



# Neutropenia Support Assoc. Inc.

P.O. Box 243, 905 Corydon Ave. Winnipeg, Manitoba R3M 3S7  
Local Winnipeg # 489-8454 Toll Free # 1-800-6-neuro (1-800-663-8876)

President: Lorna Stevens (204) 989-5000 E-mail: STEVENSL@NEUTROPENIA.CA Directors: Janis and Jim Benzelock  
Vice President/Editor: Mike Carlson E-mail: CARLSONM@neutropenia.ca Treasurer: W. Lorne Stevens Legal Advisor: R. Stefanyshyn  
Canadian SCN Disease Registry: Dr. Bonnie Cham Dr. Melvin Freedman Medical Advisor: The Hon. Dr. Jon Gerrard

## Congratulations go out to CJOB

*From the Volunteer Centre of Winnipeg...*

On behalf of the Volunteer Centre of Winnipeg I would like to congratulate you on nominating one of the successful candidates for the 1995 Golden Hand Award. The panel of judges has chosen CJOB as a worthy recipient. The award was given out April 26, 1995 at the Westin Hotel.

Ted Farr accepted this award on behalf of CJOB, Lorna Stevens presented the 1995 Golden Hand Award on behalf of the Neutropenia Support Assoc. Inc.

CJOB was nominated by both the Canadian Diabetes Association as well as the Neutropenia Support Assoc Inc.

Five years ago, the recently formed Neutropenia Support Association, was in need of exposure. There were vital issues facing those who suffered from Neutropenia and they needed to be addressed in an effective and positive manner.

CJOB was responsible for offering us the means to make our concerns known. Peter Warren has always been supportive and generous with his airtime. He deserves credit for being the first to broadcast our message and that of our medical advisers.

Although a small group, we have always had lofty goals but lacked the resources to reach a large audience. CJOB provided us with an all important first opportunity to reach not only those in the position to help us, but those who would benefit through a greater understanding and awareness of a unique and rare blood disorder.

Neutropenia is a potentially life threatening disease. Over 50% of the families that have called for information and ultimately were diagnosed with Neutropenia, came to us because of CJOB's dedication and compassion to our cause.

Thanks to your ongoing support and coverage, we continue to receive calls from across the province and as far away as the North West Territories.

The Hockey Hutch here in Winnipeg recently had a customer appreciation day in conjunction with a fund-raiser for the Neutropenia Support Association Inc. We

strongly feel that the success of the event can be attributed to CJOB's active participation.

We would also like to thank Paul Graham who provided us with the opportunity to wrap up our year with a report "Live from the Forks". ■

#### Editor's note:

Jim Benzelock of CJOB, who has also made outstanding volunteer efforts, was recognized at our last Fashion Show Luncheon. He has assisted with numerous events. Thanks to ALL at CJOB!!!



*The Golden Hand Award for Manitoba Media. Left to right: Lorna Stevens N.S.A.I., Ted Farr CJOB/97.5 FM, Susan La Bossiere, Canadian Diabetes Assoc.*

# Customer Appreciation Day at The Hockey Hutch

All of us at the Hockey Hutch would like to extend our warmest appreciation and thanks to all of our customers who took part in our Customer Appreciation Day Celebration. In total, \$1850.90 was raised for the Neutropenia Support Association. This money will be used by the University of Manitoba's cell biology program for research on neutropenia. Even more exciting news about the awareness campaign is that a family was found with a 4 year old boy who has neutropenia. This family has been put in contact with the Neutropenia Support Association and will benefit from all the services it provides.

The following people were the raffle prize winners:

**Quinn Taylor**, winner of one of the Easton aluminum sticks autographed by Wayne Gretzky.

**Peter Court**, winner of the D&R Centurion shin pads.

**Matt Walls**, winner of one of the Easton aluminum sticks autographed by Wayne Gretzky.

**Stephen Locke**, winner of the Titan hockey stick autographed by Doug Gilmour.

**Daniel Edmonds**, winner of the Winnipeg Jets Pace Setter Center Ice warm up suit.

**Thomas Law**, winner of a CCM NHL authentic jersey of his choice.

**John Matas**, winner of the Koho hockey bag.

**Adam Rodin**, winner of the Itech Sports Bag.

**Terry Barlow**, winner of one pair of Bauer 4000 hockey skates.

**Kristin Locke**, winner of the Winnipeg Jets Starter jacket.

**D. McKeigan**, winner of the Koho hockey stick autographed by Mario Lemieux.

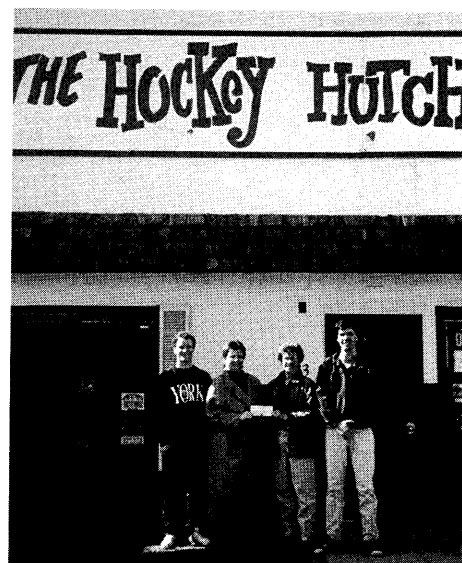
Thanks again for all your support over the last week and the last 20 years.

Comments from a letter received from The Hockey Hutch

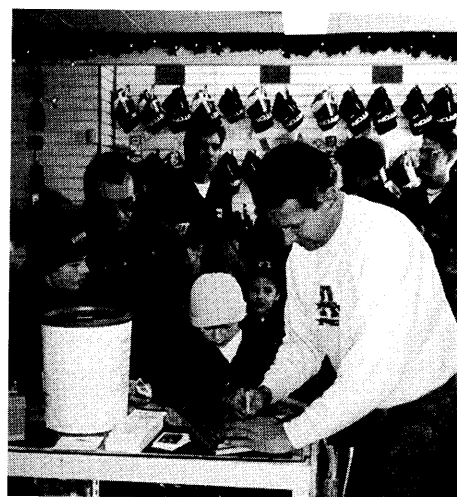
Our customers were pleased to be able to help out the Neutropenia Support Association and many expressed the fact that they had not known what Neutropenia was and appreciated learning about the disease.

Donna, Jeff and myself are especially pleased that a family with a 4 year old who has Neutropenia was found. The satisfaction derived by this fact alone, is truly a wonderful reward for the hard work that so many people did in pulling off this event. This type of promotion allows us to step outside of the retail world for a brief moment. So often in the business world what a business adds or takes away from the bottom line is the difference gained by promotional events. However, a promotion like this one for our customers and the Neutropenia Support Association offers reassurance that you can make a difference in the community when you try.

Donna, Jeff, myself and all the staff at the Hockey Hutch have been honored at having the opportunity to help support your association. We have no doubt that the new family and their 4 year old will benefit from the Neutropenia Support Association. We wish you continued success and will see you at your next event in support of neutropenia. ■



*The Hockey Hutch presents cheque to Dr. Bonnie Cham*



*Bombers and Jets alumni sign autographs.*



*Donna Smyth cooked hundreds for customer appreciation day.*

# Trucker Hockey Tournament and Winnipeg Jet Alumni Team Together for Neutropenia

On February 2, 1995 an exhibition hockey game between the Trucker All Stars and the Winnipeg Jet Alumni was held. There was no charge to watch the game but donations were accepted for the fight against Neutropenia.

Neutropenia ( new-troh-pee-nee-ah) is a side effect of chemotherapy which affects as many as one in four patients.

The neutrophils are the main white blood cell defense against infections. Chemotherapy kills these Neutrophils as well as cancer cells. There are different types of neutropenia, the most common symptoms are: fevers, mouth sores, ear infections and pneumonia. The mouth, a major entry point for bacteria, can be a real problem area.

Chemotherapy treatments may have to be delayed until the body can produce more neutrophils and a lower dosage may have to be given, which may make treatments less effective. The longer the neutrophil levels remain low, the greater the chance of infection with potentially seri-

ous results. If infection develops, antibiotics are given and hospitalization can occur, with an average stay up to 10 days for each potentially life threatening occurrence. Each occurrence of the infection saps the strength of the patient and makes it harder for the doctor to treat the disease.

New drugs have been produced which assist the bone marrow to make neutrophils and help restore the body's ability to fight against infection. These drugs may improve the lives of patients with Neutropenia by decreasing the severity of the infection and reducing hospitalizations. While these drugs are expensive, they are less expensive than hospital stays.

