The 2007 Neutropenia Family Conference, hosted by the National Neutropenia Network (NNN) and the Severe Chronic Neutropenia International Registry (SCNIR), had its “East Coast” debut in Ann Arbor, Mich., July 6-8. More than 120 people attended the annual conference. Once again, it was a great success.

The Ann Arbor location attracted families from the East and Midwest who have been unable to make the trip to Seattle, though people did come from throughout the country — some as far as California and Texas. Attendees came to share information, learn the latest research and provide support to one another.

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

Activities kicked off Friday with a reception and a story circle facilitated by life coach Trish Robichaud.

Saturday morning began with breakfast and a warm welcome from Lee Reeves, NNN president. Dr. Boxer then expressed his enthusiasm for the having the event in Ann Arbor and for seeing so many patients who made the trip to learn more about neutropenia. “It is so gratifying as a physician to know that your lives are being changed,” he said. “It was very frustrating prior to the ability to use Neupogen. All we had was supportive care.”

Dr. Newburger was first on the agenda with a presentation on “My Favorite Cell: A brief overview of neutrophil production and function.” This was a new topic for the conference and it got high marks from attendees who were thrilled to hear how this amazing white blood cell performs in the body. Dr. Newburger even included some fascinating footage of neutrophils prowling the bloodstream for bacteria to devour.

“...
Letter's From Lee

Quantum leaps in the treatment of neutropenia have been made since my daughter was diagnosed in 1978. The introduction of Neupogen has been by far the most significant. Before Neupogen antibiotics were used to manage the endless infections that plagued Leta’s life. I stopped counting her hospitalizations after the 40th. Those were lonely times with no support group to turn to. Now, nearly 30 years later with all the advances in treatment — from the advent of Neupogen to improved bone marrow transplant protocols — I believe the need for our support group is even greater.

The 2007 Family Conference drove this home in a powerful way. In the feedback forms almost everyone stated “education” as one key reason for attending: “to learn about the latest research,” “to understand the difference between cyclic and idiopathic,” “to learn how to deal with side effects of Neupogen.”

When Leta was growing up, these were not issues. The research had yielded little success; the term idiopathic neutropenia didn’t exist, and the side effects we worried about were from antibiotics. In this era of abundant information, those who keep current on relevant research findings have the best chance of getting optimum medical care. With four of the leading specialists in the country presenting at this year’s event, attendees had access to an unprecedented wealth of information.

One thing hasn’t changed in 30 years. People still want to network. That’s what inspired many to make the trip to Ann Arbor this past July: to meet others like me, to share my story, to connect with others. Throughout the weekend they laughed and cried together and told heart wrenching stories. They arrived as strangers and said goodbye as friends. The children played in a setting

Shay Jones with Lee Reeves at the 2007 Family Conference

then her heart and lungs.

“ITs taken me a long time to be able to say things happen for a reason. I hate that it had to be my baby, but I will make sure that no child will go to that hospital with no neutrophils” and it cost them their life.

“They are going to know my baby’s name.”

One week later, Brittany had to call to cancel the follow-up appointment with the hematologist. “When I called, I asked if she was sick enough now — because she wasn’t here anymore.”

Joeli was a vibrant 3-year-old who liked baby dolls and working outside with her daddy. “She liked make and animated.

A very special “Thanks” to Cyndi Lieske for doing all of the writing for this issue

Comments, contributions, and article suggestions are always welcome and encouraged. Please email Lee Reeves or Jennifer Schraag or write to: National Neutropenia Network P.O. Box 1693 Brighton, Mich. USA 48116 (810) 229-5797 www.neutropenianet.org

NNN LINKING FAMILIES PROGRAM
The NNN LINKING FAMILIES PROGRAM links individuals with similar diagnosis and life situations.

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

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

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Brittany says she showed no signs of strep throat, no sore throat, nothing. They called the doctor on call and he claimed the infection is what’s to blame for the low ANC. He ordered a shot and a prescription of oral Zithromax. “They told me to go home.”

On Monday morning, Joeli vomited directly following her first dose of the antibiotic. Brittany called her pediatrician. They said she’d be fine and to make sure she drank plenty of fluids. But on Tuesday, Brittany says Joeli was fussy and lethargic. She took her back to the doctor. “They did not do a CBC. I didn’t think to tell him to do it. Tuesday night she vomited green. She didn’t want to get up and walk because her tummy hurt. I had to carry her to the bathroom.” She called the doctor yet again, and they advised her to take Joeli off the Zithromax.

