XXIII International Congress of the World Federation of Hemophilia

From May 16-22 more than 2,500 people gathered in the Netherlands for the XXIII International Congress of the WFH. Despite a last minute change in location from Jerusalem to the Hague due to security concerns, the Israeli Hemophilia Society worked with the Netherlands Society to make this event a success. Dan Doran, Erma Chapman and myself had the privilege of attending this congress.

Sessions dealing with hepatitis, prophylaxis, gene therapy, HIV treatment, inhibitors, NMO presentations, product safety and numerous other topics were offered each day. The amount of information available at the Congress was a bit overwhelming. It was difficult to know where to begin. Each time period brought the choice of four to six different topics as well as other special presentations. There was also an exhibit with over 690 abstracts on every topic imaginable.

Over 80% of all people in the world with hemophilia have little or no treatment. Some die from a hemorrhage that would be a mere inconvenience here. Hospitals and medical treatment are available to the privileged few and even they have little access to concentrates. Twinning programs are one of the main projects of the WFH. Dr. Man Chiu Poon from Calgary, presented his project working with a clinic in China. People from the Quebec chapter met with people from the Senegal Association to begin working together. At this point, each twinning project between treatment centres or hemophilia organizations is worked out by the partners, sometimes with a great deal of misunderstanding and disappointment. Political, cultural and everyday realities are not self-evident. Poorer countries have high expectations and hopes. While developed countries have the best services and products available, they cannot simply give them away. Lack of facilities, travel and illiteracy are major problems in many parts of the world. Guidelines for twinning programs must be created so that each partner will know where to begin and what to expect from the other. This congress gave me hope for the future of treatment for people with bleeding disorders, but also the knowledge that we live in the privileged part of the world where this treatment is a reality. Let us share the wealth of our knowledge and experience.

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Canada Still Needs Blood

Events over the summer have meant large changes to Canada’s blood delivery system. As had been promised nearly since the release of Justice Horace Krever’s Report on Canada’s sick blood system, a new agency has been put in place to handle our national blood and blood product needs: Canadian Blood Services. (This agency will handle all the blood needs for Canada, except for Quebec, who will have her own organization, Héma-Québec) The Chief Executive Officer (CEO) of the CBS is Lynda Cranston, who comes to her new position from a career in both the public and private corporate world. The Executive Director of Héma-Québec is Dr. Francine Décary, who, prior to her recent nomination, held the position of Director General of Transfusion Services of the Red Cross for the Eastern Canada region.

These organizations’ lives will be full of opportunities as they try to bring reason into a public service organization that has increasingly exhibited questionable decision making practices over the past two years or so. But their lives will also be full of challenges, some of them huge.

We fear that the potentially largest problem they will face the instant they take on the responsibilities of Canada’s blood system is the issue of declining donor numbers. Over the past two years, the Canadian Blood System (under the Canadian Red Cross) has lost approximately one quarter of its donors.

Many of the other issues facing the new CBS are covered in Dr. Durhane Wong-Rieger’s article “A Consumer Perspective on the Canadian Blood Services”, printed in the Special Issue of Hemophilia Today on the Blood System, sent out with the current issue of Hemophilia Today; we urge you to read it. And we urge you all to think seriously about what we as members of the larger family of Canadians who depend on blood or blood products might do to help to alleviate this donor crisis - for it is certainly a crisis. Just the other night your Editor saw a public service television commercial dealing with this very issue, making the point that donors are still the central focus of Canada’s blood system. Without them there is no blood programme, no blood transfusions to stave off certain death after car accidents, no treatment for anaemia or leukemia. And while many hemophiliacs no longer need blood or blood products for their care, generally speaking, we are still members of that great Canadian family.

So ask yourselves, what can you do to help build the blood donor population in Canada? What can we do to make sure that the system that helped keep us alive for so many years, before it became something like an infectious agent itself, continues to provide life saving service? We’re all in this together, and unless we all work together to see the blood service grow, we will all face serious accusations if it fails. No one will be able to avoid the fingers of blame.
As I write this message, we have no idea what is going to happen at the Premiers’ Conference in Saskatoon. I continue to be optimistic that this time they’ll get it right, and everyone will be included in an HCV compensation package. As you read this message, you will know what happened at that meeting - and probably at a few other meetings too! As well as HCV compensation, we will all be watching the implementation of two new blood systems in our country, with anticipation and hope for safety and efficacy from each system. Never again, however, will we completely trust, Canada’s blood system, nor will we abdicate our more recently evolved role as watchdog.

Since this is my first message as President of the CHS Board, I would like to do three things: thank those who have worked so diligently as part of the previous Board; welcome the new Board of Directors; and, introduce myself.

Durhane Wong-Rieger is a truly amazing person. Over the past four years as President, she has worked with the Board, other volunteers and staff members to achieve higher quality blood and factor replacement products, an accounting of the past tragedy that has affected so many of our lives, and the establishment of new blood systems for Canada - systems that remarkably resemble the CHS vision for blood services. While each of us owes her an unpayable debt of gratitude for the changes to the larger systems, it has probably been the laughter and tears we have shared with her in our homes, in hospital rooms and at family weekends that we will remember most fondly and readily. Durhane is continuing as part of the new Board of Directors. In her role as Past President, she will still be sharing her expertise, her personal dynamism and her compassion with us.

We have said adieu to other Board volunteers. David Page has “retired” from the Board, but continues to co-chair our blood products/system committee. Ken Little has stepped down after several years as a member and leader of the Board and Executive, in order to give more time to family and career. While Mark Brown is no longer at the Board table, he continues in his efforts to achieve HCV compensation for all those injured through the blood system. We will miss James Kreppner at the Executive and Board meetings, but he will continue to represent us as part of national AIDS endeavors. Tanis Steffens will be missed from the finance area, but she will be busy with her new family over the next couple of years. Marie Jutras has been an unwavering advocate of the importance of grassroots representation and responsibility in the organization. Ghislaine Landreville’s passion about our responsibilities as board members to raise money will be remembered even though she has stepped down from the national board. Dr. Irwin Walker has also finished his term as the MSAC representative to the CHS Board; we will certainly miss both his professional advice and his dry wit.

