This issue of *Hemophilia Today* is focused on the difficulties of establishing and maintaining comprehensive care programs for bleeding disorders in Canada, at a time when health care dollars appear unable to cover the increasing costs of health care. In 2002, Canadian hemophilia comprehensive care clinics should be seen as models of how to manage complex patients using expensive and potentially harmful treatments, but the stories in this issue show that health care administrations are cutting randomly, as they scramble to balance budgets. The clinics have worked through the contamination of blood products with HIV and hepatitis C, have ensured that Canadians have access to modern coagulation products, and have taken part in and led the development of prophylaxis and immune tolerance therapies. In these times of financial pressures, it is critical for people with bleeding disorders to continue the fight for sustained, secure comprehensive care. continued on page 8
Welcome to the real new millennium. And with each passing year or decade, it seems to your Editor that time sometimes does not simply creep forward, it feels as if it moves in reverse, creeping backwards like some kind of cosmic ant.

What has brought on this curious feeling of déja vu? Some of the contents of this issue of Hemophilia Today are to blame, particularly those dealing with the major theme, comprehensive care. Canada has surely come a long way since the 1970s, when only two or three comprehensive care clinics existed in Ontario, Quebec and B.C., while the rest of the country waited in breathless anticipation for the benefits of care programmes that addressed all the areas of hemophilia needs—and, as you all know, there are many. But in the 1970s hemophilia care was often scattered and ineffective and sometimes worse than adequate.

We have come a long way since then, but our current fear is that we risk losing much of what we have gained over the past thirty years, and we fear that we may never gain, in some areas of the country, the kind of care that many hemophiliacs enjoy in Canada. For example, the “Profile” in this issue is dedicated to Dr. Mary-Frances Scully, the Medical Director of the St. John’s, Newfoundland Hemophilia Comprehensive Care Clinic. She has accomplished wonderful things in Newfoundland and Labrador, but she has run into a funding wall, and she and her clinic staff now have to go to quite extraordinary lengths to ensure that patients receive adequate follow up care. And if more funding is not forthcoming, many Newfoundland and Labrador hemophiliacs, those who live in the outports and in the far reaches of the western part of the province, may never receive truly adequate care.

In the case of British Columbia, underfunding of the clinic has been chronic. This necessitates the B.C. Chapter having to make funding appeals to local groups such as the Lions Club, various corporations, and even to their own members. This has worked quite well in the past, but the current fiscal climate in B.C. must have everyone worried to some extent. B.C. now faces what Alberta and Ontario have gone through during the past eight years or so.

These are merely two examples that demonstrate how perilous the state of our hemophilia clinics is, and how easy it might be for our care to be compromised. We must be ever vigilant, we must be prepared to face our legislators with cogent and well-thought out arguments if we perceive a threat to the funding of our clinics, and we must be supportive of our clinic staff as they continue to work to provide us with the best possible care—which includes keeping the clinic functioning at a truly comprehensive level through adequate government funding. The support of the hemophilia community is essential, for if government representatives or ministers of the Crown perceive the hemophilia community as unsupported of their clinics, they will surely conclude that these clinics are of little real value. We know that is not the case. And we know that we cannot afford to sit back and simply observe that cosmic ant push our care back into the horrid days of Emergency Room visits and constant hospitalizations.

(Please send any comments raised by this Editorial, or any other article, to The Editor, Hemophilia Today, 1409B 4 Street N.W., Calgary, AB T2M 2Y8. E-mail your comments to bisaac@cadvision.com or fax them to (403) 282-3295.)
Vigilance and Action are Required

The focus of this issue is comprehensive care—the idea that people with bleeding disorders are best served by a system of care that provides "one-stop shopping", not only for the bleeding disorder itself, but also for all the associated medical and psychosocial issues that affect the person living with the bleeding disorder and his or her family.

Comprehensive care is a venerable concept with a lengthy history in Canada and abroad. Yet it remains imperfectly implemented in some parts of the country. And in some of the places where it has been most fully developed, budgetary cutbacks are leading to its deterioration. These developments alarm us because the Canadian Hemophilia Society believes that high-quality comprehensive care programmes are the best way to look after the health care needs of people with inherited bleeding disorders. However, I believe that it is useful to view our current situation in the context of the broader discussion about the future of the health care system that is currently taking place in this country.

I recently attended a Conference on the Future of Health Care in Canada sponsored by the McGill Institute for the Study of Canada. As many of you know, Roy Romanow, the former Premier of Saskatchewan, is heading a Royal Commission on the Future of Health Care in Canada. His eventual recommendations, if implemented, will likely have a very significant effect on the future of comprehensive care in Canada. In his preliminary report, Mr. Romanow summarized four main perspectives on reforms to the health-care system:

**More public investment.** Advocates of this position believe that the current system is sound, but more public money is needed.

**Shared costs and responsibilities.** People with this view believe that getting more money for the system by increasing, or even maintaining, current levels of taxation reduces Canada's global competitiveness. They advocate instituting user fees or insurance premiums so that individual users pay part of the cost of their health care.

**Increased private alternatives.** People who favour this approach advocate creating parallel public and private health systems. Different versions of this perspective propose a variety of funding alternatives ranging from 100% public financing for both systems through 100% consumer funding for the private system.

**Reorganized service delivery.** Advocates of this position believe that the traditional fee-for-service model for paying doctors is outdated and propose that physicians be salaried. Some variants of this proposal involve creating medical-practice groups similar to the health maintenance organizations in the United States. Consumers would register with a practice group in their area and would then be required to use that practice group's general practitioners and affiliated specialists for all or most of their medical care.

As Mr. Romanow notes in his preliminary report, these four perspectives are neither exhaustive, nor are they necessarily mutually exclusive. Some people propose a combination of two or more of these approaches. Speakers at the McGill Conference represented a wide range of views on these issues.

One concern that we hear frequently concerns the "sustainability" of Canada's health-care system. Costs are rising, and politicians and others worry about whether governments will be able to continue financing our health-care system. Two radically different views of sustainability were elaborated at the conference.

One group of speakers noted that the percentage of Canada's gross national product (the total value of all goods and services that the country produces) that we spend on health care has remained approximately constant since Medicare was first introduced 30 years ago. Costs have increased, but so has Canada's national wealth. In addition, when it comes to considering how much Canada can afford to spend on health care, it's important to consider how the proportion of our gross national product that we invest in health care compares to the proportions of their national wealth that our trading partners and competitors invest. These days we hear a lot about the supposed efficiency of private health care in comparison to public health care; and the American health care system is often held up as a model of these efficiencies. Thus, I was astonished to learn that the Americans spend a larger proportion of their gross national product on health care than any other country in the world! And this is true despite the fact that millions of Americans who cannot afford private health insurance are deprived of access to services that we and people in other developed countries take for granted. When we compare the amount of money that we spend on health care in relation to our other trading partners, Canada is "in the middle of the pack" among other developed Western nations. In comparison to them, we spend neither an exceptionally large, nor an exceptionally small, proportion of our national wealth on health care.

Other speakers noted that in recent years Canadians have complained about high levels of taxation, particularly in comparison to tax rates in the United States; and the federal and many provincial governments have responded by cutting taxes. Speakers who maintained that Canada's present health-care system is financially unsustainable presumed that Canadians will demand additional tax cuts. In that event, governments will have to reduce the proportion of public money spent on health care in order to fund other government services, such as education. If we want further tax reductions, we will have to pay user fees, health-insurance premiums, or both.

During his speech at the McGill Conference, Mr. Romanow stressed that, before determining his final recommendations, he intends to consult ordinary Canadians about the kind of health care system they want to see in the future and how they want to pay for it. These consultations will involve a series of public hearings that will be held across the country this spring and early summer. Because changes in the way health care is financed in Canada are likely to have a profound influence on the care available to people with inherited bleeding disorders, the Canadian Hemophilia Society has submitted a brief to the Royal Commission in which we explain what comprehensive care is and why its maintenance and further development are so important and why it is crucial for factor products to continue to be provided at no cost to consumers with bleeding disorders. In addition, the CHS and our chapters are in contact with the federal and provincial ministries of health and with the hospitals in which comprehensive care programmes are located, especially those where cutbacks are occurring. All these efforts are aimed at promoting the availability of the best possible health care for all Canadians with inherited bleeding disorders.

I urge you to inform yourselves fully about the issues involved in promoting the best standard of care for people with bleeding disorders and in reforming the Canadian health care system in ways that will ensure the best possible medical care for all Canadians. Vigilance and action are required to maintain and enhance the health care system on which we, as people with bleeding disorders, depend and which is a source of pride for all Canadians.
THE CANADIAN RED CROSS SETTLEMENT

HOW DO I KNOW IF I AM ELIGIBLE?

– As a primarily infected claimant
If you received blood anywhere in Canada prior to January 1, 1986 and between July 1, 1990 and September 28, 1998 inclusively, and this blood was contaminated with the Hepatitis C Virus, you may be eligible to receive compensation under the Canadian Red Cross Settlement.

– As a secondarily infected claimant
As well, if you were infected with HCV through contact with a person who received blood, as described above, you may also be eligible.

– As a family member of infected persons
All living spouses, children, parents, siblings, grandparents and grandchildren of primarily infected claimants may be eligible.

In order to determine your eligibility, you may refer to the eligibility criteria which are clearly defined in the Distribution Protocol document, accessible via KPMG website at http://www.kpmg.ca/microsite/hepatitisc/english/distribution.html.

WHO IS KPMG AND WHAT IS KPMG’S ROLE?

KPMG, an accounting, tax and financial advisory firm, has been appointed claims administrator pursuant to the Settlement Agreement approved in the Pre-1986, Post-1990 Hepatitis C claims against the Canadian Red Cross Society (“CRCS”). KPMG will receive and process all claims in this matter as well as issue payments to accepted claimants.

A settlement has also been reached with the Province of British Columbia for persons who were infected with the Hepatitis C Virus (“HCV”) as a result of tainted blood received in this Province. KPMG Inc. has also been appointed claims administrator pursuant to this particular settlement.

MAILING ADDRESS

KPMG Inc.
2000 McGill College Avenue
Suite 1900
Montreal (Quebec)
H3A 3H8
Attention: Claims Administrator - Hepatitis C

TELEPHONE NUMBER

1-888-840-5764 (1-888-840-kpmg)

ELECTRONIC MAIL

HepatitisC@kpmg.ca
<mailto:HepatitisC@kpmg.ca>

CANADIAN BLOOD SERVICES,
HÉMA-QuÉBEC

NEW CONTRACTS FOR FRACTIONATION AND COMMERCIAL PRODUCTS IN 2003

James Kreppner, Chairperson,
Blood Safety Committee

The Canadian Blood Services and Héma-Québec perform the essential service of collecting blood throughout Canada. They then work in partnership with commercial companies to have the plasma fractionated into its various components, which are then returned to Canada to be used for a wide array of medical treatment options. The two blood services also purchase commercial products (some of which are produced by recombinant cell cultures, and some of which are plasma derived).

The current contracts are now expiring, and like any good purchaser, the two blood services are weighing their options as to which provider can give the best deal to Canadians with respect to these contracts. As part of this process, the blood services have sent out a Request for Proposal (RFP) to the various companies that have the potential to fractionate Canadian plasma, and/or sell to Canada the various blood products that we need. A number of bids have now been submitted, and at the time of this writing, a process is under way to evaluate these voluminous bids so as to compile a shortlist of potential suppliers. To assist in this process, CBS has created a Selection Advisory Committee (SAC). The committee consists of a representative of the Association of Hemophilia Clinic Directors of Canada, CBS and Héma-Québec staff, and representatives of two of the heaviest users of the blood system with respect to commercial products, namely the immunodeficiency community, and the bleeding disorder community—CHS. As the CHS representative, I am constrained by confidentiality agreements from speaking in detail about this process. I can, however, report that it is a fair and tremendously detailed process with many factors going into any final determination of the final product choice and mix. The blood services understand that they are here to provide service to those in need, and they have shown a repeated desire to take into account the desires of the communities they serve. Once the decision as to the successful candidate(s) is made, contractual negotiations will continue for the next year. It is expected that new contracts will be in place by the end of next year.

The community should know that CHS is committed to the principles that only the safest and most efficacious products should be chosen, and that the contracts should also leave some room for product choice on the part of consumers.

With respect to the fractionation contract, the CHS has noted that at the present time, Factor VIII and IX are not being processed from our Canadian plasma, and these plasma fractions are being wasted. This is a tragedy, as there are many people in the world who would gladly infuse Canadian plasma derived products. We have been strongly recommending that FVIII and IX plasma derived products be manufactured from this plasma, and provided to these less fortunate countries on a cost-recovery basis. On this last issue, there may be technical problems, and the CBS and the bidders will have to investigate whether this is a workable option.
KOGENATE FS SHORTAGE UPDATE

CONSERVATION GUIDELINES RELAXED

James Kreppner, Chairperson, CHS Blood Safety Committee

As you may recall from the last newsletter and a previous letter from CHS President, Tom Alloway, Bayer Inc. had initially predicted a time line for resolving its production problems, indicating it would be able to resume normal deliveries in the third quarter of this year. To date, Bayer’s progress has appeared to match its initially predicted time line. In a January 24 letter, Bayer indicated to the bleeding disorders community that it had “made significant progress meeting the commitments made in response to FDA inspection observations”. The company was also pleased to announce that it could now predict releases of 1000 IU vials of Kogenate FS®, and that it was on schedule for normal production levels for the summer of 2002.

While at the current time the worldwide production levels of Kogenate FS are still below normal, it is nevertheless encouraging that consumers can now rely upon Bayer’s predictions with respect to at least one product size. Moreover, if one looks only at the Canadian situation, Bayer has managed to provide the Canadian Blood Services (CBS) and Héma-Québec with the needed levels of product. It now appears that there will be sufficient product on hand at the CBS to meet Canadian needs into the 3rd quarter of 2002. Moreover, the situation is expected to improve even further as we move into the 4th quarter of 2002. It should be noted that these are Special Access Program (SAP) products, as Health Canada has not yet issued a license for the manufacturing facility in Berkeley, which had experienced some changes as part of Bayer’s re-engineering process. Health Canada has now inspected that plant, and hopefully will be in a position to issue a license in the not-too-distant future. If this occurs, it is likely that the status of the product produced by this plant will shift from SAP to normal licensure.

