

Haemophilia Awareness Week

8-14 October 2006

Haemophilia Awareness Week 2006 will be themed **Busting the Myths** and aims to dispel many myths about haemophilia. HFA will manage a media campaign and is working with State/Territory Foundations to develop activities around Australia.

You can participate in many ways during the Haemophilia Awareness Week -

- Host a "MYTHBUSTER" trivia night.
- Set up a stand in your workplace, school, hospital – busting the myths!
- Organise a fundraising event such as a casual clothes day, bike ride, walk, run or swim.
- Organise a theme day at your workplace or school – wear red and white and donate a gold coin.
- Host a luncheon, sausage sizzle or morning/afternoon tea.

HFA will provide you with promotional items for your event.

For more information please contact Natasha on 1800 807 173 or email ncoco@haemophilia.org.au or visit our website www.haemophilia.org.au.

HAEMOPHILIA



HAEMOPHILIA FOUNDATION AUSTRALIA

Busting the myths

MYTH - If a person with haemophilia cuts himself he will bleed to death.

People with haemophilia have blood that doesn't clot properly so it will bleed or seep for a longer time. With appropriate treatment, it will definitely stop. Bleeding is mostly internal. The person does not bleed faster - it just takes more effort to stop persistent bleeding. Bleeding from cuts can usually be controlled, at least temporarily, by applying pressure over the cut.

MYTH - People with haemophilia cannot play any sports.

People with haemophilia will find that sport strengthens joints and muscles, helping prevent bleeds. Care needs to be taken when choosing a sport as rough high contact sports such as football and boxing aren't usually recommended as they may cause bleeds. People learn to manage their condition and know what is good for them to do. People with haemophilia can play a wide range of sports.

MYTH - Children with haemophilia have to go to special schools.

Definitely not! Children with haemophilia thrive on being treated like any other child.

MYTH - Children with haemophilia must constantly wear protective clothing and helmets.

This is definitely not true. Everyone – whether they have haemophilia or not – should wear protective gear appropriate for their sport, e.g. a helmet should be worn by all children when cycling and standard safety gear worn when playing cricket.

MYTH - It is a royal disease.

Haemophilia has often been associated with the royal family, and is sometimes called 'The Royal Disease'. Queen Victoria (Queen of England, from 1837 to 1901) was a carrier of the haemophilia gene. However, haemophilia can occur in any family as a result of genetic mutation, or a change in the genetic code.

MYTH - Haemophilia is contagious.

There is absolutely no way haemophilia can be 'caught'. Haemophilia is hereditary, however in 1/3 of cases it appears in families with no previous history of the disorder.

For more information please contact
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Australia/New Zealand Haemophilia Social Workers' And Counsellors' Group

Activities And Events Around Australia And New Zealand

The annual meeting of the Australia/New Zealand Haemophilia Social Workers' and Counsellors' Group was held in Melbourne on 20-21 July 2006. The meeting was attended by nine social workers and counsellors from around Australia and three Haemophilia Foundation of NZ outreach workers. The agenda included full and lively discussion about psychosocial services and care for the bleeding disorders' community. The meeting, including a professional development session facilitated by Dr Paul Denborough on development stages for adolescents, chronic illness and treatment compliance was valuable.

Membership of the Australia/New Zealand Haemophilia Social Workers' and Counsellors' Group includes:

Sandy Breit, Victoria (Chair)
Leonie Mudge, Victoria
Kathryn Le Maistre, Victoria
Claire Reeves, ACT
Robert Hearn, Newcastle
Position to be filled, NSW
Maureen Spilsbury, Queensland
Kelly Brady, Queensland
Sharon Hawkins, Western Australia
Anne Till, South Australia
Helen Spencer, NZ
Colleen McKay, NZ
Drew Mackenzie, NZ

The following report of activities was written by Kathryn Le Maistre. Kathryn is the Haemophilia Social Worker at Royal Children's Hospital, Melbourne.

Many different and exciting activities have taken place throughout the year in each of the States/Territories and New Zealand. This is just a snapshot of the work that members of the Australia/New Zealand Haemophilia Social Workers' and Counsellors' Group have been doing with people with inherited bleeding disorders and their families.

Victoria:

- Annual HFV family camp April 2006 attended by Leonie Mudge
- Transition day for older boys with haemophilia moving across to the adult setting facilitated by Leonie Mudge and Sandy Breit
- Living Well with Hep C information session October 2005 facilitated by Sandy Breit
- Sandy Breit was interviewed on Hep Chat, hepatitis C community radio program about counselling issues
- Teachers seminar 2006 - presentations from Kathryn Le Maistre, Director of Henry Ekert Haemophilia Treatment Centre, Educational Advisor and Nurse Coordinator

- Newly diagnosed haemophilia parent group facilitated by Kathryn Le Maistre

ACT:

- Involvement of Claire Reeves in the Quality of Life survey produced by Dr Dipti Talaulikar
- Claire Reeves and nursing staff developed an education aid for children

Western Australia:

Sharon Hawkins facilitated a range of activities including:

- Clinic morning teas at Fremantle Hospital
- Parent/family meetings
- Carer's meetings
- Young spouse's/carers meetings
- BBQ for families
- Men's breakfasts

Queensland:

- Transition day for six young people moving across to the adult treatment centre organised by Maureen Spilsbury
- Genetic information evening
- Partners' lunch held every month with an outreach lunch planned for September on the Gold Coast

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- New families' BBQ picnic held by Kelly Brady
- Development of a social support checklist and social work pamphlet by Kelly Brady

New South Wales:

- Introduction of fortnightly case conferences in which Leola Farnell is involved
- Haemophilia family camp held in November 2005 and organised by Leola Farnell
- The writing of 'Men's Stories' by Robert Hearn

New Zealand:

- Annual Boys Day Out targeting 11-16 year old boys organised by Helen Spencer
- Helen Spencer spoke at the educational evening organised by HFNZ Northern committee in November 2005
- First 'New Families' camp January 2006 co-ordinated by Colleen McKay and assisted by Helen Spencer
- Northern Regional Camp February 2006 attended by Helen Spencer
- First National Men's Weekend June 2006 co-ordinated by Helen Spencer and assisted by Drew MacKenzie
- Central Region Winter Escape Weekend attended by Drew Mackenzie
- Play group organised by Colleen McKay in July 2006

It has been a busy year for the social workers and counsellors and they are each looking forward to planning more activities for 2006-2007. **H**

Australian And New Zealand Physiotherapy Haemophilia Group

Rebecca Dalzell

This article about the Australian and New Zealand Physiotherapy Haemophilia Group (ANZPHG) is written by Rebecca Dalzell, Physiotherapist at Royal Brisbane & Women's Hospital.

Rebecca advised of her intention to step down as one of the Co-Chairs of ANZPHG at the annual meeting of the group held on 18 August 2006 at the HFA offices in Melbourne as she will be taking maternity leave later this year. Rebecca and Wendy Poulsen have provided strong leadership over the last three years and have further encouraged the value of physiotherapy to care and treatment. We are grateful to both Rebecca and Wendy for their active participation in the bleeding disorders' community. We are also delighted that Rebecca will continue an international collaboration with her research about the use of Tai Chi and Yoga for people with bleeding disorders.

The Australian and New Zealand Physiotherapy Haemophilia Group (ANZPHG) is a professional group associated with Haemophilia Foundation Australia. The group is made up of physiotherapists from each state of Australia (except NT) as well as New Zealand.

The mission of the group is:

'To facilitate the provision of comprehensive care to people with bleeding disorders through quality physiotherapy, and to work in partnership with HFA and HFNZ to promote optimal quality of life for this population'.

We seek to be a cohesive physiotherapy special interest group in order to effectively support the community of people with bleeding disorders. Some of our goals include:

- To develop and enhance the national standard of physiotherapy care for those with bleeding disorders.
- To advocate for people with bleeding disorders in relation to condition-specific needs and opportunities at a local and national level.
- To provide ongoing education and support to people with bleeding disorders and their families.
- To provide specialist advice and education to other physiotherapists and physiotherapy students regarding haemophilia care.

- To develop comprehensive and up-to-date educational resources regarding physical activity and musculoskeletal management for people with bleeding disorders of all ages, their families, and the community.
- To promote awareness within the local and national community as to the benefits of both physiotherapy and an active lifestyle for people with bleeding disorders.
- To promote and participate in research to advance evidence-based practice, improve the quality of physiotherapy care, and enhance the health and well being of people with bleeding disorders.

As a group we strive to communicate regularly for: service development, information sharing, peer support and professional development. One way we achieve this is through annual face-to-face meetings. Group members come together each year to share ideas and work towards achieving our goals and objectives. This is kindly supported by HFA.


Since last year's meeting we have undertaken numerous projects including a group survey. The goal of this was to establish current physiotherapy work practices in each treatment centre and to use these results as a basis for lobbying. It is

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our desire that each state/territory of Australia, and numerous areas of NZ, has funding for a designated physiotherapist who is then able to have the time to develop services and enhance haemophilia care in that region. Currently there is great variation in roles and funding with many therapists only available for a couple of hours a week for haemophilia care. This is not ideal and promotes reactive rather than proactive service delivery. Achieving greater funding and support for physiotherapy services is an ongoing endeavour.