As you can see, a great deal of talent has left us and we will miss each of these people. We wish each of them all of the best, and we will see you at future CHS events!

Left behind, however, is not only what our colleagues have taught us, but new and renewed energy, ideas and talent. At the Board table, we welcome to the team Blair Myers (Alberta), Frank Figler (Manitoba), John Plater (Ontario), Sheila Comerford, Patricia Stewart (both of Quebec), and Ron McLeod (Prince Edward Island) as

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Carrier Testing: Why, When and Where

**Why Test**
The factor level in any woman who is a carrier is determined by an event called lyonization (X-chromosome inactivation) that occurs after conception and after the fertilized egg has multiplied to several cells. No one knows what affects which X-chromosome will remain active and which one will be inactivated. Studies show that 30%-50% of carriers of hemophilia A and B have problems because of low factor levels. There is no difference in average factor levels in carriers who get the hemophilia gene from their father versus those who get it from their mother. The most typical levels of factor VIII or IX in carriers are between about 30% and 70% of the average normal level. While this is enough for normal coagulation, symptomatic carriers can experience bleeding episodes, since they have mild hemophilia, when confronted with surgery or trauma. In girls, however, the monthly menstrual cycle may be the only event needed for symptoms to be recognized. Abnormally heavy or long menstrual periods are common complaints among carriers and girls with bleeding disorders.

With von Willebrand’s Disease, the types can vary within the same family. Quantitative and qualitative platelet disorders can mimic the symptoms of von Willebrand Disease. At times, members of the same family will each be diagnosed with different disorders and types. Type 3, where no vW factor is produced by the body, is treated like severe hemophilia. While some clotting factor deficiencies cause few if any symptoms (such as factor XII deficiency), others are especially troublesome for women since they can cause miscarriage.

It is important for the child’s safety to know her factor level. Carriers with low levels, symptomatic carriers, could be at risk at times of tooth extraction, trauma or surgery. While the onset of menstruation is a critical time, heavy menstrual cycles are sometimes accompanied by extreme pain and anaemia. Painkillers containing non-steroidal agents (such as Advil) and acetacylic acid (Aspirin) cause platelet problems. Girls could avoid stressful situations due to staining, painful episodes and medical problems if their status was known beforehand and treated.

**When to Test**
The daughter of a man with hemophilia is an obligate carrier and there is no need to do any genetic tests to prove this. Hemophilia breeds ‘true’: that is to say, if an affected family member has mild hemophilia, then other family members can only inherit a mild form, and there is no chance of transmitting a severe form. However, factor levels in an obligate carrier remain unknown until testing is done. Within a family, the factor levels in carriers can be very different, ranging from very low to very high. There never appears to be a “right age” to test for potential carrier status. Many families tend to delay testing as a form of denial, or to protect their child (and themselves) from “bad news”. Others test at a young age, letting the child grow up with the knowledge of her status. Still others feel it is not important since the girl has no problems. When asked whether girls under five should have their blood levels tested, only 28% of geneticists said yes compared to 67% of hematologists. There has been concern expressed about the ethical issues of screening children for conditions which are not immediately life-threatening. It is sometimes suggested that all such testing should...
be deferred until adulthood, when a woman is in a position to give full and informed consent. However, early testing allows time for the girl to come to terms with the complex nature of being a carrier.

Leaving carrier testing until the girl has grown and is in a relationship or until she is ready to conceive or already pregnant creates unnecessary stress. Not only will she have to come to terms with her carrier status and its resultant choices, but all this must also be done urgently. All too often a woman will be referred to determine carrier status in an advanced state of pregnancy, despite a well-known family history of hemophilia. Genetic testing for carrier status and genetic counseling is best done before pregnancy. Genetic studies take time to carry out, in large part because it is often necessary to track down and obtain blood samples from other members of the family. The psychological stress and decisions of prenatal diagnosis can be better dealt with if the woman is not faced with both the announcement of carrier status as well as the decisions for prenatal diagnosis of the fetus.

Because of a surge of hormones involved at the beginning of menstruation, there is a tendency for excessive bleeding at the first or second menses, sometimes requiring emergency room treatment. Dr. Gillian Oliver, a pediatric gynecologist from Toronto, has found that many young girls presenting with juvenile uterine bleeding (JUB) in the emergency room have a coagulation disorder. She suggests “anticipatory gynecological management” for young girls. A 1998 study in Moscow showed 34% of girls with JUB to have a coagulation disorder. Testing could be done at the first signs of breast development in a young girl. Another benefit of early testing is that the child can be vaccinated against hepatitis B in case she should ever need factor treatment in an emergency situation. A questionnaire for women with bleeding disorders done by Diane Wysocki found that the average age of a first bleeding episode was nine years of age and their first visit to a gynecologist was at the age of 14 instead of at 18-20 years for the average woman. Most were not tested for blood disorders until they were 25 years old and 42% had had a hysterectomy.

Many factors can affect the results of blood assays. Pregnancy, birth control pills and breast feeding raise hormone levels in some women. A woman should refrain from taking birth control pills for a month before being tested. Crying, stress, pregnancy, contraceptive pills, breast feeding, exercise, recent transfusions and recent infections can all affect the results of a blood assay. While it is impossible to avoid all of these influences, they should be taken into consideration. The time to get tested is at the time of menstruation (period) when hormone levels are lowest. In the case of Factor IX carriers however, clotting levels are not affected by hormones and testing can take place at any time.

Where to Test
The word “hemophilia” is frequently used rather loosely by non-specialists to describe an apparent bleeding tendency. While some labs will do testing for clotting times, Hemophilia Treatment Centres are the place of choice for comprehensive testing. They have the knowledge and experience to do proper testing for carrier status and to deal with the actual treatment of bleeding disorders.

Oral contraceptives are regularly used to manage heavy periods in some girls. However, for adolescents this treatment carries psychological and social implications. Moral problems may arise from this treatment. There are no easy answers. Support groups for young girls would help break the isolation and embarrassment they deal with on a monthly basis.