Due to these developments, the Blood Safety Committee of the CHS recently recommended that the treatment guidelines proposed by CHS be relaxed so as to enable the use of recombinant factor VIII for elective surgery that may have been delayed due to this shortage. This recommendation has now been endorsed by the Executive Committee. Similar positions have been taken by the CHS Medical and Scientific Advisory Committee and the Association of Hemophilia Clinic Directors of Canada. If the progress continues apace, the committee will examine recommendations on the lifting of some of the other conservation measures as well. By the time you read this article, hopefully these will have been removed. You may wish to check the CHS website (www.hemophilia.ca) to see if this is the case.

Elective surgery was the first area addressed as it was felt that the most significant impact of this shortage has been on those individuals suffering from various medical issues as they delayed surgery until the supply situation stabilized. Until the Bayer product is licensed and there is an even greater sense of confidence, CHS will continue to monitor this situation closely through biweekly teleconferences with the CBS. We hope to be able to report further good news in the next edition of Hemophilia Today.

TRAVEL ADVISORY

FLYING WITH FACTOR CONCENTRATES?

Since the terrorist attacks of September 11, several people carrying factor concentrates have experienced problems at security checkpoints in North America. In one incident involving a Canadian at the Ottawa Airport, the traveler was forced to remove the caps on the vials of both the factor VIII concentrate and the sterile water and smell them! This occurred despite the fact that the person carried a letter from his doctor certifying his condition, the need to carry blood products while traveling and providing the phone number of the doctor.

While these incidents are isolated, it now seems advisable to carry more explicit information when traveling. A letter from your doctor should include:
• information about your medical condition
• the fact that you need to carry factor concentrates when traveling
• the number of vials
• the lot numbers
• instructions that the vials should not be opened for any reason
• how to contact your doctor.

Travelers carrying needles in their hand luggage should also have the factor concentrates with them. This explains the purpose of the needles. Keeping factor concentrates in your hand luggage makes sense anyway. Checked baggage can be lost and you would be separated from important medication.

An additional precaution is to contact the airline several days before departure to find out their policy on transporting needles and medication.

The Association of Hemophilia Clinic Directors of Canada has prepared a letter to be used as a template by physicians.
The goal of the Care until Cure Research Program is to provide funding for research aimed at improving the quality of life of persons with inherited bleeding disorders. The CHS and Wyeth Genetics Institute are pleased to announce the 2001 grant recipients:

**Recipient:** Dr. Georges-Etienne Rivard  
**Co-Investigators:** Dr. Rochelle Winikoff, Dr. Claire Infante-Rivard  
**Project Title:** A Cohort Study on the Risk of Cancer Associated with Radioactive Synovectomy

**Recipient:** Dr. David Lillicrap  
**Co-Investigators:** Dr. Man-Chiu Poon, Dr. Georges-Etienne Rivard  
**Project Title:** Aminoglycoside Treatment of Severe Hemophilia

**Recipient:** Jenny Aikenhead, P.T.  
**Co-Investigator:** Jane Hagel, P.T.  
**Project Title:** Proprioceptive and Balance Training in Severe Hemophilia

**Recipient:** Dr. Manuel Carcao  
**Co-Investigators:** Dr. Brian Feldman, Dr. Victor Blanchette, Dr. Paul Oh, Dr. Paul Babyn, Dr. Douglas Hedden  
**Project Title:** A Prospective, Randomized Trial to Compare Two Regimes of Prophylaxis in Older Boys with Severe Hemophilia A (2nd Year)

### NEW CHS STAFF ON BOARD

The National Office is pleased to announce that David Page has been appointed National Blood Safety Coordinator. David will also be responsible for coordinating specific programs including the web site, *Hemophilia Today* and CHS Research Programs. We are confident that the CHS will benefit greatly from David’s experience and expertise. Clare Cecchini, who has steered *Hemophilia Today* from one comma to the next over the past ten or so years, will be concentrating on coordinating other projects, while remaining on *Hemophilia Today*’s Editorial Committee. Speaking as the Editor, I find it easy to say that Clare’s steady contributions and always wise comments have made the newsletter what it is today.

### CHS SCHOLARSHIP AND BURSARY PROGRAM

Applications for the 2002 Scholarship and Bursary Program are available at chapters, clinics or through the National Office. CHS provides 1 scholarship, 1 bursary and 1 mature student bursary in the amount of $4000 each. This program is supported through an educational grant from Baxter BioScience. The deadline for submitting applications this year is April 30, 2002. Recipients will be notified in June. For further information please contact the CHS at 1-800-668-2686.

### NOTICE

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

Saturday, June 8th, 2002 9:00 a.m. at the Delta Bessborough in Saskatoon, Saskatchewan

1. To receive the report of the Nominating Committee  
2. To acknowledge the designated Directors of each Chapter  
3. To elect and acknowledge the Directors at Large for 2002-2003 (Please note that only the designated directors are allowed to vote)  
4. To receive the audited financial statements of the Canadian Hemophilia Society for the year ended December 31, 2001  
5. To appoint an auditor for the ensuing year  
6. To transact such other business as may properly come before this Annual General Meeting of the members of the Canadian Hemophilia Society

Janice Aull  
Secretary

### CONGRATULATIONS TO OUR AWARDS RECIPIENTS

#### AWARD OF APPRECIATION –

To honour an individual who has demonstrated outstanding service to the care of persons with hemophilia or related bleeding disorders.

**Recipient in 2000 – Elizabeth-Ann (Betty-Ann) Paradis**

Betty-Ann has been working in hemophilia care for more than 20 years and has been active in hemophilia organizations at the regional, provincial, national and international levels for almost as long. She is currently President of the Canadian Association of Nurses in Hemophilia Care. People say, “Betty-Ann cares for each individual and has demonstrated outstanding service to the care of persons with hemophilia and their immediate families over and above her responsibilities as a member of the Hemophilia Care Team.”

#### PIERRE LATREILLE AWARD –

To honour staff from the national, provincial or regional offices who have contributed in a special way. The award is named for Pierre Latreille, a much-loved CHS colleague until his untimely death in 1999.

**Recipient in 2000 – Hélène Bourgaize**

Hélène is the first recipient of the Pierre Latreille Award. She began her career with CHS in 1985. Over the years, Hélène has become the pivotal point for volunteers. She makes volunteers feel that their contribution is important. Her ability to organize meetings is a sign of her professionalism. Some have said her award should really be a military citation. It is a testament to Hélène’s humanity and determination that she stuck it out with the organization and its members through some very difficult years. Her response to these events was a model of grace under pressure.
DR. CECIL HARRIS AWARD –
To honour distinguished contributions in the areas of hemophilia research or the advancement of care of patients with hemophilia or related bleeding disorders.

Recipient in 2000 –
Dr. Gershon (Jerry) Growe

Jerry became involved in hemophilia care in British Columbia at the beginning of his career and introduced the Home Treatment Programme at Vancouver General in 1971, one of the first of its kind. He went on to become Chair of the CHS Medical and Scientific Advisory Committee. He currently chairs the CHS Research Grants Review Committee, which evaluates research and fellowship applications. Jerry says that “…the affiliation between the CHS and the clinics has been a model for collaborative health care delivery and a great opportunity for those of us who enjoy the experience of team effort.”

CHAPTER RECOGNITION AWARD –
To recognize a chapter/region for an achievement in a particular area during the past year.

Recipient in 2000 –
Saskatchewan Chapter for Advocacy

Great efforts were made by Eric Stolte, Chapter President, and other dedicated volunteers to advocate on behalf of the re-establishment of a hemophilia treatment centre in Saskatchewan. Preparation of a proposal, telephone calls, letter writing and personal meetings were all part of the advocacy. Those efforts culminated in the Saskatchewan government’s decision to create and fund the clinic.

AWARDS FOR 2001
The Nomination Package for the 2001 National Awards is available through chapters, clinics or the National Office. We encourage you to take advantage of this opportunity to recognize and honour the volunteers, staff and health care providers who have dedicated their time and energy to improving the quality of life for people with hemophilia and other inherited bleeding disorders. The deadline to submit nominations is June 30, 2002. Please help us to make the Awards Program, chaired by Mr. Frank Bott, a success this year!
Comprehensive hemophilia clinics in Canada have a long and successful history, going back to the first hemophilia clinic in Montreal in 1969, through to the establishment of a comprehensive care program in Saskatoon in 2001. During this time, an effective networked infrastructure of organized comprehensive care clinics has evolved. This network has proved uniquely successful in the management of bleeding disorders, as exemplified during the recent and prolonged shortage of factor VIII. Product use was smoothly managed without major hardship to a single person with hemophilia A. Comprehensive hemophilia clinics clearly play a unique role, not just in the effective management of people with bleeding disorders, but also in the management of effective coagulation product use in Canada.

Comprehensive care programs clearly work to keep people with bleeding disorders healthy and productive. In an article entitled *The Comfort Zone - Hemophilia Treatment Centres In Canada*, Karen Creighton details the complex support needed by people with bleeding disorders, which is only possible in a comprehensive care program. This support is an effective low-tech, outpatient activity, which may not have the “glitz” of organ transplant, or trauma medicine, but is extremely effective at maintaining healthy, productive, tax-paying people. Patricia Stewart, in her article *Suggestions on the Management of Women with Bleeding Disorders*, describes the collaborative effort to create standards and treat women with these problems. She also describes how Toronto Sick Kids and Ste-Justine hospitals have successfully translated their expertise in treatment of bleeding disorders into development of centres of excellence.

Provincial support for comprehensive care programs, however, has been spotty, despite the proven success of these programs. Cyril DuBourdieu in *Newfoundland and Labrador: Difficulties in Getting Comprehensive Care Standards Recognized* describes the battle his chapter and clinic have fought to obtain adequate resources, and Pam Wilton in *The Day Ontario’s Health Care Restructuring Committee Came to Town* describes the “restructuring” faced by the South West Ontario Region Hemophilia Program. In *The Hemophilia Centre of Eastern Quebec*, a similar attack on comprehensive care at l’Hôpital Saint-Sacrement is described. Finally, Eric Stolte, in *A Long and Hard Victory Won*, tells the story of the 30-year fight to obtain comprehensive care in Saskatchewan. It is clear that the key to fighting off these assaults is the support of the regional and national societies. As Jane Bishop in Toronto, Hillary Rudd in Vancouver, Kelly Steiss in London and François Laroche in Quebec City explain, there are a variety of models of how the local society can work with and support its clinic. In Toronto, the Toronto Central Ontario Region provided the original funding to set up their clinic, and have volunteers at each clinic to help with education and support. In Vancouver, the Society manages provincial hemophilia resources, while in London, the Program Advisory Council meets with and lobbies on behalf of the clinic. In Quebec, the Chapter funds an annual meeting of the five care teams. All of these models work—the critical thing is that the local society continue to support the clinics against the restructuring onslaught.

In the care of bleeding disorders, as in much of medicine, we are victims of our success. At every level of medicine, there are new treatments that work well, that are expensive, and that the funding agencies do not know how to cope with. Restructuring has been a common response. The Romanow Commission is now touring Canada, looking for a more fundamental structural change. Tom Alloway, in his President’s Report, describes his views on the Romanow Commission on the Future of Health Care in Canada, something in which we must all take an interest. It is critical that comprehensive care of bleeding disorder patients is seen as a model of managed care, and that it does not become a casualty of cost saving strategies.

Perception is everything. Health care administrators must be made to see comprehensive care of bleeding disorders for what it is—an efficient, effective, easily studied strategy to provide health care to people with complex medical problems. I believe it is a model of how to make efficient use of limited health care resources in the setting of financial constraint. Support for this system is key to its survival. We mustn’t become complacent.
HEMOPHILIA COMPREHENSIVE CARE

A SET OF NATIONAL STANDARDS

Pam Wilton, R.N., CHS Vice President

In the spring of 1998 a consensus conference on Standards of Care of Hemophilia in Canada was held in Winnipeg, Manitoba. This conference occurred on the 20th anniversary of the first Canadian conference on hemophilia comprehensive care and has come to be known as “Winnipeg II.”

The goal of the conference was: To produce a living document, which will be continuously reviewed and updated, and which will set out the principles for the establishment of interdisciplinary standards for the comprehensive care of people with hemophilia and related diseases in Canada.

Conference participants included a broad representation from CHS, most members of the Association of Hemophilia Clinic Directors of Canada (AHCDC), nurses, physiotherapists, social workers, as well as representatives of the funders of the blood system, Health Canada and the pharmaceutical companies that provide products used to treat congenital bleeding disorders.

The group agreed on the following revised definition: Hemophilia Comprehensive Care is a health care delivery system in which individuals and families with hereditary and related acquired bleeding disorders (such as inhibitor autoantibodies to coagulation factors) receive regular preventative and therapeutic input from members of a team of specialists who represent the many medical and allied health disciplines relevant to the management of these complex disorders.

This team supervises the provision of optimal active and prophylactic hemostatic therapy, regularly assesses patients for physical and psychosocial complications,

Hemophilia in the Days before Modern Treatment

2000 years ago
Hemophilia was mentioned in the Talmud in ancient Israel. If two brothers died after circumcision, the family was told not to circumcise any other boys.

Late 1800s
Hemophilia was long known as the Royal Disease because the sons and grandsons of Queen Victoria of England were afflicted, as were a number of other royal families in Europe.

Early 1900s
Ninety percent of hemophiliacs died before the age of 20. Treatment was almost non-existent and even diagnosis of the disease was rare. Blood transfusion only became a regular part of medicine during the First World War, 1914-18.

The best known member of the European royal families with hemophilia was the tsarevich Alexei, heir to the throne of Russia. His physician, the mad monk, Rasputin, cared for the tsarevich by using hypnosis to ease his pain and help stop the bleeding.

1930s
Seventy percent of hemophiliacs died before the age of 20. Ice packs, elevation, pressure and blood transfusions were the only treatments available. The small amount of coagulation factor in a pint of blood limited the effectiveness of transfusions even for those who could afford them.

1940s
Two distinct bleeding disorders were recognized: hemophilia, transmitted by mothers but affecting only the sons; and von Willebrand Disease, discovered affecting girls in a family on a remote island in Finland. Some of the girls bled to death at the time of their first menstruations.

Early 1950s

Fresh frozen plasma was used to control bleeding episodes. Controlling a serious joint bleed often required 20 or 30 bottles of plasma over a four- or five-day period.

Snake venom injections were given for their clotting effect. However, with repeated use, resistance developed.

Eighty percent of hemophiliacs lived to adulthood, but many were crippled by the consequences of repeated joint bleeds.

Late 1950s
The first successful appendectomy on a hemophiliac in North America was performed in 1956 on a 7-year-old Montreal boy.

Hemophiliacs could require up to 100 transfusions of fresh frozen plasma in one month. This meant they had to call on family members, friends and colleagues to donate blood. Special calls for blood donors went out on the radio and in the newspapers. The hemophilia society took part in blood donor clinics regularly.

