

A Case of Mistaken Identity? Another Disease That May Mimic SCN

Editor's note: The following information may be of benefit to the older idiopathic SCN group. Please note that this is only basic information. Please consult carefully with your physician.

Large granular lymphocyte (LGL) leukemia (also referred to LGL lymphocytosis) is a clonal disorder of LGL associated with autoimmune diseases. The disease should be suspected in patients with cytopenia, lymphocytosis, or autoimmune diseases, particularly rheumatoid arthritis.

The hallmark of LGL leukemia is a high lymphocyte count and a low neutrophil count. People with LGL leukemia can report having no symptoms or many symptoms. Symptoms can include night sweats, weakness, dizziness, frequent fevers, fatigue, anemia, an enlarged spleen, and other varying symptoms.

In normal blood, 10 percent to 15 percent of the lymphocytes are LGLs. LGLs have a characteristic appearance: they are larger than normal lymphocytes and contain pink granules. LGLs may either be a T-cell or NK-cell type of lymphocyte. LGLs are part of the normal immune system and are killer cells, which fight viruses. LGL leukemia occurs when there are too many LGLs and not enough of the other types of blood cells, especially the neutrophils. In some cases, the total number of lymphocytes is not greater than normal; however, the proportion or percentage of LGLs in your lymphocytes is higher than normal.

LGL leukemia is divided into two categories depending on which type of LGL is increased in the blood. They include T-cell LGL leukemia and NK-cell LGL leukemia.

T-cell LGL leukemia is a chronic or slowly progressing disease. It usually occurs in people between the ages of 50 and 60, and about the

same number of men and women get T-cell LGL leukemia. Half of these patients have anemia, half have an enlarged spleen, and bacterial infections and neutropenia are common. Rheumatoid arthritis also is common in this patient group.

NK-cell LGL leukemia has two forms acute and chronic. For purposes of this newsletter, we will only discuss the chronic form. The clinical features of chronic NK LGL leukemia are very similar to those seen in T-LGL leukemia. It is a chronic illness and is different than the acute form of NK LGL leukemia. The chronic form does NOT change to the acute form.

LGL leukemia can be diagnosed by employing several different clinical tests. Tests can include a CBC, a flow cytometry with LGL Panel, a TCR (T-cell receptor gene rearrangement study), a bone marrow biopsy, and a spleen analysis. The cause of LGL leukemia is not currently known.

The experts at the Large Granular Lymphocyte Leukemia Registry, a national registry that collects, manages, and analyzes information on people with LGL leukemia, are available for consult. The registry is part of the Hematologic Malignancies program at the Penn State Cancer Institute located at the Penn State Milton S. Hershey Medical Center in Hershey, Penn. The Registry is directed by Thomas P. Loughran, Jr., MD, and is coordinated by Lynn Ruiz.

For more information on LGL or the LGL Registry, please contact Lynn F. Ruiz, BS, CPL, research studies coordinator and LGL Registrar at Penn State Cancer Institute, at (717) 531-7377 or lruiz@psu.edu.

Source: Loughran, TP. Overview of the LGL Leukemia Registry. Penn State Cancer Institute.

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National Neutropenia

N E T W O R K

Annual Conference to Feature Four SCN Experts!

2007 agenda expected to be the best yet ...

The National Neutropenia Network (NNN) and the Severe Chronic Neutropenia International Registry (SCNIR) will host the 7th Annual Neutropenia Family Conference July 6-8, in Ann Arbor, Mich.

Patients, families and healthcare professionals are all welcome and encouraged to attend. The meeting, this year held on the East coast, will focus on severe chronic neutropenia (SCN), and discussions will be held on optimal diagnosis and treatment options.

Participants will be briefed on the latest research and available resources for SCN patients and their families, and national SCN experts will be on hand for further detailed and individualized discussions.

The confirmed speakers for this year's event is the best line up hosted to date, and include several of the foremost American SCN researchers.

The 2007 speakers are Laurence A. Boxer, MD, SCNIR chairman and specialist from the University of Michigan Pediatric Hematology/Oncology Department; David C. Dale, MD, SCN specialist from the University of Washington and SCNIR director; Peter E. Newburger, MD, vice chair of pediatrics at the University of Massachusetts Medical School; and Mary Ann Bonilla, MD, pediatric hematology/oncology specialist from St. Joseph's Children's Hospital in Paterson, NJ.

To kick off the conference on Friday evening, onsite registration will be offered at the Meet and Greet social at the host hotel.

Attendees can pick up their name badges and program packets and get to know the other attendees. Other fun and social activities will be planned for the evening.

