the H FACTOR

Haemophilia Foundation Queensland

SUMMER EDITION

Issue 69

DECEMBER 2021

HFQ MEMBER MAGAZINE

FROM THE **PRESIDENT**

Hello everyone,

Christmas is fast approaching and we are looking forward to the break and celebrations with family and friends. It reminds me of how fortunate we are in Australia with brilliant treatments for our bleeding conditions.

When you look back in time to the 1800's things were very, very different - were life expectancy for those with bleeding conditions was around 13 years old. Where early very strange treatments were tried like - oxygen therapy, bone marrow, thyroid gland, hydrogen peroxide and even gelatin - simply amazing.

So who knows where the term haemophilia originated ….. it came from a student of Zurich University, Freiedrich Hopff & his professor, Dr Schoniein who came up with the term haemorrhaphilia which became haemophilia in 1828.

So these days we look forward in time to the real possibilities of even better curative treatments.

Regards

Dave

David Stephenson
President HFQ
president@hfq.org.au

Many people with bleeding disorders still experience down time because of bleeds and some have joint problems and/or poor mobility.

Access to wheelchairs or walking frames; and good IT and data for video conferencing and telehealth can help, but they can cost money that some of our members can't afford from within their own resources.

HFQ has a Haemophilia Equipment Loan Program (HELP) for this situation. The program is open to everyone, but all users of HELP need to be endorsed by the psycho-social work staff at the QHC's as needing assistance from the program.

Please talk to your clinic psycho-social work staff member or call the office on 0419 706 056 and have a chat with us if this is something that might help you or your family.

Inside this Issue:

ABOUT HFQ

Haemophilia Foundation Queensland Inc. (HFQ) provides representation, health promotion, education and support for people in Queensland affected by inherited bleeding disorders. The Foundation receives a grant from Qld Health and employs a part time manager and an administration assistant. It is guided by a Board of Directors which meets monthly.

We can be contacted on mobile 0419 706 056; or via email (info@hfq.org.au) or post at PO Box 122 Fortitude Valley, Qld 4006

HFQ provides financial members with support and benefits, including subsidies on:

- **Medic Alert bracelets (50% discount)**
- **Electric Shavers (up to $75 off)**
- **Supportive footwear (75% off)**

HFQ Management Committee

President	... ...	Mr David Stephenson
Vice President	...	Mr Robert Weatherall
Secretary	... ...	Mr Tony Ciottariello
Treasurer	... ...	Mr Adam Lish
Members	... ...	Mrs Belinda Waddell
		Mr Charles Eddy
		Dr Jodie Caris
		Mrs Leanne Stephenson
		Ms Shannon Gracey
		Mr Shannon Wandmaker

HFQ Delegate to HFA

Mr Adam Lish

Acknowledgements

HFQ is grateful for the support of our patron: Her Excellency the Honourable Dr Jeannette Young PSM Governor of Queensland,

HFQ programs and services are funded by the Queensland Government.

HFQ is also grateful for the support it has received

Internet

Find us on the web at *www.hfq.org.au* or at our Facebook page at *www.facebook.com/HFQLD*

QLD HAEMOPHILIA STATE CENTRES

CHILDREN'S CLINIC

PAEDIATRIC CLINIC STAFF (QCH)

Switch: 07-3068 1111 Haemophilia Mobile 0438 792 063

Dr Simon Brown – Haematologist
Haemophilia Fellow — Dr Jordan Staunton
Haemophilia Registrar – Dr Fiona Molloy
Joanna McCosker – Nurse Practitioner
Amy Finlayson / Salena Griffln – Clinical Nurse
Nathalie Holland (Mon, Tues) - Physiotherapist
Hayley Coulson (Wed, Thur, Fri) – Physiotherapist
Vacant - Senior Psychologist

Contacting the Clinic - Please call the Haemophilia mobile for urgent enquiries on 0438 792 063 (office hours 8 – 4pm).

For all non-clinical/non-urgent enquires please email QCH-Haemophilia@health.qld.gov.au

After hours — call switch and ask to speak with on-call haematology consultant or present to the emergency department

Appointments — Outpatient Bookings Office on 1300 762 831 or email QCH-Outpatients@health.qld.gov.au

Your health care team does not make these bookings or any changes to your appointments. Referrals can be sent to the Referral Centre Fax Number 1300 407 281

Haemophilia Outpatient Clinic — Dr Simon Brown — held in 3c outpatients Level 3, Thursday afternoons 1.00 – 3.30pm

Haemophilia Carrier Clinic – as needed Thursdays 1pm – 3.30pm

ADULT CLINIC

ADULT CLINIC STAFF (RBWH)

Switch: 07-3646 8111

Dr Jane Mason - Haematologist	3646-8111
Dr Sally Campbell - Haematologist	3646-8111
(Page Dr's through switch)	
Haemophilia Registrar	3646-8111
(ask to page Haemophilia Registrar on 42177)	
Beryl Zeissink - Clinical Nurse Consultant	3646-5727
Alex Connolly - Clinical Nurse (Part time)	3646-5727
After Hours - Page Haematologist	3646-8111
Scott Russell - Physiotherapist	3646-8135
Vacant - Senior Social Worker	

Contacting the Clinic Please telephone in the first instance. Appointments 3646-7752 or 3646-7751

For all non-clinical/non-urgent enquires please email RBWH-Haemophilia@health.qld.gov.au

Haemophilia and Genetic Clinic — Dr Jane Mason — Wednesdays 1.30pm New Patients Thursdays 8 - 9.30am

Haemophilia/Orthopaedic Clinic — Dr Jane Mason and Dr Brett Halliday — 9am every four weeks

OUTREACH CLINICS

Gold Coast Hospital, Toowoomba General Hospital, Nambour Hospital, Cairns Base Hospital & Townsville Hospitals: For queries email *CHQ-Haemaphilia@health.qld.gov.au* at QCH or *RBWH-Haemophilia@health.qld.gov.au* at RBWH.

