

Hemophilia TODAY

Canadian Hemophilia Society
Serving the Bleeding Disorders Community



SPRING 2007 www.hemophilia.ca VOL 42 NO 1

YOUTH FILE OPENED page 13



OUR STORIES page 14



RESEARCH SUMMIT page 5



TWINNING WITH TUNISIA page 23



Spring 2007 • VOL 42 • NO 1

Hemophilia Today
625 President Kennedy Avenue, Suite 505
Montreal, Quebec H3A 1K2
www.hemophilia.ca
Phone: 514 848-0503
Fax: 514 848-9661
Toll-free: 1 800 668-2686
chs@hemophilia.ca

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

The Canadian Hemophilia Society strives to improve the health and quality of life for all people with inherited bleeding disorders and to find a cure. Its vision is a world free from the pain and suffering of inherited bleeding disorders.

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

The CHS consults medical professionals before distributing any medical information. However, the CHS does not practice medicine and in no circumstances recommends particular treatments for specific individuals. In all cases, it is recommended that individuals consult a physician before pursuing any course of treatment.

Brand names of treatment products are provided for information only. They are not an endorsement of a particular product or company by the writers or editors.

EDITOR
François Laroche

PRESIDENT
Eric Stolte

EDITORIAL COMMITTEE
Hélène Bourgaize
Clare Cecchini
François Laroche
David Page
Chantal Raymond
Patricia Stewart

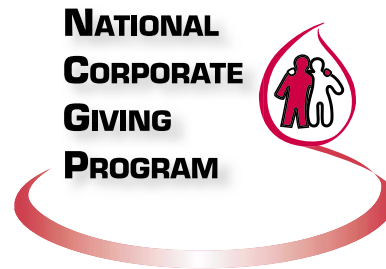
CONTRIBUTING WRITERS
Sarah Bradshaw
Clare Cecchini
Dianna Cunning
François Laroche
David Page
Andrea Pritchard, RN, PhD(c)
Jeff Rice
Nisa Renault, BSc, PhD(c)
Eric Stolte
Emil Wijnker

PRODUCTION COORDINATOR
David Page

PRODUCTION ASSISTANT AND
FRENCH VERSION COORDINATOR
Chantal Raymond

GRAPHIC DESIGN
Paul Rosenbaum

TRANSLATORS
Roy Keys
Line Ladouceur
Marie Préfontaine



We would like to thank the following companies, corporate foundations and employee fund programs for their generous support. Our way of recognizing them for their generosity is through our National Corporate Giving Program.

PLATINUM

BAYER

GOLD

BAXTER

CSL BEHRING
NOVO NORDISK
WYETH

SILVER

DEWDNEY FAMILY CHARITABLE FOUNDATION

BRONZE

DELTA HOTEL WINNIPEG
DELUCA'S SPECIALTY FOODS
EDELSTEIN DIVERSIFIED CO. LTD.
FONDATION DE BIENFAISANCE T.A. GERMAIN (LA)
JOHN BROUWER FOUNDATION (THE)
JOHN DEERE FOUNDATION OF CANADA
LILLIAN AND DON WRIGHT FOUNDATION (THE)
MANITOBA LIQUOR CONTROL COMMISSION
PRICEWATERHOUSE COOPERS
SCHERING CANADA
SCOTIABANK
TORONTO STAR FRESH AIR FUND
THE WINNIPEG FREE PRESS

We would also like to thank our numerous additional donors – individuals, corporations and foundations – who each year express their confidence in us by making substantial supporting donations.

Working together with the corporate sector in Canada helps the Canadian Hemophilia Society (CHS) accomplish its mission and vision by extending our reach and reinforcing our messages. This enables us to...

- Offer national programs of training, education and awareness.
- Support research into hemophilia and other bleeding disorders.
- Produce educational publications, periodicals (such as *Hemophilia Today*) and keep our Web site www.hemophilia.ca up to date.
- Interact with stakeholders in the health care field to promote the well being of all our families.



www.hemophilia.ca

IN THIS ISSUE

SPRING 2007
VOL 42
NO 1



WORD FROM THE EDITOR	3
PRESIDENT'S MESSAGE	4
COMMUNITY NEWS	
CHS Research Summit: Charting the way forward	5
CHS approves funding for 7 research projects in 2007	6
Notice of Settlement: Pre-86/Post-90 Hepatitis C	7
What's new on the CHS Web site?	8
Rendez-vous Québec	9
Chapter spotlight	9
Upcoming events	12
YOUTH FILE	13
OUR STORIES	14
MEDICAL NEWS	
Hep C press review	15
Exploring rural health professionals' needs in providing hemophilia care	16
THE BLOOD FACTOR	
Letter on Hemophilia Inhibitor Genetics Study ..	18
Blood factor news	18
vCJD risk from plasma-derived factor VIII "is extremely low but may not be zero"	19
Factor VIII and IX concentrates licensed/ distributed in Canada	20
THE FEMALE FACTOR	
Symptomatic carriers: Can women and girls really express hemophilia?	22
THE GLOBAL PERSPECTIVE	
Quebec Chapter – Tunisia twinning visit	23



Steve and Mike Myers, p.14

WORD FROM THE EDITOR

François Laroche




You've probably heard it said that it's easier to reach the top than to stay there. This is often true, at least in professional sport. Some teams are winners thanks to an alignment of the stars that brings them luck, helping them garner top honours one year, only to become nothing more than a good team the next year, barely able to qualify for the playoffs. It's also like this for our organization.

Over the past 50 years or more, through the hard work of volunteers and dedicated health care providers, the CHS has managed to create a state-of-the-art care system for the treatment of hemophilia and other bleeding disorders. Thanks to the unrelenting work of a handful of these stalwarts, we have access to the highest quality care that is the envy of people with bleeding disorders all over the world. For all practical purposes, we have reached the top.

In the coming years, however, we will face a big challenge. Now that we've reached the top, we have to stay there. And a number of signs I've picked up here and there as the country's blood system is being reformed tell me that it's going to be a tough fight.

In Quebec, the process of decentralizing budgets for blood and blood products from Héma-Québec to designated hospitals already poses a threat to the integrity of our care. Of course, the main advantage of this new approach is that it makes hospitals more accountable when using blood and blood products—which were being supplied to them for free—as opposed to substitutes—for which they have had to pay until now. Shifting the budgets to hospitals does, however, bring with it a number of real or possible drawbacks for people with bleeding disorders. First of all, it creates an additional line item—and a major one—in hospital budgets. Two line items, actually, since fractionated products are separate from fresh components. Any hospital administrator with an iota of sense will eventually discover that the cost of fractionated products (including factor concentrates) is very high, and try to cut back—to the detriment of users of these products. The decentralization will also create a rift between the prescriber of the products—who practices in the hemophilia treatment centre (HTC)—and the distributor—who works in the hospital or is located at the regional blood bank. It would obviously be tempting for doctors in remote regions to control prescriptions for products that are budgeted and distributed in their hospitals, with the inevitable consequences: improperly prescribed drugs, budget overruns, and eventually, loss of the expertise currently existing in the HTCs.

But another threat also lurks... Now that factor concentrates produced by genetic engineering no longer contain human biological products (2nd generation products, like Kogenate[®] FS and Niasstate[®] contain albumin in trace amounts, while 3rd generation products like Advate[®] and Benefix[®] contain none), it would be tempting for provincial health officials to reclassify them as pharmaceutical products. As a result, these would no longer be paid for from the blood products budget, but would be covered by the users' public, group or private insurance plans—with all the exasperation that would entail for someone with a chronic condition like hemophilia. The possibility of discontinuing tracking of 3rd generation products was recently brought before the *Comité d'hémovigilance du Québec*. After the CHS's position was presented and discussed, the idea of discontinuing product tracking was set aside for the time being. But it is only a matter of time before it's back on the table, either in Quebec or somewhere else in Canada.

We must remain vigilant if we don't want the quality care for which we've fought these many years to gradually wither away. We certainly don't want to return to the dark ages of the 1950s. 



PRESIDENT'S MESSAGE

ERIC STOLTE

Dream our dream

"If you lose hope, somehow you lose the vitality that keeps life moving, you lose that courage to be, that quality that helps you go on in spite of it all. And so today I still have a dream." These words of Dr. Martin Luther King, Jr. were penned in the book, *The Trumpet of Conscience*, not long before his death. When I think of *hope* in our context, I think of *cure*. That cure will come from research.

We, like Dr. King, have a dream. We dream of a world free from the pain and suffering of bleeding disorders. We strive to improve the health and quality of life for all people with bleeding disorders and to find a cure.

The Hemophilia Research Million Dollar Club was created to help ensure that cure is found. The effort to raise the endowment that has now become a \$1.8 Million Dollar Club is probably the most successful fundraising initiative ever undertaken by the CHS. This demonstrates the power of hope in a cure.

There is an ancient Hebrew proverb that says, "Hope deferred makes the heart sick, but desire fulfilled is a tree of life." A modern translation of that same proverb states, "Unrelenting disappointment leaves you heartsick, but a sudden good break can turn life around." My son recently turned thirty years of age. (I can't be that old, can I?) I remember hearing soon after his birth that a cure was only fifteen or twenty years away. Then, many times since, I have heard the same prediction. Hope deferred.

But we have had some "good breaks". Recombinant products provide safety, efficacy and reliable supply. On the near horizon are products with longer half-lives. Gene therapy holds the promise of increased factor levels for longer periods of time. All these advances mean a betterer quality of life.

The recent CHS Research Summit, held in February in Toronto, was attended by physicians, nurses, physiotherapists, social workers, CHS Board and staff members, past and current researchers, representatives from Canadian Blood Services, Héma-Québec, the U.S. National Hemophilia Foundation, the five pharmaceutical companies in the Canadian factor concentrate market and the President of the Canadian Institutes for Health Research, Dr. Alan Bernstein. It was a wonderful success. Since 1990 the CHS has channeled four million dollars into peer-reviewed research yet has never stopped to analyse that investment. Through presentations and discussion there emerged a strong sense that the CHS must give leadership in this area. Our CHS Medical and Scientific Advisory Committee will now review the input and insights from the Summit in order to determine our best path forward. My heartfelt thanks to the organizing committee of Bruce Ritchie, Tom Alloway and Craig Upshaw and the excellent staff support provided by David Page and Wendy Wong in making this a landmark meeting.

I'm also excited about the opportunity that *Rendez-vous Québec* (May 24-27 in Quebec City) presents for us. Excellent consumer workshops along with interesting medical and scientific presentations will make an event of great value. Visit our Web site, www.hemophilia.ca, for more information. I look forward to seeing you in Quebec City where, together, we can continue to hold on to hope and dream our dream! ○

Dear readers,

In the course of our subscription address update for *Hemophilia Today*, we were pleased and touched by the many appreciative comments you took the time to send us. Below are some of the messages received. In addition to your written words, many of you were extremely generous with a total of more than \$8,000 in donations! From all of us at the CHS and the *Hemophilia Today* editorial team, thank you for this extraordinary support.

READERS' COMMENTS...

- Thank you for the many printed issues of *Hemophilia Today* that you have sent me over the years. I have now subscribed to the email version.

