the H FACTOR

Haemophilia Foundation Queensland

WINTER EDITION

Issue 67

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HFQ MEMBER MAGAZINE

From the President

Hello everyone,

Does haemophilia still exist - well the answer is obviously yes - however for many that I have spoken to who have transitioned to either newer longer 1/2 life treatments or newer bypassing agents like Hemlibra, it's simply amazing to generally hear such good news stories of reduced annual bleeding rates and improved quality of life. We have our great country to thank for providing these new life changing treatments …. In contrast, it's a sober reminder of how lucky we are compared to places like Fiji and many other countries where this is currently an unattainable dream.

For me personally I am living the dream since changing to extended 1/2 life product - effectively moving from severe to mild with currently no bleeds. This has expanded my abilities and improved my quality of life as well as preventing further joint damage - simply brilliant!

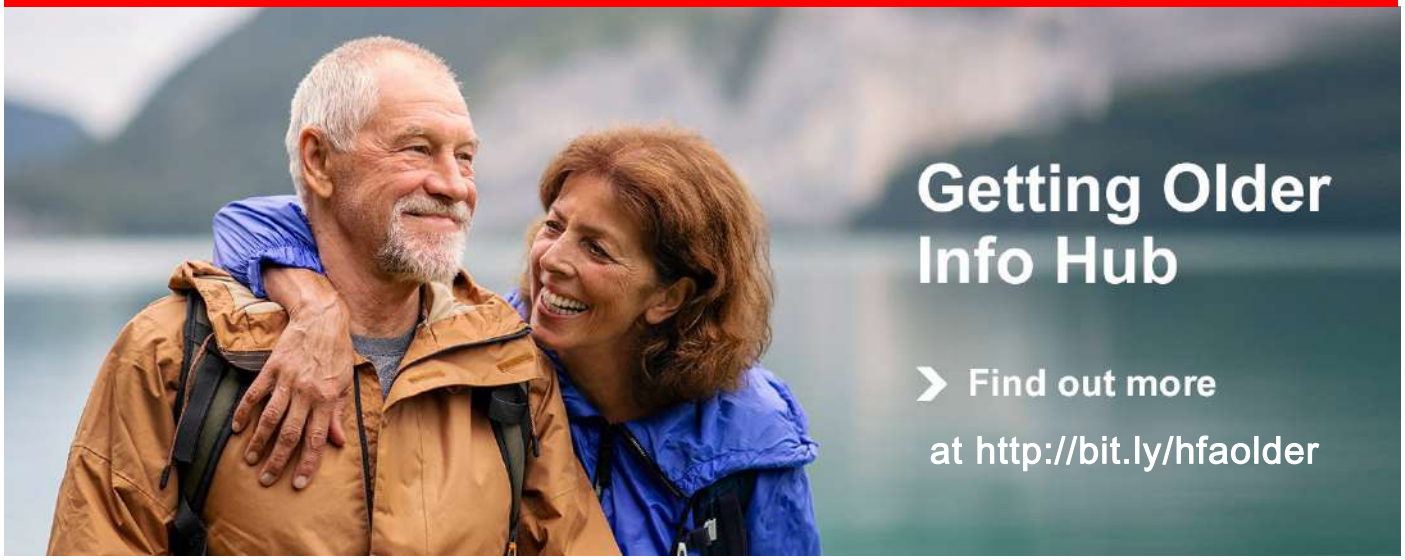
HFQ continues to provide services in partnership with Queensland Health to address identified issues that exist across the different demographics, an example is our careers service project where we are working with a masters student as well as bringing a synergy with a USA organisation (www.hemob.org) together to research, identify and look at the challenges that having a bleeding condition brings. We will let you know how this develops and what the practical outcomes / recommendations are.

Until next time – take care

Dave

David Stephenson
President HFQ
president@hfq.org.au

A Great Resource As You Get Older



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	... ...	Mrs Lauren Green
Treasurer	... ...	Mr Adam Lish
Members	... ...	Mrs Belinda Waddell
		Mr Charles Eddy
		Dr Jodie Caris
		Mrs Leanne Stephenson
		Ms Shannon Gracey
		Mr Shannon Wandmaker
		Mr Tony Ciottariello

HFQ Delegate to HFA

Mr Adam Lish

Acknowledgements

HFQ is grateful for the support of our patron: His Excellency the Honourable Paul de Jersey AC.

