

## Severe Chronic Neutropenia International Registry

Neutropenia is a condition that happens when the level of neutrophils in the blood drop below 1000/cmm (per cubic millimeter). Neutrophils are blood cells that are produced in the marrow, or core of the bones. The blood normally contains 2500 to 9900 neutrophils/cmm.

Neutrophils are very important because they fight infection. When bacteria invade the body a chemical signal is sent out and the neutrophils, like fire fighters responding to a blaze, rush to the site of infection. The bone marrow also responds by speeding up its production of neutrophils to replace those involved in fighting the infection. If, however, production of new neutrophils is suppressed or slowed down, a shortage may develop, and any infection can overwhelm the few neutrophils available. Therefore, a person with only a few neutrophils is at particular risk for developing a serious bacterial infection.

Severe chronic neutropenia is the condition where the bone marrow doesn't produce neutrophils in the blood above the 500/cmm level (on a continuing or recurrent basis, or in cycles, lasting for months or years), resulting in not enough neutrophils to fight infection. We believe there are three main groups; congenital (from birth the bone marrow produces almost no neutrophils), idiopathic (occurring some time in life but why is not known, autoimmune neutropenia is in this group), and cyclic (from birth or occurring very early in life the bone marrow starts and stops producing neutrophils in a 21 day cycle; infections, mouth ulcers, and or skin infections occur when a person is at the low point in their cycle). Infections are a serious problem for all patients with neutropenia. Neupogen is a medication that helps the bone marrow to work better, helping the marrow to produce neutrophils. Once on Neupogen congenital patients and idiopathic patients neutrophil counts generally rise into the normal range. On Neupogen cyclic patients neutrophil counts still cycle, however, the goal of treatment is to raise the lowest absolute neutrophil count (ANC) in the cycle so that the person does not develop an infection. The most common side effect with Neupogen is bone pain when the person starts Neupogen. The bone pain generally resolves within the first two months.

There is no special diet or regime that will help neutropenia. After starting Neupogen, generally neutropenic people who have been very ill or limited in their activities find they can return to activities or do things they have not done before. If someone is always tired generally that improves also because the person is not always fighting infection.

Cyclic neutropenia is one of the rarest forms of neutropenia. Very few people or doctors have much experience with it. People with cyclic neutropenia mostly have mouth sores. They might look like canker sores, but if you were to compare them to people who do not have neutropenia the mouth sores look different. Generally the mouth sores come routinely with low cycles, they tend to get big (½ inch), are deep, and have a crust that falls off. Most cases of cyclic neutropenia, especially the most severe cases, can be helped with Neupogen by raising the ANC thus reducing the illnesses that occur during the low point of the cycle.

If you would like some articles about neutropenia please send your postal address. The information will be mailed to you.

### Mission of the Registry

This international disease registry was established in March, 1994, and is directed by an advisory board of physicians who treat SCN patients and is supported by Amgen, Inc. The mission of this registry is to establish a worldwide database of treatment and disease related outcomes for persons diagnosed with SCN. The information collected will lead to improved medical care and became a focus for future research.

### To Register

Preliminary telephone screening will indicate if the patient meets the eligibility criteria; however, a final determination will be made only after an Advisory Board physician has reviewed the completed registration packet.

## Neutropenia Registries

### United States:

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Severe Chronic Neutropenia International Registry (SCNIR)  
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Fax (206) 543-3668 Web <http://depts.washington.edu/registry/>  
(We cannot guarantee the confidentiality of any information sent by e-mail)

### Europe:

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Feodor-Lynen-Str.27  
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Germany

Phone 49 511/557105  
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### Australia:

Arlene Ewan, Data Manager  
University of Ballarat, Cancer Research Centre  
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1002 Mair ST  
Ballarat, Victoria  
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Phone 61-03-53-33-4811  
Fax 61-03-53-33-4813

## **Steps to Take to Reduce Bacteria Exposure General Information for Families**

### **Diet**

- ◆ Wash all fruits and vegetables with a fruit and vegetable soap to remove bacteria. A good fruit and vegetable wash will remove fungicides, herbicides, pesticides, chemicals, waxes, dirt and oil.
- ◆ All processed foods, such as hot dogs, need to be boiled for 5 minutes to eliminate bacteria.
- ◆ All poultry needs to be cooked to appropriate temperature to eliminate bacteria.
- ◆ All non-cooked foods, such as sushi, needs to be handled in a very clean environment. Make sure everyone handling food is wearing plastic gloves.
- ◆ All purchased juices need to be pasteurized - check the label.
- ◆ Ground meat, turkey, chicken, and beef need to be cooked to the appropriate temperature to eliminate bacteria.
- ◆ Utensils, straws, and cups should not be used by more than one person. Sharing will increase the likelihood of bacteria or viruses being transferred.

