Join us at the 2012 Neutropenia Family Conference

It’s a first! We’re heading southwest for the Neutropenia Family Conference. Plans are underway for a wonderful weekend of learning and camaraderie July 13-15 in Scottsdale Arizona. If you’ve been entertaining the idea of attending, this would be a good year as we are skipping a year and will not have a Family Conference in 2013.

As always attendees will have a chance to meet in private consultations with the neutropenia experts: Mary Ann Bonilla MD, Laurence Boxer MD, David Dale MD, and Peter Newburger MD. We will have break-out sessions for different disease categories, and opportunities for guests to share stories and get to know each other. Shay Jones MA, LPC will facilitate a support group session exclusively for adults with neutropenia—last year, attendees gave Shay’s program rave reviews. In another popular sessions, parents will join together to discuss their challenges under the guidance of a trained counselor.

Shelly Uram MD, Harvard trained and triple board-certified psychiatrist and clinical associate professor of Psychiatry at the University Of Arizona College Of Medicine will open the Conference on Friday afternoon with a presentation on the impact of stress on the brain. Dr. Uram’s discussion will include a focus on how to develop a healthy perspective on a life with neutropenia in it.

Nutritionist Heather M Duquette-Wolf, Registered Dietitian will talk about the optimum diet for individuals living with a chronic condition like neutropenia. Heather specializes in medical nutrition therapy for patients with disorders such as diabetes, gastrointestinal disorders, arthritis and other auto immune disorders.

The medical community in Phoenix has been extremely welcoming. We are fortunate to have the child life staff at Phoenix Children’s Hospital to create another extraordinary Kids Camp program. Some activities currently planned include: making blood soup, face painting, pet therapy, a craft activity, and game show. The staff will provide a safe and open environment for children to discuss and better understand neutropenia, but the focus will be to have fun and make friends.

Our host hotel, The Doubletree Paradise Valley Resort in Scottsdale gave us the best room rate yet—$69 plus tax. It’s a lovely property with plenty of space. For more information on the greater Phoenix area go to: visitphoenix.com

The Conference is a rewarding and serendipitous experience. As in past years some grants will be available to help offset the costs for those families and individuals who cannot attend without assistance. Write to lee@neutropenianet.org for more information. For the many who cannot attend we will publish a follow up newsletter with highlights and details.

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The National Neutropenia Network newsletter is published several times a year. Lee Reeves, NNN Executive Director: lee@neutropenianet.org P.O. Box 1693 • Brighton, MI 48116 • 517.294.0736 • neutropenianet.org

Many thanks to Mariel Lewis, owner of Trapdoor Design, for updating the look of our newsletter. She also designed an attractive fundraising brochure which is now available to anyone wishing to raise funds for the NNN. (Contact lee@neutropenianet.org) We are most grateful to Mariel for making a difference.
Many readers know first-hand the isolation and confusion that comes with the diagnosis of a rare disease like neutropenia. Even with the benefit of caring knowledgeable physicians, and a supportive family, a diagnosis like SCN can be a lonely and difficult path. The chance to communicate with another person who has walked in the same shoes eases the isolation and uncertainty that can make living with neutropenia so difficult. Our new Peer Support Volunteer Program is designed to be a lifeline for the newly diagnosed, and a resource for someone going through a particularly rough period.

Shay Jones MS LPC, spearheaded this program guiding it from conception to final launch on our website in late February. He’s worked with a variety of volunteers with specific skills, experiences and areas of expertise. The volunteers do not dispense medical advice, but do share their own experiences with different types of neutropenia either as a patient or parent.

Referring to the benefits of the program Shay Jones said, “I believe in the value of peer support. Neutropenia has a way of influencing many aspects of a person’s life. Talking openly with someone who has walked a similar path can be a healthy and uplifting experience.” These areas can include, but are not limited to a person’s sense of self, occupation, education, business affairs, personal relationships, and overall ability to energetically enjoy life. “We have selected and trained a handful who understands,” said Lee Reeves, NNN Executive Director. “I remember how alone I felt when my daughter was growing up. It seemed that she was the only child in the world with this rare disease.”

Asking for support is not always an easy thing, but living with neutropenia requires more than just hope. It doesn’t take long to realize that the effects of neutropenia quickly expand beyond what is happening in one’s blood.

To sign up to speak with a PSV, visit neutropenianet.org. Click on the “Patient Support” tab, on the drop down menu click on “Support from Peers” where you will find a link to an application.