Dr. Newburger will kick off the educational series on Saturday morning with "My

Favorite Cell: A brief overview of neutrophil production and function."

He will be followed by Dr. Dale with a presentation on "Cyclic Neutropenia: What We Have Learned in 12 Years of Data Collection."

Dr. Boxer, SCNIR's chairman and this year's hosting physician, will present on his expertise: "Congenital Neutropenia and Related Genetics. What we've learned." Dr. Bonilla will then take the stage and cover a wide array of topics in her presentation titled, "The Clinical Challenges of Living with Neutropenia." Her topic will be all-encircling and include information on such relevant aspects as infection prevention, how to talk to physicians, understanding your counts, the need for bone marrow testing, osteoporosis, and more.

Saturday afternoon will be action packed with what was last year's biggest hit! The "Ask the Doctor" breakout sessions. The sessions will again be lined out according to disease category. The following itinerary has been developed:

- Congenital neutropenia: Dr. Laurence A. Boxer
- Cyclic neutropenia: Dr. David C. Dale
- Autoimmune neutropenia: Dr. Peter Newburger
- Idiopathic neutropenia: Dr. Mary Ann Bonilla

This year's golden opportunity comes in the form of limited individual, one-on-one sessions with the physicians. Unfortunately, not every request can be filled. Selection criteria will hinge on a first come/first serve basis as well as the severity of the need for a second opinion, and as deemed by the experts.

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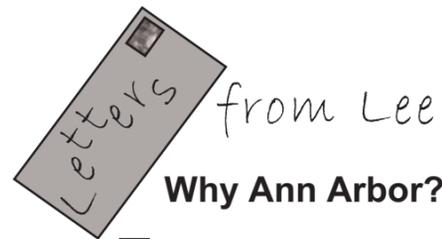
Dr. Boxer in a breakout session at the 2006 conference

RSVP Today!

Fill out the pre-registration form on page 3.

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Why Ann Arbor?

This new location for the Family Conference is in response to many requests we've had in recent years to alternate the venue of the annual Neutropenia Family Conference.

For years it has been in Seattle, Wash., where the Registry (SCNIR) is housed, and next year it will once again return to that fabulous city. But in an effort to serve those who cannot make a trip to the Pacific Northwest, Dr. Boxer has graciously offered to provide the professional support we need to present the 2007 Neutropenia Family Conference in Ann Arbor, Mich. - a charming, family-friendly, Midwestern college town.

July 6-8 promises to be an information-packed weekend with plenty of time to get to know one another and learn firsthand how others handle life with neutropenia.

Fortunately past committee members: Mara Lim, Erin Bogart, Lucy Lyman, and Jennifer Schraag will continue to invest their time and talents on the planning task force. Kristen Saleh, a Michigan resident and mom of two neutropenic boys who attended last year's conference has also agreed to help out.

And thankfully, the Registry's clinical manager Audrey Anna Bolyard, RN, BS, extends her availability, wealth of knowledge and experience to us from Seattle.

In the late 1970s and 1980s when I was raising my daughter, Leta, I felt like we were the only family in the world facing the

Lee Reeves is the president of the National Neutropenia Network. Her daughter Leta, who passed away in 1997, had congenital neutropenia.



daunting challenges of her disease. With her neutrophil count perpetually at zero, our lives were in a constant state of turmoil.

I longed to talk with others who understood what I was going through. That didn't happen until the early 1990s when Amgen reached out to patients to facilitate the start of Canadian and U.S. support groups, and later on when the Registry started the Family Conference.

NNN has worked in tandem with the Registry to assure that the Family Conference grows and develops to meet patients' needs. This year, we're blessed to have four hematologists to present the latest medical information.

We've added new workshops: one for the adult population to focus on ways to enhance quality of life while living with a chronic disease, and a children's program featuring medical educated play.

Special thanks to Dr. Boxer for facilitating the addition of Family Life specialists from the University of Michigan.

I am looking forward to the 2007 Family Conference, to putting faces to the names of those I have corresponded with over the years, and catching up with those I've already had the chance to meet.

My hope for this conference is that everyone who attends goes home armed with valuable information for optimum management of their disease, and new friends to call for support and encouragement throughout the years. **N**

*With Special
Thanks
Anne Dennis*

Anne recently retired following many years of service with SCNIR.

Anne, we not only wish you the very best in your future endeavors, we also wish to thank you for all you have done, all your kind words of encouragement, and your dedication to us over the years.

You will be missed ...

Linking Families

Feeling secluded and as if no one understands?