HFQ programs and services are funded by the Queensland Government.

HFQ is also grateful for the support it has received from the Prescott Family Foundation.

Internet

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

QUEENSLAND 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 Antoinette Runge

Haemophilia Registrar – Dr Chintaki (Chinthi) Jayasekera

Joanna McCosker – Nurse Practitioner

Amy Finlayson / Salena Griffen – Clinical Nurse

Elise Mosey (Mon, Tues) - Physiotherapist

Hayley Coulson (Wed, Thur, Fri) – Physiotherapist

Dr Moana Harlen - 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)

Dr Jane Mason - Haematologist	3646-8111
	(Page through switch)
Haemophilia Registrar	3646-8111
	(ask to page Haemophilia Registrar on 59716)
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

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 8-9.30

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

July to Oct 2021

Some of the HFQ programs and activities already planned

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

JULY

- **OBE's Monthly meeting** 7th July, East Leagues Club
- **HFQ Board Meeting** 20th July, 298 Gilchrist Ave, Herston
- **National Pain Week** 22nd to 28 July
- **World Hepatitis Day** 28 July

AUG

- **OBE's Monthly meeting** 4th August, Fitzy's Waterford Hotel
- **Dental Health Week** 2nd to 8th August
- **HFQ Board Meeting** 17th August, 298 Gilchrist Ave, Herston
- **Women's Lunch** 21st August, Venue: Everton Park Hotel

SEPT

- **OBE's Monthly meeting** 1st September, Mango Hill Tavern
- **R U OK Day** 9th September
- **HFQ Youth Camp** 17th to 19 September, Emu Gully
- **HFQ Board Meeting** 21st September (Virtual)

OCT

- **OBE's Monthly meeting** 6th October, Fernvale Bakery
- **20th Australian Conference on Haemophilia, VWD & Rare Bleeding Disorders** 8th & 9th October (Virtual)
- **Bleeding Disorder Week** 11th to 17th October
- **HFQ Annual General Meeting** 19th October, 298 Gilchrist Ave, Herston

Bleeding Disorders Awareness Week

We've just said good by to World Haemophilia Day (see photos p 16) and it's time to start thinking about the other important event in the lives of people affected by bleeding disorders. 10 to 16 October is Bleeding Disorders Awareness Week this year. This week is an important opportunity to raise awareness in your own community about inherited bleeding disorders such as haemophilia and von Willebrand disease.

In Australia there are more than 5,300 people with haemophilia, von Willebrand disease or other inherited bleeding disorders so why not **Go Red for Bleeding Disorders** + fundraise. It's easy and fun to get involved, so start planning now so that you can inform your friends and family in a fun activity utilizing the colour red.

Bleeding Disorders AWARENESS WEEK

A Fond Farewell (by Loretta)

I have been agonising over what to write for weeks. For those who haven't heard – I'm sorry I didn't get to tell you in person (or over the phone), but I have resigned from my position with the QLD Haemophilia Centre. I have and continue to be humbled by hearing your stories and I feel privileged to have been able to work alongside you during this time. I have learnt so much in the time I have been involved in the inherited bleeding disorders community, especially around the collective community's resilience and determination, the compassion and caring from peer supports and celebration over what you have each achieved.

Change happens for all of us. New treatment come along; people come into your life, whilst others leave; we change houses, or declutter them; change jobs; manage pandemics, etc. Change can be messy, but a teacher of mine recently reminded me that change can also be magical. I guess you just need to get through the messy bits to see all the new possibilities. A Life Simply Lived Psychology in rural Victoria, posted on FaceBook a lovely metaphor that I would like to share. "Sometimes life can feel a little like driving down a dirt road after some rain. You know that it will be messy and that you may even get bogged. But if you want to get where you are going, it is the only way through. And as long as you can see out the front at what is immediately ahead, and a bit out the back at what is behind you, you'll be able to keep moving forward. No matter how much mud flicks up and hits you."

So, it will be a bit muddy for me over the coming months as I move town and settle into my new job. I will no doubt see some dirt roads in the coming years – literally (through my move to a rural town) and metaphorically. My hope is that it will not be too slippery for you during the time before a new Social Worker or Psychologist starts here in the RBWH Haemophilia centre. We have all survived 100% of our worst days. Our resilience, experience, gratitude/appreciation of the things that are good and our supports will assist us to continue to manage those times when we struggle.

