



# Neutropenia Support Assoc. Inc.

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## Neutrophil Research Helps Children with Cancer

Six-year old Lindsey Stevens was born with a rare disease called Neutropenia. Simply put, patients with this condition can not produce enough white blood cells which are the body's front line of defense against bacterial infection.

Although rare, the disease can be life threatening and patients require constant medication to help them fend off infection.

"Lindsey had several infections as a baby and always responded to antibiotics but somehow we knew something was wrong" Mrs Stevens said.

At 18 months, he was admitted to Children's Hospital with croup and his white blood cell count was zero. After six stressful weeks of tests, specialists in blood disorders determined Lindsey's problem.

Now with Foundation support, Dr. Bonnie Cham is establishing a new lab to study neutrophils, the white cells primarily responsible for fighting bacterial infection. She will study the location of certain proteins within the neutrophil and the effects of a promising new drug called GM-CSF which helps patients to build up their white blood cells. Children with cancer will also benefit from Dr. Cham's research. They have low neutrophil levels as a result of chemotherapy.

Neutrophils necessarily contain destructive proteins to enable them to fight infections. But when they get out of control, they can also start to destroy joints and lung tissue.

With more understanding of how neutrophils work normally and how they produce all of these toxic proteins, researchers may be able to control their damaging effects in certain diseases such as arthritis and lung disease.

"Working with children is wonderful but it's hard when you're seeing a patient with a non-curable disorder. It helps to know that research you're doing in the lab may offer some promise for other patients in the future" Dr. Cham said.

Neutropenia, because of its rarity, is not widely known. Lindsey Stevens' parents have spearheaded a fundraising campaign so that extensive information is now available in the library at Children's Hospital for medical staff and families of affected children. Their group has also donated \$7500 to help Dr. Cham set up her lab.



### Dr. Cham shows Lindsey neutrophil samples in the lab

We feel we are very fortunate to be in Winnipeg because of the exceptional medical care available to us "Mrs. Stevens said. " We were diagnosed early and Lindsey is presently manageable on antibiotics. It scares us but it is reassuring to know that research is going on here that will improve his future". (courtesy of Teddy News)

### A Letter from Dr. Cham

At the present time, I am involved in research to study the effects of GM-CSF on neutrophil function and am also involved in studying the location of certain proteins within the neutrophil. Our goal is to be able to set up a lab in which we can investigate blood of patients with neutrophil disorders, monitor their response to therapy, and continue to do basic science research into the mechanism of action and activation of neutrophils.

As a group interested in neutrophil disorders we approach you to help us support this venture. Cheques payable to the University of Manitoba Neutropenia Research should be sent to our organization. Any support you can provide will be greatly appreciated.

## Message from the President

In 1989, the Neutropenia Support Association Inc. was incorporated in an effort to help families in need of assistance. Our focus is two-fold. First and foremost, our aim was to initiate Canadian licensing studies for the drug G-CSF which has shown miraculous results in the treatment of neutropenia patients and secondly, to raise money for research, and education.

We are happy to report that through our efforts, a great deal of progress has been made. Amgen Inc., the pharmaceutical company which manufactures G-CSF applied for licensing in Canada on June 28, 1990. The status of their U.S. application is very optimistic. Another drug that can be used for short-term emergency use is GM-CSF. This drug is produced by Schering of Canada and approval in Canada is imminent.

Donations totalling \$7500 have been presented to the University of Manitoba Neutropenia Research Grant. Recipients were Dr. Bonnie Cham and Dr. Jon Gerrard.

A further \$2,540.00 was given to Ada Ducas, Director of Educational Resources, for the Children's Hospital Paediatric Medical Library. Funds are being specifically allocated for rare blood disease information such as neutropenia.

Future Newsletters will highlight related research and be distributed world wide. We are the only Neutropenia Support group in existence.

We presently are looking for a Newsletter "Sponsor".

We wish to extend our thanks to the following:

The Transcona Optimist Club, MPIC co-workers of Brian Gamley, Contemporary Printers, Cooper Business Forms, Delbro Real Estate, Audrey Carlson, CJOB, CBC, CKY, Winnipeg Free Press, Winnipeg Sun and Gerri Thorsteinson of Teddy News, Amgen Inc., Schering and Sandos for articles and information. Dr. Nathan Kobrinsky, Dr. Jon Gerrard and to all of those whose contributions have made our worthwhile achievements possible.