What's On?

MAKE IT YOUR EVENT

Dec 2021 to Mar 2022

Some of the HFQ programs and activities already planned

Please call the office for other events, more information or to RSVP

DEC	**World AIDS Day** 1st December	**HFQ - Our Local Matters Jar at Grill'd Eagle St Pier** 1st December to 31st December	**International Day of Persons with Disabilities** 3rd December	**OBE's End Of Year Lunch** Arana Hills Leagues Club 12th December
JAN	**OBE's Monthly meeting** 12th January	**HFQ Board Meeting** 18th January	**Australia Day Event** Movie and Meal @ the Blue Room Paddington 22 January	**Australia Day** 26 January Public Holiday
FEB	**OBE's Monthly meeting** 2 February	**Women's Lunch** 6 February Everton Park Hotel	**HFQ Board Meeting** 298 Gilchrist Ave, Herston 15 February	**Rare Diseases Day** 28 February
MAR	**OBE's Monthly meeting** 2 March	**HFQ Board Meeting** 15 March 298 Gilchrist Ave,	**National Close the Gap Day** 17 March.	**Harmony Day** 21 March

World Haemohilia Day

17 April 2022 is World Haemophilia Day! The theme of the event in 2022 is Access for All: Partnership. Policy. Progress. Engaging your government, integrating inherited bleeding disorders into national policy. By raising awareness and bringing haemophilia and other inherited bleeding disorders to the attention of policymakers, we can increase sustainable and equitable access to care and treatment.

World Haemophilia Day is a day for people who have been affected by a bleeding disorder - either because they have one, or because they care for someone who does. There are many ways you can bring attention to haemophilia and other inherited bleeding disorders in your local community to raise awareness so start planning what you will do now.

Engaging your government, integrating inherited bleeding disorders into national policy.

APRIL 17 2022 | WORLD HEMOPHILIA DAY

As I see it **Pain is not the problem...** *it's the solution*

The bleeding disorder community has a pain problem, but it's not the problem of too much pain. The real problem is a misunderstanding of pain and its importance for our survival. Many people with bleeding disorders have come to devalue pain, because pain is often characterised as a mysterious and uncontrollable inevitability. A fallacious concept like "spontaneous bleeding" has sowed a seed in many minds that the actions we engage in are divorced from how our bodies respond. If the pain we experience is seen as inevitable, then we're left with no other option than to manage the best we can, or trick ourselves into thinking the pain doesn't exist.

If given the option to never feel pain again, many people would take this deal in a heartbeat. But only later would they truly understand the horrors of living without pain, because you'll be without the best warning sign nature ever created: pain. In short, your life would end much sooner. Living without pain is not a blessing - it's a tragic, brutal curse.

If you want to understand pain, know that it results from a set of factors. Pain doesn't arrive out of nowhere, and it's rarely attributable to a single cause. For example, the mild ankle pain I am experiencing as I write this is not only the result of countless past ankle bleeds, but also the result of (1) standing for two hours straight while making an elaborate meal yesterday; (2) running three days ago even though I was sore; (3) eating inflammatory foods yesterday; (4) being dehydrated today; (5) slightly twisting my ankle on a tree root while intently looking at a mailbox on a recent run; (6) not yet knowing my mileage limit while running (running is new for me since COVID-19); (7) switching to new running shoes (is it helping or hurting?); (8) walking a mile on uneven sand recently. And so on…

If you look critically at what contributes to pain, you'll see that it's possible to improve your underlying condition by changing how you navigate through this world. By examining the obvious—and subtle—contributing factors to pain, you can create a template in your mind for what you should be doing and also avoiding. In my earlier example, there are complex interactions between my physical awareness, my activity level, and my shoes that I can monitor and refine over time, as I get used to my new activity—running. This introspective, reflective process is often at odds with the objectives of using pain medications.

Sadly, many people with bleeding disorders have been offered pain medications as the only solution to their pain. Too much reliance on pain meds may be nothing more than a deception that obscures the true problem(s). While effective over the short term in damping down pain signals (the effects), pain meds also disguise the complex set of underlying factors (the causes). If we don't address the true causes of pain, and instead focus solely on the effects with pain medication, the condition or behaviours at the root of pain will get worse, not better.

So, is pain your enemy, or is it your collaborator? If you want to live a long and healthy life, pain can't be reduced to your tormentor. Don't let your pain be in vain by seeking remedy only through pain medications. You can't arrive at the underlying causes of your pain when you are literally numb to them. Our pain is not what ruins life; it is what prolongs life, but only if we listen and learn from it.

Edited for size from an article by Corey Pierce a PhD candidate in public health at Oregon State University. First published in Parent Empowerment Newsletter published by LA Kelley Communications, Inc. November 2020

Australia Day
Social Event

For our Australia Day Event this time we are off to the Blue Room at Cinebar in Rosalie on Saturday 22nd January 2022.

The movie has yet to be confirmed at the time the magazine went to press but we have asked for Addams Family 2 (and failing that Sing 2). What can be better on a hot Brisbane weekend than watching a movie while comfortably seated and enjoying a tasty meal, sweet treats, and drinks?

You don't have to stay at home to be comfortable. You can join us to experience it all at the Blue Room Cinebar in Rosalie.

This boutique cinema with semi-reclining seats and foot rests, offers us a luxurious movie experience and great service without the noise and parking hassle of a super-sized crowded cinema at a shopping centre.

And it's not just a movie, you will also get to enjoy meals and drinks with us while watching the movie and without having to leave your seat. Popcorn, of course, is also available along with coffee, tea, and sweet treats like Mary Poppins sundae, muffin, and brownie.

There is limited seating capacity of about 60 patrons so places are limited. Please reserve your place as soon as possible so you wont be disappointed.