With events such as the one mentioned above, and possibly more people and organizations getting involved, maybe one day the fight against Neutropenia will be won. ■

**Editor's note:**  
**\$712.96 was raised at this event. Thanks to all who participated..**

## "Giving is Rewarding" *Five Years a Charity!!!*

Together we have made a contribution and offered a message of hope to the children and families with Neutropenia.

The N.S.A.I. is realizing its mandate through the partnership that includes volunteers, donors, and associations. Thank You!!

We are inspired by the determination and courage of families experiencing difficulties. We want to help make their lives a little easier by providing a much needed information resource.

By supporting the efforts of our medical advisor-researchers, we will together, in our lifetime realize the ultimate dream....

## 'Meet And Greet' was a real Treat!!

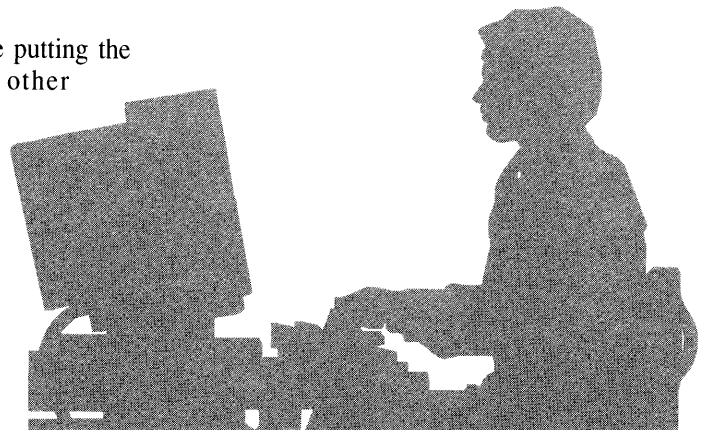
Our Meet and Greet meeting held Tuesday April 11, 1995 was a treat. Guest "celebrity" was Dr. Bonnie Cham. Thanks to you for attending, and thanks to all who participated. We welcome all diagnosed families to call so we can organize a meeting time convenient for your needs. ■

## Neutropenia Support Assoc. Inc. Goes Online

The Neutropenia Support Assoc. Inc. is now on the INTERNET or commonly known as the information super highway. If you have any comments questions, or suggestions, please feel free to send us E-MAIL at STEVENSL@neutropenia.ca. We would love to hear from you.

If you have any comments, suggestions, or articles for the newsletter, please send them to CARLSONM@neutropenia.ca. We look forward to hearing from you.

We will also be putting the newsletters and other tid/bits of information online for all to see and read. Watch for further details on this. ■



# Third Annual Fashion Show Big Hit!!

Your contribution enables us in our goal to support Neutropenia function research. We raised \$4,200.00 at our 1994 Fashion Show Luncheon. We also gave \$500.00 to the Firefighters Burn Fund.

Please accept our thanks!

Caring people like you help us to fulfill our mandate.

The Neutropenia Support Association Inc. is a registered Canadian Charity, formed in 1989 to increase awareness and understanding of neutropenia. It provides assistance to Neutropenia patients and their families and raises money for research and education.

The N.S.A.I are 100% volunteers and the only such charity in the world.

## Mark Your Calendar

*The Neutropenia Support Assoc. Inc. would like to "thank" ALL our volunteers and contributors!*

*Our 4th annual Fashion Show Luncheon is October 29th, 1995, featuring:*

*Dr. Melvin Freedman Chief Haematologist at the Toronto Hospital for Sick Children, also a variety of special guests and attractions.*

*See you there!!*

Article from the Metro, Winnipeg, Manitoba

## Event raises funds for support, research

By Brenda Fleming

When all of the figures are in, it's expected \$4-5,000 will have been raised toward the fight against neutropenia, says Neutropenia support association president Lorna Stevens.

Neutropenia is a blood disorder which can be fatal.

The Neutropenia support association was founded in 1989 and is based out of Stevens' home in River Heights.

The association recently held its major fund raiser of the year, a fashion show and luncheon at the Radisson Hotel, which sold out.

In addition to a fashion show, the event included a presentation by cancer researcher Dr. Laurence Boxer and a performance by escape artist Dean Gunnarson.

Boxer said research and education play an

important part in tackling neutropenia. Support groups like the Neutropenia Support Association also are important, he said. The association formed in Winnipeg is serving as a model for other support associations being set up in the United States, he said.

Funds raised from the event will go toward research and other Neutropenia projects.

*Photo: Escape artist Dean Gunnarson entertains at the fund raiser. Helpers are Lorne Stevens and John Isbister.*



*Above: Dr. Laurence Boxer, our guest speaker*

*The Third Annual Fashion Show "sold out"*



# Editor's Info Tid/Bits

Patients using Neupogen with a history of psoriasis may develop a flare up of his or her skin. Cutaneous vasculitis could be a similar immune system activated response. Researchers at the National Psoriasis Tissue Bank pin-pointed the location of one gene possibly responsible for certain types of psoriasis. This gene is located on chromosome 17 of human DNA. Other involved genes are believed to be located on chromosome 6.

weigh the adverse consequences. Bone marrow transplant may be an option for the patients who are at the highest risk.

Gene transfer research by Dr. Metcalf, showed that simple overstimulation of otherwise normal cells is not enough in itself sufficient to result in leukemia transformation. The Neutropenia Support Association supports repeat bone-marrow examination yearly with cytogenic analysis. Comparison with

## Up and Coming Events

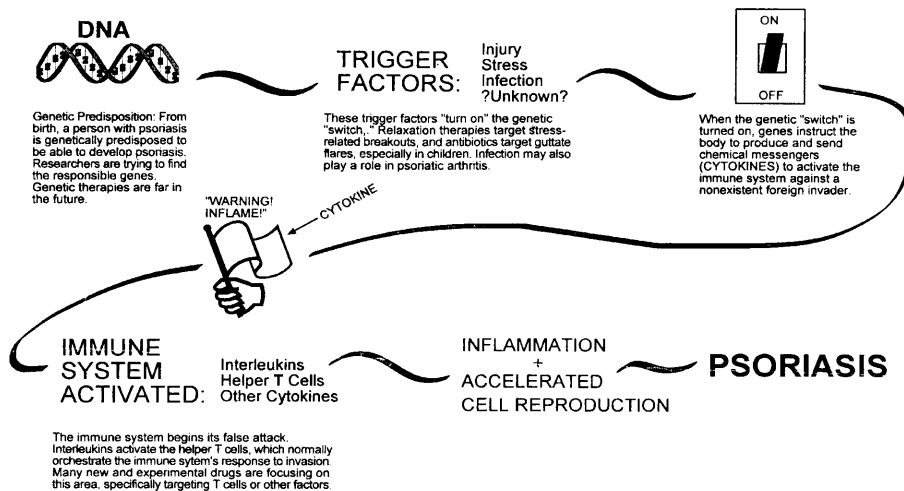
The Neutropenia and Children's Wish Foundation CJOB/97.5FM **"Poor Man's Golf Classic"** September 21, 1995...Transcona Golf Club...book your 4-some at the Pro shop 222-6171... Great prizes, fun, and karaoke!!

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Our next Family get-together is tentatively planned for September. Please call for details...

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**TARGET AREAS OF PSORIASIS RESEARCH:** Researchers are examining these factors that may cause psoriasis in their search for better psoriasis treatments.



Physicians treating chronic granulomatous disease, please call us. A support group network is forming. 1-800-6-NEUTRO.

An important question is currently being addressed in a multi-institutional clinical trial. Do hematopoietic growth factors alter the risk of evolution to AML (acute myelocytic leukemia)? The patients (5%) to date may have had high risk with an inherent predisposition of progression to AML. It is still unknown whether the pace of transformation is related to Neupogen (G-CSF) therapy. Until this question is answered the benefits of treatment for SCN patients far out-

weigh the adverse consequences. Bone marrow transplant may be an option for the patients who are at the highest risk.

Neutropenia is also a consequence of Shwachman-Diamond Syndrome, Chediak-Higashi Syndrome, congenital aplastic anemia, Fanconi's anemia, myelokathexis, lazy-leukocyte syndrome, neonatal isoimmune neutropenia, glyco-gen storage disease (see feature article in this issue). ■

**Shwachman-Diamond Syndrome Support Group formed 1995!**  
Contact: Joan Mowery, 44 Meadowlark Rd., Vernon Conn. U.S.A. 06066 Ph. 203-870-5454.

## 1995 Golf Tournament

Early Registration  
6th Annual  
Winnipeg Jets Alumni  
Heritage Golf Classic  
To Benefit Neutropenia and Kidsport

### THE TOURNAMENT:

Thursday, June 22, 1995  
Kingswood Golf course  
Shotgun Start 1:00 p.m. (be early, there will be a pre-golf snack)  
Dress Code in effect

### FORMAT:

4 players per team  
Texas Scramble  
Full Teams preferred  
Partial foursomes and individuals accommodated

### GOLF FEE: \$100.00

Included: registration, snack, golf fees, golf cart, supper, grab bag, tax receipt, Rainbow Auction, 4 par three hole prizes, and more...

