

# Hemophilia

## TODAY

Canadian Hemophilia Society  
 We're all related by blood.  
[www.hemophilia.ca](http://www.hemophilia.ca)

Newsletter of the Canadian Hemophilia Society • Winter 2001 • Vol 36 No 1

## CHS launches national von Willebrand Disease Awareness Campaign

Until a few years ago, von Willebrand Disease (vWD), and in particular its impact on women, was little recognized – even among those in the bleeding disorders community. For the next three years, with the generous support of Aventis Behring, CHS is undertaking a major campaign aimed at increasing awareness about von Willebrand Disease among health care providers and the general public. The campaign will kick off on April 17, International Hemophilia Day, with a major media blitz and local awareness activities to be presented by chapters and clinics across the country. Hopefully, as a result of our efforts over the next three years, there will no longer be a reason for persons to have to endure the symptoms of untreated vWD.

This special issue of *Hemophilia Today* will feature a variety of topics relating to von Willebrand Disease including articles of a medical nature as well as personal stories of individuals from our community who are living with vWD.

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Why do I bleed so much at the dentist?

How can I control my heavy periods?

Why do I bruise so easily?

Why do I get so many nosebleeds?

**you CAN stop the bleeding...**  
 von Willebrand Disease...  
 the most common bleeding disorder

The CHS would like to express its appreciation to Aventis Behring for making this educational program possible.





Barry Isaac, Ph.D.

## Notes of a Personal Nature

This issue of *Hemophilia Today* deals almost exclusively with von Willebrand Disease (vWD), the nearly invisible member of the hemophilia family. It is nearly invisible because it is often mistaken for some other malady, or the patient might feel that the bleeding episodes are not severe enough to warrant medical attention, and so it lies hidden, sometimes for decades, sometimes for generations. With this issue of *Hemophilia Today*, and through the undertaking of a major national awareness campaign, the CHS hopes to bring vWD into the light and thus bring a new focus on its nature and treatment. All of us at *Hemophilia Today*, and all of our members who have taken the time to write the articles in this issue, hope that our efforts will be fruitful and that the information now in your hands will help in bringing all those who suffer from vWD to a new understanding of their disorder and to new and better treatment. But your Editor has another matter that he wishes to speak about here.

It is seldom that the editorial column really does have personal opinion, even though that is what it is meant to contain. More often your Editor speaks about the content of the issue and the important issues that face the CHS from day to day.

But this column is about personal matters. Last November your Editor was the recipient of the Frank Schnabel Award and he attended the Annual CHS Awards Banquet, held in one of the prettiest resorts imaginable, the Briars on Lake Simcoe in Ontario. Upon receiving the award, your Editor gave what may have been one of the shortest "thank you" speeches on record. If truth be known, your Editor was overcome with emotion and barely even able to say a minimal "thanks". For that he apologizes deeply.

For the record, the Editor would like to repeat what he did say and expand his thanks to all members of the CHS. Two individuals have always stood at your Editor's side as he has tried to do the tasks for which he volunteered: Joan Isaac and Clare Cecchini. Joan has supported your Editor for more than 34 years, and Clare must often feel that it has been that long for her as well. Whatever your Editor might have done for the CHS over the years, he owes a huge debt to Clare, who has the patience of Job and a boundless heart. Others, too, have been helpful and supportive over the years, Hélène Bourgaize, Robert St-Pierre, Pierre Latreille, Lindee David and Bob Shearer. Hélène and Robert are the only ones still with the CHS office staff, and one of the many things your Editor sincerely misses because he is no longer on the CHS Executive is working directly with them. They are wonderful people, generous in nature and wise far beyond their years. Your Editor still misses Pierre Latreille, a man of wise and quiet counsel, who passed away at a shockingly early age.

But most of all it has been the work and aid of literally dozens of volunteers from every province in the nation to which your Editor feels indebted. He is constantly in wonder at the courage and determination you all show in your daily lives and the way you manage your hemophilia. It is this courage, this determination, this ability to laugh in the face of constant threats that often gives your Editor the means to get on in his own life and tasks. People like you give our Editor strength to accomplish the tasks for which he was awarded the Frank Schnabel Award. Thank you all for being so supportive, kind and helping over the years. And thank you for letting your Editor say here what he could not at the Awards Banquet owing to a surplus of emotion.

**(Please send any comments raised by this, or any other Editorial to the Editor, *Hemophilia Today*, 1409B 4th Street N.W., Calgary, AB T2M 2Y8. Or fax your comments to (403) 282-0786)**

# Hemophilia TODAY

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*Hemophilia Today* is the official publication of the Canadian Hemophilia Society (CHS) and is issued three times yearly.

**The Canadian Hemophilia Society exists to improve the quality of life for all persons with hemophilia and other inherited bleeding disorders and to find a cure.**

The purpose of *Hemophilia Today* is to inform the hemophilia and bleeding disorders community about current news and relevant issues. Publications and speakers may freely use the information contained herein, provided a credit line including the volume number of the issue is given. Opinions expressed are those of writers only and do not necessarily reflect the views of the CHS.

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# PRESIDENT'S MESSAGE

Erma Chapman, Ph.D.



## Final thoughts...

*Much of the success of the past three years can be attributed to teamwork within the CHS. We have forged a new way of working together, between volunteers and staff members, and among national, chapter and regional levels of the organization.*

In writing this last message to you as President of the Canadian Hemophilia Society, I looked to my first message (Fall 1998) for inspiration. While the issues that were of interest to us then bear an amazing similarity to those of today, the way we operate as an organization has changed over the past three years.

Compensation for all Canadians infected with HCV through the blood system remains a priority for the CHS. At the time that I am writing my message, I have reason to hope that an equitable solution will be found in the near future.

Canadian Blood Services and Héma-Québec have focused on safety as the blood service institutions in this country. While we have at times struggled with our partnerships with these organizations, we continue to maintain an open dialogue towards the shared goal of a safe and efficacious blood system in this country. Our position as a watchdog of the blood system was reinforced at our January 2000 strategic planning meeting. As I noted in my first message to you, however, we can never again completely trust.

Care and treatment of people with bleeding disorders have remained our focus. Beginning with the "Winnipeg II" meeting of April 1998, we have worked with the Association of Hemophilia Clinic Directors of Canada (AHCDC) and Canadian Association of Hemophilia Nurses to establish standards of care across the country. A clinic in Saskatchewan appears imminent, and strides have been made in other areas to enhance clinical services. We have dealt with the presence of CJD, and welcomed the introduction of second-generation

recombinant products. Volunteers and staff members have developed program initiatives in the areas of inhibitor management, a CD-ROM format for information designed to assist parents of newly-diagnosed children, improved access to appropriate care in emergency rooms, and services for women with bleeding disorders.

**Care and treatment of people with bleeding disorders have remained our focus.**

Currently, a major initiative is underway to ensure the appropriate diagnosis and treatment of von Willebrand Disease.

The opportunity to work with the World Federation of Hemophilia in planning for the World Congress of Hemophilia in Montreal was unique. Our CHS volunteers were among the stars of the Congress last summer. The youth experience was not only successful for the Congress, but has revitalized youth initiatives within the CHS. As well, the CHS has been able to establish a fund for international projects thanks to the revenue from the Congress. The CHS was also represented in the planning of the first global forum on the use of hemophilia products, held in Montreal during April 2000.

Research is the hope of every person with a bleeding disorder for a better life, and will remain a priority for the CHS. A positive and productive working relationship with the Hemophilia Research Million Dollar Club has helped to generate an increased funding base for research

towards a cure, and the promise of even greater research revenue over the next three years. This past year, the *Care Until Cure* research fund was introduced along with the Aventis Behring – CHS-AHCDC Fellowship in Hemophilia. In addition, we have had increased opportunities to participate in priority-setting and research initiatives with the AHDCDC, Health Canada and the Laboratory Centre for Disease Control.

A new vision for resource development within our organization opens opportunities for more diversified funding sources, as well as opportunities for volunteer development. A Chief Development Officer has been added to our staffing complement. Over the next month, staff positions will be added in Saskatchewan and in the Maritime Region.

Much of the success of the past three years can be attributed to teamwork within the CHS. We have forged a new way of working together, between volunteers and staff members, and among national, chapter and regional levels of the organization. We have developed a shared strategic vision for people with bleeding disorders and their families, and have ensured that organizational structures are in place to support the realization of our dreams.

During my time as President, I have had the chance to work with three remarkable executive directors. Lindee David's knowledge of bleeding disorders and blood systems was matched by her passion for justice, her caring for others and her hearty laugh. Pierre Latreille's quiet reassurance provided me with the direction and confidence to fulfill my leadership responsibility; I still grieve his passing and miss his gentle guidance. Daniel Lapointe brought a wealth of non-profit management knowledge, skills and experience to the position. His passion for excellence, belief in collaboration, and ability to develop effective partnerships are qualities I will continue to treasure.

It has been a privilege to meet and share with so many members across the country. And it has been an honour to serve as President of such a dedicated group of volunteers and staff.

**Thank you, and farewell,  
Erma Chapman, Ph.D.**



# NEWS UPDATE



## 1st CANADIAN CONFERENCE ON HEPATITIS C

May 1 - May 4, 2001  
HILTON MONTRÉAL BONAVENTURE

### SHARE IDEAS, TAKE ACTION

A three-track program that will include :

**BASIC AND CLINICAL SCIENCE  
PUBLIC HEALTH  
SOCIAL SCIENCE AND COMMUNITY**

Organized by the  
**Canadian Hemophilia Society,**  
in partnership with :

Canadian Association for the Study of the Liver  
Hepatitis C Society of Canada  
Canadian Liver Foundation  
Canadian Public Health Association  
Canadian Centre on Substance Abuse  
Prisoners with HIV/AIDS Support Action Network  
Hepatitis C Division, Health Canada

This conference has been made possible  
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Canadian Hemophilia Society  
We're all related  by blood.

#### NOTICE

**The Annual General Meeting of the Canadian Hemophilia Society will convene as follows:**

**Saturday, May 26th 2001 at 9:00 a.m.**  
**at the Sheraton Hotel in Halifax, Nova Scotia**

- To acknowledge the designated Directors of each Chapter;
- To acknowledge the Elected Directors at Large for 2001-2002;
- To receive the audited financial statements of the Canadian Hemophilia Society for the year ended December 31, 2000;
- To appoint an auditor for the ensuing year;
- To approve the proposed revised by-laws (Copies of the proposed revised by-laws are available at the CHS Office.  
To obtain a copy prior to the AGM please call 1-800-668-2686;
- To transact such other business as may properly come before this Annual General Meeting of the members of the Canadian Hemophilia Society.