If you would like to apply to be a Peer Support Volunteer, contact Lee Reeves at lee@neutropenianet.org.

Introducing our Peer Support Volunteer Program

Measuring and Maintaining Bone Health is Essential for Neutropenia Patients

At the 2011 Neutropenia Family Conference, Linda Anne DiMeglio MD, MPH, an associate professor of Pediatric Endocrinology/Diabetology at Riley Hospital/Indiana University in Indianapolis, offered her insights for helping neutropenia patients maintain healthy bones.

Bone mass acquired during childhood and adolescence is our natural protection against osteoporosis. X-rays do not detect bone loss until 20 percent of bone is gone, Dr. DiMeglio explained. She recommends DXA screens to detect bone loss much earlier. The DXA uses two beams of radiation to distinguish bone from soft tissue. It has a low dose of radiation. Scan time is between 3 and 30 minutes. Screenings should be done at diagnosis and every year if abnormal. However, there are no norms for children under age five, she said.

While DXA is accurate and a good choice for monitoring sequential bone changes, it needs to be done on the same machine each time. For children, it is important to have a DXA done at a center used to measuring children.

Screening for osteoporosis should include blood tests for calcium, phosphorus, 25-vitamin D (a stored form of Vitamin D), parathyroid hormone (which looks for stress on the system) and urine calcium to see if adequate calcium is being absorbed.

She encouraged people to remember that weight and bone marrow density are directly related. Avoid caffeine and excessive soda consumption; maintain adequate calcium and vitamin D intake. “Standard” supplements of 400-800 IU vitamin D a day may not be enough. Adequate calcium and vitamin D intake is essential. Good sources of calcium include dairy products; dark green, leafy vegetables; and calcium-fortified foods and beverages. Supplements can help ensure that the calcium requirement is met each day. Vitamin D supplements may be needed to ensure an adequate daily intake.

Bone is a living tissue that responds to exercise by getting stronger. The best exercises for bones include walking, stair-climbing, dancing and weight training. Exercises also strengthen muscles that support bone, enhance balance and flexibility and preserve joint mobility.

Smoking and drinking alcoholic beverages negatively affects bones. Smokers may absorb less calcium from their diets. Those who drink heavily are more prone to bone loss and fracture, she added.
Shot Talk: Try These Tips to Ease Injection Pain

Coping with the pain of injections and the best way to deal with them was one of the most popular subjects for families attending the 2011 Neutropenia Family Conference. We polled those attending to provide their best tips for shots.

Here are some ideas for easing the pain and tension with shots:

- Cut up sponges into small pieces, wet and freeze them for shot time. Apply to skin to ease the burning sensation.
- Hold syringe in hand for a few minutes to bring it to room temperature to ease the pain of injection.
- If you ever draw back blood, reposition the needle and try again. Neupogen is not harmful if you inject it in a vein but it will not be effective.
- Some patients use EMLA cream, a topical anesthetic which numbs the skin and is only available by prescription. The active ingredient is Lidocaine/Prilocaine.
- Some found relief using Cold Spray, an inexpensive over-the-counter non-medicated spray commonly used to ease the pain of burns and some athletic injuries.
- Collect little gifts at garage sales and save them for “shot prizes” on the difficult days when your child really resists taking his or her shot.
- Let your child give a pretend shot to her doll or stuffed animals first.

For those who do not have the funds to attend the 2012 Conference, there are grants available to help offset costs. If you would like to be considered for a grant, contact lee@neutropenianet.org and ask for a Conference Grant Application. Grants will be awarded based on financial need, and medical/health need. All contributions made to the Hardship Grant Fund go to helping families and individuals attend.

Hardship Grants

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Last year, nine families were able to attend the conference thanks to the generosity of the following contributors whose lives have been impacted by neutropenia.

- Mary & Mark Glazer
- Lorne & Gayle Gold
- The Jacobson Family
- Galee Jean-Louis & Dr. Vincent Mattaacci
- The Lim Family
- Kristen & Robert McGuinness in honor of The Ella Jewell Foundation
- Liz Molina
- Angela and Patrick Moore in honor of The Elia Jewell Foundation
- The Moore Family
- Lee and Louis Reeves in memory of their daughter
- Letia Reeves
- Tom Rifai MD
- Sue and Jim Rister
- Steve Ryan in honor of his son Mitch Ryan
- Mary & Carl Schroeder in memory of their sons Louis and Matthew Schroeder

Kids Camp 2012 is set for Saturday, July 14. “We are fortunate to have the enthusiasm and commitment of the terrific Child Life staff at Phoenix Children’s Hospital,” Lee Reeves said. “They understand the needs of our children and will plan a day of fun and learning in a safe and upbeat environment where children will have plenty to do.”