Let me congratulate you on your newsletter. It is beautifully done. As a former newsletter writer, I know how expensive it is to carry out this activity. I also know that it is appreciated by readers. I have greatly enjoyed having the Canadian perspective, and, tidbits of news I might otherwise not encounter. Let me wish you every success!

Carol K. Kasper, MD

Emerita Professor of Medicine, University of Southern California, Orthopaedic Hospital

- Thank you for this wonderful publication. Although I am pro-environment, the paper issue is very useful for our meetings with the parents. (free translation)

Yolaine Houle, SW

Sainte-Justine Hospital, Montreal, QC

- I am a fan of *Hemophilia Today*.

Dr. S. Kaiser Ali

Saskatoon, SK

- Thanks for a very informative magazine.

Mr. Chris Slocombe

Colchester, Essex, England

- Congratulations – publication very informative and VERY WELL done. Keep up the GREAT work.

Dr. Ron and Leni George

Calgary, AB

- On behalf of the GREEK HAEMOPHILIA SOCIETY (GHS), I would like to thank the Canadian Hemophilia Society for sending us *Hemophilia Today*. All these years that we have been receiving your publication we have shared the information given in every issue. Please be generous and continue to send it to us. We wish you every success in all your activities.

Jane Pittadaki

Executive Board Member, Chief editor of the Newsletter "OUR NEWS" of GHS

- Many thanks to *Hemophilia Today* and to the Canadian Hemophilia Society for the years I have enjoyed your excellent publication.

Dr. Campbell W. McMillan

Chapel Hill, NC, USA

ERRATUM

In the Fall 2006 issue of *Hemophilia Today*, p. 31, the article *Where do you find 25 wheelchairs?* was written by Sandra Squire, BSc, PT in the Vancouver, BC Adult Hemophilia Program at St. Paul's Hospital. We apologize to Ms. Squire for this unfortunate mistake.

COMMUNITY NEWS



CHS Research Summit: Charting the way forward

David Page

CHS Director of Programs and Public Affairs

On February 2, 2007, the Canadian Hemophilia Society brought together the Canadian bleeding disorder research community in what was called the *Research Summit*.

The Summit grew out of a desire to ensure that the CHS is getting the maximum benefit from its research programs. While the CHS has invested 4 million dollars in research since 1989, it had never stopped to analyze how we are running our programs. Can we be more effective? More efficient? Are there barriers to applying to CHS for grants that we can address? Is the focus of our research the proper one? Are there partnerships we should be pursuing? Are there other opportunities we should consider?

When the CHS asks its members, *What is our primary mission?* Almost all reply, *Research, a cure and improved quality of life.* Hope, through research, for a future free of the pain and suffering of inherited bleeding disorders is what helps many fam-

ilies cope. Research is also at the top of the list of priorities for our donors. Finding a cure is front and centre in our mission statement. Attracting bright young clinicians and researchers is key to the future of care for bleeding disorders. Therefore the research program and the results of this Summit are critical to the CHS and the bleeding disorder community.

The CHS received a very enthusiastic response to its invitation to attend the Research Summit. The attendees represented a wide variety of perspectives:

- the care providers: physicians, nurses, physiotherapists and social workers
- past and current researchers who have been awarded CHS grants
- research peer review committees without whom we could not operate our programs
- Canadian Institutes of Health Research
- Canadian Blood Services and Héma-Québec
- our counterpart in the U.S., the National Hemophilia Foundation
- our industry partners
- members of the CHS Board of Directors.

Many said they were excited by the meeting and had never attended one like it before. Many ideas were put forward as to how the CHS can maximize the benefits of its research programs, find matching funds and work even more closely with other groups interested in bleeding



disorders research. The responsibility for refining these ideas and formulating specific recommendations was given to the CHS Medical and Scientific Advisory Committee.

The proceedings of the Research Summit, including summaries of all presentations and discussions, are available on the CHS Web site www.hemophilia.ca.

The CHS thanks those who agreed to speak at the Summit. We are grateful to all participants who volunteered their time to advise us on our research programs.

The Research Summit would not have been possible without the generous support of our Presenting Sponsor, Bayer, and our Supporting Sponsor, Baxter. ○

Research,
a cure and
improved
quality of life.



Dr. Alan Bernstein, President, Canadian Institutes of Health Research, addressing the CHS Research Summit.



Participants at the Research Summit. Seated : Bruce Ritchie, MD; Lori Laudenbach, RN; Manuel Carcao, MD; David Page; Fred Ofosu, MD; Gonzalo Hortelano, MD; Greig Blamey, PT; Pam Hilliard, PT; Victor Blanchette, MD; David Lillicrap, MD; Maha Othman, MD. Standing: Lawrence Jardine, MD; Steve Green; Mark Blostein, MD, PhD; Wenda Greer, PhD; Tom Alloway, PhD; Ya-Gang Xie, MD; Anne Myerson; Pam Wilton, RN; Arlene C. Y. Ali, PhD; Jeff Bakker; Gordana Atanackovic, MD; Craig Upshaw; Eric Stolte; John K. Wu, MD; Patricia McCusker, MD; Dana Devine, PhD; Heinz Neuhaus; Paula James, MD; Connie Alvarez-Haines; Réal Lemieux, PhD; Anthony Chan, MD; Maureen Brownlow, SW; Jeff Rice. Missing from photo: Wendy Wong, Pauline Major.

CHS approves funding for 7 research projects in 2007

The Canadian Hemophilia Society is pleased to announce that seven research grants have been approved by the CHS Grants Review Committee and will receive funding in 2007. Complete reports on this research will be published in the Fall 2007 issue of *Hemophilia Today*.



Canadian Hemophilia Society
Help Stop the Bleeding



Canadian Hemophilia Society Research Grants

Supporting research towards improving the quality of life for persons with hemophilia and finding a cure have been goals of the Canadian Hemophilia Society (CHS) since it was founded in 1953. Since 1990, through funds provided by the **Hemophilia Research Million Dollar Club and the CHS**, the Society provides basic scientific research grants and studentships aimed at developing treatments for hemophilia and finding a cure.

Wenda L. Greer, PhD, FCCMG

Professor

Department of Pathology, Dalhousie University
Halifax, Nova Scotia

Project Title: **The Role of X-inactivation in the expression of hemophilia A in women**

(Second year funding)



Dr. Gonzalo Hortelano

Associate Professor

Department of Pathology, McMaster University
Hamilton, Ontario

Project Title: **Gene therapy of hemophilia A**

(Second year funding)



Dr. Maha Othman

Adjunct Assistant Professor

Department of Pathology and Molecular Medicine
Queen's University
Kingston, Ontario

Project Title: **Platelet type von Willebrand disease: An underdiagnosed cause of excessive mucocutaneous bleeding?**

(First year funding)



Dr. William P. Sheffield

Associate Professor

Department of Pathology and Molecular Medicine
McMaster University
Hamilton, Ontario

Project Title: **Factor VII(a) clearance behaviour**

(First year funding)



Care Until Cure Research Program

The **Care Until Cure Research Program** was established in the year 2000 in collaboration with **Wyeth Canada**. Wyeth Canada is engaged in the discovery, development, and commercialization of human pharmaceuticals through recombinant DNA and other technologies.

This program allows Canadian investigators to conduct research on various medical and psychosocial aspects of bleeding disorders. Grants are given for clinical research, including outcome evaluation, in fields relevant to improving the quality of life of persons with hemophilia, persons with von Willebrand disease or other inherited bleeding disorders, persons with related conditions such as HIV or hepatitis C as well as carriers of an inherited bleeding disorder.

JoAnn K. Nilson

Physiotherapist

Saskatchewan Bleeding Disorders Program
Saskatoon, Saskatchewan

Project Title: **Creating meaningful messages for individuals with mild hemophilia**

(Second year funding)



Dr. Paula James, FRCPC

Assistant Professor

Queen's University
Kingston, Ontario

Project Title: **The prevalence of symptomatic pediatric VWD**

(First year funding)



Dr. Rochelle Winikoff, FRCP

Staff Hematologist

CHU Ste-Justine
Montreal, Quebec

Project Title: **Non-steroidal anti-inflammatory drugs and menorrhagia revisited**

(First year funding)



Notice of Settlement Pre-86/Post-90 Hepatitis C Class Actions

The Government of Canada has agreed to settle several Class Action lawsuits commenced on behalf of people who became infected with the hepatitis C virus (HCV) from receiving blood and blood products in Canada prior to 1986 and after July 1, 1990. The proposed Settlement is not yet final and must be approved by the Courts in British Columbia, Alberta, Ontario and Quebec. Public hearings to approve the settlement were held in each of those provinces during the months of February and March. Class Members and their families had the opportunity to give their views on the fairness of the Settlement to the Courts. If the Courts approve the Settlement, further notices will be mailed to Class Members, posted on the Internet and published in newspapers explaining how Class Members may apply for compensation under the Settlement or choose to be excluded from the Settlement.

To settle the lawsuits, the Government of Canada has agreed to establish a compensation fund in the amount of \$962 million. People primarily or secondarily infected with HCV as a result of blood received in Canada during the period prior to January 1, 1986 and the period from July 2, 1990 to September 28, 1998 may apply for compensation under the Settlement. Estates of infected Class Members who have died, family members of infected Class Members, and the dependants of infected Class Members whose death was caused by their infection with HCV may also apply for compensation.

Under the terms of the Settlement, the Government does not admit liability.

Approved claimants will receive lump sum compensation based on their age, current disease level and the probability of disease progression in the future. Compensation is also available for people who have lost income or household services as a result of their infection with HCV, for estates of infected Class Members who have died, and for dependants of claimants whose death was caused by their infection with HCV. If the Settlement is approved by the Courts, Class Members should be able to apply to receive compensation from the Settlement in the first half of 2007.