**Ron MacLeod**  
Secretary

### RCMP Investigation into Destruction of Documents

The RCMP announced on February 26, 2001 that no charges would be laid regarding the destruction of Canadian Blood Committee documents. The RCMP confirmed that audio recordings and verbatim transcripts of the now-defunct federal-provincial government committee's meetings from 1982 to 1989 had been destroyed, but announced that they could not determine if this was done with criminal intent. CHS remains optimistic that the RCMP will uncover the truth in the larger investigation regarding potential criminal wrong doing that led to the contamination of Canada's blood supply.

### Manitoba Government Extends Compensation

On January 19th the Manitoba Government announced their decision to offer compensation to hepatitis C victims who were infected through blood products received either before 1986 or after 1990. Manitoba joins Ontario and Quebec in announcing financial assistance to victims in their province who were left out of the 1986-1990 Hepatitis C Compensation Agreement. Manitoba and Quebec are providing a lump sum payment of \$10,000 and Ontario is providing \$25,000 to all individuals infected with hepatitis C prior to 1986 or after 1990. CHS continues its efforts to convince the federal government and the seven remaining provinces to follow the leadership shown by Ontario, Quebec and Manitoba and compensate all victims of this tragedy.

### Canadian Red Cross Society Settlement Denied

On February 20, 2001 Justice Winkler denied approval of the proposed CRC settlement. The CHS believes that this recent event illustrates once more how the extension of the 1986-1990 Hepatitis C Compensation Package to all victims of tainted blood remains the best way to respond to the needs and expectations of those victims who are still waiting for compensation. CHS has reason to hope that the federal government may reconsider its position and accept to extend the 1986-1990 package to those infected outside of this period in light of recent evidence showing that approximately 5,000 victims are presently excluded, a number considerably lower than previous projections.

### First Care Until Cure Research Grants Awarded

CHS and Genetics Institute are pleased to announce the first recipients of the newly established research program aimed at improving the quality of life of persons living with hemophilia or another inherited bleeding disorder. Congratulations to the following recipients:



**Recipient: Dr. Georges-Étienne Rivard**  
**Names of Co-investigators: Dr. Rochelle Winikoff**  
**Dr. Patrick St-Louis**  
**Dr. Claire Infante-Rivard**  
**Project Title: Von Willebrand Factor and Physical Exercise**

**Recipient: Dr. Jenny Heathcote**  
**Names of Co-investigators: Dr. Jerome Teitel**  
**Dr. Ignatious Fong**  
**Project Title: Therapy for Patients with Congenital Coagulation Disorders with Hepatitis C with or without HIV co-infection**

**Recipient: Dr. Manuel Carcao**  
**Names of Co-investigators: Dr. Brian Feldman**  
**Dr. Victor Blanchette**  
**Dr. Paul Oh**  
**Dr. Paul Babyn**  
**Dr. Douglas Hedden**  
**Project Title: A Prospective, Randomized Trial to Compare Two Regimes of Prophylaxis in Older Boys with Severe Hemophilia A**

Applications for the second year of the **Care Until Cure Research Program** will be available in May. For further information please contact the National Office.

### CHS Scholarship & Bursary Program

Applications for the 2001 CHS Scholarship and Bursary Program are now available at chapters, clinics and national, provincial, and regional offices. The deadline for submitting applications this year is April 30, 2001. Recipients will be notified in May.

### Three Canadian Winners in National Art Contest

Three of the winners in the Aventis Behring 2001 Calendar Contest were from the Canadian bleeding disorders community. Annette Kavelaars-Burrows of London, Ontario was awarded First Prize for her artistic depiction of living with a bleeding disorder. Christine Czipf of Thunder Bay, Ontario was awarded Grand Prize for her artistic photograph depicting the journey of her two daughters Olivia and Jacklyn as they move down the path of life. And Timothy Glazebrook, age 6, of Greenwood, Nova Scotia was awarded First Prize for his artistic depiction of nature's beauty. Contratulations to the recipients.



## Von Willebrand Disease: Diagnosis and Management of the Most Common Inherited Bleeding Disorder

**Dr. David Lillicrap**

*Professor, Departments of Pathology and Medicine, Queen's University*

Seventy five years ago, Dr. Erik von Willebrand first described an inherited bleeding disorder that manifests as excessive bleeding from mucous membranes and into the skin in a large family in the Aaland Islands in Scandinavia. The index case in his studies, a 14 year old girl, subsequently died as a result of excessive bleeding associated with menstruation. The condition described by Dr. von Willebrand now bears his name and is recognized as the most common inherited bleeding disorder known in humans.

### Introduction to von Willebrand Disease

Over the past decade, with the increasing realization that von Willebrand Disease (vWD) was not a rare condition, but in fact a very common, usually mild, bleeding disorder, interest in the diagnosis and improved clinical management of the condition has increased significantly. Furthermore, recognition that the major health problem associated with vWD is heavy bleeding associated with menstruation, has also begun to affect the practice of gynecologists and family physicians who see large numbers of women with this concern.

The primary abnormality in vWD

*The condition described by Dr. Erik von Willebrand now bears his name and is recognized as the most common inherited bleeding disorder known in humans*

involves the deficiency or abnormal function of a large protein, von Willebrand Factor (vWF), that is produced by the lining cells of blood vessels (endothelial cells) and by the parent cells of blood platelets (megakaryocytes). Von Willebrand Factor is required for two essential components of the normal blood clotting process. The protein acts as an adhesive bridge to bind blood platelets to the site of blood vessel damage and secondly, serves as a carrier for Factor VIII in the blood. Without normal levels of vWF, Factor VIII is rapidly cleared from the blood, resulting in a significant secondary deficiency of this critical blood clotting factor.

Studies performed in Europe and North America indicate that as many as 1% of the population may have clinical signs, and laboratory test results are consistent with a diagnosis of vWD. In many of these subjects, the clinical evidence of bleeding may be subtle and amount to no more than a tendency to bruise easily or to experience recurrent nose bleeds. Nevertheless, even though their tendency to bleed spontaneously may be minor, these individuals may be prone to a significantly greater risk of excessive post-operative and post-traumatic bleeding. Despite the fact that we are now fairly confident about the high prevalence of this condition, we need to understand a great deal more about why the disorder is so common.

### The Diagnosis of von Willebrand Disease

A diagnosis of vWD requires consideration of three factors: a personal history of excessive bleeding, a family history of excessive bleeding and laboratory test results of blood clotting that are consistent with vWD.

In the vast majority of vWD cases, the spontaneous bleeding tendency is limited to excessive blood loss into the skin (easy bruising and prolonged bleeding from cuts) or from mucous membranes (nose bleeds, bleeding from the bowel and menstrual bleeding). However, in a rare, severe form of the disorder (Type 3 vWD), spontaneous

bleeding also occurs into muscles and joints as is seen in severe hemophilia. Excessive and prolonged bleeding is also frequent after dental work and other forms of surgery. For a physician to make a diagnosis of vWD, it is extremely important for them to be able to obtain a detailed bleeding history from which they can evaluate the extent of the bleeding problem. A history of nose bleeds or bruising is not uncommon in the general population, and only a detailed assessment will identify whether the extent of bleeding warrants the type of laboratory evaluation required to make a diagnosis of vWD.

### CLINICAL SIGNS AND SYMPTOMS OF VON WILLEBRAND DISEASE

- *Easy bruising*
- *Frequent and prolonged nose bleeds*
- *Prolonged bleeding from cuts*
- *Heavy menstrual bleeding*
- *Prolonged and excessive bleeding following dental work and surgery*

Von Willebrand Disease is an inherited disorder and, as such, there should be other people within the patient's close family with the condition. As the gene that directs production of von Willebrand Factor is located on chromosome 12, and everyone receives two chromosomes 12s, one from each parent, this bleeding disorder, in contrast to hemophilia, manifests in equal numbers of males and females.

Most forms of vWD have a so-called dominant mode of inheritance, whereby only one (of two) copies of the von Willebrand Factor gene needs to be abnormal to lead to the clinical manifestations of the disease. Thus, for many people with vWD, one of their parents will have signs of excessive bleeding and there may be other siblings with a similar history. However, one of the factors that complicates our understanding of this condition is a phenomenon referred to as incomplete penetrance. This circumstance results in many people that have inherited the vWD gene not showing any clinical manifestations and thus a family history for the condition is hidden.

## Laboratory Tests for von Willebrand Disease

Once there is a clinical suspicion of vWD from the clinical and family history, a standard set of laboratory investigations should be ordered to confirm or rule out the diagnosis. These laboratory tests range from a basic blood count, from which the physician can assess the patient's hemoglobin and platelet count, to relatively sophisticated studies of the structure of von Willebrand Factor (vWF) in the blood. The most important of these studies are measurements of the clotting Factor VIII level (Factor VIII is carried in the blood by vWF), a measurement of the amount of the vWF protein in the blood (the vWF antigen: vWF:Ag) and a determination of the functional activity of the vWF (the ristocetin co-factor: vWF:RCo). In most types of vWD, these laboratory tests will show a significant reduction in vWF and Factor VIII from that seen in the blood of normal subjects.

### LABORATORY TESTS FOR VON WILLEBRAND DISEASE

- **Von Willebrand Antigen (VWF:Ag)**  
*Measures the amount of vWF protein in the blood.*
- **Von Willebrand Factor Ristocetin Co-factor (VWF:RCo)**  
*Measures the function of the vWF*
- **Factor VIII clotting activity (FVIII:C)**  
*Measures the amount of factor VIII being carried by vWF*
- **Von Willebrand Factor Multimer Analysis**  
*Determines the structure of vWF in the blood. Usually abnormal in Type 2 vWD*

These tests can all be performed on a single blood sample, but, because several of the tests are time consuming and complex, most laboratories collect samples to perform the tests in batches a couple of times each month. The results of these tests will show one of several outcomes. They may show unequivocal evidence of vWD and the diagnosis is thus confirmed. However, even if these initial tests are positive, the physician will need to sub-type the disorder into one of the three major categories of vWD, types 1-3. This will require some

additional laboratory tests (such as the vWF multimer analysis) that can usually be performed on the same blood sample that was used for the initial diagnosis, and again, these tests usually take at least a couple of weeks to complete. Sub-typing of vWD is necessary to develop the most effective plan for prevention and treatment of bleeding in vWD because we know that the therapeutic responses of the sub-types vary significantly for certain forms of treatment.