While most girls will not need medication to treat their low factor levels, there are some practical things they can do to prevent problems. In Factor VIII and vWD-Type1, high hormone levels affect the quantity of clotting factor and thus impair coagulation. Knowing this, girls can arrange dental appointments (or any medical intervention) to coincide with their mid-cycle (ovulation) when hormone levels are at their highest. Exercise raises hormone and clotting levels. Pain is a common problem for girls both at the time of their period and at the time of ovulation. Data exists to show that two anti-inflammatory agents, choline-magnesium-trisalicylate (Trilisate) and salsalate (Disalcid) do not interfere with platelet function and therefore are viable options for use by persons with bleeding problems. These agents are frequently helpful in controlling the pain associated with ovulation and menstruation. The hematologists at local Comprehensive Care Centres are the persons who will prescribe any treatment necessary to counteract bleeding problems.

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Once again this year the Hemophilia Research Fund (HRF) approved the recommendations received from the Hemophilia Research Grants Review Committee. This year, with funding generated by the Hemophilia Million Dollar Club the following new grants were awarded:

**Gene Therapy Vectors based on the Factor IX Promoter:** Dr. Christopher Mueller, Queen’s University, Kingston, Ontario

**Non-Autologous Somatic Gene Therapy for Hemophilia in Mice:** Dr. Patricia Chang, McMaster University, Hamilton, Ontario

**Studentship Award: Signalling in von Willebrand Factor Secretion:** Ms. June Ma, Queen’s University

We are pleased to present progress reports on the 1998 research projects.

**Gene Therapy Vectors Based on the Factor IX Promotor**

*Dr. Christopher Mueller  
Queen’s University, Kingston, Ontario*

Through our studies of a form of hemophilia B (factor IX) called the Leyden mutations, we have developed a detailed understanding of that part of the Factor IX gene which directs its expression. Factor IX is made exclusively in the liver, and the promoter which directs its expression has many characteristics which makes it potentially very efficient for expressing any gene in this organ. Some of the main problems in gene therapy are related to getting a given gene to be expressed at high levels, for it to be expressed in the correct organ, and for its expression to persist for as long as possible. Using our knowledge of the Factor IX promoter we have designed a series of constructs which could be used to express the clotting factor VIII and IX genes in the liver. In this project we will be testing these constructs in several model systems, including cultured human liver cells and in mice. These experiments will allow us to identify what elements are important for this process, and to create more efficient ways of doing gene therapy.

Gene therapy is probably the best hope for a permanent and safe cure for hemophilia. By replacing the gene which is abnormal with a normal gene, the patient’s own body is able to produce the protein which it needs. In the case of hemophilia A and B, it is the clotting factor VIII and IX genes which are affected. Both of these genes are expressed in the liver, which then makes proteins and releases them into the blood. Thanks to a large amount of research which has been done in the past, it is becoming practical to deliver a normal copy of the clotting factor genes to the liver. This has been accomplished using modified viruses as well as DNA in different forms. The main problems that block the practical use of these systems for this therapy is having the gene expressed only in the liver, at high enough levels to be useful and for long enough to be convenient. The elements which control this type of function are called promoters, and normally ensure that a gene is expressed appropriately.

We have spent several years studying the promoter of the clotting factor IX gene in an effort to understand the basis for the Leyden form of hemophilia B. This is an unusual form of hemophilia where the children are severe hemophiliacs. However, as they become teenagers their levels of clotting factor IX increase, until they are almost normal. The mutations which cause this are in the factor IX promoter, rather than in the gene itself, which is the case for the majority of hemophiliacs. Through our studies of this form of the disease we
discovered a great deal about how the Factor IX promoter works. We then thought that for carrying out gene therapy for Factor VIII and IX hemophilia, using the natural promoter would be a big advantage as it would ensure that these genes were expressed in the way that they should be. However, because it is only possible to get a normal copy of these genes into about one liver cell in ten, we had to alter the structure of the factor IX promoter to make it stronger.

We have made a number of different types of promoter constructs, because we can only make educated guesses about what is important. In this project we will be testing these different constructs using cultured human liver cells and mice to determine what is important. This will allow us to design even better constructs in the future; it will also tell us which of the many processes involved in getting these proteins expressed are critical. By using our knowledge of the Factor IX gene we hope to develop a critical aspect of gene therapy, which can then be incorporated into existing delivery systems, and which will lead to a practical treatment for both hemophilia A and B.

Non-Autologous Somatic Gene Therapy for Hemophilia in Mice

Dr. Patricia Chang
McMaster University, Hamilton, ON
Our long-term objective is to treat hemophilia in humans by developing a safe, efficacious and cost-effective form of somatic gene therapy. We propose to implant into different recipients the same universal cell line engineered to secrete recombinant coagulation factor. These cells are protected from immune rejection by enclosure in biocompatible microspheres that allow the exit of coagulation factors but not the entry of immune mediators responsible for tissue rejection. This strategy will now be tested out in transgenic hemophilia B mice which mimic the human bleeding condition. Recombinant mouse myoblasts are engineered to secrete Factor IX and implanted into these mutant mice under various experimental conditions to assess the efficacy of this novel approach to the treatment of hemophilia.

The budget for this grant will be used in three main projects:

**Creating Factor IX expression vectors to transfect mouse myoblast cells.** Elements known to be important in controlling gene expression will be engineered into the vector encoding the cDNA for factor IX. These constructs will be used to transfect mouse C2C12 myoblasts to obtain clones secreting high levels of factor IX.

**Treat hemophilia B mice with encapsulated Factor-IX expressing cells.** The myoblast clones secreting high levels of factor IX will be encapsulated into microcapsules fabricated from a biocompatible polymer, alginate. These microcapsules will then be implanted intraperitoneally into hemophilia B knockout mice. Biochemical, functional, and clinical correction of the Factor IX deficiency will be monitored. Development of antibodies against the Factor IX may be expected in this strain of hemophilic mice, thus mimicking the human mutations that are associated with development of factor inhibitors. Titres of such antibodies will be monitored.