### **Hand Washing Technique**

- ◆ Lather the soap up between your fingers as well as the palms and backs of your hands.
- ◆ Wash hands for 20 seconds or more, until they are well covered with lather. Rinse.
- ◆ Dry your hands before turning off the water, then use paper towel to turn off water.
- ◆ In public bathrooms door handles may have bacteria, so use the paper towel to open the door.

### **Travel**

- ◆ Be prepared with a mask in case someone around you is coughing.
- ◆ Carry a small bottle of anti-bacterial hand cleaner, such as Purell.
- ◆ Carry or buy bottled water.
- ◆ Eat cooked food, and avoid fresh food in exotic places.

### **Traveling with Medication That Needs to be Kept Cold**

- ◆ Carry medication in small cooler bag.
- ◆ Insulate the medication from the ice.
- ◆ Carry the prescription label from the pharmacy for customs.
- ◆ Carry twice as many vials, syringes and/or supplies needed for the number of days traveling.
- ◆ Carry all the supplies with the medication and prescription label.
- ◆ Carry a small needle disposal unit. A small plastic bottle works well.
- ◆ Check with customs on current rules and regulations regarding taking medication out of the country.

### **Injecting Subcutaneous Medication**

- ◆ Check with your doctor regarding the best method to inject (administer) the medication.
- ◆ Some subcutaneous medications can be given into the fat of the abdomen, the leg, or the arm.
- ◆ When injecting, gently squeeze the skin together, touch the needle to the skin and then gently insert the needle. You/your child are not a dart board.
- ◆ When starting daily injections keep a record of where you have injected; the record can be discontinued after you and your physician are comfortable with your routine.
- ◆ To administer into the abdomen use the clock method. Visualize a clock around your navel. Day one, give at 12 o'clock; day two, give at 1 o'clock, etc. If injecting into the leg vary the sites in a routine manner.

## Infants

- ◆ Receiving blankets are excellent for creating a barrier between a young infant and the airborne bacteria and viruses.
- ◆ The baby should be wrapped in two blankets; the inner blanket is considered the clean blanket, the outer blanket is the barrier.
- ◆ The outer blanket can be changed easily if there is an exposure.
- ◆ Carry several clean blankets; this will allow you to change the outer blanket anytime you feel it is no longer clean. The outer blankets are then cleaned normally in the laundry and are ready to be reused.
- ◆ Bath time is an excellent time for a skin and nail check. Look for nails that need to be trimmed to prevent infection from scratches. Check for scratches, cuts, inflammation or infection. Check under arms, on hands, face, knees. Boys: check penis, scrotum, and rectal area. Girls: check labia and rectal area.
- ◆ In the case of persistent diaper rash it may be necessary to see your doctor or a dermatologist.

## Young Children

- ◆ Teach children good hand washing habits early.
- ◆ Keep toys and play areas clean. Toys and hands are always going into a young child's mouth.
- ◆ When out of the house keep baby wipes available for quick clean up of your child or the environment, ie. grocery store carts.
- ◆ Strangers love to touch and admire children, but limiting this contact reduces exposure to bacteria or viruses.

## Dental Care

- ◆ See a dentist 2-4 times a year for teeth cleaning. This reduces the plaque build up on teeth.
- ◆ An electric tooth brush and a Sonicare will reduce plaque on teeth.
- ◆ Brush after each meal. When unable to brush, rinse particulate matter from mouth after eating.
- ◆ Mouth wash will reduce particulate matter and bacteria in the mouth. Commercial mouth washes work well. Use non-irritating neutral washes, or Hydrogen Peroxide. The bottle will have instructions for a ½ water and ½ Hydrogen Peroxide mixture.
- ◆ Fluoride rinse will help prevent tooth decay. It is also has an anti-bacterial action and will reduce the bacteria count in the mouth.
- ◆ Flossing daily will reduce the plaque build up on teeth. The "glide" or "slide" type floss - Johnson and Johnson, Reach Easy Slide, Ultra Shred-Resistant Floss - will be easier to use. They slide more easily across the tooth surface, and are less damaging to your gums because less effort is needed to floss, hopefully reducing gum damage.
- ◆ Orthodontic Care.
  - Children are starting at a younger age with orthodontic care, many orthodontists are using palate expanders, i.e. Hyrax, rapid palatal, or maxillary expanders; and mandible expanders, i.e. Schwarz.
  - All appliances should have every edge checked for roughness, and be double checked for exposed wires to make sure that no wires are sticking out.
  - Expanders are a good place for bacteria and thrush to live, so they need good cleaning at least twice a day.
  - The mandible expander is made to conform to the tissue of the mouth at the beginning of the process. As it is expanded, edges may become exposed that need to be smoothed or rounded. Mouth sores will develop quickly when edges become rough. Talk with your orthodontist about the best method to prevent and reduce the rough edges. This will prevent or reduce mouth sores.
  - Appointments should be scheduled in the first few days of the week, avoiding Friday or pre-holiday appointments, so that if a problem occurs after the appointment you will be able to see the orthodontist the next day to repair or smooth the appliance. If you have to wait over a weekend or holiday with mouth sores because of a rough wire, the orthodontic experience will be more uncomfortable.