MedicAlert
When seconds count

Even if you keep great records of your bleeding condition, in an emergency this information may not be helpful if it doesn't reach the right people.

Many of our young members typically don't wear emergency medical identification, such as MedicAlert®. This type of identification, often worn as a necklace or bracelet, alerts paramedics and emergency-room staff that the wearer has a bleeding disorder.

Safety seems to be the main issue for infants not wearing them, as parents may worry that their child could choke on the bracelet or be injured by the necklace. And older kids may not like the identification because they don't want to look different. HFQ sees them as a valuable tool and if cost and difficulty getting them to fit correctly are an issue please talk to a treatment centre team member as HFQ offers a 50% subsidy on base models and may be able to assist further if your psycho social team member thinks it important.

Most practitioners, in the face of an emergency, will accept any and all input they can get. Having something well-structured, well-summarized and well-credentialed is much more valuable and will save a lot more time than having handwritten information from the patient.

There is no point in stashing medical information in wallets, purses, glove compartments or a child's car seat, because if there's an accident, the patient or parents might be injured or unable to speak.

Emergency medical providers usually only look for medical identification on the patients themselves - not in their surroundings. Having a description of you or your child's bleeding disorder in your purse or wallet is a great idea, but it's no substitute for an easier-to-find alert on a necklace or bracelet.

Ideally, your healthcare provider should create your personal health record on the ABDR, not you. We know that emergency medical providers feel more comfortable veering from their usual methods if the information comes from a doctor with a background in bleeding disorders or a HTC, rather than the patient.

Even if your child or teen objects, this information is too important to not display prominently. If you have an infant, have him wear a wrist band (or ankle band), and he'll grow up understanding the importance of keeping this information handy.

Experts agree that having crucial information on hand can provide the key to better health. This can reduce complications, reduce mortality and certainly reduce the cost time to getting care.

Preparing for ageing

Thinking about ageing and loss of independence can be difficult. Doing things like eating a balanced diet, exercising, and regularly attending your medical visits can help you maintain independence and enjoy your later years.

Aging and potential associated care needs will look different for everyone. Rather than procrastinate, proactively preparing your finances and assets and thinking through what you want for yourself can reduce stress for you and your family.

These tips can help:
Get organised. Figure out a system to keep important documents, such as bank statements, insurance policies, wills, pension records, mortgage information and car titles, in a safe, secure place for loved ones to access in case of emergency. Have passwords written down and kept in a safe place to make sure your family has access to pertinent information when needed.

Create a advance health directive and/or an enduring power of attorney. These forms changed in Queensland at the end of 2020 so make sure yours are up-to-date.

An advance health directive is a term given to written instructions regarding your preferences for medical care if you are unable to make decisions for yourself. This document guides doctors and caregivers to make the best choices for you based on your wishes. It is important to consider what choices you might make when faced with difficult healthcare decisions at the end of your life.

Depending on the type of enduring power of attorney it can be used for personal (including health) matters only; for financial matters only; or both. An enduring power of attorney allows you to appoint someone you trust (an 'attorney') to make decisions about 'personal (including health) matters' and/or 'financial matters'.

This allows them to make medical decisions on your behalf if you are unable to do so. This person does not have to be tied to your finances. They can be appointed only to make healthcare decisions. It's important to consider who you would want to make these choices for you if the circumstances called for it.

Sometimes, even if you write down your end-of-life wishes, they don't make it into the hands of your medical provider. Therefore, make sure you appoint a specific person and fill him or her in on your choices.

Get up to speed on My Aged Care. You can sign up for an assessment with My Aged Care starting when you turn 65. Navigating this program can be complicated. My Aged Care website and others have detailed information that can help.

Keep in mind that My Aged Care covers in-home care, such as help with cooking, cleaning, laundry, and other activities of daily living as well as long-term care in a nursing home or an assisted living facility. As you age, you may need these so it's better to understand what entitlements you may be able to access before you need them.

Educate your family members. As many people are switching to injections rather than infusions, it is important for family members to understand when factor might be needed, regardless of your prophylactic regimen. Educate your family members in advance about what bleeds look like so they can help you be proactive in your haemophilia care as you age.

Complementary Alternative Medicine

Pain and You

Pain is highly personal. No two people experience the same feeling of pain, even when it's the same injury, like a muscle bleed, or experience, like childbirth. A joint bleed may feel tingling to one, stabbing to another, or throbbing to someone else. Mike Birmingham, a man with haemophilia A, writes in his blog The Bleeding Edge, "Pain is pretty deeply personal. I personally have never been able to figure out what to say when a nurse asks me to describe my pain."

But it's especially personal when trying to describe the level of pain. Doctors often ask patients to rate their pain on a scale of 1 to 10. But what is a 1? What is a 10? A level 8 to one person might be a level 3 to another. Many older guys living with a bleeding disorder have got used to that level of pain and this is our 'normal.' What's difficult is when you try to explain their level of pain to their doctor. Not appreciating or understanding how much pain a person is feeling may lead to an inefficient treatment for that pain.

Because pain is so personal, medication may not be the first - or the only - option for chronic pain. Instead, both patient and physician can consider different types of complementary and alternative medicine (CAM) to learn how to handle chronic pain. And like pain, CAM can be highly personalized as well.

What Is Complementary and Alternative Medicine (CAM)?

CAM is any additional therapy, like massage, used along with conventional medicine. It's an important part of a multidisciplinary approach to pain management. It's also important in integrative medicine that focuses on the whole person and makes use of all appropriate therapeutic approaches, healthcare professionals, and disciplines to achieve optimal health and healing. Here are some of the most common CAM therapies:

Relaxation Therapies. Relaxation teaches you to relieve tense muscles, reduce anxiety, and alter your mental state. Mindful-ness meditation helps you focus attention on a specific object or your breathing patterns to induce relaxation. Guided imagery is a conscious meditation technique of relaxation followed by visual-ization of a soothing mental image, like walking on a beach at sunset.