A “cure” for hemophilia was found in 1957 by a young Louisiana physician with hemophilia. He found that he bled less if he ate peanut butter. The treatment evolved into a peanut powder that was added to milk shakes, three times a day. It turned out that peanuts contain vitamin K, a coagulant; however the amount needed to make any difference to a hemophiliac was astronomical. So the peanut cure was abandoned.

Research was being done into concentrated factors at this time. Porcine factor VIII was developed. However, it could only be used in a dire emergency as the body rejected it on the second infusion. In 1957, a dried antihemophilic factor (AHF) was developed in England but did not come into widespread use.

1963
Dr. Judith Pool from Cornell University discovered that cryoprecipitate, a sludge that forms on the surface of plasma as it thaws, was rich in factor VIII. This was the beginning of effective factor replacement therapy.

Late 1960s
Dried factor VIII and IX concentrates were developed.
and educates and counsels patients, their families, and society at large regarding bleeding disorders and their complications. Team members also share their expertise in disorders of blood coagulation by providing assistance as necessary to the administrators and regulators of the blood system. They also advance the scientific basis of the care of people with hemophilia and related diseases, through basic and clinical research.

The Conference’s primary recommendation was that hemophilia comprehensive care should be delivered according to a set of uniform national standards and that those standards should be needs based, data-driven and supported by evidence of effectiveness. The standards will be implemented locally; therefore the delivery system must be flexible and adaptable, without jeopardizing quality of care. The scope of services provided by hemophilia treatment centres includes the following:

• Expert clinical and laboratory diagnosis of congenital and complex acquired bleeding disorders
• Expert multidisciplinary management of people with these disorders
• Skilled counseling for individuals and families with these disorders
• Continuing education of patients and families, emphasizing self-treatment at home, at work and at school
• Maintenance of confidential clinical records
• Participation in the CHARMS information management system, to optimize patient care, communication, and accountability
• Educational resources and outreach to local and regional care providers
• Education for medical and other professional trainees
• Participation in collaborative clinical research and surveillance initiatives
• Participation in professional associations, and through these in the generation of guidelines.

The Ministry of Health in each province must provide the necessary resources and the host hospital must be committed to providing the service. The professionals who need to be part of the core team of service providers include a physician (with competence in hemostasis and clinical bleeding disorders), nurse co-coordinator, physiotherapist and social worker. The extended team should include an occupational therapist, rheumatologist, orthopedic surgeon, dentist, infectious disease specialist, obstetrician/gynecologist, hepatologist, geneticist, psychologist, and data manager. Laboratory expertise in coagulation and blood banking is also necessary. Specialists in the field of pain management, vocational rehabilitation, nutrition and child life may be needed for referrals.

Canada is a large country with a relatively small and dispersed population. There are considerable regional differences in needs and in funding of health services. Therefore, many Chapters of CHS have used the recommendations from the Winnipeg II Conference, as well as the CHS document Vision of Comprehensive Care 1995 to tailor proposals to government for improved comprehensive care. These proposals reflect the model of delivery which best meets the needs of their regions as well as the funding system in each province.

### Comprehensive Hemophilia Care in Canada – At a Glance

| First centre established | 1969 (Montreal) |
| Latest centre established | 2001 (Saskatchewan) |
| **Number of centres** | 24 centres, including |
| • 5 pediatric centres |
| • 18 adult or combined pediatric/adult centres |
| • 1 centre for the treatment of inhibitors |
| **Location** | Every province except PEI |
| **Bleeding disorders treated** | Hemophilia A |
| | Hemophilia B |
| | Other factor deficiencies |
| | Von Willebrand Disease |
| | Platelet function disorders |
| **Number of bleeding disorder patients registered** | 5500 to 6000 |
| **Clotting disorders treated (in some clinics)** | Thrombophilia |
| **Core resources** | Hematologist |
| | Nurse coordinator |
| | Physiotherapist |
| | Social worker |
| **Extended resources** | Occupational therapist |
| | Rheumatologist |
| | Orthopedic surgeon |
| | Dentist |
| | Infectious disease specialist |
| | Obstetrician/gynecologist |
| | Hepatologist |
| | Geneticist |
| | Psychologist |
| | Data manager |
| **Laboratory expertise in coagulation and blood banking** |

### BENEFITS OF COMPREHENSIVE CARE (COMPARED TO BEFORE IMPLEMENTATION)

- 40% reduction in mortality
- 88% fewer hospital admissions
- 73% fewer days lost from work and school
- 75% reduction in hospital and physician costs
The Comfort Zone -
Hemophilia Treatment Centres In Canada

Like many parents, my life is comforted by the support of our Hemophilia Treatment Centre at the Hospital for Sick Children in Toronto. Perhaps, too, like many parents of children with bleeding disorders, I have become too comfortable with the availability of specialized medical care and support. After all, I never fought for the establishment of “comprehensive care.”

Hemophilia Treatment Centres (HTCs) were part of the medical landscape we came to know when we became parents of children with hemophilia in the early 1990s.

Over time I have learned about the struggle to establish HTCs, and that securing funding for them has always been a battle. I have also learned—as have so many of us—that we desperately need our HTC. I would feel terribly vulnerable in a society where the medical care, support and educational benefits of HTCs were marginalized in a decentralized system.

It is a proven fact: the well-being of children and adults with bleeding disorders is best achieved through comprehensive care. Not all families will need the care and support described by the people and families in this article. We all, however, need the information and prevention skills found in our HTCs and the benefits of specialized medical care to ensure our kids lead full and happy lives. By hearing about other families’ trials and tribulations, you may also discover your clinic team has more to offer you and your child than you realized.

These four stories, featuring exceptional health care providers and courageous people with bleeding disorders facing enormous challenges, show the importance of comprehensive care in well-funded Hemophilia Treatment Centres. There are thousands of other stories to be told.

Comprehensive care is a comfort to us all. It must not, however, be taken for granted as something we are used to and as something we expect to always be there. It is both essential and precious. This means we have to be more vigilant in understanding the function a HTC serves, the specialization that has been developed within each Canadian HTC and our joint responsibility to ensure we protect and foster these incredibly precious resources.

Thank you to all the people who shared their stories and insights with me. I am only sorry I can’t write about all of you. As always, be safe and keep in touch.

Karen Creighton
creighton@idirect.com

The Nurse Coordinator –
At the Heart of the HTC

Ann Marie Stain is the Nurse Coordinator in the HTC at the Hospital For Sick Children in Toronto. This HTC is the largest pediatric clinic in Canada, providing care to 180 children with hemophilia, and 350 children with von Willebrand Disease. The clinic also provides care for kids with other factor deficiencies and platelet function disorders. You might think this caseload would place Ann Marie beyond of the reach of families. It is just the opposite. Ann Marie’s availability to the kids and their families is key to the support she renders daily in her capacity as liaison between families and members of the care team.

Whether in Toronto, Halifax or Vancouver, the role of the Nurse Coordinator is based on trust. They can be reached by pager or phone, they know our kids and they know us, too. They have a keen sense of each child, the child’s family dynamic, and each child’s medical case history. As Mike O’Grady, father of M.J., a child with hemophilia, puts it, “Ann Marie gives us peace of mind. She provides us with the information, guidance and support we need to handle the unexpected. She is our liaison, keeping on top of everything M.J. needs and arranging everything for him at the hospital. We have relied upon her to help us manage some difficult bleeds and unusual situations including M.J.’s tonsillectomy and an allergic reaction to latex. It was a frightening experience and having Ann Marie at our side meant a great deal, especially as we were trying to determine what was happening to our son. Little did we know, the swelling was an anaphylactic reaction to the latex gloves I wore to access his port. In calmer moments, Ann Marie taught us how to access and take care of M.J.’s port. Later on, she trained us to access his veins. We review his diary together and we watch out for new patterns or difficulties in resolving bleeds. She has been both an educator and friend to all of us.”

Nurse coordinators serve as educators. Nurses visit schools, family doctors, families, day care centres and community hospitals to educate about hemophilia. Some clinics provide outreach services through traveling clinics to enable greater access to comprehensive care. In Newfoundland and Labrador, where there is believed to be the highest incidence of people with mild hemophilia in the world, and distances are great, outreach services are vital. Nurses also keep lines of communication open between family doctors, pediatricians, hematologists, clinic staff and patients. They provide consistency and accountability to the patient and family.

In Toronto, Ann Marie is called upon to meet the needs of families who have recently arrived in Canada, often with very limited English and unfamiliarity with our health care system. I asked Ann Marie what types of needs these families arriving in Canada might have and she recounted a recent experience.

“A mother arriving from China brought her son into clinic for the first time. She did not speak any English, so I asked one
of our lab technicians to help me welcome her to the clinic and to provide her with some basic information. Getting her and her son to and from clinic was going to be a big challenge and, if she couldn’t get here, then how could her son receive the treatment he badly needed? They were staying in a home where no one spoke English, only Cantonese. I taught her the word for taxi and I learned a few words in Cantonese so that I could arrange for taxi service each day with the help of the Toronto Chapter of the CHS which provided the taxi vouchers for them. I took her to the curb each day until she learned which taxi was for her and her son, and how to indicate where she wanted to go. I made sure she carried our clinic contact information at all times with the identification of her son’s bleeding condition. It really comes down to identifying what a patient needs and helping them along the way. All of our nurses take the responsibility for our work very seriously. We know we are trusted and we honour that trust completely.”

Another responsibility our nurses have is the preparation for and care of our kids at summer camps. It provides the kids with the opportunity to experience camp life in a safe environment. It is also a place where our kids learn about responsibility for their own health, keeping their bleed diaries, self infusion and making safe sports choices. Ann Marie encourages the kids to be part of record keeping at home. Accurate records of treatments and observations about bleeds help all of us and provide vital information should a recall of product be necessary.

The Hemophilia Treatment Centre: Like a Second Family

Although he is only 19 years old, talking with Dale Carey is like talking with a seasoned expert on hemophilia. Dale lives in Witless Bay within commuting distance of the HTC in St. John’s, Newfoundland. In Dale’s words, “They are there for me, like a second family. They keep me focused. They always have the personal touch you need, when you need it.” Dale has severe hemophilia and has had his share of joint problems. He was diagnosed as an 8-day-old infant at which time he had a serious brain hemorrhage. At 5, the same cavity bled again, causing Dale’s first of several seizures. He broke his arm as a child and endured various joint bleeds throughout his adolescence. Dale takes great pride in his HTC. He believes the commitment of clinic staff goes above and beyond any standard of care he has ever experienced. When he was 7, Dale had a joint bleed. He was being overlooked in the emergency department so his Mom called Dr. Kaiser Ali, the clinic hematologist, at his home. Dr. Ali arrived at the hospital within 10 minutes to take charge of his care and to ensure Dale never had to wait 2 hours for treatment again.

Dale has great doctors, including Mary-Frances Scully, now the Clinic Director. He also applauds the Nurse Coordinator, Marilyn Harvey, and all the clinic staff, “They were always willing to help me in any situation—not only committed doctors, but committed nurses and other staff, too.” Dale believes the HTC’s recognition of all needs—physical, emotional, mental or psychological—ensures they are providing outstanding team care. The commitment to people, and the caring he has experienced at his HTC have empowered Dale. He is passionate about improving provision of care and making life better for all people with bleeding disorders, starting with his beloved Newfoundland. Dale is studying to become a paralegal, to get some hands-on experience and then to pursue a law degree. Issues such as availability of care, building on the care we have now, provision of funding for travel costs to the HTC, social assistance and research are close to his heart.

I asked Dale how his life would be different if he did not have hemophilia. His answer wasn’t about his physical condition, or about pain or lost time in hospitals. Dale said he would never have become the determined and passionate person that he is, nor would he have developed the skills that will guide him through his professional life if he had not experienced the compassion and support of the staff at his HTC as he faced his many challenges. Bravo!! Dale, you are a very impressive young man and a source of inspiration!
Physiotherapy—
Finding Made-to-Measure Solutions

When our children are young we can’t easily predict the needs they may have. We can, however, rely on the team members of our HTCs to help us help our kids. Kathy Mulder is a physiotherapist in the Children’s Clinic at the Health Science Centre in Winnipeg. Kathy came up with a creative solution for a 15-year-old patient with Type III von Willebrand Disease who was having recurring problems with an ankle target joint. The bleeding had led to a pattern of reduced physical activity over time. Together they determined increased physical activity was required to...

- effect weight loss
- build muscle mass
- foster a healthy body image
- improve general well being
- improve venous access and
- protect the target joint from further deterioration.

They made a list of options and determined a home exercise program that best fit her needs. Kathy suggested appropriate exercises, and had her patient try various ones. Together they selected exercises, took pictures of the patient performing the exercises correctly and created an exercise poster of the exercise program. Kathy believes the team approach to this project was key; education was vital, and having her patient select exercises, make the poster, and determine the exercise schedule gave her ownership of the solution. They made it practical and made sure it suited the patient. Kathy established baseline data including strength measurements, muscle girth, weight and body measurements. The program is working well. Kathy is monitoring results on a monthly basis. They met weekly during the formative stages. This teenager also sought the help of a nutritionist as recommended by her clinic team to provide her with more knowledge about food choices and health. Kathy is very encouraged by this experience and offers us this new book as a great resource for people in a similar situation.

Social Work—
Handling Emotional as Well as Physical Challenges

Kathy’s ingenuity helped the teen in Winnipeg emotionally and physically. Kimberly Myers, a mother of a nine-year-old boy with hemophilia from Calgary, can relate to the combination of physical and emotional needs. Her son Justin has had a “high titer inhibitor”, since he was 2. Justin’s immune system quickly destroys conventional factor replacement products. Immune tolerance therapy did not work for Justin. For 5 years Justin had an operational port. Because of infection, it was removed. He has had several “pick” lines and now is dependent on hospital staff for venous access. This involves several pokes each time a vein is needed. Justin has several target joints resulting in weekly bleeds and a lot of time spent in the emergency department and as an in-patient. Unlike many of us who moved from hospital dependency to home care, Justin has become more dependent on the hospital, introducing new stresses and difficulties for him and his family to deal with. Justin has a 2-year-old brother who does not have hemophilia. Kim’s concerns for Justin are twofold.

“First of all we had to worry about the bleeds. But at least the port helped us deal with them and it became part of parenting for us with Justin. Now we have a very upset child who is terrified of needles and is struggling to deal with all the uncertainty and pain in his life. I am very concerned about his emotional health because he has had to deal with so much.”

