Plans Underway for 2006 Family Conference

THE NATIONAL NEUTROPENIA NETWORK (NNN) will be partnering with the Severe Chronic Neutropenia International Registry (SCNIR) for an expanded Family Conference to be held in Seattle in August.

Informative breakout sessions and fun family activities will take place during the conference. Six years ago the first Family Meeting was held at the University of Washington Medical Center in Seattle and a small group of people attended. In 2005, the attendance had grown to more than 100 people.

“We realized it was time to add more speakers, topics and breakout sessions as well as have the meeting be more kid friendly,” said Erin Bogart, who is planning to help organize the conference.

Bogart is currently looking for a new facility in Seattle to hold the 2006 Family Conference.

“Our hope is to find a location that would allow the whole group to meet to hear about the latest medical updates regarding neutropenia as well as have several other rooms available for break out topics,” said Bogart.

Some ideas for break out sessions include: Q & A with Dr. David Dale; Neupogen dosing and Q & A with Audrey Anna Bolyard; a therapist to hold a special session for caregivers; break out sessions by type of neutropenia; living with neutropenia for adults; a session facilitated by a child development specialist just for children living with neutropenia; and age appropriate kids activities with adult supervision.

“We realize for many of you coming to the Family Conference is financially difficult and we will be doing our best to find reasonably priced options for meals, transportation and hotels,” Bogart said.

We would really like to hear from you about your thoughts for topics of discussion for the 2006 Family Conference. Please e-mail Erin Bogart at bogart_erin@yahoo.com with your specific thoughts about next years family conference.

We hope you will join us to make the 2006 Family Conference the biggest and best so far.
WE HAD AN INCREDIBLE RESPONSE to this summer’s newsletter, the first in many years. It clearly struck a chord with many of you who talked about how isolated and alone neutropenia can make you feel and how helpful it was to learn that others are fighting the same battles. The response underlined the need for a support organization dedicated to families and patients with neutropenia. I want to thank everyone who wrote or called.

The week the newsletter landed in the hands of the thousand-plus individuals on the Registry, I received a steady stream of powerful and touching emails. For over a month I came home to an answering machine with voices from across the country expressing appreciation, offering comments and suggestions. I hope I answered all of them but if you called and I didn’t get back with you, please call again. It was a hectic time and I don’t want anyone to think their comments are not important.

The Family Meeting was amazing. I met so many people whose lives have been profoundly impacted by the same disease that changed my life. It was wonderful to see the children laughing and playing and heartbreaking to hear some of the painful stories people shared with each other. It was great to observe first hand the comfort people derived from talking with each other about the challenges of living with neutropenia.

Several of us started a conversation about how to enhance the family meeting, how to make it more family friendly and more targeted to the specific types of neutropenia. Erin Bogart who lives in the Seattle area has offered to tackle that project. She’s exploring a new venue with activities for the kids, possibly even a play therapist to help them work through some of their issues. Erin and Audrey Anna are talking about restructuring the agenda to include break-out sessions specific to congenital, idiopathic and cyclic. If there is anyone who would like to help Erin in any way with this effort, please email her and let her know. Please refer to the story on page one.

I want to take this opportunity to thank those of you who sent donations to the National Neutropenia Network. They have made it possible to produce this winter edition of the newsletter and will help with planning for our 2006 Family Meeting. My father, William McCaffrey passed away in September and my family agreed to direct his memorial gifts to the National Neutropenia Network. My father would have done anything to help Leta and I know that he would be pleased that contributions were made on his behalf to National Neutropenia Network.

I hope this finds all who receive the newsletter in good health and spirits. I close with a quote from Viktor E. Frankl who wrote one of the books that changed my life, Man’s Search for Meaning: When we are no longer able to change a situation - we are challenged to change ourselves.

Lee Reeves
President, National Neutropenia Network
Board Member/Patient Advocate, SCNIR

Linking Families
Have you ever felt isolated or alone because of your own or your family member’s diagnosis?

LUCY LYMAN WANTS TO HELP. Lyman would like to link individuals with similar diagnosis and life situations. She would like all those who are interested to e-mail her at Lucyly3@aol.com.

“We don’t need to be alone anymore,” said Lyman.