## Pain Relief for Mouth Ulcers

(Consult your doctor before administering these medications to yourself or your child)

- ◆ Ulcer Ease (or similar product). Follow directions listed on the product.
- ◆ Maalox (one part), Benadryl (one part), Viscaine Xylocaine (one part). Rinse mouth and spit out solution.

## Skin Care

- ◆ Develop frequent and good hand washing habits early.
- ◆ Practice good nail care to prevent infections around the nail bed.
- ◆ Clean and treat cuts and scrapes with an antibacterial agent, such as betadine or hydrogen peroxide, then apply an antibacterial ointment, such as Neosporin, and cover with a Band-Aid promptly.

## Complete Blood Counts (CBCs)

- ◆ CBCs should be drawn just prior to the next shot or at least 16 hours from the last injection of Neupogen®. Using this method the CBC will show the lowest absolute neutrophil count the patient is experiencing.

Daily Injections	Every Other Day Injections	Three Times a Week Injections	Twice a Week Injections
Just prior to next injection or 16 hours after last injection	Just prior to next injection or more than 36 hours after last injection	Just prior to next injection or more than 2 days since last injection	Just prior to next injection or more than three days since last injection

- ◆ Whenever possible ask for a Manual Differential with the CBC.

## Sports and School Kits

### Sports Kit

Item	Purpose
Bottled water	clean cut or scrape
Betadine or hydrogen peroxide	clean cut or scrape
Band-aids (different sizes)	cover cut or scrape
Cold pack (that you can activate)	reduce swelling
Emergency numbers	to contact parents in case of serious injury
Sun screen	prevent sunburn
Blister pads	protect skin
Neosporin	antibacterial ointment for cut or scrape
Consent for emergency treatment	
Insurance information	

### School Kit

#### School

Emergency numbers to contact parents  
 Allergy list (if any)  
 Physician and telephone number  
 Current medication list  
 Consent for emergency treatment  
 Insurance information

### Classroom

Item	Purpose
Tooth brush / floss	remove food from under gums (popcorn)
Betadine or hydrogen peroxide	clean cut or scrape
Band-aids (different sizes)	cover cut or scrape
Cold pack (that you can activate)	reduce swelling
Sun screen	prevent sunburn
Blister pads	protect skin
Neosporin	antibacterial ointment for cut or scrape

## Effective Communication Between Parents and Professionals Managing the Medical Maze

Individuals, families and parents know themselves or their children better than anyone. Trust your feelings, enlist the help of the medical community and look for the medical health professionals who are interested in SCN.

Health care is two way communication between the patient/parent and the health care provider.

- ◆ Keep records
- ◆ Ask questions
- ◆ Be a broken record ( if you are not understanding or not understood, keep trying)
- ◆ Be the squeaky wheel
- ◆ Be self-reliant (take charge of your health care)
- ◆ Be your child's / your advocate
- ◆ Enlist your health care provider's help
- ◆ Be your doctor's partner in your health care (be knowledgeable, work together)
- ◆ Be motivated, the process may be a long one
- ◆ Trust your feelings

## General Information

Benign Neutropenia	Low WBC, low ANC, but patient stays well and is able to fight infection.
White Blood Count (WBC)	Complete white count; mature and immature cells.
Absolute Neutrophil Count (ANC)	Mature white cells; cells able to kill bacteria: This calculated value is necessary to reliably determine the body's ability to fight infections. Generally an ANC above 500 allows a person to deal with infection and stay well.
When ill	Blood work should include: WBC with manual differential (This allows the doctor and patient to know if ANC is above 500.)
How ANC is Calculated	WBC multiplied by the sum of the total number of segmented neutrophils* and segmented bands* (percentage total) multiplied by 10. *Segmented neutrophils also known as neutrophils or polys; segmented bands also known as bands or stabs.

