Haemophilia Foundation Queensland

The 'H' Factor

Issue 34 Summer 2012

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Haemophilia Foundation Queensland

HFQ Manager—Liz Fistonich

PO Box 451

Spring Hill

Phone: 0419 706056

Email: info@hfq.org.au

Facebook:

https://www.facebook.com/HFQLD

We are on The Web

www.hfq.org.au

The President's Press

Hi everyone,

It is an honour for me to be writing my first president's report for the HFQ newsletter. My family came across the HFQ 9 years ago soon after Hugo, then 11 months old, was diagnosed with severe haemophilia A. Consequently, family camps, youth camps, BBQs with newly diagnosed families and other activities have been a feature of our calendar. We have experienced the benefits of these events ourselves and seen the great support they offer others. We have also observed the selfless commitment of time and energy by committee members over the years.

Erl Roberts as my predecessor leaves very large shoes to fill, and I would like to thank him for his ongoing contribution to the Foundation. Not only did his most recent stint as president provide the steadying hand the organisation needed, he brought the wisdom and compassion of an "elder statesman" to the table; a role I trust he will continue to fulfil as vice president.

Howard Mitchell and Erl Roberts at the Wheelathon.

President's Press contd.

There are many familiar faces on the new board; Peter David (Treasurer), Craig Bardsley (Board Member), David Stephenson (HFA delegate), Leanne Stephenson (HFA delegate), Robert Weatherall (Board Member), and Adam Lish (Board Member), so I look forward to continuing to work with them this year. A particularly warm welcome goes to Sarah Hartley who joins us as our Secretary, Eva Turek who joins us for the first time as a Board Member, and we also look forward to seeing Dr John Rowell as a regular attendant at our monthly meeting. His insights will be invaluable.

Liz at the HFQ office continues to drive our events programme whilst representing the Foundation at various forums. Her boundless energy and positive "can do" approach continues unabated in spite of her recent cancer diagnosis. Liz plans to work on as she goes through her chemotherapy, and I am sure I speak on behalf of the whole community when I wish her, John and Alicia all the best over the next couple of months as her treatment progresses.

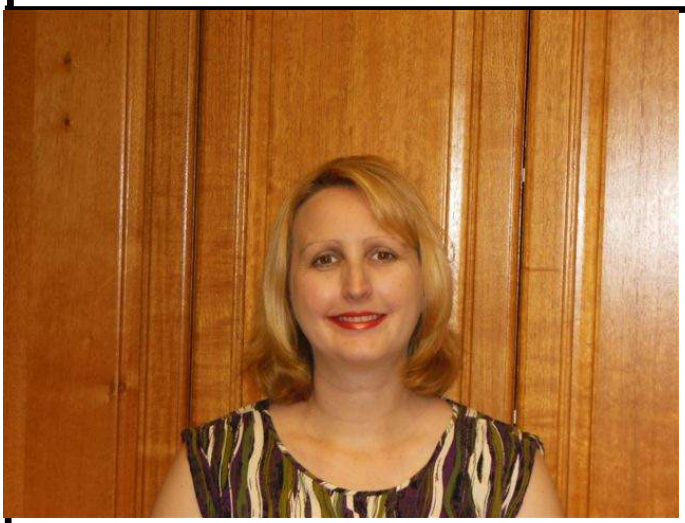
For the next 12 months, your Board will continue to pursue activities that support the needs of the community, and provide representation at various forums so that our voice is heard a loudly as possible. Further, this year we will commence a GP education programme aimed at raising the awareness of bleeding disorders with practitioners. This is an exciting new venture run in conjunction with the University of QLD and the Haemophilia treatment centre at the RBWH.

So thank-you for the honour of being the president of your organisation and wish me luck. I look forward to catching up with as many of you as possible over the next 12 months.

Regards

Howard Mitchell

Welcome to HFQ's new Secretary



Hello to the Queensland haemophilia community, I am looking forward to serving in the role of Secretary with the HFQ Board this year. I think it is important for haemophilia to have a voice and a presence in the crowded health and not-for-profit sector. As Secretary I will be assisting with the operation of the Board including correspondence, HR and event support.

