THE NEXT DECADE:
Great challenges in hemophilia
The Canadian Hemophilia Society (CHS) relies on the generosity of our donors to fulfill our mission and vision. We are fortunate to count on a group of exceptional donors who have committed to making an annual investment to support the CHS and its core programming needs.

To recognize this special group of donors we have created the BeneFACTORS Club, the CHS’ highest philanthropic recognition, which symbolizes the critical bond between our organization, the donor and every person we serve with an inherited bleeding disorder. Corporations that make annual gifts of $10,000 or more to support our organization and its core programming needs are recognized as members of the BeneFACTORS Club.

The Canadian Hemophilia Society acknowledges their tremendous effort.
Each new year gives rise to review and reflection, and new resolutions. With the arrival of a new decade, there is often an added sense of excitement and hope. It's almost as if we look forward to better times ahead, as if times passed have not met our hopes and expectations...

In fact, over the years there have been many achievements and milestones in the treatment of inherited bleeding disorders. From the giant steps undertaken in comprehensive hemophilia care over the last 30 years to the advent of prophylaxis and the identification of the different types and facets of inherited bleeding disorders, there is no lack of examples testifying to the concrete results achieved through the dedication and hard work of the hundreds of healthcare professionals, researchers, volunteers and ardent supporters of our interests over the recent decades.

But there are major new challenges ahead. Some were to be expected, while others are somewhat of a surprise. Who would have believed 50 years ago, when the life expectancy for someone with hemophilia was just 20 years, that we would be addressing the issue of aging in hemophilia? Now that prophylaxis is common practice in Canada, it is time to directly examine the "sneaky" effects of prophylactic treatment. And what about inhibitors which bring relentless trouble to some young hemophiliacs? And of course, research, particularly in terms of gene therapy and development of long-lasting products, continues to be an ongoing challenge given that success in these areas will improve the health and quality of life of people with inherited bleeding disorders and lead to a cure.

This issue focuses on the challenges that we face at the dawn of this new decade and I invite you to read the articles on pages 20 to 26. If the past in any way predicts the future, we will undoubtedly face these challenges together and work energetically to overcome them. Of this, I am certain.

So what about the post-it?

The post-it is a reminder!

A reminder about what?

A reminder to stop and think what *Hemophilia Today* means to you!

Are you still curious?

Go to the insert included in this issue!
Message from the President

Pam Wilton, RN

As I write, it is January 2010 and a whole new decade stretches out before us. We need to fill it up with a plan. We need to because we are an organization. We have policies, staff, directors, letterhead, advisors, budgets, committees, files, deadlines, and a web site. We are organized. We even have a clear mission: we know where we are going. It should not be difficult to fill the next ten years with our plans.

So the CHS is making plans. It is true that we do know where we are going; we just need to figure out how best to get there. We need to anticipate the challenges as well as the opportunities and be ready to respond appropriately.

The theme of this edition of Hemophilia Today is the next decade and the great challenges in hemophilia that we will need to work through. Some specific challenges will be addressed by our contributors, but in addition to those challenges there are a few more that I would like readers to consider. I want you to think about what has been happening in our world in the past year because we cannot address our challenges without trying to make sense of what is happening around us.

For example, as I write from my home in south western Ontario I am acutely aware of the high unemployment rate in this region. Thousands of jobs in this part of the country were necessary to support the automotive industry. Plants that made parts or materials are closing. Companies that provide software have had massive layoffs. Transport drivers are idle. The Ford plant will close next year. Those jobs are gone. I don’t need to tell you how devastating those job losses are to families and communities. I do want to remind you though, that in addition to salaries, healthcare benefit packages are gone. We need to address our challenges without trying to make sense of what is happening around us.

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The recent attempted bombing of a flight bound for Detroit has led to increased security at airports, yet again. Stories our guys have shared about the rigorous security screening they have been submitted to in order to travel for work, education, the CHS and vacation with the products and medical equipment needed to manage their chronic condition would make some people think twice about getting on a plane. Most of us are prepared to give up our dignity if it makes us safer, but not our “factor”! It will be essential to provide accurate information to our members, so that those who do travel are well prepared and have a back-up plan if and when their factor replacement products (or medications) and supplies are confiscated. We will all need to rethink how we meet.

David Page called me yesterday, seeking approval to postpone our winter fundraising campaign. Canadians have opened their hearts and given very generously to support relief efforts in Haiti. We know from our experience after the tsunami that this means donations to the CHS will be drastically reduced. It makes good sense to hold off, but the ultimate success of the next campaign worries me. We have to fund all those things in our plans. We count on donations to do our work. We also count on our human resources. We lost James Kreppner, our friend and dedicated volunteer last spring. His knowledge and expertise will be very, very hard to replace. It is already difficult to convince new parents to volunteer with us. Their kids, for the most part, are doing so well that they rarely feel compelled to volunteer to the same extent that parents used to. We need to plan carefully.

I’ve almost ruined a perfectly good Saturday morning, with my gloomy thoughts. I can’t believe you are still reading this. I do actually have a point, so instead of rambling on about the fragile state of healthcare in Canada and barriers to knowledge transfer, I’ll get to it. While it is true that we are all dealing with global challenges as well as personal challenges, there are great opportunities. It is our responsibility as leaders to make certain that we keep in mind what is happening in the world as we plan for specific challenges. At the CHS, we will utilize our experience, knowledge and resources to make good plans as we fulfill our mission, moving ever closer toward our vision.

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The challenges that lie ahead

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HEMOPHILIA TODAY

COMMUNITY NEWS

MARCH 2010 | 5

The CHS honours volunteers

At the CHS Awards Banquet, held in Toronto December 5, 2009, in conjunction with the CHS Semi-Annual Board of Directors Meeting, the CHS recognized dedicated volunteers and healthcare providers who made a significant contribution to the bleeding disorder community in 2008 or preceding years. – C.C.

2008 NATIONAL AWARD RECIPIENTS

Honorary Life Membership Award

2008 Recipient | Tom Alloway, Ph.D.

Tom Alloway has been involved with the CHS in a leadership role for over 15 years. Tom identified an important gap in CHS services and convinced the CHS that it needed to provide information and services to people with von Willebrand disease. He worked hard to help develop programs, such as the Bleeding Disorders Initiative and the 1st Canadian Conference on VWD, and provided strong leadership as a member of the CHS Executive from 1998–2007 and as CHS President from 2001–2004. Tom served as chair of the CHS Program Committee from 1998–2000 and the CHS Resource Development Committee in 2007. He has been involved with the Network of Rare Blood Disorder Organizations (NRBDO) since its inception in 2004, and remains an active member of the CHS Blood Safety Committee and the CHS Scholarship and Bursary Committee.

Although VWD was the issue that compelled Tom to join CHS, he has been a tireless, dedicated, reliable volunteer who has helped the CHS achieve many of its goals.

The CHS National Awards Program will become a biannual program, with awards presented every two years in May at the Rendez-vous joint meeting hosted by the CHS and the four medical groups. It was felt that the Rendez-vous meeting would be a more appropriate occasion to recognize the recipients of CHS National Awards. The next deadline for submitting nominations is January 31, 2011. For further details about the National Awards Program please visit the CHS Web site at www.hemophilia.ca/en/about-the-chs/to-volunteer/volunteer-awards.

International Contribution Award

2008 Recipient | Dr. Brian Luke

Dr. Luke, a pediatric hematologist, was director of the hemophilia clinic in Ottawa from its inception in the 1970’s until 2005. He has been active with the WFH project in China since 2000, when the Ottawa-Guangzhou centre twin (a second Canada-China twin) was established. Dr. Luke immediately subscribed to the idea of having the Calgary-Tianjin and Ottawa-Guangzhou twins collaborate to promote hemophilia care in China. The WFH grant obtained for the Ottawa-Guangzhou twin in 2001 resulted in the initiation of three priority projects (nursing, registry, laboratory diagnosis).

Dr. Luke subsequently visited all the centres to consolidate collaboration and provide educational sessions and advice on the projects. This resulted in a 3rd Canada-China twin (Ottawa/Calgary with Shanghai) in 2002, and the formal establishment of the Hemophilia Treatment Centres Collaborative Network of China (HTCCNC) in 2004. Two other projects were subsequently introduced: physiotherapy (2005) and prophylaxis (2006).

There have now been significant achievements in the five priority projects. The patient group collaborates with the HTCCNC. Together, they have convinced the governments of some of the more economically advanced cities to provide medical insurance to partially cover inpatient/outpatient use of concentrates.
Dr. Luke played an important role in motivating the Beijing Children’s Hospital to develop a Children’s Hemophilia Union of China involving pediatric hematologists around the country. The focus of the WFH hemophilia conference in Beijing in 2007 was children with hemophilia. The Children’s Hemophilia Union held its first forum/conference in July 2008, and Dr. Luke played an important role as advisor and lecturer.

Dr. Luke’s dedication, expertise, advice and hard work have been instrumental in advancing hemophilia care in China.

**Award of Appreciation**

**2008 Recipient | Ann Marie Stain, RN**

Ann Marie is an outstanding nurse. For the past 15 years she has served with distinction at the local, provincial, national and international levels. She has been the nurse coordinator for the Pediatric Comprehensive Care Hemophilia Program at the Hospital for Sick Children in Toronto since 1993. She is a kind, compassionate individual who always puts the well-being of patients and families first, but who also finds the time to contribute to educational initiatives and participate in research in inherited and acquired bleeding disorders in children. Her commitment to individuals with hemophilia extends beyond the adolescent years, and she played a leadership role in setting up a transition program for patients with hemophilia transferred to the adult hemophilia clinic at St. Michael’s Hospital. Another example of Ann Marie’s commitment has been her involvement with Camp Wanakita for the past 14 years.

The teaching Ann Marie provides to children and families with bleeding disorders is an essential component of the Sick Kids vast program. With colleagues in London, Ontario and Montreal (Hôpital Sainte-Justine), Ann Marie was production coordinator of the two original videos about hemophilia in school and home infusion. She is co-author of the chapter “Comprehensive care” in the CHS resource, *All About Hemophilia: a Guide for Families*. At the international level her leadership skills and knowledge of pediatric bleeding disorders are well recognized. Ann Marie is author/co-author of a number of peer-reviewed publications, and has presented research findings at national and international meetings.

**2008 Recipient | Linda Waterhouse, RSW**

Linda Waterhouse has been the social worker at the hemophilia program of the Hamilton Health Sciences Centre for over 15 years. She has been an active member of the hemophilia social work group (CSWHC) and served as co-chair from 2006–2009. She has a deep understanding of the issues faced by her patients and their families and is dedicated to promoting improved comprehensive care through her work as co-chair of the CSWHC group. Linda (along with Ruanna Jones) played a key role in moving the social work group forward as part of the multidisciplinary bleeding disorder team. She is organized and efficient, but always warm and friendly. The experience and insight she brings to CSWHC is deeply valued.

As co-chair of CSWHC, Linda worked in partnership with the CHS to facilitate attendance by as many social workers as possible at national annual meetings and CHS symposiums. She represented CSWHC on the CHS National Program Committee from 2006–2009. She has volunteered on many CHS national projects, including the 2006 Adult Inhibitor Workshop, and was one of the reviewers of *Challenges, Choices, Decisions, an Orthopedic Surgery Decision Making Guide for Adults with an Inhibitor*. She participated on the Advisory Committees for the revised *Hemophilia and School* video and the booklet on *Finding Childcare*, and was also involved in the chapter on comprehensive care for the revised edition of *All About Hemophilia: A Guide for Families*.