Over the weekend, I was going through quotes I had kept over the last few years and came across this one. I am embarrassed to say it is from a tv character, however, it serves as a reminder for me and maybe will resonate or be helpful to someone else. "So, do it. Decide. Is this the life you want to live? Is this the person you want to love? Is this the best you can be? Can you be stronger? Kinder? More Compassionate? Decide. Breathe in. Breathe out and decide." (Meredith Grey – Grey's Anatomy).

I hope we can all be the best versions of ourselves for ourselves, not for anyone else, just yourself. Treat yourself with the kindness and compassion that you would give your best friend. Notice the good around you.

Thank you so much for being a part of my life and allowing me to be a part of yours.

Loretta

Talking to Dave on Extended Half Life Factors

What is your bleeding disorder?

As a child my parents took me to hospital with a knee the size of a party balloon. The doctors didn't understand why this was happening so the just decide to syringe the blood out. It was much later when I was 6 that a GP put the puzzle together and I was then diagnosed with severe haemophilia B.

Having haemophilia B has had a huge impact on my life. In the early years I spent more time in hospital than I did in school. I had to wear a calliper to stop any movement in my knee which meant I stood out from the other kids, and some of the bullying was quite extreme. The treatment back then was on-demand where I had to travel over an hour to Melbourne to go to the emergency department where I waited in pain for five or six hours before being seen, unfortunately significant joint damage resulted.

In my early 20s I wanted to become a teacher, but when I got in the classroom I found dealing with kids wasn't for me, so I changed to an electronic technology pathway, which was a passion of mine that led to quite a good career. Haemophilia was a secret because of promotion opportunities. People can't help being judgemental for good or bad reasons, I kept it quiet.

But then I met Leanne, and along came marriage. Leanne thought that I'd probably be dead by 30, because that's what people read back then. But she continued on, and we're still together today 38 years later.

What treatment are you on?

I'm on Aprolix which is an EHL (extended half-life factor). EHL's have been around overseas for a while but relatively new here in Australia.

I had concerns about changing treatment as I knew there was a small chance of an inhibitor developing which in Haemophilia B can possibly result in life threatening anaphylaxis – very different to Haemophilia A. Given my standard ½ life treatment was working ok I initially declined to change. As time passed there was additional evidence in Australia that it was indeed safe – no one had developed an inhibitor.

Have you noticed any changes, since going on to EHL's.

I really wish this happened ten years ago. This is brilliant stuff. Since the change I can stand longer, I can walk further, I can lift things and most of those niggly pain like issues & micro bleeds have predominantly gone. So it's been a significant and positive change, it's a shame that it didn't come along a little earlier because it would have prevented some joint damage.

What would you tell other people with Haemophilia B about EHL's?

For me, it's an amazing treatment, I would tell them that given my great outcomes, I wish it was available much earlier. It's effectively moved me from severe to mild and you know those micro bleeds, they're gone. An improved quality of life is what I have now, and I can see how this will benefit everyone in all sorts of ways.

I'd also tell people to discuss any questions with their haematologist, but before you do that, write down all your questions beforehand. Think about it overnight and have any further questions answered, so that you really have all the facts in front of you. We all know people don't like change, so it's often challenging with fears and concerns. But look at the experiences of others, the medical advice and be supported by that.

Our Bleeding Experiences

OBE's (Our Bleeding Experience) is an enjoyable social gathering normally held on the first Wednesday of each month for Men in the Bleeding Disorders Community. Most members of our community who attend these gatherings are no longer working so this day works for them.

HFQ aims to hold an OBE event on the weekends at least twice a year to open it up to more men in our community who are unable to make it to the Wednesday gatherings due to work commitments, etc.

The next **WEEKEND OBE** event is scheduled for **Sunday 8th August 2021 at Fitzy's Tavern - Waterford.**

So, if you are male who has a bleeding disorder, or maybe a Dad who has a child with a bleeding disorder, or you may be a bloke who has a father with a bleeding disorder, please feel free to come and join us for a meal, beer, coffee and a good old yarn.

This is a social gathering to enable men in our community to catch up in a casual environment and meet with other men who may be facing the same challenges or issues as yourself in day to day life.

HFQ is part funded by Queensland Health. While we welcome donations on the day to help fund future events like this, please don't let finance stop you coming.