### SUB-TYPES OF VON WILLEBRAND DISEASE

#### Type 1 vWD

~ 85% of all cases.  
*Mild/moderate reduction in vWF and Factor VIII levels.*

#### Type 2 vWD

~ 15% of all cases.  
*Production of functionally abnormal vWF.*

#### Type 3 vWD

~ 50 cases in Canada.  
*Total absence of vWF from the blood. Severe clinical disease.*

A second potential outcome from the initial set of laboratory studies is that the test results are "borderline" or normal. In this instance, the physician will need to re-evaluate the clinical bleeding history and, if this still appears to be excessive, they will very likely ask for a repeat set of tests to be performed. Unfortunately, this circumstance is not unusual in attempting to make the diagnosis of vWD. The blood levels of vWF and Factor VIII are well known to fluctuate over time, and unless testing has been repeated at least twice (and some physicians will ask for three sets of tests) the diagnosis of vWD cannot be ruled out with confidence in someone with a significant bleeding history. Even after several sets of tests, some individuals still show "borderline" values for vWF and Factor VIII. These people may be told that they probably have vWD and that at the current time no additional studies can clarify this situation. These observations emphasize that the major problems in dealing with vWD are the initial consideration of the condition as a diagnostic possibility and, probably most difficult of all, confirming the diagnosis.

## The Prevention and Treatment of Bleeding in von Willebrand Disease

In contrast to the challenges in making the diagnosis of vWD, the treatment of this condition is effective and relatively straightforward. Apart from the very few individuals with the severe type 3 form of the disorder, most patients with vWD do not require regular treatments with transfusions. For the past 20 years the mainstay of treatment in most vWD patients has been the use of the drug Desmopressin (chemical abbreviation DDAVP). This drug is effective in the prevention or treatment of bleeding in approximately 80% of patients with vWD. Desmopressin works by inducing the release of vWF from stores of the protein that are maintained in the cells in which it is produced. In most patients, a treatment of Desmopressin will increase the blood level of vWF (and Factor VIII) by 3 to 5-fold over the baseline values, at one hour after administration. The drug is

**In contrast to the challenges in making the diagnosis of vWD, the treatment of this condition is effective and relatively straightforward.**

usually given by intravenous or subcutaneous injection, but a new concentrated form of Desmopressin, administered as an intranasal spray, has also recently been introduced. The administration of Desmopressin is sometimes accompanied by mild side effects that may include some facial flushing and small transient changes to the pulse and blood pressure. In contrast, the serious side effect of seizure induction, is extremely rare and can be effectively prevented by limiting fluid intake in the 24 hours after Desmopressin treatment. These issues are routinely discussed with patients by their physician or nurse co-ordinator at the time of drug administration. Desmopressin is a safe and very effective treatment for the prevention of bleeding at the time of dental work, minor surgery and for minor or moderate bleeding episodes. It can also

Treatment Options for von Willebrand Disease	
• <b>Desmopressin (DDAVP)</b>	– Raises vWF and Factor VIII levels by 3 to 5-fold in ~80% of patients
• <b>Humate-P</b>	– Plasma-derived vWF concentrate for treatment of major bleeding
• <b>Hormones (oral contraceptive) and clot stabilizing drugs (Cyklokapron and Amicar)</b>	– Useful in treating heavy menstrual bleeding and for prevention of bleeding with dental work and minor surgery

to investigate the underlying genetic changes in the common type 1 form of vWD. Over the next year, patients with type 1 vWD will be approached by clinics to invite their participation in this study, in which the vWF gene will be analyzed in over 100 families with this form of the disorder. It is hoped that this study, in conjunction with a similar research project

be used to reduce heavy bleeding associated with menses.

For the treatment of more serious bleeding episodes and in patients that respond inadequately to Desmopressin, a von Willebrand Factor concentrate, derived from plasma, is available for transfusion. The most frequently used concentrate for this purpose is Humate-P, which has been shown in studies, both in Canada and elsewhere, to be safe (no evidence of virus transmission) and very effective at replacing vWF function in the clotting process.

Overall, vWD produces more long-term problems in menstruating women than any other patient group. Studies completed recently in both Europe and North America have indicated that 10-20% of women with excessive menstrual bleeding have vWD and would clearly benefit from the treatment of this disorder. In this clinical situation, many health care centers are now beginning to initiate programs that provide combined hematologic and gynecologic expertise. This will enable women with a diagnosis of vWD to receive the best advice concerning the use of hormone treatments (forms of the oral contraceptive) and clot stabilizing drugs (e.g. Cyklokapron and Amicar) that are among the most effective methods of treating this problem.

### Von Willebrand Disease Projects in Canada

There are several ongoing efforts in Canada that relate directly to the diagnosis and treatment of vWD.

#### CHS National Awareness Campaign

The Canadian Hemophilia Society is in the first year of a three year project to raise public and physician awareness

about vWD. This project will involve extensive media communications and a variety of targeted educational events that will re-introduce the current concepts of vWD diagnosis and management to gynecologists and family physicians.

CHS has established an advisory committee composed of physicians, nurses and consumers to provide guidance into the planning and implementation of the project.

CHS has hired a part-time public relations consultant, Natalie Byk, to coordinate the awareness campaign.

#### Association of Hemophilia Clinic Directors of Canada Sub-committees

- **AHDCDC Women's Bleeding Disorders Scientific Sub-committee**
- **AHDCDC von Willebrand Disease Scientific Sub-committee**

The hemophilia physician's group, the Association of Hemophilia Clinic Directors of Canada (AHDCDC), now has two scientific sub-committees that are dealing with issues concerning vWD. A recently formed "Women's Bleeding Disorders" sub-committee (Chairperson, Dr. Christine Demers) will be addressing vWD problems that are exclusive to women, while the pre-existing vWD sub-committee (Chairperson, Dr. David Lillicrap) is involved in several long-term studies of vWD. These projects include the maintenance of a Canadian vWD Registry in which anonymous, encrypted data has been collated on over 800 vWD patients being followed by Canadian Hemophilia Clinics. The other major project that this Group is embarking on is a three year study funded by the Canadian Institutes of Health Research

ongoing in 7 European countries, will improve the complex and often unresolved diagnostic strategies currently in use for this subtype of vWD.

#### Canadian Type 1 von Willebrand Disease Research Project

*Funded for 3 years from the Canadian Institutes of Health Research*

**Objective:** *To determine the genetic changes associated with Type 1 vWD and, in doing so, to improve the diagnostic efficiency for this disorder*

**Study Plan:** *To perform coagulation and genetic testing on 150 families with proven Type 1 vWD*

**If you have Type 1 vWD and are interested in participating in this project, please speak to the staff at your local Hemophilia Clinic.**

### Concluding Remarks

There is now uniform agreement that vWD represents the commonest inherited bleeding disorder in humans. Although in most instances the disorder does not result in major spontaneous bleeding, it does produce long-term problems in menstruating women and can result in significant and prolonged bleeding after dental work and surgery. Confirming the diagnosis of vWD remains a difficult challenge in many cases, but once the diagnosis has been documented there are several safe and very effective means of preventing and treating bleeding. Future research will hopefully further enhance the choice of treatment options for vWD and also provide a more effective means of establishing the diagnosis.

# Profile

## Dr. David Lillicrap

by Barry M. Isaac, Ph.D.

Born in the United Kingdom, Dr. David Lillicrap received his medical education at St. Mary's Hospital Medical School, University of London, the very place where Dr. Fleming discovered the "miracle" drug, penicillin. But his interest in medicine was sparked years earlier by an aunt, Muriel Roffey, who was a Nursing Sister and who served as Dr. Lillicrap's role model. He says that, like her, he "was lucky; I never wanted to do anything else" than medicine, and this was reinforced each year when he and his family would make a Christmas visit to his aunt at the hospital where she worked. This early interest was married to a desire to know the puzzles as to how the human organism works: "I was always curious – wanted to know the answers." And Canada was the eventual lucky nation to benefit from his curiosity and desire to know how the human genome worked.

Dr. Lillicrap moved first to Edmonton and the University of Alberta in 1978, but then moved to Queen's University, Kingston, Ontario, in 1980. Dr. Lillicrap has lived in Kingston since then with his wife, Mary, and 3 children – Tim, (19) Kate, (18), and Anna (16).

He says that he moved to Queen's to "avoid" the subject of blood clotting, but that he came under the wing of Dr. Alan Giles, already doing groundbreaking work with his colony of hemophiliac dogs. His interest in Hematology had grown since medical school where he discovered that it was a fascinating blend of clinical and laboratory sciences, offering challenges in both patient care and laboratory research in unique ways. So Dr. Giles' interest in hemophilia, the colony of hemophiliac animals, Dr. Lillicrap's interest in factor VIII, first realized when he worked with Drs. Arthur Bloom and Ian Peake on



*Dr. Lillicrap's current research interests in the genetics of von Willebrand Disease have naturally grown out of his work in clinical and research medicine.*

hemophilia, combined with the discovery of the factor VIII gene in 1984 all made for a serendipitous moment in Dr. Lillicrap's life. "I was terribly lucky," he says, to have been in the right place at the right time, working with the right people. One cannot help but wonder if this really was luck, or was it, like so many other "lucky" events, the result of hard work and intense curiosity.

Dr. Lillicrap's current research interests in the genetics of von Willebrand Disease have naturally grown out of his work in clinical and research medicine. But his work does not stop there, as he is also working on genetic therapy for factor VIII hemophiliacs. He feels that this may be available on a small scale in five or six years because the factor VIII gene is "middle aged," its genetic structure having been known for some 15 years. Factor VIII has been around quite a while as a model for a genetic disease, and advances in its therapeutic use should not be long in arriving. "We're busy," he says enthusiastically, remarking that it is an "exciting time" to be working in these fields and on these projects. The therapy will consist of injections of a dead virus that will carry the factor VIII genetic material and allow it to be produced in the patient's body and thus provide the missing clotting factor. But Dr. Lillicrap advises that the first factor to be made available through gene therapy in all likelihood will be factor IX simply because of its relatively small size. Dr. Lillicrap has been and is uniquely and fortunately situated for research into both factors as Dr. Giles' animal colony afforded "an extremely valuable model," he says.

When asked what has been the greatest advance in care for hemophiliacs, Dr. Lillicrap quickly replies, "the genetic revolution." Both the diagnosis and treatment of the disorder have been made more effective and safe by developments in genetic therapy and research. It is now easier to diagnose the disease and to treat it with genetically recombinant clotting agents, which are far safer than simple human clotting agents, all as a result of the genetic revolution. But "the icing on the cake," says Dr. Lillicrap, will be gene therapy for hemophilia. In fact, it will be easier to "sell the human genome project," says Dr. Lillicrap, once an effective genetic therapy is developed for hemophilia. That project, now in its late stages, is a map of the entire human genetic content, a description of the complete human genetic picture. Once this project is complete, it should be possible to provide genetic therapy for any disease the human organism has. That is the current research thought on the subject.