**Chapter Recognition Award**

**2008 Recipient | New Brunswick Chapter Development**

Over the past four years, the New Brunswick Chapter Board has made significant efforts to increase membership and expand programs and services for people living with bleeding disorders in New Brunswick. Under the leadership of president Aline Landry, the chapter has organized family educational weekends for the past four years. Various resource people have been invited to present educational sessions. It is hoped that people who attend the educational sessions will realize “first hand” the importance of a strong organization at the local level, and become members. Ongoing efforts are underway to increase membership.
Chapter Leadership Award

2008 Recipient | Jeff Beck

Throughout his many years of involvement at the local, provincial and regional levels, Jeff's goal has been to build and ensure a strong society. Despite health challenges, Jeff has remained determined to bring stability to the Ontario Chapter. He has participated in almost every provincial committee and working group, helped guide the development of the Standards of Care, and piloted the province through uncertain times.

In 2008, during his final year as President, several of his initiatives were adopted. As part of his One Organization vision, new initiatives were implemented, beginning the provincial transformation. The Working Together and Volunteer Summit weekends, developed to move the organization's strategic planning forward, were the perfect way to engage members as part of the transformation process.

Jeff, a quiet leader, is always engaged and active in one aspect of the Society or another, and has made a significant difference in the lives of those living with inherited bleeding disorders in Ontario.

Did you know about...?

The National Endowment Fund Scholarship - University of Ottawa

The objective of this award is to provide financial assistance to students registered in a post-secondary program in a recognized Canadian institution whose lives or families were affected by the tainted blood tragedy between 1980 and 1989 inclusively. Value: variable, depending on tuition fees of the post-secondary institution, to a maximum of $3,000.

The deadline to submit an application is April 30, 2010.

For more information, visit the University of Ottawa at www.admission.uottawa.ca/Default.aspx?tabid=2687 or call 613-562-5734.

NOTICE

The Annual General Meeting of the Canadian Hemophilia Society will convene as follows:
Saturday, May 15, 2010 - 8:30 a.m. at the Delta Montreal Hotel, Montreal, Quebec.

1. To receive the report of the Nominating Committee.
2. To acknowledge the designated directors of each chapter.
3. To nominate candidates for the director-at-large positions on the CHS Board for 2010-2011.
4. To receive the audited financial statements of the Canadian Hemophilia Society for the year ended December 31, 2009.
5. To appoint an auditor for the ensuing year.
6. To approve the recommendation from the Board of Directors to amend Section IV, article 4.03 of by-law no.1 as follows:

   The ex-officio directors for each term shall be the immediate-past president of the Corporation, the medical advisor of the organization and the chair or a co-chair of the National Youth Committee to be selected by the National Youth Committee. Each ex-officio director shall commence his term as director of the Corporation at the end of the Annual General Meeting of members of the Corporation and shall continue until the election by the board of a new president of the Corporation, the nomination of a new medical advisor, or a new chair or a co-chair of the National Youth Committee as applicable. For purposes of clarity, the ex-officio directors shall not be entitled to vote to elect directors-at-large but shall be entitled to vote on all other issues of the board. If qualified, an ex-officio director shall be eligible for re-appointment.

7. To receive the report of the Hemophilia Research Million Dollar Club.
8. To transact such other business as may properly come before this Annual General Meeting of the members of the Canadian Hemophilia Society.

Aline Landry
Secretary
**Chapter Spotlight**

**Hemophilia Saskatchewan**

The Steak Night and Silent Auction held in November was well attended and a great success.

**South Western Ontario Region (SWOR)**

**Holiday Wreaths and Pots**

Thank you to everyone who supported SWOR by purchasing a holiday wreath, pot or door swag. Together we sold 165 wreaths/swags and 72 pots raising approximately $1,400 profit for our region. You have made a difference with your support.

Thank you to Kathleen Hazelwood ( grandma to Jake, Wyatt & Luke) and Sam Davis for making time to bring this all together in one day. You were both invaluable, and we thank you for helping make short work of a long day.

**Youth Speakers Bureau**

In October SWOR held a Youth Speaker’s Bureau to train speakers to speak on behalf of our region and help spread awareness of bleeding disorders. Several of our members participated in the event. Ryan Kleefman, Paul Travaglini, and Marco Valdez-Balderas showed their stuff, and their speaking capabilities are reminiscent of a young Barack Obama, Jerry Seinfeld, or David Suzuki.

Charlie Pangborn and Paul Wilton have been trained in coaching potential speakers and are interested in holding additional training for those who may be interested. Our goal is to have ten speakers in total trained within SWOR by the end of 2010. Please contact Paul Wilton at pwilton3@uwo.ca if you are interested.

**Toronto and Central Ontario Region (TCOR)**

The Toronto Marathon took place on October 18, 2009. The event coursed through 42.2 kilometers of Toronto’s finest scenery. Hemophilia Ontario was entrusted with managing a hydration station located on the Rosedale Valley Road route.

Hemophilia Ontario would like to extend its sincere gratitude and thanks to Pfizer for their ongoing contribution to the Toronto Marathon, including their participation in the Relay race event. We would also like to extend a hearty thank you to Bayer HealthCare, Baxter BioScience and Novo Nordisk for their contribution to this year’s event.

As an important resource development program, Hemophilia Ontario would like to expand participation in the Toronto Marathon for 2010. For more information please contact Stephanie Darroch at 416-972-0641 ext. 20 to learn more about being a runner/walker and/or volunteer.
Central West Ontario Region (CWOR)

A magical holiday event!

Horse-drawn carriages, log cabins, jingle bells, and the smell of sweet maple syrup are only a few of the highlights from CWOR’s 2009 Holiday Event.

This year saw around 40 families gather together to celebrate the season at Mountsberg Conservation Area’s Christmas Town. This year’s activities included a pancake lunch with home-made maple syrup from the sugar bush, the jingle bell craft project, cookie decorating, storytelling, and a surprise visit from Santa and Mrs. Claus.

Families and children also got a chance to play an interactive and surprisingly competitive game of Jeopardy with physiotherapist Cecily Bos and Regional Service Coordinator Alex McGillivray. The categories which got the kids buzzing in to answer included Bleeding Disorder Care, Muscles & Joints, Sports, Famous People, and Service Providers. Everyone had a blast playing this great trivia game.

Many of the families said that the day was magical. Sincere thanks are extended to the Sutton Family who volunteered their time on the day of the event to make sure everything went off without a hitch. Thank you, and we’re looking forward to next year’s event already!

Quebec Chapter (CHSQ)

Dance for Life

The 3rd edition of the fabulous Dance for Life benefit show was held on November 14 at Espace Dell’Arte in Montreal in front of 200 people. In addition to raising $20,000, the event was purely entertaining and completely dazzled the spectators!

Volunteer Recognition Banquet

Forty people gathered to attend the Volunteer Recognition Banquet held on November 28, at the Auberge Handfield in Saint-Marc-sur-Richelieu. This great evening was a perfect opportunity for the CHSQ to sincerely thank its volunteers for the time they give to the organization and to highlight their generosity as human beings. It was also an opportunity to honour Donald Pouliot and present him with a CHSQ Life Member plaque.
Quebec creates no-fault compensation program for products supplied by Héma-Québec

David Page, CHS national executive director

QUEBEC – NOVEMBER 18, 2009

The Quebec National Assembly has adopted draft Bill 24, creating a no-fault compensation plan for those injured by products distributed by Héma-Québec.

As a result, Quebec becomes the first province to accept recommendation #1 of the 1997 Commission of Inquiry on the Blood System in Canada—the Krever Commission—which stated, “It is recommended that, without delay, the provinces and territories devise statutory no-fault schemes for compensating persons who suffer serious, adverse consequences as a result of the administration of blood components or blood products.”

The Canadian Hemophilia Society has been calling on provinces and territories to pass such legislation since the mid 1990s.

In his final report, Justice Krever wrote, “Under both the common law and the Civil Code, the claimant must prove fault before being entitled to compensation from the ‘wrongdoer’. Even if fault actually existed, if no fault can be proved, the claimant must bear the entire burden of the injury, both financial and non-financial. It is the opinion of many legal scholars that this mechanism, the ‘tort system’ or the ‘delict-based system’, is unsatisfactory as a means of compensation for harm. Its disadvantages, including the cost, delay, and adversarial nature of the proceedings, are especially pronounced for a plaintiff who is seriously ill or dying.”

He also wrote, “Shifting the focus from finding fault to compensating the injured party will not compromise the safety of blood... Rather, the safety of the blood supply may best be achieved through strict regulation.”

Quebec’s Bill 24 calls on... “The Minister to compensate, regardless of any fault, all victims of bodily injury caused by a defect or contamination by a pathogen, known or emerging, in a product distributed by Héma-Québec.”

The compensation plan is based on the Quebec Automobile Insurance scheme, also no-fault. It includes provisions for income replacement, a care allowance, lump-sum indemnities, reimbursement of medical costs and re-training. Access to compensation is intended to be universal, quick and easy.

The no-fault scheme also constitutes a benefit for Héma-Québec. Since its creation in 1998, the blood service has spent millions of dollars to insure itself against potential claims, though not one claim was actually made.

Canada’s other nine provinces and three territories have given no indication they are following Quebec’s lead. Consequently, Canadian Blood Services (CBS) has had to take out insurance against injuries due to its products. This is costing CBS, and indirectly taxpayers, many millions of dollars annually. Nor, in the absence of legislation, is it clear how quickly and universally compensation would be paid out in the event of a catastrophe.

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Hemophilia 2010
Saturday, July 10 - Wednesday, July 14
Buenos Aires
Argentina

organized by: Hemophilia 2010 Congress
hosted by: Argentina
visit: hemophilia2010.org
Progress in comprehensive care for rare blood disorders conference

Silvia Marchesin, past president, Aplastic Anemia and Myelodysplasia Association of Canada

In 2004, a group of organizations representing patients with rare blood disorders came together as the Network of Rare Blood Disorder Organizations (NRBDO) under the mentorship of the Canadian Hemophilia Society (CHS). Early on it became clear that the topic of comprehensive care was a key one for all of the groups. Although the hemophilia patients had numerous clinics across the country, virtually none of the other patient groups had access to such resources.

In 2006, we held our first conference, with national and international presenters describing comprehensive care models that worked well. At this conference we developed a list of the principles which comprise comprehensive care. The organizations were then to work in their respective communities in order to further the cause of comprehensive care.

We felt it would be good to have another conference to see what progress has been made in this area since that first gathering. So, on November 13-15, 2009 in Toronto the Progress in Comprehensive Care for Rare Blood Disorders Conference — Presented by CSL Behring was held. The focus was on patient registries and comprehensive care for rare blood disorders. It was also an opportunity to reinforce and build partnerships. The delegates included leaders from patient organizations, healthcare providers, and members of industry.

Friday was dedicated to the topic of patient registries, a cornerstone of comprehensive care. Registries contribute to the understanding of these rare disorders, help quantify the number of people affected, measure treatment outcomes and facilitate research. Seven excellent examples of registries were presented. These included discussion on how data is collected, how patients are enrolled, outcomes that have been made possible by the registries, privacy, security, governance and funding. There were also presentations on the many challenges in implementing national registries.