For more information or to RSVP please call 0419 706 056 and speak to Graham or Samantha.

COVID-19 Treated in a Patient with Haemophilia

The course of infection with SARS-CoV-2 virus in patients with congenital bleeding disorders does not differ from the general population. COVID-19 in these patients can be mild but also can progress to severe pneumonia, respiratory failure and death. The treatment of patients with COVID-19 and haemophilia A, receiving emicizumab for bleeding prevention is particularly challenging because of the need to balance the increased risk of thrombotic events and bleeding.

A 70-year-old male patient with severe haemophilia A on emicizumab prophylaxis developed severe COVID-19. Prior to COVID-19, he was on emicizumab prophylaxis at a dose of 3 mg/kg once weekly and his weight was 85 kg; he had no history of bleeding episodes nor thromboembolic events. His comorbidities include hypertension and HCV infection successfully treated in 2008.

He was admitted to hospital with SARS-CoV-2 infection, the patient's respiratory status deteriorated requiring intubation and ICU admission. Chest X-ray showed numerous bilateral ground-glass opacities and consolidations. The patient received treatment for COVID-19. The clinical examination revealed abdominal tenderness, and an increase in total bilirubin levels and drug resistant staph. A diagnosis of ventilator-associated bacterial pneumonia and secondary sepsis was made. Emicizumab at weekly dose of 3 mg/kg was continued.

During ICU treatment, including invasive procedures (intubation, central and intravenous catheter placement, bladder catheter placement, tracheostomy, enteral feeding tube), no bleeding or thrombotic incidents have been observed. By day 20 the patient had two negative SARS-CoV-2 test results. Contra indications gradually decreased and the clinical condition improved. On day 30, the patient was breathing independently and at present, the patient is at home, and is again on emicizumab prophylaxis.

Patients with severe congenital bleeding disorders are less susceptible to clotting associated with SARS-CoV-2 infection. On the other hand, additional factors, such as sepsis which was diagnosed in our patient may enhance different prothrombotic pathways, resulting in the activation of coagulation factors and may worsen the course of infection. Finally, patients with severe haemophilia on regular prophylaxis with FVIII and/or emicizumab have their baseline haemostasis improved and therefore seem to be at higher risk of thrombotic complications as compared with severe haemophilia patients not treated prophylactically.

It is not entirely clear which of used drugs or complex supportive care had a decisive impact on the cure of this patient highlighting the need for additional research to optimize COVID-19 therapy.

Edited for size from "Successful treatment of COVID-19 in a patient with severe haemophilia A on emicizumab prophylaxis in the intensive care unit" by Donata Urbaniak-Kujda et al., and published in the Haemophilia journal https://doi.org/10.1111/hae.14326

Talking to Natalie about Rocky and Hemlibra

Tell me about life before hemlibra.
Rocky has severe haemophilia A. When he was little, we opted for a port even though we knew that there would be the chance of port infections, which we didn't want. At some point, his port stopped working, so he had to have that one removed and another one put in. But he went from being on Advate three times a week to every other day because he kept getting ankle issues. So, I was accessing his port every other day. And that was right up until he started his hemlibra in December of last year.

How did you find out about hemlibra?
We'd heard about it a few years back at a community camp with a boy on compassionate access. And I was just like "let's get this stuff over here as soon as possible, so we can all start having it". So, when funding for Hemlibra was announced I was jumping up and down and wanting to get Rocky on it, basically advocating for his needs, not to mention his port was playing up anyway, so it was really good timing.

Have you seen anny differences him?
We've already had one visit to the hospital since being on it with a toe bleed, but nothing really dramatic has changed.

He's been doing it in his leg as he's not game enough to do it in his belly yet and that's completely up to him. I've not done one thing with him since hemlibra. So a lot has changed for Rocky. He's been very empowered. He's able to take control of himself and how he wants it to go. Except for one time when I told him no, he has to do it today, we were kind of in a rush and it had to happen before school starts.

Before you'd have to put aside an hour or two each time, but he does it himself now too. Before it was Michael or me having to do it. Now it's about reminding him, and picking it up from the pharmacy. Besides that he asks us to put it under our armpits to bring it back to body temperature so it's not going in cold.