By Wednesday night things were getting worse. Another call to the doctor, another “if she is drinking, she will be fine” reply. Brittany became frantic on Thursday morning when Joeli’s pulse was almost 200 when she checked it. They had been up all night. The doctor said to meet them at the office when they open, but when they arrived they were told the doctor was running late and to take Joeli straight to the ER.

“I was crying, I knew something was wrong. They made us wait when we got there. She arrested at 10:17 that morning, and they couldn’t get her back.” Joeli had gone septic from a pseudomonas aeruginosa infection. The deadly bacteria had infiltrated her bloodstream, shots, considered normal. They came out of their programs smiling and animated.

The annual conference is a bittersweet experience for me. It calls up difficult memories but it also reminds me that bringing people together to share their stories and celebrate their victories is a perfect way to honor my daughter’s life.

This year many people stepped in to help make the conference a success. At the risk of leaving some deserving person out, here are their names: Audrey Anna Bolyard, Tracy Marrero, Kim Blanz, Debbie Dicesare, Mara Lim, Shelly Fiscus, Tammy Loader, Kristen Saleh, Jennifer Schraag, Sharon White, and the many good Michigan friends I persuaded to volunteer for the children’s program.

A very special thank you goes to Erin Bogart, a driving force throughout the planning stages and for the entire weekend. Here’s an excerpt from a note she sent me that sums up how many of us felt after the final gathering on Sunday: “I can’t stop thinking about all the amazing people I met over the weekend. I’m excited to see what we can do with all the new energy created.”

In closing I wish to thank Audrey Anna Bolyard and Dr. David Dale for recognizing the need for a Family Conference years ago and for stepping up to host the first one in 2000. They started something big!

Lee Reeves is the president of the National Neutropenia Network.
Mary Ann Bonilla, MD, remembers the first neutropenic patient to receive G-CSF (Neupogen). Doctors gathered around her waiting to see what would happen. They didn’t know if she would immediately respond or if she would ever respond at all.

“For seven days we were really crestfallen,” Dr. Bonilla recalled. “On the eighth day we got a call from the lab.”

The patient’s counts were up for the first time in her life.

Dr. Bonilla, who has been with the Severe Chronic Neutropenia International Registry (SCNIR) since its inception, was able to attend her first Neutropenia Family Conference this year.

“I was very impressed by the enthusiasm and the openness of the families and the interest in what we had to share with them,” she said.

Dr. Bonilla was among the researchers who worked with Dr. Karl Welte in the early 1980s at Memorial Sloan Kettering Cancer Center/Cornell New York Hospital. The pre-clinical studies of G-CSF in chemotherapy-induced neutropenic patients were performed in association with Dr. Richard O’Reilly.

G-CSF’s ability to induce increased neutrophils then led to clinical trials in patients with congenital neutropenia. The original study grew to 12 people and then a later phase included 100 people.

“It is important to highlight that it was a project developed for only 3 or 4 people,” Bonilla asserts. “It has grown tremendously to be a movement.”

Dr. Bonilla is proud of the advances that the National Neutropenia Network and the SCNIR have made. She was encouraged to see so many people offering support, education and research information.

“It would be great not to see the support for the group dwindle again,” she said. “With every budget cut we have, we lose support.”

Currently, Dr. Bonilla holds a faculty position at Columbia University and St. Joseph’s Children’s Hospital in Paterson, N.J. She continues to follow many of the original group of SCN patients as well as practices general hematology/oncology.

Dr. Bonilla gave a presentation on “Beyond the ANC: Other Clinical Aspects of Neutropenia” during the 2007 Neutropenia Family Conference. “I realized it was a great way to clarify a lot of misunderstandings,” she said.

Later in the afternoon, she met with the idiopathic group during a breakout session to answer patients’ question and discuss the particulars of their type of neutropenia. Following the breakout sessions she met with five families for one-on-one consultations.

“I think that in general people were interested in how to alleviate symptoms such as bone pain, issues that come up with pregnancy, and the affects of G-CSF. (These) were among their main concerns,” she said, adding, “Finding a doctor to work with is a concern. This is a rare disorder a lot of physicians are not familiar with.”

**Tips From Dr. Bonilla’s Presentation**

1. Clean and treat cuts with an antibacterial cleanser such as Betadine.
2. Apply antibacterial ointment such as Neosporin, Bactracin, or Bactroban (requires a prescription)
3. Schedule dental cleanings two to four times per year. Fluoride rinses are helpful. Mouthwashes such as Biotene or those with a peroxide base are good to use.
4. Keeping areas clean is important, but it is not necessary to sterilize houses.
5. Take the child’s temperature. Don’t just say they “feel warm.” If necessary, ask medical staff the proper way to take a temperature. Use ear, mouth or under arm thermometers. No rectal thermometers.