### MULLIGANS:

\$5.00 per, a must for each player

### REFRESHMENTS:

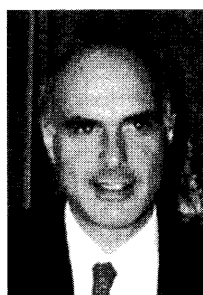
Available around the course

### ALUMNI CONTACTS:

|                 |            |
|-----------------|------------|
| Jordy Douglas   | 1-885-6865 |
| Gordie Tumilson | 233-8558   |
| Duke Asmundson  | 474-1336   |
| Ron Loustel     | 943-9526   |
| Gerard McDonald | 982-5413   |
| Perry Miller    | 956-2333   |

Feature Article from Dr. Melvin H. Freedman

# Recombinant Human Granulocyte Colony Stimulating Factor for Treatment of Neutropenia and Recurrent Infection in Patients with Glycogen Storage Disease Type 1B



*Dr. Freedman*

Glycogen storage disease type 1B is an inherited disorder. The consequences of this disease are low blood sugar when fasting with seizures, often occurring early in infancy, liver enlargement, poor growth, and poor development if the low blood sugar level and seizures are not treated early and aggressively. These patients also have Neutropenia and/or neutrophil dysfunction resulting in frequent recurrent pyogenic infections. The exact etiology of the neutrophil defects in this disease are not clearly understood. Frequent hospitalization with life-threatening infections have been a way of life for these patients.

Until recently, the management of these patients has been entirely supportive. Most patients have a stomach tube inserted in order to receive continuous feeding overnight to avoid low blood sugar. Attempts have been made, to treat bacterial infections early and aggressively. Prophylactic antibiotics have also played a role.

Roughly 18 months ago at The Hospital for Sick Children, we initiated a trial of recombinant human G-CSF to treat Neutropenia and neutrophil dysfunction. Patients from around Ontario and as far away as Calgary and Vancouver have been enrolled on this trial. A total of eight

patients are currently enrolled, six of whom have been on trial for at least one year. Results to date on those six patients are extremely encouraging. We have noted improvements in neutrophil counts with most patients achieving neutrophil counts in the normal range. We have also noted improvement in vitro in neutrophil function, as marked by bacterial killing. This has translated into improved clinical status in these patients. Many of them have not had a serious infection in the year while on therapy, and the one patient who has continued to have episodes of cellulitis feels subjectively that these are much less severe than they have been in the past. In addition, these infections seem to resolve much more rapidly on antibiotics. Compliance with drug therapy in all patients has been excellent. No patient has experienced a significant discomfort or side effect from the treatment to date. The families are uniformly pleased with the results they have seen in their own children.

Based on these early results, the trial will continue for three years for each patient enrolled. If you should require any additional information about this study, or know of a patient who may benefit from this trial, please contact either the principal investigator of the trial, Dr. Stan Calderwood at 416-813-5872, or Dr. Mel Freedman at 416-813-6152. The trial is open to adults as well as pediatric patients.

■

## Doctor Freedman sends thanks!!

I have just received the generous cheques in support of Neutropenia research. I have been gearing up to specifically address the issues of SCN and the development of leukemia to determine who, when, and why. The funds will be a valuable resource for this important research initiative. Thanks so very, very much for your continuing efforts and support.

### Editor's Note:

The 1994-1995 funds to date sent to Dr. Melvin Freedman from Manitoba, Ontario, and Newfoundland total \$8110.00..

## Also from Dr. Melvin Freedman

Suggestion: as you raise increasing amounts of money you may wish to give some thought to having a medical or scientific advisory component that can offer advice as to the best way to distribute the funds. If you raise enough, you may want to open up the funding for competition so that researchers apply with their proposals. ■

## ONTARIO NEWS

# Ontario Drug Program Branch meet with Neutropenia Support Assoc.

**L**orna Stevens, as President of the Neutropenia Support Association has attended a series of Ontario Drug Programs Branch meetings. She provides these comments and observations.

The Neutropenia Support Association Inc. wants to see the successful implementation of the "Catastrophic Coverage" of medicines for the 20% of Ontario families without insurance benefit. We were surprised to find out 45 million has been "grandfathered". The remaining 75 million may not be an accurate assessment for this plan plus management (extra staff, accounting, pharmacy technicians). With catastrophic coverage available through this Ontario government Trillium Plan, employees may "opt out" of their company sponsored health plan. This could create a negative impact and greater deficit for Ontario to manage.

Cost effective treatment is being ignored because the pharmacists are being asked to cut costs (period) without taking the success of "High Tech" medicines that work for a common problem into account.

We have recently heard a "pharmacological impact" study will be funded by Federal and Provincial Ontario governments. Neupogen is one of the major drugs being reviewed.

Government bureaucrats suggest the need for double blinded chemotherapy induced pediatric studies to provide proof of cost effective treatment with cytokines. Challenging further the lives of children to provide more data is not acceptable. As an example, over 200,000 patients have used neupogen. It is an approved drug in Canada (3 indications), United States and other countries around the world.

The need for a pediatric expert advisory committee to assist the Ontario Government with information has definitely become apparent.

The 1995 Neupogen Guidelines are very general, are primarily for adult disorders which aren't for the most part for children, and thus, not much use. The physicians to our knowledge want spelled out in as much detail as possible when and when not to use (G-CSF) Neupogen for children. We have also requested that severe chronic Neutropenia be included in the "Box" as an approved use. Neupogen is the only treatment presently approved by the Health Protection Branch for SCN patients.

*Can it be that the use of  
"life saving" drugs are  
being slowed down by  
making these drugs  
less accessible?*

P.O.G.O. and Candlelighters join with us in addressing the needs of chemotherapy induced Neutropenia and cytokine treatments. Dr. Eleanor Pask (Candlelighter's) and Dr. Corin Greenberg (P.O.G.O.) joined Lorna Stevens (NSAI) at the March 1995 ODPB meeting in Toronto.

All three associations are part of the Ontario Childhood Cancer Alliance, created March 29th, 1994.

They will continue to communicate closely with the Drug Programs Branch and monitor the implementation of "Trillium".

Many issues have been outlined by Dr. Corin Greenberg (P.O.G.O.) and presented to Ms Mary Catherine Lindberg,

Assistant Deputy Minister, Ontario Ministry of Health.

Can it be that the use of "life saving" drugs are being slowed down by making these drugs less accessible?

In one particular instance, a patient was hospitalized for a total of 13 weeks experiencing coma and blood poisoning. Her husband works, but does not enjoy the benefit of health insurance. The physician applied for use of Neupogen under the Section 8 Individual Request Ontario Drug Benefit Program.

The Drug Quality and Therapeutics Committee carefully considered the clinical information described in the physician's request. The Ministry approved the request October, 1994. The letter sent back to the physician requested "eligibility" from ODB with no instruction as to how to accomplish this process. As of March of 1995 "Eligibility" had not been determined. Most case workers with Welfare Canada and ODB employees when contacted are unaware of the "Special Benefits", "Special Assistance" category. This category is "an exception to the rule" to assist "life saving" financial needs. The patient's family is "audited" by a case worker to determine "eligibility". The physician needs to supply the estimated cost of the treatment to the case worker as part of the "formula" for "eligibility".

Hospital stays can average \$1,000.00 a day. The drug treatment (expedited) could have saved taxpayers as much as \$72,000.00 by avoiding the extensive hospital stays.

Thankfully, her brother found the Neutropenia Support Association Inc. listed in the Toronto Yellow Pages under "charities".

Lorna Stevens, while in Toronto for an ODPB meeting, visited her in hospital. The paperwork was completed and "eligibility" attained within 48 hours. The



## ONTARIO DRUG PROGRAM (cont'd)

patient is now out of hospital.

We strongly recommend that you contact our association if you are experiencing any delays or difficulties with your application under Section 8.

*"Thank you for your  
help in getting us  
through the bureau-  
cratic jungle"*

"Trillium" should help... With the expectation of the Trillium Plan receiving an estimated 240,000 applications yearly, "Support Group" advocate assistance may be needed. We recently received a letter from a physician stating "Thank you for your help in getting us through the bureaucratic jungle".

We are careful to maintain patient and physician confidentiality. We are only a toll free number away, **1-800-6-NEUTRO!** We welcome your calls! ■

# Pediatric Oncology Group of Ontario

*The following are excerpts from P.O.G.O. to N.S.A.I.*

Congratulations on your efforts thus far regarding the drug safety-net consultations with Ms. Mary Catherine Lindberg of the Ontario Ministry of Health Drug Reform Secretariat.