Any actual side effects or risks using it that you've noticed?
Initially, there was a red mark around the needle site which went down and possibly it was a little bit itchy at one time, but that was only the first four or five weeks and probably only happened two times out of the five. Other than that, he's not had any reactions to it at all.

Have you been able to get your heads around that concept that taking hemlibra is like going from being severe to mild.
I don't think we have, I don't think I ever will, but that's just being a mum, because we did without hemlibra for ten and a half years. I think Rocky is still as cautious as he was before, and same with us as well. However, I would say though, if he has got a little niggle, we kind of watch and wait and see. and if it becomes worse, we are then a bit more proactive and call someone, particularly that time with his toe We kind of watched and waited and he just sort of said, nah, this is not good, mum. So for him to say that, and know that he had to go to the hospital to have a needle in his arm, was my indication to say, that's not good.

So, we take him to the hospital if we need too because Rocky doesn't have a port anymore and I can't do vein access for him. Rocky has to go to hospital to get treatment. The only downside for Rocky and me, is that I haven't been able to practice and do vein training where we could just treat at home.

What would you say to someone thinking about starting hemlibra?
It's life changing. It really is. it's an hour, almost two hours of your day not having to use the port. First, you've got to add the hour on for the anaesthetic cream, and then it's time to make sure the area that you're doing is clean and then you've got to set up and then you got to actually do it and then take it away. So, it takes time.

even though our heads haven't probably wrapped around the fact that he's no longer supposed to be severe haemophilia, but he's not. It's definitely great to see, and I wouldn't change it for the world, and I can't wait to see what it does for my dad as well.

It's already life changing for us and just think of the possibilities that we haven't even tried yet. And for my dad too, every time I call my mom at the moment, he's got bleed. So, for him to not even have that will be an amazing change. Like Rocky, his life span will be even longer.

Did CSL abandon people with Haemophilia?

HFQ member Greg Ball has lost nearly everything after being infected with Hepatitis C following routine treatment in the 1980s for his haemophilia. Greg developed cancer as a result. He was about to receive a lifesaving liver transplant in March last year but that was cancelled after doctors found that tumours on his liver had spread to his bowel.

Greg ran a successful business as a house painter but when his health started failing, he had to stop work, sell the family home and go on the disability pension. Now he is in a fight to stay alive.

Greg is just one of an estimated 1750 Australians with haemophilia who received contaminated blood products manufactured by CSL in the 1980s. Some 80% of Australia's haemophilia population acquired Hepatitis C; about 1400 people with haemophilia have since died.

Up to 20,000 Australians were infected, often following routine surgery. And the closest Australia has come to a national reckoning was a Senate inquiry in 2004, which found that the best scientific advice at the time was followed, a conclusion that helped head off demands for a judicial inquiry. Yet such a conclusion is debatable.

CSL and Factor Products

Key to the scandal is the blood products, plasma derived Factor VIII and IX because treatment was made by pooling plasma from up to 40,000 donors. Just one infected donor would contaminate the entire batch.

CSL introduced Factor VIII into Australia in the late 1970s. A key question is whether CSL mixed foreign-sourced plasma with Australian-sourced plasma to make its Factor. In the 2004 Senate inquiry, the medical and research director of CSL Bioplasma stated that Australian plasma had been mixed only with New Zealand plasma, and that this practice ceased in 1984.

However, according to publicly available information, CSL was still mixing Australian plasma with New Zealand's in 1986. CSL was also reportedly mixing Australian plasma with foreign-sourced plasma, including from countries in Asia that were considered high risk because Hepatitis C was rife.

As reported by the Canberra Times in 1994, the Red Cross wrote to CSL in 1986 requesting that Australian plasma be processed separately from plasma collected from Asia, Papua New Guinea and New Zealand because of concerns about Red Cross' legal liability in the case of plasma mixing. CSL's managing director, replied that CSL was "dismayed and perplexed" by the request because abandoning the pooling practice would cause "prohibitive cost increases".

The Commonwealth Auditor-General reported that in October 1998, on a visit to the US, a Health Department officer discovered that CSL had breached safety regulations by importing and processing plasma from at least one US source without the Health Department's knowledge. Health Department officials later raided CSL's Broadmeadows facility on November 24, 1998 and confirmed breaches of safety protocols.

The 2004 Senate inquiry found that in 1985, the UK began a new heat treatment that cleared plasma products of the virus that became known as Hepatitis C. Yet Commonwealth Serum Laboratories did not introduce this treatment into Australia for another four years, until the end of 1989.