Beyond these steps, Dr. Lillicrap feels that the future holds some answers to questions that currently puzzle researchers, questions such as, why does type I von Willebrand Disease occur? He also suggests that some advances in treatment, such as a "super factor VIII" and an increase in the half life of clotting factors, will be available "within two to three years." Advances such as the "super factor VIII" will result from using types of the molecule other than the normal one, once again a product of the genetic revolution.

Dr. Lillicrap's enthusiasm and warmth makes anything seem possible, and we are once again most fortunate that he, like so many other care givers and researchers, has chosen Canada as his home, a home for both his family and his research interests.



David Page, Chairperson, CHS Blood Safety Committee



## Octostim® Spray Available, Not Widely Used

*Effective treatment for Type 1 von Willebrand Disease and mild hemophilia A*

A new treatment for Type 1 von Willebrand Disease (vWD) and mild hemophilia A has been available in Canada since late 1998 but is not yet widely used. Octostim® Nasal Spray, manufactured by Ferring Pharmaceuticals, is the brand name for desmopressin acetate delivered through a convenient nasal spray, ideal for home use.

Other forms of desmopressin acetate have been licensed in Canada since 1993. These include DDAVP® Injection and Octostim® Injection. As the names imply, both these products were administered by injection, either intravenously or subcutaneously. None of these products are made from blood.

All three forms of desmopressin act by releasing von Willebrand Factor (vWF) stored within the body. Von Willebrand Factor is one of the proteins necessary in blood clotting. In Type 1 von Willebrand Disease, characterized by low levels of vWF, the vWF levels are temporarily raised high enough to stop bleeding. In mild hemophilia A, the increased amount of vWF carries additional quantities of factor VIII, the protein lacking in this condition, to the site of bleeding. Desmopressin is of no value in Type 2B or Type 3 vWD, nor is it useful to treat severe hemophilia A. However, some people with Type 2A vWD do receive some benefit.

Octostim® Nasal Spray comes in a spray pump bottle with 25 doses. A single treatment usually involves 1 dose sprayed into each nostril. The treatment has been proven effective in a number of situations including

minor surgery, dental work, menorrhagia, nosebleeds, bleeding in the mouth, muscle bleeds and joint bleeds. Because it is so easy to administer, people quickly learn how to use the product at home. This means many fewer trips to hospital or to the doctor.

Octostim® Nasal Spray is regularly prescribed by physicians across Canada's network of Hemophilia Treatment Centres.

Octostim® Nasal Spray can be used for children as young as 1 year of age.

The drug is not without side effects.

It causes the body to

retain water. Consequently, people taking desmopressin are advised to decrease their intake of fluids. This is especially important for young children and older people. Other side effects can include flushing, headaches and sleepiness. People with a history of high blood pressure or thrombosis (blood clots) may not be able to take desmopressin.

One reason why Octostim® Nasal Spray has not achieved wide acceptance is the cost. One spray pump bottle of 25 doses, or 12 treatments, costs almost \$400. Some provinces cover

the cost of the drug for people receiving social assistance. Also, people with private drug insurance plans can get a large percentage of the bill paid. However, this leaves many people across the country with no coverage. Some cannot afford the high cost.

Tom Alloway, CHS Vice-president, told *Hemophilia Today*, "One of the priorities for the CHS is services for people with vWD. This includes affordable treatment for everybody right across the country."

The Canadian Hemophilia Society has adopted the position that desmopressin is a blood alternative and should be made available in the same way as factor concentrates – free of charge to the user, distributed by Canadian Blood Services and Héma-Québec, and paid for by the provinces. Indeed, the alternatives to using desmopressin are even more costly – plasma-derived factor concentrates costing hundreds of dollars per infusion and frequent hospital visits. A failure to treat vWD leads to increased absenteeism from work and the possibility of serious complications from bleeding. The CHS position is supported by the Canadian Blood Services mission statement which says: "Canadian Blood Services operates Canada's blood supply in a manner that gains the trust, commitment and confidence of all Canadians by providing a safe, secure, cost-effective, affordable and accessible supply of quality blood, blood products and their alternatives."

**For more information on von Willebrand Disease, see *All About von Willebrand Disease*, published by the CHS or visit the CHS website at [www.hemophilia.ca](http://www.hemophilia.ca)**

Tom Alloway, CHS Vice-president, told *Hemophilia Today*, "One of the priorities for the CHS is services for people with vWD. This includes affordable treatment for everybody right across the country."

# THE FEMALE FACTOR



by Patricia Stewart

This section is related specifically to women with bleeding disorders and their families. All articles are reviewed by physicians to ensure medical accuracy. If you have any questions, comments or ideas, feel free to contact me, Patricia Stewart, at the following addresses:  
Phone & Fax: 418-884-2208 or e-mail: [davpage@zone.ca](mailto:davpage@zone.ca) or simply put pen to paper and mail to: 389, R.R. # 4, La Durantaye, Quebec GOR 1W0

In this issue, the *Female Factor* deals with the way women live with vWD. The first article is written by Renee Paper, a woman who was diagnosed with vWD in her twenties. She has been working for many years advocating for better vWD diagnosis and treatment. Ms. Paper works in Nevada as an ER nurse and is President of the Nevada Hemophilia Society. The second article presents the viewpoint of a young woman, Céline Cyr, who, although she had some major problems, also was not diagnosed until she was in her mid-twenties. The experiences of these two women shows the need for information on vWD both for the general public and for medical practitioners.

## Like Mixing Oil and Water?

### Women with Bleeding Disorders in Emergency Departments.

By Renee Paper, RN, CCRN

Reprint of an article which appeared in the July 1998, Vol 3 Issue 3 of HEMAWARE, published by National Hemophilia Foundation.

When I was asked to write this, I thought about the irony of having to write such a specialized article. The fact that such an article is even needed is a sad example of the care many women with bleeding disorders have reported receiving in emergency departments (ERs) throughout the country. Accounts such as “the physician didn’t believe a woman could have a bleeding disorder” or “the doctor had never heard of von Willebrand Disease” are all too common. In fact, reports about mistreatment at the hands of ER personnel throughout the country when managing males with hemophilia continue to abound, so one is not surprised to hear that women with bleeding disorders face the same mismanagement. For a woman, however, the issue of gender bias enters the picture.

Despite the facts that von Willebrand

Disease (vWD) is the most common bleeding disorder, at least 100 times more common than hemophilia, and that it affects males and females, hemophilia still has the most name recognition among not only the general public, but the healthcare community as well. Since hemophilia is a sex-linked recessive disorder affecting mainly males, most physicians automatically think of hemophilia when they hear about a patient having an inherited bleeding disorder. Those without knowledge or expertise in vWD may even assume a male with vWD actually has a form of hemophilia and that a female with “alleged” vWD is lying. Add to that a woman who presents with some of the rarer coagulation disorders, or even the symptomatic carrier state, and her chances of being believed and receiving appropriate management will probably decrease accordingly. After all, many hematologists do not readily acknowledge the symptomatic carrier state (which is actually no different clinically from mild hemophilia), so why expect clinicians less versed in coagulation disorders to do so?

Obviously, such behaviour and ignorance are unacceptable, but the cold, grim reality is that they still persist. So what can a woman with a bleeding disorder do to increase her chances of adequate and appropriate management at the hands of ER personnel? The following are some steps I propose every woman or caregiver of a girl with a bleeding disorder think about prior to having to utilize ER services.

#### Be realistic about your expectations for care in the ER

Why are you seeking ER care? Is it for the usual emergent conditions such as broken bones, lacerations, or burns are

you looking to the ER personnel to figure out why you keep having excessive menstrual bleeding and prescribe therapy to stop it? Such complaints are more appropriately handled by your hematologist in consultation with your gynecologist. ER personnel are not specialized in either. ER personnel are truly “jacks of all trades, but masters of few.” The reason for this is they see and

treat such an incredibly wide array of illnesses and injuries that they are likely to have true expertise only in those they see frequently, such as heart attacks, strokes, asthma, trauma, diabetes, fractures and lacerations. Assuming or even desiring that ER personnel have expertise in managing bleeding disorders is probably unfair and unrealistic. They cannot possibly know everything about every disorder. ER personnel increasingly are looking to patients with rare chronic disorders for guidance on how to manage them. For instance, if you need to go to the ER to have a wound sutured, they may look to you to guide them on how to stop or prevent the bleeding that

Hemophilia, vWD, and related bleeding disorders are chronic, lifelong disorders, just like diabetes. People affected by these disorders should learn early on how to manage them on a day-to-day basis.

may be associated with the injury or its treatment. They expect you to have knowledge of how to treat such bleeding episodes. And you should.

#### Look for alternatives whenever possible

Hemophilia, vWD, and related bleeding disorders are chronic, lifelong disorders, just like diabetes. People affected by these disorders should learn early on how to manage them on a day-to-day basis. Do not become dependent on ERs to do so. Diabetics do not go to the ER for their daily insulin injection, and also, people with bleeding disorders should not count on ERs to manage their clotting factor or DDAVP infusion needs. If you need frequent infusions of clotting

factor or DDAVP, either learn how to do them yourself, have a family member learn, or employ the services of a home health company that can send a nurse to your home to perform the infusion. This solution is quicker, cheaper, and less stressful than the ER. It returns some control back to the patient and their family and ultimately causes less disruption in their lives. Also, if you use DDAVP and have never tried the high potency nasal spray formula called Stimate, ask your physician about it. Most people who respond to the intravenous (IV) form respond adequately to the nasal spray form too. Your physician will give you a test dose to measure your response, and, if adequate, you may be able to avoid the need for IV infusions.

#### **Be prepared to educate the ER staff about your disorder and its management**

Most people with bleeding disorders who have had to utilize an ER can recount the infamous question by an ER physician, “so how long have you had hemophilia?” That is your first warning sign that you are in trouble. If the physician does not know that hemophilia and von Willebrand Disease are inherited disorders and that you were born with it, do not expect him or her to know how to treat it. Know enough about your disorder and its treatment to be able to explain it to the personnel. Make it short and sweet. ERs are busy places, and the ER staff will not have time to listen to the basics of hemostasis and the differences between intermediate purity and recombinant factor concentrates. Just tell them what they need to know to care for you. Have a letter or wallet card from your physician outlining your treatment regimen, including dosage, so they can administer the necessary treatment without delay. Make sure the letter includes your physician's name and number if the ER physician needs to contact him or her. Be sure to wear a Medic Alert bracelet that lists your disorder for those situations in which you may be unable to speak. The treating physician can call the number on the back of the bracelet and get your individualized treatment recommendations provided by you when you obtained the bracelet and which you have periodically updated.