Over the past year many group members have also participated in research projects, sat on committees and provided new and improved services within their region. Educational articles and materials have also been produced, and presentations delivered at camps and conferences.

Fifteen physiotherapists, including two from New Zealand attend the 2006 annual meeting in Melbourne on 18 August. We are excited that each year we seem to be establishing more contact with NZ physiotherapists. At the meeting this year we had opportunity to discuss and plan for future pilot projects and the development of new educational materials. We discussed other research projects that are currently underway and looked at how we can continue to enhance our services through new programs and protocols. We had an afternoon of energetic discussion regarding treatment approaches and management strategies, and discussed numerous case presentations and complex scenarios. It was overall a very worthwhile day and we now have new plans to move forward within the coming year. It is our desire that as we continue to meet, plan, and achieve our objectives that the community will benefit through enhanced haemophilia physiotherapy services throughout Australia and New Zealand.

If you would like further information on physiotherapy services offered in your area, or if you have any physiotherapy related questions, you are encouraged to contact your local haemophilia physiotherapist. If you do not have a name or contact details for a physiotherapist, contact Heather at HFA on the freecall number 1800 807 173. 

Australian Haemophilia Nurses' Group

The annual meeting of the Australian Haemophilia Nurses' Group was held in Melbourne 7-8 September. The meeting was attended by 17 haemophilia nurses from around the country and 1 from New Zealand. Further information about the activities of the group will be published in a later newsletter. 



Australian Haemophilia Nurses' Group



Australian and New Zealand Physiotherapy Haemophilia Group

Staff Changes Around The Country

Heather Lauder

There are some staff changes in the haemophilia scene around Australia.

Queensland Haemophilia Centre

Rebecca Dalzell, physiotherapist for adults at the Queensland Haemophilia Centre will leave the position she has occupied for five years on 13 October to take maternity leave. Carley Ekert will commence in the position on 16 October and will be contactable on the same telephone and contact details.

Kelly Brady commenced as the Haemophilia Social Worker in mid-April and works with Maureen Spilsbury, Senior Haemophilia Social Worker and the team at the Queensland Haemophilia Centre. Kelly works predominately with paediatric patients and their families at the Royal Children's Hospital, Brisbane. In addition, she will provide cover for adult patients and their families of the Royal Brisbane and Women's Hospital during times when Maureen is away on holidays and other occasions.



Kelly Brady

Haemophilia Foundation Victoria

Julia Broadbent has commenced as Communications Coordinator for Haemophilia Foundation Victoria. Julia's background is in office management, having had nine years' experience in responsible administrative roles. Just before she joined HFV, Julia was working for a pre-school non profit organisation preparing duty rosters, conducting training sessions and designing a number of business web sites. Prior to that Julia was an office manager in Boston (USA) for a modern telecommunications company – this role included assessing fraudulent activities and general office administration. Julia has also managed a busy dental surgery, and worked as an events organiser in Birmingham, UK.



Julia Broadbent

Women's and Children's Hospital, Adelaide

Dr Ben Saxon from the Women's and Children's Hospital in Adelaide will be leaving to work in Toronto for two years in mid September. In the first instance Dr Tom Revesz will cover clinics, however a clinician is expected to take on a longer term role late this year.

Royal Prince Alfred Hospital, Sydney

Leola Farnell, haemophilia counsellor at the Royal Prince Alfred Hospital recently left the position after several years' service. A replacement counsellor has not yet been appointed.

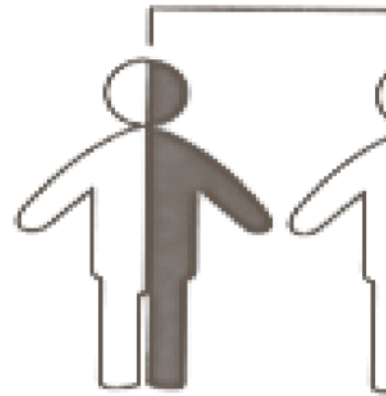
WORLD AIDS DAY


Friday, 1 December 2006

AIDS Awareness Week –
Friday, 24 November to
Thursday, 30 November 2006

HIV/AIDS - Let's talk about it:
many faces, different stories

HFA Speaker's Kit



HFA has completed a Speaker's Kit which will be distributed to all State/Territory Foundations for use when they represent their organisation or HFA at a speaking engagement. The kit, which is compiled in a display folder, includes speaker's notes, powerpoint presentations to go with the notes, further information about bleeding disorders and issues affecting the community, school project sheets and activities to encourage audience participation. The resource is designed to assist people in their preparation for speaking engagements, and to ensure we portray consistent information and appropriate messages. 



Haemophilia Foundation Australia ~ Speaker's Kit



Global Feast 2006



in support of people worldwide
with bleeding disorders

Haemophilia Foundation Australia will join other international Haemophilia Foundations including New Zealand, Ireland, United Kingdom, Portugal, Iran, Argentina and Netherlands for GLOBAL FEAST 2006.

You are invited to be an official GLOBAL FEAST host. Time your event around the month of September 2006. Invite your family, friends and work colleagues. Ask them to bring a donation instead of flowers, wine or a gift. Explain that the proceeds will go to HFA programs and services and to support people worldwide with bleeding disorders. Make sure you add that every dollar donated will make a difference to the lives of people struggling with lifelong and often disabling disorders.

Did you know that 75 % of the global population of people with haemophilia is not diagnosed and that 25 % do not receive adequate treatment

If a dinner isn't your "cup of tea", any type of festive event will do — a pancake breakfast, pizza party, backyard barbeque, afternoon tea or

picnic lunch. Be creative and have fun!

If you are interested in participating in GLOBAL FEAST please contact us. We will send you everything you need to make your event a success; brochures, promotional items, posters and tax deductible receipts for your guests.

It's so easy, but it will make a huge difference to the lives of others.

What can you do?

- Organise a morning or afternoon tea at your workplace.
- Organise a sausage sizzle outside your local supermarket (please seek permission).
- Contact your local restaurant and ask them to participate. Invite your closest friends for a meal.
- Organise a dinner with members at your Haemophilia Foundation.
- Host a BBQ street party.
- A cooking demonstration party to share the secrets on how to prepare a favourite dish.

For more information or to register please call Natasha Coco on 1800 807 173, email ncoco@haemophilia.org.au or visit the official GLOBAL FEAST website www.globalfeast.org. 

HAEMOPHILIA FOUNDATION AUSTRALIA WILL CELEBRATE GLOBAL FEAST AT THE CITY SQUARE IN MELBOURNE WITH A SAUSAGE SIZZLE TO RAISE FUNDS FOR WFH AND HFA.

COME DOWN AND BUY A SAUSAGE!

Friday 6 October 2006

Melbourne City Square, Corner Swanston and Collins Street
11am-2pm

Did you know that 75 % of the global population of people with haemophilia is not diagnosed and that 25 % do not receive adequate treatment

VP MEDICAL WRITES

Review of Hemophilia 2006 medical sessions

Dr. Paul Giangrande
WFH Vice President Medical

Medical experts share knowledge in state of the art medical program.



Particular thought went into the selection of the topics and speakers for the plenary sessions in the congress medical program, which aims to provide an insight into the very latest developments in hemophilia treatment.

Orthopedics

We heard from Luigi Solimeno (Italy) that elective orthopedic surgery in patients with inhibitors is now feasible, although the outcome may not be as good as in non-inhibitor subjects, as the incidence of hemorrhagic and infectious complications appears to be higher. The very high cost of blood products needed to cover the surgery is often a significant obstacle.

Goris Rosendaal (Netherlands) provided insight into how hemophilia arthropathy develops in the first place, explaining that the liberation of iron from red blood cells released into the joint space associated with joint bleeds leads to the generation of destructive oxygen metabolites, which in turn causes premature death (apoptosis) of cartilage cells.

Prophylaxis and hemophilia care

The role of prophylaxis in preventing development of joint damage was emphasized in a session chaired by Victor Blanchette (Canada). Although the efficacy is now clear from several long-term studies, many questions still remain unanswered, such as when to start, exactly what dose regimes should be employed, and whether it is ever feasible to stop in adulthood. In a separate session devoted to pharmacokinetics, chaired by Martin Lee (U.S.), it was emphasized that there is considerable inter-patient variation in the elimination profile of both factor VIII and IX. Pharmacokinetic evaluations may have an important role in optimizing prophylaxis regimes in individual patients.

Bruce Evatt looked at how hemophilia care has developed in the last few decades. He emphasized the danger of complacency in the hemophilia community and the need for vigilance to maintain what we have already achieved. Evatt highlighted the difficulties in recruiting medical personnel with an interest in hemophilia care in many countries and he recommended broadening the scope of work to encompass thrombosis as well.