My son was diagnosed with haemophilia two years ago at age five following surgery. We began prophylaxis this year which has been very challenging, but we have found the practical benefits and peace of mind to be invaluable.

I am a solicitor, usually I advise at community legal centres in the area of family law. I am currently working as an advocacy officer with a not-for-profit organisation for people with a disability. I worked for the state government for many years in a variety of marketing, planning, program and contract management roles. I hope that my skills and personal stake in the welfare of the haemophilia community will benefit the work of the Foundation.

Hope to catch up with as many as possible of you at our Christmas party (see the back page for details)

Sarah

ABOUT HFQ

The Haemophilia Foundation of Queensland (HFQ) provides advocacy, education and support for people in Queensland affected by inherited bleeding disorders. The Foundation employs a fulltime Co-ordinator and is guided by a voluntary Management Committee which meets monthly.

Members of HFQ are entitled to benefits, including subsidies on:

Medic Alert bracelets (50% discount)

Electric Shavers (up to $75 off)

Cool Relief patches (special trial offers)

Supportive footwear (75% off)

DATES TO REMEMBER

17th November	HFQ Christmas Party at Ipswich Rail Museum
24th November	HFQ and UQ General Practitioner Training Day
25th December	Santa Visits
21st December to 7th January HFQ closed	Christmas Holidays

HFQ Management Committee

President	Mr Howard Mitchell
Vice President	Mr Erl Roberts
Secretary	Mrs Sarah Hartley
Treasurer	Mr Peter David
Members	Mr Craig Bardsley
	Mr Robert Weatherall
	Dr John Rowell
	Mr Adam Lish
	Mrs Eva Turek
HFQDelegates to HFA	Mrs Leanne Stephenson
	Mr David Stephenson

Queensland Haemophilia Centre Contact Details

ROYAL BRISBANE AND WOMEN 'S HOSPITAL

Dr John Rowell—Haematologist	3646-8067
Beryl Zeissink—Nurse	3646-5727
After Hours—Page Haematologist	3646-8111
Emma Patterson—Physiotherapist	3646-8135
Maureen Spilsbury—Snr social worker	3646-8769

ROYAL CHILDRENS HOSPITAL

Dr Simon Brown-Haematologist	3636-9030
Joanna McCosker—Nurse	3636-9030
After Hours—Banksia Ward	3636-7472
Prue Ramsey - Physiotherapist	3636-8506
Mona Chong – Psychologist	3646-7937

HAEMOPHILIA CLINICS

RBWH

Appointments 3646-7752 or 3646-7751 or speak to Beryl

Haemophilia and Genetic Clinic—Dr John Rowell—Wednesdays 1.30pm

Haemophilia/Orthopaedic Clinic—Dr John Rowell and Dr Brett Halliday—9am every four weeks

RCH

Phone Joanna about appointments

Banksia Outpatients—Dr Simon Brown—Thursdays at 2.30pm

OUTREACH CLINICS

GOLD COAST HOSPITAL, TOOWOOMBA GENERAL HOSPITAL, NAMBOUR HOSPITAL, CAIRNS BASE AND TOWNSVILLE HOSPITALS:

Book through Joanna at RCH and Beryl at RBWH

HFA Report

Hi Everyone

The World Haemophilia Congress was held in Paris this July. I am pleased to day that the HFA booth was extremely busy and that people were genuinely excited about eh next one being held here in Melbourne in 2014. Start saving your penny's as this will be an event you will not forget.

This is an opportunity for Australia to show the world that we can host such an event which they will never forget. Hope to see all of you there

In Paris I met many people from all over the world and from all walks of life. It opened my eyes and made me realize how lucky our children are to receive their factor when needed.

There are many in the world who either cannot get to a hospital because it is too far away or do not have the money to pay for it.

We look forward to being able to keep you up-dated on the progress of the Melbourne Congress.

Regards

Leanne Stephenson

HFA Delegate

Physiotherapy research for Haemophilic Arthritis in the knee and

Volunteers are being sought for a new research project which aims to provide practical information for management of joints for individuals with haemophilia.