There is no need to feel alone!

Others are facing all the same worries, woes, and challenges as you, and Lucy Lyman can help you find them.

Through *Linking Families*, Lucy is creating a strong network for people with all types of neutropenia. She is linking individuals with similar diagnosis and life situations.

If you are interested in contacting individuals and families with the same condition as you or a loved one is facing, please email Lucy at: lucyly3@aol.com.

"We don't need to be alone anymore," Lucy reminds us.

Lucy Lyman, a mother of three, has cyclic neutropenia and serves as a member of the Board of Directors for the National Neutropenia Network.

The National Neutropenia Network newsletter is published three times a year.

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Avoiding Those Almighty Germs

Taking a proactive role in learning to stay safe with severe chronic neutropenia (SCN) is at the forefront of all our minds. The challenges we face not only differ from the general public in that SCN patients are more susceptible, but in many cases, our healthcare needs land us in the very places we should be avoiding.

It is no secret that healthcare acquired infections cause nearly 90,000 deaths among hospital patients per year, with an additional \$4.5 billion in medical expenses. National organizations have begun to recognize this and they are working to fight it, but we have to do our part too.

Let's discuss the basics of bacteria and modes of transmission for a moment. There are two distinctive types of bacteria: gram-negative and gram-positive. Gram-negative cell walls are not as tough as the gram-positive cell walls. It's harder to kill gram-positives than gram-negatives because the cell walls are tougher in gram-positives - they have a thicker peptidoglycan layer.

Although the two types of bacteria don't have a true "preference" for a moist or dry environment, gram-negative bacteria will survive longer on a moist surface because their cell walls are more vulnerable to drying out. On skin, you mostly find gram-positive organisms, unless the skin is moist. If skin is moist, then you can find both gram-negatives and gram-positives. There are both gram-negatives and gram-positives in human feces, but around the rectum, unless it's moist, the organisms that end up staying on the skin are for the most part gram-positives (i.e. enterococcus, staphylococcus, streptococcus, etc.).

Our natural skin flora can be a source of infection. You will sometimes hear the hematologists speak of our "danger zone" with our ANCs of being under 200 or so. This is because we can then react to our body's own natural flora.

This is where good hand hygiene comes in. We can't sterilize skin, but we can reduce the bacterial load our skin carries. According to the Institute for Healthcare Improvement's (www.IHI.org) guidance document, transmission of healthcare associated pathogens most often occurs via the contaminated hands of healthcare workers. Today's hand washing compliance rates for healthcare workers hover around an average of 40 percent. What's more shocking is this is an exceptional rate compared to that of the general public. Always request that your doctors and nurses wash their hands before touching you. You too must wash frequently.

You may think well, many healthcare workers wear gloves. The CDC Web site reads: "The use of gloves does not eliminate the need for hand hygiene. Likewise, the use of hand hygiene does not eliminate the need for gloves."

Transmission

There is no argument that infectious organisms can be spread from surfaces to hands to patients. A good example is an outbreak of norovirus reported last year among residents and employees of a 240-bed long term care facility in Pennsylvania. Over half (52 percent) of the residents and nearly half (46 percent) of surveyed employees reportedly had gastroenteritis symptoms during the reported infectious period.

Researchers took environmental surface swabs from resident rooms, a dining room table, and an elevator button in an elevator used only by employees to test the method of transmission in the breakout. All samples tested positive for norovirus and both the environmental and clinical norovirus sequences were found to be identical.

Another study released this summer tested the hypothesis of infection transmission from surfaces to hands. In this project, researchers inoculated the upholstery, flooring, and wall coverings in a hospital area with vancomycin-resistant enterococci (VRE) and *Pseudomonas aeruginosa*. To aid in the assessment of potential for transmission, volunteers touched the inoculated surfaces and then palm plates for later review to

test for growth.

Twenty-four hours following inoculation, all surfaces had recovery of VRE, and 13 of 14 surfaces had persistent *P. aeruginosa*. After cleaning (following manufacturers instructions for each surface), VRE was recovered from seven surfaces, and *P. aeruginosa* from five surfaces. The plate cultures from the volunteers' hands all tested positive for VRE.

Environmental sources of infection transmission

If you culture your carpet, at any given moment you'll get about 10,000 bacteria per square inch. The environment is just covered with bacteria and some forms can survive for up to 72 hours on some surfaces.

A *Chicago Tribune* investigative report solidified the importance of environmental contamination when investigators alleged that in 2000, 75 percent of an estimated 103,000 patients' deaths linked to hospital acquired infections were due to unsanitary facilities, unwashed hands, and unsanitary instruments.