**Physician Responsibilities**

- Stay informed of medical research
- Stay interested
- Communicate in a clear, compassionate manner
- Provide education – written and verbal
- Have adequate support staff

**Patient Responsibilities**

- Keep records
- Be compliant
- Be an advocate, not an adversary
- Ask questions and ask again
- Schedule family conferences
- Seek valid sources of information
- Be realistic

*Dr. Bonilla currently practices pediatric hematology/oncology at St. Joseph’s Children’s Hospital in Paterson, NJ.*
Dr. Dale followed Dr. Newburger with a presentation on “What We Have Learned About Neutropenia” an overview of the Registry, the data collected over the past 10 years with a look to the future.

Dr. Boxer’s presentation, “Severe Congenital Neutropenia: Pathogenesis and Treatment” provided a thorough review of the congenital neutropenias, detailing the various genetic components and risk of leukemia associated with this type of neutropenia.

The morning program for the children included fun activities supervised by Jessica Porter, recreational therapist for the Cancer Center at C.S. Mott Children’s Hospital in Ann Arbor. Children learned about blood, what it is made of and what it does. They used every day materials like marshmallows and Red Hots candies to make their own baggie of “blood.”

Before the adults broke for lunch, Jessica brought three young students into the adult conference room where they stood at the podium and explained the role of the red blood cells, white blood cells and platelets.

After lunch, Dr. Bonilla covered a wide array of topics with her presentation “Beyond the ANC: Other Aspects of Neutropenia.” Her talk covered such information as infection prevention, how to talk with physicians, understanding your counts, the need for bone marrow testing, osteoporosis and a helpful vocabulary of terms.

She encouraged families experiencing neutropenia not to isolate themselves. “No boy in the plastic bubble,” she said. “Participate in school activities, go shopping, go to the movies, plan for college and careers, date, marry and enjoy life.”

Saturday afternoon was busier than ever. Conference planners decided to sandwich in a session where all four doctors answered questions asked by the very animated audience.
"Why do I have neutropenia?"

"Why do I have neutropenia?" is what one little girl asked Jessica Porter during the children’s program at the 2007 Family Conference in Ann Arbor.

Jessica Porter, recreational therapist at the Cancer Center at C.S. Mott Children’s Hospital in Ann Arbor, thought carefully about her answer to the 5-year-old girl. Jessica related the story to Lee Reeves in an e-mail she sent Lee after the conference.

"Every parent talks differently with their child about why they might have an illness or diseases," she said.

“So I first talked about how some children are born with neutropenia, just like being born with blue eyes or brown hair. I then asked others, that were comfortable, to raise their hands if they also had neutropenia. The little girl felt a little more comfortable when she saw the hands in the air.”

Jessica talked with the children about how neutropenia is not something to be ashamed of and it is not because they or others did something wrong.

“Finally we talked about what it is and the great medicines we have to help keep us safe and healthy,” she said. “Many of the kids wanted to talk about how often they get shots and what helps them cope with the pain of the injections.”

“At the end of the day we talked about how ‘keeping clean’ (handwashing, covering cough and sneeze) and how that helps keep those around us safe … with or without neutropenia.”
Kim Blanz of Franklin, Tenn., had someone to provide backup for her when she attended the 2007 Neutropenia Family Conference in Ann Arbor — her son’s pediatrician Dr. Shelley Fiscus.

Dr. Fiscus, attended the conference with Kim to learn more about caring for four-year-old Collin who was diagnosed with idiopathic neutropenia in December 2006. After talking with the doctors attending the event, both Kim and Dr. Fiscus now believe that Collin fits better in the autoimmune category.

While Dr. Fiscus said she attended the conference to support doctors attending the event, both Kim and Dr. Fiscus now believe that Collin fits better in the autoimmune category.

When she became dissatisfied with her prior pediatrician, Kim started looking for a new doctor for Collin. A friend recommended Dr. Fiscus, and Collin became her patient in November 2006.

“Shelley was the first one that found that his ANC was way below normal,” Kim said. “She was the first one that started collecting all the information. There was a symptom here, a symptom there, but they never put them together as signaling something bigger.”

Dr. Fiscus was worried about Collin, Kim recalled. “When she looked at the ANC she said something is definitely wrong. She had never seen an ANC so low on a child of four years old.”