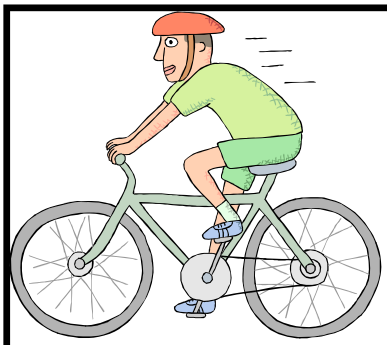
The Queensland Haemophilia Centre Physiotherapist Emma Paterson is coordinating a short trial looking at the benefits of gentle hands on therapy (referred to as manual therapy) for haemophilic arthritis in the knee and ankle.

Volunteers with moderate or severe haemophilia are being sought to attend one appointment with a follow up phone call one week later. The appointment should take approximately 30 minutes, and the follow up phone call 10 minutes.

Please note that this is a research project that aims to obtain information that may or may not benefit individuals with haemophilic arthritis. If you are interested in more information about this project please contact Emma Paterson at the Queensland Haemophilia Centre on (07) 3636 8135.

What HFQ did for...

The Wheelathon



Great fundraiser and snakes for free!

Twenty four keen riders embarked on the inaugural Ride for a Cure wheelathon on Sunday 7th of October.

It was a great turn-out with lots of support by many families at Eagle Junction State School as well as a few of Hugo's keen cousins, aunts, uncles and grandparents.

Kids and a few dads cycled the 1.2km track at the Nundah Criterium Circuit seeing how many laps they could complete in 30 minutes. The maximum achieved was 9 which was a mammoth effort on such a hot morning.

Extra excitement was created by a red belly black snake that decided to scoot across the track right in front of Hugo and his cousin Portia. Not something you would expect to see on a bike track on a Sunday morning!

The amount raised hasn't been tallied as yet however we expect it to be well into the thousands.

The response was overwhelmingly positive so we plan to run the event again next year. We hope to see you then!

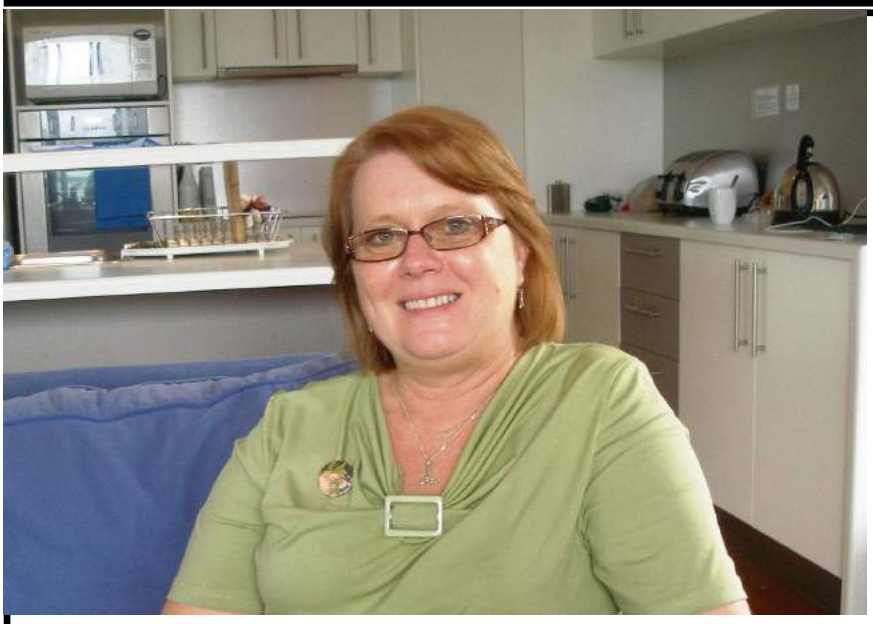
Howard, Danielle, Hugo and Poppy Mitchell

Haemophilia Awareness Week

Ready Set Go

Erl and June Roberts—the support crew. (Yes Erl is having a treatment!)