Hemophilia and the elderly

This was the first congress to have a session devoted to the problems of elderly patients with hemophilia. Alison Street (Australia) highlighted that colleagues in disciplines such as geriatrics will now have to become familiar with the condition and become members of our multidisciplinary team. Her key message was that the life expectancy of people with hemophilia in affluent countries is now very similar to that of the rest of the population. Balance dysfunction in elderly patients needs to be assessed. The management of myocardial infarction (heart attacks) can be particularly problematic in older patients with hemophilia, as conventional treatment typically involves the use of potent drugs that inhibit blood coagulation. As patients live longer, attention is rightly turning to measurement of quality of life as well as longevity.

Quality of life

A session chaired by Alessandro Gringeri (Italy) explained the application of new questionnaires specifically designed to measure quality of life in hemophilia, and showed how the results can be used to explore data for clinical research. The data could be used, for example, to determine cost-benefit analysis as well as to determine patient treatment preferences and satisfaction. Gringeri argued that periodic quality

of life measurement should become part of the routine review of patients.

Inhibitors

Charles Hay (U.K.) chaired a debate between Claude Negrier (France) and Keith Hoots (U.S.) on whether the incidence of inhibitor development is higher among recipients of recombinant products as compared to those who receive fewer, pure plasma-derived products. Several small studies suggest that there is a difference but it is clear that further prospective randomized studies will be required to address this important issue. The WFH is also keen to encourage sharing of data between databases in different countries in order to provide the statistical power to answer this important question.

Jan Astermark (Sweden) explored the fundamental question of why patients develop inhibitors. It is not disputed that the underlying genetic defect plays a major role in determining predisposition. Astermark reviewed other contributing factors including data from the Malmö International Brother Study (MIBS), which suggests that immune response genes are also important. He explained that certain polymorphisms of interleukin-10 and tumour necrosis factor (TNF α) are associated with a high risk of inhibitor formation.

New treatments

David Lillicrap (Canada), chosen to present the prestigious Arosenius Lecture, described his work in the field of gene therapy (see page 12). This was complemented by a session on ethical aspects (chaired by Donna DiMichele, U.S.), in which aspects relating to gene therapy was one of the topics considered. There was also debate on the place of gene therapy in the context of the current state of hemophilia care in which safe and effective treatments are already available.

Speakers emphasized that safety must be the first and foremost consideration in clinical trials. Steven Pipe (U.S.A.) explained in his plenary lecture how targeted modification of the structure of the factor VIII molecule could yield recombinant products with enhanced properties such as a prolonged half-life or reduced immunogenicity. Another potential strategy to prolong the effective half-life of factor VIII would involve reducing clearance from the circulation through pharmacological blockade of hepatic receptors, which normally take up factor VIII from the blood.

Akira Yoshioka (Japan) chaired a separate session on novel therapies. We heard of a new treatment under development in his department, consisting of an antibody capable of imitating the activity of factor VIII by modulating the interaction between factor IXa and factor X. In an *in vitro* experiment, the addition of the bispecific antibody to factor VII deficient plasma shortened the clotting time but without impairing factor VIII activity, which suggests an additive effect.

The session also included a presentation of preliminary data relating to the efficacy, immunogenicity, and pharmacokinetics of recombinant porcine recombinant factor VIII, which is under development as a treatment for patients with inhibitors. Some data on the use of recombinant factor VIII formulated in polyethylene glycol-treated liposomes was also presented. This appears to prolong the action of the factor VIII and this study reported considerable prolonga-

tion in bleed-free periods with prophylactic infusions. The new formulation provided 13 days of protection as compared to seven with conventional product.

Other bleeding disorders

The recent WFH strategic review recognized the need to broaden the scope of our work to encompass both von Willebrand disease as well as the rarer disorders of coagulation. The program amply covered both themes with a session on rarer disorders chaired by Flora Peyvandi (Italy). There is a need to pool experience on the rarer disorders and an international database has been established (www.rbdd.org).

There is also a need to develop specific coagulation factor concentrates for many of these conditions, as in many cases the only treatment available is still only fresh frozen plasma. Much is known about the genetic basis of these conditions and this could represent an important tool for prevention through prenatal diagnosis and counselling; many of these conditions are inherited in an autosomal recessive fashion and arise because of parental consanguinity.

Laboratory sessions

Laboratory support is a vital component of hemophilia care. A separate session on platelet disorders, chaired by Catherine Hayward (Canada), focused on new diagnostic tools, such as the use of flow cytometry. Recombinant activated factor VII may be of value in the treatment of some of

these disorders. Robert Montgomery (U.S.) gave a plenary lecture on von Willebrand disease in which updates on the classification, laboratory diagnosis, and molecular basis were presented.

Clinical aspects were explored further in a session chaired by Peter Kouides (U.S.), in which the potential impact on women was explained. Many women can suffer from menorrhagia and it is important to at least consider the possible diagnosis of vWD in women with this problem. Intranasal desmopressin is effective treatment in many cases if the response to tranexamic acid alone is not sufficient.

Another session devoted to vWD, chaired by Augusto Federici (Italy), explored the role of desmopressin in the various subtypes of vWD. No recombinant VWF concentrate exists yet and the session also reviewed the various plasma-derived products available. These all contain a good VWF multimer profile are subjected to viral inactivation processes, and have proved effective in surgery. However, it is now appreciated that there is the potential for venous thromboembolism in the setting of surgery with these concentrates and laboratory monitoring is recommended in order to ensure that levels of factor VII do not exceed 150 iu/dl.

I would like to thank all members of the medical program committee for their tireless and valued work in helping to develop the program for the Vancouver congress. ■

Doctors review the congress program.



New directions in diagnosis and care

Maureen Spilsbury
Chair WFH Psychosocial Committee
(with other reports)

Participants from both developing and developed countries benefited from an extensive range of psychosocial, nursing, and other congress sessions on inherited bleeding disorders.



Delegates discuss a previous session

A highlight of the psychosocial program was Parents Empowering Parents (PEP), a pre-congress workshop that focused on the unique problems faced by parents of children with bleeding disorders. Designed by U.S. social worker Danna Merritt, PEP is presently used only in the U.S., but hopefully will be piloted globally over the next year.

Miriam Rodriguez from Uruguay looked at programs to increase the quality of life of children and families by increasing their self-esteem and involving them in activities and workshops.

All speakers agreed that burnout affects many families and paid helpers of people with hemophilia. In response to reports of overwhelming stress, one congress session examined the causes of burnout as well as strategies to deal with it.

The effects of HIV and HCV

Dejan Krstic from Serbia was given a standing ovation for his account of the impact of HIV and hepatitis C (HCV) on his community, where approximately 20% of hemophilia patients are affected by HIV and 45% by HCV. Delin Kong from Shanghai also described the local implications of viral infection and his attempts to raise public awareness of the situation.

Research by U.S. social worker Susan Cutter finds that although HIV and HCV can have the same clinical effects on individuals no matter where they live, significant differences occur between developing and developed countries in

the incidences of infection, available treatment options, perception of the infections, and impact on quality of life.

Brazilian psychologist Frederica Cassis has found that emotional support and individual and group psychotherapy for people with HIV and HCV help participants to cope with the reality of constant treatment and live a productive life.

Robert James presented a moving session based on the U.K. website: "Living Stories: the haemophilia and HIV life history project. (<http://www.bl.uk>) This resource provides a unique insight into individuals' experiences of living with hemophilia and terminal illnesses. "I am always touched by hearing accounts of family members and worker's own experiences and can never get enough", said one social worker. "In a very powerful way, it connects me to other people who are in similar situations – it's healing!"

Nurses focus on awareness and early diagnosis of rare bleeding disorders

Better awareness and knowledge about rare bleeding disorders, coupled with earlier screening and diagnosis will improve care, medical outcomes, and patient quality of life, according to panellists at a pre-congress nursing workshop.

Several case studies demonstrated that diagnosis of disorders such as factor XII deficiency is often slow. In a U.K. case cited by Mary Edgar, it took 26 days and multiple severe bleeding incidents before the patient was referred for appropriate care. Even after diagnosis, Edgar said, "We're often working

in the dark; there's very little pharmacokinetic data, so a lot of the time it's trial and error with a bit of science applied."

"It's also important to understand the patient's particular circumstances and challenges to provide effective treatment," according to Nadine Alliaume, from France. "Family situation, cultural considerations, distance from treatment, and problems with compliance all affect care."

Nathalie Aubin directed participants to www.hemophilia.ca for a series of information booklets published jointly by the Canadian Hemophilia Society and the Canadian Association of Nurses in Hemophilia Care.

Women with bleeding disorders 'a silent majority'

Women with bleeding disorders are a "silent majority" who lack proper diagnosis and care, with those in developing countries facing particular challenges due to social, economic, and cultural barriers, according to Roshni Kulkarni of the U.S.

Beyond von Willebrand disease, bleeding disorders in women also include coagulation factor deficiency, immune thrombocytopenic purpura, platelet function disorders, and acquired hemophilia.