The entire team at the Calgary HTC is working hard to help Justin. He is getting a great deal of help and support from Ruanna Jones, the clinical social worker. Ruanna has become a special friend to Justin. He is able to open up to her, giving him a consistent outlet to express his feelings. Kim appreciates their relationship and the added support of a professional to monitor his ability to cope and to help him deal with anger, fear and pain. Justin also has to contend with the struggles of Attention Deficit Hyperactivity Disorder, which can make learning, focusing and coping very difficult. Kim believes Ruanna is able to help Justin because she understands. She believes taking care of the whole child is very important.

Ruanna put a great deal of effort into finding Justin a day care solution to enable Kim to keep working. Although she was not successful, her help and support were invaluable. Ruanna remained involved in Justin’s care and was instrumental in arranging for a full time RN to be with Justin so he could enroll in the local primary school instead of attending the hospital school facility. Ruanna helped Justin make the transition go well by offering her expertise to all the parties involved. Kim continues to work 4 days a week. Ruanna has also helped Kim get counseling for Justin to deal with his fear of needles. They still have a lot to accomplish in this regard. Kim hopes Justin will be able to have a new port in the future. She soon hopes to visit the Quebec Reference Centre for the Study of Inhibitors, where even more expertise relating to port surgery and inhibitors is available, to evaluate Justin’s options. We wish all the Myers family members a speedy and healthy outcome.

Kathy Mulder, Physiotherapist, Health Science Centre, Winnipeg

Justin Shenher and Ruanna Jones, Social Worker, Alberta Children’s Hospital, Calgary

Peak Performance Fitness: Maximize Your Fitness Potential Without Injury or Strain, by Jennifer Rhodes, M.S. PT, published by Hunter House, ISBN 0-89793-296-X, 130 pages, approx. $25.00 (Tel: 1-800-266-5592)
The Day Ontario’s Health Care Restructuring Committee Came to Town

Pam Wilton, R.N., CHS Vice President

It was a dark and stormy morning when physicians, staff and administrators of St. Joe’s crammed into the auditorium to hear the decision of the Health Care Restructuring Committee. They were shocked to hear that their hospital would cease to exist as it had for over a century as an acute care teaching facility. Instead, it would become an Ambulatory Care Centre. The room was silent, as the decision sunk in. The CEO must have said something inspiring… he always does, but no one heard him. It simply couldn’t be true. Within a few minutes, pagers sounded and they hurried back to the wards and clinics to do what they do—save lives, and ease pain and suffering. Someone else would figure this out.

There is no Canadian model for an Ambulatory Care Centre. St. Joe’s, London, would be the first. Some were excited and looked forward to being a part of the planning and implementation, while others were skeptical and began looking for other paths.

The South Western Ontario Regional Hemophilia Program (SWORHP) was based at St. Joe’s. One would expect that a Hemophilia Comprehensive Care Centre would be a natural in a new Ambulatory Care Centre, but a decision was made to transfer the program to the other hospital, London Health Sciences Centre (LHSC). Many hematologists decided to leave London following the decision and it was felt that the Hematology/Oncology program was no longer viable at St. Joe’s. The program transfer was moved forward and planning began. Through the South Western Ontario Regional Hemophilia Program Council, members of the bleeding disorders community insisted that they be part of the transfer planning. The transfer would actually situate SWORHP at the pediatric site, but the Pediatric Clinic Director would leave before the transfer was complete. There were several concerns raised by the members of Program Council: adults would have access to emergency services at a separate site, adult admissions would be spread over four sites, some special hematology lab services were only available at St. Joe’s and the blood bank was separate from the program site. To further complicate care issues, many nurses experienced in hemophilia care had been “bumped” out of positions with the transfer of services and programs, the on-call hematologist was covering four sites and the physician specializing in care of patients with AIDS, who also had a special interest in hemophilia, left. Assurances were given that the problems would be overcome during transition phases. An experienced pediatric hematologist/oncologist was recruited. The Clinic Director, Nurse Co-coordinator and secretary worked hard to preserve the integrity of the Program.

Soon, patients and families began to report problems with care. There were further cuts to health care and erosion of services in regional hospitals with resulting pressures carried by physicians, nurses and other health care professionals. The Medical Director resigned and went into private practice in another province. The Chairperson of SWOR wrote to the London Health Sciences Centre, the District Health Council, the University of Western Ontario and the Ministry of Health asking them to accept their responsibility to provide health care.

Administrators of LHSC called a meeting. The problems were identified by each of the stakeholders and a task force was struck to review the resources and services provided by SWORHP. The completed report was submitted last fall. The resources necessary to deliver a hemophilia comprehensive care program, which will meet national standards, have been identified. It’s now up to LHSC to do the right thing.

For many, the day the Restructuring Committee came to town still seems like a nightmare. For members of the bleeding disorders community of Southwestern Ontario, it is very hard to understand how a world-class hemophilia program could be so vulnerable. More of that same determination, courage, commitment and vision that helped to establish a centre of excellence will be needed to guide SWORHP through the difficult decisions and changes in health care that lie ahead.

Models of Cooperation Between Clinics and Hemophilia Organizations

SOUTHWESTERN ONTARIO
Kelly Steiss, Regional Service Coordinator, Southwestern Ontario Region

The South Western Ontario Regional Hemophilia Program (SWORHP) formally includes the client organization—Hemophilia Ontario’s Southwestern Ontario Region—as a member of the Program’s integrated model of care. The Comprehensive Care Team includes a physician, nurse, physiotherapist, social worker, specialized laboratory technicians, hospital administrator and the client organization.

The client organization was involved in getting the clinics started. The Program Advisory Council was developed as the vehicle for client participation. The Comprehensive Care Team listens to and responds to the concerns, queries and recommendations of the clients. The team members have demonstrated their commitment by continuing participation in the Program Advisory Council meetings and by welcoming the participation of clients in developing and reviewing hospital policy and various documents. The Council recently reviewed the Hemophilia Program and made recommendations to the London Health Sciences Centre administration, which resulted in the hospital confirming support for the Hemophilia Program, securing social work services for our adults and expanding the role of our Nurse Clinician to that of Nurse Practitioner!

The client organization plays its role within the Comprehensive Care Team as well as the London Health Sciences Centre by...

- ensuring representation at the Program Advisory Council meetings;
- responding with letters, phone calls and requests for meetings when issues or concerns arise;
- ensuring that the Regional Service Coordinator attends the adult and pediatric clinics, which allows clients to put a face to a name, vitally important in developing a relationship. Subsequently, this allows the client organization to keep a pulse on the needs of clients.

This partnership continues to exist and function because members of the team hold each other in high regard, whether they are patients or care providers.
NEWFOUNDLAND AND LABRADOR

Difficulties in Getting Comprehensive Care Standards Recognized

Cyril DuBourdieu, President of the Newfoundland & Labrador Chapter

We in the Newfoundland and Labrador Chapter, as part of our mission, have been trying to improve the care of those with bleeding disorders for as long as I can remember. My personal involvement with the Society only goes back ten years or so, after my son was first diagnosed with hemophilia. The original clinics—adult and children—were set up in the late 1970s and early 1980s and were independent in their funding. It has been difficult to make progress, with the staff and budget cuts and health care restructuring Newfoundland faced throughout the 1990s. We saw the independent funding of our clinic being swallowed up by hospital and regional budgets. The staffing we had, which at one time was dedicated for hemophilia, became part of global hospital staffing. These changes did not move the level of care forward; in fact, we continued to lose ground in our efforts to improve the care for those with bleeding disorders.

Let me be clear—the staff at our clinic work hard and have gone beyond the call of their job descriptions in the past and present to provide the services we have. An example of this is that our clinic's medical director is not paid for giving of her expertise in this role and is considered to be acting in a voluntary capacity. If you look at a map of Newfoundland and Labrador, you will see our province covers a large area with a small population. However, the largest number of those with hemophilia and other bleeding disorders live outside of St. John's, the capital, where our clinics are located. In the mid 1990s, we at the Newfoundland and Labrador Chapter started a new round of lobbying to reinstate dedicated funding and access to outreach or traveling clinics, which we had identified as a priority some years before. Although we were listened to by those in power, we did not seem to make much progress until Dr. Mary-Frances Scully arrived on the scene here in Newfoundland. She started doing the outreach clinics without much support from either the Health Care Corporation of St. John's or the government. We suddenly had biannual clinics in both Twillingate and Corner Brook, thanks to this dedicated woman (For a profile of Dr. Mary-Frances Scully, please see page 22.). We continue to struggle to gain access to such things as increased nursing, physiotherapy and social services.

As many readers know, in 1998 a plan for comprehensive standards of care was put together by the CHS, clinic directors and other health care providers. We in Newfoundland provided input to reflect some of the needs here in Newfoundland and Labrador. We then presented the plan to the Newfoundland and Labrador Minister of Health of the day, the present Premier, Roger Grimes. Both we and the clinic team felt positive about the initial meeting and the clinic team went on to continue lobbying the Health Care Corporation of St. John's.

Although good things were said about our proposed Comprehensive Plan of Care, it became stalled in its implementation in 2000. A meeting was arranged with all the stakeholders. With so much at stake, we were pleased to get the support of Pam Willon, CHS National Vice-president, and Dr. Bruce Ritchie, chairperson of the Association of Hemophilia Clinic Directors of Canada, who came to Newfoundland to voice their support. Again, we had a great reception from the representatives of both the Department of Health and the Health Care Corporation. However, after several months, we learned that no action had been taken and the senior health care official at the meeting had retired.

We have continued letter writing and lobbying efforts over the past year, but the government, health care officials and the St. John's Health Care Corporation have all shirked responsibility for making changes to the service and passed them on to the next guy. The government officials stated they gave funding to the hospital corporation and the hospital decided on how it was spent. The hospital officials claim they do not receive enough funding to make the needed improvements to the program. And around we went!

In February 2002, we decided to refocus our efforts and a new campaign, please see page 22., was embarked on with input from National CHS and the clinic directors. Five hundred postcards were distributed to members of the Newfoundland Chapter and everyone was encouraged to mail these cards to their local government representatives and the Minister of Health. These cards asked the government to meet the standards laid down in the comprehensive care document. This was followed by a revised copy of the comprehensive care document sent with letters of support for the improved plan from Dr. Bruce Ritchie, Dr. Tom Alloway, CHS National President and our chapter.

While none of these efforts have resulted in increased resources for clinic activities or outreach, they have succeeded in raising the profile of the work done by Dr. Scully, the nurse coordinator, and the rest of the staff. Mr. George Tilly, chairperson of the Health Care Corporation of St. John’s, awarded them a Team award for their dedication and hard work. Imagine, Mr. Tilly, what they could do with the proper resources!

We will be following up our efforts with a request for meetings with the Minister of Health and the Health Care Corporation. We have shown that the implementation of this plan is not only in the best interests of patients but is also in the best interests of the administrators of the health care system. It would result in more effective delivery of care, services and products. Most importantly, it would guarantee a standard of comprehensive care that those with bleeding disorders in our province expect and deserve to have.
THE STORY OF COMPREHENSIVE CARE IN SASKATCHEWAN

A Long and Hard Victory Won

Eric Stolte, Past President, Hemophilia Saskatchewan

Sir Winston Churchill in his first address as newly appointed Prime Minister spoke the words, “…victory however long and hard the road may be…” It has taken almost 30 years of innumerable hours of volunteer time to achieve a funded, provincial comprehensive care program for persons with bleeding disorders here in Saskatchewan. Yesterday—March 10, 2002—during Hemophilia Saskatchewan’s 2002 AGM, the Bleeding Disorders Program Team was introduced. This was our victory celebration. But the work is never completely done. Continued effort to preserve and improve what we have will be necessary.

We’re deeply grateful for the commitment of doctors, nurses and other health care professionals who, despite the lack of dedicated funding, have provided their best care for hemophiliacs and their families.

Hemophilia Saskatchewan was founded in 1965 and it wasn’t until 1972 that the initial Bleeding Clinic supervised by Dr. J. B. McSheffrey was held. Three years later a temporary clinic RN was funded through research monies obtained by the College of Medicine. In 1976, after advocacy work by the society, the government funded a half-time Nurse Coordinator through the hospital budget of Royal University Hospital (RUH). But in the following two years, the government refused the requests from Hemophilia Saskatchewan (HSK) for $8,000 to help fund home care. Victory is never achieved without defeats.

Through the rest of the 1970s and 80s, a comprehensive care clinic was organized in name only. Team members were seconded to the clinic without a specific mandate or funding. In the 1990s, health care was reorganized and RUH became a site in Saskatoon District Health. This, plus health care cutbacks, led to a reduction of the RN position to less than 4 hours/week. The absence of a mandate for other positions and the lack of provincial status led to an overall program reduction. Comprehensive care became a very hollow label.

In March of 1994 I was elected to the Presidency of HSK. With the Krever Commission well under way but with our chapter in financial distress, I determined to move on three priorities. 1) Achieve financial stability through thrift and fundraising, 2) Continue to build on our strong sense of community through programs and 3) Regain and maintain a provincial comprehensive care program. Priority three took all seven years of my terms as President.

Dr. Robert Card, our Clinic Director, was due for a sabbatical soon so I wrote a letter to then Chair of Saskatoon District Health (SDH), Cliff Wright, alerting him to the “crisis” of non-funded hemophilia care. He then asked the Associate Minister of Health, Lorne Calvert, for provincial funding. Of course, Minister Calvert replied that funding for this was the SDH responsibility.

Without HSK’s knowledge, a funding proposal for a “Saskatchewan Comprehensive Care Hemophilia Program” was prepared over the summer of 1994 by a SDH health care student. In August, this proposal was submitted to Saskatchewan Dept. of Health by SDH President, John Malcolm. One of the flaws of this proposal was having no place for consumer input.

As this was occurring, a CHS task force in which I participated had been set up to produce a document on our vision for comprehensive care. In May of 1995, the CHS “Red Book”, A Vision of Comprehensive Care for Persons with Inherited Bleeding Disorders was printed. However, from August 1994 until March of 1996, the original proposal sat on a government shelf gathering dust. I inquired of the Health Minister, Lorne Calvert, but have no record of a response. Bureaucratic gears grind ever so slowly.

I suspect that some time in early 1996, the Canadian Blood Agency (CBA) exerted some pressure on the province to accurately track product usage primarily for accountability and efficiency purposes. This created a flurry of activity resulting in a landmark meeting on March 11, 1996 of 10 stakeholders including CBA rep. to Sask Health, Bill Dorsett; CHS President, Durhane Wong-Rieger; Sask Health rep. Al Johnston and others, including me.

The interest in a provincial comprehensive care program was renewed and a working group was formed to revise the proposal for immediate submission. Finally, in November, a reworked proposal, incorporating both the CHS “Red Book” and AHDCD guidelines was given to Al Johnston of Sask Health.