The privatisation of CSL was one of the worst deals ever done by an Australian government, according to respected economist John Quiggin in September last year. It was helped by the Australian government granting CSL an indemnity against claims by "any person who becomes HIV-positive or contracts an AIDS-related condition or hepatitis as a result of … a CSL product derived from Australian plasma."

Some would say that a proportion of that success was built on the back of the deaths and serious illnesses of thousands of Australians.

Edited for size from an article that appeared in Michael West Media https://www.michaelwest.com.au/in-cold-blood-how-privatisation-of-csl-abandoned-the-victims-of-australias-public-health-tragedy/

Covid-19 Vaccinations FAQ (from AHCDO and

With the rollout of the COVID-19 vaccine commencing in Australia, members of the community have asked us about how this will impact on people with bleeding disorders.

The Australian Haemophilia Centre Directors' Organisation (AHCDO) has endorsed the joint COVID-19 vaccination guidance for people with bleeding disorders, produced by the World Federation of Hemophilia (WFH). This has detailed information and is available on the AHCDO website (*https://bit.ly/3oKvkDU*)

AHCDO has advised HFA on some answers to some common questions.

These FAQs may be updated as more information becomes known.
This information was reviewed in April 2021 and AHCDO has confirmed there is no change to this advice.

Frequently Asked Questions

Q1 – Is the COVID-19 vaccination safe for people with bleeding disorders?

A - In general the COVID-19 vaccine is as safe and effective for people with bleeding disorders as for anybody else without a bleeding disorder. As with all immunisations, there are some steps you may need to take before being vaccinated. See Qs 4,5 and 6 below.

The Australian Government has a careful and thorough process to check that the COVID-19 vaccines in Australia are safe and effective before it makes them available to the community. You can find more information about this on HealthDirect, the Australian Government-funded health information website - www.healthdirect.gov.au/coronavirus

Q2 – Am I in a priority population because of my bleeding disorder?

A - People with bleeding disorders are not at greater risk of contracting COVID-19 or developing a severe form of the disease, so they are not considered a priority group for vaccination.

The Australian Government will roll out the vaccine in phases, starting with priority populations. Some groups have been prioritised because they will be the most affected if they become infected with COVID-19. Information on the phases for the vaccine rollout is on the HealthDirect website (*https://bit.ly/3c9ArlX*) and click on Who will get the COVID-19 vaccine first?

Q3 – Where will I receive my vaccination?

A - The Australian Government has not yet announced specifically where Australians in the community will receive their vaccines and more will be known when this information becomes available. This may include hospital hubs, some general practitioners (GPs) and community pharmacies. Haemophilia Treatment Centres may not be accredited for vaccination. HTCs will update you about their position as they are advised of this information.

You can find more information about getting the vaccine on the HealthDirect website website (*https://bit.ly/3c9ArlX*) and click on Getting the COVID-19 vaccination

Q4 – Do I need treatment for my bleeding disorder before I have the vaccine?

A - Both of the currently approved vaccines require 2 intramuscular injections over a number of weeks for full vaccination. They cannot be given sub-cutaneously (under the skin) like the Fluvax.

You may also need to have treatment beforehand to prevent bleeding from the injection. Please contact your HTC to discuss this.

If you have a moderate or severe bleeding disorder, such as haemophilia or VWD or a rare clotting factor deficiency:

- If you are on prophylaxis with clotting factor concentrate, time it to have it on the day of your vaccination before the injection
- If you do not routinely give yourself factor, please contact your HTC for advice
- If you are taking emicizumab (Hemlibra®), whether you have inhibitors or not, just follow your usual treatment plan - you do not need to take any extra treatments

HFA)

before the vaccine injection.

If you have mild haemophilia or Type 1 or Type 2 VWD:

- Usually you will not need any special treatment with factor concentrate or DDAVP before the vaccine. Please follow the general precautions for immunisations - see below.
- However, if you have ever had a problem with bleeding from an injection in the past, please contact your HTC or haematologist for advice before you have the vaccine.

Q5 – How do I prevent bleeding with the vaccine injection?

A – As you would do with any immunisation, let the health care provider who is giving the vaccine know that you have a bleeding disorder.