As you suggest, it is valuable to coordinate our efforts, and the ongoing consultations offer a very important opportunity to identify the generic issues of drug costs for families of children with cancer - where out-of-pocket expenses, chronic course of care and frequent loss of one income are well documented and add up to a very unique financial stress.

One fairly recent Ontario study (based on 1986 data) estimates that, for families of children with cancer, one-third of the average family's after-tax income may be expended on out-of-pocket cost related to the child's disease and its treatment. This finding is consistent with earlier studies emanating from the U.S.A.

Among the drugs that are a major financial concern to parents and, therefore, candidates for this fund in the pediatric oncology context are:

- cytokines, which promote survival by decreasing septic episodes and allow-

ing chemotherapy to go ahead

- anti-viral agents, e.g. acyclovir which is life giving in the event of herpes/chicken pox

- anti-fungal agents, e.g. fluconazole, which allow children to survive fungal infections occurring during chemotherapy

- anti-emetics which reduce adverse reactions to chemotherapy, and decrease dramatically the amount of nursing care required, as well as the time off work it is necessary for parents to take

- symptom control medications such as specialized mouth washes to prevent oral infections

- antibiotics given at home are not easily covered

- oral chemo-therapeutic agents delivered in the home are also candidates for safety-net coverage. ■

**"Trillium packages" available  
to our Ontario families  
1-800-6-NEUTRO**

**From The Neutropenia Support Association Inc. Ontario Chapter  
Submitted By Shirley Cox.**

## ONTARIO UPDATE

Just a short note to keep in touch. Hope all are well and are enjoying the spring season.

1994 was fairly successful from a fund raising point of view. A very big thank you goes out to Mrs Doris Cassan, whose daughter has Neutropenia, and in the past year has raised more than \$1500.00 with a promise of more to come. In addition to her wonderful fund raising

through the Portobell Market, which is a monthly Antique Show held at the Thornhill Community Centre 7755 Bayview Ave (at John St.), she has been handing out our Neutropenia - Causes, Consequences and Patient Care booklets. Great job, and a sincere thank you from all of us, to all of those who support her.

Another big thank you goes out to Glenn and Corrie Rogers and the wonder-

ful people at C.N.. The Rogers will be featured in an up and coming article about Neutropenia in one of the Scarborough papers and across Canada in the C.N. employees newsletter. Did you know that each time there is media coverage of any kind at least one new family is identified? Please contact your local newspaper, you could save somebody's life.



## BIG NEWS;

Eric Nagler a very well known children's performer, who appears with Sharon, Lois and Bram has agreed to help our cause. Eric will do as many concerts as we can organize for him. If anybody is interested in organizing a concert in your area, please contact us for further details. Your local children's clothing store usually will help sell the tickets! People with children in the elementary schools can approach the school about sending out notices to all children. A large ad in the local paper is also important to help sell tickets at the door.

## LIFE INSURANCE PREMIUM REDUCTION

London Life Insurance Company's underwriters and company doctors have been asked to review and re-assess its policy for insuring Neutropenia sufferers, and has elected to downgrade Neutropenia patients who are being successfully controlled by neupogen treatment. This means a substantial reduction in monthly premiums for those who already hold a policy with London Life and the availability of affordable insurance for those not able to obtain insurance in the past because of the underlying medical problem. London Life's controlled Neutropenia patients will be now classified and categorized the same as a smoker. All insurance companies should, when asked, be following London Life's lead.

## DONATIONS

The Rotary Club of District 7070 in Alliston, Ontario recently invited Shirley Cox to talk to them about Neutropenia and the support group's activities. The Rotary Club shows a real desire to make life better in communities by supporting various international and local programs. Proceeds from an upcoming urban community night will be donated to the Ontario Chapter of the Neutropenia Support Association Inc. Several business owners volunteered to house donation cans in their various establishments. Their warmth and willingness to listen, learn and support is greatly appreciated.

St Joseph's Church Women's Guild of Canadian Forces Base Borden also invited Shirley Cox to speak about Neutropenia. By the end of the evening she had gained support of the Women's Guild. Information packages were hand delivered to the director of medical training for the two Canadian Medical Training Hospitals located at Base Borden and Base Trenton. A package of information was sent to the attending physician of one of the guild member's parents who was hospitalized out of province and not responding to treatment for severe abdominal infections which occurred after chemotherapy.

Thank you to all of the wonderful people who have financially supported the Ontario Chapter of the Neutropenia Support Association, the Everett Legion's \$1,000.00 donation from a community of 800, and the \$1200.00 donation from the Alliston Street Toys Car Club. We also sincerely appreciate all of the donations received by family, friends and business associates. ■

## A Hardy Well Done Gang!!

Our Manitoba Chapter would like to extend special recognition to the Rogers, Cox, and Cooper families. Their efforts have been extraordinary!! We send our best wishes and a hardy "Thank-you" handshake to you all. ■

# Generous Donation Received from C.N.



We received a pleasant surprise at our C.N. North American Christmas Dance, (Dec.10-94). The Toronto office donated \$910.00. The members of the Christmas party committee decided to give the money left over to a charity. They picked the Neutropenia Support Association because they were aware that our son Matt has Severe Chronic

Neutropenia. Many thanks to our friends on the Christmas party committee for this generous donation.

Corrie & Glenn Rogers

*Above: Matt Rogers, age 13*

*Below: Mary Evans presenting cheque to Corrie and Glenn.*



## NEWS FROM NEWFOUNDLAND

### News Update from Newfoundland!!

Hi Everyone,

Hope every one is doing well. Krista is staying well. We took her to the Janeway this weekend to find that Dr. Ingram may not be coming back to Nfld. Krista hadn't many fighters at all, but still no infections. When we think back to the days before Neupogen my, my how on earth did we hang in there? Thank you goes out to all the experts who are fully dedicated in research and to you Lorna for being just a 1-800-NEUTRO away.

I've still had much help in and around my community. Our firemen again this year held a dart tournament and raised \$1000 for neutropenia. Special thanks to the crew who spear headed this event, and to all the firemen who donated food. Truly, you are special people in our community. Also, an army of people from Leading Ticks, too many to mention by name, raised a total of \$1113.45. People who never met our family just went crazy raising money. "Tickle Retreat" held a dance and some of the ways they raised funds were just unreal.

I've been quite busy myself stuffing pillows. I've had material donated from the hospital for covering (thanks Ron and Bev) and pillow fill donated from Superior Glove. Also, I've been selling, or you could say renting, youth books at a local store here. Books sell for 50 cents and most of the buyers return the books to be sold again. Thanks Glen for the use of your store for these purposes. Botwood area raised over \$400, Vivian Elliott organized most of the work here. I have a sticker on my refrigerator, its a heart with a bandage down through it. It says "kids need more than medicine to get better". With the help you've all given its seems there's a light at the end of the tunnel.

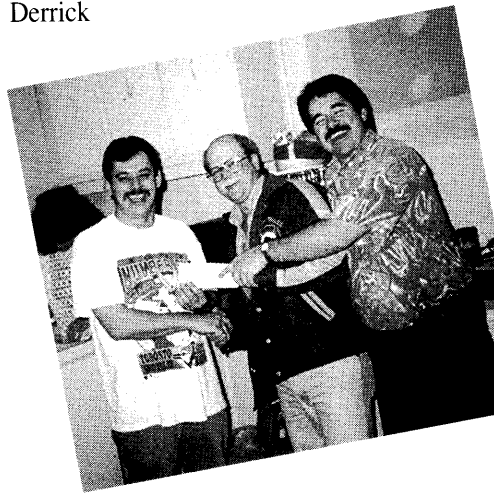
Sincerely Yours, The Cooper Family

**The above news was sent to us via the Internet, and was our first E-Mail message. Wow! has the world ever gotten smaller!**

### A few excerpts from local Newfoundland papers!!

A fund-raiser was held to give support to the Neutropenia Society. On Friday night various events such as a jelly eating contest, cracker eating contest, grape-fruit in the pantyhose race, and a dart shoot off was held. Special thanks to Molson who donated items like caps and sweatshirts to be used for prizes.

A dart tournament was held Saturday, organized by Derrick



*Johnny Rouse!! (fire chief) presenting owners of club a check.*

Chippett. Teams came from Point Leamington, Glover's Harbour and Leading Ticks to join in the fun. Harold Hemmeon was at the club cooking up a big traditional scoff. Many thanks to Harold and his wife Velma for preparing such a delicious meal. Thank you also to Bill Rolfe who donated all the vegetables.