Social Work News RBWH



This month we are delighted to welcome Sarah Anticich to our team. Sarah comes to us with a wide range of skills and experience. Sarah will be working mainly at the RCH but will also provide cover for staff leave for social work/ psychology issues at the RBWH. Sarah has been appointed in a full-time permanent capacity in the role previously filled by Kelly Brady who resigned after taking Maternity Leave.

We are pleased that Mona Chong also stays as part of our team. Mona now works with the RBWH team on Fridays. We hope to be able to be creative in using the range of skills and experience of our new team.

Here is an outline of our working days:-

Maureen Spilsbury (**Monday to Thursday** – RBWH – ph 3646 8769) - Advanced Social Worker

Mona Chong (**Fridays only** – RBWH – ph 3646 8769) - Advanced Psychologist

Sarah Anticich (**Monday to Fridays** – RCH – ph 3646 7937) – Senior Psychologist

You may get to meet all of us at some point as we provide cover for each other during leave.

2. Unfortunately the Federal Government has announced the **closure of the Medicare Chronic Disease Dental Scheme from 1 December 2012**. We have been advertising this scheme for a few of years and a number of people in the haemophilia community have been able to utilize the process to have dental work completed. Debate continues in parliament regarding plans for community dental care in the future.

3. At this stage **Medicare Rebates** are still available for people who have chronic and /or complex medical conditions. Rebates are available for treatment through a range of allied health professionals. Management of the scheme is through your local GP. It can include visits to Audiologists, Chiropodists, Dieticians, Occupational Therapists, Podiatrists etc..

4. The Commonwealth and State Governments provide funding for a wide range of support services to encourage older Australians to remain independent in their own home and active within their community. **HACC (Home and Community Care) providers and ACAT (Aged Care Assessment Teams)** can provide assessment, information and referral regarding support services within your local area.

5. Please note that all RBWH phone numbers have now changed to (07) **3646**---- You will need to change our contact details wherever you have them saved.

For further information on any of these matters call Maureen Spilsbury/Mona Chong on (07) 3646 8769

Social Work News RCH

I would like to introduce myself, my name is Sarah Anticich, I am very excited to join the Queensland Haemophilia Centre team in a full timer permanent capacity. I am a registered Clinical Psychologist who specialises in the treatment of children and adolescents with extensive experience providing psychological services to your children, adolescents and families within both public and private practice. I am a full member of the Australian Psychological Society, Australian and New Zealand College of Clinical Psychologists.

I moved to Brisbane with my young family from Christchurch, New Zealand in June 2011 and have been working with the department of neonatology at the RBWH until my recent appointment to Haemophilia. I am currently in the final stages of my PhD through the University of Queensland which is focused on anxiety management and enhancement of resilience in children.

My role as a psychologist in this team is to provide psychosocial support, counselling, information and education to children with haemophilia and other inherited bleeding disorders and their families to maximise quality of life. These may include helping children to manage their fear of needles and anxiety about their condition, working with parents to assist with the challenges of parenting, stress and personal issues. I look forward to meeting you over the course of the year.

In the meantime, please do not hesitate to contact me on

07 3646 7937 or

sarah_anticidh@health.qld.gov.au if you require any assistance.

General Practitioner's Training Day for Haemophilia

The HFQ in conjunction with the Qld Haemophilia Centre and the HIV & HCV Education Project at the University of Queensland, School of Medicine are planning a one day training forum for ***General Practitioners*** *in November this year. Haemophilia Foundation Queensland is sponsoring the Forum which will be held in Brisbane. Please make contact with the Maureen Spilsbury or Mona Chong (07) 3646 8769 if you think your GP might be interested in attending. The forum will cover a range of health issues relevant to people in the haemophilia community.*

Sleep Wellness in Older Adults

By Mona Chong

Changes in sleep patterns are normal as we get older. However, sleeping well is still essential for our physical health and emotional well-being. Older adults who do not sleep well are more likely to suffer from depression, attention and memory problems and excessive daytime sleepiness.

How much sleep do older adults need?

This varies from person to person but most healthy adults require between 7.5hr to 9 hours of sleep to function at their best. However, it is important to focus more on how you feel following a night's sleep than the specific number of hours of sleep you need. Frequently waking up not feeling rested or feeling tired during the day are better indications that you're not getting enough sleep at night and may have a sleep problem that needs to be addressed.