While this new proposal was gathering new dust on government shelves, a two-year-old boy, after an incredible ordeal related to complications from surgery for a port-a-cath and the development of severe inhibitors costing the province well over $1 million, passed away. Finally, in August 1997, nine months after the submission of the revised proposal and 17 months after the March 1996 meeting, Dr. Card, having heard nothing from SDH sent a memo to Susan Bazylevski, VP of Medicine for SDH, informing her of the “crisis state of hemophilia care” and that the existing “program” would dissolve at the end of the year unless funding was found. In September she replied that she was “…surprised to read your concerns regarding the “crisis state of the existing program”. Not only do bureaucratic gears grind slowly, but bureaucratic bubbles are hard to burst! After a few more months of inactivity, a
third revision of the proposal was submitted to Sask Health in April, 1998. In November we heard that Regina District Health had agreed that only one provincial program was needed, based in Saskatoon. I suppose seven months was needed for such complex plans to be determined!

After another year of frustration and inactivity, CHS supported HSK in January 2000 with letters calling for a meeting with Health Minister, Pat Atkinson. Three months later, on April 12, 2000, a seven-month-old boy died from what is suspected to be an undiagnosed intra-cranial bleed. CHS President, Erma Chapman, mentioned to media that where there are comprehensive care programs, mortality rates of hemophiliacs are reduced. Without any meeting or communication, the Minister of Health announced that a Provincial Comprehensive Care Bleeding Disorders Program would be implemented after meetings with Sask Health officials.

Although we cannot directly link the boys’ deaths to a lack of care, it certainly speaks to the importance of comprehensive care programs. With an excellent program in place, we would have had assurance that these deaths were unavoidable. Now all we have are unknowns. The issue is not the care shown by the treaters who did all that they could to help the boys. The issue is that comprehensive care adds so much more to what treaters can do individually during a crisis.

It took from April until October to finally have the meeting and then six more months until a revised budget could be approved. In August 2001, a year and a half after Pat Atkinson committed to funding, a full-time Nurse Coordinator, Cindy Olexson, was appointed. Others have been added to the team. Now, in March 2002, we’re investigating with Sask Health the right timing for an official launch.

Although we cannot directly link the boys’ deaths to a lack of care, it certainly speaks to the importance of comprehensive care programs. With an excellent program in place, we would have had assurance that these deaths were unavoidable. Now all we have are unknowns. The issue is not the care shown by the treaters who did all that they could to help the boys. The issue is that comprehensive care adds so much more to what treaters can do individually during a crisis.

Lessons from Saskatchewan
1. Keep at it no matter how dim the prospects. You never know when the climate may change.
2. Try to create win-win scenarios—don’t become enemies of government.
3. Always remember the cause; it’s worth all the hard, slow and continuing effort. Realize that even though our cause is of great importance and the benefits are obvious to us, this is not clear to others, especially those in government. They must respond to requests from many areas. It is our advocacy role to be persistent in making the case.
4. Be patient and “nag” periodically. It’s not always a direct frontal assault that will produce results.
5. Remember, CHS national is part of the team. They can bring helpful pressure that provincial chapters can’t always produce.
6. Remember, too, that physicians and other health care workers are part of our team. Include them in your plans and work together for the common goals we share.
7. When good things happen, applaud and congratulate, even when things seem to be too little, too late.

Models of Cooperation Between Clinics and Hemophilia Organizations

BRITISH COLUMBIA

Hillary Rudd

The British Columbia Chapter of the Canadian Hemophilia Society was formed in the 1960s and, in 1972, the Hemophilia Assessment Clinic began operating in Vancouver. The main goals of the clinic are to evaluate patients’ needs through annual or semi-annual assessments, distribute factor concentrates, prepare patients for home care, educate doctors and other health care workers, as well as advocate for the clinic with the government.

The Society initially played a major role in convincing the provincial government that there was a legitimate reason to fund the Clinic’s activities. The Clinic’s philosophy and operations were developed with the help of Society members.

The BC Chapter, and thus the clinic, are funded under the Community Grants Division of the provincial government. This arrangement helps to ensure the continued operation of the Clinic since the funding goes to an organization and the clinic is not funded directly from the health region. The BC Board contracts out with the Mary Pack Arthritis Centre at Vancouver General Hospital, where the clinic is located. With the grant, the Chapter pays for all of the salary of the nurse and part of the salaries of the physiotherapist, secretary and social worker. Because we are underfunded by the province, we rely on corporate sponsors, community service organizations (e.g. Lions Club), and donations from individuals and members of the hemophilia community to complement the provincial grant.

The BC Chapter works closely with the clinic to ensure that the medical and emotional needs of the province’s hemophiliacs are met.
HÔPITAL STE-JUSTINE, QUEBEC

A world leader in comprehensive care

Patricia Stewart

In 1979 Hôpital Ste-Justine, affiliated with l’Université de Montréal, was designated by the Quebec government as one of four official Hemophilia Treatment Centres in Quebec. The centre has been under the direction of Dr. Georges-Etienne Rivard from that day on. Ste-Justine has since become a leader in the diagnosis and treatment of inherited bleeding disorders including hemophilia and von Willebrand Disease, and the management of inhibitors.

The team at the hemophilia centre includes a director, an associate hematologist, two part-time nurses, a research nurse, and part-time physiotherapist, social worker and psychologist. The inhibitor centre has a full-time nurse coordinator.

Over 600 patients with various types and severity of bleeding disorders are registered at the centre. Because there is no official adult hemophilia centre in Montreal, adults continue to be followed at Ste-Justine through a cooperative effort with an associate hematologist, Dr. Jean St-Louis. He is present during the annual checkup and consults with the team to assure continuity when an adult needs to be hospitalized elsewhere. He can count on the support and expertise of the team at Ste-Justine.

Staff offer support for the child and the immediate family as well as documentation and videos for families to use as educational tools with teachers, babysitters and other care providers. The team acts as a resource and, when needed, holds outreach information sessions.

The team at Ste-Justine participates in a number of cooperative research projects. One of the latest is a primary prophylactic study with 1- to 2 1/2-year-olds, to discover the minimum quantity and frequency of infusions needed to completely prevent bleeding and joint disease. Another cooperative project is a study of the genetic markers for von Willebrand Disease in families, headed by Dr. David Lillicrap in Kingston. Ste-Justine, and particularly Dr. Rivard, are well-known for research into radioactive synovectomies. A fourth research project, recently completed, evaluates the use of a visual evaluation tool—a pictogram—to measure actual menstrual blood loss as compared to blood loss as perceived by the woman herself.

In December of 2001, Ste-Justine and the Hospital for Sick Children in Toronto, each received a three-year commitment of $400,000 from Bayer Inc. that will enable them to continue as leaders in hemostasis research and treatment. At Ste-Justine, three specific programs will especially benefit.

With the creation of the Hémostase au Féminin program, initiated by Drs. Georges Rivard and Michèle David in May 2000, Ste-Justine became the first Hemophilia Treatment Centre in Canada to create a team of specialists offering services specifically designed to diagnose and treat gynecological/obstetrical complications directly related to a problem with hemostasis. The team includes two hematologists, an obstetrician/gynecologist, a pharmacist, a laboratory technician, an anesthetist, and a nurse coordinator. A number of research projects are presently underway, including a study on the effect of exercise on VW factor levels and menorrhagia led by Dr. Rochelle Winikoff, resident in hematology.

Ste-Justine is designated as a Mother–Child University Hospital Centre. One of the projects underway is a study of the genetic and environmental factors contributing to the roles and responsibilities of each group are not clearly defined. She feels that the relationship between the CHS and CHSQ is excellent although sometimes she does not know who to approach (CHS National? Quebec Chapter?) in certain situations. She feels that you are an important pressure group because of your involvement in the blood system today. The CHS is still considered to have an important role in the blood system. “We know that you are an important pressure group because of your involvement in the blood system in Quebec, and this is an asset if we need it,” declared Dr. Demers. She says the establishment of proper standards of care for hemophiliacs.

Today the CHS is still considered to have an important role in the blood system. “We know that you are an important pressure group because of your involvement in the blood system in Quebec, and this is an asset if we need it,” declared Dr. Demers. She says the relationship between the CHS and CHSQ is excellent although sometimes she does not know who to approach (CHS National? Quebec Chapter?) in certain situations. She feels that the roles and responsibilities of each group are not clearly defined.

Most of the time cooperation involves the exchange of information and here also it is excellent on both sides. In addition, the Association of Hemophilia Centre Directors of Quebec invites the CHSQ to its annual meeting, and the CHSQ in turn invites and/or hires staff from the treatment centres (hematologists, nurses, physiotherapists, etc.) for some of its information workshops and various activities such as the family weekend and summer camp for young hemophiliacs.

Models of Cooperation Between Clinics and Hemophilia Organizations

QUÉBEC
François Laroche

Traditionally, hemophilia treatment centres in Québec and the CHS have always worked together. When the four centres were opened in 1979, the CHS and its Quebec Chapter (CHSQ) supported the hematologists when they pressured the Minister of Health to establish proper standards of care for hemophiliacs.

Today the CHS is still considered to have an important role in the blood system. “We know that you are an important pressure group because of your involvement in the blood system in Quebec, and this is an asset if we need it,” declared Dr. Demers. She says the relationship between the CHS and CHSQ is excellent although sometimes she does not know who to approach (CHS National? Quebec Chapter?) in certain situations. She feels that the roles and responsibilities of each group are not clearly defined.

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to thrombophilia in pregnant women. Thrombophilia may contribute to growth retardation in the fetus due to a progressive coagulation/obstruction of placental blood vessels.

The third program which will benefit from this funding is the Inhibitor Centre. Officially designated by the Quebec Ministry of Health on March 30, 2000 as the Quebec Reference Centre for the Treatment of Patients with Inhibitors, the centre offers specialized care in this most difficult to manage aspect of hemophilia. This program is unique in Canada and offers care for patients with inhibitors, both inherited and acquired. A 24-hour emergency line allows patients to receive help within ten minutes of calling. Approximately 30 patients are treated throughout the year, half with acquired hemophilia. An increasing number are being referred from across Canada and requests for information and consultations have begun to come in from around the world.

In all these cases, the new funding will help solidify infra-structures, allowing a work environment favourable to the continuation of research and clinical services. Salaries for nurses, funds for researchers in molecular biology, data collection, fellowships and the education of residents, nurses and other medical personnel through participation in conferences and congresses will all be made possible.

Obviously, the hemophilia program can only profit from these projects which will enhance the education and training of personnel in related fields. “While recruiting health care professionals specializing in hemostasis is a challenge everywhere, it is particularly so in Quebec where individuals must be fluent in French,” notes Dr. Georges-Etienne Rivard. “Bayer’s generous funding is critical to ensuring our ability to employ young and bright research staff and to embark on research projects that will build Quebec’s leadership in the understanding and treatment of bleeding disorders.”

Sick Kids participates in a number of nation-wide studies, including a primary prophylaxis study with children from 1 to 2 1/2 years of age. This is a patient-driven, dose escalation study whereby the number of treatments is increased in frequency and dosage depending on the response of the child. The purpose is to find a minimum treatment regimen that will prevent joint bleeds and avoid the need for insertion of a central line (port). A similar carry-over project to study the management of treatment for boys from 3 to 5 years is underway. Another study is being done in collaboration with the neurological department comparing neurological functionality in children with hemophilia who have had a head bleed with those who haven’t. A fourth study of young girls referred to the treatment centre due to menorrhagia is also taking place. And finally, Sick Kids participates in a cross-Canada study of genetic markers in Type 1 von Willebrand Disease headed by Dr. David Lillicrap at Queens University.

In June 2001, Sick Kids and Ste-Justine in Montreal each received a three-year, $400,000 commitment for funding from Bayer Inc. that will enable them to continue their leadership roles in the development of expertise in the fields of hemostasis and thrombosis.

Sick Kids pediatric hematology services include other conditions like...
thrombophilia. Children with this disorder suffer strokes, heart attacks and deep vein thrombosis. The Bayer funding will help train fellows who will specialize in hemostasis and thrombosis. “These two specialties are wedded in children due to the limited number of pediatric hemostasis specialists and the limited number of children’s hospitals,” states Dr. Blanchette. In most centres, especially the smaller ones, hematologists already deal not only with bleeding and clotting problems, but also with oncology. Sick Kids is able to concentrate specifically on bleeding and clotting disorders because of the large population in the Toronto area and the presence of specialists at the HTC. Sick Kids also treats rare and difficult cases from across the country.

Funding for clinical mentorships will enable staff to be partnered with recognized clinical experts, further building expertise in the care of children with bleeding and clotting disorders. The new funding will also enable specialists from the Centre to train medical personnel at conferences and scientific meetings as well as training visiting medical personnel (nurses, physiotherapists, doctors, geneticists) in the care and treatment of children suffering from these disorders. “Bayer’s involvement is critical to ensuring The Hospital for Sick Children not only grows the overall knowledge base in the areas of pediatric hemostasis, but also that we are also able to transfer this knowledge and skill to others,” said Dr. Victor Blanchette. It will support the salaries of nurses, pharmacists, and physical therapists in the field of pediatric hemostasis, and will support basic and applied research in this new and rapidly expanding area of pediatric medicine.

Funding for clinical mentorships will enable staff to be partnered with recognized clinical experts, further building expertise in the care of children with bleeding and clotting disorders.