A new study by University of Arizona microbiology professor Charles Gerba ranks the TV remote control as the highest carrier of bacteria in a patient's hospital room compared to the toilet bowl handle, bathroom door and call buttons, among others. Even more disturbing is the detection of methicillin-resistant *Staphylococcus aureus* (MRSA) on the remote control.

The total average bacteria on sites in the 15 hospital rooms Gerba studied were 91, compared to the average for the remote controls at 320. The sites tested included the hand rail, call button, tray table, door knob in/out, bath door out, faucet handle, and flush handle. Additionally, the study involved 20 samples of newly-opened disposable remotes. In this case, the average total bacteria for the newly opened disposable remote controls were significantly less at 8.35.

What You Can Do

Request extra alcohol swabs and wipe down high touch areas in the room ... the nurse call button, the remote, the bed rails, etc. It's a good idea to wipe down the IV pump as well; especially the keypad. Make sure you take shoes, socks or slippers off before getting in the bed or crib and always wear foot coverings when you get out of the bed; even if you're just walking to the bathroom and back.

Limit traveling through the facility as much as possible. Try to avoid treatment rooms - many facilities, especially in the pediatric wards, prefer patients go to a designated treatment room on the floor for procedures such as starting or changing an IV, etc. Ask if the IV team can simply come to your room to do the procedure. The more areas of a hospital you visit, the more organisms you will come in contact with.

When visiting labs and other testing rooms such as an X-ray room, ask that the technician wipe the equipment down before using it on you. These things circulate patient after patient all day, every day. This too could be a huge vector of transmission.

The best piece of advice that I can offer, besides asking you to regularly wash your hands, is the next time you are hospitalized to ask for a visit from the facility's infection control practitioner. When they come, ask them to share with you some pointers on what you can do to protect yourself from infection during your stay. They'll be more than happy to oblige, as this is their job. The lower the infection rate in their facility, the better they themselves look. They will be happy to help in any way. **N**

Jennifer Schraag is a healthcare journalist and mother of three residing in Chandler, AZ. Her two-year-old daughter Brooklyn, pictured above, has autoimmune neutropenia.

The Prevention zone



Crossing the Finish Line: Celia Franklin Races Thru Life Despite SCN

Empowerment, drive, determination, strength ... these are but a few words one might use to describe 20-year-old communications major, Celia Franklin.

Another is *idiopathic*.

Celia's battle with severe chronic neutropenia (SCN) began two and half years ago during her freshman year in college. She had just moved from her hometown of Wilmette, Ill., to attend Northwestern University. Shortly after the school year began, she fell ill with mononucleosis, a viral infection that causes fever, a sore throat and swollen lymph glands, among other symptoms. Celia hit bottom, health-wise, and had a really hard go at it. Even after she should have been well over the mono, she still continued to get more and more ill.

The repetitive CBCs the doctors were doing kept reflecting a dwindling ANC (absolute neutrophil count). Celia says her counts bottomed out at around 200 to 300.

Her physicians decided it was time to start her on Neupogen to try to get her well. She says she experienced a lot of bone pain when she first started the medication, but when she tried to cut back the dose, she again became sick all the time.

She finally decided what was best for her was to take a very small dose every day. "It is much more stable now," she says. "Pre-Neupogen, I was sick all the time. It is amazing what a drastic difference there was. I haven't been sick in a while - probably since I've been on the Neupogen."

An interesting aside to Celia's story is that her father, Cory Franklin, is a physician. He too has been baffled by her illness and worries for her wellbeing - as any father would.

"He doesn't really know much about it. Nobody really does," Celia shares. She adds that he dislikes her daily dosing of Neupogen and often asks her to cut back. She says she feels like she often struggles for her parents' understanding of her personal necessity for the medication. "I know right away if I miss a dose," she explains. "By 4 p.m. the next day I am just dead."

Such a lack of energy is bad for anyone, but for Celia, it would mean not pursuing what she most enjoys - running.

"I ran track in high school and by my senior year I did cross country. I came to Northwestern, it was near where I lived, and I made the team."

Before she could even get a good start on the team that year, however, she fell ill. She was out of her team activities for much of that season - battling the illness and the SCN. Even through all the challenges she faced that year,

she ran in the 5K Forester Invitational and finished 10th overall.

"When I got better, I was able to increase my training," she said. She trained really hard over the summer following that dreadful freshman year, and it paid off. The following season, she became the best runner on her team.