How can we improve sleep?

Here are some simple tips and strategies for you.

Improve daytime habits

- **Be engaged.** Social activities, family, and work can keep your activity level up and prepare your body for a good night's sleep. Try volunteering, joining a seniors' group, or taking an adult education class.
- **Improve your mood.** A more positive mood and outlook can reduce sleep problems. Find someone you can talk to, preferably face-to-face, about your problems and worries.
- **Exercise regularly.** Exercise releases endorphins that can boost mood and reduce stress, depression, and anxiety.
- **Expose yourself to sunlight.** Bright sunlight helps regulate melatonin and your sleep-wake cycles. Try to get at least two hours of sunlight a day. Keep curtains and shades open during the day or move your favourite chair to a sunny spot.
- **Limit caffeine, alcohol, and nicotine.** All are stimulants and interfere with the quality of your sleep.

Keep a regular bedtime routine

- **Make sure your bedroom is quiet, dark, and cool**, and your bed is comfortable. Noise, light, and heat can cause sleep problems. Try using an eye mask to help block out light.
- **Maintain a consistent sleep schedule.** Go to bed and wake up at the same times every day, even on weekends.
- **Block out snoring.** If snoring is keeping you up, try ear plugs, a white-noise machine, or separate bedrooms.

Sleep Wellness in Older Adults contd.

By Mona Chong

- **Go to bed earlier.** Adjust your bedtime to match when you feel like going to bed, even if that's earlier than it used to be.

Develop bedtime rituals. A soothing ritual, like taking a bath or playing music will help you wind down. Relaxation and stress management techniques, such as deep breathing and progressive muscle relaxation, take some practice but their benefits can be substantial.

Bedtime diet tips

- **Limit caffeine late in the day.**
- **Avoid alcohol before bedtime**. Don't use alcohol as a sleeping aid. It might seem to make you sleepy, but will disrupt your sleep.
- **Satisfy your hunger prior to bed**. Have a light snack such as crackers, cereal and milk, or yogurt or warm milk.
- **Minimize liquid intake before sleep**. Limit what you drink within the hour and a half before bedtime.

Getting back to sleep at night

It is normal to wake briefly during the night but if you are having trouble falling back asleep, the following tips may help:

- **Make relaxation your goal, not sleep.** Try a relaxation technique such as deep breathing or meditation, which can be done without getting out of bed.
- **Do a quiet, non-stimulating activity.** If you've been awake for more than 15 minutes, try getting out of bed and doing a non-stimulating activity, such as reading a book. Keep the lights dim so as not to cue your body clock that it is time to wake up, and avoid TV and computer screens.

Postpone worrying. If you wake during the night feeling anxious about something, make a brief note of it on paper and postpone worrying about it until the next day when you are fresh and it will be easier to resolve.

Include exercise into your daily life

Studies have shown that participating in moderate endurance (aerobic) activity can have the greatest impact on improving sleep. **Always consult your doctor** before embarking on any new fitness programme. Some ideas include

- **Swim/Water Exercises** – Swimming laps is a gentle way to build up fitness and is great for sore joints or weak muscles. Many community pools have swim programs just for older adults, as well as water-based exercise classes such as water aerobics.
- **Take up lawn bowling** – Variations on throwing a ball on an earthen or grassy court are gentle ways to exercise.
- **Walk—**The more you walk, and the brisker the pace, the more aerobic benefit you'll experience.

Sleep Wellness in Older Adults contd.

By Mona Chong

Reduce mental stress

Stress and anxiety can easily get in the way of a good night's sleep. Everyone has worries and lists of things to do, but it is important to teach yourself to let go of these thoughts when it's time to sleep.

- Keep a journal to record worries and concerns before you retire.
- On your to-do list, check off tasks accomplished for the day, list your goals for tomorrow, and then let go!
- Listen to calming music.
- Read a book that makes you feel relaxed.

Seek opportunities to talk with a friend or therapist about what is troubling you.

If you still having trouble sleeping, make sure you go to your local GP for more help.