#### **Bring your own factor or DDAVP**

If your treatment requires infusion of factor concentrate or DDAVP, it is best to bring it with you to avoid delays in

obtaining it. If you do not have factor with you, know where to obtain it in your community. If you are travelling to rural areas, it is best to carry factor with you because it may be completely unobtainable in a reasonable period of time in small rural communities. Know how to mix and administer the product. Again, this may be the first time those personnel have given clotting factor concentrates; the more you help them, the more efficient the procedure will be. Be sure the

letter from your physician tells them to round off the dosages for factor VIII or IX in order to utilize the entire vial contents, or the personnel are likely to draw up only the dose ordered and dispose of the rest. In almost all cases, it is cheaper if you provide your own product from home, as hospitals routinely mark up the prices of medicines to exorbitant levels to offset the costs of operating facilities 24 per day. Sure, some ERs will not want you to bring

**Continued on page 12**

## One Woman's Story

*Céline Cyr, Montreal, Quebec*

Growing up, I was always covered in bruises, big and small. I love being active, so I never deprived myself of sports and got plenty of bruises. Luckily I never suffered a serious injury.

I have always had heavy periods and have always had to be less active at that time. I had no choice because the bleeding was always too heavy for me to do anything, and I was always afraid my clothes would get stained, especially in public. My periods became heavier and heavier, and a few times I was bleeding so much that I had to go to the emergency room. I was even hospitalized for a hemorrhage (a period that did not end). When the doctor found no cause, such as miscarriage or fibroids he ordered blood tests and referred me to a hematologist.

I was 25 when I was diagnosed with Type 1 Von Willebrand Disease. I had already spent 2 years trying to find out what was causing my problem and it took me another 2 years, after the diagnosis, to find a doctor who really knew about the disease.

When I was diagnosed, my first reaction was “What is vWD, how did I get it and what should I do?” Then I wondered “Is there a cure?” My way of dealing with it was to get lots of information from books, the Internet and my doctors. I found out everything about the illness and I also got to know myself better, which is what enables me to live with it today. Nobody else in my family has vWD.

After I was diagnosed, the bleeding was brought under control, and I am now more careful about what sports I take part in. On days when the bleeding is heavy, I stay home from work and watch a movie or immerse myself in a book.

I am now on birth control pills all the time, to stop me from menstruating. I have a period about every 2 months and when the bleeding is excessive, I take cyklokapron and inject myself with DDAVP under the skin. This greatly reduces my problems.

Although I have not yet decided whether I want to have children, I know that vWD will be a big factor in my decision. I have one chance in two of passing it on, and if I do I know I will feel guilty.

The disease also has a big psychological impact. One lives in constant dread of being covered in blood in public or even just waking up in the morning in a blood-soaked bed. Even now, and in spite of the treatment, the fear is still there.



Continued from page 11

in your own product. They are likely to tell you it is against policy. Always ask to see a copy of such a policy. They rarely exist. If you know how to administer your own product and just need them to perform the venipuncture, then after they perform the “stick” go ahead and infuse yourself or your family member. They cannot stop you from medicating yourself or family member. You do not lose your rights just because you are in an ER. The best thing to do, however, is to infuse before you leave home. For example, if you need to use an ER for sutures, infuse yourself at home first so when you arrive in the ER you are a “non-hemophiliac.” You have managed that which you are able to and are now utilizing the ER for that which you cannot do, namely suturing.

### Stay informed and in charge

Be careful about consenting to invasive procedures unless you fully understand them, are sure of their necessity, and pre-treat ahead of time. For example, if the ER physician recommends an arterial blood gas (ABG) because you are complaining of a cough and shortness of breath, ask if there are any alternatives. An ABG involves drawing a blood specimen from an artery, usually in the wrist. This procedure can cause excessive bleeding and hematoma formation in persons with blood clotting abnormalities. A less invasive test is the pulse oximetry, wherein they attach a monitoring device to your index finger which tells them the percentage of oxygen saturation in your bloodstream. It does not involve any needles and often gives the physician enough information so that the more invasive ABG is not needed. Again, I am not expecting you to know the alternatives, just to ask if there are any, and fully understand the risks and benefits of a procedure before you consent to it. If ultimately you feel the procedure is warranted, be sure to pre-treat if the likelihood for bleeding post-procedure exists. An example of procedures that will usually require pre-treatment include lumbar puncture (spinal tap), naso-gastric tube insertion (if at all possible ask for the tube to be inserted orally instead of nasally), angiogram, chest tube insertion, and peritoneal lavage (a good alternative to this is a CAT scan which will reveal bleeding in the abdomen without having to insert a needle into it). The insertion of a peripheral (arms or legs) intravenous line usually does not require pre-

treatment, but the insertion of a central line into the chest or groin does because the risk of bleeding with that procedure is higher. Obviously, if you are in need of any of these procedures, you are probably seriously ill or injured, and making such decisions under those circumstances is difficult at best. If at any time you are unsure of what to do, insist that your hematologist or the nearest HTC be consulted prior to proceeding.

### Do not consent to unnecessary testing

This includes unnecessary blood testing. It always amazes me to hear about people with known hemophilia allowing an ER physician to order coagulation screening studies on them such as a PT and PTT. What did they expect to find? These tests will only confirm the fact that the person has some sort of bleeding disorder; it will not confirm the type! What was the purpose of such a test? The person already told the physician he had hemophilia. Sometimes even the doctor is not sure when asked why he or she ordered the test. There is no benefit to performing such tests. They prolong the visit and raise the cost. Refuse them. The same goes for X-rays of routine joint bleeds. In the absence of some sort of traumatic event preceding the bleed, there is little information the ER physician will garner from X-rays of the joint.

### Remember you can catch more flies with honey than vinegar

Always treat ER personnel with the same respect and dignity with which you wish to be treated. Behaving in a threatening or adversarial manner will not help the situation; it will only worsen it. You are in a crisis, and these professionals are there to help. You will need to understand and accept the fact that they will not have the expertise in managing bleeding disorders that you may desire. What they do possess, however, is the knowledge and skill to help you in a true emergency. Go in prepared to help explain the bleeding disorder and its treatment so that they can proceed to deliver the necessary care.

ER visits need not be horrible, frightening, powerless experiences. The power truly does rest with the patients if they will just assume that role. But remember, if you can avoid the ER in the first place, that is always best.

## WEAR A MOUSTACHE

Occasionally I do the walrus thing. You know, long white moustache hanging down – at least, half a moustache. It comes right out of my nostril. It is, in fact, a Kleenex. Okay, you caught me: it’s toilet paper. A big wad of toilet paper, jammed right up my nose and hanging down, getting in the way of lip-related activities. This is called doing the walrus thing. It’s also called having a nose-bleed.

My brother did it. I do it. It’s disgusting – but it’s also brazen, which is, I think, a good thing to be in the face of a morale-zapping medical condition. I do, by the way, know better. A nosebleed is meant to be faced patiently, sitting down, a cold cloth pressed firmly to the active area. I subject my daughter to the squeeze treatment, and it works – for her.

Personally, I can’t wait. I bleed slowly and persistently. And impatiently. Faced by gushing, I’ll give it a good squeeze. I’ll sit pretending the fingers on my nose are my mom’s, reminiscing about degrees of pressure and how nice it felt to have her hand resting on my cheek. But the results are so indefinite – it seems the blood never completely stops. There’s always a stubborn trickle: too big to ignore, but enough to make me think the grand squeeze is overkill. Cue the walrus.

Did I say it’s disgusting? It is! It’s one of those things that makes you appreciate marriage. My husband is used to me. It also helps to work at home. My computer is used to me, too.

But it’s one of the ways I cope: get up, get on with it. Laugh about it. There are worse things to be than ridiculous. Even disgusting isn’t necessarily bad. It’s all in the attitude.

The nose-squeeze is good; it has its place, and it’s an important one. So we all squeeze. But I suspect there’s also an army of secret moustache-wearers out there, can’t-hold-em-down walruses, dangling toilet paper in the privacy of their own homes. At least, I hope there are. I can’t be the only one.

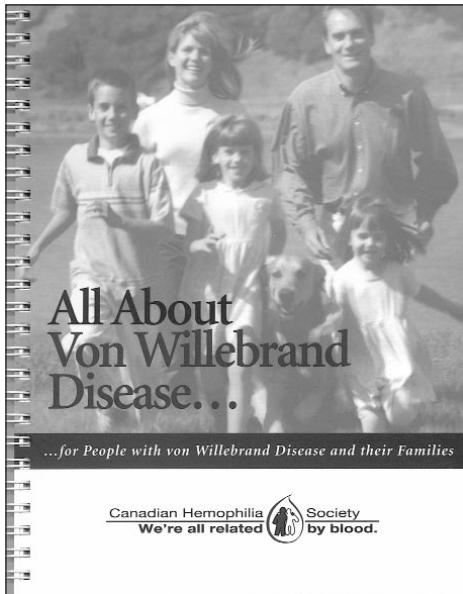
Can I?

Ania Szado, Toronto, Ontario

VON WILLEBRAND DISEASE RESOURCES AVAILABLE FROM THE CHS

VON WILLEBRAND DISEASE RESOURCES AVAILABLE FROM THE CHS

VON WILLEBRAND DISEASE RESOURCES AVAILABLE FROM THE CHS



Comprehensive guide book for people with vWD and their families

How can I control my heavy periods?

Why do I bleed so much at the dentist?

Why do I get so many nosebleeds?

Why do I bruise so easily?

**Your symptoms could be caused by von Willebrand Disease**

- von Willebrand Disease (vWD) is an inherited bleeding disorder caused by a problem with a blood clotting protein
- vWD affects over 300,000 Canadians
- most people affected are not diagnosed
- symptoms include:
  - heavy menstrual periods
  - easy bruising
  - frequent or prolonged nosebleeds
  - prolonged bleeding after injury, surgery, childbirth, or dental work
- vWD affects both females and males
- effective treatment is available

**If you think you might have von Willebrand Disease** you should be tested and treated by a hematologist – a doctor who specializes in blood. Ask your family doctor or gynecologist for a referral. Or contact the Canadian Hemophilia Society for more information. We're the largest national consumer organization working to ensure that, for people with bleeding disorders, the bleeding stops.