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### The Major Milestones of Hemophilia Care in Canada

<table>
<thead>
<tr>
<th>Year</th>
<th>Event</th>
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<tbody>
<tr>
<td>1947</td>
<td>Whole blood and fresh frozen plasma became widely available in Canada. This marked the beginning of factor replacement therapy for people with hemophilia.</td>
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<tr>
<td>1953</td>
<td>The Canadian Hemophilia Society was founded in Montreal.</td>
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<td>1964</td>
<td>Cryoprecipitate was discovered. Effective treatment for hemophilia A became possible.</td>
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<tr>
<td>1968</td>
<td>The first factor VIII and IX concentrates were introduced. The first experiments with home infusion began.</td>
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<td>1969</td>
<td>The first Canadian hemophilia treatment centre offering comprehensive care was opened in Montreal.</td>
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<tr>
<td>1970s</td>
<td>Hemophilia treatment, based on factor concentrates and home infusion, began to be available in larger centres across Canada.</td>
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<td>1979</td>
<td>The Quebec government recognized the concept of comprehensive care by designating 4 treatment centres with dedicated funding.</td>
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<td>1980</td>
<td>The Winnipeg Conference, organized by the Canadian Hemophilia Society, was held to discuss comprehensive care. The conference served as the springboard for the creation of a network of hemophilia treatment centres across Canada.</td>
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<td>1985</td>
<td>Heat-treated factor concentrates, effective in eliminating HIV, were introduced in Canada.</td>
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<td>1988</td>
<td>Factor concentrates, manufactured with enhanced viral inactivation methods, effective in eliminating hepatitis C, began to be used in Canada.</td>
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<tr>
<td>1993</td>
<td>Genetically engineered (recombinant) factor VIII concentrates were introduced in Canada. The network of hemophilia clinic directors, associated to improve standards of care and facilitate hemophilia research since the 1970s, was incorporated as The Association of Hemophilia Clinic Directors of Canada.</td>
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<tr>
<td>1997</td>
<td>The network of nurse coordinators, which had been meeting to provide a forum for professional development since the 1970s, was recognized as the The Canadian Association of Nurses in Hemophilia Care by the Canadian Nurses Association.</td>
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<td>1997</td>
<td>Genetically engineered (recombinant) factor IX concentrates were made available in Canada. Canada became the first country in which all people with hemophilia A and B had access to recombinant factor concentrates.</td>
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<td>1998</td>
<td>The Winnipeg II Conference on comprehensive care, bringing together health care workers and patients, was held to re-define standards of hemophilia care.</td>
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<td>2000</td>
<td>Second-generation genetically engineered (recombinant) factor VIII concentrates, without human or animal protein as a stabilizer, were introduced.</td>
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<tr>
<td>2001</td>
<td>The Saskatchewan government formally designated a Hemophilia Treatment Centre to provide comprehensive care to people with hemophilia in that province.</td>
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Hemophilia Centre of Eastern Quebec

François Laroche

The Hemophilia Centre of Eastern Quebec is located in the Saint-Sacrament Hospital (an affiliated hospital: CHA) in Quebec City. It is one of the four centres for the treatment of hemophilia designated in 1979 by the Quebec Minister of Health to deliver comprehensive care to all Quebec hemophiliacs. This includes managing and prescribing clotting factor concentrates to anyone with a blood clotting disorder in Quebec.

As soon as it opened in 1979, the Hemophilia Centre of Eastern Quebec became a model, an example to follow for other treatment centres in Canada. The doctors there from the outset, including Dr. Jean-Marie Delâge and Dr. Agathe Baril, built a solid foundation that still functions very well today. The comprehensive care provided, and especially the control of the inventory and distribution of blood products by the centre, make it one of the leaders in this regard.

The Centre Director, Dr. Christine Demers, a hematologist who specializes in hemostasis, and the staff that work there full-time (Nurse Coordinator and senior secretarial/administration clerk) or who are on call when required by patients (physiotherapist, pediatrician, orthopedic specialist, infectiologist, gastroenterologist, rheumatologist, gynecologist, dentist, etc.) are all top-notch. Unfortunately, in recent years, with the transfer of the hemato-immunology unit, the change in St-Sacrement Hospital’s role and the CHA’s accumulated deficit, some of the services have been eroded.

For the past two years, hemophiliacs and hematology patients can no longer be hospitalized at Saint-Sacrament Hospital but have to go to the affiliated hospital, Enfant-Jésus. Outpatient clinics, however, are still held at Saint-Sacrament. Doctors, therefore, have to divide their practice between the two hospitals. Also, when hemophiliacs are hospitalized and their condition requires a continuous infusion, for example, the nurse-coordinator is not there to supervise it.

Other problems include the position of social worker which was created in 1985 and eliminated in 1994 for budgetary reasons. For the same reasons, when the dentist assigned to the centre took sick leave in May 2001, he was not replaced and his position has still not been filled. But the most disturbing development occurred in 2001 when the position of physiotherapist for CHA outpatients was eliminated. Hemophiliacs were thus deprived of a specialty that is essential to maintaining the health of their joints. The director of the hemophilia centre and the Quebec Chapter of the CHS, through some of the centre’s patients, mobilized to apply the necessary pressure to reverse the situation. “The situation had become unacceptable. We were losing the services of a specialist with recognized expertise and interest in hemophilia. He was the one who did the annual assessments of our patients’ joints, before determining if they should be seen by an orthopedic or other specialists,” explained Dr. Demers. “Hospital management did not seem to understand that cutting this type of position would result in much higher health care costs (therapy, hospitalization, etc.) than the specialist’s salary,” she continued. After prevaricating for several weeks, CHA management agreed to re-hire the same physiotherapist (who had found another place to practice in the meantime) but only part-time. He is now available to do annual assessments and is on call. “Although this situation is not ideal, it’s working very well. The physiotherapist is available enough to see our hemophiliacs quickly, often the same day,” said Dr. Demers.

Despite these problems, Dr. Demers thinks the treatment centre is still a model, especially in regard to the control of blood products. The system at Saint-Sacrament was created when the hemophilia centre opened and it ensures that all clotting products sent to hemophiliacs in eastern Quebec go through the treatment centre before being distributed. This means that in order to receive replacement therapy, someone with a clotting disorder must first be registered at the hemophilia centre. “This tightly controlled system means products can be traced quickly and efficiently. Thus, if a product is recalled or withdrawn, we can easily contact the patients concerned directly because we know exactly who has which batch of the product,” said Dr. Demers.

Also, since it introduced annual assessments, the centre asks hemophiliacs to make an appointment at least once a year to have their condition reassessed. They see the nurse-coordinator for routine blood tests, and also the physiotherapist in the majority of cases. They are then referred to other specialists if necessary. About seven times a year, pediatric days are organized for young hemophiliacs, which also gives them the opportunity to meet with the pediatrician and hematologist. “The full-time staff at the centre are competent and dedicated, which is essential if our structure is to work well,” concluded Dr. Demers.

Transfer of the immuno-hematology department to Enfant-Jésus Hospital

According to Denis Carbonneau, Director General of the CHA which combines two Quebec City hospitals, Saint-Sacrament and Enfant-Jésus, “The Saint-Sacrament hemophilia centre will soon be moved to Enfant-Jésus Hospital, together with the rest of the hematology and immunology department (including staff, laboratories and other equipment).” Although this has been dragging on for nearly seven years, the transfer date is still not known, although every indication is that it will happen this spring or, at the latest, this fall.

Mr. Carbonneau says his mandate is “to organize the transfer in a way that attempts to satisfy everyone—users, physicians and other staff involved.” Meetings were held in this regard in February with the institution’s director of professional services, Pierre Laliberté, and the centre’s hematologists.

It is reassuring that CHA management seems to consider the hemophilia centre to be inseparable from the rest of the hemato-immunology department. Thus the expertise of the hematologists already in place and the expertise accumulated over the years will continue to be accessible to hemophiliacs in eastern Quebec.

According to Dr. Demers, “Moving the hemophilia centre to Enfant-Jésus Hospital will provide hemophiliacs with better support since all related specialties will be available under one roof.”
In Canada, an effective network of Comprehensive Care Bleeding Disorder Clinics has evolved under the guidance and support of Canadians with bleeding disorders. This network of comprehensive care clinics has been fabulously successful in maintaining the health of people with bleeding disorders, moving their health care out of hospitals, and into their own hands in their homes, where it belongs. However, despite the initial savings and obvious benefits of keeping people out of hospitals, costs have increased regularly as the use and price of coagulation products have increased. This has happened worldwide, not just in Canada, and funding agencies are scrambling to cope. The cause of this increased cost is complex, but is for the most part based on increased use and cost of coagulation products.

The increased cost of coagulation products is partly dependent on the introduction of improved products, but more critically, on the costs of meeting changing regulations imposed by the regulatory agencies. Team Biological of the US Food and Drug Administration has been visiting biopharmaceutical companies, issuing notices, warning letters and Consent Decrees, as the companies scramble to bring their production in line with the rest of the pharmaceutical industry. This has been a painful experience for the biopharmaceutical companies and for the patients served by these companies. The shortage of factor VIII, and the measures imposed as a result of this over the last eighteen months have been a clear result. The costs of meeting changing regulations are passed on to the consumers as increased costs of coagulation products.

In addition, the use of coagulation products has increased by 25% per year for a number of years, driven by prophylactic use of coagulation products and immune tolerance therapy protocols, which call for regular infusions of large amounts of coagulation products. These treatments clearly work. Prophylactic use of coagulation factors protects the joints of patients with bleeding disorders, keeping them healthy, productive members of society. Immune tolerance therapy can nearly normalize the life of a bleeding disorder patient with the life threatening complication of an inhibitor. Both strategies are now established as the standard of care throughout the Developed World. Up until now, Canada has been able to keep up with the costs of these useful treatments, but provincial governments are looking closely at all aspects of medicine to reduce costs.

The solution is to justify the increased costs. One way to do this is to begin to audit Comprehensive Care Bleeding Disorder Clinics, before health care administrators do it for us. This audit would be done by a multi-disciplinary group, including: the Canadian Hemophilia Society, Health Canada, Canadian Blood Services, Héma-Québec, the Association of Hemophilia Clinic Directors of Canada, the Canadian Association of Nurses in Hemophilia Care and international representatives from the Centers for Disease Control, and possibly Europe. Such a system would use people with a unique background in the care of bleeding disorder patients to optimize treatment at each centre and provide the ammunition for these clinics to lobby for improved resources.

Quality control and quality assurance are key to maintaining effective comprehensive care for people with bleeding disorders. A regular audit system is a mechanism to maintain quality assurance, and demonstrate this quality assurance to the funding agencies. Justification of what we do is key to maintaining comprehensive care in a competitive environment.
pursue internal medicine. She tutored in Pediatrics at the Royal College of Physicians and Surgeons, and she planned to study and work at Great Ormond Street, the world famous hospital in London, England.

But her sense of duty to humanity was too strong, and she became interested in the peace movement and the work of individuals such as Gandhi and the Dalai Lama. She studied at the Menjushri Institute in England where she learned the principles of meditation. In 1984 she went to India where she worked for a month in Darjeeling at a small cottage hospital, and then in the district of Karnataka, where she trained local doctors.

In 1985 Dr. Scully applied to both the University of Toronto and Great Ormond Street Hospital for a Fellowship in Pediatrics. She was awarded a place at the University of Toronto and it was there that she met and worked with Dr. Blanchette. In 1987 she applied to Queen's University to study hematology and wound up working with Dr. David Lillicrap. Dr. Scully says that she “had a fantastic year there,” working with Dr. Lillicrap and learning about hemophilia. Between 1988 and 1990 she worked with Dr. Giles at McGill University on laboratory hematology, where she gained considerable knowledge about the clotting system and factor IX hemophilia. From 1990 to 1991 Dr. Scully received further training with Dr. Georges-Etienne Rivard in the Hemophilia Clinic at Ste-Justine Hospital in Montreal. But from 1991 to 1997 she was actually able to put down some roots after she arrived in Saint John, New Brunswick, where she worked with Dr. Sheldon Rubin and Dr. John Pond in helping to establish a Hemophilia Comprehensive Care Clinic in Saint John. A clinic had been previously established in Moncton, but that had left a large area of New Brunswick without adequate hemophilia care.

While in New Brunswick, Dr. Scully had the opportunity to meet a family of three men, all hemophiliacs, all suffering to one degree or another from inadequately treated bleeds. Their method of treating a joint hemorrhage was to pack the affected joint in bags of frozen peas. Their fridge was virtually full of frozen peas, but there was not a speck of coagulation product of any kind to be found.

Dr. Scully began work to improve hemophilia care soon after she arrived in Saint John. She began a screening programme for von Willebrand Disease and set up a pediatric oncology programme for children suffering from cancer, but by 1997 cutbacks in funding medical care made further programmes impossible to set up. The hospital administrators suggested that they close the Hemophilia Clinic to save money. So, in 1997 Dr. Scully took up a position in St. John’s, Newfoundland where she soon became Medical Director of the Hemophilia Comprehensive Care Clinic.

The clinic had begun under the leadership of Dr. Kaiser Ali in the 1970s, but by 1997 over 70% of the hemophilic population received no follow-up care simply because of the geography of the province. Many of the patients lived great distances from St. John’s. In fact, some who lived in Cornerbrook had to travel for ten hours by car to attend Clinic, or they had to fly, at great expense. And neither the government nor the chapter had the funds to assist these patients. To add to the problems, the hemophilia nurse became ill and the continuity so necessary in a programme featuring follow-up services was lost. In response to these problems, Dr. Scully and her team began a series of Travelling Clinics to rural central Newfoundland and held all day Clinics to attract the hemophilic population, many of whom were fishermen, at sea when the weather permitted. This programme has been a grand success. The rural physicians have been very supportive, and the patients, while sometimes a bit apprehensive about the connections between HIV, HCV and hemophilia, have also taken the programme to heart. Dr. Scully has found a new Nurse Coordinator, Marilyn Harvey, who is really supportive of the programme. All in all, the Travelling Clinics have been a “big hit”, as Dr. Scully says, even though there are problems with weather and with patients who can’t attend because fishing is their livelihood. As long as there are funds enough to keep the Travelling Clinics on the move, it looks as though Newfoundland’s hemophiliacs face a much more promising future. Dr. Scully is also considering a video conferencing programme for follow-up with patients who live in the outports.

The Travelling Clinics have had unexpected results. Growing out of an interest that began when she worked with Dr. Lillicrap, and working with a group of physicians and researchers, Dr. Scully has participated in identifying a significant cohort of people related to each other, numbering over 1,800, of whom 112 have mild factor VIII hemophilia. They suspect there are more members in this group and more mild hemophiliacs to be found within it. Of most interest is that they have identified a different mutation, and they now think that the effects of mild hemophilia can be as severe as those of moderate or severe hemophilia, particularly when the patient’s diagnosis has been missed and he needs surgery.

As if all of her accomplishments are not enough, Dr. Scully has plans to go to Labrador and establish a Travelling Clinic programme there. How she has managed to do all she has over the past few years leaves one gasping for breath just trying to keep up. And one sincerely hopes that she is able to keep up her work as long as possible, for the Atlantic Provinces, surely, have greatly benefited from Dr. Scully’s efforts.
Back-up Recall Notification System
Announced by CBS, Héma-Québec

CHS to Sit on Advisory Board

In November 2001, the Canadian Blood Services and Héma-Québec finalized an agreement with the Plasma Protein Therapeutics Association to provide product recall information for Canadians using plasma and recombinant products. Called the Patient Notification System (PNS), this voluntary service has been in place in the United States for several years. (See NEW SYSTEM FOR PLASMA PRODUCT RECALL INFORMATION for a complete description.)