At the Settlement approval hearings, Class Counsel requested that their legal fees of approximately \$37.29 million (plus GST and PST, where applicable) be approved by the Courts. The Government of Canada has agreed to add that amount to the settlement fund, as well as an amount for the administration of the settlement, so that no legal fees or administration expenses will be deducted from the \$962 million negotiated for Class Member compensation. ○

To read a complete version of the Settlement Agreement, go to: www.reko.ca/html/hepc_settleagreement.pdf

For more information:

Residents of British Columbia: www.kleinlyons.com

Residents of Alberta: www.marshallattorneys.com

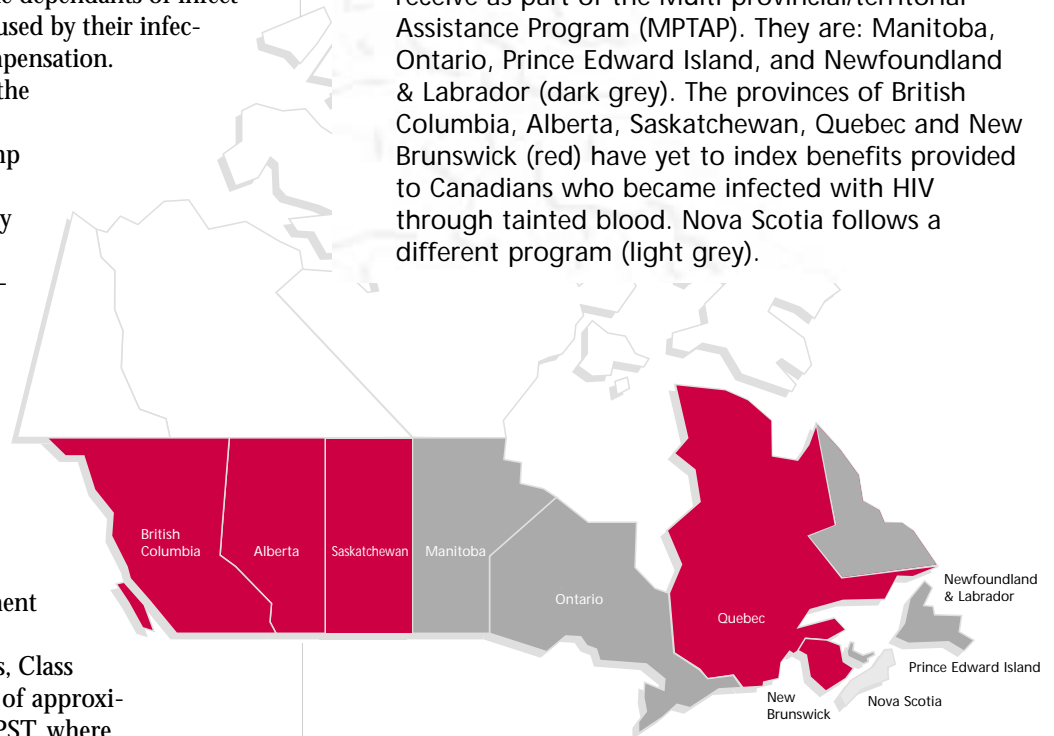
Residents of Quebec: www.lauzonbelanger.qc.ca

Residents of Ontario, Saskatchewan, Manitoba, the Atlantic Provinces, the Territories, and residents residing outside of Canada: www.reko.ca/html/hepatitisc.html

For updates check the CHS Web site at: www.hemophilia.ca

More work to be done on MPTAP indexation

Across Canada, four provinces have already indexed to the cost of living those benefits individuals receive as part of the Multi-provincial/territorial Assistance Program (MPTAP). They are: Manitoba, Ontario, Prince Edward Island, and Newfoundland & Labrador (dark grey). The provinces of British Columbia, Alberta, Saskatchewan, Quebec and New Brunswick (red) have yet to index benefits provided to Canadians who became infected with HIV through tainted blood. Nova Scotia follows a different program (light grey).



Representatives from the CHS, who are involved with Chapters in those provinces where benefits have not yet been indexed, continue to press their governments to act.

NOTICE

ANNUAL GENERAL MEETING

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

Saturday, May 26, 2007 - 8:30 a.m. at the Delta Québec Hotel, Quebec City

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 2007-2008.
4. To receive the audited financial statements of the Canadian Hemophilia Society for the year ended December 31, 2006.
5. To appoint an auditor for the ensuing year.
6. To approve the recommendation from the Board of Directors to amend the CHS by-laws to allow the Chair or a Co-Chair of the National Youth Committee to sit on the National Board of Directors as an ex-officio member.

That Section IV, Article 4.03 (c) of by-law no. 1 be amended as follows:

The ex-officio directors for each term shall be the immediate Past President of the Corporation, the Chairman of the Medical and Scientific Advisory Committee 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, a new Chairman of the Medical and Scientific Advisory Committee 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 transact such other business as may properly come before this Annual General Meeting of the members of the Canadian Hemophilia Society.

James Kreppner
Secretary

What's new on the CHS Web site?



Rendez-vous Québec – Find out about the Canadian Hemophilia Society Annual General Meeting and Medical and Scientific Symposium that will take place in Quebec City, May 24-27, 2007.

www.hemophilia.ca/en

Red, White & You event – Learn more about this outstanding fundraising and awareness program intended to educate people about inherited bleeding disorders while generating needed revenue to carry out our mission.

www.hemophilia.ca/en



A new expanded Web site section for *Hemophilia Ontario* and its regions with news of recent and upcoming activities

www.hemophilia.ca/en/8.1

A full report on the CHS *Research Summit*, held February 2, 2007 in Toronto

www.hemophilia.ca/en

The new CHS *Case for Support*

www.hemophilia.ca/en/1.8

A new section describing the international work of the CHS and its Chapters

www.hemophilia.ca/en/11.6

Our Stories – The “Bruise Brothers”

www.hemophilia.ca/en/2.9

Regular updates to the home page highlighting “Upcoming events” in the bleeding disorders community.

www.hemophilia.ca/en

www.hemophilia.ca



CHS Annual General Meeting and Medical & Scientific Symposium and Consumer Workshops

DELTA QUEBEC HOTEL, QUEBEC CITY

SCHEDULE OF EVENTS

THURSDAY, MAY 24, 2007

- 8:30 - 20:30 New Team Workshop
16:00 - 22:00 Registration

FRIDAY, MAY 25, 2007

- 8:00 - 9:00 Registration
9:00 - 17:00 CHS Medical & Scientific Symposium
(simultaneous translation)
17:00 - 18:30 Reception - Exhibit Area
19:00 - 21:00 Youth Session
19:30 - 20:30 Board Orientation

SATURDAY MAY 26, 2007

- 8:30 - 17:00 AHDCDC Annual Meeting
CANHC Meeting
CPHC Meeting
Social Work Group Meeting
CHS Annual General Meeting
9:30 - 9:45 Break
9:30 - 12:00 Twinning Workshop
(for those responsible for current
WFH/CHS organizational twinning)
9:30 - 11:00 Volunteer Recruitment Workshop
11:00 - 12:00 Fundraising Workshop
12:00 - 13:00 Lunch
13:00 - 17:00 CHS Board of Directors' Meeting
13:00 - 17:00 All About Carriers Workshop
or
13:00 - 15:00 Navigating the ER Workshop
15:00 - 15:15 Break
15:15 - 17:00 Hepatitis C Information Session
19:00 CHS Banquet

SUNDAY, MAY 27, 2007

- 8:30 - 14:00 CHS Board of Directors' Meeting
(continuation)
8:30 - 12:00 AHDCDC Meeting (continuation)
Medical Meetings (if required)
9:00 - 12:00 "Hands-on" Fundraising Workshop
12:00 Lunch



PRINCIPAL SPONSORS

CHAPTER SPOTLIGHT

■ Nova Scotia Chapter

Nova Scotia Chapter
to be in 2007 pumpkin race!

Dianna Cunning

I am very excited to announce that this year the Nova Scotia Chapter will be entering the Windsor/West Hants Pumpkin Festival. Over 40,000 people attended in 2006. This is a large media event. People from all over the world come to see this regatta. The Farmer's Almanac comes annually from Atlanta, Georgia with a media crew. The CBS Sunday Morning Early Show and even Martha Stewart were scheduled to come last year but immigration and legalities prevented it. This event is huge. Numerous articles and photos have been in publications world wide. CBS provides full coverage of the festival along with corporate sponsors. Nova Scotia Chapter will be a corporate sponsor. Three Metro Halifax radio stations do a complete 2-week promotion prior to the pumpkin festival. There are also 3000 brochures in which we will be named. The media attention that this event and its corporate sponsors receive is enormous.

We will receive a massive pumpkin for the regatta, carve it out and paint it with the Red, White and You theme. Wim Peters has agreed to paddle the pumpkin across the Windsor River with about 40 other competitors.

We will have a float in the parade with a Nova Scotia Chapter banner. We will pass out candies and brochures about hemophilia and bleeding disorders. On the back of the float will be our pumpkin, Wim, chapter members and children. I truly feel that we will get excellent exposure for our Nova Scotia Chapter and the CHS, and greater awareness of bleeding disorders. Our participation will promote education about bleeding disorders and help to raise donations. This will be the most coverage our Nova Scotia Chapter has ever received. Let's all be there in October to celebrate... together!



We will receive a massive pumpkin for the regatta, carve it out and paint it with the Red, White and You theme. Wim Peters has agreed to paddle the pumpkin across the Windsor River with about 40 other competitors.

■ Quebec Chapter

The CHSQ's spotlight activity, the annual members family weekend, was held on March 16 to 18 at Auberge Matawinie in St-Michel-des-Saints, and once again this year was a resounding success. More than 180 people attended.

The program featured a variety of workshops that offered something for everyone (coffee klatches for youngsters or parents, workshops on physiotherapy, blood product safety and supply, and even relaxation). One of our young members organized a fund-raising activity, the Factor Auction, as part of a school project. The event was a lot of fun, and managed to bring in a total of \$912 for the organization.

The organization's Annual General Meeting, the focal point of the weekend, was held on Saturday afternoon. A number of presentations on results for the year 2006 and possible new paths of development for the CHSQ were made by volunteers and staff. The presence of three delegates from the Tunisian Hemophilia Association provided an opportunity to update members on our twinning project and raise awareness of the quality of services provided in Canada, as well as the importance of international cooperation projects between hemophilia associations. In his presentation, David Page explained how vital cooperation between CHS and provincial chapters is, as he reviewed a number of joint projects currently underway.

In short, the 2007 weekend was enjoyable, useful, a great forum for discussion and exchanging information, and spawned many promising ideas for fundraising, programs, and the future of our organization. Mission accomplished!



The Lifetime Member Award was presented to François Laroche by Patricia Stewart for his outstanding contribution to the organization's advancement.



François Laroche (far right) presented the delegates from the Tunisian Hemophilia Association with "dreamcatchers" as a memento of their visit to Quebec. Left to right: Taoufik Raissi, Secretary-general; Professor Raouf Hafsia, Vice-president; and Islem Nafti, President.

■ Hemophilia Ontario

Hemophilia Ontario has moved!

As of April 2, 2007 you can find Hemophilia Ontario at a new address. We've moved up in the world, to the eighth floor! Our new address is:

Hemophilia Ontario
45 Charles Street East, Suite 802
Toronto, ON M4Y 1S2

If you are in Toronto, please drop by and see our new digs!

New Staff

Hemophilia Ontario has more than a few new employees to announce. **Aaron Cantor** is the new Executive Director for Hemophilia Ontario. **Justine Casserly** is working as Administrative Associate for Hemophilia Ontario and TCOR. **Andrea Lajdecki** is our new Regional Service Coordinator for TCOR. **Terri-Lee Higgins** is our new Regional Service Coordinator filling the maternity leave in SWOR. *Welcome to all the new staff in Ontario!*



The staff of Hemophilia Ontario/TCOR: (from left to right) Administration and Finance Coordinator Andrée Delisle, HIV/HCV Program Manager Sarah Crymble, Executive Director Aaron Cantor, Regional Service Coordinator (TCOR) Nigel Small – Front Row: Regional Service Coordinator (TCOR) Andrea Lajdecki, Administrative Associate Justine Casserly.

Winter Wanakita 2007

Sarah Bradshaw,
Member of TCOR, National Youth representative

On February 2-4, 2007, eleven brave young guys and girls set off to Camp Wanakita on Koshlong Lake in Haliburton, Ontario. For six of the young men, it was their first time on this winter trip; for five, it was their first time ever on a Hemophilia Ontario Youth (HOY) trip. We did everything from cross-country skiing, to snowshoeing, to our annual broomball game. This year we challenged an all-girls school from Oshawa. After a tough fight, Team Hemo prevailed, winning 2-0. We can't forget about those who braved the high ropes in the middle of winter and the same group who lived through the almost six-hour drive home in the storm after having our chicken lunch... leave it to the boys to be boys!