Saturday was an opportunity to share experiences and best practices in comprehensive care. We heard from many speakers who described how their clinics were attempting to become more “comprehensive” or expanding to include other patient populations. It was very encouraging to hear about the successes across the country. Indeed, progress is being made, and we recognized the many achievements, as well as the areas that still need improvement. We were also fortunate to have a session on advocacy led by a local patient advocate.

Sunday was a time for the delegates to discuss what we can do within our respective roles: for example, how patient groups, healthcare providers and industry can work together.

We also discussed the barriers and challenges to moving toward comprehensive care. From this list of challenges, we then identified four priority areas to be addressed in the short and long term. Finally, we proposed concrete tasks and strategies that could be used to create an action plan to accomplish these goals.

The NRBDO as a whole will now have to prioritize this list and decide how to move forward. The network and the individual associations will advocate and work to push initiatives forward together and in our separate communities.

The NRBDO is a coalition of the following patient groups:

- Aplastic Anemia and Myelodysplasia Association of Canada
- Canadian Association for Porphyria
- Canadian Hemophilia Society
- Canadian Hereditary Angioedema Network
- Canadian Immunodeficiencies Patient Organization
- Canadian Neuropathy Association
- Canadian Organization for Rare Disorders
- Canadian Sickle Cell Society, Sickle Cell Association of Ontario, Sickle Cell Disease Patient Support Group of Ottawa, Quebec Sickle Cell Anemia Association
- Thalassemia Foundation of Canada

The conference was an occasion to welcome a new group into the NRBDO: the Canadian Association of Paroxysmal Nocturnal Hemoglobinuria (PNH).

For more information on the NRBDO, please visit the CHS Web site www.hemophilia.ca and click on the NRBDO icon on the left hand side of the page (under Our Partners). For more information on the conference, select 2009 Progress in Comprehensive Care for Rare Blood Disorders Conference — Presented by CSL Behring. There you will find almost all of the presentations posted as well as the conference proceedings.

Thanks to the sponsors who made this event possible: Alexion Pharma Canada, Canadian Blood Services, Celgene Corporation, CSL Behring, Héma-Québec, Novartis Canada, and Shire Canada. Also, thank you to the CHS for their continued support of this network, and to David Page, its first coordinator, and Michel Long, its current one.
As in past years, parents had the opportunity for their child to have a consultation with experts on inhibitors, including Dr. Georges-Etienne Rivard, Dr. Manuel Carcao, Dr. Vicky Breakey and Claude Meilleur, RN.

On Saturday morning the children enjoyed a trip to the Biodome...while their parents attended sessions from the experts about treatment options and the management of inhibitors.

On Saturday afternoon the children worked on an art project facilitated by Suzanne Douesnard, psychologist, and Marianne DuFour, art therapist, about living with inhibitors. The children then presented their feelings to the parents.
A Saturday evening dinner and dance was enjoyed by all...

Dr. Norman Buckley and Angela Forsyth, PT, spoke about medical options and gadgets and gizmos for managing pain.

During the closing session a panel consisting of parents and adolescents shared their experiences, and the children presented skits about living with hemophilia and an inhibitor.

Thanks to the CHS, the sponsors, the planning committee and the families for making the 4th CHS Family Inhibitor weekend such a success!
As a follow-up to ceremonies and tree plantings held at the Canadian Blood Services headquarters in Ottawa in the fall of 2007 to mark the 10th anniversary of the publication of the Krever report and the one held in 2008 at Héma-Québec in the presence of the Quebec Minister of Health and Social Services, Dr. Yves Bolduc, numerous CHS chapters planned and held commemorative events in 2009 in order to continue to honour those who died in the tainted blood tragedy and those who are still living with its consequences. The events also served to remind Canadians of the importance of keeping our blood supply system safe, and about the watchdog role that our organization continues to play in the hope that no such tragedy happens again.

We are pleased to share with you a glimpse of some of the events which occurred in 2009. Thanks to all the volunteers who made these happen! Planning for events in 2010 has already begun and we encourage our members to contact their respective chapters to find out how they can help and contribute to the growing forest of commemorative trees. – M.L.

Wascana Park, Regina, Saskatchewan
October 27, 2009

Tuesday, October 27, 2009 Hemophilia Saskatchewan commemorated the tainted blood tragedy by dedicating an evergreen tree on the grounds of the Legislative Building. Seven MLA’s, representatives from Hemophilia Saskatchewan, Canadian Blood Services, Wascana Centre Authority, and the media gathered near the tree where Faye Katzman conducted Saskatchewan’s first Tree of Life ceremony.

After a moment of silence, Elaine Zech, vice president at Hemophilia Saskatchewan, presented the Canadian Blood Services with a commemorative plaque. Everyone then gathered at the Canadian Blood Services Clinic for a tour of the facilities and lunch before members of HSK returned to the Legislative Building, where they were introduced to the Legislature.

The commemoration event, a Hemophilia Saskatchewan initiative, was supported by Canadian Blood Services, Wascana Centre Authority, and the Canadian Hemophilia Society.
Prince Edward Island Legislative Assembly grounds Charlottetown, Prince Edward Island
October 27, 2009

On October 27, 2009 the Prince Edward Island Chapter of the Canadian Hemophilia Society (CHS) and Canadian Blood Services held a special tree-planting ceremony at 11 a.m. near the Coles Building on the grounds of the PEI Legislative Assembly as a reminder of PEI’s commitment to ensuring the safety of Canada’s blood supply. In attendance were representatives from Canadian Blood Services and several federal, provincial and municipal politicians and community leaders, including members of the CHS PEI Chapter.

St-Paul’s Hospital Roof Garden, Vancouver, British Columbia – October 27, 2009

On October 27, the British Columbia Chapter of the CHS, in collaboration with the local CBS office, held a tree planting ceremony on the roof garden of St-Paul’s Hospital. Dr. Michael O’Shaughnessy, director of the BC Centre for Excellence in HIV/AIDS, who has established an international reputation for his AIDS research, his care of people living with HIV, and his advocacy of human rights, was also among those who planted a tree of life. Another tree is scheduled to be planted in front of the hospital later this spring.

2009 Commemorative ceremonies in Ontario
A number of events were held in Ontario, including:

South Western Ontario Region (SWOR) October 27, 2009

A tree was planted on the grounds of the University of Western Ontario and a ceremony was held. Thank you to Marion, Jeff and Maureen who shared their poignant stories and Councilor Paul Hubert for speaking on behalf of the City of London. Thank you to the friends and families who attended, honouring their loved ones. And a special thank you to the University of Western Ontario for allowing us the opportunity to share their grounds for the planting of our first tree.

North Eastern Ontario Region (NEOR) – October 27, 2009

An existing tree in Kawartha Park, which is already dedicated to AIDS as a memorial, was given to TCOR. A plaque will be placed on World Hemophilia Day, April 17, 2010.

Ottawa and Eastern Ontario Region (OEOR) – October 27, 2009

A beautiful Sugar Maple Tree was planted at The Niagara Lions Club Community Park in Niagara Falls to commemorate those who suffered from the tainted blood tragedy. The tree was planted along the memorial walkway in the park. We encourage those who live in the area to visit.

Central West Ontario Region (CWOR) – October 27, 2009

A number of events were held in Ontario, including:

South Western Ontario Region (SWOR) October 27, 2009

A tree was planted on the grounds of the University of Western Ontario and a ceremony was held. Thank you to Marion, Jeff and Maureen who shared their poignant stories and Councilor Paul Hubert for speaking on behalf of the City of London. Thank you to the friends and families who attended, honouring their loved ones. And a special thank you to the University of Western Ontario for allowing us the opportunity to share their grounds for the planting of our first tree.

North Western Ontario Region (NWOR) October 27, 2009

A tree was planted in a provincial park.

North Eastern Ontario Region (NEOR) – October 27, 2009

An existing tree in Kawartha Park, which is already dedicated to AIDS as a memorial, was given to TCOR. A plaque will be placed on World Hemophilia Day, April 17, 2010.

Ottawa and Eastern Ontario Region (OEOR) – October 27, 2009

A beautiful Sugar Maple Tree was planted at The Niagara Lions Club Community Park in Niagara Falls to commemorate those who suffered from the tainted blood tragedy. The tree was planted along the memorial walkway in the park. We encourage those who live in the area to visit.
Upcoming Events

Canadian Hemophilia Society
- April 30 – May 2, 2010 – PEP Workshop | Saskatchewan and Manitoba in Winnipeg, Manitoba.
- May 15, 2010 – CHS Annual General Meeting in Montreal, Quebec.
- May 19, 2010 – World Hepatitis Day | This is hepatitis...

Hemophilia Saskatchewan
- March 27, 2010 – Hemophilia Saskatchewan AGM Linger Longer Day - AGM, Lunch and Educational Sessions. From 11:00 a.m. at Mayfair United Church, 902 33rd Street West, Saskatoon. Childcare will be provided.
- June 2010 – Annual BBQ.

South Western Ontario Region (SWOR)
- March 27, 2010 – SWOR Annual General Meeting and Educational Conference, Hilton Hotel, London, Ontario. This full-day interactive conference will provide pertinent information for you about your bleeding disorder.
- May 20, 2010 – 3rd Annual Golf Tournament for Hemophilia and Inherited Bleeding Disorders, Highland Golf and Country Club, London, Ontario. Funds raised through this event provide training, education, financial assistance and moral support to more than 300 children, youth, and adults with an inherited bleeding disorder in south western Ontario.
- August 24 to 29, 2010 – Pinecrest Adventures Camp, Camp Menesetung Goderich, Ontario. This five-day residential camping experience for children with a bleeding disorder and their siblings aged 5-16 years provides respite to parents while providing the campers an opportunity to experience camp as all children do.

Central West Ontario Region (CWOR)
- May 1, 2010 – Mothers Day. An educational and bonding experience for our female membership.
- June 5, 2010 – Biking to Stop the Bleeding. Raising awareness and funds for Hemophilia Ontario (provincial).
- June 6, 2010 – Fathers Day. An educational and bonding experience for our male membership.
- June 26, 2010 – Summer BBQ. Education and networking for new and old families.
- July 1, 2010 – Lions Club Canada Day Carnival. Raising awareness and funds in support of CWOR.
For more information regarding any of these events, please contact Alexandra at 905-522-2545 or by e-mail at aplumb@hemophilia.on.ca.

Quebec Chapter (CHSQ)
- April 24, 2010 – Bowl-a-thon in Montreal.
- May 2010 – Survey to members on the CHSQ Web site. Prizes to win in return for your opinion.
- May 1, 2010 – Bowl-a-thons in Victoriaville and Quebec City.
- June 6, 2010 – The CHSQ youth group will attend the Montreal Grand Prix.

World Hemophilia Day activities

Canadian Hemophilia Society

Toronto and Central Ontario Region (TCOR)
- April 17, 2010 – On this day YOU CAN HELP us by selling tulips to your neighbours, colleagues, family and friends. It is a simple three-step procedure, and your help will make a huge difference.
 Also, TCOR announces a creative competition: Express your thoughts in a creative way!! Think outside the box. Create an art piece which is primarily visual in nature, such as ceramics, drawing, painting, sculpture, architecture, and printmaking.
 Topic: Your life. Age groups are: 6 to 11; 12 to 16; 17 to 25.
 Submit your art work by April 1, 2010.
 For all the details on either activity, please contact Manisha at 416-972-0641 ext. 23 or 1-888-838-8846 or by e-mail: mramrakhani@hemophilia.on.ca.