WBC x total neutrophils (seg neutrophils% + seg bands%) x 10 = ANC

Example

WBC	5.0	WBC x (neuts + bands) x 10 = ANC
Seg neuts	3%	5.0 x (3+1) x 10 = 200
Seg bands	1%	5.0 x 4 x 10 = 200
		ANC = 200

Example Blood Report

		Manual Differential	
WBC	5.1	Segmented Neutrophils	5
RBC	4.00	Segmented Bands	5
Hemoglobin (Hgb)	11.0	Lymph	63
Hematocrit (Hct)	32.1	Monocytes	10
MCV	80.3	Eosinophils	16
MCH	27.4	Basophils	1
MCHC	34.1		
RDW	13.6		
Platelet (plts)	233		

WBC x (neuts + bands) x 10 = ANC  
 5.1 x (5+5) x 10= ANC  
 5.1 x 10 x 10= ANC  
 510 = ANC

## First Doctor Visit

Checklist

A 3 ring binder works well to hold and organize information

- ◆ Referral letter from general practitioner or prior health care professional
- ◆ Medical history of patient and family
- ◆ Past medication history
- ◆ Reports from past bone marrow and cytogenetic evaluations
- ◆ Other significant evaluations: e.g. MRIs, CT Scans, Bone Density Evaluations
- ◆ Current medication
- ◆ Allergies (medication, food, tape, other)
- ◆ Question list
- ◆ Insurance information



**Doctor Visit**

Suggestions for questions

- ◆ Labs – obtain results and ask about implications
- ◆ Medications – should there be any changes
- ◆ Precautions to take
- ◆ Medical status – any changes
- ◆ Treatments – anything new

Questions for the Health Care Professional	Parents Notes
1.	
2.	
3.	
4.	
5.	

To Whom it May Concern:

I am writing in order to clarify any confusion about the health of one of your students, \_\_\_\_\_  
\_\_\_\_\_. This child has severe chronic neutropenia. This is a non-contagious blood disorder. There are three types of neutropenia, congenital, cyclic, and idiopathic. Congenital and cyclic neutropenia can only be passed from the parent's DNA to the child's DNA at conception (people inherit congenital and cyclic neutropenia the same way that they inherit red hair or blue eyes, it is part of their genetic make-up). Idiopathic neutropenia is not inherited and develops in children in a variety of ways, but it is still not contagious. There is no risk of any teachers or classmates contracting this disease. Neutropenia is treated through regular injections of Neupogen (in the same way that diabetes is treated by regular injections of insulin). Although regular treatment with Neupogen allows the neutropenic individual to live a relatively normal life, it is important that adults working with neutropenic children are aware of these children's special needs.

These children are at a higher risk of infections, therefore adults need to watch for cuts and scrapes. It is important that all cuts and scrapes be cleaned and treated with an antibacterial agent such as betadine or hydrogen peroxide, antibacterial ointment applied (Neosporin) and covered with a Band-Aid promptly. In case of high fevers or sudden fatigue, parents should be notified immediately.

Neutropenic children treated with Neupogen may experience bone pain. The bone pain the child experiences is normal. The pain is generally located in the long bones, pelvis and / or forehead. Once Neupogen is injected and starts working the marrow expands causing pressure within the bone, creating bone pain. Children are not good at describing the pain, and behavioral difficulties that result are best dealt with kind and firm behavior management. These children truly hurt. It is our hope that the pain will diminish as the child becomes more stable on Neupogen. The most severe bone pain has been described by adults as "having my leg broken over and over". Adults take Ibuprofen or narcotics for the pain. It is our hope that children will experience mild bone pain and will only need Ibuprofen or Acetaminophen to receive relief.

If you have any questions about this disorder, please feel free to contact the Severe Chronic Neutropenia International Registry at 1-800-726-4463.

Sincerely,

Audrey Anna Bolyard, R.N., B.S.  
Clinical Manager

To Whom it May Concern:

I am writing to ask for your help with \_\_\_\_\_ who has severe chronic neutropenia, which is a non-contagious genetic blood disorder. There are three types of neutropenia, congenital, cyclic, and idiopathic. In congenital and cyclic patients it can only be passed from the parent's DNA to the child's DNA at conception (it is inherited in the same way that they inherit red hair or blue eyes - it is part of their genetic make-up). Idiopathic neutropenia is not inherited and develops in children in a variety of ways, but it is still not contagious. There is no risk of any coaches or teammates contracting this disease. Neutropenia is treated through regular injections of Neupogen (in the same way that diabetes is treated by regular injections of insulin). Regular treatment with Neupogen allows the neutropenic individual to live a relatively normal life. It is important that adults working with neutropenic children are aware of these children's special needs.