**Enhance the effectiveness of the above treatment with immune suppression.** Using the most clinically effective protocol developed above, we shall treat the hemophilic mice, but concomitantly, administer one of the three immune suppressive reagents known to be effective in suppressing inhibitor development. These reagents are anti-CD4 monoclonal antibodies, FK506 or cyclophosphamide. The biochemical, functional and clinical improvements will be compared with those without the immune suppression, to verify if there is reduced inhibitor development and improvement of clinical efficacy.

**Expected advances:** The proposed work on this mutant model of hemophilia represents our most exciting development towards the treatment of hemophilia with the non-autologous approach to gene therapy. The recent creation of the hemophilic-B knockout mice and their generous provision to us by our US collaborator, Dr. D. Stafford, now permit us to examine many of the issues related to the therapy of hemophilia in greater detail. Insight gained from this study will be important for the application of this method of gene therapy to humans.
MSAC Recommendation Regarding Hepatitis A vaccine

The CHS Medical and Scientific Advisory Committee (MSAC) recommends that individuals who are hepatitis A antibody negative receive hepatitis A vaccine, particularly those who have hepatitis C infection and those who might in future receive plasma derived concentrates.

Comment:

Individuals in the above groups need to have their hepatitis A antibody status determined, and receive the vaccine if their antibody status is negative. Individuals who are infected with hepatitis C should receive vaccine even if they are taking recombinant concentrates because hepatitis A infection may occur via other routes. The case for vaccination of individuals who are not infected with hepatitis C and who are taking recombinant products is weak; however, such individuals might conceivably receive plasma derived products if recombinant product is not available.

History

In 1993 MSAC recommended that “those individuals negative for HAV-IgG should be vaccinated with hepatitis A vaccine when this becomes licensed and available in Canada”. This recommendation was driven by the outbreaks of hepatitis A in Europe transmitted by plasma derived concentrates. In 1994 the vaccine became available and the National Advisory Committee on Immunization (NACI) included persons with hemophilia among those who they recommended should receive it. A letter was sent to CHS by the MSAC announcing the availability of the vaccine, and recommending that a) this information be disseminated to members and b) that the society consider lobbying for its funding. An article on hepatitis, including information about the hepatitis A vaccine appeared in the 1995 spring issue of Hemophilia Today. Since then many individuals have been vaccinated on recommendation of the clinics, but many have not, due probably of combinations of cost and perceptions that risk and consequences of hepatitis A infection were low. However, there is now an additional reason to reinforce the original recommendation to vaccinate. In the New England Medical Journal, January 29, 1998 (1998;338:286-90) there is a report of 41% fulminant hepatitis and 35% mortality in HCV infected individuals superinfected with hepatitis A. An editorial in The Lancet subsequently called for hepatitis A vaccination in all HCV infected individuals.

We suggest that clinics reinforce the recommendation for hepatitis A vaccination in light of this new information. We note the letters to the editor of the New England Journal of Medicine June 11 disputing the degree of risk; however, these should not detract from what would seem to be a logical, simple and safe recommendation.

Editor’s Note: The CHS would like to express their sincere appreciation to Dr. Irwin Walker who will be stepping down as Chair of the MSAC this fall. For the past four years, Dr. Walker has generously shared his time, expertise and knowledge with the Board of Directors, staff and individuals. We thank him for his dedication, interest and support of the hemophilia and bleeding disorders community. Dr. Sue Robinson, Clinic Director at the Q.E.II Health Sciences Centre in Halifax, Nova Scotia, will be taking over as Chair of the MSAC on September 1, 1998.
Fifteen Physiotherapists representing seven provinces had the opportunity to participate in Winnipeg II, Standards of Comprehensive Care for People with Hemophilia and Other Inherited Bleeding Disorders Conference in Winnipeg. Following the Conference, the Physiotherapists met for discussion, education sessions, and strategic planning.

Considerable work had been done since the 1997 meeting in Mississauga. Several therapists are involved in the Prophylaxis study. An assessment form was developed for discussion and revisions, and will be ready for use soon. A document entitled “Standards of Care for People with Hemophilia and Other Bleeding Disorders”, modeled after work done by the Chartered Society of Physiotherapists in Britain was presented; after some revisions, it will be forwarded to the Canadian Hemophilia Society and the Canadian Physiotherapy Association for approval. Abstracts of recent articles on hemophilia were collected and distributed at the meeting.

Education sessions dealt with joint replacement surgery, chronic arthropathy, and pseudo tumors. Plans were made to continue to submit articles to Hemophilia Today, and to continue to gather and disseminate information that physiotherapists can use to educate themselves and the families dealing with bleeding disorders of all severities.

The Physiotherapists are grateful to the CHS for supporting this meeting. We hope to be able to continue our work and put the Standards of Care into action across the country.

The Blood Task Force continues to draw upon all resources of the RCMP, nationally and internationally (i.e. Analytical Services, Canadian Police College, Community Policing Branch, Foreign Services Branch) towards the expeditious resolve of this investigation. We have benefited from the assistance, experience and cooperation of policing partners, as well as private and public organizations, locally, nationally and internationally.

Over 500 calls have been received on the TIPS hotline since its implementation in February and in excess of 500 interviews of witnesses, victims and complainants have been completed. The investigative team has been augmented by the addition of a civilian member of the RCMP, who is a lawyer, in order to assist in this very complex endeavor.

The Task Force continues to travel extensively throughout Canada, in addition to traveling to Germany, the Netherlands and the United States. The magnitude of information accumulated to date has permitted the investigation to become increasingly focused. However, the sheer volume of information requires considerable time to be effectively accrued and analyzed. The criminal investigation is now into its sixth month.

The direction of the investigation is being maintained on the structures and systems relating to the decision-making processes within the Canadian blood program. Anyone with information can call the national TIPS line at 1-888-530-1111.
Hemophilia, An Evolution

by Karen Creighton

As I reflect upon our lives as a family comprising children both with and without hemophilia, I think it is fair to say that it has impact on all of our lives sometimes, in unpredictable ways. Three of our five children have moderate, type A hemophilia. Our oldest son, Brendan does not have hemophilia. Our six year old fraternal triplet boys: Ryan, Sean and Dylan are hemophiliac. I try not to feel sorry for our situation as we experience the ever changing facets of hemophilia. My husband Ted and I believe the value of information and support we have received from the local, provincial and national hemophilia chapters have been outstanding. Combined with the medical expertise and support of our comprehensive care team, we are prepared to handle our challenges. Why then has the evolution of hemophilia in our lives been such a surprise?