Early diagnosis of women's bleeding disorders is critical to avoid unnecessary and invasive procedures such as hysterectomies, while early treatment can prevent severe manifestations such as athroplasty, Kulkarni said. ■

Musculoskeletal sessions explore new treatments

Kathy Mulder
Chair WFH Musculoskeletal Committee

The latest research from 19 countries presented at congress.

Musculoskeletal sessions at the 2006 world congress included assessment tools, surgical and non-surgical treatment techniques, and guidelines for making decisions about the best treatment. Formal feedback from participants was highly positive.

Pre-congress workshops were held for physiotherapy/rehabilitation and orthopedics.

Physical therapists learned that bone density and levels of physical activity are important factors in addition to traditional measures such as joint range of motion and muscle strength.

A diverse panel, including consumers, shared views on "compliance." These included the necessity of communication and negotiation between client and care giver for more realistic expectations on both sides. Speakers from different backgrounds discussed physiotherapy management of contractures, which continue to be a challenge in the developing world. Finally, an open session enabled people to share their current and planned research activities. Many exciting projects are underway and a database will be developed to enhance information sharing.

An orthopedic pre-congress day, held for the first time, reviewed current knowledge and best practice in the areas of synoviorrhesis, joint surgery, bone substitutes, and surgery for people with inhibitors. It was standing room only when more than 130 orthopedic and physiotherapy/rehabilitation participants came together for a session on diagnostic imaging.

Musculoskeletal session participants also heard how bone grafting is being used successfully for the treatment of hemophilic pseudo tumours and cysts. Bone substitutes, genetic transduction, and autologous chondrocyte implantation are seeing success in controlled experiments, but are still far from being practical or readily available as a treatment technique for the tissue damage caused by hemophilia. Nevertheless, they do offer hope of some exciting possibilities.

Elsewhere, orthopedic surgery for people with HIV and/or hepatitis C presents some unique challenges. Risk of infection and anesthesia issues must be considered, and surgery for this group must be conducted in specialized hemophilia centres.

"Synovitis: what is the best time to treat?" is always a difficult question. The topic was explored by six speakers on the

second day of the congress. The short answer is: prevention is best but not always possible. The next best solution is early and as aggressive treatment by whatever means available in each treatment centre.

Other speakers presented updates on five important assessment tools: International Joint Health Scores (Clinical and MRI), Functional Independence Score for Hemophilia, and the Hemophilia Activities List. As these tools are refined, they will become the international standard for assessment of people with hemophilia.

Much of our understanding of management of hemophilic joints comes from other diseases. However, we are now developing evidence specific to hemophilia about some of the treatments we use. Dr. Goris Roosendaal summarized experimental work that shows that blood causes direct damage to cartilage. The damage increases if the cartilage bears weight in the presence of blood. This finding emphasizes the need for prevention or very early treatment of all bleeding episodes, and reinforces the importance of rest.

There is good experimental evidence that ice is an effective pain reliever, but other experiments show that ice may slow clotting. The hemophilia team in Pretoria, South Africa, designed a study to measure the direct effects of ice on bleeding in acute hemarthroses. At present there are some methodology issues, but the project is important to help us understand whether ice should be a regular part of hemophilia care.

In all, 30 speakers from 19 different countries presented original work. Topics ranged from laboratory work with cultured chondrocytes, to gait analysis and physical activity levels. While many of these studies are small and inconclusive, the authors are commended for trying to enhance the base of evidence for our practice. ■

Executive officers of the WFH musculoskeletal committee.

Left to Right: Tariq Sohail, Rachel Tiktinsky, Pier Luigi Solimeno, Kathy Mulder, Adolfo Llinas.



WFH General Assembly embraces vision of Treatment for All

Miklos Fulop
WFH CEO and Executive Director

With 109 National Member Organizations, the WFH's reach continues to grow.

Following a highly successful world congress, the 28th WFH General Assembly at the Morris J. Wosk Centre for Dialogue in Vancouver brought together delegates from national hemophilia organizations to review recent accomplishments and set future directions.

Great progress has been achieved on many fronts through the WFH since the last general assembly. This could not have been done without the leadership of WFH president Mark Skinner, invaluable direction from the executive committee, and the hard work of our staff and volunteers.

The network continued to expand, with Sri Lanka achieving full membership, while Bahrain, Japan, and Lesotho joined the fold to bring the total number of WFH National Member Organizations to 109.

Treatment for All

The general assembly heard an overview of the WFH's new strategic plan focused on making available treatment for all those with inherited bleeding disorders, regardless of where they live.

"Treatment for all means ensuring care for everyone in our community — that all people with hemophilia have access to a safe and adequate supply of products and care by a multidisciplinary team," said WFH president Mark Skinner.

"It also means expanding services to von Willebrand disease, rare deficiencies, and inherited bleeding disorders. Another focus of the strategic vision is to ensure continued development and sustain treatment where it is already well established."

Over the past 18 months, WFH hemophilia care programs reached more than 75 countries. Efforts to help build local capacity expanded, with 33 ongoing country programs in 2006, while another 42 countries were involved in general program activities. The first three-year cycle has been successfully completed by the Global Alliance for Progress (GAP) project, which aims to close the gap in hemophilia care between developing and developed countries. To date, six agreements have been signed with governments to establish national hemophilia care programs. The WFH also supports 49 active twinning partnerships worldwide.

Delegates wholeheartedly supported the vision of Treatment for All, providing the WFH with a plan for the future focused on improving standards of treatment for all rare bleeding disorders. The plan also involves monitoring, advocacy, research, training and education, and resource management.

New executive committee members

The general assembly saw four executive committee positions up for election this year: Dr. Angelika Batorova (Slovakia) and



Dr. Grace Phiri of Lesotho accepts WFH membership on behalf of her national hemophilia organization.

Dr. Nigel Key (U.S.) were elected as medical representatives, and re-elected as lay members were Gordon Clarke (U.K.), and César Garrido (Venezuela).

Departing executive members

This year, several executive committee members completed their terms. Distinguished hematologist Bruce Evatt ended his 10-year term of office as VP programs. Bruce has been a tireless WFH volunteer and his outstanding scientific achievements have contributed immensely to the world hemophilia community. The general assembly also acknowledged the service of Mohamed Aris Hashim as a lay member on the executive committee, and medical representative Mammen Chandy.

Congress 2010

Representatives from the Argentinean and French hemophilia societies gave presentations for hosting the 2010 world congress. The winning vote went to Argentina, and Buenos Aires will be the site of the 29th WFH World Congress.

With a new blueprint for success, the WFH is energized for the work and challenges ahead. "We have made great progress on all fronts," said Skinner. "We must sustain the many gains we have achieved thus far. Through focused and deliberate implementation of this plan, we will move closer to making our vision a reality." ■

Recent advances in the search for a hemophilia cure

Professor David Lillicrap
Queen's University, Kingston, Canada

Experts 'cautiously optimistic' about the prospects of gene and cell-based therapy for hemophilia.

The goal of a long-lasting treatment for hemophilia using innovative gene and cell-based strategies remains a feasible objective, as seen from papers presented at three independent meetings last spring. Recent advances were described at the U.S. National Hemophilia Foundation workshop on gene therapy in Philadelphia, the WFH Congress in Vancouver, and the annual meeting of the American Society of Gene Therapy (ASGT).

The best results to date from clinical hemophilia gene transfer have occurred in the Avigen-sponsored trial in the U.S. using a recombinant adeno-associated viral (AAV) vector to deliver factor IX to the liver. (A vector is a method to transfer DNA sequences from one organism to another.)

In one patient studied, a plasma factor IX level of 12% was achieved and therapeutic levels were expressed for a month before returning to baseline levels, accompanied by evidence of liver cell toxicity. Subsequent immunologic analysis in another patient showed that liver damage following vector delivery is most likely due to a host immune response against components of the vector protein coat. In other words, the patient's immune system recognized the vector from previous virus exposure and attacked it.

Despite this setback, there is still well-founded enthusiasm for the AAV delivery system. Recent studies in hemophilia A dogs show that long-term expression of therapeutic levels of factor VIII can be achieved with this vector, while two upcoming human clinical trials will use AAV delivery in hemophilia B patients.

A study coordinated by Dr. Kathy High in the U.S. intends to use a temporary course of immunosuppression to abolish the host immune response to the vector. A second study involves centres in the U.K. (Drs. Amit Nathwani and Ted Tuddenham) and the U.S. (Dr. Andrew Davidoff). In this second study, researchers are trying to circumvent the host immune response by switching to a vector with a different protein coat (AAV8), which may be less likely to be recognized and attacked. The trials may begin to enroll patients later this year.

There has also been progress with the preclinical application of certain types of retroviral vectors. Retroviral delivery systems have the capacity to insert their clotting factor gene "cargo" into the patient's chromosomes, and



Professor
David Lillicrap
in Vancouver.

are thus well suited for the modification of stem cells that can then serve as depot sites for clotting factor expression. (Stem cells are cells that retain the ability to become specialized and take the place of cells that have a specific task.)