Lyman gathered information from those attending the family meeting in Seattle in August and is working to make connections between people.

Lyman, a mother of two, has Cyclic Neutropenia. She is interested in creating a strong network for people with neutropenia. She also serves as a member of the Board of Directors for the National Neutropenia Network.
Grant Beall Enjoys an Active Life After Tough Times

GRANT BEALL SPENDS HIS DAYS like any other active 8-year-old. He rides a scooter, jumps on the trampoline with his brothers and attends the third grade. He also swims competitively and likes to watch his older brothers play football.

But it wasn’t always this way.

When Grant was younger, he was often sick, and it was noted that he had neutropenia. His life was a series of one illness after another. He was either in a doctor’s office or hospitalized 120 days before he turned 5.

For Grant, having neutropenia meant many antibiotic injections during the course of a year. In April 2002, after having received over forty injections, the leg where he had the injections became sore. His mother, Sheri, brought him into see his pediatrician who recommended that he go to the emergency room. Throughout the day the infection, which began as a small red splotch began to grow. While at Stanford’s emergency room Grant was diagnosed with necrotizing myofasciitis, or flesh-eating bacteria. His parents were told that to save Grant’s life, doctors would have to remove his leg.

“The doctor told us he had seen five cases in his 25 years as a doctor he had never seen anything move that fast,” Kent said.

While in the hospital Grant took Neupogen. He continues to take the Neupogen and his health problems have abated. “It is remarkable, the contrast,” said Grant’s Father, Kent Beall. “It is just amazing. No more sickness or ailments.”

Kent Beall is now serving as a member of the National Neutropenia Network Board of Directors. He tries to help others in his community by helping them learn more about neutropenia.

Kent remembers what it was like to try desperately to find information about neutropenia. It was a struggle to find medical journals and other information he was seeking.

“I went to my local library,” he recalled. “I discovered that they didn’t even have the publications I was looking for. I went to Stanford Medical library. I was reading everything I could read. I was looking up all the words I didn’t understand.”

Kent now works to help other parents who are trying to learn how to best care for their child with neutropenia.

“For people who have questions about what they are experiencing and what questions they should be asking the doctors - I would certainly welcome helping people,” he said. “I would be willing to talk with them and help anyway I could.”

Recently the Beall family became acquainted with the mother of a young child that Kent believes has a severe and chronic form of neutropenia. The boy’s mother attended the most recent Family Meeting with Kent and Sheri.

“You definitely feel a need, a calling to help others,” he said.

Kent Beall would like to see the National Neutropenia Network attend the American Society of Hematology annual meeting and communicate the body of knowledge the SCNIR has produced over the past eleven years. He said that he thinks the National Neutropenia Network can help to educate others about severe and chronic neutropenia. “For the residents, fellows, and others, it may be the first time that they have heard of the registry,” he said. “That is what I would like to do. I would love to relay the SCNIRs knowledge to others, and be able to help a doctor connect with the experts in severe chronic neutropenia.”
National Neutropenia Network’s 6th Annual Family Meeting

Anne Dennis, Research Assistant, SCNIR

Frank Roberts, DDS

Peggy Murie addresses concerns at family meeting.

Madison Winters, Isabella Archibald and Mackenzie Winters

Erin Bogart (right) gives token of appreciation to Audrey Anna Bollard (left) at family meeting.

Dr. David Dale talks to family members
Tips for Social Security Applicants

SOME INDIVIDUALS WITH NEUTROPENIA who experience recurring infections and other debilitating complications find it impossible to maintain steady employment. We have received calls from several individuals seeking information on accessing Social Security benefits. Melissa Serra whose daughter has Severe Congenital Neutropenia is familiar with processing disability applications. Here are some tips she offered to those seeking such benefits.

Two examples of guidelines Social Security uses for chronic granulocytopenia include:
A. Absolute neutrophil counts repeatedly below 1,000 cells/cubic millimeter and
B. Documented recurrent systemic bacterial infections occurring at least three times during the five months prior to adjudication.

Melissa urges all applicants to remember the following.

1. Everyone is entitled to apply for disability. If you think you may qualify, you should apply and don’t assume you will or will not be eligible.
2. First, Social Security makes a financial determination. If the person qualifies financially, then a medical determination is made.
3. This is a process. While Social Security has very specific guidelines to follow, there are many other areas to be considered. Mostly a person’s disability must prevent them from working and performing basic daily living activities.