Biofeedback Training. You can learn how to recognize and change your biological reactions to stress and pain by using electronic equipment to monitor your physical responses: brain activity, blood pressure, muscle tension, and heart rate.

Behaviour Modification. Some people with severe chronic pain may become anxious, depressed, homebound, dependent, or bedridden. Behaviour modification helps you create a step-by-step approach to confronting challenges by changing your behaviour and shifting your attitude. Matt Barkdull, a man with haemophilia B says he finds if he deliberately engages in deep-breathing exercises and stays connected while avoiding his mind to wander and unhinging from false perceptions, the pain is much better controlled.

Stress Management Training. If your pain level is high, your stress levels probably are too. This training helps you maintain a routine schedule for activity, rest, and medication. It incorpo-rates exercise or physical therapy into your daily routine, and trains you to keep a positive outlook.

Hypnotherapy. Therapeutic or medical hypnosis directs your focus inward to help you relax and reduce pain and anxiety. You can learn self-hypnosis from a trained hypnotherapist.

Counselling. Individual, family, or group counselling with a professional trained in pain management can provide emotional support and guidance. Matt Rollins says, "Speaking with a mental health professional and learning meditation helped me the most. I can't tell you how at peace I became when my mind accepted the fact that pain is part of my life

and I can turn it into power and motivation to help others."

Acupuncture. Many patients report pain relief from this ancient Chinese technique of inserting and manipulating thin needles into specific points on the body known to control pain pathways. If you're considering acupuncture, first talk to your haematologist or the staff at the HTC first.

Dozens of other therapies, including acupressure, massage, and chiropractic manipulation, may help control pain. Transcutaneous electrical nerve stimulators (TENS) deliver electrical impulses to interfere with pain transmission. Ultrasound therapy warms joints internally to provide pain relief, and laser treatments may provide relief in a similar way.

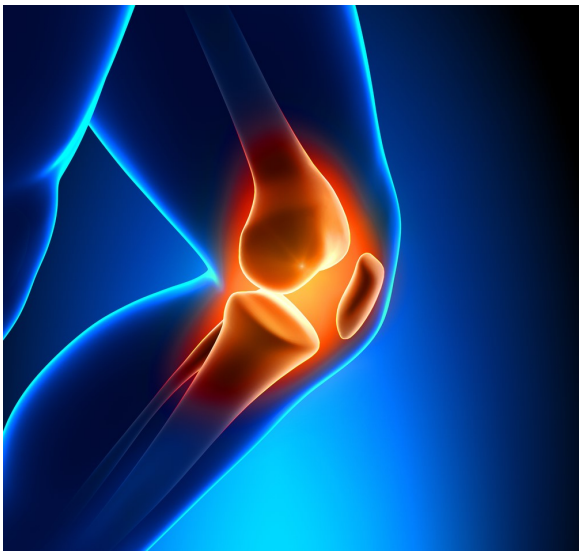
A good management plan for chronic pain must be personalised. It should use a multimodal approach, which addresses the psychological component of chronic pain by treating depression and reducing anxiety and stress. A multimodal approach includes adjuvant therapies (antidepressants and anticonvulsants); an exercise and/or physical therapy component; and some form of CAM, which allows the person to manage moderate to severe chronic pain with the lowest possible dose of painkillers.

Edited for size and Australian content from Chronic Pain, CAM and you. By Laurie Kelley. First published in Parent Empowerment Newsletter by LA Kelley Communications, Inc. November 2020

Meet your Child's Joints

Joint disease is still the most common complication of bleeding in severe haemophilia. Repeated bleeding into joints eventually leads to haemophilic arthropathy - a degenerative joint disease caused by the slow deterioration of cartilage in the joint, resulting in a painful form of arthritis and eventual destruction of the joint. To try to prevent joint disease prophylaxis, or "prophy" is used. It was hoped that prophy would greatly reduce or even prevent joint disease.

Yet some young men who have been on prophy all of their lives still have joint bleeds and are showing some signs of degenerative joint disease, sometimes in joints where they haven't reported any major bleeds.



A joint is where two bones meet. In haemophilia, it's the freely movable joints in the body (including the elbow, knee, and ankle), also called synovial joints, that are most likely to bleed.

These joints are surrounded by a tough, thick sheath of connective tissue called a joint capsule, which helps stabilize the joint and also seals and isolates it from surrounding tissues. Lining the inside of the capsule is a very thin layer called the synovial membrane.

Synovial fluid, coats the cartilage that covers the bone ends within the joint capsule, and serves as a lubricant to reduce friction between the moving bones.

Unfortunately if you damage cartilage, your injury may not heal, and will probably get worse over time.

Joint bleeds begin when one of the many tiny blood vessels of the synovium ruptures, spilling blood into the joint capsule. Untreated, the ruptured blood vessel fills the joint cavity with blood. Eventually, the pressure of the blood inside the joint capsule increases until it equals the pressure inside the blood vessel, so that the bleeding stops.

In the first stages of a bleed, the joint may feel warm, bubbling, or tingling. As the pressure of the blood within the joint increases, the swelling joint becomes increasingly painful. Later symptoms of a joint bleed often include stiffness, warmth to the touch, reduced range of motion, and inability to bear weight.

After a bleed has stopped, the body begins to remove blood from the joint. Cells in the synovium secrete enzymes that break down the blood, so it can be absorbed and removed by white blood cells. It may take four to six weeks after a joint bleed for the blood inside to be completely removed. Chronic inflammation of the synovium causes the synovium to overgrow and thicken, and this sets the stage for more bleeds.