**Take the first step today to stop the bleeding**

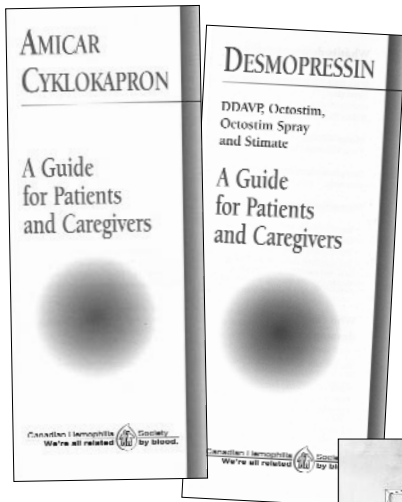
Call us toll free at 1-800-668-2686, contact us by E-mail at [chs@hemophilia.ca](mailto:chs@hemophilia.ca) or visit our web-site at [www.hemophilia.ca](http://www.hemophilia.ca)

**you CAN stop the bleeding...**  
von Willebrand Disease...  
the most common bleeding disorder

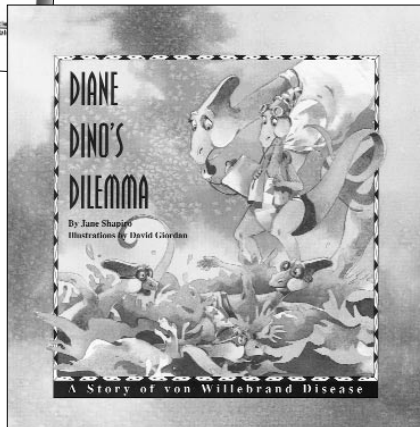
Aventis this public awareness campaign is made possible through an unrestricted educational grant from Aventis Behring.

Canadian Hemophilia Society We're all related by blood.

New vWD Awareness Campaign material

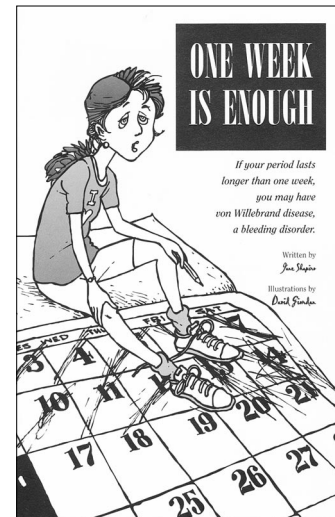


Patient guides on products for the treatment of vWD developed by Canadian Association of Nurses in Hemophilia Care (CANHC)



Children's storybook about vWD

Brochure for teens published by the National Hemophilia Foundation





## Providing Support to Families

In the Spring of 1992, my husband Ted and I discovered our fraternal nine month old triplets each had hemophilia. That day will forever more remain our personal definition of “overwhelmed”. I also remember the lack of educational materials available to help us cope and to understand more about hemophilia. Literature for children and parents, teaching videos, and support groups were not in abundance. Focus on new parents and young children was yet to come – even *Hemophilia In Perspective*, had not yet been published. So many great resources have been developed and made available over the years and it keeps getting better.

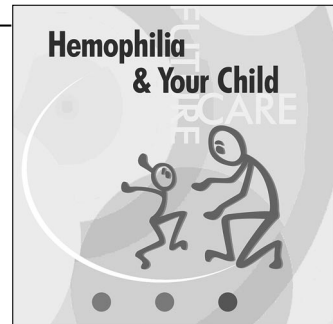
The plethora of available materials on hemophilia is a great accomplishment, as the needs have been identified, and resources have been utilized to produce great educational materials for children with hemophilia and their families. A new and exciting tool for parents is being launched entitled, *Hemophilia & Your Child*. It is an interactive, educational CD-ROM, which was developed by a group of hemophilia nurses, physiotherapists, physicians, parents and Bayer, in collaboration with the Canadian Hemophilia Society. It was created to address the immediate concerns of raising children with hemophilia and to help parents take an active role in the management of their child's hemophilia.

This free educational CD-ROM will be available in April 2001 through your local clinic or the Canadian Hemophilia Society. As one of the parents who have worked on this project, I hope it helps many families.

*continued on page 15*

## HEMOPHILIA & YOUR CHILD

*Bayer, in collaboration with CHS, Presents A New Interactive CD-ROM for Parents*



As parents, we want our children to be healthy and happy at all times. The reality is – every child encounters challenges. It's all part of growing up. That's why children need parents to help and support them through difficult times.

As parents of a child with hemophilia, you'll face many difficult challenges. The initial diagnosis can cause tremendous anxiety as you worry about your child's future and the risks of everyday life. As your child grows, you must constantly adjust and allow them the freedom and independence to mature, while ensuring their safety and well being.

“Fortunately, the anxieties, worries and frustration can be alleviated through education about how to manage the bleeding disorder,” said Erma Chapman, President, Canadian Hemophilia Society. “By being supportive and informed, parents can help their child – in fact, the whole family – deal with hemophilia in a healthy and positive way. Knowledge helps you to feel some control, the kind of control you need to enjoy family life on a day-to-day basis.”

There are many formal resources available to parents, including the hemophilia clinic team, The Canadian Hemophilia Society, parent support groups and books. A new and exciting tool for parents is the interactive *Hemophilia & Your Child* CD-ROM, which was developed by a group of hemophilia nurses, physiotherapists, physicians, parents and Bayer, in collaboration with the Canadian Hemophilia Society.

*Hemophilia & Your Child* is an interactive, educational CD-ROM designed to promote effective parenting skills. It was created to address the immediate concerns of raising children with hemophilia and to help parents take an active role in the management of their child's hemophilia.

“Caring for a child with hemophilia is a team effort involving the nurse coordinator, the hematologist, parents and most importantly – the child. We are all working together to help give the patient the best quality of life,” said Nora Schwetz, Bleeding Disorders Nurse Coordinator, Winnipeg Health Sciences Centre. *Hemophilia & Your Child* will certainly be a worthwhile tool to help parents learn more about their child's hemophilia. We are extremely appreciative to Bayer for their ongoing support and funding of education programs in Canada.”

Developed by Digital Rain, a new media company with expertise in computer oriented patient education tools, the CD-ROM was designed so that the program has no specific beginning or end. The viewers explore the program, choosing links to topics of immediate interest while progressing at their own rate.

There is a main menu with eight major topic headings such as Growing Up with Hemophilia, Living Safely, and Understanding and Assessing Bleeds. Each section provides a carefully chosen collection of sub-topics that are hot-linked so that when the viewers click on a link, they can jump to different types of related information, such as the Glossary and Resource Section. Simple animation and video footage of parents discussing their experiences help to illustrate certain topics. Although targeted primarily to parents, there is also a section for children with Hemophilia-related puzzles and games.

“Given Canada's dispersed population, it is often difficult for families to attend clinic programs. In such cases, an effective learning resource is an interactive program that will make accessing information easy. *Hemophilia & Your Child* is portable and can be used in any computer with a CD-ROM drive – at home, work, at the library or school,” said Rena Battistella, Market Manager, Hemostasis, Bayer. “A tremendous amount of work went into developing *Hemophilia & Your Child*, and the Bayer team wants to extend a heart-felt thank you to everyone who was involved.”

This free educational CD-ROM will be available in April 2001 through your local hemophilia clinic or the Canadian Hemophilia Society.

*“Caring for a child with hemophilia is a team effort involving the nurse coordinator, the hematologist, parents and most importantly – the child. We are all working together to help give the patient the best quality of life.”*

I can easily understand how parents of children with von Willebrand Disease have struggled with the lack of understanding about this condition and the lack of available resources to support them. Historically, little had been done to gather families and children together to discuss their experiences and concerns. That changed when a mother of 2 children with von Willebrand Disease decided to get involved with her local hemophilia chapter, where she subsequently tabled her idea to have a workshop for families about von Willebrand Disease. The first von Willebrand's workshop was held in Hamilton in 1997. For many it was the first time they met other families dealing with von Willebrand Disease.

Caroline Mulder-Sutton is the mom who became involved at CWOR (Central West Ontario Region). She continues to support her chapter, is a Board member at CWOR and remains involved in many continuing projects for families and children with von Willebrand Disease and hemophilia.

I spoke to Caroline about Desiree's article. She and Alan are very proud of Desiree's initiative and what she wants to share. For the Sutton family, living life is a key lesson and not letting the bleeding disorder live your life for you is something they want both their children to accept.

I asked Caroline what has helped her cope so positively with the added challenges of parenting children with bleeding disorders. Caroline works part time, so making sure the schools are well informed is key; a cell phone gives her peace of mind and freedom to not have to be with the kids or at home. She and Alan have helped the kids choose activities that make sense for each of them, and in her own words,

"You get tired of saying no, so we have taught the kids that they have to take responsibility for their own actions and their own bodies. We have made sure they know all about first aid and the appropriate responses for injuries they incur. We have tried to teach them how to cope with their bleeding disorder and to make sure they lead normal lives. I also think I have learned so much by being involved at CWOR and with the CHS von Willebrand Advisory Committee. How we cope with things teaches our kids to cope too."

Congratulations Desiree. Thank you for sharing your article with us. Congratulations to Mom and Dad also. It seems your efforts are paying off! Perhaps Brendan will write to us some day too with his thoughts on having a bleeding disorder?

Take care and keep in touch.

Karen Creighton  
creighton@idirect.com



The following article is written by Desiree Sutton. Desiree is 12 years old and is a grade 7 student. She and her brother Brendan who is 9, both have von Willebrand Disease. They live in Cambridge with their mom, Caroline and father Alan, who also has Type 1 vWD.

*Having a bleeding disorder can be fun. A bleeding disorder can not be seen unless you are bleeding. You might bleed for a couple of minutes, or for a day. If you know what to do, and the right way to do it, then nobody needs to know about it, or even care too much about it. It's a part of you, and me for that matter. You can look at the disorder in two ways, the many things you can't do, or the many things you could do. You might not be able to do boxing or things like that, but you might play an instrument or take up hiking or canoeing. You can find something that interests you and devote your time to it.*

*The first thing you should do when you start school and have gym is tell the teacher about it. The sooner you get it over with, the better. I should know. If the teacher fusses over you, just tell him or her it is no big deal. Unless, of course, it is a big deal to you and to them. You because you didn't tell them. Them, because if you told them, they might have prevented the injury from happening to you.*

*Another thing that might help with your disorder is to not be afraid of blood. The colour of blood, the sight of blood, the taste or even feel of blood. You name it (if you can find anymore senses other than hearing, but I don't think you can hear blood. Can you?) Most people might not think this has anything to do with a bleeding disorder, other than you get to see more of it than other people.*

*Don't hold yourself back too often, or you might miss a lot of the things that make life worth living. What's a little scrape or two if it meant that you get to be with your friends. Also, don't over extend yourself. Know where your boundaries are and if you don't know where they are, find out. You might find something about yourself you never knew before. I'm finding new things every week. I tried playing the flute at school and ended up in the band. I'm a 'fish out of water', in the summer. You almost have to drag me out of the pool. In the winter, I go skating. My real talent seems to be in the water, not on it, even if it is frozen. I keep on asking for pads to go on my butt for all the times I fall down. I heard somewhere that you learn something new every day. You might not think so, but look back at what you learned at school. You would not even be able to read this if you didn't go to school. You hopefully didn't even have to put too much effort into reading, you just do it, you weren't born able to read, you had to learn it. I hope you learned something reading my thoughts, because I certainly did, writing about them. Thanks for reading my thoughts.*

Desiree Sutton

# YOUTH FILE

## Living with vWD



Jean-Daniel flying high at Club Med circus school

### The Guys's Perspective

**Jean-Daniel Beaubien**  
19 years old – Type 2A vWD  
Montreal, Que.

Jean-Daniel was diagnosed with Type 2A vWD as a baby. His mother and sister also have it. Since he has always lived with the knowledge that he has this disease, he simply accepts it.