For more information about sleep wellness, you can visit the

- Sleep Disorders Australia (http://www.sleepoz.org.au/)
- Sleep Health Foundation (http://www.sleephealthfoundation.org.au/)

Australasian Sleep Association (http://www.sleep.org.au/)

Information for this article was adapted from http://www.helpguide.org/life/sleep_aging.htm

"If you have trouble falling asleep, lick your feet for a few minutes. It works for my cat!"

HFQ 2012 Youth Camp

By Adam Lish

Recently Haemophilia Foundation Queensland held their annual youth camp, at the Emu Gully Adventure Park just outside of Toowoomba. HFQ's youth camp aims to get boys of ranging ages, from about 8 years old to 25, together for a great weekend of fun and education. The aim of this camp is to show boys that Haemophilia is in no way a barrier to life and they can do anything (including all the physically challenging activities we did at camp), while educating them on managing their Haemophilia (specifically self cannulation and being away from home and parents).

HFQ received its best attendance rate since the revival of their youth camp last year. In 2011 HFQ held their first youth camp after about 10 years. 2011 was a great success, yet 2012 surpassed 2011 as there were more boys there, more challenges overcome and tons of friends made.

I'd like to add in my own personal reflection on the weekend. I'm 18 years old now and have been a part of the HFQ Board for the past two and a half years, as well as being an active member of HFQ for the past 18 years. After 2011's and 2012's camp, I saw great results and benefits and potential for the future. From my point of view I saw boys achieve and overcome challenges in their life, a lot of which was based on confidence issues. I witnessed people take part in activities which they were either scared to do (one activity was to crawl through pitch black tunnels) or

Boys also overcame their fear of self cannulating, as some of the younger boys and even a couple of older ones needed to make that jump to being (or knowing how to be) independent. I also saw from my point of view the older boys mentoring the younger ones, almost like an older brother, which is a brilliant thing as it shows the younger ones that they can deal with Haemophilia and not let it stop them (as some of the older boys were a good example of how to do anything- but safely)

All in all, HFQ was very pleased with the results and benefits of their youth camp. I can say on behalf of the board and other participants, that it was an excellent time. Everyone was very happy with what each individual drew from the weekend. To say it was a great weekend, would be an understatement, it was just priceless.

By Joanna McCosker

What can I say; it was one fun filled, non stop and very muddy adventure!

From walking across unstable planks with the cold water below waiting for us to fall in (none of us did) to crawling in dark and cramped tunnels, sliding down a massive slide in the pitch dark, then ripping it up on the quad bikes and finally wading chest deep in mud: WE achieved a lot…..

It took a lot of team work, mateship and courage to "survive" the challenges that we were faced with! I was most impressed that not one person in the group thought that Haemophilia was an obstacle to achieving their goals for the weekend. Everyone who went achieved one thing that they thought they couldn't do or was fearful of….including me! Not a fan of the wobbly boards over water!

The older boys were great role models for the younger boys as they demonstrated their skills of self infusion. Many friendships were made and I hope that the older boys will continue to "buddy' the younger ones and lead the way in treating their haemophilia well. Remember that you can do anything in life and haemophilia is not an excuse. The only thing that can stop you achieving your goals is you (or your bad attitude)

Thank you to all the boys who participated this year, I loved sharing the experience with you all and I am proud of what you achieved.

One final thing to say "Go Eeemmmuuu" (Emu)

xx Joanna

A Step Back in Time.....by David Stephenson

22 Year Old Haemophiliac Shot

A 22 year old shop worker was shot in the loin by mistake by a customer who was trying out a new gun. The man from Norwich required surgery and was initially treated with bovine material to stop the bleeding. This was initially successful in controlling bleeding but he soon developed a severe allergic reaction and bleeding was no longer controlled due to inhibitory antibodies against the bovine material. Desperate measures were then employed and pig plasma was used which saved the young mans life.

This happened back in 1954 when treatment for bleeding was very limited, this young man was lucky to survive. Interestingly the porcine material was better tolerated than bovine material, although allergic and resistance caused by formation of antibodies was still a problem after repeated transfusion. This remained one treatment in the late 50's and early 60's until the development of cryoprecipitate in 1965.