Encouraging preliminary data has shown that this approach might be feasible using patient blood stem cells, with subsequent re-introduction of the genetically modified cells into the patient for long-term factor delivery. The retroviral strategies will still need to demonstrate a lack of immune activation and, depending upon where the cells have been re-implanted, long-term viability.

As several different strategies for hemophilia gene transfer are explored in pre-clinical studies, the challenge of the host immune response continues to limit long-term success. Evidence suggests that the delivery of clotting factor genes to specific parts of the body, such as the liver, may facilitate the development of immunologic tolerance to the newly expressed clotting factor protein.

Furthermore, recent studies presented at the ASGT meeting indicate that gene transfer approaches using the cell's own regulatory machinery can limit protein expression to defined cell types, and thus minimize the risk of adverse immune responses such as clotting factor inhibitor development.

Overall, the hemophilia gene transfer community remains cautiously optimistic about future advances towards the goal of long-term correction of the bleeding tendency in hemophilia. The next year will undoubtedly provide new information about the rate and extent of this progress. ■

The following information about two promising strategies for advancing gene therapy in haemophilia is reprinted from information provided by WFH on its web site www.wfh.org.

The information is based on material that was presented at the US National Hemophilia Foundation's Eighth Workshop on Novel Technologies and Gene Transfer for Hemophilia, in Philadelphia 31 March - 1 April 2006. The NHF workshop was supported with funding from the Katharine Dormandy Trust, the US Centers for Disease Control, the Coalition for Hemophilia B, and the World Federation of Hemophilia.

The WFH plans to provide ongoing reports on developments in gene therapy including news of new clinical trials on its web site and in publications. Readers are also referred to the article by Professor David Lillicrap in this edition of "National Haemophilia".

Gene Therapy

Although researchers have already used gene therapy to effectively treat mice, dogs, and other animals that have haemophilia, transferring these innovative therapies to humans has proven more difficult.

In the clinical setting, the human immune system eventually rejects the gene transfer and, as it attacks the delivery system (called "vector"), factor levels fall below what is acceptable. Two methods of overcoming this problem may provide exciting results in the next couple of years.

People with haemophilia have specific genetic defects that prevent their bodies from producing sufficient levels of clotting factor. So far the most promising method of gene therapy in haemophilia has been the use of viral vectors – modifying a virus so that it carries genetically modified cells for factor delivery.

Viruses are very effective at tricking the body into replicating their genetic contents, and haemophilic dogs treated with this method have maintained therapeutic levels of factor for years. Although it has been shown that factor levels can be corrected by using viral vectors to deliver corrected genetic material, the human immune system seems to recognise the vector as harmful from previous exposure and attacks, eliminating the positive outcome.

Scientists are trying to overcome this rejection of the viral vector in a couple of ways. One is to use temporary immunosuppression to prevent the body from rejecting the

vector when it is administered. The idea is to eliminate the immune response so that the vector will continue to work. A second strategy is to try different vectors in the hope that the human immune system may not react in the same way to slightly different viral delivery systems. Both of these methods are now being attempted in clinical trials in the US and UK.

An alternative strategy to using viral vectors to deliver gene therapy is the use of retroviral vectors. These vectors work to insert the clotting factor gene into the patient's chromosomes. One version of this therapy uses the patient's own stem cells: the retroviral vector is inserted in the lab and the genetically modified stem cells are then re-introduced into the patient. (Stem cells are undifferentiated "blank" cells that do not yet have a specific function.) This therapy is in much earlier stages of development but it may potentially be effective in delivering long-term therapeutic levels of clotting factor.

For further information and recent publications on gene therapy see:

Blood, March 2006; Vol 107 No 5

Blood, April 2006; Vol 107 No 7


Nature Medicine, 2006 March 12(3):342-7 

Our Corporate Partners

HFA is delighted to have commenced a new three year relationship with Corporate Partners - Baxter, CSL Bioplasma and Wyeth. HFA recently developed a Memorandum of Understanding with each of these companies to recognise the common objectives of improving care and treatment to the bleeding disorders' community and the special relationships, opportunities and responsibilities the relationship creates.

HFA initiated its Sustaining Patrons' Program in 1998 and the program has been a valuable resource that has enabled HFA to meet its objectives ever since.

In 2006 the Program was renamed the HFA Corporate Partners' Program and Memorandums of Understanding were established to reflect the framework of cooperation and understanding and recognise our shared objective to improve the health and welfare of individuals and groups of people in the Australian bleeding disorders' community.

The partnership also enhances opportunities to strengthen the knowledge and information needs of the bleeding disorders' community by education of patients and their families, and multidisciplinary health professionals through conferences, special projects, and other activities. 

Baxter

CSL Bioplasma
Biotherapies for Life™

Wyeth



**National Hepatitis C Awareness Week will be held
1-7 October 2006.**

The 2006 Awareness Week aims to promote general public awareness of hepatitis C, with a special emphasis on effective self-management practices for people living with hepatitis, their friends and family, and those involved in their care.

The Australian Hepatitis Council established a Project Reference Group to work with State/Territory Hepatitis Councils, Haemophilia Foundation Australia (HFA), Australian Injecting and Illicit Drug Users' League (AIVL), Tasmanian Council on AIDS Hepatitis & Related Diseases Inc (TASCAHRD), Australasian Society for HIV Medicine (ASHM), Northern Territory AIDS and Hepatitis Council (NTACH) to develop material and events for the week.

The Hepatitis C Awareness web site will have a range of valuable information about hepatitis C for people with hepatitis C and their families and there is a national toll free telephone number for inquiries.

<http://www.hepcawareness.net.au/>

**Call the national toll free hotline number
on 1300 437 222.**

The following information about stress is abridged from material compiled by the NSW Association for Mental Health from Strategies for Stress (Dr David Lake) and Coping with Stress (Leichhardt Women Health Centre) for a Fact Sheet published by the Hepatitis C Council of NSW in April 2004.

Stress And Hepatitis C

Stress is an area in our life that we all need to keep under control. If stress is allowed to increase, unchecked, the impact on one's physical and mental health can be severe. Some typical signs of stress are insomnia, headache, neck, shoulder and back pain, heart palpitations, fatigue, irritability, panic attacks, loss of concentration and low self esteem. There are a number of areas in our lives, though, where we can take action to control our levels of stress.

The positive sides of stress

Stress is not necessarily always negative. Without the action of positive stress, many people would not attempt to reach their full potential. For example, competitive tension usually keeps a footballer or salesperson working to their best. There is an optimal level of stress at which a person is likely to be most productive. Knowing one's particular stress level is useful.

The negative side of stress

When pressure is continual and when the feeling that you are not in control stays with you over a long period of time, you're probably suffering the negative effects of stress. When this happens you'll feel overworked but will actually be contributing less and less. Physically, pulse rate will increase, blood pressure will rise, perspiration increases and there is an increase in blood adrenalin levels.

Symptoms

You may feel irritable, anxious, nervous or fatigued. There is often a change in appetite and perhaps an increase in cigarette smoking or drinking alcohol. Other symptoms may include diarrhea, abdominal cramps, insomnia, trembling, urinary infrequency, increased heart rate and headaches or backaches. As mentioned previously, palpitations, fatigue, irritability, panic attacks, loss of concentration and low self esteem may occur as well. To date, surprisingly little long-term research has been done on the effects of stress - although it has been linked to ulcers, infertility, heart disease, mental illness and cancer.

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Hepatitis C Awareness Week
1-7 October 2006

• lifestyle • health care and treatment • symptoms
• family, friends and work • support and services

Stress and lifestyle

Stress is a normal response to lifestyle and work, and the general challenges of living. A certain level of stress is fine and even necessary in order to be productive. However, too much stress can be a health hazard. People need to learn to be aware of when their stress level is unhealthy. Once stress overload is recognised you can assess your current lifestyle, but there are skills you can use to address stress - see below.

Nutrition

When we experience stress and nervous tension, our bodies use up large amounts of nutrients (vitamins and minerals.) This can cause dietary deficiencies which in turn leave us tired, rundown, irritable and less able to cope with stress-back to square one!

Nutrients which are particularly related to stress are Vitamin C, the B complex vitamins, and the minerals calcium, magnesium, phosphorus and zinc. These nutrients can be supplemented by eating healthy foods high in vitamin and mineral content, or by taking tablets or medicinal herbs.

Diet considerations

The best diet to cope with stress contains fresh wholesome foods:

- lots of fruit and vegetables, beans, lentils, bread & cereals
- moderate amounts of dairy foods, lean meat, skinless poultry, fish & nuts
- small amounts of oils and fats
- salt should be avoided

Counselling

We all live and interact with other people and often cope better with our problems and life stresses by talking and sharing our feelings with other people. This can be as simple as talking with a neighbour or friend over a glass of juice or mineral water or writing a letter or story for your favourite newsletter!

Some people may need to phone a friend or relative specifically to talk about a particular problem. Additionally, many people choose to go to a trained counsellor, social worker or therapist.