South Western Ontario Region (SWOR)
- April 17, 2010 – Polar Bear Dip. This SWOR event will celebrate World Hemophilia Day by educating the community about living with an inherited bleeding disorder, as well as raising funds for regional programs.

Central West Ontario Region (CWOR)
- April 17, 2010 – Raising awareness and funds for Hemophilia Ontario (provincial) and CWOR.

Quebec Chapter (CHSQ)
- April 15 and 16, 2010 – Awareness stands will be held at the Quebec National Assembly, Sainte-Justine Hospital and L’Enfant-Jésus Hospital.

Newfoundland and Labrador Chapter
- April 17, 2010 – Awareness events will be held across the province. Volunteers are needed.

Nova Scotia Chapter

Prince Edward Island Chapter
- April 2010 – Run For It, Red White & YOU at local schools.

Newfoundland and Labrador Chapter
- June 2010 – 7th Annual Community Walk-A-Thon. Proceeds support research. Date to be confirmed.
- July 8–11, 2010 – Annual Family Weekend- Lions Max Simms Camp, Bishop Falls, Newfoundland and Labrador.

Nova Scotia Chapter
- May or June 2010 – Staff training weekend for summer camp. Many other meetings will be held over the next few months in order to get camp details ironed out for the 2010 camp year.
- June or July 2010 – Nova Scotia Chapter Annual General Meeting.

Prince Edward Island Chapter
- May 2010 – Prince Edward Island Annual General Meeting. Date to be confirmed.
Volunteer File

"Volunteers do not necessarily have the time; they just have the heart."
–Elizabeth Andrew

Marion Stolte
Chair, National Volunteer Development Committee

The Volunteer Binder

Time constraints – we all face them in our lives, whether at work, at home, or in our volunteer life. Those constraints can be viewed negatively or as boundaries that actually can give greater focus to our work. Sometimes having a deadline can be just the impetus needed to write that article or to sharpen our minds to do a presentation! What time constraints are you facing? How can we assist each other as volunteers in the roles we have taken on?

The Volunteer Binder, which has been distributed to each chapter, has a variety of tools available for you to use. These range from job description templates, to project event worksheets which will assist you in planning your events, to volunteer recognition ideas. By utilizing these resources you’ll be able to more easily manage the deadlines and time constraints you face. Often we’re not quite sure where to turn for suggestions or advice on some hurdle we are facing or endeavouring to overcome or some initiative we want to implement in our chapters. The National Volunteer Development Committee (NVDC), which has members throughout the country, is available as a resource. Feel free to call or e-mail any one of us. In fact, to simplify contact, here are the names and e-mail addresses of the current members of the NVDC:

Colleen Barrett
snowbirds@nf.sympatico.ca

Sarah Bradshaw
sarahbradshaw_@hotmail.com

Mylene D’Fana
mylenedfana@videotron.ca

David Pouliot
david.pouliot@gmail.com

Jamie Pytel
jpytel@telusplanet.net

Ryanne Radford
ryanneradford@hotmail.com

Suzanne Shaw-O’Leary
shawoleary@bellaliant.net

Marion Stolte
mstolte1972@yahoo.ca

Elaine Zech
dandezech@sasktel.net

As you think about various events, fundraising, awareness and volunteer recognition activities your chapter will be holding over the next few months, be sure to utilize the resources in the Binder. Be prepared to recruit new volunteers to assist in organizing your events. Be ready with time frames and responsibilities for jobs needing to be done, which will assist you in coping with time constraints and allow you to plan more effectively. Seek out someone who has been a competent volunteer and ask questions, learn from him or her. There are many people in our chapters who can be resources and mentors.

We are most effective as we work together, as we learn from each other. Over these next few months, consider your own volunteer role and ask how you can do your job better. Ask how you can volunteer in such a way that the upcoming events are better than ever. Remember, as volunteers we do have the heart – we do care deeply – so let’s keep on making a difference!
Great charitable organizations are rare. Few have strong track records of improving lives, like the Canadian Hemophilia Society (CHS). But how do we do it? One of the vital ingredients, in our case, is philanthropic support from our committed corporate partners... our BeneFACTORS.

The Greek word, *philanthropos,* combines two words: *philos,* or "loving" in the sense of benefitting, caring for, nourishing; and *anthropos*—"humankind", "humanity", or "human-ness". This is the aim of philanthropy... improvement to the quality of human life.

For the bleeding disorder community and the CHS, the magic of philanthropy is how we are able to provide programs and services that would not otherwise be possible without gifts from our transformational donors – Baxter, Bayer, CSL Behring, Novo Nordisk, Octapharma and Pfizer.

As we all know, our industry partners are engaged in the discovery, creation, development and manufacture of medications that are designed to improve the health and quality of life of patients. But their commitment to the CHS and its members does not stop there... it extends to:

- **Contributing to the improvement of care** for all people suffering from an inherited bleeding disorder.
- **Increasing knowledge** about state of the art developments in the care of people with inherited bleeding disorders.
- **Empowering people** with bleeding disorders, at all stages of their lives, to maximize their quality of life.
- **Increasing knowledge and competence** of patients and families of people with rare blood disorders.
- **Providing fellows** in hematology or relevant fields the opportunity to acquire clinical or research skills.
- **Providing parents with crucial information** and support during the different stages of their child’s development.
- **Funding research** to improve the quality of life of our members and to find a cure for bleeding disorders.

The spirit of philanthropy is deeply imbedded in the culture of all of our industry partners. What we have seen over the last decade, over and over again, has been a solid desire to help us address needs that may seem so large or intractable that no one else will even try, or sometimes, so small that no one else will even bother. But we rise to these challenges because we know we can – with the strength and philanthropic commitment of our partners behind us.

We are grateful for our BeneFACTORS Club partners’ passion and commitment to the CHS. Together we are accomplishing things that we never could alone – we are building a stronger, healthier bleeding disorder community where we can live, play, work and dream of a better life.

Philanthropy is not reserved for the super-elite or corporations of the world. Philanthropy is for everyone... please give generously to the CHS. 

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**The magic of philanthropy**

Deborah Franz Currie and Pauline Major, CHS resource development
Fundraising

Pauline Major
CHS National Corporate Philanthropy Manager

2010 Dare to Dream for a Cure presented by Pfizer

Let's face it; most of us will probably never get the chance to drive a Formula One race car. Only the privileged few get to sit behind the wheel of a Formula One car. BUT on September 17, 2010 you can experience the thrill of driving a race car at the Canadian Hemophilia Society's Dare to Dream for a Cure national fundraising event!

The Dare to Dream driving experience is as close as it comes to a Formula One, and the renowned Bridgestone Racing Academy's circuit in Mosport, Ontario, is the ideal place to test your racing skills. Shift your driving skills up a gear and experience the speed and handling of a single seater Van Diemen race car! Be prepared to handle the power of a car capable of 0-60mph in less than 6 seconds!

This could be you on September 17, 2010!

Dare to Dream for a Cure is an exciting and innovative national fundraising event that captures the spirit and excitement of racing while raising awareness and funds for research. In 2010 we are continuing our partnership with our chapters as we work together to make this event the most successful to date!

On September 17, 2010, experience the

Thrill of a Lifetime

▪ Travel from your location to the Bridgestone Racing Academy, Mosport, Ontario;
▪ Hotel accommodation for one night at the Marriott Residence Inn;
▪ In-class session of driving instructions; track and car orientation;
▪ Fifteen-minute warm-up; 2 twenty-minute lapping sessions on the track;
▪ Photograph and certificate of course completion.

Don't miss your chance!

Contact your local chapter for more information and to purchase raffle tickets!

Tickets are only $20 or 6 for $100
THE NEXT DECADE:
Great challenges in hemophilia

There are numerous challenges to be overcome in the coming ten years. The next pages offer a brief portrait of some of them. Identifying these challenges is an initial step towards overcoming them. Here we explore possible solutions towards finding the answers to meet the challenges ahead. – C.R.

CHALLENGE: Inhibitors

David Page, CHS national executive director

Since the advent of an adequate supply of safe clotting factor concentrates in the 1990s, it has generally been accepted that the major challenge in the treatment of hemophilia is inhibitors.

The body’s immune system perceives an infusion of factor VIII or IX protein as a foreign substance. It mounts a defense and develops antibodies to quickly destroy the clotting protein. This is called an inhibitor.

Out of 100 patients with severe factor VIII deficiency, approximately 30 will develop an inhibitor. In 10 of these 30 patients, the inhibitor will soon disappear on its own. In 10 other patients, the inhibitor will disappear after large, frequent doses of FVIII. This is called immune tolerance therapy. In 10 others, the inhibitor will persist; treatment becomes more complicated and less effective.

Inhibitors are approximately 10 times less frequent in hemophilia B (factor IX deficiency) but can nevertheless occur and, when they do, be even more serious.

“I believe that the best hope to improve on the issue of inhibitors,” says Dr. Georges-Étienne Rivard, director of the Quebec Centre for Inhibitors of Coagulation, “is to prevent their formation. We need to better understand what contributes to their development. There are a number of possible causes: the individual’s factor VIII or IX mutation, the concentrate infused, the time of initiation of factor exposure, the intensity of first treatments, and infection or inflammation associated with treatment or vaccination.”

“The best way to improve on the issue of inhibitors,” says Dr. Georges-Étienne Rivard, director of the Quebec Centre for Inhibitors of Coagulation, “is to prevent their formation.”

Dr. Manuel Carcao, co-director of the hemophilia program at The Hospital for Sick Children said, “There is so much still to learn! We need to know how best to eradicate inhibitors. What is the best immune tolerance regimen?”

According to Dr. Carcao, there are some promising areas of research that may change the portrait in the coming decade. These include large cohort studies to determine the risk factors for inhibitor development (the CANAL and RODIN studies), and the SIPPET study comparing the risk of inhibitor development between plasma-derived and recombinant factors when administered to previously untreated patients.

To address the issue of early introduction of prophylaxis leading to less inhibitor formation, Dr. Carcao suggests undertaking a “randomized study comparing an approach of very early introduction (by six months of age) of low dose, infrequent prophylaxis (once per week or even once every two weeks) vs. standard care.”
The good news is that this is no different for the bleeding disorder population, and we are now observing the aging of people with hemophilia. Advances in hemophilia care beginning in the 1950’s have contributed to an overall improved life expectancy and quality of life. The improved life expectancy brings new challenges to bleeding disorder healthcare providers. It will be important to recognize the new reality of improved life expectancy early on during the management of hemophilia. Perhaps prophylaxis treatment will take on a new meaning if a person realizes that he will need healthy joints to carry him into his eighties. There may be no guidelines for the provision of care for health issues not previously recognized in this population. We will need to collaborate in order to build data to ensure that the care offered to our patients is evidence-based. It is important that we continue to be strong advocates for our patients to ensure that they receive high quality care in the health and community sectors. We will need to establish new partnerships and educate specialists in fields such as gerontology, oncology, cardiology and home care nursing who may be unfamiliar with bleeding disorders. As one patient states: “Because I am a seventy-two (72) year old woman with a rare factor V deficiency, I carefully assess, with my healthcare team, the risks and benefits of treatment options for recently diagnosed cardiovascular disease. My medical management has been adjusted due to increased bleeding side effects of some prescribed drugs.” We need to meet this exciting challenge together and promote healthy aging of the bleeding disorder population. Aging is a process that starts at birth!