We'd like to welcome MJ, Trent, James, Greg and Korey to the group of select few who are brave enough to come up North with us in February.

Special thanks go out to Julia Sek and Dane Pederson for taking the time to join us on our trip and planning everything so wonderfully for us.



Hemophilia Ontario – Winter Youth Trip



■ Hemophilia Saskatchewan

Hemophilia Saskatchewan AGM

Saturday, March 10, 2007 was a busy day for Hemophilia Saskatchewan. The Annual General Meeting brought everyone up to date with the business side of the organization. Twelve members were elected to the board.

Following the AGM, there was good fellowship over a catered roast beef dinner.

After dinner, Darla Walz gave a presentation, *Live Well with Chronic Conditions*, showing how you can control how you live with a chronic condition.

A wonderful skit by Colleen Beuhler and Janice Kelly from the Saskatchewan Bleeding Disorders Program illustrated how action plans can be very beneficial to the patients who attend clinic.

In conjunction with the AGM there was a very successful cookie dough sale.

■ Alberta Chapter

Since our change in governance several years ago, the regions have strived to collaborate and create a single strategic direction for the province, develop a synergistic approach to dealing with the health authorities, as well as expand our support and education programs into small rural communities outside of Calgary and Edmonton. This collaboration started under the direction of Craig Upshaw and Tony Niksic, the regional presidents, but has an increased focus with the expansion to include Clara Penner representing the Medicine Hat/Lethbridge region, and with the recently elected executive in the southern region. In November, during the Southern Region's Annual General Meeting and Christmas gathering, a new executive was elected consisting of Darlene Gates (President), Stacey Johnston (VP), Crystal Verbeek (Treasurer) and Susan Anderson (Secretary).

As part of our reaching out to all communities in the province, the Northern Region extended an invitation to all members of the Alberta Chapter to attend the annual "beach party" at West Edmonton Mall. This very popular event brings families together to share experiences and connect in a warm environment during the harsh February Alberta winter. Families attended the beach

■ Manitoba Chapter

2007 Bayer Healthcare's Gala Dinner of Culinary Inspirations from Around the World

The 2007 Manitoba Chapter Gala held on Saturday March 3, 2007 was a huge success! Our 230 guests enjoyed a sumptuous meal prepared by 5 of Winnipeg's best Chefs. Thank you to the committee who worked so hard for 8 months to ensure that the evening was perfect. Wendy Wong, the Canadian Hemophilia Society's National Director of Resource Development, came from Toronto to observe and was a huge help! This year we raised over \$25,000. The 2007 Gala Committee is working to create a manual for the event and hope to share the information with other Chapters that might like to try this fundraiser. ◊



2007 Gala Head Table, left to right: Shane Keilback; Christine Keilback, 2007 Gala Chairperson; Wendy Wong, CHS National; Guest MC Jon Ljungberg from *CityTV Breakfast Television*; Christine Ljungberg; CHS-MC President Tony Tavares; from Platinum Sponsor Bayer Healthcare, Tom Hsu; and Joanne Ukno, Business Relations Manager, Bayer Healthcare, Pharmaceutical Division.



Back row, left to right: Bruce Cameron, Shane Keilback, Cory Prestayko, John Rogasky. Front row, left to right: Lynda Regan, Katherine Cameron, Christine Keilback, Wendy Wong, Donna Rogasky, Colleen Cario.

UPCOMING EVENTS

NOVA SCOTIA CHAPTER

- MAY 2007 – Chapter Fundraising and Maritime Family Weekend Planning Committee Meetings. Royal Artillery Park Officer's Mess, Halifax, Nova Scotia.
- MAY 25, 2007 – Deadline for articles to be included in the June issue of the Nova Scotia Chapter newsletter.
- JULY 28-29, 2007 – Maritimes Family Weekend 2007 to be held at Saint Mary's University, Halifax, Nova Scotia.
- JULY 2007 – Commencement of NS Chapter Quarterly Executive meetings. Cambridge Military Library, Royal Artillery Park, Halifax, Nova Scotia.
- AUGUST 2007 – Maritimes Adventure Camp 2007 to be held at Scotian Glen, Nova Scotia.

QUEBEC CHAPTER

- MAY 5, 2007 – Bowl-a-thon (Quebec City)
- MAY 6, 2007 – Bowl-a-thon (Montreal)
- MAY 12, 2007 – Strategic planning day at the Estrimont Suites & Spa.
- JUNE 1-3, 2007 – *Just the Gals* weekend at the Estrimont Suites & Spa.
- AUGUST 5-10, 2007 – 2007 Summer Camp at Camp Portneuf, located in St-Raymond-de-Portneuf.
- SUMMER 2007 – Youth camping weekend
The youth committee of the CHSQ is preparing its annual event. This year, young people aged between 15 and 28 are invited to go on a camping trip.
- SEPTEMBER 15, 2007 – Deadline to send in applications for student bursaries.

HEMOPHILIA ONTARIO

There are always many exciting developments in the works at Hemophilia Ontario and the regional offices. Plans are being made for a tri-regional *Just the Guys* event, a provincial educational summit for volunteer development, as well as the *Toronto Marathon*. All of these events will be held in the fall. The staff is working hard to make these events a success!

- JULY 1-15, 2007, Camp Wanakita 2007 – As we ramp up for another year at camp, we are feeling prepared and ready to make this the best year yet! We have put in place a new registration process, post camp committee meeting and Camp Committee. We are ready to do this right!

We are looking forward to providing a camp experience for 55 campers with inherited bleeding disorders. Hemophilia Ontario is excited about this new opportunity and look forward to a great partnership.

If your child is attending camp or you are a past camper and want to volunteer, please let us know. We need people with camping experience for feedback to make our camp program focused and most of all FUN!

If you require any information about Hemophilia Ontario's Camp Wanakita, please contact Justine Casserly, 1 888 838-8846.

- AUGUST 15-19, 2007, Summer Youth Trip – The next scheduled HOY trip is a Canoe Trip. We hope to see you this summer for our annual summer trip. Don't delay in contacting Sarah Crymble (Hemophilia Ontario) or your local Regional Service Coordinator for more information, and keep your eyes open for the flyers! Summer is just around the corner!

HEMOPHILIA SASKATCHEWAN

- JUNE 2, 2007 – Regina BBQ
- JUNE 9, 2007 – Saskatoon BBQ
- JULY 21-22, 2007 – Guys Getaway

ALBERTA CHAPTER

- JUNE 2007 – Northern Region: Family Picnic and Education event
- JUNE 30, 2007 – Southern Region: Zoo, Picnic and Education event
- SUMMER/FALL 2007 – Southern Region: Alberta Children's Hospital new clinic open house and *Passport to Fitness* Educational event
- SEPTEMBER 7-9, 2007 – Alberta Chapter: Annual General Meeting and Family Retreat, Goldeye

Keep an eye out for Alberta Chapter announcements with registration and confirmation of event dates.



Calling all youth!

Emil Wijnker

In the fall of 2006, the Canadian Hemophilia Society identified the need for a committee to address the issue of youth involvement in the organization, and so the National Youth Committee was born! The CHS-NYC consists of very excited and energetic individuals who represent almost all regions of Canada who are working together "to support, promote and address youth involvement within the organization." We are currently working on acquiring a representative from British Columbia to complete our coast-to-coast committee! The Committee is co-chaired by Sarah Bradshaw and me and is supported by H el ene Bourgaize, CHS Director of Volunteer Development and Human Resources, and Julia Sek, Ontario Hemophilia Provincial Coordinator.

At our first meeting held in Montr el last February, the committee identified the various roadblocks in our various communities to youth involvement. We are currently working towards overcoming them and engaging Canadian youth with bleeding disorders in order to support each other and develop amazing local, provincial and national youth programming. We recognized that some regions already had very developed youth programming and some regions wanted to do this, but needed some help getting the ball rolling. That is one of our main goals and we have begun working together on an unprecedented national level towards this end! Keep your eyes and ears open for awesome youth-oriented programs coming to a Chapter/Region near you!

Our first All-Canada event is going to be a Youth Forum at Rendez-vous Qu ebec where we will be sharing our plan of action for 2007. Youth will have the opportunity to meet others from all across Canada, network, attend a Medical Symposium on bleeding disorders and celebrate with the CHS-NYC as we officially launch our all-new Web page! If you want to attend Rendez-vous Qu ebec, con-

tact your local youth representative or your Chapter's or Region's Board of Directors.

If you want to get involved, want to help plan/organize an event, or just want some information about CHS's new youth initiative, feel free to contact your local youth representative at the following addresses! They will be excited to hear from you!  

MEMBERS OF THE NATIONAL YOUTH COMMITTEE

- Crystal Verbeek** (Alberta) crystal.verbeek@uleth.ca
- Sheri Jevne** (Alberta) sherijevne@hotmail.com
- Jesse Katzman-Beimuts** (Saskatchewan) jesse.katzman@gmail.com
- Silvana Moran** (Manitoba) spanish_raddish@hotmail.com
- Jaime Villeneuve** (Ontario – Ottawa/OEOR) pyro_jaime@hotmail.com
- Adam Del Gobbo** (Ontario – Toronto/TCOR) b3457_88@hotmail.com
- Sarah Bradshaw** (Co-Chair) (Ontario – Toronto/TCOR) sarahbradshaw_@hotmail.com
- Greg Stutz** (Ontario – Hamilton/CWOR) greg_stutz@hotmail.com
- Emil Wijnker** (Co-Chair) (Ontario – London/SWOR) ewijnker@gmail.com
- David Pouliot** (Qu ebec) david.pouliot@umontreal.ca
- Martin Kulczyk** (Qu ebec) kulczykalpha@hotmail.com
- Patrick Syriani** (Qu ebec) patricksyriani@hotmail.com
- Robert Maye** (Newfoundland) tapionn@mail.com
- Meech Kean** (New Brunswick) jackojim@hotmail.com
- Katie Hines** (Nova Scotia) katieakalips05@hotmail.com



Members of the newly created National Youth Committee met for the first time in Montreal on February 10 and 11, 2007.



The “Bruise Brothers” play the blues

Clare Cecchini, CHS Program Coordinator

It has often been said that to play the *blues* you have to have *lived the blues*, or at least overcome challenges or hard times at some point in life. In the case of Steve and Mike Myers, blues musicians from Ottawa, Ontario, their experiences growing up with mild hemophilia in the 1960s and 70s led them to their own special style of Mississippi hill-country juke joint blues.

The Myers brothers were diagnosed at a young age with mild hemophilia when they almost bled to death after having their tonsils removed. There was no history in the family. After the diagnosis, they were discouraged from participating in active sports like hockey and picked up instruments instead of baseball bats.

“Without realizing it at the time, this was probably our first real experience with the blues,” Steve remembers.

Even though the boys could not play hockey or go roller-skating, their father kept them busy doing outdoor activities like camping, canoeing and fishing and never allowed them to use their hemophilia as an excuse.