Take some control over the thoughts that go through your mind. Talk yourself out of repeating negative or unhelpful self thoughts.

Acupuncture

Acupuncture has been shown to be a very effective treatment for various conditions as well as a stress management tool. It's best to see a qualified practitioner who belongs to a professional body.

Massage

Whether done by a trained professional or just a friend, a massage is a simple and inexpensive way of releasing tension from our bodies. If done regularly, a massage can also make for a welcome break from a daily routine.

Meditation

Meditation is a technique that offers a structured way to reduce tension and stress. Although it may not provide an immediate release of stress, over time a meditation program will prepare you to handle physical and mental problems and recharge your system. Structured exercise and concentration programs such as Tai Chi and Yoga are also very effective in reducing stress.

Choose your attitude

Take some control over the thoughts that go through your mind. We all have such thoughts, some of us more so than others. Talk yourself out of repeating negative or unhelpful self thoughts. You will be surprised at how much difference this can make after a short period of time.

Relaxation


Relaxation can mean spending time at the beach or local park, doing hobbies or listening to your favourite music. The big obstacle to relaxation is physical tension. Relaxation is often the end result of stress management exercises such as those mentioned above.

Exercise

Physical exercise can often help to release tension and improves your overall level of health. The most helpful thing is to do exercise moderately and regularly. Any physical exercise such as jogging, swimming, gardening and playing

team sports is helpful. It might help to talk to your doctor about the suitability of new activities if you have a bleeding disorder or other health problems.

Some tips to remember

- Be aware of physical and mental stress and recognise the signs. Stress is only a problem if it is excessive or if you are not handling it properly.
- You are not the only one who experiences the problems that lead to stress - such problems are a normal part of everyday life.
- Try not to make problems any bigger than they actually are. Identify situations that are stressful for you - deal with them or avoid them.
- Practise relaxation techniques.
- Find interests that really involve your concentration.
- Plug into support networks - formal or informal.
- Grow a garden and get a pet if practical.
- Take regular time out.
- Talk to others about your feelings.
- Live in the present and not in the past.
- Cultivate regular stillness and quietness in your life.
- Set daily routines, giving yourself time for some of the above ideas. 



Angela Kelly's experience earlier this year in China when she was working with people with HIV and her encounters with people with haemophilia reminds us of the importance of having people to talk to and share difficult experiences and fears with others.

The Power Of Story

Angela Kelly

As I walked down the corridor in the infectious disease department of Beijing You An Hospital I saw the sign to the Mangrove Support Group (MSG) which was the first support group for people living with HIV/AIDS in China. We knocked on the door and we were invited in. I was introduced as an Australian Youth Ambassador for Development working as an HIV educator in the hospital, specifically within the organisation called Home of Loving Care which provides care and support to people with HIV in hospital. I was welcomed in and offered a seat and a warm glass of water. I shook the woman's hand as I wanted her to know that I was not afraid of HIV. Soon after, a man came into the room on crutches. He had a firm hand which I found refreshing. He sat, and the woman took his crutches. I thought nothing of it.

Within a short time he indicated that he uses the crutches because his right leg does not have full movement. As I should have already guessed, but did not, he has haemophilia. If I were in Australia I may have thought of it. I recall many images of Andrew Knox, my late partner, on crutches and we always had a pair in the cupboard. In a foreign land where I had heard so much about the plasma infected with HIV, I had not turned my attention to those with haemophilia. There is little discussion here of people with haemophilia infected with HIV, but there is much general discussion about infections via blood transfusions.

The exact number of those with haemophilia who have died of complications associated with HIV/AIDS is unknown due to

He had never met anyone with either haemophilia or HIV. He had no friends who knew of his HIV status. HIV was a secret burden. His mother slept in the hospital bed next to him during his stay in hospital. Each time I saw her she cried.

inadequately developed surveillance systems. MSG is currently preparing to conduct a survey on the needs of people with haemophilia and HIV in China and use it as an advocacy tool.

During my six month stay in China I met three men with haemophilia and HIV. I also met the mother of one of the men and the wife of the other. I had not anticipated meeting so many people affected by haemophilia in my work here in HIV yet it was these meetings that were the most powerful for me. They were powerful interactions because they reinforced for me the importance of the haemophilia community both at formal and informal levels in Australia and abroad. It is within the community of those affected by haemophilia that we can most easily share stories and experiences that make living with both haemophilia and HIV possible. It is what makes the transition from dying with HIV to living with HIV possible.

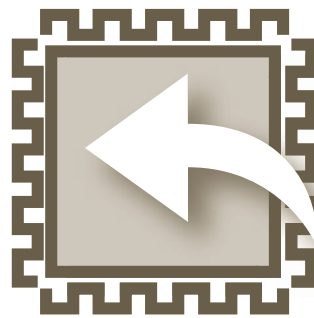
The youngest boy I met with haemophilia and HIV was a 16 year old who lived in rural China with his parents and brother. He had never met anyone with either haemophilia or HIV. He had no friends who knew of his HIV status. HIV was a secret burden. His mother slept in the hospital bed next to him during his stay in hospital. Each time I saw her she cried. The mother pulled up her son's trouser on his left leg to reveal a

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When I shared with her my own story of being the partner of a man with haemophilia and HIV she began to cry. What was most important was sharing my partner's love for his mother and his appreciation of her for giving him life.

knee badly affected by his haemophilia bleeds. Unsure of how I could reach this young man whose face was permanently solemn, I shared with him that I had friends with haemophilia and HIV in Australia. Showing the first bit of interest in my visit, he looked at me and asked if I would tell him the story of one of them. I told him about a young man in Australia who I know. When I finished he said 'hen hao' which means it was a good story. This young man's story was an example of what is possible. After that I asked if he would like to meet another person with haemophilia and HIV. He was still afraid, but he said he wanted to do so. I introduced him to one of the men from MSG and they talked for two hours. Already proficient in using the computer to play games, he was shown how to send an email so they could stay in touch. On a practice email the young man wrote 'I hope we can become good friends'. This young man's isolation was no longer. He had a living example of what was possible in his life – that he could live with haemophilia and HIV.

That young boy's isolation was lessened by that interaction, but it was the isolation of a mother I met in later weeks whose isolation concerned me most. She and her family lived in a province north of Beijing where services for people living with HIV/AIDS were poor and where there was little acknowledgement of the virus. She showed me her son's test results, his CD4 and viral load tests over the past few years. He was doing well



YOU'VE
GOT MAIL

medically with a stable CD4 count of 350. His hepatitis C was under control and best of all he was happily married. Her son was now 32 and had been diagnosed 10 years ago. The tragedy of this family's story did not reveal itself naturally. It was only when I asked how his wife accepted a boyfriend, then husband with HIV that her secret was shared. Grief stricken with the guilt that she caused her son to have haemophilia she could not bear to tell her son that he was infected with HIV although he knew about his hepatitis C. With the support of the doctors, they had colluded to keep her son's secret from him and now his wife. It is a secret she thinks spares him pain and lets him live, but it is one that is killing her.

When I shared with her my own story of being the partner of a man with haemophilia and HIV she began to cry. What was most important was sharing my partner's love for his mother and his appreciation of her for giving him life. A life for him with haemophilia and HIV was better than no life at all. I shared that her guilt was not unique and that other mothers shared this in Australia and other countries, China too. She drew strength from sharing this story and I can only hope it may enable her to

face her son honestly and tell him that in spite of all of her love for him there were things that she could not protect him from and because of that love she could no longer go on protecting him from it. It is only by sharing with her son that she will be able to transition into a phase a 'living' with the knowledge of her son's HIV status. And, in turn, it will only be then that her son and his wife can learn to live with HIV and take the necessary precautions if it is not already too late. **H**

WFH Twinning Program

The WFH Twinning program, a strong example of the WFH vision of *Treatment for All*, has enjoyed much success and growth in recent years. The WFH Twinning program is sponsored by Wyeth and there are now 48 twins in 51 countries throughout the world.

Linking haemophilia organisations and treatment centres in developing and developed countries is a positive two-way experience for the twins, and results in improved diagnosis and treatment and care, staff and volunteer motivation, and strong relationships which encourage and improve care – often in a stepwise manner, according to the situation. Planned twinning activities included outreach, education programs, and training in fundraising and advocacy. WFH works with twins to clarify objectives and provides support to twinning partners.

In 2005-2006, several of the new twinning partnerships have extended the program into new territories with the inclusion of Cape Verde-Coimbra (Portugal); Guatemala-Colombia; Honduras-Georgia (USA); Tunisia-Quebec (Canada); South Africa-Canada; Shiraz (Iran)-Milan (Italy); Oran/Sidibelabbas (Algeria)-Strasbourg (France); Dharwad (India)-Detroit (USA); Armenia-United Kingdom; San Salvador (El Salvador)-Houston (USA); and Vientiane (Laos)-Brest (France).