These children are at a higher risk of infections, therefore adults need to watch for cuts and scrapes. It is important that all cuts and scrapes be cleaned and treated with an antibacterial agent such as betadine or hydrogen peroxide, antibacterial ointment applied (Neosporin) and covered with a Band-Aid promptly. As the child's coach, if you would clean scrapes and cuts at the time of the injury this would prevent infection and allow the child to get right back into the game. In case of high fevers or sudden fatigue, parents should be notified immediately.

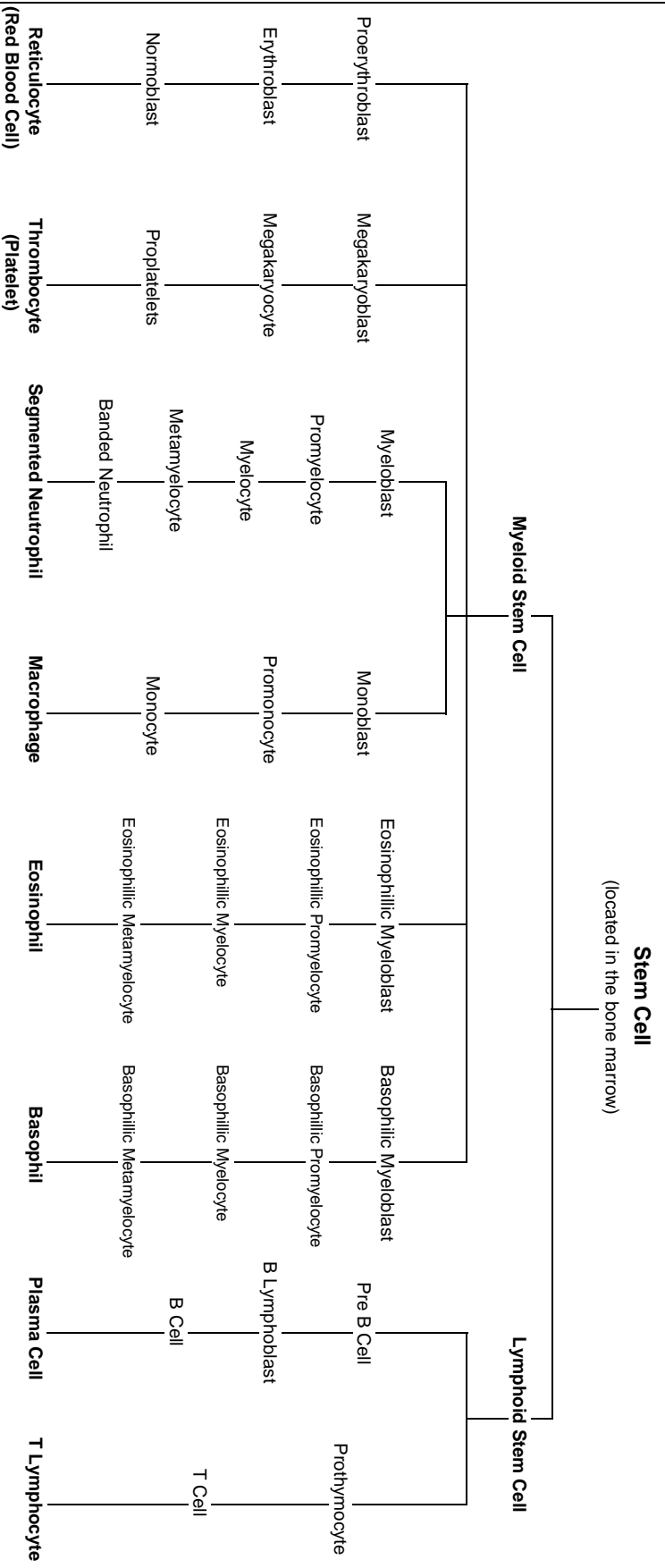
Thank you for your help. If you have any questions about this disorder, please feel free to contact the Severe Chronic Neutropenia International Registry at 1-800-726-4463.

Sincerely,

Audrey Anna Bolyard, R.N., B.S.  
Clinical Manager

## Hematopoiesis in the Bone Marrow

Hematopoiesis is the formation and development of blood cells



**Neutrophils, Eosinophils, and Basophils are also known as granulocytes.**