CHALLENGE: The “sneaky” effects of prophylaxis

François Laroche, CHSQ president and Hemophilia Today editor-in-chief

At this point, most people aged 25 and under with hemophilia have been treated from a very early age with long-term primary prophylaxis (preventative factor replacement therapy), considered the gold standard of care for hemophilia since the late 1980s. The majority of these individuals generally experience only minor bleeding and have few or no major bleeding episodes into their joints, muscles or soft tissues. As a result, many feel a sense of invulnerability to their disorder, and sometimes they can be too quick to return to their activities (such as walking, running or sports) or make inappropriate activity or even career choices that can have serious short-, medium- and long-term repercussions for their musculoskeletal health. According to Dr. Christine Demers, director of the hemophilia centre at l'Hôpital de l'Enfant-Jésus in Quebec City, “We can’t blame young people for age-appropriate behaviour or for behaviour in keeping with that of their peers. The education that the team at the hemophilia treatment centre (HTC) offers to parents of a newly diagnosed child with hemophilia should be followed up throughout the child’s youth and adolescence, to make sure that physical activities and career choices are suitable. In
According to Dr. Christine Demers, director of the hemophilia centre at l'Hôpital de l'Enfant-Jésus in Quebec City, “We can’t blame young people for age-appropriate behaviour or for behaviour in keeping with that of their peers.”

practice, this is not always easily achieved, as a result of the lack of resources in the centres (nurses, physiotherapists and social workers), as well as by the distance, in some cases.”

For Nichan Zourikian, a physiotherapist at Sainte-Justine University Hospital Centre (CHU Sainte-Justine) in Montreal, the introduction of home treatment and prophylaxis is a double-edged sword. “People with hemophilia have gained a lot of autonomy and their health status has been substantially improved, however, now some young people with hemophilia only come to the HTC to pick up their coagulation products. They neglect the importance of being monitored by the HTC’s multidisciplinary team, particularly when they regard the injuries as being minor. For example, minor, persistent, but often painless synovitis (joint inflammation), resulting from a minor or moderate hemarthrosis (joint bleed), can lead, in a hidden or sneaky fashion, to premature joint damage.”

The crux of the problem is education. Dr. Demers adds, “It is important to understand that even with prophylaxis two or three days a week, there will be variations in the level of the replacement factor circulating in the blood, which is connected in particular to its half-life. Take for

example factor VIII, which has a half-life of 12 hours in the case of a young person with hemophilia A given 30 units of factor VIII per kilo. Immediately following the infusion, his factor level increases approximately 60%, but declines rapidly to 30% about 12 hours later, then to 15% about 24 hours after infusion, and 7.5% about 36 hours after infusion. After 48 hours, with a factor level of 3.75%, bleeding can occur if major exertion is put on the joint.”

Nichan Zourikian emphasizes the preventative approach. “In full action, the infused replacement factor circulating in blood gets used to stop minor bleeding resulting from tiny injuries and trauma, which is inevitable with any intense or repetitive activity. This leaves much less replacement factor available should there be a major trauma. It is therefore essential to select appropriate sports and physical activities and wear proper protective equipment (helmets, braces, knee pads, tape, etc.). It is also important to adapt one’s physical activities according to the physiotherapist’s recommendations to protect joints that are at risk, or to simply abstain from playing certain sports or engaging in certain activities if the risk outweighs the benefits.”

Another consideration is that during intense physical activity, the body has the capacity to double or even triple the level of natural factor circulating in the bloodstream—it isn’t unusual for a person’s factor level to temporarily rise by 300%. However, it is not possible to double or triple factor level in a person with hemophilia through infusion of replacement factor.

“When a professional or sport activity forces overuse of the same muscles or joints, it generally results in bleeding, sometimes significant but often minor or moderate, which causes medium- or long-term damage because a person with hemophilia is not protected in the way people normally are. When you factor in the fact that some adolescents and young adults reduce their prophylaxis for a variety of reasons, some justified, it is not surprising to still see damaged joints at the onset of adulthood,” says Dr. Demers.

Nichan Zourikian adds, “The equation also needs to include the ability to heal, which is different from one person to the next. We see a greater capacity for tissue regeneration and healing in some people than in others. We take a case-by-case approach. For this reason, among others, choices concerning sports and activities need to be discussed with the multidisciplinary team, which can provide guidelines that are well adapted to the individual’s specific circumstances. We recommend that children, parents and the HTC team begin this discussion fairly early, rather than waiting for the arrival of adolescence.”
The next decade: great challenges in hemophilia

One of the five components of the Canadian Hemophilia Society’s Passport to Well-Being program is entitled Charting Your Course. It aims to help participants understand how we can improve the treatment of people with bleeding disorders through innovative monitoring of product usage. Charting Your Course provides a wonderful overview of how “information gathered at home, patient charts at hemophilia treatment centres, collective Canadian data on care and treatment, can contribute to the well-being of individuals and of the entire bleeding disorder community.” (see www.hemophilia.ca/files/Charting_ang.pdf)

A key component of the system for collecting data is the Canadian Hemophilia Assessment and Resource Management Information System (CHARMS). It was developed and implemented across Canada in 1998 as the result of an agreement between the Canadian Blood Agency, the Association of Hemophilia Clinic Directors of Canada, and McMaster University. Twelve years later this Microsoft Access® based program is in need of an overhaul, and the opportunity this process presents makes it timely to review the potential and pitfalls of record-keeping in the electronic age from our perspective as people with bleeding disorders.

As a good friend of mine used to say, “If it weren’t for patients there would be no need for healthcare.” Medical records have to be understood from a patient’s perspective. For the person with a bleeding disorder, improved quality of life is always the end goal. Tracking your blood product use at home and providing the information you collect to your hemophilia treatment centre (HTC) creates a medical record which is tracked by the HTC in the CHARMS database at the clinic along with other information relevant to their care for your bleeding disorder. They also use CHARMS to record what product has been delivered to you at home. Recording product use has two important functions. First and foremost it is about tracking bleeding episodes and the amount of product used to treat those episodes. By reviewing trends with HTC staff, care can be managed to optimize product use and minimize damage resulting from bleeding. The tainted blood tragedy of the eighties also awakened us to the importance of good record management for tracking products in the system when safety becomes a concern.

However, the present structure of CHARMS is not optimal. The information patients collect, whether using old paper “bleed sheets” or newer electronic systems based on the Web or handheld devises, has to be manually inputted into CHARMS, as does information about product distribution. Also, any upgrades to the CHARMS software have to be installed independently at each HTC. All of this work is unnecessarily time- and human resource-intensive given 21st century technology.

So why don’t we have a seamless system where patients can input their records in an electronic format of choice, transmit in real time to the HTC for review and feedback, order product, and receive appointment reminders and urgent notices with one set of key strokes? Privacy has been a concern since the introduction of electronic record-keeping, acting as a weight on the shoulders of progress. Centralizing records always raises eyebrows as patients can Google® “misplaced patient records” and read themselves to sleep. However, thanks to strong privacy legislation in place across the country and highly sophisticated web-based database software that allows for detailed access logging and data encryption, privacy concerns can be resolved.

The other critical issue has been resources. The prospect of making a significant change means new programming and diligence to ensure compliance with various legislation and healthcare facility protocols, both of which require funding and human resources. When the need for a patient-friendly electronic input option became apparent to replace written bleed sheets, the original CHARMS partnership did not provide a solution, so private industry stepped in. Two downsides became apparent. A multitude of options may be a good thing in terms of product choice, but in record-keeping it has been problematic in the absence of unified standards for data collection. We have also heard concerns about data privacy and questions about motivation when medical records management services are supplied in conjunction with the supply of blood products themselves. To what degree these concerns are real or imagined remains to be seen, but it is clear that a revamp of CHARMS is an opportunity to address the need for proper data management standards, tighter integration and increased patient involvement in planning from the ground up.
The next decade: great challenges in hemophilia

In June 2007, the Canadian Hemophilia Society and the four professional organizations representing physicians, nurses, physiotherapists and social workers adopted the *First Edition of the Canadian Comprehensive Care Standards for Hemophilia and Other Inherited Bleeding Disorders*.

The purpose of national standards is to encourage hemophilia treatment centres (HTCs) to adhere to uniform practices that are desirable, accountable, transparent and organized.

Since their adoption, these standards have informed the work of HTCs and hospital administrations whose mandate is to ensure adequate care for patients. In addition, in 2009 HTCs performed a voluntary self-assessment of their ability to meet the standards.

The standards were developed with a view to an eventual external audit of HTCs as is currently the practice in HTCs in the United Kingdom and Ireland, where the external audit provides a valuable and objective evaluation of the quality of services. In the event they fall short of the standards, the audit provides clear recommendations for improvement and a powerful incentive to clinic and hospital administrators, and Ministries of Health, to take corrective measures including additional human resources.

Dr. Irwin Walker, co-chair of the Canadian Hemophilia Standards Group and Director of the Hamilton Niagara Regional Hemophilia Centre, said, “I see tremendous value in standards. Combined with Standard Operating Procedures (SOPs), standards take away uncertainty, permit quality assessments and audits, promote training and ensure consistency of care across the country. Essentially, standards help a clinic to say what it does, do what it says, and prove it.”

Dr. Walker speaks from experience, having also participated in the development of standards and SOPs for bone marrow transplantation. The Hamilton Bone Marrow Transplant Program at McMaster University, which he directs, is one of four accredited programs in Canada.

Following the 2009 recession, governments across the country face significant budget deficits. Health care represents close to 50 percent of provincial expenditures so clearly it will be a target for cuts. Relatively small cuts in already under-staffed HTCs could have a huge impact on their ability to provide optimal care. Already, some HTCs do not have dedicated resources in physiotherapy and social work. Others have such small percentages of full-time positions that devoting time to monitoring, prevention and rehabilitation is difficult, if not impossible.

More than 95% of the cost of care for bleeding disorders goes to paying for expensive clotting factor concentrates. It would seem unwise not to invest in the human resources—physicians, nurses, physiotherapists, social workers and other healthcare providers—who can ensure that these therapies are used optimally and that all the ancillary services are available to complement the provision of clotting factor therapy.

In a context where an ever-increasing majority of bleeding disorder care is provided in the home by the patient or caregiver, far out of the sight of hospital administrations, a critical challenge in the coming decade will be to ensure that the highly trained healthcare team needed to support home infusion in a comprehensive care model is supported and enhanced.

Challenges:

**Standards of care and support for bleeding disorder clinics**

David Page, CHS national executive director

**Longer half-life products and gene therapy**

David Page, CHS national executive director

"The major change in hemophilia therapy in the past decade, " according to Dr. David Lillicrap, researcher at Queen’s University and director of the hemophilia program at South Eastern Ontario Regional Inherited Bleeding Disorders Clinic in Kingston, Ontario, “has been the growing realization that we can prevent chronic musculoskeletal damage through the use of prophylactic concentrate infusions. This paradigm change in treatment strategies has prompted the current search for ways in which clotting factor levels can be sustained at therapeutically relevant concentrations without the need for frequent intravenous infusions. This goal can be attained through two approaches: by engineering clotting factor proteins that have a prolonged circulating half-life or through the delivery of a clotting..."
factor gene that expresses sufficient protein to achieve a therapeutic effect for a prolonged period of time.

Half-life is the amount of time it takes for half of the active clotting activity to disappear from the bloodstream. The half-life of factor VIII in people with hemophilia A is, on average, 12 hours. The half-life of factor IX in people with hemophilia B is closer to 24 hours.