At its November meeting, the CHS Board of Directors voted to inform its members about the service through Hemophilia Today and to continue to sit on the PNS Advisory Board. Board members felt that the new service may provide information on recalls/withdrawals that some people with bleeding disorders would not otherwise get. In addition, Board members stated that some people with bleeding disorders may want to have a secondary source of information on recalls/withdrawals in addition to the primary source in hemophilia treatment centres.

At the same time, given its voluntary nature, Board members stated very clearly that they see the PNS as a back-up to the primary notification done by treating physicians and/or hospital officials as per their legal responsibility. (For a description of this primary notification system, see Recalls, Withdrawals and Quarantines of Clotting Factor Concentrates on page 25.)

Patient Notification System

New System for Plasma Product Recall Information

(Article provided by Canadian Blood Services)

A confidential Patient Notification System (PNS) is now available to Canadians interested in information on plasma products. The system is a 24-hour communication system that provides up-to-the-minute information on the status of recalls and withdrawals of plasma-derived and recombinant products at no cost to the consumer.

The PNS was originally designed in the United States by the Plasma Protein Therapeutics Association of Washington, D.C. (made up of manufacturers of life-saving plasma protein therapies) and several U.S.-based consumer groups. It has been adapted for Canadian use by an advisory panel of Canadian companies, and relevant medical and patient groups who worked with Canadian Blood Services and Héma-Québec. In Canada, the system is offered with funding from plasma and protein therapy producers and distributors.

The system is entirely voluntary. Registering or choosing not to register does not change an individual’s right to be informed and does not replace normal patient notification done by treating physicians and hospitals. Existing communication methods between patients and hospitals will not be affected, nor will existing communication methods between Canadian Blood Services or Héma-Québec and their hospital customers be affected. In Canada, in the event of a recall or withdrawal of blood or blood products, it is the primary and legal responsibility of the treating physician and/or hospital officials to inform the patient and ensure that any products still in circulation are not used.

If you would like to register, you can do so by visiting the web site at www.patientnotificationssystem.org, calling the toll-free phone number (1-888-UPDATE-U), or by sending an application form by mail to NNC Group, Attention: PNS Manager, 5250 West 76th Street, Indianapolis, IN, 46268. Pamphlets containing application forms are available upon request through your organization. There is no cost to the consumer and you will be asked to provide your name, contact information, password, password hint and a list of the product(s) you would like information on. You can choose to be contacted by express mail, telephone, fax or e-mail. You will also receive a follow-up notification by regular mail. Detailed information can be found on the web site.

The Patient Notification System is completely confidential. It is operated by NNC Group, an independent organization that specializes in pharmaceutical notifications. All patient information is held in strict confidence by NNC Group. Every attempt will be made to reach you within 24 hours of any recall or withdrawal.
Recalls, Withdrawals and Quarantines of Clotting Factor Concentrates

David Page
CHS Blood Safety Coordinator

The last recall of factor VIII or IX concentrates in Canada occurred in September 2001. One particular lot of a recombinant factor concentrate was recalled because the manufacturer, through its regular quality control procedures, found that the lot had lost some of its potency—it was no longer as effective as it had been. The potency had, in fact, fallen to 78% of that indicated on the label. Health Canada regulations require that when the potency of a factor concentrate falls below 80%, the manufacturer must notify Health Canada and recall the product. That is exactly what happened with this one lot.

Understandably, recalls cause concern. Patients and their families wonder if the product they have been using is unsafe. They ask if there might be a continuing problem with the replacement products. They question whether or not they have all the information. This article is intended to explain recalls, withdrawals and quarantines—all of which are different—and describe the procedure from manufacturer to patient when a recall, withdrawal or quarantine occurs.

Recalls – At some time a Hemophilia Treatment Clinic (HTC) may inform patients that there has been a recall of clotting factor concentrates. This means that there is a concern that the product may not be safe, or that it may not be entirely effective in stopping bleeding. Such recalls occur very rarely. The recall may come about because the manufacturer has become aware of a problem with efficacy, as in the September 2001 case described in the introduction. It may happen because there has been a report of an unexpected adverse reaction in a patient which, by law, because of safety concerns, must be reported to the manufacturer.

In the case of factor concentrates in Canada, there is a chain of communication. The manufacturer of the product informs Health Canada of the concern and what actions it has taken to deal with the problem. If Health Canada is not satisfied with the actions taken by the manufacturer, it can take action on its own. The manufacturer also informs the distributors of the product—either Canadian Blood Services (CBS) or Héma-Québec. To ensure notification, Health Canada also informs the distributors. CBS and Héma-Québec in turn contact the hospitals or HTCs to which the product was sent. Although CHS has no official role to play in the chain of communication, it sends copies of the recall information to HTCs and to its own chapters, as soon as it is informed. Finally, patients are notified by their doctors or nurses. This is often done by telephone, especially if it is an urgent safety issue. Staff will work until everyone has been notified.

When a HTC receives notification of a recall, the staff take it very seriously. Once notified by the distributor which product and lot numbers are affected, they check their records to see who received them. Nowadays, HTCs have computerized records. This means that it takes very little time to find out who received the products being recalled—that is, as long as the patient is registered in a HTC and the HTC is aware of the lot numbers distributed to him. The HTC staff will explain the concern to the patient and advise what to do. Often, replacement products will be issued. If necessary, information sessions will be organized to provide more explanations and answer questions.

People are often anxious when a recall occurs and it is important that they ask questions and get reliable information. If people are worried, they should contact their HTCs for information.

It is important, in case of a recall, that records with details of all infusions of clotting factor concentrates—date, product name and lot number—be immediately available. When a person receives treatment at a HTC or hospital, the doctor or nurse giving the infusion records this information. When products are distributed for home use by the staff at a HTC, the lot numbers are recorded. When a person is on a home infusion program, many clinics require that he (or a family member) record all infusions in the bleed diary provided by the HTC.

Withdrawals – A product withdrawal occurs when the manufacturer decides to remove the product from use. Although both Health Canada and the manufacturer consider the product to be safe and effective, the manufacturer feels that the product does not meet its own standards.

Quarantine – A quarantine occurs when a product is held back and not used for a short period of time because of a possible problem with its safety or efficacy. During this time, manufacturers and Health Canada investigate. If the problem is found to be real, the product is recalled. If it is found that there is no problem, the product is released from quarantine. Many readers will remember the “Utah donor” case in late 1998. Because of concerns over Creutzfeldt-Jakob Disease (CJD), some recombinant factor concentrates were quarantined for a period of one week. However, after investigation, Health Canada determined that there was no risk to safety. The products were released.

In hemophilia care, people have clotting factor concentrates in their homes. As a result, they have to be notified of a quarantine so as not to use the products. The staff at the HTC will advise patients to return them to the clinic in exchange for another supply. Usually, other products are used until the safety of the “held back” product is decided.

Recalls, withdrawals and quarantines are very different. But from the point of view of the patient on home care, they are hard to tell apart. That is why it is important to talk to the staff at the HTC. They can give the full story.
Standardized Barcodes for Coagulation Products

Bruce Ritchie, M.D., Chairperson,
Association of Hemophilia Clinic Directors of Canada

Over the last five years, the Association of Hemophilia Clinic Directors of Canada (AHCDC) has developed a secure computerized database program known as CHARMS (Canadian Hemophilia Assessment and Resource Management Information System). This program functions as an electronic chart, a system to track blood products, a database to collect summary data about blood product use, and as a tool for projections of future use. Great care and vigilance are taken by the hemophilia comprehensive care clinics to ensure CHARMS data are secure and available to the individual, as required by Freedom of Information and Privacy Protection (FOIPP) legislation. The product tracking system of CHARMS has become a key part of the Canadian blood system, functioning securely and seamlessly in the recall of coagulation products. It can also be used to track the outcomes of blood products to look for more subtle problems that may not be apparent in individual people receiving blood products. This post-marketing surveillance is not well done by anyone yet; CHARMS holds promise to do it well. In an effort to make CHARMS work more efficiently and smoothly, we are investigating the use of standardized barcodes on coagulation products for rapid and accurate entry of information into the CHARMS database, hospital blood bank databases and other databases—in fact, a standardized barcode could be used by anyone who handles these products, including consumers.

Barcodes were introduced in 1974 in an Ohio supermarket, and now 5 billion barcodes are scanned each day in 140 countries. These barcodes identify classes of products, which are entered into inventory databases. More sophisticated tagging is under development in the form of radio frequency tags, which can identify individual manufactured items. These chips are powered by a scanner, using radio signals or electrostatic charge. The scanner powers the chip, then receives the information stored in the chip. The advantage of these devices is that they do not need to be in a specific orientation, but only need to be brought close to the tag. As the price of the tags and scanners becomes economical, it will become possible to track products closely enough to link the supply chain from raw material right through to consumption. In preparation for this, a consortium of companies (Gillette, Proctor and Gamble, International Paper, Sun Microsystems, and the Uniform Code Council) has come together to develop a system, AutoID, to name trillions of items, and track them through the Web.

In the meantime, until such a system is in place, the tracking of coagulation products must use technology that is at hand. The barcode can function to track coagulation product distribution and consumption, identifying product in producer warehouses, distributor warehouses (CBS), hospital blood banks/pharmacies and patients’ homes. A system, known as ISBT 128, has been developed for red blood cells, although implementation has been slow and the distance tracked is much less than with coagulation products. Barcodes are common on coagulation product boxes, although the format of the code and the information stored is variable from company to company, and in general does not include the critical information required for tracking, such as lot number, and expiry date.

With support from Health Canada, the AHCDC has organized a meeting of the blood product industry, regulators, Canadian Blood Services and other interested parties to establish a standard format of information labeling on coagulation products for automated/electronic data capture. Encoded data will include: product type, product name, lot number, unit size and expiry date. Once a common format is agreed upon, such data will be encoded on the product boxes and vials in barcode, radio frequency chip or using any other technology that is agreed upon—the data will always be in the same format so that the particular system is not critical. For now, a standard for barcode will be agreed upon. The ongoing harmonization of European, American, and other regulations provides a unique opportunity to set a global standard, leading to global success at monitoring production, distribution and use of these expensive, time dated, and critically needed products. Tracking these products and their use is critical to security of supply, and the safety and efficacy of these products.

Q&A

Third-Generation Recombinant Factor VIII in Clinical Trials

This column has previously reported on the development of new recombinant factor concentrates such as BeneFIX® (summer 1999), Kogenate® FS (winter 2000), and ReFacto® (fall 2001). This issue will focus on the third-generation recombinant factor VIII concentrate manufactured by Baxter BioScience currently in clinical trials. The product is called rAHF/PFM which stands for recombinant Anti-Hemophilic Factor/Protein Free Method. The name under which rAHF/PFM will be marketed has not yet been approved.

Hemophilia Today interviewed Dr. Gordon Bray and Mr. Michael Hamilton, both of Baxter BioScience. Dr. Bray is the company’s Global Medical Director for the rAHF/PFM development program and works in Glendale, California. Before joining Baxter, he was Director of the Coagulation Disorders Program at the Children’s National Medical Center in Washington, D.C. Mr. Hamilton is Director of Sales and Marketing with Baxter BioScience in Mississauga, Ontario. He has worked in a number of different divisions of the company over the last 10 years.

Hemophilia Today: You refer to rAHF/PFM as your “next-generation” product. Our readers have heard about first-, second- and third-generation recombinant products. Could you explain the differences?

Gordon Bray: First-generation products were the first to be licensed. These were Kogenate and Recombinate. First-generation products were stabilized with pasteurized human albumin. People refer
to second-generation products as those that do not contain proteins of human or animal origin as stabilizers in the final formulation. However, these second-generation products, like Kogenate FS and ReFacto, do include pasteurized human albumin as a component at one or more stages of the production process. Third-generation products, like rAHF/PFM, are products made without the addition of human or animal proteins at any stage in the production process from cell culture medium all the way to final formulation. We refer to rAHF/PFM as our “next-generation” recombinant factor VIII.

**Hemophilia Today:** If albumin is no longer used as a stabilizer in the final formulation, what has replaced it?

**Gordon Bray:** We used a combination of sugar, salts and amino acids. We believe this will meet our stability requirements—storage at 5°C for longer than 18 months.

**Hemophilia Today:** You say that rAHF/PFM is made without the addition of human or animal proteins at any stage in the production process. Does this mean that there are no human or animal proteins at all used in the processing of rAHF/PFM?

**Gordon Bray:** When I say “no addition of protein”, I am not referring to proteins derived from Chinese Hamster Ovary (CHO) cells, which are the cells that express human factor VIII in cell culture. Even this new product continues to contain trace amounts of CHO cell protein and mouse antibody left over from the purification process. These have been present in the Recombinate product throughout its 14-year history and there have been no unanticipated safety issues associated with the presence of these low-level proteins.

**Hemophilia Today:** Does rAHF/PFM contain bovine protein?

**Gordon Bray:** No. In rAHF/PFM that, too, has been removed.

**Hemophilia Today:** How have you gotten around the addition of albumin to the cell line with rAHF/PFM?

**Gordon Bray:** We use a proprietary cell culture medium which has allowed the cell line to adapt to an environment that does not require human or animal protein.

From a genetic standpoint, the cell line is identical to that used to make the Recombinate product.

**Hemophilia Today:** Does that mean that the factor VIII protein is identical to the protein in Recombinate?

**Gordon Bray:** All of the testing we have done so far including biochemical comparisons as well as toxicology, pharmacokinetics, hemostatic efficacy in relevant animal models reveals that the two proteins behave in a virtually identical manner.

**Hemophilia Today:** Have human clinical trials for rAHF/PFM begun?

**Gordon Bray:** The clinical trials began a little over a year ago. We are conducting a number of them. Enrollment in our pivotal study of pharmacokinetics, neo-antigenicity, safety and efficacy in previously treated patients (PTPs) was completed in June 2001. There are 111 PTPs enrolled. We’re also doing a continuation study which allows patients in the pivotal study to continue with rAHF/PFM until regulatory action is taken on the license application. We’re doing a surgery study, and soon, a pediatric study and a previously untreated patients (PUPs) study.

**Hemophilia Today:** Are there any Canadian patients enrolled in these studies?

**Michael Hamilton:** Yes, there are currently three Canadian sites participating in the surgery study—Dr. Bruce Ritchie’s clinic in Edmonton, Dr. Houston’s program in Winnipeg and Dr. Irwin Walker’s program in Hamilton. The second trial in which Canadians will participate is the pediatric study. This has not yet begun. It is designed to look at the pharmacokinetics, safety, efficacy and neo-antigenicity in children under 6. The pediatric centre involved is the Hospital for Sick Children in Toronto under the direction of Dr. Victor Blanchette.