Soon after, another doctor diagnosed Collin with idiopathic neutropenia, which Dr. Fiscus and Kim now say is probably inaccurate. “He belongs more in the autoimmune category,” Kim said.

“His neutropenia is in the destruction process not in the production process. His bones make the neutrophils, but there is an antibody that is killing off the neutrophils in greater numbers than he can produce.”

Finding someone to support her decisions and give Collin more options has been important to Kim. Through caring for Collin, Kim and Dr. Fiscus have developed a close relationship. Their sons also are friends. Dr. Fiscus’ son, Collins, is also four, and he and Collin go to the same preschool.

Dr. Fiscus has helped Kim sort through the recommendations that doctors had previously made for Collin. “We knew that some of the recommendations were not going to be beneficial,” Dr. Fiscus said. “They were suggesting things that from a quality of life standpoint did not make sense. Their lifestyle did not need to be disrupted as much.”

For instance, it was recommended that Collin’s counts should be as low as possible; close to neutropenic. “They wanted to take him closer down to neutropenic with each dose of medicine which would then open him up to infection,” Kim said.

“When he had really low counts he couldn’t go to preschool. I feared that he would catch something at that point. It really severely limited what I could do when I thought his counts were down.”

— “It was kind of unsettling to know that there was something wrong, but you couldn’t put your finger on it. It was very scary.”

Simple activities such as running errands, attending birthday parties and going to playgrounds were out of the question because Collin’s counts were so low, according to Kim.

“He couldn’t even play with the neighborhood kids,” she said. “We live in a cul de sac that has 25 kids. I would have to keep him in. I think that truly hurt him more than anything else.”

Now that Dr. Fiscus and Kim have worked out a plan for Collin’s care, Kim does not have to spend as much time taking Collin to see other physicians for his blood tests.

“I had been taking him up to Vanderbilt Children’s hospital an hour away,” she said. “Then we would have to wait for the results. It pretty much killed an entire day.”

Now, Collin goes to see Dr. Fiscus for the blood draws. “Her office is about four miles away and that is much easier,” she said. “We go in for a CBC and within 15 minutes we are out the door and on our way back home. That has allowed Collin to form a relationship with Shelley,” Kim explained. “She actually does the blood draws on him.”

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!
**Conquering a Lifetime of Neutropenia**

In the 1980s, when Mitch Ryan was diagnosed with congenital neutropenia, doctors told his parents he did not have a good chance of celebrating his second birthday.

“When he was born studies showed that 80 percent of kids who had it did not live until age 2,” his father, Steve, recalled. “It was new territory. He was a guinea pig for them.”

Now 22, Mitch is a senior with a double major in English and history, and who plays rugby for Santa Clara State University in California. When he graduates he hopes to attend law school in Arizona.

Despite the dire early predictions, he’s rarely been sick and has only been hospitalized once in his life. He was hospitalized two years ago when he had an infection. He spent three nights in the ICU and a total of seven days in the hospital, Steve recalled.

“That’s really only the kind of close call in 20 years,” Steve said. “He’s never had a real serious life threatening situation.”

They recently learned that Mitch has developed osteoporosis but it has not changed his life significantly. He still plays ice hockey and rugby. “I just try to stay healthy and active,” he said of learning he has osteoporosis. “I have not had any problems with broken bones.”

Mitch knows he has been fortunate and was happy to talk with others during the 2007 Neutropenia Family Conference in July. “Some might not be able to break out of a cycle of despair with a disease such as neutropenia,” he said.

“I don’t like to let that hold me back. I recognize my limitations, but I try to push myself as much as I can. Despite it, I try to do as much as I can to enjoy life. I use it as a reason to try and do more.”

Steve said he knows Mitch is someone that other parents see as a shining example of how things can get better. “The thing that we have learned, of course, is that there is hope,” Steve said.

“We also know to take the dosage regimen seriously. I think he’ll tell you the only time he has really gotten down or is in trouble is when he has been lackadaisical about his dosage.”

While Mitch knows there are children and parents that look up to him, he said he does not think he deserves praise for being healthy.

“It’s like I have earned something. I don’t feel like that. I feel it is the disease I was born with and I deal with it and everything has gone well for me.”

“There are kids who had it a lot worse than me and have gone through a lot more. I am happy being able to be a visual image for younger kids so that they can see someone can grow up to have a normal lifestyle.”

He knows that talking with parents also helps them to have perspective. “It is definitely one of those diseases that can really make a parent distraught and disheartened,” he said. “They see me and they know you can still live and function and be normal or near enough.”