The day ended with a dance, with music provided by Jason Parsons and Steve Hobbs. Thank you both. A total of \$1118.45 was raised. A note of thanks go out to the Fire Brigade, Ladies Fire Auxiliary, ACW, Gloria's Variety, and Newman's Supermarket for their contributions. Very special thanks to Derrick



*(L-R) Jason Parsons and Steve Hobbs provided free music for the dance.*



*Members of the Recreation Committee (L-R) Hazen Chippett, Cliff Chippett, Rodney Rowsell, Dean Cooke, Morley Alcock, Derrick Fudge*



*Jerry Stuckey donated a lobster to Neutropenia. We sold tickets and raised about \$70.00.*

Fudge and Bailey Noseworthy owners of the Tickle Retreat, for hosting all of the events.

Thank you to all the members of the Recreation Committee and the Economic Development Team, for putting together this worthwhile event. To all those people who participated in the games, bought hot suppers, tickets, or went to the dance, your thoughtfulness and your support was greatly appreciated.

**Editor's Note:**

**Dianne Cooper has on hand \$5,000.00 and will present on behalf of the N.S.A.I. to the Janeway during the T.V. June Telethon. This is an outstanding effort by all volunteers, participants, and donors. Dianne continues to make pillows and can be reached at 1-709-484-3592**

## **FROM THE ADVERTISER, GRAND FALLS, NFLD.**

**By Wendy Houlihan**

A ray of hope has finally come for two Grand Falls-Windsor boys. David Rose, 14 and brother Danny, 9 have been living with a rare blood disorder all their lives, and with the help of an international research team, the boys may see an end to their illness.

In 1981, 18-month-old David was diagnosed with cyclic neutropenia, a rare blood disease. His brother Danny was diagnosed in 1985 with the same disease. Not much is heard about neutropenia, but it can affect anyone - infants, children or adults. Some people are born with it, or it can happen after a viral infection. In some cases, the cause is a drug side effect, or exposure to certain poisons.

People can also get Neutropenia when treated for cancer with chemotherapy drugs.

Neutropenia exists when the number of neutrophils in the blood falls to a low level. People with the disorder get infections easily, and often. Most of the infections occur in the lungs, mouth and throat, sinuses and skin.

Certain types of Neutropenia are very rare like the one Danny and David have. Because this disorder is diagnosed in more than one member of the family, it is also considered multiple.

"What cyclic means is the boys suffer severe Neutropenia about every 21 days," said Bev Rose, the boys' mother. "The spell may only last three to six days, maybe weeks. During this time they are susceptible to infections."

Some infections can be severe or life-threatening, she said.

In 1981, when David was diagnosed, very little was known about the disease. At the time, there was only one known case in Newfoundland. Today, approximately 300 cases of cyclic Neutropenia have been recorded in North America. However, there is still very little known about the rare blood disorder.

### *Before and after*

Ms. Rose remembers the hard time she had before her first child David was diagnosed. David was given a blood test, and the results came back with a scare of leukemia. After more tests in St. John's, Doctor Kaiser Ali identified the child as having the rare blood disorder.

Before Danny, the youngest son was diagnosed, in 1985, the doctors thought it was a stomach tumor. He too had neutropenia.

"It was like beating my head against a wall," said Ms. Rose. "We were always at the hospital, with our boys, not knowing if this bout of infections would be the last. Doctors didn't know much about the disease at the time either."

It was frightening to know that each time one of the boys became ill, it might be fatal, she said.

Because of their illness, the boys missed a lot of school and kept to themselves a lot. David is now 14 years old, and has seen a big change in his illness. He has outgrown the symptoms and has been actively involved in school activities, and sports!

"He is a different person," said his mother. "His personality has totally reversed from a shy, sick boy to an outgoing and happy Teen."

The problem with Neutropenia is the symptoms can reoccur at any given moment without warning. David could be back where he started 14 years ago, she said.

### *Treatment*

Research is ongoing to develop treatments for neutropenia. Drugs are being tested, but to date, have not been authorized for use. (see correction)

"The drug treatment that has been most effective so far is antibiotics," said Dr. Lewis Ingram, of the Janeway Children's Hospital, "Because David and Danny are diagnosed as multiple cases, they are going to be involved in International research, conducted by Dr. Melvin Freedman, Hospital for Sick Kids in Toronto, and Dr. David Dale, from Seattle.

In 1994 Bev Rose attended a conference on childhood cancer and the family in Halifax. She got to meet the two doctors in charge of the international research project.

The project has begun and involves an extensive family history study. Blood work results should tell which gene caused the disorder and which side of the family it came from.

In the meantime, funding for the research project will be aided through Dominion Stores. Shoppers are asked to drop their receipts in a box in the store. Dominion stores will donate \$1 for every \$500 in receipts received.

### **Correction:**

**Neupogen (G-CSF) was approved as a treatment for severe chronic Neutropenia (congenital, cyclic, and idiopathic) October 28, 1993 by the Health Protection Branch Canada, December 22, 1994, Food & Drug Administration, U.S.A. Should you require information please call 1-800-6-NEUTRO.**



## Cox...a familiar Newfoundland name, a common link with Neutropenia.

Shirley Cox writes; our only daughter Stacey died suddenly at the age of 5 from a bacterial infection that her body couldn't fight effectively. Her husband and 2 of 4 boys also have Neutropenia. She asks our readers to help in locating other Cox families in helping her develop their family tree. The Cox Family has participated in a study for diabetes at the Memorial University for Research. Shirley has already determined a number of families also with Neutropenia are related. She states "Please help us to help ourselves, so that we can in turn help others".

Shirley and Michael Cox can be reached at:

11 David Drive RR #1  
Lisle, Ontario, Canada  
L0M 1M0  
Telephone (705) 424-1285 ■



1985 Stacey died age 5

# "Holding My Breath"

Part two of feature story (volume 6.1)

## Through the Wall

**T**he blare of an electric guitar makes me smile, even at full volume! The inevitable shout from the sister and the inevitable retort from the brother... I am still smiling. The "normality" of our lives envelope me like a warm blanket.

A year ago we were leaving the hospital, my son having completed the last chemotherapy session of an intense protocol for Burkitt's Lymphoma. Our story last fall "Holding My Breath", recounted our experiences, frustrations and fears as my son battled through a situation that had originally been deemed "hopeless" while he fought for his life in intensive care. Now, our visits to the clinic are every four months, and happily, uneventful and brief. We walk away with renewed hope as each milestone is passed.

My son missed most of Grade 6 in the 1993/94 year, present only in September and parts of the last term. There was no question in my mind that he would be put forward to Grade 7 as the difficult year with cancer had been "punishment" enough. It would have been too stressful for him to be separated from his friends. A supportive teaching staff and tutoring in Math helped immensely, but even with the confidence boosters, Grade 7 has proved to be quite a challenge.

I worried about the stress. Frustration levels were high, the demands of the rotary system new to him, and the load of daily homework and projects all became major challenges. The first few months were extremely difficult and he often wondered if he should have been held back. His concentration faltered, his self-esteem was low, he felt pressure and incompetent (and he was making his sister's life hell, at home!). The fact that he had missed so much school and that he should be proud of being in grade 7 was of no importance to him. That was then and this was now. He wanted to move on, and consequently, was very hard on himself and unhappy.

I knew he needed additional help.

We had been connected to the child psychiatrist while at the hospital, had seen him regularly while there, and continued to do so through the spring until summer. He had helped my son immensely (and my daughter) and my son still uses the tape they made together whenever he has problems relaxing or sleeping.

This doctor recommended another child psychiatrist in our area, for follow-up purposes whom we began to see in the Fall. Our visits are only occasional now. We saw him separately, and together as a group. It was very helpful to have this third party voice and the strategies he offered for lowering the stress levels, at school and at home.

By December, the situation eased. He felt less pressured. Assignments were getting done. The routines were familiar. Math wasn't a cinch, but less of a burden.

And then, without noticing the day-to-day changes, something happened—I got my boy back! I can't pinpoint exactly when, but early in the new year, there it was—that smile, the infectious laugh, the quirky, wonderful humour. This was my boy, physically and spiritually healed. He had begun to reclaim his life, to the fullest.

His current schedule includes organizing a garage band (Where he plays lead guitar/keyboard and vocals), a professional band (lead guitar and vocals) their first concert was in January, piano lessons, saxophone from school, the basketball team, Hebrew and Bar Mitzvah lessons, and general goofing around with good buddies, listening to music, playing tennis, skating, rollerblading, shooting hoops... he is making up for lost time, with relish!

As we take each step further away from that hospital life, we talk about tomorrow and plan far beyond tomorrow, with the growing confidence that we will all share in it.

The healing process affects each member of the family in a different way, at a different pace. I battled with myself to let go and not worry too much as he bounded toward any and everything there was to do. My daughter, 10, is still in the

healing process, not fully believing that it is over. This is her year to recover and to feel all the aches and pains that "were not" allowed, while her brother was sick. She is happier, but reticent to venture far from home. She needs the attention now.