An enlarged and chronically inflamed synovial membrane is called synovitis. Synovitis causes the joint to remain swollen and "spongy," even after treatment. Chronic synovitis contributes to more frequent bleeding into the joint, and this speeds cartilage damage, eventually causing haemophilic arthropathy, a crippling form of arthritis. Adding to these destructive changes in the joint, the muscles surrounding the joint often atrophy (weaken) because the patient uses them less, as a result of pain. Weak muscles provide less support for the joint, which also leads to more frequent bleeding. And the painful cycle of joint bleeds continues.

The only way you can prevent degenerative joint disease is to prevent joint bleeds and studies show that primary prophy has dramatically decreased (but not eliminated) joint damage in people with severe haemophilia without inhibitors.

Edited for size from an article of the same name by Paul Clement that appeared in Parent Empowerment Newsletter published by LA Kelley Communications, Inc. May 2020

3 Simple ways to be savvy on-line

Technology can be overwhelming if you didn't grow up with it, and now computers are everywhere. Whether you want to see your children's Instagram updates, have video chats with your grandkids or send emails to friends, you'll need some tech savvy.

1. Start simply.
Get a simple device with an internet connection. A computer can be hard to figure out, and a phone's small screen can be challenging to read so a tablet, such as an iPad is a good option. Then, focus on learning a few simple applications that help you accomplish your goals. This may include emailing, video chatting, streaming TV or movies, and checking social media.

2. Be very cautious online.
Older adults are common targets of scams, unfortunately, "because they are more trusting in general". They also tend to have financial savings that online scammers are after. One popular tactic is phishing. A phishing email looks like a real email from a legitimate source, such as your bank. Scammers want you to click links or provide personal information so they can access your financial or other accounts. If you are not sure about the source of a request, do not respond - and you can always ask someone you know to help you check something out.

3. Build your tech skills.
Turn to trusted sources to learn more about technology, such as classes on how to use specific applications (Zoom or Facebook, for example) or how to set up a tablet or computer. Classes directed to seniors move at a pace for people who did not grow up with technology all around them. Of course, tech-savvy family members and trusted friends can be helpful as well.

By starting with easy technology, taking steps to be safe online and building on your tech knowledge, you can enjoy the benefits of a more connected world.

Edited for size from "3 Simple Ways to Be Savvier and Safer Online" by Stephanie Conner https://hemaware.org/life/3-simple-ways-be-savvier

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After our research into the employment and career planning needs of members HFQ is developing small workshops and/or mentoring focused on career planning, employment, disclosures in the workplace and transitioning between different work environments.

To do this we need stories of successful careers and to hear from people willing to share that story and even spend some time mentoring participants if they are interested in a career path like that one you have (or did). This is not a commitment on your part just an expression of interest in sharing your experiences in small groups or one-on-one mentoring situations.

If this is you, please call the office on 0419 706 056 and let us know or email info@hfq.org.au

5 Minutes with Brett

What was your family like?
I grew up in Holland into a family with two older sisters. My father had a severe form of haemophilia with an inhibitor, so when I grew up, he was in hospital most of the time. He passed away when I was nine years old. Haemophilia was always a big part of the family life growing up because when he was in hospital, my mother would often be there and my sisters and I would take care of ourselves. And when he was at home, he was in pain very often, so we had to be quiet in the house.

The amazing thing was that my dad was quite a positive character and I've very fond memories of when he would be at home. He might be bedridden, but we would have Sunday night's dinner around his bed and afterwards we would watch television together.

Whenever he could, he would go to work. He would also take me to school and that would be our quality time together. When he could, when his mind wasn't clogged up with pain and painkillers, he would definitely be there for us and in a positive way.

He really focused on my academic skills and my mother played a big role fighting for me too. For example having birthday parties, even when I would be walking around with a brace on my leg. She fought really hard to give me a youth that was as normal as possible.

Now that I'm a mother myself, I'm quite impressed with how much freedom she gave me to experiment. I definitely admire that in her, and I'm very grateful for that because I know I needed it. But I know I understand now how hard that could have been for her.

They both wanted you to have a normal life. Does that mean you knew that you had Hemophilia quite early on?
I got diagnosed when I was one year old so I never knew anything else. In my first year I had quite a lot of bruises and behaviour where my mother thought it might be a bleed. But when she went to see my father's haematologist, he would laugh it away and say, Oh no, that's not possible and dismiss it. And when I was about one year old, they did do the factor VIII level test. And when he rang her, it was just like he was in shock too. They said that at that time, I was one of three women that were known in the world to have such low levels of factor VIII.

Have you had any issues about being a woman with a haemophilia diagnosis?
When I was about 11, I started administrating the FVIII myself, intravenously. . But before then, every time we would go on holiday, I would be dependent on the hospitals we would go to if I had a bleed. It would mean long conversations with the doctor on-call how it is possible that I have a bleeding disorder and that my mother had been tested negative for the gene. And yes, I have quite severe Hemophilia with levels below one percent. Those would be endless conversations. Even though I would bring my own factor VIII and was just asking them to administer it for me. The questions were tiring but understandable as it wasn't in the medical curriculum and no doctor wants administer something that might not be needed or harmful for their patient.

Have you had treatment decisions that you thought could have been better for you?
Moving to Australia I found it interesting that there's only a couple of treatments available.

Talking with Ira as a Woman with Haemophilia

I was surprised that the tenders would go out and a government body would decide which products would be available and you as the patient might have to switch. Whereas I grew up thinking the one thing I shouldn't be doing is switching products because that could increase the risk of inhibitors. So I found it quite surprising having to switch because of 'political' reasons. That's not something that I was used to in Holland. Having said that, I'm now on Hemlibra®, and that's definitely a game changer. How good is that?

What do you miss about being here compared to being back home in Holland?
My friends and family. Children are a bit more independent in Holland from a younger age, and that's something that I wish for my kids. Here kids are dependent on their parents bringing them everywhere. And I don't think that's necessarily good, especially not for teenagers.

Did you have any problems with your qualifications being recognised here?