The Hemophilia Treatment Centre, and Hemophilia Organisation twinning awards were presented in Vancouver at the World Hemophilia Congress in May 2006. Twin of the Year Awards for 2004 were presented to three sets of twinning partners:

- Dr Robert Reid, Cabral Children's Hospital, Dominican Republic and the National Hemophilia Centre of Venezuela
- Lebanese Hemophilia Association and the University Hospital of Geneva Hemostasis Unit, Switzerland; the Foundation of Support to Hemophiliacs
- Dominican Republic and the Venezuelan Association for Hemophilia

Twin of the Year Awards for 2005 were presented to

- Fairview University Medical Centre, Minneapolis, USA and the Institute of Hematology, Yerevan, Armenia
- Pakistan Hemophilia Patients Welfare Society and the Haemophilia Society of Malaysia
- Centre d'hémophiles, hôpital de Caën, France, and the Moroccan collaboration of the Hopital d'enfants, Casablanca, and the Hopital d'enfants 

Linking haemophilia organisations and treatment centres in developing and developed countries is a positive two-way experience for the twins, and results in improved diagnosis and treatment and care, staff and volunteer motivation

The following is abridged from an article which appeared in the *State of the Art Book, XXVII International Congress of the WFH, "Haemophilia" journal, July 2006, Vol 12, Supplement 3, pages 13-21. For the full text and references please refer to the journal or request a copy from HFA.*

Dr Bruce Evatt ended his ten year term of office on the WFH Executive Committee as Vice President Developing World, in May 2006. A standing ovation for his scientific achievements and outstanding contributions to the global haemophilia community at the Awards Ceremony at the Vancouver Congress in May 2006 was a special way to acknowledge his tireless work and dedication.

The Natural Evolution of Haemophilia Care: Developing And Sustaining Comprehensive Care Globally

Dr Bruce L Evatt

Comprehensive care is vital for patients with haemophilia to prevent early death and free patients from the complications that inhibit living normal lives. Experience has shown that once introduced in a country, there is a progressive restoration of normal healthy lives to the haemophilia community.

Accompanying this progress is a gradual decreased dependency on the haemophilia comprehensive centre – except during brief periods when expertise contained within the comprehensive centre is mandatory for life-saving clinical management or to prevent severe morbidity. During each stage of the natural evolution of comprehensive haemophilia care in a country, challenges to the existence of the centre occur, which threaten the comprehensive treatment concept. The haemophilia community must understand this natural evolution and be prepared to work collaboratively with governments, physicians and other patients to ensure that centres retain the expertise to meet the emergent needs when they arise.

Prior to the 1960s, when no comprehensive care was available, individuals with haemophilia suffered a similar fate worldwide. Severe joint disabilities appeared in early teens, and most patients died before the age of 20. Haemophilia was treated primarily with fresh blood

transfusions, and as a result haemophilia associations were established for the purpose of recruiting the donors.

Cryoprecipitate was discovered in 1964 and subsequent development of clotting factor concentrates dramatically increased clinical management options. As concentrates could be easily stored, administered at home and carried with patients during travel, home therapy was adopted. In developed countries, early treatment of bleeding episodes and home therapy quickly evolved as the primary management option. Training and education of patients about disease management became necessary with the increasing popularity of home therapy. Specialised centres soon delivered services to meet these needs. These approaches on patient care produced significant effects on patient general health and survival, and as a result, the haemophilia community requested and received support for networks of haemophilia treatment centres from governments.

The comprehensive care centre model provided comprehensive services based on an integrated public health approach and has been one of the most successful public health programs in many developed countries, resulting in significantly improved health for patients with haemophilia and reduced use of

health care services. These programs have been so successful at reducing the complications of haemophilia that the dynamics of haemophilia care have changed significantly. However, these changes are now producing other challenges for sustaining the current model.

Comprehensive care for haemophilia differs from routine patient care for several reasons. First, it is very specialised and affects many other areas of the patient's physical and mental health and is best met through a multidisciplinary team approach. Appropriately trained and experienced medical staff are needed to avoid poor therapeutic decisions that can lead to severe disability and mortality. The tasks of the comprehensive care team, which are critical to preventive care include diagnosis and assessment, education of the patient and his/her family, management of acute bleeding episodes, initiating and supervising home therapy, routine follow-up, preoperative assessment and postoperative management of the patient when surgery is necessary. In addition, the treatment centre provides support to the patient and family to prevent social or psychological distractions from interfering with health-care outcomes.

A primary treatment team is organised in each centre to maximise the effect of the resources available. In most centres, this consists of a

nurse coordinator, a medical director (usually a haematologist), a physiotherapist and a social worker. A referral team is organised to meet the specialised needs of individual patients and may include specialists in dentistry, orthopaedic surgery, clinical genetics, rheumatology, infectious diseases or other areas of clinical medicine.

Second, maintaining trained and experienced health-care providers can often be achieved only by concentrating patient care into specialised centres. The comprehensive care team works collaboratively with other centres, and these networks enable continual evaluation of treatment trends and modify treatment standards. To maximise the efficiency of the network, it is often divided into two to four layers or levels of treatment centres depending on the number of patients in a geographic area and the resources available. For example, a country may form a basic clinic level, a haemophilia treatment centre level and a comprehensive treatment centre level.

Third, priority is placed on preventive medicine, because complications of haemophilia, such as infection with hepatitis viruses, HIV and progressive joint disease, are severe and extremely difficult and expensive to treat. Successful modern haemophilia care depends on the principle of anticipating and preventing the complications of haemophilia rather than treating those complications following their occurrence. This principle of management of haemophilia has come to be termed comprehensive care and requires more resources and organisation of care delivery than the standard physician-patient relationship used for the management of many diseases. However, it yields dramatic results by producing productive individuals living normal healthy lives without the crippling effects and early death

produced by the complications of haemophilia.

Fourth, maintaining a coordinated network of speciality centres enables patient access to clinical research and evaluation needed to make well-informed decisions. Good prevention requires that patients and their families make well-informed decisions regarding their physical, psychological and social health, which is consistent with the most up-to-date medical knowledge. Under the comprehensive care model, the team has responsibility for assisting patients and their families with these decisions fall into the comprehensive care team.

Careful structure of the speciality centres enables optimal delivery of care based on the allocation of limited resources.

The Economics Of Haemophilia – The 'Nine Hundred Pound Gorilla'

Economics affects how countries of different economic development approach comprehensive care with their governments. Over 20 years, the cost of care has risen exponentially as improved safety of treatment products has produced a 5–10 times increase in cost. Optimum care is beyond the reach of individuals with haemophilia, so others governments or insurance must bear the cost. As a result, the haemophilia community must continually convince others to accept this financing responsibility. By nature, the payer's focus is on the bottom-line economics, ie 'the most served with the least cost'. This attitude complicates the efforts of the haemophilia community. In countries with emerging economies, economic issues will be the primary force determining how far patients and physicians can push their goals. In the developed countries, only constant pressure from patient groups prevents reductions in services for haemophilia patients, when these countries attempt to reduce rising health-care costs

In fact, if cost alone were the deciding factor in haemophilia care, it might be very difficult to convince governments of the developed countries to fully finance haemophilia care. For countries with emerging economies, the prospect is even worse. Cogent arguments against

considering the cost factor as the decisive one must include a consideration of the government's view of the cost factor on the nation's total health expenditures.

Many countries with low GNP may believe haemophilia care is not affordable in their country because they would need to spend significant (and often unrealistic) amounts of their total health-care budgets to provide adequate care for their relatively few patients with haemophilia.

To counter this, proposed programs must be compatible with each country's economy and it must be argued that the comprehensive care model can produce significant improvement in health status and quality of life without the expenditures used in the developed countries and relatively small expenditures used for the organisation of the care delivery, education of medical personnel and patients and modification of blood bank practises yields large benefits to patients. Addition of modest amounts of concentrates greatly amplifies this trend. As more resources become available, additional services such as

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orthopaedic surgery, prophylaxis and immune tolerance can be added, but these will increase the cost and benefit for fewer patients.

Rather than being discouraged by such economic realities, the haemophilia community must use this information to clearly argue for the benefits of comprehensive care, based on scientific data, advocating for a health-care plan in line with their country's economic capacity. Stepwise improvements in the delivery system should subsequently be requested, as the economy improves and resources become available. In the developed countries, where cost cutting is a constant threat, the haemophilia community (patients and physicians) must understand the changes that these benefits have produced in health-care utilisation, speak with one voice of the benefits of maintaining high-quality care and advocate for changes in the haemophilia treatment centre to ensure its continued existence.

The Argument For Comprehensive Care

In developed countries comprehensive care leads to substantial socioeconomic benefits, increased employment, decreased health-care resource utilisation and lower cost of care. Patients and decision makers might ask whether comprehensive care is necessary when they see happy and well people with haemophilia, but outside the comprehensive care environment costs increase significantly.

In countries with emerging economies with limited resources and restricted access to clotting factor concentrates, even modest expenditures to modify the structure and organisation of the care delivery (with emphasis on prevention), patient and doctor education and modified blood bank practises to improve the safety and supply of therapeutic products yield huge economic and quality of life benefits.