Living through the tainted blood tragedy in the 1980s and seeing the impact on the bleeding disorders community was another challenge. “Mike and I count our blessings for having made it through these tough times and our personal experiences are reflected in our blues.”

Music was always a big part of their lives while growing up in Ottawa and the Gatineau region. Their mother’s French Canadian family was musical and many of their uncles and cousins played instruments, mostly old-time music and country & western. Their first contact with blues music occurred in the early seventies when they saw BB King performing on CBC television. As they began to explore playing blues music they were influenced by musicians like Johnny Winter, John Lee Hooker and Muddy Waters.

In the early 1990s, the TV documentary *Deep Blues/A Musical Pilgrimage to the Crossroads* had a profound impact on their musical direction. They began a new musical and spiritual journey, and developed their own style of “hypnotic trance-blues and rockin’ boogie”. By the late nineties they were travelling to the Mississippi delta on yearly pilgrimages and performing in “juke joints” in places like Memphis and other delta towns, including a club owned by Morgan Freeman.

As Steve was proud to point out, “He asked Mike and I to autograph a poster for the wall of his club.”

In 2005 they recorded their CD *Straight From the Woods* with the legendary delta drummer Sam Carr of the *Jelly Roll Kings*. In 2007 they will be returning to the delta to record another session for their upcoming CD release.

The brothers have factor VIII levels of 20 to 30% and, since they have mild hemophilia, are not on home infusion. They receive treatment as needed at the Hemophilia Treatment Centre in Ottawa and in emergency situations rely on their *FactorFirst* cards. When asked if hemophilia has affected their lives as musicians, Mike mentioned that he suffers from arthritis in his hands and has had to adapt the picking style he uses when playing the guitar. He also recalled the time that Steve bit his tongue minutes before a show and it was bleeding badly during the performance. Mike joked, “It’s lucky that he doesn’t do the vocals!”

Steve and Mike are both married and Mike has a ten-year-old son. Like many musicians, the brothers work by day and play the blues by night. Mike is involved in a family-run pollution abatement environmental business and Steve recently completed a degree in criminology at Carleton University. They plan on retiring within ten to fifteen years, which would give them the opportunity to realize their dream of going on the road for several months a year to perform the blues.

Members of the Ottawa and Eastern Ontario Region (OEOR) of Hemophilia Ontario had a unique opportunity to hear the Myers Brothers, or as they sometime refer to themselves, the *Bruise Brothers*, perform on March 24, 2007 at the Region’s Annual General Meeting and Family Weekend. In past years, Steve and Mike volunteered on the OEOR Board and attended several meetings and retreats, but this was the first time that they had performed for the CHS.

To find out more about the Myers Brothers Band, please visit their website at www.quazimojoblues.com. ☺



Mike Myers, seated on left; Steve Myers, standing on right.



Hep C press revue

Jeff Rice,

CHS Hepatitis C Program Coordinator

■ Effectiveness of complementary and alternative medicine in hepatology

Clinical Gastroenterology and Hepatology. January 13, 2007 – Use of complementary and alternative medicines (CAM) is increasing among individuals with liver disease, with the most popular CAM currently in use being herbal therapies. The single most commonly used herbal agent is silymarin, a mixture of the active ingredients of the milk thistle plant. In animal models, many of the frequently used CAM agents have shown anti-inflammatory and antifibrotic effects.

Although many human studies have shown improvements in subjective symptoms (well being) and liver biochemistry, “there are no convincing data to suggest a definite histologic and/or virologic improvement with most of these agents.”

“Poorly designed studies, heterogeneous patient populations, lack of standardized preparations, and poorly defined non-objective end points may partly explain the conflicting reports in the literature,” write the authors.

It has been well documented that liver toxicity and/or unwanted drug interactions may occur from the use of many herbal medications (e.g. St. John’s Wort adverse interaction with HIV protease inhibitors). Physicians are urged to talk to their patients about their potential use of CAM in order to help avoid use of any toxic agents or those that might cause deleterious drug interactions.

“Only well-designed, randomized, controlled trials will be able to ascertain whether CAM has any role in the management of patients with acute or chronic liver diseases. Until such time, the use of CAM cannot be recommended as a therapy for patients with liver disease.”

■ Occult HCV infection a milder disease than chronic HCV

Journal of Viral Hepatitis 14(1): 36-40. Jan. 2007 – Occult or “hidden” HCV infection is a term used to describe individuals who have HCV RNA in their livers, but no detectable HCV RNA or anti-HCV antibodies in their blood.

Spanish researchers previously determined that occult HCV was sometimes present in patients with persistently abnormal liver function of unknown cause. Investigators sought to compare the characteristics of 68 patients with occult HCV infection and 69 untreated patients with typical chronic HCV (detectable HCV RNA and anti-HCV antibodies in the blood), matched for sex, age, body mass index and duration of abnormal liver function tests.

The results indicate that AST and ALT levels were higher in patients with chronic HCV than in those with occult HCV. Chronic HCV patients also had higher levels of gammaglobulin, iron, and alpha-fetoprotein, a marker associated with hepatocellular carcinoma. In contrast, cholesterol and triglyceride levels were significantly higher in patients with occult HCV infection. The rate of necro-inflammatory activity and fibrosis was higher among chronic HCV patients. The mean percentage of infected hepatocytes (liver cells) was higher in chronic HCV patients than in those with occult HCV.

The authors wrote, “Occult HCV infection is a milder disease than chronic HCV, and this could be related to the significantly lower number of infected hepatocytes observed in occult HCV.”

■ Liver transplantation in HIV-HCV co-infected patients: A case-control study

Transplantation 83(3): 354-358. Feb. 15, 2007 – Traditionally, people living with HIV were not considered suitable candidates for liver transplantation, but in the era of highly active antiretroviral therapy (HAART), studies have shown that HIV positive individuals with well-controlled virus and relatively preserved immune function can have post-transplant outcomes nearly as good as those of HIV negative individuals.

For HIV positive and HIV negative transplant recipients alike, however, HCV is associated with poorer outcomes, since HCV nearly always infects the new liver graft.

Researchers compared outcomes for 9 co-infected liver transplant patients and the HIV negative HCV-infected patients who received transplants before and after each co-infected patient. Immunosuppressive regimens (used to prevent organ rejection) consisted of tacrolimus with steroids or mycophenolate mofetil.

Results indicate that acute cellular rejection occurred in 44% of the HIV-HCV co-infected patients and 22% of the HIV negative patients. 3-year actuarial patient survival rates were 87.5% for the co-infected group and 93.7% for the HIV negative group. Acute HCV re-infection occurred earlier (2.3 vs 4.3 months) and was more cholestatic (mean bilirubin 10.8 vs 1.6 mg/dL) in the co-infected patients. 8 co-infected patients (100%) and 9 HIV negative patients (64.3%) received antiviral treatment with pegylated interferon plus ribavirin. 1 co-infected patient (20%) and 1 HIV negative patient (11.1%) achieved sustained virological response.

The authors wrote, “Short- to mid-term results of liver transplantation in HIV-HCV co-infected patients were excellent and similar to those of non-HIV -infected patients.”

■ Shorter, simplified treatment option may encourage more patients to seek treatment

BASEL, Switzerland, March 6 – Some hepatitis C patients with difficult-to-treat HCV genotype 1 who respond quickly to treatment with a combination of pegylated interferon alfa-2a plus ribavirin can benefit from a shorter and simplified course of therapy. Following a European Union (EU) decision, a subset of patients with genotypes 1 and 4 HCV who receive rapid viral response can now receive a shortened, 24-week duration of treatment.

The EU approval is based on data from two pivotal clinical trials, with results showing that among patients who achieved a rapid viral response

continued from p. 15

(undetectable viral load at week 4) in the first month of treatment, up to 93% of patients with genotype 1 HCV and with a low pre-treatment viral load, and 83% of patients with genotype 4 were cured following only 24 weeks of therapy – a similar cure rate to that seen following 48 weeks of therapy.

“This is excellent news for patients with hepatitis C,” said Dr. Peter Ferenci, Professor of the Department of Internal Medicine IV, Gastroenterology and Hepatology at the University of Vienna. “This means that patients can find out within one month of starting therapy if they have an excellent chance of being cured and can benefit from a shortened treatment duration.”

■ Bill would let men with HIV use reproduction techniques

SACRAMENTO, March 6 – California State Senator Carole Migden (Democrat, San Francisco) has introduced legislation that would allow couples to use reproductive services when the man has HIV. Before a couple could try artificial insemination or in vitro fertilization, the man’s semen would first be treated by sperm washing to minimize the chance of transmitting HIV.

Migden said current law discriminates against HIV-positive men and that her legislation, Senate Bill 443, would “ensure equal reproductive rights for women,” regardless of their partner’s HIV status.

One HIV-positive man who lives in the Sacramento area said he and his wife have risked trying to conceive on their own, to no avail. Their last hope, he said, is in vitro fertilization, but California law precludes them from trying.

“She cannot get pregnant without reproductive assistance, but because I have HIV there is no way for them to use my sperm. It’s unfair.”

California is one of two states where couples with an HIV-positive male can’t take advantage of so-called assisted reproduction services. Many of those couples try to conceive instead through intercourse, increasing the risk of infecting the woman and, if that

happens, potentially the child.

The risk of transmitting the virus from an infected man to uninfected female through intercourse depends on the amount of virus in the blood stream. It generally ranges from 1 in 1,000 to 1 in 8,000 per act of intercourse, Cohan said, but those odds multiply the longer it takes to get pregnant.

By contrast, the 3,800 reported cases outside California in which couples with an HIV-positive man have used reproductive assistance hasn’t turned up a single instance of transmission to the female partner, said Cohan, an assistant professor in the University of California-San Francisco’s Department of Obstetrics, Gynecology and Reproductive Sciences. ○

Exploring rural health professionals needs in providing hemophilia care



Andrea Pritchard, RN, PhD(c);
Faculty – Mount Royal College, Calgary,
Alberta Nurse Coordinator,
Calgary HTC, 1993-2005
apritchard@mtroyal.ca

Rural hemophilia care

According to the Public Health Agency of Canada (2003), “rural Canada occupies ... around 95% of Canada’s territory... [and] about 30% of Canada’s population live in rural and remote areas of the country. In rural Canada, as in urban Canada, good health is a major resource for social, economic, community and personal development.” According to Taylor (1999) and UBC (2001), Canadians living with bleeding disorders identified difficulties associated with accessing rural healthcare services with required expertise. Furthermore, the urgent nature of bleeding and the relative rarity of hemophilia presents an obvious challenge for those healthcare providers practicing in geographically isolated rural communities. This is compounded by the fact that urban-based HTC providers may not be fully aware of the unique chal-

...the urgent nature of bleeding and the relative rarity of hemophilia presents an obvious challenge for those healthcare providers practicing in geographically isolated rural communities.

lenges that exist in the rural health care setting (MacLeod, 1999). Given the nature of bleeding disorders and coordination of specialty care in a rural setting, this issue was identified as a priority to explore and address. In particular, the question arose if rural providers had adequate preparation and support for safe and effective hemophilia care of HTC families.