Hat-making and brick decorating were two of the most popular activities during SCN Camp for Kids 2011.

The group of about 18 children ranging in age from 4-12 kept busy throughout the morning and afternoon with a full schedule of activities that included making “bone marrow snacks,” drawing pictures of what having neutropenia means to them, and learning about laughter Yoga. Children also practiced proper hand washing and learned about how blood is manufactured in the body. Jessica Delisle BS, CTR and Marianna Loevski, BS, CCHS, from CS Mott Children’s Hospital led the children in several activities along with a team of volunteers.

Dagnor Moore of Ann Arbor volunteered to help out with the children and said she enjoyed it so much she can’t wait to help out next time the conference is in Michigan.

“I was totally charmed by the children and amazed how well they responded to the art projects and the games,” she said. “It was a wonderful experience. I’d volunteer again tomorrow.”

Moore said during her time helping out she became a “champion paper hat maker.” Using butcher paper, packaging tape and various odds and ends the children created hats that they could customize with ribbons and other scraps.

“Parisian hats to cowboy hats—you name it we made it.” Moore said.

Art facilitators brought in bricks and allowed the children to paint and decorate them, Moore said. “The kids really liked that they got to take them home,” Moore said.

After lunch children shared pictures and stories about living with neutropenia whether as a patient or a sibling of a patient.

Some of the drawings they shared with the adults gathered in the main conference room included a white blood cell eating a germ, a person in the hospital, a girl with an IV and many injections.

Comments from the children included:

“Neutropenia affects me and my family because we have to go to the hospital a lot for my sister. My family has to take a lot of safety precautions more than others because of neutropenia.”

“I have a lot of shots. I used to have to take one every other day, now I take one every day.”

“We should always wash our hands when we start to eat and we should use a paper towel to open the doors. We should cover our mouth when we have a cough.”

“If someone gets sick, we might get sick too. If you don’t have any white blood cells you might get sick and you might get dead, so we should wash our hands.”

“Neutropenia affects me because we can’t spend as much time as a family.”
Twelve-year-old Devin Reid can’t remember what it was like to be ill. His mother, Yvonne Bradley-Reid, remembers the years when Devin took shots daily and was frequently ill. Devin was born July 8, 1999 three weeks premature—his parents initially thought his frequent illnesses might have been due to his premature birth. But by the age of six months his poor health continued and several very high fevers had spurred them to take Devin to the hospital. His mother noted that he’d had pneumonia seven times by the time he was a year old. She was frustrated and worried. “I kept saying ‘something’s not right’” said Yvonne Bradley Reid.

Blood tests confirmed her suspicions. At nine months Devin’s neutrophil count was recorded at 50. During an extended hospital stay in April, she finally got an answer for her son’s medical problems. A physician told the family that Devin had autoimmune severe chronic neutropenia. With his counts being so low and numerous serious infections, Devin was put on Neupogen.

Not long after his diagnosis, the family had an opportunity to transfer to Florida where Devin did very well under the care of hematologists at All Children’s Hospital. For three years Devin took Neupogen injections and his father took him to the hospital for routine blood tests. Around the age of four Devin’s family got the good news that his neutrophil counts were beginning to climb. By the time he was ready for kindergarten his counts were normal and he no longer had to take shots.

His parents were ecstatic and deeply grateful for their son’s good health. “No one would ever know that he was sick as a kid. He is well and healthy. He’s an athlete,” said Yvonne Bradley-Reid.

His parents still keep an eye on Devin’s health but he now leads a normal active life. “He doesn’t let it impact him in any way,” his mother added.

What is autoimmune neutropenia? It is a disease in which the neutrophil count is low because the person’s immune system makes an antibody, directed against his or her own neutrophils, that destroys them as rapidly as they are made. Another important characteristic is that the frequency and severity of infections is generally lower, at any given absolute neutrophil count, in an autoimmune neutropenia than in other forms of neutropenia due to failure to produce cells in the bone marrow. The reason is that the neutrophil count represents neutrophils in the blood at a moment in time. In autoimmune neutropenia, the neutrophils may transit through the blood more quickly than normal, but they are still able to get to the site of an infection in tissue outside the bloodstream. There is a delicate balance between the destruction by the antibody and production by the bone marrow, so the neutrophil count may fluctuate widely. Many people with autoimmune neutropenia do quite well clinically. Part of their risk is that they end up with unnecessary hospitalizations from minor infections. Although they still need to be evaluated for fever and infections, as it may be hard to distinguish what is minor and what is major. This may be why there is often variation among physicians in the management of fever, as autoimmune neutropenia is quite different from the more common neutropenias that follows chemotheraphy.