It is every parent's worst nightmare to have a child fall ill, and when it is a life threatening illness we are helpless, spinning in the void of our own worst fears, dealing with painful realities and clinging to hope while we try to hold the family together and soothe and comfort the sick child.

While it is a path no parent or child should ever have to take, when we emerge on the other side there is great joy. I am thankful for all the prayers that were answered, for the support of family and friends and well wishers, the technology, knowledge and medications that are available today, my perseverance in exploring every avenue for his physical and psychological care, and my son's will to survive.

For those of you who have reached this plateau, you understand. For those of you still in treatment, I wish you strength. It may seem as though life will never be the same, it will not. It will be different, but if you have each other, it will be enough. ■

## Home Away From Home

Ronald McDonald House is a residence for families from outside Winnipeg, Manitoba, Canada, whose children are being treated for life threatening diseases and other serious medical conditions. We are a non-profit charity, and are able to limit your costs to \$10.00 per room per day. Where possible, we ask that you make reservations in advance by calling our House Manager, Ms Kerry Mushumanski at 204-774-4777. For first-time residents a health care referral is necessary.

Ronald McDonald House provides a home-away-from-home atmosphere to families in times of stress. Please call us with any questions you may have.

Check for your "home away from home" in your area. ■

## A letter from The Synapse Group

I would like to take this opportunity to thank you for your tireless efforts in assisting me on the various projects we have worked together on over the past few years. I have been greatly impressed by your dedication in getting the Organization going, and assuring it meets the needs of the patients and their families, while at the same time developing strong relationships within the medical and government communities. What you have accomplished is truly remarkable.

James J. Moriarty  
The Synapse Group  
Toronto, Ontario ■

## French Translation Now available!

We sincerely appreciate the assistance of Dr. Jean-Marie Leclerc, Chief of Hematology/Oncology and Clinical Director of Oncology, Hopital Sainte-Justine. We also wish to acknowledge the efforts of M. Camille de Varennes representing Leucan and Candlelighters.

We also appreciate the assistance of M. Ronald Davidson, executive director of Leucan, Montreal. Please call Leucan 1-514-731-3696 or the Neutropenia Support Assoc. Inc. 1-800-6-NEUTRO. They are provided free of charge.

1) La Neutropenie  
causes, consequences et conduite a tenir

2) Chimiotherapie et Neutropenie  
Renseignements a l'intention des patients atteints de cancer et de leurs familles

Phone or send in your order provided free of charge. ■

### U.S. SCN NEWS RELEASE

## NEUPOGEN (Filgrastim) LICENSED FOR USE IN REDUCING INFECTIONS IN PATIENTS WITH RARE BLOOD DISORDER

THOUSAND OAKS, Calif. — December 22, 1994 — People with a rare, potentially life-threatening blood disorder in which the body fails to manufacture sufficient infection-fighting white blood cells can now be treated with NEUPOGEN (Filgrastim), a biotechnology-derived product manufactured and marketed by Amgen Inc. NEUPOGEN, a recombinant granulocyte colony-stimulating factor (G-CSF), was licensed for its third indication today by the Food and Drug Administration (FDA) as the first therapy indicated for treating the condition, severe chronic Neutropenia (SCN), the company announced today.

Canada was approved October 28, 1993.

SCN affects some 1,000 to 2,000 children and adults in the United States. Prior to the introduction of NEUPOGEN, no effective treatment was available for this condition. Medical management of SCN had principally been driven by episodic management of infection, relying mainly upon regular or continuous antibiotic use and frequent hospitalization.

"Children with the congenital form of SCN often died from severe infection before adolescence. Doctors would switch them from antibiotic to antibiotic as a short-term bandage to prevent superficial infection," said David Dale, M.D; the lead investigator in the clinical trials that led to the licensing of NEUPOGEN (Filgrastim)

for SCN. "But in just the first few tests with NEUPOGEN, we knew this was a treatment that worked. The results were dramatic," said Dr. Dale, professor of medicine at the University of Washington Medical Center in Seattle.

NEUPOGEN has been in use since 1991 for use in cancer patients on various chemotherapy regimens and was approved for use in bone marrow transplantation (BMT) in July of 1994. In these settings, NEUPOGEN reduces the duration of Neutropenia (dangerously low levels of neutrophils) and neutropenia-related clinical conditions in patients with non-myeloid malignancies and those undergoing myeloablative (destruction of myeloid cells) chemotherapy followed by BMT.

"In the last three years since the original licensure of NEUPOGEN, our product has been demonstrated to be both safe and effective in treating hundreds of thousands of cancer patients receiving chemotherapy worldwide in an acute treatment setting. The clinical trial that led to today's FDA approval have now shown that NEUPOGEN is safe and effective in treating severe chronic neutropenia, as well," said Amgen Chairman and CEO Gordon M. Binder.

## Clinical Trials Document Fewer, less Serious Infections

In people with SCN, the bone marrow fails to produce sufficient neutrophils — one of the five major types of white blood cells and the one the human body deploys in the greatest number to keep bacteria and other microorganisms at bay. NEUPOGEN is a genetically-engineered version of a natural human hormone that increases production of neutrophils.

Though not a cure, in SCN clinical trials that evaluated the effect of daily subcutaneous injections of NEUPOGEN (Filgrastim), it has been shown to be safe and effective in decreasing the number of infections by causing a sustained increase in the neutrophil count. In the phase 3 trial conducted in the United States, investigators evaluated the efficacy and safety of NEUPOGEN in people with SCN over a four-year period. In this trial, 120 people with a median age of 12 years were studied and found to have a significant reduction in the incidence (60 percent

reduction) of Infections as well as a 59 percent reduction in the incidence of required antibiotic use.

"Our findings show that nearly all people with SCN can benefit substantially from this new therapy," Dr. Dale said. "This is the only agent that has been proven effective for use in SCN patients."

The trials were sponsored by Amgen and conducted by Dr. Dale along with eight other U.S. researchers from institutions including Memorial Sloan Kettering Cancer Center, University of Michigan, St. Jude Children's Research Hospital and Duke University Medical Center.

## SCN Result of Insufficient Bone Marrow Activity

The cause of SCN is not known. According to Dr. Dale, there are both acquired and hereditary forms of the disease. Dr. Dale and other clinical investigators and researchers are exploring the genetic makeup of SCN patients in the hope of finding its precise cause.

People with SCN contract infections easily and often, frequently in the mouth and throat, sinuses, skin, lungs and anus. These infections vary in severity and can be life-threatening. Ongoing mouth ulcers, ear infections and periodontal disease are common and adversely affect the patient's day-to-day life.

Treatment is generally well tolerated and high compliance was one of the most significant observations in the phase 3 trial, Dr. Dale said. Adverse reactions may include mild to moderate bone pain readily controlled with non-narcotic analgesics, general musculo-skeletal pain and enlarged spleen. Less than 3% of the 325 patients tested developed myelodysplasia or myeloid leukemia. However, acute myeloid leukemia or abnormal cytogenetics have been reported to occur in the natural history of SCN without cytokine therapy.

Because the natural history of SCN is not well understood, an international registry which will collect information on patient health and course of treatment has been established. This Registry, sponsored by Amgen, is under the guidance of a scientific advisory board headed by Dr. Dale. Amgen has agreed to provide NEUPOGEN (Filgrastim) to the Registry for those patients who require treatment.

In addition, because so few patients and families have met others with the disease, the need for a national support network was identified. With support from Amgen and the Canadian Neutropenia Support Assoc. Inc., the National Neutropenia Network (NNN) was formed by several U.S. families who have relatives with SCN. The group's charter is to "promote awareness, education and research about SCN and provide a support system for patients with Neutropenia and their families through a national resource network."

In February, 1991, NEUPOGEN was licensed as the first hematopoietic growth factor indicated to decrease the incidence of infection as manifested by febrile Neutropenia in patients with non-myeloid malignancies receiving chemotherapy regimens with the potential to compromise the body's immune system.

Amgen (NASDAQ:AMGN) is a global biotechnology company that discovers, develops, manufactures and markets human therapeutics based upon advanced cellular and molecular biology. The company is located in Thousand Oaks, California. ■

## International SCN Registry

## Dear Neutropenia Families

Dr. Melvin Freedman of the Hospital for Sick Children in Toronto is currently in the process of establishing the Canadian Neutropenia Registry.

Please read the attached information and encourage your physician's participation in this important medical resource. A successful participation rate will help researchers to better understand Neutropenia and hopefully pave the way toward managing your disease more effectively.

If you have any questions we welcome your calls to the Neutropenia support Association Inc. at 1-800-6-NEUTRO (1-800-663-8876) or you may contact Dr. Freedman directly at 1-416-813-6152.