Rural hemophilia study

This topic was formally studied as my Masters thesis research project with the assistance of my classmate and rural health expert, Kari Simonson, and faculty advisor, Dr. Kathleen Oberle; this article is dedicated in memory to Dr. Marlene Reimer, Acting Dean-Department of Nursing at the University of Calgary who was also an instrumental part of our research team. Our study was recently published in October 2006 by the

Australian Journal of Rural Health in an effort to share hemophilia related information to an audience that has widespread international rural readership. The following summary has been adapted from that article (Pritchard, Reimer, Simonson & Oberle, 2006).

Our study had two phases: focus groups (phase I) and telephone interviews (phase II). We invited participation by 20 interdisciplinary healthcare professionals or 'providers' who worked in rural communities that had at least one family living with hemophilia. These rural providers were from five different hospitals within southern Alberta and southeastern British Columbia, and included nursing, medicine, lab technology, social work and physiotherapy. Learning and resource needs, capacities and related barriers, and facilitators to rural hemophilia care were identified as 5 main themes represented in our "C-STOP"* model:

1. Communication network - Facilitate multidirectional, connection of families, rural providers and specialty clinic staff.
2. Subjective knowledge - Provide readily available individual care plans for family, rural providers and specialty clinic staff.
3. Team roles - Establish clarity of responsibilities for family, rural providers and specialty clinic staff.
4. Objective knowledge - Make available succinct resources such as protocols, practice guidelines and standards.
5. Partnerships - Recognize shared expertise, affiliation, and accountability amongst families, rural providers and specialty clinic staff (Pritchard, Reimer, Simonson & Oberle, 2006, p. 188).

Rural healthcare providers identified these five key elements as requirements for safe and effective rural hemophilia care.

Comments by study participants revealed their direct insights (Pritchard, 2004, p. 58-61):

1. Communication - Rural providers... "need to know what the family knows... need to be as informed... when we don't know these things it causes discomfort and difficulty... sometimes we use a different solution than at the specialty clinic but we have

our rationale. We are still competent but different."

2. Subjective knowledge – "The actual care plan starts with the basics of hemophilia care and objectives of assessment. Then as the staff get to know the family they start to fine tune the care plan."
3. Team roles – "Our staff take care of many patients at once... hemophilia just happens in this larger picture. This needs to be respected: what we can do and what we are faced with."
4. Objective knowledge – "We need drugs used, dosages, facts about hemophilia, what to watch for when they come in, quick reference tools, guidelines and protocols. We need specific information about hemophilia care... We don't need pages, just basics."
5. Partnerships – "The hemophilia clinic is like the hub of the wheel with the spokes going out to everyone involved. It's not a quick fix – it comes as a whole package."

Using findings in practice

We used study findings to develop rural resources at the Calgary HTC including an interdisciplinary education day. This event focused on rural healthcare provider education for nurses, physicians, lab technologists, and physiotherapists from hospitals throughout southern Alberta and southeastern British Columbia, including 34 onsite participants, and telehealth links to 5 rural hospitals. Presentations given by HTC, CHS and CBS representatives included topics such as emergency care, factor replacement therapy, care of muscles and joints, and care of the person and family. Pre- and post-test surveys revealed that participants had an increased overall readiness to provide rural hemophilia care regarding all 5 key elements of the C-STOP model.

Now what?

As discussed with CHS program planners, study findings and C-STOP model may support the implementation of the *FactorFirst* program. I look forward to continuing my work with the CHS on their commendable initiative. Also, I will be presenting this study in May 2007 at

the upcoming Alberta provincial conference for registered nurses, physicians and pharmacists. I hope that sharing these findings will help interdisciplinary health care professionals learn more about bleeding disorders and collaborative partnerships in safe and effective care.

Acknowledgment

Sincere appreciation goes to Bayer for its generous support of this study through an unrestricted grant. I would also like to give heartfelt thanks to the Calgary HTC families and staff – I miss you all very much and you have inspired me to continue my doctoral studies in bleeding disorder care.

* **C-STOP** is an acronym for **C**ommunication, **S**ubjective knowledge, **T**eam roles, **O**bjective knowledge, **P**artnerships. ○

References

1. MacLeod M. "We're it." *Issues and realities in rural nursing practice*. W. Ramp, J. Kulig, I. Townshend & V. McGowan (Ed.,) Health in rural settings: contexts for action (165-178). Lethbridge: University of Lethbridge, 1999.
2. Pritchard, A.M. (2004). *Learning and resource needs and capacities of rural hemophilia health care providers*. Masters Thesis, University of Calgary.
3. Pritchard, A.M., Reimer, M.A., Simonson, K. & Oberle, K. (2006). *Learning and resource needs and capacities assessment of rural hemophilia healthcare providers*. *Australian Journal of Rural Health*, 14(5), 184 -8.
4. Public Health Agency of Canada (2003). *Rural Health*.
5. Taylor G. (1999). *The emerging needs of persons living with haemophilia and HIV/AIDS in British Columbia*. *Haemophilia*, 5(1), 1-8.
6. University of British Columbia, The Institute of Health Promotion Research (2001). *Psychosocial needs assessment of persons with hemophilia and von Willebrand's disease (with or without hepatitis C)*. Montreal: The Canadian Hemophilia Society.

THE BLOOD FACTOR



David Page,
CHS Director of Programs and Public Affairs

In the news

■ CSL Behring and Bayer extend agreement on Helixate FS through 2017

KING OF PRUSSIA, Pa., February 1, 2007 – CSL Behring (formerly ZLB Behring) has announced an agreement with Bayer HealthCare to continue the world-wide supply and distribution of Helixate®FS for the treatment of hemophilia A. Helixate FS is identical to Kogenate®FS except for packaging and distribution. Both are manufactured by Bayer, and both are approved for use by Health Canada. Under the contract extension, Bayer will continue to supply Helixate FS to CSL Behring through 2017.

More than six billion units of Kogenate FS and Helixate FS have been infused over the last nearly two decades.

■ Wyeth latest company to initiate research into longer-lasting factor concentrates

MADISON, N.J., February 5, 2007 – Wyeth Pharmaceuticals has become the latest manufacturer of clotting factor concentrates to announce research into longer-lasting products. Over the last two years, both Bayer and Baxter have initiated research into products with a goal of reducing the frequency of prophylactic or on-demand therapy for hemophilia.

Wyeth and Nautilus Biotech in Evry, France, have announced the signing of a research collaboration and license agreement. The goal is to develop novel recombinant factor IX proteins with an extended half-life for the treatment of hemophilia B. The technology makes minimal and specific changes to amino acid sequences in order to slow the breakdown of the protein in the body.

CHS supports promotion of inhibitor study

An article in the Fall 2006 issue of *Hemophilia Today* described the Hemophilia Inhibitor Genetics Study.

In April 2007 the CHS Blood Safety and Supply Committee Co-chairs, Michael King and Bill Mindell, wrote to Dr. Manuel Carcao, President of the Association of Hemophilia Clinic Directors of Canada, to encourage maximum participation in the study. That letter is re-printed below.



Canadian Hemophilia Society
Help Stop the Bleeding
Société canadienne de l'hémophilie
Arrêtons l'hémorragie

Executive Committee Members/
Membres du comité exécutif

President/Président
Eric Stolte

Past President/
Président sortant
Tom Alloway, Ph.D.

Vice Presidents/Vice-présidents
Mike Beck
Heather Moller
Pam Wilson

Treasurer/Trésorier
Norman Locke

Secretary/Secrétaire
James Kreppner, LL.B

Directors/Directeurs
Steven Bardini
Colleen Barrett
Mylène D'Fana

Dan Doran
Martin Kulezyk
Aline Landry
Bea Bohn-Meyer
Tony Niksic
Dane Pedersen
David Pouliot
Bob Steffens
Tony Tavares
Craig Upshaw
Sandy Watson
Ann Wood

MSAC Representative/
Représentant du CCMS
Bruce Ritchie, MD

Acting Executive Director/
Directeur général par intérim
David Page

Honorary Life Members/
Membres honoraires à vie
Frank Bott
Leni George
Ronald George, Ph.D.
Martin Inwood, MD
James Kreppner, LL.B
John R. (Jack) McDonald, Ph.D.
David Page
John Plater, LL.B
Kenneth E. Poyser, CA
Joyce Rosenthal
Elaine Woloschuk
Durhane Wong-Rieger, Ph.D.

Founding President/
Président-fondateur
Frank L. Schnabel

Affiliation
World Federation of Hemophilia
Fédération mondiale de l'hémophilie

Charitable registration/
Association de bienfaisance
11883 3094 RR 0001

April 3, 2007

Dr. Manuel Carcao
President, Association of Hemophilia Clinic Directors of Canada
Div. of Hematology/Oncology
Hospital for Sick Children
555 University Avenue
Toronto, ON M5G 1X8

Dear Dr. Carcao,

We are writing on behalf of the Canadian Hemophilia Society Blood Safety and Supply Committee to encourage maximum Canadian participation in the *Hemophilia Inhibitor Genetics Study* (HIGS). As you know well, the objective of the HIGS is to determine genetic factors, other than mutations within the factor VIII gene, that are associated with the development of inhibitors in severe hemophilia A. The chief investigator is the renowned Erik Berntorp, MD, PhD, of the Malmö University Hospital, Malmö, Sweden.

With the advent of safe clotting factor concentrates, it is now accepted that inhibitors currently represent the most important complication of hemophilia. As you are well aware, up to 30% of people with severe hemophilia A develop inhibitors at one time or another in their lives. Up to 10% of these patients are affected by a persistent inhibitor which makes treatment very challenging.

Unfortunately, the causes of this serious complication remain unclear. Inhibitors are rare, their causes likely varied. It is only through large, international, multi-centre studies with many participants that the mystery will be unravelled.

As of October 31, 2006, investigators from 63 centres in Europe, North America and Australia have agreed to participate in the HIGS study.

To the best of our knowledge, only three Canadian centres, Hôpital Ste-Justine in Montreal, Hospital for Sick Children in Toronto and the Dr. John Akabutu Comprehensive Centre for Bleeding Disorders in Edmonton, are among the 63 centres that have agreed to participate. Hôpital Ste-Justine, under the leadership of Georges-Etienne Rivard, is one of the six centres around the world to have enrolled 60% of the 130 families currently participating.

As recruitment of families and centres is continuing, we would request that the members of the Association of Hemophilia Clinic Directors of Canada actively promote participation in this study among families with severe hemophilia A and a history of inhibitors. The upcoming meeting, Rendez-vous Québec, would be an excellent opportunity.

For its part, the Canadian Hemophilia Society has publicized the importance of the study in a Fall 2006 article in *Hemophilia Today* (See Vol 41, No 3, page 25) and has agreed to publish this letter in the Spring 2007 issue.

If there is any more the Canadian Hemophilia Society can do to promote this very valuable research with its members, please let us know.

Sincerely,

Michael King, MD
Co-chair, CHS Blood
Safety and Supply Committee

Bill Mindell
Co-chair, CHS Blood
Safety and Supply Committee

■ Bloodbot installed in BBPSP lab

EDMONTON, February 15, 2007 – In February, a unique robotic device was installed in the Blood Borne Pathogens Surveillance Project (BBPSP) laboratory at the University of Alberta. Researchers are using the *bloodbot* to increase the number of blood samples from people with bleeding disorders that can be stored.