Is the cause known? No. It can sometimes be associated with systemic autoimmune diseases such as lupus, but in children it tends to be isolated. In children, it is generally transient, resolves completely by age 3-5, and does not come back. There are outliers in whom it lasts a bit longer. In adults, it may be much more chronic or lifelong.

How is it diagnosed? Diagnosis is difficult. It relies primarily on a test for anti-neutrophil antibodies. However, the quality of that test is very variable. It is important that it be done by a laboratory that is experienced with the test. Also, the test is susceptible to both false positives and false negatives. Most physicians use the test, but not as the only basis for the diagnosis. Some may also do a bone marrow examination.

Unlike severe congenital neutropenia, which typically presents with severe illness and infection, autoimmune neutropenia may be discovered as an incidental finding on a lab test.

Is a bone marrow biopsy necessary? It is often helpful, but not always necessary.

Is there a group that is predisposed to autoimmune neutropenia? No.

Does the presence of the antibody always indicate that a child has autoimmune neutropenia as opposed congenital? No. It may be hard to distinguish between the two. Having a positive antibody test does not rule out other forms of neutropenia.

Do children always outgrow it? Children usually outgrow it, although we can never say that 100 percent of them will. It is likely to be chronic only when associated with an underlying immunodeficiency or autoimmune disorder, or when combined with other autoimmune blood disorders such as immune thrombocytopenia (ITP) or anemia. Adults are much more likely to have long-lasting or life-long neutropenia and it is more often associated with an underlying disorder like lupus.

What is the recommended treatment? There are multiple treatment options all of which are valid. One is simply close observation and treatment of any infection that does arise. That is usually my first choice. Another is to use prophylactic antibiotics to reduce the frequency of infections. That’s usually not necessary, because most of these patients don’t have frequent infections. The last is using G-CSF or Neupogen, for the rare autoimmune neutropenia patient with frequent hospitalizations or emergency room visits. These patients usually respond to very low doses, which can sometimes be taken every other day.

Is autoimmune neutropenia extremely rare? Autoimmune neutropenia is rare, although more common than congenital neutropenia. Some cases may go undiagnosed because the affected individuals never become ill.

Is it associated with developing leukemia? Unlike congenital neutropenia, autoimmune neutropenia is NOT associated with leukemia, and G-CSF (Neupogen) can be administered without fear of this complication.

Some people with autoimmune neutropenia are treated with steroids, is that generally recommended? No. Steroids have many side effects including suppression of other parts of the immune system so they actually make the patient more susceptible to infection. They also can have effects on growth, bone development, blood sugar and blood pressure. They may be used in some adults, such as those with lupus. In children, steroids should be used only for treating an underlying disease (e.g. lupus) and not for neutropenia alone.

How about Intravenous gamma globulin therapy? That may sometimes be helpful, but then only very transiently. The treatment is ineffective in the long run, and is both uncomfortable and expensive, so it is very rarely used.
Welcome Katie Bottinger

We are pleased to welcome new board member Katie Bottinger to the National Neutropenia Network Board of Directors. Katie, who has had idiopathic neutropenia for twenty years, resides in Wayzata, MN with her husband and two children. She comes to the NNN board with rich and varied experience in the not-for-profit management world. Since 2000 she has served as the Director of Volunteer Resources for Hammer Residencies which has been providing quality services for adults and children with developmental disabilities since 1923. From 2006-2010 she served as the organization’s Program Director, opening four new homes and supervising six managers. “We are delighted to have someone with Katie’s experience and dedication join the board,” said Lee Reeves. “She will be a great addition.”

Katie, understands firsthand the challenges of trying to manage life with neutropenia as well as a job and family. She is excited about the opportunity to serve on the board, “I would really love to become more involved and help others with neutropenia—whether its education, advocacy setting up support groups, e-newsletter, etc”

ER Card Available on NNN Website

Visitors to the Network’s website, neutropenianet.org, who click on the Resource tab, can now download an Emergency Room Protocol Card. This card, designed to be presented in hospital emergency rooms, guides the ER physician on the recommended protocol for treating a febrile patient with severe chronic neutropenia. We recommend downloading several copies and laminating them to assure you get the best care in emergency situations.