Her comeback involved a full season with the NU Wildcats, finishing first among NU runners in two events and second in another. She led over all of her team's runners at the Big Ten Championships that year with a time of 22:45 and did it again just two weeks later at the Midwest Regional Championships with a time of 23:03.

She set a few personal records that year as well, finishing the 5K Sean Earl Invitational with a time of 18:52 and the 6K Pre-National Invitational with a time of 22:55. She wrapped up the year becoming NU's recipient of the Big Ten Sportsmanship Award.

Celia again ran in all six races for the NU Wildcats this past year. Her dedication paid off when she won the individual title in NU's sweep of the season-opening Chuck Carroll Invitational last Sept. with a 6K time of 23:14. Among other very impressive stats, she ended the season finishing third for NU and 95th overall at the NCAA Midwest Regional Championships.

"I have one more season next year, and I am hoping it will be a good one."

Celia has had her fair share of challenges including a stress fracture in her right femur. A bone density test following the fracture shows low in her spine, so she has begun taking calcium supplements to help keep her bones strong.

Because her legs are quite muscular, she administers her daily Neupogen shot in her back near her hip. Luckily, she doesn't experience the bone pain on her current dose.

Striving and succeeding through all she has faced hasn't been easy, but she has prevailed. "I just had to push thru it," she recalls. "I developed a close relationship with our trainer. She would say not to train as much, to cut back, and eventually, I figured out the Neupogen and I figured out a good training regimen. I am faster now than I've ever been."

She says she's not sticking to any strict plans for the future, but is thinking about law school. She hasn't yet decided whether to go

straight to grad school or break for a couple of years and work.

She did sign up for a few summer classes to "get ahead a little," and is still training for next season with the Wildcats. A marathon may very well be in her future as well. She says she has a friend that dares her to run with him in the New York marathon, and she mentioned possibly trying out the Chicago marathon as well.

"I'm hoping to have enough energy and be healthy enough to," she adds. "I need all the energy I can get right now. Between finals and my running, it takes a lot of energy. During my freshman year, I was a wreck. It's not easy. I don't know, the Neupogen is a lifesaver - literally. Well, between that and coffee; I started drinking coffee this year," she adds, jokingly.

As for her SCN, she says those challenges remain. "There are two sides to it, I think," she explains. "Physically it is so hard. I was sick all the time, and it really drains your energy. Then, emotionally, I felt like there was something wrong with me. No one understands or knows what it is. I thought, 'Am I freak or something?'"

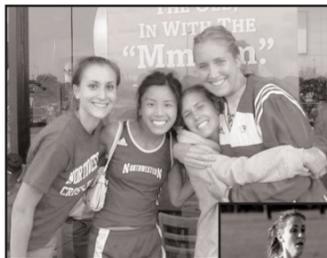
"People are like, 'Oh, you have a blood disorder?' or they see me giving myself a shot. You have to explain those things and they don't

really understand. Plus, all my friends were going out and partying. I couldn't party, I didn't have the energy; which was probably better for me but ... It was definitely hard. Especially just coming in my freshman year. It was hard."

She said she hopes for more awareness and education about SCN, and says she feels not many are well-versed on the condition because, "not enough people have it."

"I get frustrated when I go to the dentist or the dermatologist and try to explain it all to them. Sometimes they know, but most of the time they don't." She says that frustration can sometimes even extend to her family, as no other family member has neutropenia. She says they all mean well but, "They don't understand how I am feeling."

Overall, she keeps a stellar attitude and an even more pristine outlook on life. "Don't give up hope," she offers others facing SCN. "My freshman year; I was at my lowest point. Then, I got better and now I am the No. 1 runner on my team. You have to continue taking care of yourself. Things will get better." **N**



20-year-old Northwestern University student, Celia Franklin, doesn't let her idiopathic neutropenia keep her down.



....continued from pg 1

For initial sign-up for this opportunity, please include the following information with your conference registration form or email nnnconference@yahoo.com with:

- * SCN diagnosis - if one has been established (i.e. cyclic, congenital, idiopathic or autoimmune)
- * Main concern you would like to discuss
- * Recent health history
- * Current treatment (i.e. Neupogen, etc.)
- * Physician name and contact information

Please include any other information you deem of importance. You will be notified if more information is necessary. Every effort will be made to accommodate as many requests as possible.

Saturday afternoon will wrap up with concurrent sessions. The two topics will include: "Living with Chronic Illness" and "The Challenges of Parenting a Child with Chronic Illness."

In addition, a new workshop titled "Key Strategies to Living Well With Chronic Illness" will be added to this year's educational program and will be presented by Trish Robichaud, a motivational speaker and life and disability awareness coach. The session will focus on proactive living.