In the past, Ted and I have deliberately avoided drawing attention to our family’s situation. It truly surprises me now to willingly consider writing about our kids and our circumstances. If, however, we can help any parent better relate to our experiences or anticipate issues, then we will continue to keep writing and sharing. We welcome input from all families out there. We need to grow and learn from each other, to get more involved in our local chapters and to enjoy the support we can share as families of children with bleeding disorders.

In the Spring of 1993, when our boys were diagnosed we found there to be a shortage of written information available to parents. As there was no family history of hemophilia we were starting from a weak position. Overall, we were in total shock and had no idea of what to expect. Despite our best efforts to educate ourselves, the various stages in the evolution of hemophilia have been surprising, and often difficult to deal with. The good news now is the present situation we find ourselves in and the generosity of people who have gone out of their way to help us over the last five years.

The Diagnosis
Ryan, Dylan and Sean were nine months old when they were diagnosed with moderate hemophilia A. As they learned to crawl, they began to develop large bruises on their ribs and chest. A visit to our local pediatrician confirmed to us that this was not a particularly normal thing to see. Thankfully, we were quickly referred to The Hospital For Sick Children in Toronto for tests. A diagnosis was made within a few days. I realize many families endure great difficulties getting to this stage, especially in relation to vonWillebrand’s disease. We live in Mississauga, west of Toronto, where medical resources are excellent. Within the first month following our introduction to hemophilia it was decided that the kids should be fitted with helmets.

Helmets
I know many parents dislike helmets. We, on the other hand, didn’t debate the issue too much considering the fact we had three kids in the same situation. Unlike some families that believe in padding the home environment and making the home as safe as possible, we padded the kids and let them roam wherever they chose. The custom fitting and fabrication of these helmets took place at the Hugh MacMillan Rehabilitation Centre in Toronto. I felt I had lost a part of each of the boys the day they tried their helmets on for the first time. They were in a triplet stroller at Hugh MacMillan. I wanted to bolt out the front door and leave the hardware behind. Over time, though, I learned to befriend the helmets. I washed and scrubbed them, kept a good supply of stickers around to decorate them and...
took them wherever we went. In time, they became so much a part of our lives I couldn’t understand why we drew so much attention to ourselves at playgrounds and parks. I learned to deflect a lot of the attention and ignore those driven by curiosity.

Despite these best efforts, we had one head injury through the toddler years. For us, helmets made good sense. In many ways, it gave us the sense of security we needed to deal with some of the injury risks related to hemophilia. After all of the initial debate we did find that it was really difficult to wean ourselves away from them. In fact, when we moved last year neither Ted nor I could bear to toss the helmets out. More for sentimental reasons than for practical ones. They remain in a box in the basement, a testament to days gone by.

**Teething**
We didn't have trouble with new teeth. We have had and continue to have some problems with baby teeth coming out.

**First Injuries**
Our first trip to Sick Kid’s occurred in July of 1993, when Ryan fell out of his high chair and bumped his head. It wasn’t serious, yet I remember it as an anxious time. Previously, I worried: Would I recognize the need to treat an injury? Who would need the first treatment? How would it go? How often would the boys need treatments?

All these questions were answered over time. We didn’t really experience typical hemophilia issues until the boys turned five. At that time we encountered from zero to seven or eight treatments a month based on various pre-schooler events. We have seen: bitten tongues, stubbed toes, banged elbows, twisted ankles, bumped heads, doors shut on fingers, a cut hand, winter nose bleeds and the odd unique injury such as one where Sean jumped off a diving board with a plastic oar in-between his legs. Add to this the fact that the boys each had a bad dose of chicken pox last summer with Sean needing a treatment to help him through the worst of it.

Our treatment pattern mostly matched an injury/response pattern. The injuries seemed random and typical of any child’s potential injuries.

**Treatment Plans**
We became motivated very early on in our experience to eliminate the need to travel to Sick Kid’s for treatments. In heavy Toronto traffic, it could take upwards of one and a half hours to reach a busy emergency waiting room. With the help of the Sick Kid’s team and an outstanding pediatrician at our local hospital in Mississauga, a Care Plan was established to enable us to use The Credit Valley Hospital emergency department for treatments. The boys were one year old at this point.

After many months at Credit Valley, we had become ‘regulars’ in Emerg which had its pluses and minuses. We again became motivated to improve our treatment routine as we frequently experienced lengthy emergency room visits. Our pediatrician began to meet us at the hospital during her office hours to facilitate the process of providing treatments. She then began treating the boys at her office during the day. Through Sick Kid’s, I received and brought Kogenate to her office.

We had outstanding day support and continued to rely on our local hospital for evening and weekend coverage. In the Fall of 1994 we became part of the Homecare Program. This involved a nurse from the V.O.N. organization visiting our home to provide treatments as required. For head injuries, however, we continued to see a Doctor. During this period I often spoke to our local Doctor and contacted the Sick Kid’s team to seek advice. We took the kids to our Pediatrician whenever we encountered something new or felt the need to seek her advice.

We came to know one nurse in particular who kept her pager on for us during her non-working hours. Her commitment to the children and to our family was beyond our most extreme hopes. She knew the children intimately and was instrumental in training me to know their veins and treatment techniques.

**Independence**
Is there such a thing? Considering the boys are moderate, we never expected to set the patterns we did. Last Spring the boys had repeated ankle bleeds. It seemed a trip to the park would automatically yield repercussions. It was hard to watch them experience the pain of their first joint bleeds. They had injuries that took longer to heal, often reoccurred and required more treatments.