He played a lot of sports when young. Of course he had more bruises than every other kid, and when he'd get hit in the face with a hockey stick, he'd often end up in the hospital, but he'd go back out and do it all over again...

As a child, there wasn't any synthetic treatment and the blood-based coagulants were available but the risk of HIV was too scary. So he would either tough it out or go to the hospital where they would pack his nose. They would fill his nose up to the sinuses with cotton bands (lubricated with Vaseline). "It's horrible, one of the most painful procedures I ever got in my life" says Jean-Daniel. Today he uses DDAVP nasal spray.

"I rarely think about the fact that I have vWD and it has never caused me a life-threatening problem. Most of the time when I think about it, it's because I'm already bleeding. I snowboard regularly; I train at a gym and

basically do anything I want. I always wanted to take boxing classes, but I decided I'd be better off not to."

His friends are all aware of his bleeding problem, although only a few have ever seen him bleed. "I've had the same friends for maybe 13-14 years now. I guess they're surprised sometimes

when I start bleeding in front of them, they aren't used to that kind of bleeding. But", he says, "we still have no problems punching each other's arm until it hurts..."

Jean-Daniel adds: "Perhaps the only thing I'd like to add is this. When I was little, the doctors told me I should never ski, play football, etc, etc... Skiing turned out to be the most fun I ever had in my life, perhaps. Unless you have a really serious case of vWD or hemophilia, go out, and try things, even if they say you shouldn't... just be a bit more careful I guess.

**William Dutot**  
17 years old – Type 3 vWD  
Port-aux-Basques, Nfld.

As a baby, William had problems with teething, actually drooling blood and he had so many bruises that there were rumours that he was being abused. He was diagnosed with Type 3, the severe form of vWD, when he was about nine months old. His mother has been diagnosed with a milder type of vWD.

His earliest memories are of being all "banged up" just by running around the house. William has always lived with the knowledge that he has vWD. He has accepted the limits that this type of vWD puts on him and avoids rough contact sports. He also skips

physical education classes. His interests include curling, swimming ("if only there was a swimming pool around") and he enjoys art.

He says his friends know that he has a bleeding disorder because it's so evident. He explains what vWD is to them. William has learned to live with Type 3-vWD and places his own limits on his activities, but feels that it doesn't take over his life. He just deals with it and goes on with everyday living.

**Matt Strmic**  
12 years old – Type 3 vWD  
Boulton, Ontario

Matt was diagnosed with hemophilia at the age of two when he fell and bit his tongue and it wouldn't stop bleeding. Both his parents were then tested and, 3 months later, the diagnosis was changed to Type 3 von Willebrand Disease. His sister has a mild form.

He wasn't a very adventurous kid and preferred playing with toys. He says this was probably lucky for him and his parents. He had few problems until he was six when he had a gastric bleed, a frightening experience. It was then that he began to realize that something was wrong.

His everyday life is affected only when he can't join his friends in certain activities. And at times he has to explain all his bruises. However, this doesn't stop Matt from taking part in many sports. But his passion is swimming. Matt would rather swim than do anything else. He feels that it has helped limit his bleeding episodes because it develops muscles which help cushion the joints. He joined the Vaughan Aquatic Club 5 years ago and now swims almost daily. He has close provincial times in the 100m and 200m backstroke and regularly participates in swim meets. His coaches know about his condition

and are very encouraging. However, Matt had a big disappointment last June when a hip bleed prevented him from participating in a meet where his team won the gold. But one of the older boys sent him a medal that he'd won. That's true friendship!

Matt attributes his success to a close family and supportive friends. It helps him get through difficult times. His mother is his medical advocate, his father "is in charge of the fun department", while his sister, Katie, comforts him when he's hurt, is the first one with the ice pack ready and always cheers him on.

"Our family has a kind of philosophy or attitude which I feel has really helped me to keep positive about things. We accept things we cannot change and learn to adapt to situations supporting each other every step of the way."

## A Young Woman's Perspective

**Heather Carlson**  
24 years old - Type 1 vWD  
Toronto, Ontario

I was diagnosed at the age of 17. My younger brother had some other medical issues and one of his doctors had recognised a problem with bruising and suggested that he be tested for a bleeding disorder. When testing resulted in a diagnosis of vWD, the doctor suggested that my mother should be tested due to her history of bleeding problems. She didn't even consider the possibility of my having vWD as well. We were just chatting about her upcoming appointment to be tested and what symptoms she had when I suggested that I showed all the same symptoms. I accompanied her to her appointment and shortly after we were both diagnosed with vWD Type 1. I think the word relieved is the best to describe my reaction. All of a sudden things made sense.

Prior to being diagnosed I had had a bleeding problem when my tonsils were removed and when baby teeth fell out. And I have always had problems with my periods being very heavy and painful. In fact shortly after I started menstruating my family doctor sent me

to a gynecologist and I was put on birth control pills in order to lessen the duration and pain involved. At that time I would be off school for 2-3 days each month. I continue to have problems with periods like mid-month spotting, 2-3 week periods a couple of times and the most embarrassing for a young girl, bleeding through clothing at times. I learned early to wear dark colours at that time of the month.

I now use DDAVP and Octostim and I do my injections myself at home. I must admit I avoided learning how to do this for quite some time. My mother is a nurse so it was easier to get her to do it. The nurse co-ordinator at the hemophilia clinic finally talked me into learning to inject myself. I use a little needle that goes just under the skin and I inject a fairly small amount of liquid. All the hemophiliacs I know are quite jealous of the ease of my injections. I don't need them often enough to get really comfortable giving them, but after a few times I was more relaxed about it.

Von Willebrand Disease doesn't really affect my every-day life now. I just have to be a little careful. My biggest issue now is that I'm a klutz. I'm famous for hitting my head or walking into things. Anyone who knows me can attest to this. I have to be careful to ice any good bumps to my head and pay attention if I get any headaches. If I think I'm really going to bruise I'll take an injection. Some times if I cut myself it doesn't want to stop seeping a little bit of blood. I'll hold pressure on it a little longer than the average person. For me I'm just being a little more careful when I hurt myself.

Most of my friends know about my diagnosis but only because I've chosen to tell them. In my case, and like most



Heather Carlson and Karttik Shah

persons with vWD, if I didn't tell people they would never know.

I guess the reason I was so willing to share my story is to let other young women with vWD know that there are others out there like them. And, girls, I'm beginning to feel a little outnumbered by all of these hemo guys. Now

don't get me wrong, guys; you're all great, and some of you are my dearest friends. But I would like to see more young women with von Willebrand involved in the Society. In my 4 years as a member I've been involved in weekend retreats, canoe trips and I was fortunate enough to go to Montreal for the World Federation of Hemophilia Congress. At the youth pre-congress meeting, only one other vWD female was present, Anna from Sweden. She became a friend and I hope to see her at the next conference in Spain 2002. Come on, girls; we can't continue to let the guys

I guess the reason I was so willing to share my story is to let other young women with vWD know that there are others out there like them.

have all the fun.

I would like to hear from any other young women with vWD. You can contact me at the address below:

Heather Carlson  
552 Birchmount Road, # 704  
Scarborough, ON  
M1K 1P4

or forward your comments to Karttik Shah, editor of the Youth File at [karttik.shah@sympatico.ca](mailto:karttik.shah@sympatico.ca)

## CHS HONOURS PAST AND PRESENT AWARD RECIPIENTS

By Frank Bott,  
Past Chair, National Awards Committee

The Briars on Lake Simcoe was the setting for the annual **CHS Awards Banquet** on November 25th, 2000. Frank Bott, retiring Chair of the National Awards Committee, was Master of Ceremonies. President Erma Chapman announced the appointment of Barry Isaac as the new chair of the committee. Frank Bott introduced the previous year's committee – Barry Isaac, Normand Landry, Patricia Stewart, Eric Stolte, and Pam Wilton. He also acknowledged the excellent work of Clare Cecchini, the staff resource person during the five years he had been committee chairperson.



**The Chapter Recognition Award** was presented to Sheila Comerford and Patricia Stewart of the Quebec Chapter by Erma Chapman, in recognition of their fundraising which resulted in a 50% increase in revenues. Successful fundraising initiatives

included a Corporate Campaign, a golf tournament, a lottery, and sale of Christmas colouring books.



**The Chapter/Region of the Year Award** was presented by Erma Chapman to Pam Wilton and Mike McCarthy of the Southwestern Ontario Region for its outstanding efforts, in collaboration with the clinic team, to

ensure the maintenance of the comprehensive care program during its transfer to another institution. The Program Council continued to be an effective mechanism to facilitate collaboration between consumers and health care providers.



**The Award of Appreciation** went to Nora Schwetz, nurse-coordinator in Winnipeg who has, over and above her professional dedication and devotion, contributed volunteer hours to education, family camp, and group facilitation. As well, she has been Co-Chair of the CHS Nursing Group and helped to develop educational resources for patients. She has also been

active on the international scene. The award was presented to her by the President of the Manitoba Chapter, Frank Figler.



**The Dr. Cecil Harris Award** was presented by Dr. Sue Robinson, Chair of the Medical and Scientific Advisory committee to Dr. David Lillicrap of Kingston, recognizing 15 years of devoted work to improve the quality of life for persons with hemophilia. Dr. Lillicrap has an international reputation as a researcher in the area of gene therapy. He is one of Canada's

leading authorities on von Willebrand Disease and serves as Chair of the AHCDC sub-committee on vWD and is a valued member of the CHS vWD Advisory committee. In 1997 he created the Canadian registry for patients with von Willebrand Disease. He has served for many years on the CHS Medical and Scientific Committee.



**The Chapter Leadership Award** was given to two sisters, Lois Bedard and Joyce Rosenthal, who were very much a team in the founding and early development of the Ontario Chapter, including setting up the structure of the first provincial chapter, and early educational and

advocacy efforts on behalf of persons with bleeding disorders. As well, they were both actively involved in the national organization, Lois at one point being on the national Executive Committee, and Joyce as the first national Executive Director of the CHS when the office was in Toronto. Erma Chapman and Tom Alloway, representing Hemophilia Ontario, jointly made the presentation.