The BBPSP consists of a secure collection of blood samples collected through Canadian comprehensive bleeding disorder clinics, with a link to clinical data from the Canadian Hemophilia Assessment and Resource Management System (CHARMS) database. The collection or archive of blood samples is made available to laboratories looking for known and emerging blood-borne pathogens, as well as labs that look for the mutation causing the bleeding disorder, and other genetic changes that affect the bleeding tendency. The collection undergoes regular audits and oversight to assure that it functions in a transparent and responsible manner. See the Summer 2005 issue of *Hemophilia Today* (Vol 40, No 2) for more information on the project.

“We’re very pleased with how the robot is working,” said Dr. Bruce Ritchie, professor in the University of Alberta Division of Hematology and the director of the BBPSP. “Where we could only process about 25 samples a day, with the robot we’re now able to do more than 50 samples, and we hope to increase this to more than 100 in the near future. One of the main things this machine does is eliminate the possibility of human error in some minute, repetitive work.”

The robot does the tedious work of separating the components of the blood samples, registering them according to a barcode reading, and then preparing them to be frozen until researchers can analyze them.

Dr. Ritchie hopes the increased production will allow the BBPSP program to expand its scope and accept blood samples from more people in Canada who require frequent blood transfusions, such as thalassemia and sickle cell anemia patients.

The BBPSP laboratory is sponsored by the Public Health Agency of Canada and the Association of Hemophilia Clinic Directors of Canada. Researchers at the University of Alberta teamed up with Baxter to commission a company in California to create the robot. ○

vCJD risk from plasma-derived factor VIII “is extremely low, but may not be zero”

David Page,

CHS Director of Programs and Public Affairs

In December 2006, the U.S. Food and Drug Administration (FDA) released its risk assessment for transmission of variant Creutzfeldt-Jakob disease (vCJD) by plasma-derived factor VIII products. Results of the risk assessment model suggest that the risk of vCJD infection from plasma-derived FVIII currently manufactured from American plasma... “is extremely low, but may not be zero.”

The FDA risk assessment finds that the risk for users of U.S. plasma-derived concentrates is between 1 in 105,000 and 1 in 9,400,000 per patient per year.

The FDA documents note that donor deferral criteria in place since 1999 have reduced the risk of blood donation by people exposed to vCJD through ingestion of infected beef. As well, manufacturing processes for human plasma-derived FVIII products likely reduce the quantity of vCJD agent, if present, but the level of reduction through manufacturing steps is not precisely known.

The risk analysis concerns only plasma-derived factor VIII products manufactured from American plasma since the introduction of donor deferral criteria for visitors to and residents of the U.K. and France. It does not attempt to calculate risk from products manufactured previously.

There has been no known case of transmission of vCJD by plasma products anywhere in the world. This includes in the U.K. where hundreds of thousands of bovines were stricken with Mad Cow Disease in the 1980s and more than 150 people have died from its human form, vCJD. Four cases of vCJD, however, have been linked to blood transfusion in the U.K. These people contracted the disease after transfusion of red blood cells from donors who were later diagnosed with vCJD.

In a statement issued to the FDA, World Federation of Hemophilia President, Mark Skinner, noted, “There are a number of uncertainties in the risk assessment models and it is important to continue to follow developments closely as more is learned about issues such as prevalence of vCJD, clearance of prions, or other factors.”

He went on to say, “The WFH continues to view both recombinant and plasma-derived products as important treatment options for the global bleeding disorders community. The potential remains for adverse events including inhibitor development or unknown pathogen risk, thus continuous learning and communication are required.”

Canadians with hemophilia A use recombinant factor VIII. These products are not considered to be at risk. A very small number of Canadians, however, have used plasma-derived factor VIII over the last several years. These products were all manufactured from American plasma. In addition, those people with von Willebrand disease who use clotting factor concentrates with factor VIII and von Willebrand factor use products manufactured from American plasma. The risks cited by the FDA apply to these products.

No risk assessment has been completed for plasma-derived factor IX; however, because infectious vCJD prions are concentrated in cryoprecipitate and not in the fraction of plasma used to manufacture factor IX, the risk is thought to be lower.

For more information, including the complete FDA risk analysis, see the CHS Web site (www.hemophilia.ca/en/4.2). ○

“There are a number of uncertainties in the risk assessment models and it is important to continue to follow developments closely as more is learned about issues such as prevalence of vCJD, clearance of prions, or other factors.”

Factor VIII and IX concentrates licensed/distributed in Canada

The products available to treat hemophilia A, hemophilia B, and von Willebrand disease in Canada are in constant evolution. New ones are introduced; older ones are replaced. *Hemophilia Today* has produced this chart to provide some basic information on the recombinant and plasma-derived factor concentrates currently licensed and/or distributed in Canada by Canadian Blood Services and Héma-Québec. For more complete information, see the product monographs that accompany the products. Most of these monographs are available via the Internet.

In the next issue of *Hemophilia Today*, we will publish a summary of the factor concentrates available to treat some of the rare factor deficiencies.

Recombinant factor VIII

Product	Generation*	Albumin in cell culture	Albumin as stabilizer	Viral inactivation	Vial size	Storage	Availability	Comments
Recombinant Manufactured and distributed by Baxter	1	Yes (bovine albumin)	Yes	None	250, 500, 1000 IUs	2-8°C, room temperature for up to 6 months	Licensed, currently being removed from Canadian market	Full-length factor VIII molecule, no von Willebrand factor, human albumin added as stabilizer
Kogenate FS Manufactured and distributed by Bayer	2	Yes (human albumin)	No (stabilized with sucrose)	Solvent detergent: TNBP/ Polysorbate 80	250, 500, 1000 IUs	2-8°C, room temperature for up to 3 months	Licensed, distributed in all provinces	Full-length factor VIII molecule, no von Willebrand factor
Helixate FS Manufactured by Bayer Distributed by CSL Behring	2	Yes (human albumin)	No (stabilized with sucrose)	Solvent detergent: TNBP/ Polysorbate 80	250, 500, 1000 IUs	2-8°C, room temperature for up to 3 months	Licensed, not currently distributed in Canada	Identical to Kogenate FS, Full-length factor VIII molecule, no von Willebrand factor
ReFacto Manufactured and distributed by Wyeth	2	Yes (human albumin)	No (stabilized with sucrose)	Solvent detergent: TNBP/ Triton X 100	250, 500, 1000, 2000 IUs	2-8°C, room temperature for up to 3 months	Licensed, not currently distributed in Canada	B-domain deleted factor VIII molecule, no von Willebrand factor
Advate Manufactured and distributed by Baxter	3	No	No (stabilized with sucrose)	Solvent detergent: Polysorbate 80	250, 500, 1000, 1500, 2000 IUs	2-8°C, room temperature for up to 6 months	Licensed, distributed in all provinces	Full-length factor VIII molecule, no von Willebrand factor

* Generation 1: Human or animal albumin as both nutrient in cell culture and as stabilizer in final bottle.

Generation 2: Human albumin as nutrient in cell culture but not as stabilizer in final bottle.

Generation 3: No human or animal albumin either as nutrient in cell culture or as stabilizer in final bottle.

Plasma-derived factor VIII with/without von Willebrand factor

Product	Plasma source	Fractionation	Viral inactivation	Vial size	Storage	Availability	Comments
Hemofil M Manufactured by Baxter	USA: volunteer, remunerated plasmapheresis donors	Monoclonal antibody affinity and ion exchange chromatography	TNBP/Triton X 100	250, 500, 1000 IUs	2-8°C, room temperature for up to 6 months	Licensed, provided on a case-by-case basis	Albumin added as stabilizer, no von Willebrand factor
Humate P Manufactured by CSL Behring	USA: volunteer, remunerated plasmapheresis donors	Multiple precipitation	Pasteurization at 60°C, 10 hours	- 250 IUs of FVIII (600 IUs of Ristocetin co-factor) - 500 IUs of FVIII (1200 IUs of Ristocetin co-factor) - 1000 IUs of FVIII (2400 IUs of Ristocetin co-factor)	2-8°C, room temperature for up to 6 months	Licensed, distributed in all provinces	Albumin added as stabilizer, contains von Willebrand factor
Wilate Manufactured by Octapharma	USA: volunteer, remunerated plasmapheresis donors	Size exclusion high pressure liquid chromatography	Solvent detergent & dry heat (100°C, 120 minutes)	- 450 IUs of FVIII (400 IUs of VWF) - 900 IUs of FVIII (800 IUs of VWF)	2-8°C	Licensed, not currently available in Canada	Albumin added as stabilizer, contains von Willebrand factor

Recombinant factor IX

Product	Generation*	Albumin in cell culture	Albumin as stabilizer	Viral inactivation	Vial size	Storage	Availability	Comments
Benefix Manufactured and distributed by Wyeth	3	No	No (stabilized with sucrose)	Nano-filtration & solvent detergent (Polysorbate 80)	250, 500, 1000 IUs	2-8°C, room temperature for up to 6 months	Licensed, distributed in all provinces	

* Generation 1: Human or animal albumin as both nutrient in cell culture and as stabilizer in final bottle.

Generation 2: Human albumin as nutrient in cell culture but not as stabilizer in final bottle.

Generation 3: No human or animal albumin either as nutrient in cell culture or as stabilizer in final bottle.

Plasma-derived factor IX

Product	Plasma source	Fractionation	Viral inactivation	Vial size	Storage	Availability	Comments
Immunine Manufactured by Baxter	USA: volunteer, remunerated plasmapheresis donors	Ion exchange and hydrophobic interaction chromatography	Polysorbate 80; Vapour heat, 60°C, 10 hr @ 190 mbar; then 80°C, 1 hr @ 375 mbar	250, 500, 1000 IUs	2-8°C, room temperature for up to 3 months	Licensed, distributed in all provinces	Albumin added as stabilizer
Mononine Manufactured by CSLBehring	USA: volunteer, remunerated plasmapheresis donors	Immunoaffinity chromatography	Sodium thiocyanate & ultrafiltration	500, 1000 IUs	2-8°C, room temperature for up to 1 month	Licensed, currently available in Canada on a case-by-case basis	Albumin added as stabilizer

THE female FACTOR

Patricia Stewart



Symptomatic carriers: Can women and girls really express hemophilia?

Nisa Renault BSc, PhD candidate,
Dalhousie University
Halifax, Nova Scotia

In the Spring 2006 issue of *Hemophilia Today*, JoAnn told the story of her daughter, Olivia, who has severe hemophilia A. Her symptoms were unexpected because, typically, only males express hemophilia A. Most females who inherit one altered hemophilia A gene are asymptomatic carriers. This is the usual pattern because of the genetics of hemophilia A.