**Hemophilia Today:** Are there any preliminary results of these trials?

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**SUGGESTIONS ON THE MANAGEMENT OF CARE FOR WOMEN WITH BLEEDING DISORDERS**

*New Guidelines Published*

by Patricia Stewart

Because women were considered simply ‘carriers’ of the gene for hemophilia, specific medical problems that many of these women endure due to this condition went undiagnosed and untreated. Women with von Willebrand Disease (VWD) are often treated by their local doctor or gynecologist who has little or no experience with hemostatic problems. However, since 1995, great strides have been made and many women are now registered at Hemophilia Treatment Centres (HTC). Two HTCs have official clinic programs designed specifically for women with bleeding disorders: Hôpital Ste-Justine in Montreal and Hôpital du St-Sacrement in Quebec City.

To meet the growing demand for treatment by women with bleeding disorders, especially vWD, the Association of Hemophilia Clinic Directors of Canada (AHCDC) created the Women with Bleeding Disorders Subcommittee. This group has developed a document entitled Suggestions on the Management of Women with Bleeding Disorders to use as a basis for treatment. The members of this group, led by Dr. Christine Demers from Hôpital du St-Sacrement, include Drs. Georges-Etienne Rivard, Michèle David, Mary-Frances Scully, David Lillicrap, Sara Israels, Bernadette Garvey, Linda Vickars and Diane Francoeur.

The introduction to the document reports on the prevalence and consequences of hemostatic problems for women. This document is offered as “a practical approach for physicians caring for these patients”, but not as a complete review of the subject. Nonetheless, it can be used as a working document for Hemophilia Treatment Centres wishing to offer services specifically designed for women. There are four sections to the document: creation of a clinic, lab tests, medical treatment of menorrhagia, and the management of pregnancy in women with bleeding disorders.

**SETTING UP A MULTIDISCIPLINARY CLINIC FOR WOMEN**

The importance of a multidisciplinary team is stressed. As a minimum, a nurse, a hematologist and a gynecologist are required in the basic team for clinical consultation. Regular meetings of the team should occur to discuss patients and standardise approaches. The ideal multidisciplinary team has a broader representation of expertise, with a laboratory hematologist, an obstetrician-gynecologist, an anesthetist, a family physician, a pharmacist, a laboratory technician and/or other interested health professionals. Designated secretarial support is essential as well.

All patients should be referred by a physician. Before any coagulation investigation, patients should have a complete personal and family history and physical examination, including a gynecological examination. A woman’s perception of her menstrual flow is often not reliable. The introduction of a graphic scoring system for menstrual bleeding has resulted in a more accurate
LAB INVESTIGATIONS

The document outlines the types of coagulation tests necessary for proper diagnosis, ranging from a simple PT to the rarer factor tests. The procedure for these tests is explained, as well as the difficulties in sample taking and exterior factors that affect the results of these tests.

MEDICAL TREATMENT OF MENORRHAGIA

“Management needs to be individualized and is best undertaken in a coordinated fashion by both hematologists and gynecologists. This approach should result in a substantial reduction in hysterectomies and a significant enhancement in the quality of life.”

This statement is the essence of the problems that women with bleeding disorders often have to live with. The effect of a bleeding disorder on a woman’s quality of life is underestimated.

This section proposes treatment options for menorrhagia. A list of medications and their side effects are included. However, surgical options are not addressed here.

MANAGEMENT OF PREGNANCY IN WOMEN WITH BLEEDING DISORDERS

From conception to post partum, women with bleeding disorders should work closely with both their hematologist and obstetrician. It is important that women with bleeding disorders deliver in a hospital where there is access to hematologists, obstetricians and pediatric consultants. The document covers treatment options both during delivery (including epiduals) and post partum, as well as precautions for the newborn.

The Suggestions on the Management of Women with Bleeding Disorders will be available through CHS. It can be used as a working document for Hemophilia Treatment Centres and will also be distributed to gynecologists or hematologists who participate in continuing medical education sessions dealing with this topic.

CHS Hepatitis C and HIV Educational Workshop

In the Summer 2001 issue of Hemophilia Today a young man living with hepatitis C shared his impressions of the first Canadian Conference on Hemophilia C. Since that conference, the CHS Youth Committee has been working on a plan for a hepatitis C workshop aimed specifically at young people infected with hepatitis C. This workshop, entitled CHS Hepatitis C and HIV Educational Workshop, will address some of the key issues that were brought to light during the hepatitis C conference and will also address many issues surrounding co-infection with HIV. It will be held June 6, 2002 to coincide with the CHS Annual General Meeting in Saskatoon. The workshop will transmit information specifically applicable to hemophiliacs infected with hepatitis C and HIV.

One of the issues that the workshop will cover will be that of fertility options and sero-discordant couples. (See the Fall 2001 issue of Hemophilia Today for a report on research on this question.) The issue of anti-viral regimens for individuals infected with HIV and their impact on the progression of liver disease associated with hepatitis C will be discussed. A session on diet and natural therapies will also be given.

Many hemophiliacs infected with hepatitis C (and HIV) also face additional life-stressors. Many life-stressors arise from trying to balance treatment regimens with one’s schooling or career and the constant struggle to ensure that one’s liver stays as healthy as possible for as long as possible. This can be challenging, especially since consumers are often subjected to conflicting medical information. In order to address some of the issues surrounding positive disease management, the CHS Youth Committee will be exploring the possibility of incorporating psycho-social sessions to complement the medical and scientific presentations that will be made during the workshop. Many of the topics at the 2002 Youth Hepatitis C- HIV Co-infection workshop will be derived from recent needs assessments completed by CHS, provincial chapters and/or regions.

In the Spring of 2001 Hemophilia Ontario completed an HIV-HCV co-infected client needs assessment. This needs assessment was different from the one funded by Health Canada. Its aim was to ensure that programs and services offered by Hemophilia Ontario were appropriately addressing the issues of HIV and hepatitis C co-infection for members of Hemophilia Ontario. One of the primary needs identified was that of quality counselling for individuals living with multiple medical conditions. The other issue that emerged from the Hemophilia Ontario AIDS Advisory needs assessment was that of fertility options for sero-discordant couples. Hemophilia Ontario will soon be embarking on a research project to address the issues of fertility treatments and options for sero-discordant couples. Counseling and fertility options have been designated as priority issues for young hemophiliacs living with hepatitis C and HIV co-infection.

This summer was yet another busy one for Hemophilia Ontario youth activities. The Hemophilia Ontario Youth Committee hosted another successful summer canoe trip. This year the canoe trip brought together individuals from outside Ontario. It is fantastic to see youth from across Canada sharing and participating in youth events that were previously limited to young hemophiliacs residing only in Ontario.

The Quebec Chapter of the CHS is embarking on developing a Youth Working Group to address the issues of youth with hemophilia in that province. The format would be similar to the one already established by the Hemophilia Ontario Youth Committee. The lead individual responsible for the Quebec Youth Working Workshop is already thinking about establishing a partnership between the Ontario and Quebec groups.

If you wish to comment on a specific item that you have read about, or wish to become involved in a youth program planning process, please do not hesitate to contact me at this e-mail address – karttik.shah@sympatico.ca.
Mongolia to Ukraine to China

What does it mean to be a responsible “World Citizen” as a hemophilia organization in our global society? The exact answer is still elusive, but the International Projects Committee hopes to find some answers to give guidance to both CHS and possibly other “First World” countries. We’ve been given much and so we have greater responsibility to help those who have less. That’s what twinning is all about!

After an exciting first assessment trip to Mongolia, reported in the Fall 2001 Hemophilia Today, Hemophilia Saskatchewan is doing the “grunt work” of twinning—trying to develop an action plan. We will be submitting this to the Mongolian Association of Hemophilia for their input, and then submitting it to the World Federation of Hemophilia for their official approval. We’re hoping that what we can offer will result in improved care and greater life quality and expectancy for those born with hemophilia in Mongolia.

Hemophilia Manitoba’s Joel Hershfield, a volunteer, is leading their efforts in establishing a twin with the Hemophilia Society in the Ukraine. They are in the process of submitting their application to the WFH and are hoping for a first assessment visit in late summer. Accompanying Joel will be a physiotherapist, Greig Blamey. Greig will also be attending the 2002 World Congress in Seville, Spain and will have a chance to meet some of the Ukrainians there.

If your hemophilia chapter is interested in exploring the opportunities of twinning, and doesn’t know where to start, request the publication entitled, Reaching Beyond Our Borders from the World Federation of Hemophilia (Tel: 514-875-7944 or E-mail: wfh@wfh.org) or download it in PDF format from their web site: www.wfh.org in the Publications section. After reading this you’ll have a good idea of what’s involved.

Then contact Karine Frisou, Program Officer at WFH (514-394-2818) with your thoughts and ideas. She’ll be delighted to assist you in finding an appropriate twinning partner.

CHINA CONNECTIONS

Late November found Dr. Man-Chiu Poon (Calgary) and Dr. Brian K.H. Luke (Ottawa) in Guangzhou, China and beyond. They were there continuing to facilitate their own clinic twinnings (Calgary – Tianjin and Ottawa – Guangzhou) and create an even greater network of cooperation between various treatment centres in China. Their efforts have produced some very promising results.

November 24-26 was the Guangzhou Hemophilia Conference with attendees including 14 physicians from centres in Beijing, Tianjin, Hefei, Jinan, Shanghai, Guangzhou and Hong Kong as well as centres from outside China, namely Ottawa, Calgary and London (UK). Also attending were the Chair of the Hemophilia Union of China, Prof. Z. Han, and 5 patient leaders of the Hemophilia Home of China.

In summary, three needs were targeted for immediate attention: 1) accurate diagnosing, 2) hemophilia nurse training and 3) a national patient registry. A very important resolution was also reached: unanimous support to form a network of Hemophilia Treatment Centres in China!

This will be a pivotal milestone in the development of hemophilia care in China.

Following this conference, Drs Poon and Luke visited centres in Shanghai, Tianjin and Beijing. This was enabled by a grant from the CHS International Projects Committee. Their purpose was threefold: 1) to better understand the expertise and needs of the centres and explore how the priorities established at the Guangzhou Conference could be implemented, 2) to facilitate the working of the newly established network and 3) to move hemophilia care forward in China.

They determined that the Shanghai Hemophilia Centre should be brought in to a twinning relationship fairly quickly and be recognized as a Centre by the WFH. They would prefer a Canadian centre (anyone out there interested?) to better coordinate efforts in China. A current proposal is to jointly twin Calgary and Ottawa until another Canadian centre is found. Other recommendations included forming a committee with hemophilia physicians from other key centres to begin to establish a registry and encouraging the Physical Medicine and Rehabilitation Department in Beijing to develop expertise in hemophilia physiotherapy and rehabilitation care and then go on to become a trainer for hemophilia physiotherapy for other Chinese hemophilia centres.

There were many other exciting accomplishments and future plans that include a nursing exchange, coagulation laboratory practice and management training, participation of Chinese doctors in WFH fellowships and more. But the real fulfillment is knowing that all these efforts contribute to the quality and longevity of life for those with hemophilia in China. Certainly this is a vital part of what it means to be a Global Citizen in the world hemophilia community—little by little doing what we can to equalize hemophilia care around the world. Thanks, Dr. Poon and Dr. Luke, for all the extra effort you put forward on behalf of those without the level of care we receive here in Canada. You leave a lasting legacy of benefit to those in need in China.
Send a Postcard to your Minister of Health (see enclosed postcard)
This issue of Hemophilia Today has focused on the importance of maintaining or enhancing comprehensive care for people with bleeding disorders available through Hemophilia Treatment Centres.

Some articles have described examples of wonderful success stories, where care for people with bleeding disorders rivals the best in the world. Other articles have described clinics where, despite the best efforts of dedicated health professionals, care does not meet the high standards people deserve. Still other articles have documented the dedicated efforts of chapter volunteers and health care providers to convince provincial governments to improve services.

The postcard inserted into this newsletter is an easy way to have your voice heard. If you think the services in your comprehensive care centre need to be improved, write to your Minister of Health or MLA. Tell him/her what you think needs to be changed or improved.

These are some of the ideas presented in the various articles. Some of them may apply to your clinic.

- Dedicated funding
- Official designation by the provincial government
- Increased funding
- A full-time Nurse Coordinator
- Attracting young hematologists and Research Fellows to ensure long-term medical expertise
- More access to a trained physiotherapist
- More social work services
- Services for women with bleeding disorders
- Having all services under the same roof
- More resources to do outreach
- Better care at the ER
- Tracking of coagulation products by clinic personnel
- A standard of care in your centre that is comparable with other clinics in Canada
- Improved lab services in the same hospital as the clinic
- An annual audit by people knowledgable about hemophilia
- More access to prophylaxis
- Access to specialized resources
- An end to cutting costs

Here are the names and addresses of the current Ministers of Health across the country.

**BRITISH COLUMBIA**
The Honourable Colin Hansen
Minister of Health Services
PO Box 9050
STN PROV GOVT
Victoria BC V8W 9E2

**ALBERTA**
The Honourable Gary Mar
Minister of Health and Wellness
Legislature Office
#323, 10800 - 97 Avenue
Edmonton, AB T5K 2B6

**SASKATCHEWAN**
The Honourable John Nilson, Q.C.
Minister of Health
Room 361
Legislative Building
Regina, Saskatchewan S4S 0B3

**MANITOBA**
The Honourable Dave Chomiak
Minister of Health
Room 302
Legislative Building
Winnipeg MB R3C 0V8

**ONTARIO**
The Honourable Tony Clement
Minister of Health and Long-Term Care
Hepburn Block, 10th floor
80 Grosvenor Street
Toronto, ON M7A 2C4

**QUÉBEC**
Monsieur François Legault
Ministre d'État à la Santé et aux Services sociaux
1075, chemin Sainte-Foy, 15e étage
Québec (Québec) G1S 2M1

**NEW BRUNSWICK**
The Honourable Elvy Robichaud
Minister of Health and Wellness
P.O. Box 5100
Fredericton, New Brunswick E3B 5G8

**NOVA SCOTIA**
The Honourable Jamie Muir
Minister of Health
Province House
1726 Hollis Street
Halifax, NS B3J 2Y3

**PRINCE EDWARD ISLAND**
The Honourable Jamie Ballie
Minister of Health and Social Services
Second Floor, Jones Building
11 Kent Street
PO Box 2000,
Charlottetown, PEI C1A 7N8

**NEWFOUNDLAND**
The Honourable Julie Bettney
Minister of Health and Community Services
Confederation Building
P.O. Box 8700
St. John's, NFLD A1B 4J6