He said he is happy to talk with others who have questions about how he has lived with taking Neupogen for most of his life.

“I feel like I have a decent amount of experience with Neupogen and what it does,” he said. “If anyone still has questions in terms of the medication or side effects, they can e-mail me. I am certainly not an expert, but I have been through it and I can give them answers.”

To contact Mitch, e-mail him at mryan1@scu.edu.

**Kids Get Glowing View, Learn Proper Handwashing**

Children were amazed at the “germs” still on their hands after a cleanup game during the Saturday children’s program at the 2007 Neutropenia Family Conference.

Recreational therapist Jessica Porter used a product called Glo Germ™ to get the children enthused and curious about washing their hands thoroughly.

Each child received a little squirt of Glo Germ™ to rub on their hands. Then they went to the bathroom to wash it off. After they washed their hands Jessica used a black light to show them where the “germs” were.

“The Glo Germ™ represents what was still underneath their fingernails, on their wrists or on the backs of their hands,” she explained. “They could see that there still were germs in certain spots that they didn’t focus on too much.”

The children then were able to go back to the bathroom and wash their hands again to see if they got all the “germs” cleaned off.

Glo Germ™ is made by a company in Utah and sold over the Internet. It simulates the way germs spread when in contact with skin.

“It’s a good teaching tool because it helps the kids understand that germs don’t come off in running water, they have to scrub,” Jessica said. “A lot of them wanted to do it again because they thought it was cool.”

**Good Handwashing Techniques**

1. Start running the water.
2. Get a good father going. Scrub with soap for at least 15 seconds which is about how long it takes to sing the “Happy Birthday” song.
3. While singing the song, either out loud or in your head, clasp your hands and rub your palms together, let your fingers slide back and forth. Work the soap between your fingers, up to your wrists and across the back of your hands. Scrub under your fingernails if you have a nail brush.
4. Leave the water running a little while longer after you rinse off and get a paper towel to dry your hands.
5. After you use the towel to dry your hands use it to turn off the water and to open the door to leave the bathroom.

Source: Jessica Porter
Using guided imagery and art projects can help children express how they feel about having a chronic illness, according to Lorrie Carbone, a social worker from the University of Michigan and speaker at the 2007 Neutropenia Family Conference.

Lorrie talked with a number of parents about ways to get through difficult days with their children. During the session, many parents shared information and talked about what they had learned while living with a child with neutropenia. “The beauty of the conversation was that people were learning more from each other than they were from me,” Lorrie said.

Most parents told Lorrie that the hardest things to deal with are frequent shots and bone marrow biopsies. She had some advice for them. “For the younger ages, the helpful tools are imagery and medical play,” Lorrie said. With guided imagery exercises, parents can talk their children through stressful situations with relaxation techniques. Through medical play with dolls, children can act out the problems they are facing.

Using guided imagery, meditation and rewards, parents can help children through major medical events, Lorrie said. Books that tell stories of children coping with illness can also be very soothing, she added.

“Communicating about the illness and what is happening is helpful. When you read a book that shows another child going through the same thing, it presents an opportunity for the child to share their feelings and thoughts about it in a non-threatening way.”

One man who attended the session has had neutropenia since he was a child, Lorrie said. He expressed some of the things he felt growing up and what parents might need to understand for teens.

“He was very eloquent,” she said. “He said ‘I felt like I had a monster living inside of me. It is so hard to predict when you will get sick and what is going to happen.’ Older children and teens need parents to understand how they view having neutropenia. The man who shared his experiences with other parents gave some good examples of things he had gone through,” Lorrie explained.

“He would ignore symptoms and not tell his mother because he wanted to live a normal life. I think that speaks strongly to children not wanting to be different. It is often a fine line that parents have to walk with their children in doing the medical care that is necessary and living a life that is normal.”

Reva Jones knows all about struggles that come as children become teenagers. It can be difficult to get her 14-year-old son, Davon Jones, to take his twice-daily shots. She said there are moments where the two of them end up crying together on the couch about his struggles.

“She knows the importance of it, but he just gets frustrated,” she said. “He gets in his moments … he is getting older and he is getting rebellious. ‘I am sorry mom, but I just don’t want to do this. I feel hopeless.’ Then I say, ‘I understand, but you are going to take your shot.’”

Davon was diagnosed with Kostmann’s at five months old and has been hospitalized many times in his life. “It’s been really rough, but we made it through it,” Reva said. “It was just the grace of God that got us through it.”

Conference Session Teaches Parents How To Cope