however, take measures to avoid infection. The most important thing a family can do is –WASH THEIR HANDS! A thorough 10-second hand wash can significantly help prevent spread of infection. (If you don't want to count to ten as you wash your hands, try singing a simple song like Happy Birthday!) Some families install hand sanitizers in conspicuous locations throughout the house to remind the child, family members, and visitors that clean hands are essential to avoiding infection.

Other basic measures to help your child avoid an infection are making sure your child gets enough rest, eats and drinks adequately, and avoids ill people. Remember -- these basic safeguards apply to all members of the family!

Depending on where your child is in treatment, your child's oncology team may or may not want your child to be immunized against influenza. If they do, the flu vaccine would be administered as a shot and not as a nasal mist. Every member of the child's household and anyone else in frequent contact with the child should be encouraged to get a flu shot. However, be aware that your child should not be near another person who received the nasal flu mist vaccine or any live virus vaccine.

Other measures that can be taken in order to try to avoid infection are:

- avoid large crowds and ill people, especially those who have chicken pox, measles, shingles, or the flu;
- avoid day care;
- avoid hot tubs and hot showers as this dries the skin;
- wear shoes at all times;
- avoid using razors or handling sharp objects;
- clean any cuts immediately;
- practice good oral hygiene with a soft tooth brush;
- avoid fresh flowers or plants;
- avoid cleaning up after pets;
- avoid swimming in ponds, lakes, or rivers;
- avoid sharing glasses, utensils, or towels with other people;
- avoid constipation;
- apply sunscreen and avoid sunburn;
- keep the house, especially the patient's room, very clean;
- minimize stuffed animals in the patient's room;
- teens with cancer who are menstruating should avoid tampons;
- have the child follow any dietary restrictions or guidelines recommended by your oncology team; common neutropenic dietary guidelines are to avoid raw foods including raw fruits and vegetables; cook and clean foods thoroughly; avoid unpasteurized food or liquids; and eat no leftovers or food that the parents do not prepare (unless the child is hospitalized).

Some parents have their neutropenic child wear a face mask when being in contact with the general public cannot be avoided, on public transportation, for example.

Indeed, the possibility of infection should be considered whenever the child comes into contact with

the general public - at home, at school, and any public places – whether or not the child is neutropenic! Naturally, you want your child to enjoy life to the fullest even though he or she is in cancer treatment, but avoiding unnecessary illnesses is always a concern to be considered for a child in NB treatment, and especially during neutropenia. Some decisions about safety precautions are very personal, and each family should discuss the necessary safety precautions for their child's particular situation with their oncology team.

Please contact info@cncfhope.org with any comments