Melissa’s contact information by e-mail is dmserra4@comcast.net and her home phone in Maryland is (410) 357-5475. Here is a link that may be helpful: http://www.ssa.gov/disability/professionals/bluebook†
ERIN BOGART LEARNED of her adult onset idiopathic severe chronic neutropenia amid training for her first triathlon.

She was experiencing severe fatigue at the time. Tests showed she had a very low white blood count and she was referred to a cancer specialist for consultation.

“I remember being so shocked when I called to make the appointment and realized the specialist was at a cancer center and I got even more scared when I was told to bring someone with me to the appointment!” she said. “It was the first time in my life I waited for the doctor in his office, not in an exam room.”

Bogart was told the low white counts might mean she had leukemia, an infection or virus, or aplastic anemia. A bone marrow biopsy was done that day. The week passed by slowly while Bogart waited to learn the results.

“When the test results came back my diagnosis was still a mystery . . . not cancer; that was good, but still no answer as to why my white blood cell counts were so low,” she said.

A few months later she developed a cold with a high fever and was hospitalized with a blood infection. At that time she was given Neupogen, to promote the growth of white blood cells to help to fight the infections. After the hospitalization Bogart, who lives in Seattle, was sent to a specialist at the University of Washington who quickly diagnosed her.

She learned she had adult onset idiopathic severe chronic neutropenia.

Bogart was relieved to learn about the Severe Chronic Neutropenia International Registry (SCNIR) and that by qualifying to be on the registry she would receive Neupogen for free from the drug-maker Amgen.

“It all seemed like something that was happening to someone else, it just didn’t seem real,” she said. “I had a rare blood disorder, was going to have to give myself a daily injection of a drug that cost more than $90,000 a year, I needed to go to the emergency room if I ever got a fever over 101, and at the time I was one of only about 1,000 other officially diagnosed people in the world.”

She considered the marathon her “last hurrah” as a runner. In the months and years that followed she developed several other conditions mostly falling under the category of autoimmune disorders: poly-arthritis, fibromyalgia, TMJ, carpal tunnel, IBS, and chronic fatigue. Her daily Neupogen injections are mostly able to keep her white blood cell count high enough so that she does not struggle with chronic bacterial infections.

Early on Braeden seemed to be fine, but a few days after his birth his blood counts began to drop from 6,000, all the way down to 142 neutrophils.

“We were referred to a wonderful specialist at Children’s Hospital and with his help and Audrey Anna at the SCNIR we began to monitor Braeden’s blood counts on a regular basis,” she said.

Bogart has been involved with the Neutropenia Network and will work with Audrey Anna Bolyard to plan the 2006 family meeting in Seattle.

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MORGAN SERRA was first diagnosed with Kostmann’s Syndrome when she was three months old. Her mother felt uneasy and sensed something was wrong earlier than that. When Morgan was six weeks old Melissa Serra took her daughter to the pediatrician. Morgan was fussy and seemed to be sick though there were no specific signs of illness.

The doctor ordered a blood test. “It shows she has no white blood cells.” He told Melissa, “That’s impossible. This must be a faulty reading.” A second test produced identical results. Once again the doctor said it couldn’t be right.

A few weeks later Melissa came home from work to discover a huge sore on Morgan’s eyelid. “I came home at 10:30 that night,” she said. “Morgan had a raging fever. Her eye looked like someone had punched her. My husband said, ‘I put her to bed three hours ago and she was fine.’ That’s how fast the whole process was.”

Melissa and her husband Derick learned Morgan had orbital cellulitis, which was an infection on her eyelid. She also had an irregular heartbeat. The young family soon found themselves at the University of Maryland Pediatric Intensive Care Unit.

“It was frightening,” Melissa recalled. “Everyone took really good care of us, but it was several days of not knowing.” Finally, they learned their daughter had severe congenital neutropenia. The irregular heartbeat, they were told, was Morgan’s reaction to the high fever.

“At the time they told me she would probably be 4 to 5 years old before she would outgrow it,” she said. “They assured us that Morgan would outgrow it in time.”