My husband had finished his training as an emergency physician, and we had young children when he was offered a job for a year here in Brisbane. He had a lot of problems, as his training wasn't recognised here. So he had to jump through quite a lot of hoops, and by the time we decided we would stay longer, although I'd worked as a consultant psychiatrist for a couple of years before coming out, I decided that I would start a life coaching company rather than get my medical registration here.

It took us almost 10 years to get permanent residency and it was quite frustrating as Haemophilia often counts against migration so we had difficulty there, too, but by then I had my successful wellbeing program for junior doctors running and they could see that my contribution to Australian society was equally as big as the cost I would be.

Did Haemophilia shape your career choices.

To be honest, I didn't want to study at all. Then my sister said, If you don't study, then you have to work immediately and you can do that for the rest of your life. So I thought, OK, so if I go and study, what would I like to study? At first I studied year Italian because I wanted to do European studies, but it quickly became apparent that wasn't really my calling. And so my second choice was medicine.

At the start it was that I had enjoyed sciences at school and I thought medical school would teach you interesting topics. Maybe I was partly influenced because with Haemophilia I felt I had grown up with not much control over my body and treatments I received. . And part of it would be, gaining some control by at least being a doctor myself. I when I did my internship in psychiatry all the pieces of the puzzle came together and I thought, it just holds all the parts that I find important in being a doctor. Other specialties I found, were too narrowly focused. Because of my own experiences, I knew how each symptom or complaint can impact on the rest of your body and mental wellbeing. For example, If the patient is depressed, it impacts their overall quality of life, so that's why I chose psychiatry as my specialty.

For a young person with a bleeding disorder leaving school and thinking, What do I do for a career? How would you advise them?

So much has changed since I grew up. I've never been on prophylaxis. Which had a negative impact on my joints. Today you've got medication that can treat you as prophylaxis, so there shouldn't be any reason to stop you from doing anything (except perhaps a rugby player). But apart from that, I don't see any reasons why a person with Haemophilia wouldn't be able to do anything they wanted.

I learned some really valuable lessons from my Haemophilia. From the very first moment, I had to learn to read bodily sensations and act upon it to get treatment if it was a bleed. This is a skill that I now recognise as Mindfulness, It also taught me gratitude and to focus on the moments that I wasn't in pain or could walk, rather than remain stuck in the bad days. My father was a very good role model in having a positive mindset, trying to focus on the positives rather than the negatives in life. And I think it's not very surprising that the first programme I've written when I started my life coaching company was Mindfulness for Busy People because I knew first hand how having a mindful presence can be beneficial

The other thing I had to learn is to accept myself with my vulnerabilities. That also meant becoming more self-compassionate, this might be a big part of the same process. Self-compassion is treating yourself as your own best friend, being able to look at your own behaviour or your own actions without being overwhelmed by your own critical internal voice. That negative self-talk, the harsh, critical inner voice can be completely disruptive and cause a lot of negative outcomes, such as anxiety, depression, burnout, you name it.

I now use these insights of both sides (as a patient and a doctor) to help others, both patients, doctors and the general public finding healthier ways to deal with stress. So, there is something good that can come out of the Haemophilia as well.

Keeping Summer Activities Safe for kids

It's summertime, and kids are out of school and excited to have fun in the great outdoors. After a year of remote learning, social isolation and so much uncertainty, probably more than ever!

But if you're the parent of a child with haemophilia or another bleeding disorder, your eagerness for your child to get outside and be active is tempered by the concern that the sport he is playing or another activity he's doing might put him at risk for a serious injury. But there are things you can do to help make this summer an enjoyable and active one for your child while also keeping them safe from injuries.

Check with your QHC

A good first step is to talk to your child's haemophilia treatment centre (QHC) team. You can find out if your child has a problem joint and determine if there are specific sports that would be safer for them than others.

Clearly, it's a no-brainer that everybody should use the appropriate safety gear for each sport, but you can also talk about any measures you can take to make the sport or activity safer.

Steer Clear of high velocity and Activities with a high risk for collision

For more information about specific sports and haemophilia, ask the QHC staff for a copy of the book Boys will be boys by Brendan Egan. This includes details on injury prevention and appropriate protective equipment.

BMX, boxing, and football (including rugby & AFL) are sports that can be dangerous for anyone, regardless of whether they have a bleeding disorder, because they are considered high velocity and/or at high risk for collision. The most important thing we want to prevent is a blow to the head, which can cause a concussion and at the very worst, a head bleed.

Be Cautious with Trampolines

Sales of backyard trampolines jumped last summer as families looked for ways to keep their kids occupied at home during the pandemic.

A report on trampoline safety for the Australian Competition and Consumer Commission found that guidelines and recommendations on supervision, age of child and number of children using trampolines are being disregarded and are major risk factors for trampoline injuries in Australia.

If you choose to allow your child to be on a trampoline, the things that are going to make it safer are ensuring there's adult supervision and no flips or somersaults and, most importantly, having only one child jump at a time. You want to avoid the risk of two heads hitting each other really hard.

Don't Overlook Neighbourhood Games

While football in all it's form is a definite high-risk sport, what about a friendly backyard game of cricket of footie? These kinds of impromptu games can often be a lot riskier than you realise, because there's usually little to no adult supervision. With supervised team sports, you're going to have refs that are watching, but if it's just a bunch of kids playing up the street, it's possible that a player could go rogue and do something really dangerous.

Keep Your Child's Interests in Mind

Don't make the mistake of limiting your child's activities to only those considered "low risk. It's important to allow kids to have a say in the sports or activities they're interested in.

Some activities are actually less risky for kids with bleeding disorders than parents might think. Take rock climbing, for instance. While outdoor rock climbing can have risk, indoor rock climbing, with proper safety equipment such as ropes and harnesses, is probably OK.