Hemophilia A is caused by insufficient activity of the factor VIII protein that is needed for blood clotting. Changes in the structure of the factor VIII protein can affect its activity and lead to hemophilia A. The DNA encoding the factor VIII gene is found on the X-chromosome. Females have 2 X-chromosomes, and males have one X- and one Y- chromosome in each cell. The amount of genetic material on one X-chromosome is just the right amount needed for each cell, so each cell with 2 X-chromosomes needs to inactivate one of them. Genes on the inactivated chromosome are not available for use. X-chromosome inactivation is a normal and necessary process of development. Genes on the X-chromosome are called X-linked genes, and conditions resulting from alterations in those genes are called X-linked conditions. For males to express X-linked conditions like hemophilia, they only need to inherit one altered copy of a gene. Because females have two copies of X-linked genes, they typically need to inherit one altered copy from each parent. Because Olivia only inherited one altered factor VIII gene, she should be an asymptomatic carrier. So what happened? That's where we come in.

Thanks to the enthusiastic participation of Olivia and her extended family, and funding from several institutions

(including the Canadian Hemophilia Society, CIHR, CDHA, IWK, Killam), our research team in Dr. Wenda Greer's lab (Pathology Department, CDHA and Dalhousie University, Halifax, Nova Scotia) has been able to answer a few questions. Females sometimes express X-linked recessive conditions (like hemophilia A, hemophilia B, Duchenne Muscular Dystrophy, colour-blindness) because of unbalanced X-chromosome inactivation. The "choice" of which X gets turned off in each cell has typically been viewed as a random process, like flipping a coin. Heads – the X-chromosome inherited from your father gets turned off, tails – the X from your mother gets turned off. This process is thought to occur near the 8-cell stage of development. So, each embryo flips a coin 8 times to determine which of the two X-chromosomes in each cell will be inactivated. Most get a balanced ratio of 4 heads to 4 tails, but some get 3 heads to 5 tails or even 8 heads to 0 tails. An unbalanced ratio of heads to tails (or father's X off to mother's X off) is only a concern if one of those Xs has an altered gene.

We were able to show that, in hemophilia carriers in Olivia's family, the more cells that contained a functional copy of the factor VIII gene, the higher the factor VIII blood clotting activity. The fewer the cells activating the normal factor VIII gene, the lower the factor VIII activity and the more severe the hemophilia symptoms. For example, in Olivia's sister, half of her cells express the normal factor VIII protein, and half are expressing the altered factor VIII protein. Consequently, she has half the normal factor VIII activity. Fortunately, half the normal factor VIII activity is sufficient to protect her from clinical symptoms of hemophilia. In Olivia, however, most of her cells are expressing the altered factor VIII gene (92%). To continue the analogy, 7 out of 8 coins came up tails! Consequently, she has very low factor VIII activity (<2% normal) and severe symptoms. Many women in Olivia's family have unbalanced ratios (at least 6 of 13 have ratios exceeding 70 to 30). Some of these family members have turned off mostly Xs with altered factor VIII protein and have nearly

normal factor VIII activities. Others have turned off mostly normal Xs and have low factor VIII activities and symptoms of hemophilia. So, the hemophilia symptoms in the carriers in Olivia's family can be explained by unbalanced, or skewed, X-chromosome inactivation. That's one question answered. However, our findings pose a new problem to solve: why do Olivia and many of her female relatives have skewed X-chromosome inactivation?

It is possible that Olivia has skewed X-chromosome inactivation by chance. However, it is unlikely that many women in the same small family would all have skewed X-chromosome inactivation by chance (a probability of less than 5% in our case). We are exploring the possibility that the choice of which X-chromosome



Olivia Craig

to inactivate may not be random, but rather is genetically influenced.

We have several lines of evidence supporting this idea. First, the same skewed X-chromosome inactivation was observed in different types of cells from each family member. Also, high resolution studies revealed no X-chromosome structural abnormalities that can sometimes cause X-chromosome inactivation skewing. Lastly, our review of published studies indicates that X-chromosome inactivation skewing may be more common in the general population than can be explained by random models. We are currently in pursuit of a possible gene influencing the choice of which X-chromosome to inactivate. Such a gene may be influencing X-chromosome inactivation in Olivia's family. If we can find it, we will learn a great deal about X-chromosome inactivation, and could potentially use this information to predict the severity of many X-linked conditions, like hemophilia, in female carriers. Eventually, this information could lead to ways of relieving or preventing symptoms in carriers. ○



Quebec Chapter - Tunisia twinning: January 2007 visit

François Laroche, *President of the CHSQ*

The partnership between the Quebec Chapter (CHSQ) and the Tunisian Hemophilia Association (ATH) produced some memorable moments from January 18 to 25 during the second CHSQ visit to Tunisia. The Quebec delegation was made up of the following members: Dr. Georges-Étienne Rivard, Director of the Hemophilia Treatment Centre (HTC) at Sainte-Justine Hospital in Montreal; Nichan Zourikian, physiotherapist attached to the HTC at Sainte-Justine Hospital; David Page, Vice President, Communications and Public Policy with the World Federation of Hemophilia (WFH); Patricia Stewart, Chair of the International Projects Committee at the CHSQ; and François Laroche, then Vice President of the CHSQ.

On Friday, the day after our arrival, there was a medical symposium entitled: Tunisia-Quebec Hemophilia Day. Under Honorary Patron Her Excellency Mrs. Leila Ben Ali, wife of the President of the Republic of Tunisia, the symposium was officially opened by Mrs. Naziha Cheikh, Secretary of State in the Tunisian Ministry of Public Health, and Mr. Bruno Picard, Canadian Ambassador to Tunisia. The presence of these two dignitaries, and above all the fact that the wife of President Ben Ali had agreed to act as Honorary Patron for the event, drew a large number of reporters.

The main goals of the symposium were as follows:

- Raise public awareness about hemophilia through the media
- Increase the number of cases of hemophilia recorded by raising health professionals' awareness about hemophilia
- Improve care and treatment provided to hemophiliacs through knowledge acquired from the talks, especially those given by hematologists
- Improve care provided to hemophiliacs through the workshop for physiotherapists.

A total of sixty physicians (hematologists, orthopedists, general practitioners, etc.) took part, along with laboratory technicians, biologists, physiotherapists,



An example of synovitis, caused by frequent hemarthroses, in the knee of a young Tunisian with hemophilia.

dentists, and representatives of the blood product supplier (Pasteur Institute), mainly from the four major cities in Tunisia (Tunis, Sousse, Sfax, and Gafsa), in addition to representatives of the two associations. At the end of the day, the participants said they were delighted with the wealth of information and quality of the discussions. From the opening talks to the closing reception, everything went very smoothly, all to the credit of the Organizing Committee. The mere fact of securing the patronage of Mrs. Ben Ali was a great accomplishment by the ATH.

Even though the following day, Saturday, was the Muslim New Year and a national holiday, we were still received at Aziza Othmana Hospital by the Head of the Clinical and Biological Hematology Department, Dr. Balkis Meddeb.

We were able to visit the clinical and biological haematology facilities, the new haematology treatment centre (in preparation), and meet with the hospital director. David Page gave a talk on the various WFH programs, such as the Global Alliance for Progress. In the afternoon, François Laroche gave a talk on strategic considerations in an organization, and Patricia Stewart followed with another talk on basic principles of fundraising.

At the end of the day, the participants said they were delighted with the wealth of information and quality of the discussions.



The opening of the Tunisia-Quebec Hemophilia Day, with representatives of the Tunisian Hemophilia Association, the Tunisian Ministry of Public Health, the Canadian Hemophilia Society (Quebec Chapter) and the Canadian Embassy.

continued from p. 23

On Monday, January 22, we had two activities on the program for which we had to divide the delegation in two. François and David accompanied several doctors and member of the ATH to meet the Minister of Public Health, Dr. Rihad Kechrid. After explaining the situation of

Our general impression of this visit is extremely positive. We once again noted a strong will to improve living conditions for Tunisians with hemophilia on the part of both health professionals and ATH volunteers.

people with hemophilia in Tunisia and discussing with each of the members of the delegation, the Minister made a commitment that “Tunisians with hemophilia should have access to the best treatment available and adequate comprehensive care by the State.” This commitment was certainly one of the high points of our visit, and it is very encouraging for the future of care to those with hemophilia in the country. Physiotherapy was the other item on the day’s agenda, with a theoretical training ses-



Three members of the Tunisian Hemophilia Association brave the Montreal winter cold in March 2007. Left to right: Professor Raouf Hafsia, Vice-president; Islem Nafti, President; and Taoufik Raissi, Secretary-general.

sion in the morning for Tunisian physiotherapists, led by Nichan Zourikian. The afternoon was devoted to case studies with young people with hemophilia who had come to consult the experts with their parents.

Our general impression of this visit is extremely positive. We once again noted a strong will to improve living conditions for Tunisians with hemophilia on the part of both health professionals and ATH volunteers. The impending opening of an official hemophilia treatment centre in the Hematology Department of Aziza

Othmana Hospital will make a tremendous contribution toward this goal. The involvement of a number of hematologists together with volunteers who are dedicated to the cause gives the organization a lot of credibility with government officials. In the ATH there are people with remarkable lobbying skills, and the organization is gradually discovering the almost limitless possibilities this can open up.

At present, 270 people with hemophilia have been registered in Tunisia, out of a population of 10 million (therefore out of a potential total of 1000 people with hemophilia). Of these 270 people with hemophilia, 130 (or 48%) have the severe form of the disease. Since our last visit in 2005, Tunisia’s consumption of factor VIII has increased from 0.15 International Units (IU) per capita of total population to a little more than 0.3 IU/capita. Factor VIII concentrate has tripled from 0.1 IU/capita to 0.3 IU/capita (largely due to a better tendering process resulting in lower prices), whereas the standard for adequate treatment of hemophilia, according to the WFH, is 1.0 IU/capita. Consumption of factor VIII in Canada is 5.0 IU/capita.

Since our last visit in 2005, Tunisia’s consumption of factor VIII has increased from 0.15 International Units (IU) per capita of total population to a little more than 0.3 IU/capita.

The CHSQ is very enthusiastic about the care that can be provided to Tunisians with hemophilia in the future. A very promising solidarity has emerged between the two organizations. Tunisians have a reputation as a warm and welcoming people, a reputation they well deserve. The hospitality we received throughout our long stay was superb, and the CHSQ hopes to be able to extend its partnership with the ATH for several more years. ○



The Quebec, French and Tunisian delegations during the visit to the Aziza Othmana Hospital in Tunis. (Standing): Dr. Georges-Étienne Rivard (hematologist, Montreal), Dr. Sondes Mseddi (hematologist, Sfax), Dr. Balkis Meddeb (hematologist, Tunis), Mr. François Laroche (President of CHSQ), Mr. David Page (Vice-president of WFH), Dr. Viviane Guérin (hematologist, Bordeaux), M. Hédi Moulahi (Directeur-general of the hospital), Dr. Aïcha Hafsia (Honorary President of ATH), Dr. Ségolène Clayssens (hematologist, Toulouse), Mr. Taoufik Raissi (Secretary-general of ATH), Mr. Hamma Amdouni (member of the Board of ATH) et Ms. Nejia Grichi (member of the Board of ATH). (Kneeling): Dr. Emna Gouider-Belhadjali (hematologist, Tunis), Ms. Kaouther Zahra (physiotherapist, Tunis), Ms. Patricia Stewart (Chair of the International Projects Committee of CHSQ) et Mr. Habib Chouikha (member of the Board of ATH).