At the start of this decade, there is a great deal of research activity related to the development of longer half-life clotting factor proteins. Pharmaceutical companies in North America and Europe have either begun or are close to beginning clinical trials in longer-acting factor VIII in hemophilia A, factor IX in hemophilia B and factor VIIa for the treatment of inhibitors. If successful, these new therapies could decrease the frequency of prophylactic infusions in hemophilia A, for example, from three times per week to once a week.

“Currently," estimates Dr. Lillicrap, "the engineered protein strategies appear to be winning this race, with the arrival already in the clinic of a couple of candidate proteins in which the half-life has been extended through distinct engineering approaches. These early phase clinical studies should provide key data to allow the evaluation of these approaches within the next couple of years."

While significant success in gene therapy and hemophilia has been limited to mice and dogs, human clinical trials continue. Gene therapy has made dramatic breakthroughs in treating other chronic diseases, notably severe combined immunodeficiency (boy in the bubble syndrome), though not without important complications.

"While we have wonderful therapies," said Mark Skinner, president of the WFH, "we don't have a cure, and the therapies we have still require infusions two or three times a week and can be costly.

while much progress is occurring through the development programs of the WFH, global challenges remain huge. In his plenary address to the last WFH World Congress in Istanbul in 2008, Mark Skinner identified five research challenges to attaining the WFH goal of Treatment for All.

▪ Inhibitor formation – identifying risk factors for the development of inhibitors, including the possible immunogenicity of different products, as well as optimal treatment strategies to overcome them.
▪ Evidence-based standards – validating and/or establishing optimal clinical management and treatment standards.
▪ Rare factor deficiencies – development of treatment products and protocols for rare disorders.
▪ More effective treatment – development of treatment products with a longer half-life.
▪ A cure – achieving a cure for bleeding disorders, including gene therapy.

“Answering these questions,” Mark Skinner said, “necessitates global collaboration due to the limited number of patients eligible for research protocols combined with the outcome data required. Working together is the key to finding the answers.”
For this issue our co-chairs took a well-deserved break from the YOUTH FILE column, giving me the opportunity as staff coordinator of the National Youth Committee to talk to you about the future challenges for the Canadian Hemophilia Society.

This issue of Hemophilia Today focuses on the important challenges ahead for the hemophilia community in the next decade. Reflecting on the growing needs of our members, several questions come to mind: How will the organization prepare to face the challenges? What resources will be available in order to adequately respond to the needs? Will our youth be ready to take charge of the challenges from our aging members? How can we get more young people involved now and prepare them for leadership succession?

I asked about 40 youths actively volunteering with the organization at different levels what motivated their involvement. Many said that the Canadian Hemophilia Society is an important resource to them. In fact, many feel that the CHS is a vital source of information that allows them to take better charge of their hemophilia. Others are driven by the motivation to preserve the gains achieved through the tireless work of previous generations of CHS volunteers, and ensure that future generations will also have access to a range of safe and affordable therapies.

A Native youth member said that through his involvement he has been able to meet other people with bleeding disorders beyond his family circle. Volunteering with his provincial chapter has also been an exceptional opportunity for him to develop leadership skills.

Allow me to share other thoughts provided by two youths. First, a young hemophiliac living in Toronto gives us an urban perspective:

“I chose to get involved with the hemophilia society for both selfish and selfless reasons. The selfish reasons were largely a realization that there were objectives I wanted to see out of the society, and I realized quickly that it was not going to meet those objectives without my help. My voice had to be heard, and that was only going to happen if I started to speak up. As Ghandi put it best: ‘Be the change you want to see in the world.’ The selfless reasons were that I was speaking loudly not just for myself, but on behalf of so many others I felt were being left on the sidelines. I felt that the crowd I represented (the ‘non-camp’ crowd) was almost being marginalized by the society, through no one’s fault in particular; it was simply that we did not have a loud enough voice. I needed to bring a voice of diversity to the society so that everybody else who felt left out of the crowd, so to speak, had someone speaking on their behalf.”

And a British Columbia student with von Willebrand disease explains why she feels it’s important to volunteer:

“I would love to get more involved with the organization. Growing up, I didn’t know anyone with von Willebrand, besides people in my own family. As I’ve become more involved in the past couple months, attending conferences and camps, I’ve come to realize the great impact we as youth can have as role models and leaders. Deeper involvement for me would mean helping to give young people what I didn’t get to experience growing up— an environment in which young people of the same age can share stories and experiences with others who understand what it is to live with a bleeding disorder.”

Now I ask you, dear readers of the YOUTH FILE column:

Does the CHS meet your current needs? Are there specific issues that affect young people that are not being addressed in CHS programs? How do you envision the organization’s future? What are your main concerns? Research? Long-acting factor products? Gene therapy? Latent effects of prophylaxis?

We need to hear from you. The Canadian Hemophilia Society is your organization, it exists for you. We need to know about your concerns to be able to respond. We need your involvement to carry on the excellent work achieved by your parents, grandparents and, in some cases, great-grandparents through the CHS over the past 50 years. Please contact me at hbourgaize@hemophilia.ca to share your comments or to learn more about the different ways you can get involved with your organization.
The Canadian Association of Nurses in Hemophilia Care (CANHC) recognizes the need to ensure a high standard of nursing practice, education and research. In order to achieve quality nursing care for people affected by hemophilia and other bleeding disorders, we strive to enhance professionalism through partnerships, collegiality and mentorship.

Nurses have played an integral role in caring for patients with inherited bleeding disorders at Canadian hemophilia treatment centres (HTCs) for over 30 years.

The first Canadian HTC was established in 1955 in Montreal, Quebec. In 1978, the Canadian Hemophilia Society held a National Hemophilia Symposium. Nurses from eight HTCs met informally in what was the first meeting of Canadian hemophilia nurses.

The nurses shared ideas to support one another and develop educational resources for patients, families and members of the comprehensive care team.

HTC’s were being established throughout Canada, and today there are 26 centres across the country serving people with bleeding disorders.

In the mid 1980’s, hemophilia nurses were in the midst of the “Tainted Blood” tragedy. By 1987, hemophilia nurses from across Canada were meeting annually as “The Canadian Hemophilia Nurses.”

In 1997 we sought membership within the Canadian Nurses Association (CAN) and became the “Canadian Association of Nurses in Hemophilia Care” (CANHC), an associate member of CNA.

There are 37 hemophilia nurses in Canada.

CANHC has a constitution that is reviewed and amended by the membership at the annual meeting of the association.

CANHC developed a generic job description and established a mentorship program to help nurses new to hemophilia care.

Job description

The role of the hemophilia nurse coordinator is dynamic, comprehensive and expansive. The hemophilia nurse coordinator is the link between the patient and members of the comprehensive care team, the community and secondary service members. He/she plays a key role in patient care management, and provides care and education to the hemophilia population and those who care for them. Thus, the nurse plays a critical role in assisting patients’ personal health management and in improving/maintaining their quality of life.

There are five key areas of practice that assist the hemophilia nurse coordinator in the role: clinical practice, communication, outreach services, administration and research.

CANHC mentorship program

A mentor is simply someone who helps someone else learn something that he or she would have learned less well, more slowly, or not at all if left alone. Mentors are learning coaches – sensitive, trusted advisors.

Goals and objectives

- Accelerate the time required for the new CANHC member to acclimate to the position of hemophilia nurse coordinator.
- Increase the applicant's level of competency to fulfil the unique and complex role of hemophilia nurse coordinator.
- Enhance the likelihood of retaining hemophilia nurse coordinators new to the role.
- Improve the quality of care provided to patients with bleeding disorders.
- Facilitate networking within the profession and community at large.

CANHC continues to work toward fulfilling our mission and supports the philosophical statement developed by the Canadian Nurses Association.

The HTC nurse coordinators look forward to taking up the challenges of the next decade and to successfully meeting those challenges. This can only be done with input from patients and all members of the HTC team.

N.B. A copy of the CANHC constitution, job description and mentorship program can be found on the Canadian Hemophilia Society Web site www.hemophilia.ca. Care and Treatment: Nursing or click on the CANHC logo. ☞
Hepatitis & HIV Press Review

Michel Long  
CHS National Program Coordinator  

and Dr. Elena Vlassikhina  
Volunteer collaborator

- HIV vaccine shows promise for first time  
  An experimental vaccine has prevented HIV infections for the first time, a breakthrough that has eluded scientists for a quarter century. A U.S.-funded study involving more than 16,000 volunteers in Thailand found that a combination of ALVAC and AIDSVAX cut infections by 31.2 percent in the people who received it compared with those on a placebo. The latest result will transform future research.  

- More new studies confirm that daily coffee consumption is linked to less severe liver fibrosis  
  Daily caffeine consumption of about 2 cups of coffee was associated with reduced liver fibrosis, according to results of a study published in the January 2010 issue of *Hepatology*. The protective effect persisted after controlling for age, sex, race, weight, liver disease, and alcohol intake, though it was more pronounced in people with hepatitis C virus (HCV) infection. The researchers found, however, that consumption of decaffeinated coffee—or of caffeine from sources other than coffee—was not associated with reduced fibrosis.  

- Quebec docs tout custom AIDS vaccine  
  Doctors in Montreal have made an important breakthrough in AIDS research by developing a vaccine that could eventually replace the cocktail of drugs currently used to fight the HIV virus. The new vaccine is different from past efforts in that it is based on each person’s individual condition, and the approach is unique in the world. There was an 80% drop in the virus in each patient, and patients were then able to stop taking antiretrovirals.  

- Once-daily Schering hepatitis C drug shows promise  
  An experimental new treatment for hepatitis C being developed showed impressive effectiveness and was well tolerated in an ongoing mid-stage study. The pill, narlaprevir, is the first once-a-day member of an emerging new class of treatments for hepatitis C called protease inhibitors. It has potential to eventually become the preferred drug because of its once-daily dosing, a major advance in terms of convenience, which will help patients comply with their treatment regimens.  

- More than 80% of HCV Genotype 1 treatment-naive patients achieved sustained virologic response with twice-daily Telaprevir-based regimen.  
  Telaprevir could allow shortened treatment duration and increased cure rates in people with HCV [compared to standard of care], offering a new approach to the treatment of HCV.  

- First hint of a hepatitis C vaccine?  
  A novel vaccine, IC41, has produced a modest, but apparently continuous, viral load decline in patients who already have hepatitis C. Although this is a therapeutic rather than a preventative vaccine, this is the first time that any vaccine designed to enhance the body’s natural immune response to HCV has had a significant effect on viral load. This therapeutic vaccine might serve as a way of enhancing other therapies, and demonstrates that it is possible to engineer an effective immune response against HCV, a virus whose hyper-variability is such that some researchers thought a vaccine could not work.  

- New drug technology produces marked improvement in hepatitis C therapy in animals; may be useful for wide range of diseases  
  In a dramatic finding, SPC3649, a new drug for hepatitis C virus infections that targets liver cells, produced a substantial drop in blood levels of the virus in animals, and continued to work up to several months after treatment. This proof of concept study suggests that the technology might also prove useful in treating many other diseases, such as HIV, cancer, and inflammatory diseases. The new therapy could potentially replace interferon in future cocktails, since it provides a high barrier to resistance. It may also be a good therapy to use after transplant, since it may help suppress HCV in the new liver. It has no toxic or adverse reactions and this is critical in the transplant setting.  
Hepatitis C patients can have good outcomes after transplants using HBV+/HCV+ liver grafts, but donor status may affect HCV recurrence

Due to a severe shortage of donor livers, researchers have investigated transplants using suboptimal or marginal—known as "extended criteria donor"—organs, including giving patients who already have hepatitis B or C livers that are infected with these viruses. Hepatitis C patients who receive donor liver grafts infected with both hepatitis B virus (HBV) and hepatitis C virus (HCV) had similar survival rates to patients who received uninfected livers, although they were slightly more likely to require a second transplant. Another transplant study found that patients who received livers from donors after cardiac death experienced more rapid and severe HCV recurrence than those who received grafts from brain-dead donors, though survival rates were similar.

www.hivandhepatitis.com/2009icr/aasld/docs/112009_b.html

Liver issues...