There will be several networking and story-sharing opportunities for attendees, and fun-packed, educational activities for the children scheduled throughout the event. Jessica Porter, a certified therapeutic recreational specialist with the University of Michigan Child Life Program will facilitate children's activities on Saturday morning with the assistance of several Child Life professionals.

The children's program will feature medical educated play. One activity planned uses every day materials to simulate the making of blood. For example white rice will serve as white blood cells. The format is fun and interactive, and allows children a comfortable atmosphere to play and talk. Other esteem-building activities are also planned. The children's afternoon will be more open play focusing on art projects and games.

Sunday's activities will wrap up the weekend beautifully with

networking and camaraderie, as well as an update on the NNN presented by Lee Reeves, president. The day's activities will end at 1 p.m. to allow for travel time.

Registration Information

The pre-registration fee for the conference is \$65 for adults and \$30 for children under age 10. Onsite registration is \$75 for adults and \$30 for children under age 10. This fee includes all presentations and several meals throughout the three-day event.

If this fee presents a hardship, please contact Lee Reeves. Thanks to donations from several families, funds have been allocated to help with conference fees.

To pre-register send a check or money order to the NNN at P.O. Box 1693, Brighton, Mich. 48116. Please be sure to include the registration form below with your payment, and don't forget to include your request for the expert consult mentioned above if you are signing up for that.

Hotel Accommodations

A limited number of rooms are being held at the Holiday Inn North Campus, in Ann Arbor. If reservations are made prior to the June 15 reservation deadline, the group rate for conference attendees is \$82 plus 8 percent room tax. Ten portable mini-refrigerators are available on a first come, first served basis, and the hotel staff has agreed to make a refrigerated space available for all NNN guests. Ice machines also are readily available on each floor.

Call (734) 769-9800 to reserve your room or visit the hotel's Web site at www.hiannarbor.com for more information. The property is 25 minutes from Detroit Metropolitan Airport and just minutes from the University of Michigan Medical Center. For more information on the Ann Arbor area, visit www.annarbor.org.

For more information about the 2007 Neutropenia Family Conference, email us at nnnconference@yahoo.com, or contact Lee Reeves, NNN president, at (810) 229-5797 or email her at leereeves99@comcast.com.

Watch our Web site for updates leading up to the event: www.neutropenianet.org. **N**

2007 Family Conference Registration Form

Name (please print): _____

Address: _____

City: _____ State: _____ Zip: _____

Day/evening/cell phone: _____

E-mail address: _____

Number of attendees: _____ adults _____ children

Please list age of each child: _____

Names of each attendee: _____

Person with SCN: _____ Type of SCN (if known): _____

_____ Please check here if OK to include SCN type on name badges

Any food allergies or other special needs?: _____

_____ Check here if you are willing to volunteer to help with the conference.

_____ Please check here if including sign-up information for the one-on-one appointments with the experts.

Please detach and mail with full payment for total number of attendees listed above to P.O. Box 1693, Brighton, Mich. 48116

REGISTRATION COST

	Pre-registration	Onsite
Adult:	___ \$65 each	___ \$75 each
Child:	___ \$30 each	___ \$30 each

Total for Adults: \$ _____

Total for Children: \$ _____

Total enclosed: \$ _____

Please make checks payable to the National Neutropenia Network.

NEWS
~
IN
~
BRIEF

The Five-Second Rule De-Mythed!

We've all heard of it and most of us and even lived by it as small children.

The golden five-second rule that tells us that if we pick up that edible whatever-we-dropped within five seconds than it is still OK to eat.

Well, researchers at Clemson University investigated this long running childhood standard. Their results may surprise you - or at least make you rethink your dedication to the rule.

Experiments were conducted to determine both the survival and transfer of the common bacteria *Salmonella Typhimurium* from three separate surfaces: wood, tile and carpet. The test subjects used were single slices of bologna and bread.

Ultimately, the researchers found that Salmonella can survive for up to four weeks on dry surfaces in populations high enough to be transferred to foods. Moreover, Salmonella could be transferred to the foods tested almost immediately upon contact.

The scientists also note that over 99 percent of the Salmonella cells were transferred from the tile to the bologna after five seconds of the bologna being exposed to the tile. Transfer from carpet to bologna was much lower than that of its counterparts: wood and tile.

Ick!

Source: Dawson P, et. al. Residence time and food contact time effects on transfer of Salmonella Typhimurium from tile, wood and carpet: testing the five-second rule. J Appl Microbiol. 2007 Apr;102(4):945-53.