What a contrast to the range of products we currently have, not to mention those that are on the development conveyor belt. Blood products from pigs have been in use for half a century (1954 – 2004) and a recombinant porcine factor VIII is now at an advanced stage of clinical development.

In a bit of a twist to this story it appears pigs may be able to provide us with human coagulation proteins with the development of transgenic animals. This future possibility could see the milk from pigs providing any amount of clotting factor the world required by simply scaling up milk production and at relatively low cost – each sow can produce 200 to 300 litres of milk per year. There is currently no evidence of transmission of any pathogens to humans and in particular pigs are not subject to 'mad cow disease'.

Dr John Rowell adds -:

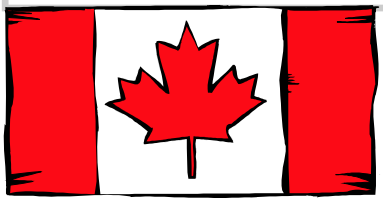
In the 1950s bovine plasma was used to treat Haemophilia as it was thought this would be a ready source of replacement product - unfortunately allergic reactions to the crude product and inhibitory antibodies made treatment ineffective. Porcine Factor VIII was developed and the latter product was freer of allergic reactions and it was used as there was no cross reactivity (interaction) between the porcine FVIII and FVIII antibodies particularly in those with antibodies to FVIII. Unfortunately the isolation of porcine parvovirus - although not known to affect humans - stopped production of this useful product. Recently porcine FVIII is being produced with recombinant technology (artificial and free of viruses) and being used in clinical trials in those with FVIII deficiency in particular those with inhibitors.

Information from the Paris WFH congress

As many of you will be aware, David and Leanne Stephenson represented HFQ at the recent WFH congress in Paris. David has bought back many articles of interest for us to reprint into our regular newsletter, The 'H' Factor. The following four pages are excerpts from our Canadian colleagues, The Canadian Haemophilia Society.

THE NEXT DECADE: Great challenges in hemophilia

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



CHALLENGE: Aging with hemophilia

Lucie Lacasse, RN, *Regional Comprehensive Care Centre for Hemophilia and Hemostasis, Ottawa Hospital*

This is truly a success story!

In the general population, individuals are now living longer and having healthier lives. In almost every country, the proportion of people aged over 60 years is growing faster than any other age group, as a result of both longer life expectancy and declining fertility rates. In 2006 there were 500 million individuals worldwide over 65 years of age. By 2030, it is projected that this number will double.

The good news is that this is no different for the bleeding disorder population, and we are now observing the aging of people with hemophilia. Advances in hemophilia care beginning in the 1950's have contributed to an overall improved life expectancy and quality of life, while previous generations of patients with either hemophilia A or B died prematurely from bleeding complications because factor concentrates were not readily available, or from complications relating to HIV and/or HCV infection. While patients living with severe hemophilia still have a reduced life expectancy, the overall life expectancy is nearing that of the general male population (Dolan et al., 2009). As a result, people with hemophilia are expected to experience agerelated clinical problems that were not previously observed in this population, as well as suffer from the long-term effect of existing complications such as liver problems secondary to hepatitis, or further mobility problems related to hemophilic arthropathy.

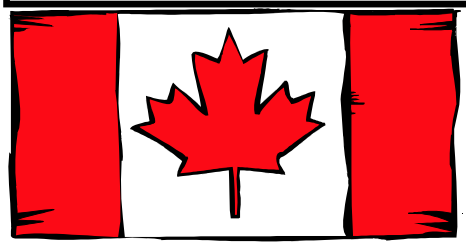
The improved life expectancy brings new challenges to bleeding disorder healthcare providers. It will be important to recognize the new reality of improved life expectancy early on during the management of hemophilia. Perhaps prophylaxis treatment will take on a new meaning if a person realizes that he will need healthy joints to carry him into his eighties. There may be no guidelines for the provision of care for health issues not previously recognized in this population. We will need to collaborate in order to build data to ensure that the care offered to our patients is evidencebased. It is important that we continue to be strong advocates for our patients to ensure that they receive high quality care in the health and community sectors. We will need to establish new partnerships and educate specialists in fields such as gerontology, oncology, cardiology and home care nursing who may be unfamiliar with bleeding disorders.