A large number of individuals in our community were infected with HIV and/or hepatitis C because of tainted blood. Many have passed away, while many others continue to live with the viruses and their consequences, including liver disease. Increasingly, people are thinking about or are being considered for liver transplantation. At present, access to liver transplants in Canada for people infected with HIV is limited to non-existent. The CHS HIV–Hepatitis National Committee has been working to address this issue. Among other things, the CHS has been collaborating and advocating with the Canadian Treatment Action Council (CTAC) to make solid organ transplants available for HIV-positive people in Canada. To further this work, the CHS HIV–Hepatitis National Committee is trying to assess the number of infected individuals from our community considering a liver transplantation, either here in Canada or abroad. The CHS also wants to remind all members that the organization can provide support and information to those considering a transplant. If this is your case, or if you would simply like to help advance our work to obtain better access to liver transplants, please feel free to contact Michel Long in all confidentiality at mlong@hemophilia.ca or 1-800-668-2686. – M.L.

Criminalization of HIV non-disclosure

The criminalization of HIV non-disclosure is one of the most pressing issues facing people living with HIV. To help address this issue, people living with HIV, community activists, AIDS service organization staff, lawyers and others have formed the Working Group on Criminal Law and HIV Exposure (CLHE).

Given that hemophiliacs have recently been charged and/or prosecuted, the CHS has been following developments around this issue very closely.

The CHS supports the CLHE’s position that HIV/AIDS is an individual and public health issue first and foremost, and should be addressed as such. The CHS supports CLHE’s position that the increasing criminalization of HIV non-disclosure has happened without sufficient debate and critique. To ensure that the criminal law will not be used inappropriately and/or in a discriminatory manner in cases involving allegations of HIV non-disclosure, the development of guidelines for police and criminal prosecutors is being examined. Police and prosecutors need guidelines to ensure that decisions to investigate and prosecute such cases are informed by a complete and accurate understanding of current medical and scientific research about HIV, and take into account the social contexts of living with HIV.

The CHS is very interested in hearing your views on this matter, answering questions and providing support to those who may be facing charges.

Please feel free to contact Michel Long in confidentiality at mlong@hemophilia.ca or 1-800-668-2686. – M.L.
May 19 is World Hepatitis Day!

For the third consecutive year, the CHS and its chapters will join with groups around the world to raise public awareness of the life-threatening liver diseases hepatitis B and C. For more information and to find out how you can take part in this effort, go to: www.hemophilia.ca/en/hcv-hiv/world-hepatitis-day.

This is **hepatitis**...

- **Get Protected** – Knowing the risk factors for hepatitis B and C is the most important step in preventing new infections.
- **Get Tested** – Liver disease is long and complicated. Getting tested is quick and simple.
- **Get Vaccinated** – In the case of hepatitis B there is a vaccine and the disease is preventable.
- **Get Treated** – In many cases, treatment for hepatitis B and C is effective.

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**HIV-positive individuals can now legally visit and migrate to the U.S.**

WASHINGTON – JANUARY 4, 2010

U.S. immigration regulations that severely restricted travel to that country by HIV-positive individuals ended today. Previously, those wishing to travel to the U.S. were required to reveal their status to U.S. officials and apply for a special waiver before leaving their own country. Such requests were often denied. The ban was introduced during the late 1980s.

President Barack Obama announced the end of the ban in October 2009. The 60-day waiting period before the change came into effect ended today.

With confirmation of the end to the ban, the International AIDS Society confirmed that the 2012 International AIDS Conference will be held in Washington, DC.

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**REMINDER | IMPORTANT NOTICE**

**JUNE 30, 2010 is the FIRST CLAIM DEADLINE** for ALIVE primarily-infected persons for the following compensation programs:

- Pre-1986/Post-1990 Hepatitis C Settlement Agreement
  www.pre86post90settlement.ca

- Hepatitis C (HCV) January 1, 1986-July 1, 1990 Class Actions Settlement
  www.hepc8690.ca
From January 28 to February 3, 2010, Mylene D’Fana and I, as delegates of the Canadian Hemophilia Society – Quebec Chapter, visited our partners at l’Association tunisienne des hémophiles (ATH) to participate in a number of medical and organizational events with our Tunisian “twins.”

Friday, January 29 was set aside for a roundtable with representatives of the CHSQ and the ATH, as well as representatives from the national hemophilia organizations of Morocco and Algeria. The agenda focused on the experiences of each organization with regards to patient registries and fundraising activities. It was very interesting to learn a little more about the experiences of hemophilia organizations from the Maghreb in these two areas, the strengths and weaknesses, and areas needing improvement. Participants agreed on the importance for hemophilia organizations to have patient registries, independent of those existing in hospitals, with restricted but useful information on members in order to better represent their needs. This information is also valuable for program planning and development.

In terms of fundraising, each organization has had good results, with good ideas for special events including some with pharmaceutical sponsorship. However, more emphasis could be placed on developing financial autonomy (through benefit events, sale of promotional items such as reusable bags with the organization’s logo, etc.), and obtaining government support through advocacy and lobbying.

The afternoon was devoted to the Third National Hemophilia Day, featuring a medical symposium focused on prophylaxis, organized by the ATH under the auspices of the Tunisian Hematology Society and in collaboration with Bayer. Following presentations by Drs Emna Gouider (Tunisia), Georges-Étienne Rivard (Canada), Phu Quoc Lê (Belgium) and Assad Haffar of the World Federation of Hemophilia, there appeared to be consensus regarding the application of prophylaxis in Tunisia according to the following principles:

Prophylaxis must be started after the first joint bleed and thereafter given on a treatment schedule suitable for the child, using the recommended dose based on the number of units per vial (between 20 and 35 IU/kg), which is once weekly, with the possibility of twice a week for recurrent bleeds.

The option of daily prophylaxis for adults at a dose of 10 IU/kg was put forward. In terms of type of product to use, the suggestion was to begin therapy with plasma-derived product because it contains von Willebrand factor, and furthermore because some studies have demonstrated that plasma-derived products have a two to three times less likelihood of inhibitor development. However, after 50 days of exposure to plasma-derived factor, the risk of developing inhibitors is very low, at which point switching to recombinant product is recommended because it offers the best safety from pathogens, particularly emerging ones.

The next two days were devoted to training sessions for the ATH Board of Directors. On Saturday, Mylene D’Fana gave a presentation on recruiting volunteers and keeping them involved. There were good discussions on how to adapt strategies for the Tunisian context. This was followed by a working session on program planning and development. The ATH already has a solid foundation in this regard so we focused on refining certain aspects, in particular establishing deadlines and individual responsibilities to facilitate organizational logistics. We also stressed the importance of succession planning so as not to wear out key volunteers and thereby ensure a dynamic organization.

Sunday was reserved for a presentation and workshops on governance. Starting with the ATH’s mission and values, my presentation focused on the key conditions of good governance. I stressed the importance for the ATH to establish clear general regulations and internal policies (on pharmaceutical relations, conflicts of interest, member support, etc.) in order to provide a framework for the work to be done by members of the organization and their respective roles and responsibilities. Other aspects addressed included the creation, role and mandate of working committees, financial statements, codes of procedure and rules of order to ensure the proper conduct and functioning of the organization. Once again, ATH members actively participated and the discussions were very productive. Some points were simply a matter of review, while others were signposts for the future work.

In closing, on behalf of the CHSQ, Mylene D’Fana and myself, I would like to thank the members of the ATH Board of Directors for the wonderful welcome, their attentive participation, committed involvement, and above all for the outstanding personal qualities that they emanated throughout our visit.

Chokran (Thank you) Emna Gouider, Taoufik Raissi, Kaouther Zahra, Amel Derwaz, Rania Kammoun and Amdouni Hamma. Beslama (Goodbye and see you soon).
Kogenate Liplong Study stopped
Frankfurt – January 25, 2010 – Bayer announced that it has stopped an international phase II trial (Kogenate Liplong Study) to compare an experimental long-acting recombinant factor VIII formulation, BAY 79-4980, with a current product, Kogenate® FS. The company stated that early results indicated that the therapy has proven less effective than hoped. An independent Data and Safety Monitoring Board did not raise any safety concerns. Several Canadian centres were enrolling patients in the study.

Biogen Idec and Biovitrum move forward with trial for long-acting hemophilia B therapy
Cambridge, Mass. and Stockholm, Sweden – January 25, 2010 – Biogen Idec and Biovitrum AB announced that the first hemophilia B patient was dosed in the companies’ registration, open-label, multi-centre trial into a long-acting, recombinant factor IX Fc fusion protein (rFIXFc). The decision to advance the program is based on promising data from a Phase I/IIa open-label, multi-centre, safety dose-escalation and pharmacokinetic study of intravenous rFIXFc in severe, previously treated hemophilia B patients. In the study, rFIXFc was well tolerated and demonstrated a prolonged half-life compared to existing therapies.

The global trial is being designed to assess the safety, pharmacokinetics and efficacy of rFIXFc in hemophilia B patients. rFIXFc has received orphan drug designation for the treatment of hemophilia B from both the European (EMEA) and US (FDA) authorities.

Inspiration Biopharmaceuticals and Ipsen partner on range of hemophilia products
Paris (France), and Laguna Niguel (California, USA) – January 21, 2010 – Inspiration Biopharmaceuticals and Ipsen announced that they are partnering in the development of a range of recombinant proteins for the treatment of hemophilia. The most advanced products are Inspiration’s IB1001, a recombinant factor IX for the treatment of hemophilia B, and Ipsen’s OBI-1, a recombinant porcine factor VIII for the treatment of patients with an inhibitor to factor VIII. Both IB1001 and OBI-1 are expected to enter phase III clinical trials in 2010.

In addition to OBI-1 and IB1001, Inspiration is developing a portfolio of products that include recombinant factor VIII for hemophilia A and recombinant factor VIIa for those with hemophilia A or B and inhibitors.

Biovitrum takes novel factor VIII long-acting hemophilia A therapy into clinical trials
Stockholm, Sweden – December 15, 2009 — Biovitrum AB announced that the first patient was dosed in a phase I/llla study of its long-acting, fully recombinant factor VIII Fc fusion (rFVIIIFc) protein. The phase I/llla open-label study will assess the safety, tolerability and pharmacokinetics of rFVIIIFc in severe, previously treated, hemophilia A patients. The rFVIIIFc program and international study are partnered with Biogen Idec.

FDA approves Wilate® for VWD treatment in U.S.
Lachen, Switzerland – December 4, 2009 – Octapharma AG announced that it has received approval for Wilate® from the U.S. Food and Drug Administration (FDA). The designation was granted for the use of Wilate for the treatment of bleeding episodes in patients with severe von Willebrand disease (VWD) as well as in patients with mild or moderate VWD in whom the use of desmopressin is known or suspected to be ineffective or contraindicated.