NEUTROPENIA EDUCATIONAL DVD NOW AVAILABLE!

The NNN is proud to announce the availability of the SCN educational DVDs. The DVD is of select sessions from the 2006 Neutropenia Family Conference. It includes all of the following sessions:

- >> Dr. David C. Dale ~ "Neutropenia: What We've Learned in 12 Years of Data Collection."
- >> Dr. Laurence A. Boxer ~ "The Genetics of Severe Congenital Neutropenia Based on an Unusual Set of Events."
- >> Dr. Frank Roberts ~ "Dental Hygiene for the Neutropenic Patient."
- >> Jennifer L. Schraag ~ "Infection Prevention in the Healthcare Setting."

Cost is \$30, which includes shipping and handling. For more information or to submit an order, contact Lee Reeves by email at leereeves99@comcast.net; by mail at P.O. Box 1693, Brighton, Mich. USA 48116; or by phone at (810) 229-5797.

Bank On It!

Many cord blood banks offer their services free of charge to families in need. For example, CBR (Cord Blood Registry), located in Tucson, Ariz., offers a "Designated Transplant Program" as a public service.

CBR provides the Program (also referred to as DTP) free of charge to families wishing to store their newborn's umbilical cord blood stem cells for a sibling or other family member diagnosed with a life-threatening disease that is currently treatable with stem cells.

This includes congenital neutropenia due to its increased risk of acute myelogenous leukemia (AML).

Facts from the CBR Web site: "Studies have shown that related stem cell transplants provide double the survival rate over unrelated stem cell transplants - with far less rejection issues, thereby reducing the need for a lifetime of drugs."

There are over 1,000 patients currently enrolled in the DTP.

To qualify:

The family member in need must be diagnosed with a disease that is currently treatable with umbilical cord blood stem cells; be a first- or second-degree blood relative; have a hematologist/oncologist who agrees that the stem cells can be used for treatment

CBR requires little paperwork to be filled out by the parents and the treating hematologist. Once completed and returned, CBR will decide to accept or decline each case as they see fit.

If approved, they will send the collection kit free of charge and will process and store the newborn's cord blood sample for an undetermined amount of time at no cost to the family.

In addition, should the physician who collects the cells charge, CBR will reimburse that expense up to \$150. The only fee the family is required to pay is the courier charge for the cells to be transported from the place of collection (i.e. the hospital where you give birth) to the CBR facility. This charge is generally \$150.

For more information on the Designated Transplant Program, please contact Cord Blood Registry at (888) 932-6568 or visit www.cordblood.com/dtp/index.asp.

For other companies that extend this courtesy, visit: www.parents-guidecordblood.com/content/usa/medical/caseofneed.shtml.

FOR MEDICAL QUESTIONS CONCERNING NEUTROPENIA OR NEUPOGEN, OR TO FIND A KNOWLEDGEABLE PHYSICIAN IN YOUR AREA, CALL ONE OF THE SEVERE CHRONIC INTERNATIONAL NEUTROPENIA REGISTRY (SCNIR) OFFICES :

USA	Europe	Australia
Audrey Anna Bolyard, RN, BS Seattle, Wash. (206) 543-9749 (800) 726-4463	Hannover, Germany Beate Schwinzer, PhD 49-511-557105 Connie Zeidler, MD 49-511-546-0918	Ballarat, Victoria Australia George Kannourakis, MD 61-353-33-4811

Wanna Help Out?

The National Neutropenia Network is currently seeking to expand our board of directors. We are looking for volunteers with leadership experience on nonprofit boards and expertise in one or more of the following areas: accounting, law, fundraising, business, technology, medicine. If you have skills in these areas and are committed to seeing the NNN grow and develop, please contact Lee Reeves at leereeves99@comcast.net. Thank you!

Research Review:

Dan Link, SCNIR Board Member and Researcher Shares His Thoughts on Today's Advances, Tomorrow's Hope, and What We Can Do to Help

Daniel Link, MD, associate professor of medicine at Washington University School of Medicine, in St. Louis, and board member of the SCNIR is a dedicated researcher of SCN.

He works closely with the Registry and with Dr. Laurence Boxer to uncover the many intricate details encircling this insidious disease that threatens our loved ones.

He points out that over the last three or four years, SCNIR researchers have come to understand the genetic mutations that cause most types of congenital neutropenia. For example, he mentions that for severe congenital neutropenia, there are two dominant mutations: ELA-2 and the more recent findings by Karl Welte and the German SCNIR group that identified the HAX-1 mutation.