The recipient of this year's **Frank Schnabel Award** was Barry Isaac of Calgary. Frank Bott made the presentation, citing Barry's achievements over at least 30 years, including his influential involvement in upgrading and developing comprehensive care in

Calgary; years of serving at regional, chapter, and national levels of the CHS; his valuable advisory role in HIV and hepatitis C compensation, the Krever Inquiry, and blood safety issues. For the past ten years he has been editor of *Hemophilia Today*. He has been a member of the National Awards committee since its inception, developed the criteria for the CHS Scholarship and Bursary Program, and is the current chair of the Scholarship Review Committee.

## NOMINATIONS FOR YEAR 2000 NATIONAL AWARDS

The nomination package for the year 2000 National Awards has been sent out to chapters and clinics or may be obtained by calling the National Office. This year a new award, in memory of Pierre Latreille, is being introduced to recognize CHS staff.

Please consider nominating an individual from one of the following categories. The National Awards are:

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### CHAPTER / REGION OF THE YEAR AWARD

The award is designed to recognise a chapter or region that has demonstrated excellence over all, during the past year. It is awarded to that chapter which has shown outstanding efforts in a variety of areas such as advocacy, fundraising, patient services, education, or chapter/regional development.

Previous Recipient  
Manitoba Chapter

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### CHAPTER RECOGNITION AWARD

This award is designed to recognise chapters that have demonstrated an achievement over the preceding year in a specific area such as fund-raising, patient services, education, or chapter/regional development.

Previous Recipients:

B.C. Chapter  
CWOR  
Hemophilia Ontario  
Manitoba Chapter  
Newfoundland  
Labrador Chapter  
Nova Scotia Chapter  
PEI Chapter  
Quebec Chapter  
Saskatchewan Chapter  
TCOR

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### AWARD OF APPRECIATION

This award honours an individual who has demonstrated outstanding service to the care of persons with hemophilia or related bleeding disorders.

Previous Recipients:

Ann Harrington, *Nurse Co-ordinator*  
Jane Neil, *Physiotherapist*  
Lois Lindner, *Nurse Co-ordinator*  
Muriel Girard, *Nurse Co-ordinator*  
Jack McDonald, *Ph.D., Social Worker*  
Brenda Blair, *Nurse Co-ordinator*  
Lorraine Bernier, *Nurse Co-ordinator*

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### DR. CECIL HARRIS AWARD

This award honours distinguished contributions in the areas of hemophilia-related research or the advancement of the care of patients with hemophilia or related bleeding disorders. It is named after the late Dr. Cecil Harris, in recognition of his contribution as one of the pioneers in the care and treatment of hemophiliacs in Canada.

Previous Recipients:

Dr. Irwin Walker  
Dr. Georges-Étienne Rivard  
Dr. Martin Inwood  
Dr. Kenneth Shumak  
Dr. Man-Chiu Poon  
Dr. Kaiser Ali

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### CORPORATE LEADERSHIP AWARD

This award honours outstanding corporate leadership which fosters commitment to the spirit of voluntarism on behalf of hemophilia on a national level.

Previous Recipient:  
Bayer Inc.

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### CHAPTER LEADERSHIP AWARD

This award is given to an individual who has merited special national recognition for having provided exceptional leadership and devotion to a specific CHS Chapter over many years and for outstanding efforts to further the growth and development of the chapter.

Previous Recipient:  
Normand Landry, New Brunswick

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### HONOURARY LIFE MEMBERSHIP AWARD

This award is given to an individual who has demonstrated exceptional leadership and devotion to the CHS over many years. The recipient has merited special recognition for continuing efforts, particularly at the CHS Board level, to further the growth and development of the mission and objectives of the CHS and the development of public recognition of the CHS and its goals at the National and Chapter level.

Previous Recipients:

James Kreppner  
Durhane Wong-Rieger, Ph.D.  
Pierre Fournier  
David Page  
Jack McDonald, Ph.D.  
Elaine Woloschuk  
Joyce Rosenthal

Kenneth E. Poyser  
Dr. Martin Inwood  
Pat Harris  
Dr. C.E.C. Harris  
Ronald George, Ph.D.  
Leni George  
Frank L. Schnabel

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### FRANK SCHNABEL AWARD

This award was initiated to honour the outstanding service of Frank Schnabel, the founder of the Canadian Hemophilia Society, for his valued role in the growth and development of the CHS, the education of and care of hemophiliacs, and the education of the public regarding hemophilia needs. The award is presented in his name to honour a volunteer who, over a number of years, has rendered distinguished services and noteworthy contributions to the mission and objectives of the Canadian Hemophilia Society.

Previous Recipients:

Erma Chapman, Ph.D.  
John Plater  
Ken Little  
Durhane Wong-Rieger, Ph.D.  
Frank Bott  
David Page  
Peter Wachter

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### PIERRE LATREILLE AWARD

This new award for excellence will be given to a staff member of the CHS working at either the national, chapter or regional level who demonstrates the qualities that were shown by the dearly appreciated CHS staff member for whom it is named, Pierre Latreille. Pierre worked as Finance Manager for the Society from September 1991 to October 1999 and served as acting Executive Director from November 1998 to July 1999. He passed away on October 29th, 1999 in the CHS office while at work at his many duties.

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### SPECIAL AWARDS OF APPRECIATION

Special awards in recognition of outstanding contributions to the Canadian Hemophilia Society have been awarded over the years to the following persons:

Previous Recipients:

Robert Gibson  
George Moody, MPP, Nova Scotia  
Paul Ramsey, MPP, BC  
Russel Williams, MNA, Quebec  
Bonnie Tough  
Kathy Podroberak  
Ed Kubin

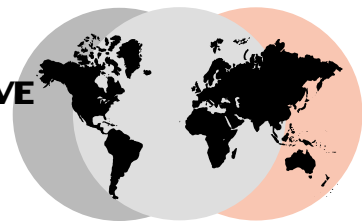
Over the past several years there has been an increasing sense that the CHS needs to step up to the challenge of what it means to be part of a global community. There are countries where the average life expectancy of a person with hemophilia is under 25 years. Still, over 75% of people with hemophilia around the world die from bleeding. Being so well resourced, we must help those in our world-wide bleeding disorder community whose resources are severely low.

To keep our readers informed about international issues we are introducing a new column, The Global Perspective, which will feature topics of interest about the global community. This first issue will include an article about the newly established CHS International Projects Committee and a report on the Quebec Chapter's recent visit to Senegal. Please forward any comments or suggestions for future topics to me at [estolte@home.com](mailto:estolte@home.com)



By Eric Stolte,  
Chair, International  
Projects Committee

## THE GLOBAL PERSPECTIVE



### CHS INTERNATIONAL PROJECTS COMMITTEE

Having helped to host the WFH's World Congress last summer, an agreement was struck that will give the CHS somewhere around \$150,000. It was decided to put those funds to use outside our borders. To enable that, and to give longer term direction to our global responsibilities, the CHS struck what has come to be called the International Projects Committee that I have the privilege of chairing. It's exciting to think of the difference we can make as we help, both organizationally and medically, people with bleeding disorders in needy countries.

Our first goal is to figure out how best to distribute the funds we've been given. Already requests have come both from Canadian chapters who are working with other countries as well as from other countries themselves. Exactly what our priorities are and how we can strategically contribute to long term improvement in bleeding disorder care will be the committee's first challenge. We're eager to begin to put funds in the hands of worthy initiatives as soon as possible. We'll look forward to keeping you abreast of future developments as we get going.

## QUEBEC FORMALIZES TWINNING AGREEMENT WITH SENEGAL

**François Laroche**, Vice-President  
Canadian Hemophilia Society,  
Quebec Chapter

From November 12 to 20, 2000, the Canadian Hemophilia Society - Quebec Chapter (SCHQ) undertook a field trip to Dakar to formalize its Twinning Agreement with l'Association sénégalaise des hémophiles (ASH).

I had the opportunity of heading this mission. Accompanying me were Claudine Amesse, Hemophilia Nurse Coordinator, Sainte Justine Hospital in Montreal, and Louis Gibeau, representing the World Federation of Hemophilia (WFH).

Our trip corresponded with one of the main objectives of the WFH, namely to promote the advancement of hemophilia associations in developing countries. This in turn will enable a network to be forged between the associations, leading to bilateral cooperation and productive cultural exchange.

SCHQ first made contact with ASH during the International Congress of the WFH in Dublin in 1996. More intensive discussions took place at the 1998 Congress in The Hague and in Canada, at the Montreal Congress, in 2000.

Why did we choose ASH? Because

Quebec and Canada have always had extensive cultural and commercial relations with Senegal. In addition, ASH seemed to be a solid organization run by dynamic people dedicated to improving the quality of life for hemophiliacs. Last but not least, synergy and a feeling of trust soon built up between everyone involved, especially the president, Ms. Anta Sar.

We spent a busy week with friendly people who made us feel very welcome. Our colleagues in Senegal did everything to make things easier and treated us as VIPs.

Among the highlights of our program were a tour of the national transfusion centre, a meeting with ASH administrators and members during Hemophilia Day, a visit to the Canadian embassy in Dakar, a meeting with the director of the office of the Minister of Health, interviews with the Senegalese press and electronic media, a tour of Dakar's three hospitals, a short excursion outside the capital and, of course, the signing of the Twinning Agreement.

The goals of the partnership are: to disseminate the operational model of the SCHQ and the hemophilia treatment centres, including their development, possibilities and limitations; to support ASH in raising the awareness of Senegalese deci-

sion-makers in order to ensure hemophiliacs receive adequate treatment from health professionals with expertise in this type of treatment; and to organize bilateral exchanges conducive to developing our mutual skills and knowledge.

It was agreed that the Twinning Agreement would be signed for two years and then re-evaluated. For this type of project to succeed, both parties need to have the same expectations and common, realistic goals.

This field trip taught the SCHQ a lot about life and customs in Senegal. It gave us the chance to make friends with wonderfully warm, welcoming people who are easy to work with. Our partnership promises to be extremely productive for the people of Senegal and Quebec.

My thanks go to WFH representative Louis Gibeau for his sound advice and support, which were indispensable on such a mission abroad.

I also salute the contribution of Claudine Amesse, Nurse Coordinator of the hemophilia clinic at Sainte Justine Hospital. In projects like this, being able to rely on the support and expertise of a hemophilia treatment centre is paramount.

Finally, I acknowledge the tireless dedication of ASH president Anta Sar; her kindness and generosity during our stay knew no bounds. Thank you, Anta, we hope to meet again soon.