At this time I was taught to infuse Dylan and Ryan myself. The learning process was odd. I wanted to learn yet I dreaded doing it. Shortly afterwards, Sick Kid's recommended prophylaxis as a result of the bleeds the boys continued on page 18
The CHS is pleased to announce the recipients of the 1998 Scholarship and Bursary Program. This year a new category of award has been added specifically for mature students enrolled in post secondary vocational training or retraining programs. The Scholarship Review Committee reviewed eight applications and selected the following recipients:

**Scholarship**

Christina Halliday is a Ph.D. student now entering her third year in the Faculty of Education at York University, Toronto. Christina comes from a background in the sciences, literature, philosophy, and fine arts. For her doctoral thesis, Christina intends to study the relationship between long-term illness, perception of body and self, and dance movement therapy. On the side, Christina works as a fundraising consultant for two Toronto, inner-city projects: A Home for Creative Opportunity and Canadian Children’s Dance Theatre. For fun, she’s into urban cycling and hiking, swimming, reading, and creative writing.

**Bursary**

Louis-Philippe Gagnon is a student in the Bachelor of Business Administration Program at the Université de Sherbrooke. The Bachelor of Business Administration program allows him to obtain simultaneously a general training in administration based on the knowledge of concepts, principles, administrative methods and specialized training. The specialization aims at elaborating and implementing an information system in addition to the connected problem analysis.

**Mature Student Bursary**

Rob Friesen is a forty-three year old member of the Manitoba Chapter. He is a husband and a father of five children who also lives with moderate FVIII hemophilia and complications from Hepatitis C. Because his hemophilia was not diagnosed until he was about eight years old, the lack of early treatment has resulted in some permanent damage to joints. As a result of these limitations he was unable to continue in his career in furniture refinishing. He decided to capitalize on his art background and twelve years of business experience and is pursuing a new career by studying computer graphics at Red River Community College.

The Scholarship Review committee extends their congratulations and best wishes to Christina, Louis-Philippe and Rob for continued success in their chosen field of studies.
This summer I was fortunate enough to be able to attend my second world AIDS conference in Geneva, Switzerland. My first experience at one of these conferences was in Vancouver in 1996.

I see now that the Vancouver World AIDS Conference was not a typical conference. I remember feeling a lot of positive energy there, and on a more personal level, I cherished the hope of the new drugs and the conference’s thematic promise - “One World, One Hope.”

I also remember being very conscious of the lack of discussion in Vancouver around the accessibility of the antiretroviral drugs. Although there were delegates from developing countries in Vancouver who spoke informally about having no opportunity and no resources to get these drugs (I remember, in particular, an HIV+ man I met from Uganda who voiced these concerns to me in the PLWHA Resting Lounge), the urgent needs of these conference delegates were clearly second fiddle to the glory of the “drug cocktail” discovery and the men behind it.

The overall tone in Geneva, however, was a bit different. There was a sense of “going back to the drawing board” - of examining and making restitution for what critics believed the Vancouver conference was really about, that is “Many Worlds, but only One with Hope.”

The 12th World AIDS Conference in Geneva hoped to create an environment where the PLWHA community and specifically, those people affected by AIDS and HIV in the developing South, would have a strong and united voice. The theme of this conference, “Bridging the Gap,” brought with it a naive optimism -- that is, that the grave concerns and issues of PLWHA in the South would be adequately heard and responded to. Moreover, the conference theme suggested that the grave gap in access to antiretroviral drugs, between North and South, could be bridged.

It was interesting to see how this theme of “bridging gaps” got played out in Geneva. Possibly the most fundamental mechanism for “bridging the gap” at the conference was “The Geneva Principle.” This was a principle in conference organization and development that sought to make links between science and the PLWHA community by involving both in all aspects of conference planning.

Some examples of “The Geneva Principle” at work were the presence of PLWHA on Plenary Session panels (the first and most comprehensive session of the conference day) alongside research scientists and medical professionals. Bridging Sessions were another highlight in Geneva - also interdisciplinary but less formal and consisting of debates and round tables that were designed to forge links between science and community and between different regions of the world.

The daily conference newspaper, The Bridge, posed itself as the voice of the conference and was another extension of “The Geneva Principle.” Concerns and issues of the PLWHA community took centre stage in The Bridge but I felt that the publication lacked complexity in the voices of community and science that it represented. Most articles only reiterated the mantra of the conference, “Bridging the Gap,” with its implication that the gap could indeed be easily bridged.

And this brings me to the drug companies who occupied a huge airplane hanger at the conference, turning “The Geneva Principle” and “Bridging the Gap” into a giant hypocritical joke. While sessions happened and community forums met to struggle with the gross lack of access to antiretrovirals in the South, the drug companies brashly continued their business. They gave out free stuff which delegates scrambled for (bags, pens, CDS, cappuccino, chocolate) and they had representatives stand around in pretty, fabricated offices to chat up doctors. But nowhere in their midst did I see any thoughtful awareness or action related to the problem of continued on page 16.
Greetings Canada! This season has been filled with a variety of programs and events within the hemophilic community. During the spring the Canadian Hemophilia Society hosted the second Standards of Care Conference in Winnipeg. This conference was designed to bring Hemophilia specialists and consumers together from across Canada to participate in a variety of groups. The ultimate goal was to improve and continue the evolution of Hemophilia care in Canada. Hemophilia Care varies greatly province to province and this conference was important in bringing together ideas and providing insight into overcoming challenges faced by many provinces. This conference will definitely bring about changes in the way Hemophilia care is delivered in the next century as the millennium is fast approaching and Health Care delivery systems at large are changing to meet new realities.

As I mentioned in the previous edition of Hemophilia Today, Hemophilia Ontario Youth Events hosted its annual Winter Retreat in March. This weekend of relaxation saw the participation of twenty youth from across Ontario. There was cross country skiing, tobogganing, broomball, as well as a chance to make new friends and rekindle old friendships.