"We now know the genetic basis for most types of congenital neutropenias," he says. Link shares that the eponym "Kostmann's Syndrome" is now being reserved for those cases of neutropenia with mutations of HAX-1. "We don't know for sure, but probably 10 to 20 percent of all cases of severe congenital neutropenia are HAX-1-mutant; and probably about 60 percent are ELA-2-mutant. So we think we know about 80 percent or so of the causes."

Now armed with the knowledge of these genetic mutations, Link says the question he and other researchers are asking is "Can we use this information to help determine prognosis?"

"We have a study where we are going to be genotyping people with congenital neutropenia for all the common and even uncommon mutations," he explains. "Then we are going to ask, if you have an elastase (ELA-2) mutation, are you more or less likely to get leukemia? Are you more or less likely to respond to GCSF? And so on. So it may be possible that when somebody is diagnosed with congenital neutropenia, we can provide some prognostic information and possibly even modify treatment." This research is ongoing, he says, adding that another future hope is to gather enough information to one day develop a targeted therapy.

They have a current hypothesis they are now working on, (please note that this is just in the developmental stage at this time), where they think that the ELA-2 mutations cause disease by activating the unfolded protein response (UPR). A paper covering their initial findings is currently under review and should be published soon. Link explains, "They (the mutations) cause proteins to fold poorly and this causes the cell to die. Importantly, there are drugs that can modify the UPR pathway. "We

are currently testing whether these drugs in bone marrow cells from patients with severe congenital neutropenia. If these drugs can reverse the block in neutrophil development, we may have a novel therapy for this disease. However, I don't want to get people too excited because it is still just a hypothesis," he asserts.

Another hot topic of current research is the GCSF receptor mutation that has been found among the congenital group and has been linked to leukemic transformation. In fact, they have noted recently that fl of children with congenital neutropenia that went on to develop leukemia had this mutation (another Welte study, he notes). "The evidence that G-CSF receptor mutations directly contribute to leukemia is becoming quite convincing"

He says it is his personal belief that patients should be screened for this mutation at their annual bone marrow biopsy. But challenges lie within this aspect in that while this screening can actually be done through either blood or bone marrow samples, where to have the testing done is the big challenge. Link says that currently, there are no clinical labs that perform this testing (only his and other research labs, but they are not at liberty to do the "clinical" screening for patients).

Link goes on to say that one of their biggest pushes in their current research is determining who exactly is at risk for leukemic progression. "I mentioned the GCSF mutations, but there are others, and we are trying to identify those mutations. So that is ongoing," he adds.

Link says the one thing that is needed most

is bone marrow samples.

"That's probably the single most important thing. There is a real need to take these samples and use them for research." He explains that the annual procedure many SCN patients undergo is used for clinical reasons, and with consent, there's usually enough left over to do research.

"If families could encourage their doctors to do that that would be helpful for many different people's (SCN researchers) research."

He said both he and Boxer have been working on getting fresh samples sent. "What I mean by fresh is, a lot of times the samples are sent frozen. For purposes of our research, we need living cells. If we had more of them, we could push the pace of science faster."

He continues, "The leukemia aspect, to me, it is one of the key issues. There are almost no samples available. I have collaborative efforts in place with the German Registry, the Australians, the French, and we've collected only 18 samples. So it is the bone marrow of the kids that unfortunately transform that we are really missing. We're getting very few of those and it's really slowing down research."

While Link conveyed his utmost compassion to the families facing such daunting times, he also is hopeful that recognizing the need will convince them to help.

"The pace of science, from my perspective especially, is not going as fast as it could because we are not getting samples. If families can help donate bone marrow samples, maybe in the long run it will help us come up with a better therapy."

Know Your Dose

The following conversion chart is for Neupogen at 300 micrograms per milliliter (300 mcg/1mL) concentration. Please note: other concentrations are available, please check concentration of yours carefully. It is located on the label on the vial.

micrograms (mcg)	cc or ml syringe	insulin syringe
3 micrograms	0.01 milliliters	1 unit
6 micrograms	0.02 milliliters	2 units
12 micrograms	0.04 milliliters	4 units
18 micrograms	0.06 milliliters	6 units
24 micrograms	0.08 milliliters	8 units
30 micrograms	0.1 milliliters	10 units
60 micrograms	0.2 milliliters	20 units
120 micrograms	0.4 milliliters	40 units
180 micrograms	0.6 milliliters	60 units
240 micrograms	0.8 milliliters	80 units
300 micrograms	1.0 milliliters	100 units

chart courtesy of SCNIR