Back in the day before we had such good treatment, every kid was encouraged to swim because it's not a high-impact sport, but what if your kid doesn't like swimming? Forcing your kid to do a low risk sport they hate is not helpful, and they're not going to get any fun out of it.

Edited for size from "How to Keep Summer Activities Safe for Kids with Bleeding Disorders" by Donna Behen. https://hemaware.org/mind-body/how-keep-summer-activities-safe-kids-bleeding-disorders

Liver Cancer Risk Lingers

Although the risk for liver cancer diminishes for patients with hepatitis C virus (HCV) who have eliminated the virus with direct-acting antiviral (DAA) drugs, you may not be out of the woods yet.

With the high success of the new treatments we often consider HCV to be sorted, but there are consequences on the liver from the virus and follow up / monitoring is often required.

A presentation of study findings from the PITER Cohort Study (a prospective, multicentre observational study of a representative sample of patients with HCV) at The Liver Meeting 2021: American Association for the Study of Liver Diseases (AASLD), showed that these patients are not out of the woods even though the risk for liver cancer diminishes for the large percentage of patients who have eliminated the hepatitis C virus with direct-acting antiviral drugs. A small percentage who do not have a sustained viral response are at substantially higher risk of developing hepatocellular carcinoma (HCC).

Not having a sustained viral response was associated with a more than sevenfold higher risk for liver cancer. The researchers said that failure to achieve SVR after DAA treatment is strongly associated with the probability of HCC development. Older age, [HCV] genotype 3, and low platelet counts and albumin levels are independent factors of HCC development despite viral eradication.

Long-term Follow-up Required

Dr Raymond Chung, director of the Hepatology and Liver Centre at Massachusetts General Hospital, Boston, was not involved in the study. He says it's useful to think of hepatitis C as a viral infection on one hand and a liver disease on the other.

In terms of thinking of elimination, most patients can eradicate the virus with virtually 100% success with antiviral treatment, but this may give a false sense of comfort because despite the elimination of the virus, patients may still have significant liver fibrosis or cirrhosis and it is imperative that patients be monitored for signs of cancer.

This is why it's important to understand how severe your liver disease is, because if there is advanced fibrosis, bridging fibrosis, or cirrhosis, you are probably going to need long-term oncology care. The infectious disease is eliminated, but the liver disease remains so always discuss your situation with your specialist team - don't ignore it.

Edited for size from Medscap e Conference News from: The Liver Meeting 2021: American Association for the Study of Liver Diseases (AASLD). https://www.medscape.com/viewarticle/963101#vp_1

Is it **Possible** to **'Cure' HIV?**

Dr. Xu Yu, a principal investigator at MIT and Harvard, has published a report which suggests hints that HIV might be able to be cured. The study says that a women patient who tested positive for HIV, was somehow able to control the virus with her body's immune system and block it from places like the lymph nodes – all without being on anti-HIV drugs.

If that's the case, she would be only the second case of a patient curing themselves of HIV. There have been previous reports of patients who stopped taking anti-HIV medications and achieved undetectable viral levels for years but these patients benefited from stem cell transplants. And they probably continued to harbor latent reservoirs of HIV, which means they haven't been entirely cured, but what scientists call functionally cured.

That doesn't appear to be the case with Dr Yu's patient. And while this and another similar San Francisco case are encouraging, Dr Yu cautions that the findings may not be generalisable to most HIV patients. Both of her patients belonged to a group known as elite controllers, or people who are able to suppress HIV at very low, often undetectable levels, with their immune systems without the help of anti-HIV drugs. Researchers are studying these people intensively, looking at everything from the antibodies they make to the highway that immune cells use that includes lymphocytes.

Dr Yu is hopeful that these patients, and others like them will help answer critical questions such as whether tweaking current combinations of anti-HIV drugs could help others mirror the response her two patients have had. One thing she is hoping to do, for example, is compare the immune response among these two patients to that produced by people who are taking anti-HIV drugs. It's possible that the drug cocktails produce an immune response weaker than what's generated by these two patients, and scientists could find ways to bolster that response.

Edited for size from A Possible Sterilizing Cure of HIV-1 Infection Without Stem Cell Transplantation
Gabriela Turk, PhD et al. https://doi.org/10.7326/L21-0297

Rapid ageing and **inflammation** source in **PLWHA identified**

Researchers have found that elusive white blood cells called neutrophils play a role in impaired T cell functions and counts, as well as associated chronic inflammation that is common with the virus.

In a study, published in the journal PLOS Biology, Prof Elahi and his team ran comprehensive sequencing on all the genes expressed in the neutrophils from a group of people with HIV and a control group to determine any differences between them.

Not all HIV-infected individuals have similar types of neutrophils. As the disease progresses, neutrophils become more activated and more potent, and in turn activate the body's T cells, which likely causes some of the problems associated with HIV infection such as inflammation and rapid ageing.

One of the proteins released by neutrophils is galectin-9, which can lead to a hyper-immune response and inflammation in PLWHA. Based on his findings, Dr Elahi said preventing galectin-9 shedding might be a powerful tool in reducing many of the negative effects of HIV infection.

https://www.ualberta.ca/folio/2021/11/landmark-study-points-to-source-of-rapid-aging-chronic-inflammation-in-people-living-with-hiv.html

Health Updates

Potential Gene Therapy SPK-8011 Fares Well in Haemophilia A Study

A single dose of Spark Therapeutics' investigational gene therapy SPK-8011 increased the levels of factor VIII (FVIII) and, in 16 of 18 male patients, those levels were sustained for up to four years, according to results from a Phase 1/2 clinical trial and its extension study.

Trial findings were detailed in the study, "Multiyear Factor VIII Expression after AAV Gene Transfer for Haemophilia A," published in The New England Journal of Medicine.

Notably, the sustained FVIII levels allowed the discontinuation of prophylaxis, or preventive FVIII therapy, and resulted in a pronounced reduction or complete elimination of bleeding episodes.