The Serras soon learned the doctors were misinformed, that congenital neutropenia is a lifelong condition. Over the next 18 months Morgan was hospitalized three or four times for cellulitis and pneumonia. She would start to get an infection and not let anyone know about it, Melissa said.

“She would get these infections on the bottom of her feet and not tell us,” Melissa said. “The cellulitis would move very quickly through her body. We wouldn’t know about it until it was crisis time.”

Even now that she is 10, the Serras struggle with getting Morgan to tell them when she is having a problem. She has started orthodontia work and her retainer irritates her gums. Recently Melissa checked Morgan’s mouth and found a massive infection.

“I was frustrated,” Melissa said.

Morgan’s high tolerance for pain, coupled with her dislike of going to the hospital has made for some difficult situations. Through the years the Serras have come up with ways to help Morgan cope with neutropenia.

For instance, when Morgan resisted having her nightly dose of Neupogen, a doctor suggested they set a timer. When the timer would go off then Morgan would have her shot. The timer took the pressure off Derick and Melissa and helped Morgan, Melissa said.

Melissa has often wished she could talk with other parents who have been through the same thing. “For 10 years I have wanted connect to another mom who knew what I was going through,” she said. “We come in contact with other people in our life who have children with health problems but it is not the same. I felt like I was out here on my own.”

After receiving the Neutropenia Network Newsletter in June, Melissa was spurred to start asking some questions of her daughter’s hematologist. She asked about the correlation between the gum infections Morgan was having and neutropenia. She also had questions about what she should do as Morgan approached adolescence. She had questions about whether or not acne would be a problem for her daughter. Would it bring about infections that would cause problems for Morgan? When she didn’t get a response to her questions, she researched her options and found another hematologist.

“I feel more reassured now that the information is out there and that I am going to get it,” she said. “I feel that I have people who are advocating for us and with us.”

Thank you to the following individuals for gifts to the National Neutropenia Network.

Ron and Vickie Bloxham
Vic Lopez
Lucy Lyman
Friends and Family of William P. McCaffrey
Lee and Louis Reeves
Ron and Tammy Winters
Mistaken Identity in Severe Congenital Neutropenia

RECENT STUDIES BY LAURENCE BOXER, M.D. have shown that antineutrophil antibodies can lead to misdiagnosis in infants with severe congenital Neutropenia.

Dr. Boxer said it is important to be aware that positive tests for anti-neutrophil antibodies can occur with severe congenital neutropenia. He recommended that infants with severe neutropenia and recurrent or severe bacterial infections should have a bone marrow examination and ELA2 evaluation to establish the underlying pathogenesis of the neutropenia.

Autoimmune Neutropenia of Infancy (ANI) is generally regarded as a benign condition. Diagnosis is determined by a finding of Neutropenia and a positive test for antineutrophil antibodies. Dr. Boxer evaluated SCNIR data from young children with positive anti-neutrophil antibodies in an effort to determine whether the occurrence of the antibodies in infants predicts the more benign expression of the disease.

The study evaluated eight children who presented with severe infections including: cellulitis, recurrent ear infections, pneumonitis, bacteremia, abscesses and mouth ulcers. The children tested positive for antineutrophil antibodies and were thought to have the more benign autoimmune Neutropenia. Further studies however, showed a mutation in the neutrophil elastase gene (ELA2) in some and bone marrow patterns in all consistent with severe congenital neutropenia which indicated the diagnosis of autoimmune neutropenia was not valid.

“It is important to evaluate the bone marrow when a child presents with the severe infections and a positive antibody test. We can’t assume that the child has the less severe autoimmune Neutropenia of infancy,” Dr. Boxer said.

Erin Bogart

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Bogart and her husband were told Braeden probably had a “transient” form of neutropenia that would most likely disappear within 3-6 months.

“His first real cold and ear infection didn’t come until around his first birthday and his body was able to mount a response to the infection and his blood counts raised high enough to fight off the infection,” she said.

Although his counts still run low, he has never had to be treated with Neupogen.

“He is extremely healthy and able to fight off the few illnesses he has had,” she said. “We still hope and pray his counts will normalize and he won’t have to deal with neutropenia for life. He is now 21 months old and a happy little guy.”

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Erin Bogart

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