One of Dr. Freedman's primary goals is to classify patients into categories ranging from mild to severe chronic neutropenia, therefore, all individuals suspected of having Neutropenia in whatever form should respond to this request. It is essential for the future well being of all. ■



# Dr. Freedman NEEDS OUR HELP!!

The International Severe Chronic Neutropenia Registry has been established to develop a worldwide database of treatment and disease-related outcomes for persons diagnosed with severe chronic neutropenia. Collection of this information will lead to improved medical care for patients and become a focus for future research. The Registry is directed by an advisory board of physicians with clinical and research expertise in Neutropenia and therefore those patients enrolled in it have access to up-to-date information from the experts. The Registry also ensures that patients who require treatment with G-CSF (Neupogen) receive it whether they can afford it or not.

In September, 1994, Dr. Bonnie Cham and I sent the enclosed letter to all Pediatric Hematologists and selected adult Hematologists in Canada. We urged our colleagues to enroll all appropriate patients under their care into the Registry. The response was encouraging at first but then declined sharply. Our estimate is that only 10% of Canadian patients are currently enrolled, and since we are aiming for a 100% national registration, a different strategy is needed.

Therefore, we are requesting your help by way of the Neutropenia Support Group Newsletter. If you would be kind enough to publish this letter as well as the information letter we sent to the Canadian Hematologists, patients with severe chronic Neutropenia who receive your Newsletter will be advised of the existence of the Registry.

The patients can then approach their physicians and urge them to initiate enrollment into the Registry.

We suspect that the patients and their families will respond more vigorously and enthusiastically than the physicians!

Thanks for your help with this important initiative. ■

## *Letter sent to Physicians*

September 26, 1994

Dear Colleague:

We invite you to participate in an international disease registry that has recently been established in Australia, Canada, the European Community and the United States for patients diagnosed with severe chronic Neutropenia (SCN). SCN is a disorder originating in the bone marrow that results in a chronically low peripheral blood neutrophil count (less than 500/mm<sup>3</sup>), which is not due to leukemia, aplastic anemia, viral infection (e.g. HIV) or a recognized autoimmune disease. SCN is estimated to affect 2,000 to 5,000 patients; in the United States and proportionately more worldwide. The diagnosis of SCN includes patients with genetic forms of neutropenia, cyclic neutropenia, and idiopathic forms.

The Severe Chronic Neutropenia International Registry is directed by an independent advisory board of physicians who treat SCN patients. The mission of the Registry is to establish a worldwide database of treatment and disease-related outcomes for persons diagnosed with SCN. Collection of this information will lead to improved medical care and become a focus for future research.

All patients with SCN over three months of age are encouraged to enroll in the Registry. The Registry will operate under a protocol which describes the standards for care and therapy as determined by the Scientific Advisory Board. However, all treatment decisions will be left to the discretion of the referring physician. Upon enrollment, patients and their physicians voluntarily agree to provide information about patient health and course of treatment. This data will be accessible only for research and treatment purposes.

G-CSF (Filgrastim; Neupogen) is approved for use in Canada by the HPB regulatory agency in Ottawa. The cost of G-CSF is handled in a variety of ways that vary from province to province. Some provincial Ministries of Health have established a budget for patients with SCN so that the product is prescribed without charging the patients. In other provinces, for example Ontario, about 60% of patients have a drug insurance plan, and 20% have a Drug Benefit Plan whereby the provincial Ministry of Health pays for the G-CSF. For the remaining patients, Amgen Canada has agreed to provide the product to the patient if all other sources of funding have been exhausted.

You may participate in the Registry by enrolling your eligible patients. All participating physicians will become part of a physician network designed to increase the understanding of SCN, and will be provided with periodic summaries and analyses of aggregate data from the Registry. Participating physicians may also request information from the Registry and are eligible to submit research proposals to the SCN International Registry Advisory Board for evaluation.

We believe disease registries are an important tool for understanding the etiology, pathogenesis and clinical course of rare diseases. We encourage you to enroll all of your patients with SCN. To facilitate this, you can contact either Dr. Melvin H. Freedman in Toronto or Dr. Bonnie Cham in Winnipeg who can confirm your patients' eligibility and facilitate the enrollment process.

Sincerely,

Dr. Melvin H. Freedman  
Hospital for Sick Children  
555 University Avenue  
Toronto, Ontario  
M5G 1X8  
Tel (416) 813-6152

Dr. Bonnie Cham  
Manitoba Cancer Treatment  
and Research Foundation  
100 Olivia Street  
Winnipeg, Manitoba  
R3E 0V9 Tel (204) 787-2188



# Consent for Release of Medical Information

## Physician Name

Melvin Freedman, M.D.

## Institution

Hospital for Sick Children

## Telephone

416-813-6152

## Background and Purpose

Because we know very little about the natural history of severe chronic Neutropenia (SCN) we, are asking you to participate in this Registry. We do know that treatment with Filgrastim (r-metHuG-CSF), or G-CSF, has been used for about five years to treat a number of patients, and it has decreased their fevers and incidence of infections. We still need to learn much more about this condition and how it should be treated. The objective of this Registry is to collect information that will allow for evaluation of various methods of treatment, and to monitor patients who chose, with the advice of their physician, to be treated with recombinant human granulocyte colony stimulating factor (referred to hereafter as G-CSF). The Registry will permit monitoring of the effectiveness and the safety of drug therapy for SCN and will help physicians determine the best means of treating this disease. The purpose of this Consent Form is to explain the Registry so that you, as a patient, can make an informed decision whether to participate, in the Registry.

## Procedures

As a participant in the, SCN Registry you will need to have regular medical checkup and tests (blood counts) as prescribed and directed by your physician. You may be, asked to maintain a calendar to report if you are ill and if you are hospi-

talized. You may also receive treatment with G-CSF, a new drug which has been approved by the Health Protection Branch for the treatment of severe chronic neutropenia. As an approved drug, G-CSF has significant therapeutic benefits as well as risks. In the event your physician prescribes G-CSF for you, then your physician should discuss with you in advance the risks and benefits of G-CSF. Similarly, if your physician prescribes a different therapy for SCN, then you should discuss that course of treatment with your physician. You and your physician are required to release information regarding your medical history and medical tests to the Registry office at the University of Washington, Seattle, Washington, U.S.A. You are also asked to provide the Registry with information regarding your health, tests and treatments as prescribed by your doctor on a long term basis. This will require completion of some standard forms when you are enrolled and about every six months thereafter expecting the Registry to be ongoing for several years.

## Other Information

The medical information released to the Registry will be kept confidential, but may be released to the. Canadian Health Protection Branch, other regulatory agencies and Amgen, the manufacturer of G-CSF. In addition, the Registry will forward identifying Information to the pharmacy if G-CSF is prescribed for your treatment. If prescribed by your physician, G-CSF will be provided if necessary, to participants in the Registry, free of charge for at least one year from enrollment, with annual renewal at the discretion of Amgen. You may refuse to participate or may withdraw from the Registry at any time. If you chose not to participate in the Registry, the quality of your medical care will not be affected. Circumstances may also arise which

would cause the operation of this Registry or your participation in it to be terminated, if this should occur these changes will be explained to you as promptly and completely as possible.

SIGNATURE OF INVESTIGATOR

DATE

### SUBJECT'S STATEMENT:

THE REGISTRY DESCRIBED ABOVE HAS BEEN EXPLAINED TO ME. I VOLUNTARILY CONSENT TO PARTICIPATE IN THIS ACTIVITY. I HAVE HAD AN OPPORTUNITY TO ASK QUESTIONS. I UNDERSTAND THAT FUTURE QUESTIONS I MAY HAVE ABOUT THE REGISTRY OR ABOUT MY RIGHTS AS A SUBJECT WILL BE ANSWERED BY ONE OF THE INVESTIGATORS ABOVE.

SIGNATURE OF SUBJECT

DATE

SIGNATURE OF PARENT  
OR LEGAL GUARDIAN

DATE

CC. SUBJECT  
INVESTIGATOR'S FILE

# Recommended Reading!

## **Chemotherapy and Neutropenia: Information for persons with cancer and their families.**

Booklet explains causes and basis of neutropenia, fevers and infections and provides practical suggestions for dealing with Neutropenia when it occurs. Produced by the Neutropenia Support Association and Sponsored by Amgen Canada. Price: No charge.

## **The Candlelighters Guide to Bone Marrow Transplants in Children.**

An excellent collection of articles by parents and professionals dealing with the many aspects of bone marrow transplants. It is designed to help parents make informed decisions and to gain some sense

of control. Edited by F. Leonard Johnston and Ellen I. O'Donnell. Published by U.S. Candlelighters, 1993. This book is made available through a generous donation from the U.S. Candlelighters. Price: no charge for parents.