Wilate is a newly developed, high-purity, double virus inactivated (solvent/detergent plus special terminal dry-heating) von Willebrand Factor/Coagulation Factor VIII Concentrate, derived from human plasma, that has shown efficacy for all types of VWD including pediatric patients in four prospective clinical trials.

Wilate is approved for use by Health Canada for the treatment of factor VIII deficiency.

Advate® now distributed in 3000 IU dose size
Toronto – December 3, 2009 – Baxter Corporation has announced that Advate® [Antihemophilic Factor (Recombinant), Plasma/Albumin Free Method (rAHF-PFM)] will be distributed in Canada in the 3000 IU size starting in January 2010. Advate is therefore available in six dosage strengths: 250, 500, 1000, 1500, 2000 and 3000 IUs.

Researchers discover that “Royal Disease” is hemophilia B
Moscow – November 6, 2009 – Researchers from Moscow State University and University of Massachusetts Medical School have identified the cause of the “Royal Disease” transmitted from Queen Victoria (1819-1901) to the British, Russian, German and Spanish royal families.

Analysis of recently discovered skeletal bone specimens from members of the Romanov family, murdered at the time of the Russian Revolution, showed that the Tsarevich Alexei suffered from hemophilia B and both his mother, Alexandria, and sister, Anastasia, were carriers.

continued ➤
Case of Freeman vs. CBS & Attorney-General of Canada ends – Decision expected in summer

David Page
CHA National Executive Director

The proceedings in the civil case of Freeman vs. Canadian Blood Services (CBS) and the Attorney-General have now ended in Ontario Superior Court after 35 days of expert and factual testimony, lasting from September to December. During the week of January 4-8, final legal arguments were presented by Canadian Blood Services, the Government of Canada, counsel for Mr. Kyle Freeman, the Canadian Hemophilia Society, Egale, a national organization committed to advancing equality and justice for lesbian, gay, bisexual and trans-identified people, and the Canadian AIDS Society. The judge in the case, the Honourable Madam Justice C. Aitken, has indicated a decision can be expected this summer.

The case originated in 2002. Kyle Freeman had donated blood on 18 occasions between 1990 and 2002; however, after the last occasion in June 2002, he informed Canadian Blood Services in an anonymous e-mail that he had lied when responding to the questionnaire, specifically with regard to Question #18, Male donors: Have you had sex with a man, even one time, since 1977? CBS was able to discover his identity and sued Freeman in civil court for negligent misrepresentation and damages. In subsequent discovery proceedings it was established that Freeman had lied to four questions of the blood donor questionnaire, including one related to his sexual history of having had sex with other men (MSM). According to Health Canada regulations, men who have had sex with other men, even once, since 1977 are permanently deferred from giving blood. Question #18 was introduced by the Canadian Red Cross in the 1980s to protect the recipients of blood and blood products against infection with HIV. Had Freeman replied honestly to these questions, he would have been ineligible to give blood. Mr. Freeman countersued CBS and Health Canada on grounds that the question violated his right not to be discriminated against based on sexual orientation, a right guaranteed in Section 15 of the Canadian Charter of Rights and Freedoms. He asked the court to find that "he need not answer truthfully" and that the court should order a change to the MSM question.

Three key questions the judge must answer are:

Is CBS, as a private not-for-profit corporation at arms-length from government, subject to the Charter of Rights and Freedoms?

Does the question concerning men who have had sex with men violate charter provisions on equality (Section 15 of the Charter)?

If the question does violate Charter rights, can this discrimination be justified for reasons of public health (Section 1 of the Charter)?

The evidence is unequivocal that recipients of blood and blood products are particularly vulnerable and use blood products out of necessity. It is respectively submitted that when the rights in this case are balanced, the rights of recipients must prevail.

The Canadian Hemophilia Society argued that the deferral is justified in the interest of safeguarding the blood system. Current epidemiology shows that sexually transmitted diseases are many times more prevalent in the population of men who have had sex with men, compared to those who haven’t. The CHS also contended that the MSM deferral protects the blood system against new and emerging pathogens that would be sexually transmitted.

The CHS’ final written submission concludes as follows: "The MSM deferral serves to protect recipients from blood-borne pathogens that threaten to kill them or seriously impact their quality of life. The evidence is unequivocal that recipients of blood and blood products are particularly vulnerable and use blood products out of necessity. It is respectively submitted that when the rights in this case are balanced, the rights of recipients must prevail. Ultimately, it must be remembered that the recipients bear 100% of the risk of blood-borne pathogens and the donors (whether MSM or otherwise) bear none... The CHS submits that if this court should find that the CBS is bound by the Charter and that the current MSM deferral is a violation of Mr. Freeman’s section 15 rights, that such violation is justified under section 1 of the Charter."
Our Stories

Living with factor XIII deficiency

Lyman Keeping, Garnish, Newfoundland and Labrador

I have a very rare bleeding disorder called factor XIII deficiency. Writing about my disorder brings back many memories of living in an isolated outport with very few medical facilities and coping with a disorder that caused me much pain and my parents extreme periods of grief.

I was born and spent the first 15 years of my life in the small outport community of Point Rosie, which is located on the Burin Peninsula in Newfoundland. When I was very young, our only means of travel was either by open boat to Garnish, or by coastal boat (steamer) to Grand Bank. Not a great place to be living with a bleeding disorder.

My life with factor XIII deficiency started the first few days following birth. I started bleeding through my belly button almost immediately following the cutting of the umbilical cord. Although at first it was not a cause for alarm, as the days passed, my parents started to worry because the bleeding would not stop. A scab would appear, but after a short while, the bleeding would start again. Eventually the bleeding stopped.

My first memories of this disorder were when I was about six years old. While playing with some friends, I fell and landed on the side of my face. Two of my teeth were busted up pretty bad. I was taken to the Grand Bank Cottage Hospital (a two-hour ride by coastal boat) where the two broken teeth were extracted. Then my troubles began. I started to bleed and the bleeding continued for the next several days. Doctors were both worried and mystified.

It was not long before my parents began to suspect that all was not right with me. My legs and arms were constantly full of bruises and I began having painful episodes with my
hips and knees. For weeks I would be hobbling around, barely able to straighten my leg at all. Very often I would be confined to the house for two or three weeks.

When I was about eight or nine years old, I had an experience that almost cost me my life. Being very active, I was constantly roughhousing with my friends. On one occasion, I cut my hand on a broken bottle. It was a very deep gash in the crevice between the thumb and forefinger. That night, Mom noticed that my arm was starting to swell, so she removed the band-aid. The blood squirted out of the cut with such force that it almost hit the ceiling.

All that night the bleeding continued and I became very weak. Mom and Dad realized that this was an emergency. To make a long story short, by the time I arrived in Grand Bank, I was unconscious and an emergency blood transfusion was necessary to save my life. I was in a virtual coma for three or four days.

As a side note here, I owe my life to the late Captain Brown of the coastal boat, Bar Haven. He kept a constant vigil over me during the whole trip, and made every effort to keep me awake. Fortunately, his blood type was the same as mine, and it was his blood donation that helped save my life.

After recovery I was sent home. My bleeds, however, as well as my trips to the hospital, were becoming more frequent. Finally, the doctors in Grand Bank decided they could do no more for me and sent me to St. John’s.

At the Grace Hospital in St. John’s I was put in the care of a Dr. Neary, who was a very well-known surgeon. I was given every test imaginable from spinal taps to every type of x-ray known to man. Finally, Dr. Neary informed my mom that he was still not certain what was causing my many bleeding episodes. There was no evidence of classic hemophilia (factor VIII) and my blood did not appear to lack any of the factors known to medical authorities at that time.

Dr. Neary then decided to perform surgery to take out my spleen and appendix. I have no idea why these two organs were removed because it was never fully explained to my mother. Needless to say, I had a bleeding episode following the procedure. After a number of transfusions and an extended stay in hospital, I finally recovered and was sent home.

For the next two or three years, there were no major incidents, however, I was still bruising easily, cuts on certain parts of my body took a long time to scab over and heal.

Another significant incident occurred when I was about 14 years old. One day I stumbled on the steps of the school and landed on my stomach. By the time I got home, I had a terrible pain in my abdomen and it was beginning to swell to the point where I could barely do up my pants. As the night went on, the pain worsened and I also began to run a fever. I was rushed to Grand Bank and the next day, the doctors decided to send me to St. John’s. It was one of the most painful rides I have ever experienced. The ride from Grand Bank to Goobies was all on gravel road and here I am in terrible pain sitting up in taxi for several hours.

I was admitted to the new Janeway Hospital and the next day Dr. Neary performed emergency surgery. It was discovered that I had a massive bleed in my tummy. A draining tube was inserted into the affected area. Again, I was sent home and no one appeared any the wiser as a result of my ordeal.

The next year, 1969, my parents resettled to Garnish. One night in early fall, an excruciating pain developed suddenly in my right hip. There appeared to be no apparent reason for this pain as I had not had any type of blow in that area. As the night wore on, the pain became so intense I had to bite into the pillow to keep from screaming.

I was rushed to Grand Bank Hospital that continued on page 36
night, and as per the routine, sent to St. John's the next day. By this time my mother was beside herself with frustration and I was becoming increasingly more depressed.

After being admitted to Janeway, I was put in the care of Dr. Wally Ingram, a leading hematologist. My mother demanded that I not be sent home until the medical specialists had determined what was causing this most unusual and painful condition.

Over the next several weeks, I was carted around to every hospital in St. John's, subjected to a number of different tests.

Then, as if out of the blue, Dr. Ingram informed me that I had a very rare bleeding disorder called factor XIII deficiency. I was now 15 years old and was told in no uncertain terms that I was lucky to be alive.

To replace the missing factor, I was to receive two units of fresh frozen plasma once every four to five weeks or as needed. Needless to say, I was ecstatic to finally not have to endure extreme pain and long stays in the hospital.

Since 1969, I have only experienced one major bleed. It occurred in 1974 while I was teaching in an isolated outport and was not getting my infusions of plasma on a regular basis. Since that time, however, I have had no major bleeds and my life has been pain free. Plasma has since given way to Fibrogammin as the means to keep my bleeding under control.

I run four to five times a week and have completed five 26.2 mile marathons with no adverse effects. Over the years, I have become very health conscious and make every attempt to control my weight and stay in shape.

The only bleep has been a battle with hepatitis C. However, that has also been eradicated from my system — another journey, to be shared at some other time.

I am now 55 years old and living life to the fullest.

The one person I feel I owe my life to is my mother. Her indomitable spirit and stubborn will are two of the reasons I am healthy today. She would not give up on me and bugged the medical people until they finally decided to dig deeper into what was causing me to bleed so much and causing me to have such a miserable life. During one twelve-month period, while we were still living in Point Rosie, she and I made TWELVE trips to St. John's. Each journey involved a two-hour ride in an open boat and a taxi ride of anywhere between 7 to 10 hours. I will always respect and love her for that.

The other important person to whom I owe a debt of gratitude is my wife, Yvonne. Even though my disorder was already under control by the time I met her, she has been with me through some very tough times. Several years ago, when I was undergoing an aggressive treatment therapy for hepatitis C, she accompanied me to all my doctor's appointments and looked after me during the days when I was too sick to help myself. It's important to have a strong person by your side in times of need.

I hope this narrative will help others persist when faced with a long-term illness. Never give up! Keep the faith!

Running up Signal Hill, St. John's, NL, at the end of the Cape to Cabot 20 km road race.

My wife, Yvonne and I, at home in Garnish.

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