In May I attended the World Federation of Hemophilia Congress in the Netherlands. This well-organized and informative event was once again a great learning experience. I had the chance to participate in a variety of informative sessions ranging from effective communication to the psychological aspects of HIV/AIDS. However, one subject that really attracted my attention was inhibitor management. There are many effective treatments available; however, as good as they may be many treatments lack the potency of traditional Hemophilia treatments. However, there may be some more good news in sight. Novo Nordisk’s NovoSeven product has been tested and used with success in many individuals with inhibitors. NovoSeven contains activated recombinant factor VII. Currently this product is unlicensed in Canada and is only available through Emergency Drug Release Program. When it becomes licensed this fall in Canada it will be called Niastase as the name Novo is owned in Canada by another company. The recombinant version of factor VII is very similar to plasma-derived factor VII. Bleeding episodes can be effectively controlled using Novo Seven depending on the type and severity of the hemorrhage. The recommended dosage is 90mcgs/kg, IV every 2 hours. Where Novo Seven becomes slightly different than traditional treatment is that it must be administered initially every second hour until there is marked improvement. However, once improvement is noted the dosage intervals may be increased. A major bleeding episode may be treated in two to three weeks. Mild bleeding episodes would require less aggressive treatment and homeostasis could be achieved with as little as one to three doses administered every three hours. Though this treatment regimen may sound time consuming and tedious, home therapy is entirely possible for this product. The reconstitution is similar to other concentrate products. In fact I personally know of one hemophiliac who effectively manages his bleeding episodes with Novo Seven on home therapy. However, it is important to note that Novo Seven is not yet licensed for use in Canada and there may be adverse reactions with its use. However, it is good to know that there are innovations being made in hemophilia care in order to meet new realities.

Overall the XXIII International Congress of the World Federation of Hemophilia brought me increased knowledge and I had a great opportunity to make new friends and see people who I have not seen in a
couple of years. I am happy to say that the congress for the year 2000 will be hosted in Montreal. I have been appointed liaison on the Youth Division of the World Hemophilia Federation to Montreal. I am sure many of our great successes in Ontario can be shared the world over. In the coming month Ontario will be printing a special publication regarding the events and detailed information of the XXIII international congress in a newsletter. If you live outside of Ontario and would like to obtain a copy, please e-mail me and I will mail you a copy of the newsletter.

On the programming front, 1998 has seen a record number of Hemophilia programs for youth in Ontario than in any other year. Recently, Hemophilia Ontario Youth Events held its first ever JAY DAY for younger children. This included a day at the ball game and a chance to bring younger hemophilic youth together within the community. In August, Hemophilia Ontario will be hosting its annual Wilderness Getaway and the number of participants is expected to be high. Stay tuned for the next issue of Hemophilia Today for news regarding the Getaway. Take note of my new e-mail address and if you have any comments or questions please let me know. On behalf of the Canadian Hemophilia Society I would like to wish everyone in Canada a safe and happy return to school and I hope you all had an enjoyable summer.

Karttik Shah
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Update on Hepatitis C Compensation Negotiations

The following update was provided by Bonnie A. Tough, LLB, who is representing those hemophiliacs infected with Hepatitis C who received blood products between 1986 and 1990.

There have now been four negotiating sessions with the governments. While everything continues to move ahead slowly, we have progressed through the preliminary and organizational matters to the stage where substantive issues are being discussed with a view to ultimately determining a compensation proposal. The negotiations will not likely reach any significant inroads over the summer months as a result of individuals’ vacation schedules, but they will resume in September.

Currently, we are pursuing an agreement with the federal government that it agree to certification of the action commenced in Ontario upon a settlement being reached. The agreement would include all hemophiliacs residing in provinces other that British Columbia and Quebec. In British Columbia and Quebec there would be separate consent certifications.

We have established a co-ordinating committee made up of the representative plaintiffs from the various provinces, or their nominated representative, to discuss issues and a national strategy on a weekly basis. The members of the co-ordinating committee are Pauline Fournier, Edmundston, New Brunswick, Allan Gruhlke, Edmonton Alberta, Barry Isaac, Calgary, Alberta, James Kreppner, Toronto, Ontario, Normand Landry, Moncton, New Brunswick, Jim Love, Winnipeg, Manitoba, Ron MacLeod, Charlottetown, PEI, Chris Mitchell, North Vancouver, B.C. and David Page, La Durantaye, Quebec.

If anyone wishes more information or wants their name to be added to the registry they may contact Bonnie Tough at 1-800 342-7924.
Over 100 countries were represented at the table during the AGM on the last day. The 2002 Congress will take place in Seville, Spain. While simultaneous translation is always available in English and Spanish there is a need to add more. The meeting in Montreal in June 2000 will, naturally, have simultaneous French translation available.

Representatives from eight French speaking countries met in the Hague to begin a program of co-operation and exchange for documentation and information in French. This makes the accessibility to understandable information for their members easier for some of the French speaking African and mid-Eastern nations. It will also lower the cost of translation for all. The WFH has promised to develop this program.

During the Congress I focused mainly on the sessions dealing with women’s issues. The sessions were so numerous that it was impossible to attend them all. I jumped from one session to another to hear speakers deal with issues related to carrier testing, prenatal diagnosis, obstetrical issues, and treatment for women with bleeding disorders of all kinds. Every presentation on women’s issues reinforced the fact that bleeding disorders in women are often more complicated than the same degree of a disorder in men due to menstruation and childbirth. Dr. Peter Kouides of Rochester, NY emphasized the need for standardized guidelines and procedures for the diagnosis and treatment of women with bleeding disorders. He also stated the need for standardized guidelines for the management of pregnant women with bleeding disorders as well as neonatal guidelines. Presentations on molecular genetic testing showed that there are numerous options enabling accurate carrier detection. In his presentation Dr. P. Giagrande (UK) stated that prenatal diagnosis deals not only with the medical procedures, but also the psychological aspects. Quality of life for women with menorrhagia and the psycho-logical, physiological and financial effects was a common topic in numerous presentations. I gained a great deal of information which I will be sharing in the Female Factor in future.

A program for parents was presented called PEP (Parents Empowering Parents) to train parents to deal with discipline, siblings and independence for children who have a major health problem. Other presentations dealt with port-o-caths, prophylaxis for young children including the optimal age to begin treatment versus on demand treatment and clinical results for both.

Many of the presenters or moderators at the sessions were doctors or nurses from Canada. It was nice to see familiar faces and to get to know them away from the clinics. Ann Harrington, the nurse-coordinator from St. Michael’s Toronto, was elected as Chair of the Nursing Committee for the World Federation.

In the year 2000, the next Congress of the World Federation of Hemophilia will take place in Montreal, Quebec. It’s your chance to get an incredible amount of information and to meet people from all over the world who are part of the family of bleeding disorders, be they patients or caregivers. We all are truly related by blood.