## **Chemotherapy and Neutropenia: Information for persons with cancer and their families.**

Booklet explains causes and basis of neutropenia, fevers and infections and provides practical suggestions for dealing with Neutropenia when it occurs. Produced by the Neutropenia Support Association and Sponsored by Amgen Canada. Price: No charge.

## **EMLA Colouring Book: A Fun and Games Book to Make You Smile**

A colouring book for the preschool and early school age children designed to help them learn how the proper application of EMLA can prevent the pain of invasive procedures. Includes games to help pass the time while waiting. Produced by Astra Pharma. Price: No Charge.

## **FILGRASTIM (r-methHuG-CSF) IN CLINI- CAL PRACTICE**

edited by George Morstyn. T. Micheal Dexter. Price: \$125.00 (U.S.)

To Order: Call 1-800-228-1160  
Or Write to:  
Marcel Dekker Inc.  
270 Madison Ave. New York,  
N.Y. U.S.A. 10016

## **HELPFUL HINTS**

# Needles Can't be Avoided: Fear and Pain Can

## *EMLA Gets Even Easier to Use*

39% of all Canadians admit they dislike needles; 11% are scared, absolutely terrified of needles 38% say the worst part is the anticipation; 25% avoid needles wherever possible; and 38% of those with children say it can be traumatic to take their child for a needle.

How often have you heard someone say to a child "this won't hurt a bit" or "this will only take a minute." We all know that children feel that having needles is a miserable experience and that the fear of needles can make seconds or minutes seem like for ever.

Children no longer need to experi-

ence the pain of needles or the fear associated with them. EMLA when applied to the skin as directed anaesthetizes the skin making procedures such as blood tests, port access, starting IV's and lumbar punctures free of pain.

*Children no longer need  
to experience the pain  
of needles*

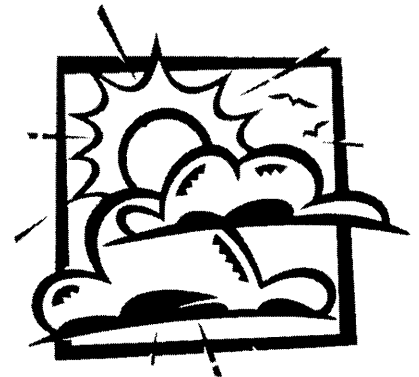
The new EMLA patch is even easier to use. The key is to apply the patch firmly to the skin so that is airtight. This will allow the maximum absorption of the medication. It can be used where you would have applied the cream before. It is hoped that the ease of application will enhance the use of EMLA and it is our hope that children will not suffer from needless pain which can effectively be prevented.

EMLA cream will still be available in both sizes - the 5 gram tube for two procedures or the 30 gram tube for mul-

tiples or larger applications.

The EMLA patch is available from your pharmacist but you will have to request it directly. The product number is #974 and two patches should retail for approximately \$8.50

The wholesale cost to the pharmacist is \$4.65.) Product information is available from your oncologist or pharmacist or by calling the manufacturer Astra at 1-800-668-6000. You may also write to EMLA Information Service" Astra Pharma Inc., 1004 Middlegate Road, Mississauga, Ontario. L4Y 1M4.



## Doctor's Corner



*Reprint from the  
National  
Neutropenia  
Network*

*We receive questions from families regarding Neutropenia and we welcome your questions for future issues. This issue's question is being answered by Dr. David Dale, Professor of Medicine, University of Washington Medical Center, Seattle. Dr. Dale is the lead investigator in the clinical trials which led to the licensing of NEUPOGEN (Filgrastim) for SCN.*

**Question: Should a woman who has had a child with severe chronic Neutropenia have other children?**

**Comment:** This is not a simple or easy question to answer. Some forms of congenital Neutropenia are inherited, and others probably are not. At present we know that childhood onset/congenital cyclic Neutropenia is inherited in an autosomal-dominant pattern. This means that a person who has the disease, who marries unaffected person, has a 50/50 chance of each of their children being affected. The normal brother or sister of a patient with cyclic neutropenia, however, can expect to have normal children. It is important to note that cyclic Neutropenia occurs with equal frequency in males and females, and it can be inherited from either the mother or the father. Thus a man who has had a child with cyclic neutropenia, or a woman who has had a child with cyclic neutropenia, have equal chances of their children also being affected.

For other forms of congenital neutropenia, our information is less secure.

There are now a substantial number of patients with severe congenital Neutropenia who have had affected children, but usually only some and not all of their children are affected. This suggests that these other forms of Neutropenia may also be inherited in a dominant pattern and from either parent. When hematologically normal parents have a child with severe neutropenia, this suggests mutation in the developing tissues for the child. In this situation, it is most likely that other children would not be neutropenic. There are, however, families in which Neutropenia is inherited in a recessive pattern; in this situation, having an affected child does suggest that additional children from the same two parents may also be neutropenic. The best way of trying to understand this possibility is to see a geneticist who can carefully review the medical history of the whole family, to try to give the best advice in this special situation. ■

**Editors note: We welcome your questions and can submit them to the International Neutropenia Registry Physicians for answers.**

*From The Manitoba Cancer Treatment and Research Foundation:*

*Thank you for generously agreeing to purchase a textbook for the Manitoba Cancer Foundation Library. The textbook is entitled "Neoplastic Diseases of Childhood" and its cost is approximately \$430 US. It is a comprehensive oncology text and includes sections on prevention of infection and treatment of the febrile neutropenic child and the use of hematopoietic growth factors in children with cancer. You may be personally interested to know that one of the contributing authors is Dr. N. Kibrinsky.*

*Thank you again for your ongoing interest and support.*

**The Royal College of Physicians & Surgeons of Canada, Montreal, Quebec. September 13 -17**

**N.S.A.I. invited to exchange with Healthcare Practitioners Neutropenia Registry and general info.**

## Candle Lighters Childhood Cancer Conference 1996

**"Forward to the Future"**  
**Calgary, Alberta August 9-11, 1996**  
**Neutropenia Session**  
**Canadian and U.S. families are invited.**  
**Call for details**  
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# SCN Disease Registry

## For Patients in Europe:

Prof. Karl Welte  
Med. Hochschule Hannover  
Abt. Kinderheilkunde IV  
Konstanty Gutschow Str. 8  
G 3000 Hannover  
Germany

Tel: 49511 5326020

Fax: 49511 5326120

## For Patients in Canada:

Dr. Bonnie Cham  
Manitoba Cancer Treatment and Research  
Foundation  
Cell Biology  
100 Oliva Street  
Winnipeg, Manitoba  
Canada R3E 0V9

Tel: (204) 787-2188

Fax: (204) 783-6875

or

Dr. Melvin Freedman  
Hospital for Sick Children  
555 University Avenue  
Toronto, Ontario  
Canada M5G 1X8

Tel: (416) 813-6152

Fax: (416) 813-5327

## For Patients in Australia:

Dr. George Kannourakis  
Department of Hematology  
Royal Children's Hospital  
Flemington Road  
Parkville, VIC 3052

Tel: (613) 345-5656

Fax: (613) 349-1124

## For Patients in the United States:

### West

Dr. David C. Dale  
University of Washington  
Department of Medicine  
AS 522 RG22  
1959 Pacific Street, N.E.  
Seattle, WA 98195

Tel: (206) 543-7215

Fax: (206) 685-4458

### East

Dr. Mary Ann Bonilla  
Memorial Sloan Kettering  
Cancer Center  
1275 York Avenue  
New York, NY 10021

Tel: (212) 639-8451

Fax: (212) 717-3447

### Central

Dr. Laurence Boxer  
University of Michigan  
F6515 Mott Children's Hospital  
Box 0238  
Ann Arbor, MI 48109-0008

Tel: (313) 764-7126

Fax: (313) 936-8520

*The Neutropenia Support*

*Association Inc.*

*has gratefully received*

*many "In Memory of"*

*donations.*

*May the knowledge that*

*this gift will aid others be*

*of comfort.*

*The families have received*

*acknowledgements of the*

*generous donations.*

*We continue our efforts*

*with help from your tax*

*deductible donations.*

***Thank You!***

# SCN Registry in Canada

**Re: Canadian SCN disease registry. Registration is facilitated by the two Canadian registry physician contacts, Dr. Melvin Freedman, Toronto Sick Children's Hospital (ph: 416-813-6152 fax: 416-813-5327), and Dr. Bonnie Cham, Manitoba Cancer Treatment & Research Foundation.**

**Registration forms will be provided to the referring physician. Call your physician today to begin this process. Information on reimbursement possibilities will be outlined by Dr. Freedman and Dr. Cham.**



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- ☐ copies of the Neutropenia Support Association Newsletter and back issues (as available)
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