This FVIII level stability stands out from results of previous trials of Roctavian, another experimental gene therapy showing that, following a sharp increase, FVIII levels started to decline after a year and up to four years.

These findings offer compelling data to support the current approach to haemophilia A gene transfer can indeed confer stable factor VIII [production] over multiple years for near disease ameliorating effect.
https://www.nejm.org/doi/full/10.1056/NEJMoa2104205

Catalyst Halts MarzAA Program for Hemophilia, Despite Positive Results

In an unexpected turn, Catalyst Biosciences is discontinuing the clinical development of marzeptacog alfa activated (MarzAA), its experimental under-the-skin therapy for hemophilia A and B patients with inhibitors, which was being evaluated in an international Phase 3 clinical trial.

The company said that based on several factors including a recently updated feasibility assessment, They determined that they cannot continue to develop MarzAA through completion of the ongoing trials.

Enrolment in these trials had been adversely impacted by several factors, including pandemic-related logistical challenges, competition for subjects, and increasing availability of prophylaxis [preventive] therapy globally, making it impossible to deliver top-line data in 2022.

Catalyst plans to share Crimson-1's results obtained to date, showing that they have successfully treated bleeds with [under-the-skin] MarzAA and have not observed any treatment-related adverse or thrombotic [blood clotting-related] events.

The trials had been expected to conclude in March 2022.
https://ir.catalystbiosciences.com/news-releases/news-release-details/catalyst-biosciences-announces-change-corporate-strategy

One in five women with heavy periods have hidden blood clotting disorder

Thousands of young women who experience heavy periods are unknowingly suffering from a genetic disorder that puts them at high risk of life-threatening complications during childbirth.

Women can also be carriers of the haemophilia gene and be mildly affected – something many doctors are unaware of. There are other blood-clotting disorders that affect men and women equally that are more common but, paradoxically, are less well known. While in men they often cause few symptoms, women who suffer them often experience problems with menstruation.

According to international studies, one in five women who seek medical advice for heavy periods will have a bleeding disorder but aren't tested, which experts say is a missed opportunity. Heavy periods are linked to anaemia, a lack of red blood cells in the body that leads to symptoms including debilitating exhaustion, feeling extremely cold and pale skin.
https://www.nation.lk/online/one-in-five-women-with-heavy-periods-have-hidden-blood-clotting-disorder-149715.html

Immune system-stimulating nanoparticle could lead to more powerful vaccines

A common strategy to make vaccines more powerful is to deliver them along with an adjuvant -- a compound that stimulates the immune system to produce a stronger response.

Researchers from MIT have designed a new nanoparticle adjuvant that may be more potent than others now in use. A study on mice published in Science Immunology, showed it significantly improved antibody production following vaccination against HIV, diphtheria and influenza.

The researchers tested the adjuvant by injecting it into mice along with a few different antigens, or fragments of viral proteins, including two HIV antigens. They compared the adjuvant to several other approved adjuvants and found that the new nanoparticle elicited a stronger antibody response than any of the others.

The researchers now hope to incorporate the adjuvant into an HIV vaccine that is currently being tested in clinical trials, in hopes of improving its performance.
https://news.mit.edu/2021/vaccine-immune-nanoparticle-1203

Phase 3 Trial of Gene Therapy SB-525 for Haemophilia A on FDA Hold

The U.S. Food and Drug Administration (FDA) has placed a clinical hold on the Phase 3 trial evaluating SB-525 (giroctocogene fitelparvovec), an investigational gene therapy for haemophilia A.

After being treated with SB-525, some patients showed FVIII levels greater than 150%, and higher-than-usual factor levels raise a risk of blood clots. No adverse events attributed to these elevated levels have been observed, and some patients were given oral anticoagulants (blood thinners) as a "precautionary measure".

Pfizer voluntarily paused patient screening and dosing to implement a change in the trial's protocol that gives guidance for managing people with elevated FVIII levels.

Following their decision to pause the study, the FDA informed Pfizer that the program was placed on clinical hold on November 3rd. Further trial updates from the companies are expected after the FDA's review of the proposed protocol amendment.
https://www.ehc.eu/pfizer-hemophilia-a-clinical-program-update_november-5th-2021/

Important Dates for HFQ Members

Covid-19 is still a concern and all HFQ activities are subject to any restrictions that may apply at the time of the activity.

OBE Men's Meetings:
Sunday, 12th December 2021
Arana Hills League Club

Wednesday, 12th January 2022
Venue: TBA

Women's Lunch :
Sunday, 6th February 2022
Everton Park Hotel

Australia Day Event:
Saturday, 22 January 2022
The Blue Room Cinemas, Rosalie

Rare Diseases Day
28 February 2022

Please ask for events and activities happening in your area.

Please call Graham or Sam at the office on **0419 706 056** for more info on any of these events and other activities.

The HFQ office staff are taking a break from the office over Christmas but you can still get us if you need us via the office mobile 0419 706 056. Our last day in the office will be 24 December 2021 and we will return on 4 January 2022.

The festive season can be a fun and an exciting time for catching up with family and friends, attending parties and events, buying gifts, and preparing yourself for social times. While it can be a busy and stressful time for many, for others this can bring about feelings of isolation and loss.

We wish all our readers a well-deserved rest and a safe and joyful holiday season. If you find yourself caught up in the hype of festivities, take a few moments to check that other people you know are OK and don't forget about taking care of yourself too. We look forward to seeing you at our events in the new year.

About The H' Factor

The 'H' Factor is published four times each year by HFQ by the HFQ manager and assisted by Brett Williams, our communications volunteer. We occasionally send important information and updates on local and relevant events for people affected by bleeding disorders to subscribers of our email list. If you would like to be on the HFQ Email List, please register your interest by sending through an email with the subject title The 'H' Factor email list' to info@hfq.org.au. You can be removed from the list at anytime.

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