Patricia Stewart
designated directors. In addition, Francois Laroche (Quebec) is new to the Board as a director-at-large.

Appreciation and a hearty “welcome back” are due to Sue Gibson, Janice Aull, Bob Vereau, Eric Stolte, Mike McCarthy, Tom Alloway, Dave Mitchell, Pam Wilton, Donald Pouliot, Normand Landry, Neil Van Dusen and Cyril DuBourdieu as designated directors, and Dan Doran and Ron Boghean as designated directors.

So who am I? Many of you may know me, or at least my name, from a variety of activities that I have been involved with other the past 10 years or so as a CHS volunteer. Certainly one of the greatest gifts I have received during that time has been our friendships. For those of you to whom I am still a stranger, here is a short biography. I was born and raised in Toronto, and moved to Winnipeg to pursue my graduate degrees. After finishing my Ph.D. in developmental psychology, and enjoying the freedom of no school and long-distance family responsibilities, I met my future husband, James Love. Jim had severe factor 8 hemophilia. Up until that point, hemophilia to me was part of historical movies and books or a condition to help teach inheritance of sex-linked traits. Now it was real, with all of the chronic arthritis, joint problems and trips to the hospital for treatments that anyone could do without. The switch to home infusions was a blessing!

Time moved on to diagnosis and hospitalizations for HIV-related illnesses, more recently resulting in full blown AIDS. Then came the fights for HIV for national and provincial/territorial compensation, the stark and horrendous truths from the Krever Inquiry relayed to us daily through the media, the HCV diagnosis and the fight for HCV compensation. And guess what? There seems to be an inherited bleeding disorder within my own genetic history. In the midst of the work shone the joy - a daughter arrived in 1995, truly the center of our small family.

Through all of the years, I have continued to marvel at the goodness and strength of people who were harmed so needlessly and, for some, recklessly. Although there have been diverse and sometimes divergent ideas about how to react and to cope, we have continued to listen and learn from one another. Anger has been used to build bridges to one another in such a way that there always seems to be hope and joy. I walk this road everyday. Sometimes it’s up, sometimes down, and usually both several times a day. And these people, their hope, and the path I have walked will define my approach to the coming year of my presidency.

Over the past years with the CHS, I have valued the way in which we work. Each and every staff member has contributed enormously to furthering our mission and goals, and it is through the synergy of staff and volunteer efforts that we have forged a strong, effective organization. Mutual respect, value and collaboration will be evident in the way we conduct our business.

Working as a team, respecting the differences among us and building on the similarities, I see us addressing several challenges: implementing the standards of care developed at Winnipeg Two in April/May 1998; safeguarding access to highest quality factor replacement products; reaching out to ensure that people with a bleeding disorder receive appropriate diagnosis and treatment; renewing our resources for those who are newly diagnosed with a bleeding disorder; monitoring the implementation of Canada’s two new blood systems; and defining our role in a world in which 80 per cent of those with hemophilia have the same standard of care as Queen Victoria’s son in the 19th century.

As your new president, I want to thank you for the opportunity to serve you in this capacity. I can promise you that I will listen to you. And I am looking forward to getting to know more of you, to sharing with you and your families, and to working together to advance hemophilia care.

Erma Chapman, Ph.D.
Helping girls to deal with carrier status

Consider the readiness of your daughter to deal with the information, taking into account her age, emotional maturity and level of interest and understanding. Recognise that denial is a way of dealing with carrier status. Even if she has known that she may be or is a carrier, it is in adolescence that the implications will be more fully understood. Adolescents feel sensitive about personal image and undergo rapidly changing emotional states. Allow for strong reactions and be supportive. Use any available help from counsellors and genetic counselling centres at the CCC. The attitudes of fathers or brothers with hemophilia to their disorder usually provides a model for the carrier daughter’s attitude toward being a carrier and to having a son with hemophilia. A daughter’s anxiety may be made more acute by the suffering of a father or brother with hemophilia. This anxiety may be further compounded by feelings of anger towards a brother who requires special attention for a condition which not only causes her to be ignored, but also for which she, herself, may one day be responsible in deciding her own reproductive choices.

Science now provides a diversity of genetic options for people who are known carriers and their partners. Our next article will deal with some of these options.

Two things then happened. The injuries stopped and we ran into trouble treating all the boys. Based on three regular treatments a week I had difficulty treating Ryan and Dylan. It took me one month to successfully access secondary sites and give their best veins a rest. Sean became close to untreatable via venous access. The Sick Kid’s team monitored us closely and helped us see Sean required a port. This was not a solution I accepted readily.

In early July, Sean had surgery to implant his port. I had great difficulty accepting that he needed one and seeing one of our boys moving towards a treatment option I never envisioned he might need. I am pleased to say it went well - so far. I am now treating all three cherubs myself. This statement seems so simple to write yet it is the culmination of much stress, learning, emotions, dread, pain and people’s help. It is their story in that the boys are the ones with hemophilia. Our family, however, has been shaped by their health issues.

Let’s Celebrate

We now have our fifth child, Colleen, who is two. The triplets are now six and our oldest, Brendan, is nine. For the first time we are packing up and going on a family cottage holiday. We will be traveling with more medical supplies than the average family. We, however, are not the average family.

Ted and I take little for granted and assume very little about life. We both feel, all in all, we are at a great stage in our lives and we are definitely at the best point we have been so far, relative to the day in April back in 1992 when we were told the children had hemophilia.

There is a great deal to be grateful for in our lives starting with five amazing children. Just as we encourage the boys to deal positively with the ups and downs of hemophilia, we do the same.

Involvement

Now more than ever we enjoy our connection to other families with children with bleeding disorders. We have more to contribute based on what we have experienced and more to learn as we think about new issues. The Toronto chapter is enjoying the influx of some new people and new ideas. We had a parent get together recently with our physiotherapist from Sick Kid’s. It was a great opportunity to discuss injuries and treatments in a family environment.

Please let us know if you have any questions or experiences to share. If the interest is there we will seek medical responses to questions sent in. Keep in touch and keep healthy.