Impact Of The Success On Clinic Utilisation In The Developed Countries

The success of comprehensive care has reduced the need for frequent or

day-to-day management of patients and a shortage of trained health professionals with experience and this may lead to funding challenges.

Impact On Trained Physician Availability

Another problem resulting from the decreased dependency on treatment centres is a growing shortage of doctors with training to manage bleeding disorders. Many doctors need to treat patients other than bleeding patients (eg oncology or haematological malignancies) to make a living, haemophilia and blood coagulation becomes secondary and young doctors are discouraged or unable to seek training in the area. Many young doctors will be attracted to the growing area of oncology than haemophilia. This threatens the existence of haemophilia treatment centres.

Options For Maintaining Care

It is necessary to attract young physicians by providing training opportunities and a young physician training in haemostasis must feel that he/she will practise his/her speciality more than 20–25% of the time. We need to stimulate the research environment for the research of

blood coagulation disorders. Clinics need to be expanded to include other people with bleeding disorders and those with clotting defects that produce thrombosis.

Role Of Patient Advocacy

Patient advocacy has been critical in the past to improve safety and supply of products and needs to be re-energised. To ensure the gains made in the past are not lost.

Future Role Of Patient Organisations

Patient organisations must recognise and adapt to the changing environment and define a new purpose for their organisations. They must understand political processes and health economics processes in their country. Patient organisations will have more credibility and power increases when working collaboratively with clinicians. Objectives must be set to meet needs and national plans must be based on sustainable goals. **H**

The comprehensive care centre model provided comprehensive services based on an integrated public health approach and has been one of the most successful public health programs in many developed countries, resulting in significantly improved health for patients with haemophilia and reduced use of health care services.

The bleeding disorders' community is becoming increasingly concerned about ageing. Several sessions at the recent WFH Congress addressed ageing and related topics. Clearly, in Australia we are faced with an ageing bleeding disorders' community and our approach to services and care needs to adapt to reflect need. HFA will explore how we might address the needs of our members and will work with State/Territory Foundations and special interest health professional groups to ensure we respond appropriately. Haemophilia Foundation Australia has funded the authors below to undertake research to evaluate balance dysfunction and falls in people with haemophilia (aged >30 years), and to determine the feasibility of a home exercise program targeting balance training for this group.

The following piece has been extracted from an article by A Street, K Hill, B Sussex, M Warner and MF Scully published in the State of the Art book, XXVII International Congress of the WFH, Haemophilia journal, July 2006, Vol 12, Supplement 3, pages 8-12. The full text with references is available from HFA.

Haemophilia and Ageing

The paper addresses falls in the older man with haemophilia, their causes and consequences and cardiovascular problems in particular. Very little literature has been published about these common problems. We need to be aware of the ageing issues in haemophilia and develop 'wellness' programs which are directed to the early identification of disease as well as preventative strategies to reduce the physical and psychological impacts of ageing.

Men with haemophilia have not only the challenges of living with HIV and/or HCV infection and premature arthritis as complications of their disorder, but they also confront the other ails of ageing. These include genitourinary problems such as prostatic hypertrophy, prostatic cancer and renal stone disease, and arterial disease for which haemophilia is not protective. Progressive arthritis and declining fitness may lead to loss of independence which causes great concern.

Associated with the physical aspects of ageing, many patients also suffer from psychological symptoms which may be precipitated by changes in work such as early retirement and altered family dynamics.

Many older men with haemophilia may never have consulted primary care physicians because of the rarity and complexity of their disorder. Haemophilia centre staff often assume responsibility for the identification and management of all health problems of their patients. Even when other clinicians are involved, patients require their centre's involvement in the investigation and support of many procedures such as coronary artery surgery and urological surgery.

Advances in haemophilia care; beginning in the 1950s led to a worldwide movement to concentrate haemophilia care in designated treatment centres. Initially, haemophilia treatment focused principally on the musculoskeletal manifestations of the disease, which required expertise in Physiotherapy, Rheumatology and Orthopaedic Surgery available through comprehensive treatment centres. New biological therapies to manage haemophilia promised huge success and a normal life expectancy for many patients until the enormous tragedy of contamination of the blood supply with HIV and hepatitis C in the 1980s and 1990s. During that period, haemophilia care often concentrated on the treatment of these dreaded complications. With the advent of recombinant coagulation concentrates, the issues of inhibitor formation, optimal dosage and duration of factor support and treatment of an ageing cohort of patients with haemophilia have gained importance.

The new challenge is how to provide excellent care to an ageing population, including women, with hereditary bleeding disorders. With age, people with haemophilia develop medical and surgical disease previously not seen in this group. The care of older people with haemophilia may require input from services inexperienced in the management of patients with congenital coagulation disorders, such as Cardiology, Cardiovascular Surgery, Urology and Oncology.

Management requires early planning with all members of an extended haemophilia team with particular emphasis on preparing inexperienced hospital units for the care of these unique patients.

Diseases such as osteoporosis, hypertension and diabetes are seen with increasing frequency as the patient cohort ages. Lifestyle issues and common screening programs need to be included in the management of many older patients attending haemophilia treatment centres with referrals and guidance to appropriate specialties.

Balance dysfunction and falls becomes increasingly common with age in the non-haemophilia population. Many falls also cause a loss of confidence in mobility, which often results in activity curtailment, and subsequent deconditioning and further risk of falls. Recent research has shown that identifying and managing risk factors for falls among older people can be effective in reducing falls rates. Exercise interventions, particularly those with a balance-training component, have been shown to be effective in reducing falls in this high-risk group.

Haemophilia is associated with bleeding episodes, which occur often internally into the joints or muscles. Clinical staff at the Ronald Sawers Haemophilia Centre at The Alfred Hospital, Melbourne, recently identified that falls appear to be a common problem for their clients with haemophilia and other bleeding disorders. Falls in this client group were considered particularly important because of the high risk of serious bleeding associated with injuries, potential impact on the musculoskeletal system and function, and costs associated with treatment for these fall-related injuries and consequences. However, there has been virtually no research to date evaluating falls and balance dysfunction in people with haemophilia and other bleeding

disorders other than investigations around bone strength in children with haemophilia, and potential benefits and harms associated with exercise (primarily strength and fitness exercise, not on balance training).

In the Melbourne study results from a comprehensive assessment battery have been used as a basis for a home exercise program prescribed by a physiotherapist, based on the successful Otago program (targeting functional balance and strength training to address problems identified on the assessment). This has been shown to be successful in improving balance, strength, and significantly reducing falls among older people. Results may indicate the need for a greater focus on preventative balance-training exercise programs for persons with haemophilia.

Hereditary bleeding disorders are rare lifelong chronic illnesses, which require episodic treatment, making it very difficult to develop an evidence-based clinical trials approach to therapy. Developing an evidence-based approach to the management of patients with hereditary bleeding disorders as they age will be a challenge.

The publication goes on to consider haemophilia and atherosclerosis, increasing age and the risk factors for cardiovascular disease in people with haemophilia. Management of cardiovascular interventions in those with haemophilia remains challenging and requires a team approach. A number of successful approaches have been described for angiograms, angioplasty, bypass surgery and valvular surgery. Haemophilia is associated with an increased risk of hypertension. This is, in part, related to the increased risk of renal disease.

As haemostasis is central to the evolution and treatment of cardiovascular disease, it is to be expected that disorders of haemostasis, such as haemophilia, will have a major impact on morbidity and mortality among other risks.

Calendar

Hepatitis C Awareness Week

Australia 1-7 October 2006

ph 03 9385 9107

email heather@hepcvic.org.au

Haemophilia Awareness Week

Australia 8-14 October 2006

ph 03 9885 7800

fax 03 9885 1800

email hfaust@haemophilia.org.au

Australasian Sexual Health Conference

Melbourne 9-11 October 2006

ph 02 8204 0770

fax 02 9212 4670

conferenceinfo@ashm.org.au

www.sexualhealth2006.com.au

18th Annual ASHM Conference

Melbourne 11-14 October 2006

ph 02 8204 0770

fax 02 9212 4670

conferenceinfo@ashm.org.au

www.ashm.org.au/conference

Youth Leadership and Mentoring Program

Launceston, 12-15 October 2006

ph 03 9885 7800

fax 03 9885 1800

hfaust@haemophilia.org.au

14th National Symposium on Hepatitis B and C

Melbourne 18 November 2006

eleanor.belot@svhm.org.au

14th Australian and New Zealand Haemophilia Conference

Canberra 4-7 October 2007

ph 03 9885 7800

fax 03 9885 1800

hfaust@haemophilia.org.au

World AIDS Day

1 December 2006

www.worldaidsday.org.au



Hemophilia 2008

Istanbul, Turkey, 1-5 June 2008

www.wfh.org

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Haemophilia Foundation Australia acknowledges the funding and assistance received from the Commonwealth Department of Health and Ageing which makes this publication possible.