As one patient states: *"Because I am a seventy-two (72) year old woman with a rare factor V deficiency, I carefully assess, with my healthcare team, the risks and benefits of treatment options for recently diagnosed cardiovascular disease. My medical management has been adjusted due to increased bleeding side effects of some prescribed drugs."*

We need to meet this exciting challenge together and promote healthy aging of the bleeding disorder population. Aging is a process that starts at birth!

CHALLENGE: Charting CHARMS' Course: Recording your treatment for yourself and for the rest of us

John Plater, *Heathcote, Ontario*



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

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

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

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

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

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

CHALLENGE: Longer half-life products and gene therapy

David Page, CHS national executive director

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

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

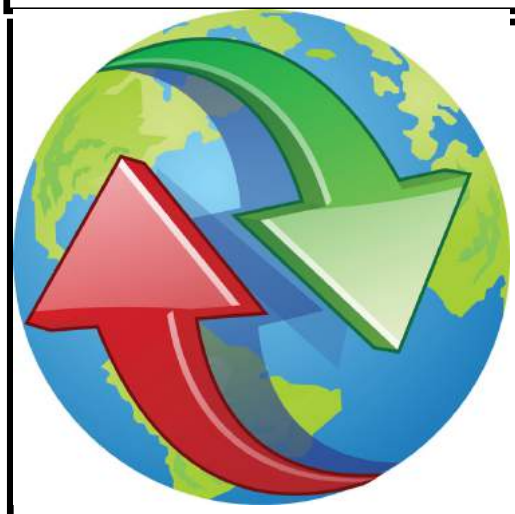
At the start of this decade, there is a great deal of research activity related to the development of longer half-life clotting factor proteins. Pharmaceutical companies in North America and Europe have either begun or are close to beginning clinical trials in longer-acting factor VIII in hemophilia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

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

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

CHALLENGE: The global challenges

David Page, CHS national executive director



According to the World Federation of Hemophilia (WFH), 70 percent of the estimated 400,000 people living with hemophilia around the world are not yet diagnosed. And of those that are, 75 percent are not receiving what we in Canada would consider adequate treatment. Many children simply die young or grow up severely disabled. The challenge is one of education, outreach and diagnosis. Then, once this groundwork is laid, the goal becomes access to treatment, including affordability of therapy.

"While we have wonderful therapies," said Mark Skinner, president of the WFH, "we don't have a cure, and the therapies we have still require infusions two or three times a week and can be costly. So we're looking for enhanced therapies that would reduce the burden on daily living and make treatment more affordable and accessible for more patients in more parts of the world. Ongoing investment in evidence-based medicine is also crucial. We need to build the body of science that will identify optimal dosing and prophylaxis protocols, improve treatment regimens and support comprehensive care."

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

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

Hot off the press

This was released as I was finishing the final touches to our newsletter.

www.sciencedaily.com/releases/2012/10/121009160706.htm

Many of you will be interested in this as many of our Queensland community participated in this study.

Latest News

Outreach clinic dates for the remainder of 2012

Nambour October Friday 26th 2012

Contact the nursing staff at RCH and RBWH if you have questions about attending these clinics

HFQ Christmas Party. Saturday 17th November 2012.

Ipswich Rail Museum—be there to meet old friends and of course Santa will be there with gifts for all the kids.

HFQ Christmas Holidays

HFQ will be shut over the Christmas Break from December 21st 2012 to the 7th of January 2013. If you require any assistance over this period please ring the staff at either RBWH or RCH.

Excuse me, God. Hector wants to show you an idea he has for a new animal.

HFQ 2012 Christmas Party

Date: 17th November 2012

Time: 10.30am at the front gate

Address: North St, North Ipswich

Come along and enjoy a fantastic day with your family and friends from the Haemophilia Foundation Queensland.

If you would like to attend and visit the railway and share a lunch with Santa and friends RSVP by 6th November 2012.

RSVP to: Liz info@hfq.org.au or call 0419 706056