## Words from Dr. Jon M. Gerrard

I write to thank you for the tremendous effort you and others in the Neutropenia Support Association have made in helping to increase the awareness and understanding of a very important white blood cell, the neutrophil. As you are well aware, when neutrophils are deficient, a condition called neutropenia occurs. Children and adults with this condition are very susceptible to infection. This is a rare congenital problem, but it is a problem that we face also daily as a result of the chemotherapy that we give to children with cancer. There are also a variety of neutrophil abnormalities in which the function of the neutrophil is below normal and which are associated with an increased predisposition to infections. I think your efforts have been very important in highlighting these conditions, and in facilitating the development here of a neutrophil function laboratory run by Dr. Bonnie Cham. As you are aware, this is in its very early stages at the moment, but I think we can be very optimistic, in part because of the wonderful support that you have provided.

## Important News FLASH !!

### **DRUG CAUSES COMPLETE REMISSIONS IN DEADLY BLOOD DISEASE STRIKING CHILDREN, U-M RESEARCH FINDS**

An experimental treatment at the University of Michigan Cancer Center and seven other centers around the country was nearly 100 percent effective in causing complete remissions in children with a rare, deadly blood disorder.

The results of the 1 1/2 year study, according to the director of the program, are "phenomenal."

Boxer and other physicians treated the children with injections of a drug called Granulocyte-Colony Stimulating Factor, or G-CSF., which raises the body's white cell count to normal level.

Without this drug, he said, many of the children in the study would have died from overwhelming infection.

"This is probably the most exciting bit of clinical research I've ever done," said Boxer. "The results are stupendous. It is not often in medicine that you can completely turn around a patient's life.

Researchers are now awaiting approval of the drug from the Food and Drug Administration, which may come in about one or two years. Once the FDA gives the OK, the drug will be available for general use by haematologists throughout the country.

## Letter from Brian Gamley

Around the beginning of January, 1990 I was fortunate to catch a C.B.C. 24-Hours broadcast involving the newly formed Neutropenia Support Group. I heard families and a doctor talking about the disease I had been diagnosed as having - Neutropenia - because it is a rare blood disease with few known sufferers, I called C.B.C. and they referred me to Dr. Gerrard who in turn referred me to the Group.

The story doesn't start there, however. It started when I was roughly 12-13 years old. I had a multitude of oral and rectal canker sores, colds developing into pneumonia, gum infections and blood infections which were brought upon by something as tiny as a scrape. Now, at the age of 37, after seeing a total of 17 doctors, including general practitioners, internists, surgeons, dermatologists, neurologists, immunologists, haematologists, dentists and denturists; after a wide assortment of medicines, many of them with harmful and dangerous side effects, after rectal surgery and after having all of my teeth removed, I was referred to the Mayo Clinic. There, they diagnosed Neutropenia in January, 1988. They told me at that time there were only five known cases in North America.

Under the monitoring of a Manitoba haematologist, I have been getting by - taking two types of medicine to keep the infections down. While that may be okay to some, one of the medicines is prednisone known to have severe long-term side effects; the other can cause liver damage.

After seeing the C.B.C. broadcast though, and attending the Neutropenia Support Association meetings (now a registered charitable organization), I have new hope. There is research being done at the Children's Hospital in Winnipeg - specifically in research of neutrophils. Dr. Gerrard, although a paediatrician, has taken an active interest in Neutropenia and has been helping me with my case. We have news of a new drug being tested and soon to be approved in the U.S.; a drug called GCSF and another similar GM-CSF, which will help neutropenics develop their own neutrophils which their own bone marrow cannot seem to do. But equally important, this same drug may ultimately help cancer and Aids patients in that this medicine may bolster up the immune system.

So there is hope and what is more, there are more sufferers out there with Neutropenia - it is very often misdiagnosed and mistreated, as in my case. The medical community must be made aware of the symptoms and, hopefully soon, the proper treatment will be approved and implemented. The general public could become enlightened through education. If I had not watched C.B.C. and caught that broadcast, I would never know what I know today.