- Ask them to use the smallest gauge needle that is available for the vaccine. Some COVID-19 vaccines must be administered with the needle and syringe package provided and a smaller gauge needle may not be possible.
- Apply pressure on the injection site for 10 minutes after the injection to reduce bleeding and swelling
- Check the injection site several minutes and 2-4 hours after the injection, both visually and by touching it, to make sure bleeding and swelling (haematoma) has not occurred
- You may have discomfort in the arm for 1-2 days afterwards. If it becomes worse and there is swelling, contact your Haemophilia Treatment Centre (HTC)
- Do not lift anything heavy with that arm for 24 hours, eg, shopping bags, gym weights, handbags.

Q6 – Does my bleeding disorder mean I am more likely to have an allergic reaction?

A - No.

It is rare, but some of the vaccines are known to cause allergic reactions in people who have a history of severe allergic reaction.

If you have ever had an allergic reaction to any vaccine or drug (for example, a severe allergic reaction to PEG or other vaccines) or have had other severe allergic reactions, you should talk to your doctor before you have the vaccine.

If you experience an allergic reaction after the vaccine injection (fever, warmth, redness, itchy skin, rash, shortness of breath, or swelling of the face or tongue), contact your doctor immediately and go to the nearest hospital emergency department straight away as it can be life-threatening.

Q7 – Do I need to have the Fluvax as well as the COVID-19 vaccine?

A - Current advice is that people should still have a Fluvax this season as well as the COVID vaccination.

Ask your doctor about having Fluvax and the timing of having it if you are also having the COVID vaccination.

If you have any questions about your bleeding disorder in relation to the COVID-19 vaccine, contact your Haemophilia Treatment Centre or your treating haematologist.

Important Note: This information was developed by Haemophilia Foundation Australia for education and information purposes only and does not replace advice from a treating health professional. Always see your health care provider for assessment and advice about your individual health before taking action or relying on published information.

5 Minutes with Brett

This time instead of 5 minutes with Brett, I thought I would tell you where I have been for the last four months and NO it was not on a sunny beach on some island in the Whitsundays.

It all started on 22 December last year. I have one of those beds where you can raise the head, or feet, or the entire bed. The night of the 21st I raised the feet end as my feet were very swollen. After taking my prescribed cocktail of drugs for pain, depression and anxiety. I drifted off to sleep.

The next thing I remembered was falling and I said to myself Oh F*!k this is going to hurt. Then I hit the floor. My mum and youngest nephew came running into my bedroom with mum saying, "Brett, hat have you done?" Then mum called the ambulance, which took me to Royal Brisbane hospital, where they found that I had broken my right femur.

The doctors initially decided that they were going to operate on Christmas Day but decided to operate on Boxing Day instead when they inserted a rod and screws to mend the break. After three weeks in at the Royal I was transferred to Redcliffe hospital for rehab. I was admitted to the orthopaedic ward while waiting for a bed to be free in the rehab ward and eventually after spending 2 weeks in the orthopaedic ward a bed because free in the rehab department.

After only 3 days in rehab, the physio and OT said that I could go home, so having missed Christmas I was pleased to be out. However roughly 7 days after I got home my right leg was getting more painful every day and it became so unbearable that I decided to take myself back to Redcliffe hospital where I had an X-ray and found out that the rod and screws had come out due to my osteoporosis.

I was re-admitted and spent just over 2 weeks in Redcliffe hospital before being transferred back to the Royal, waiting to be operated on which was sometime towards the end of February. I was given a big dose of factor (Eloctate) just before going into the theatre, but halfway through the operation the surgeons had to abort the operation as I had lost 1.5 litres of blood. The surgeons had to quickly stitch the cut up and sent me to the high dependence ward for a day before moving me back to the orthopaedic ward where I was given numerous blood transfusions.

Within days my entire body became swollen so much that the stiches come undone. So I had numerous doctors and nurses around my bed deciding what they should do to stop the blood. At this point my inhibitor level went to 23.

A few days later I went back to theatre to finish what the surgeons had started. This time they inserted a much stronger and bigger rod which was hammered into the bone and numerous screws attached. Once again after the operation I went to the high dependence ward.

A few weeks later back up on the ward, my leg was getting sorer and sorer and it turned out I had a massive infection and haematoma in my leg. After a few weeks back on the ward to get that under control I was transferred once again to Redcliffe hospital for rehab, but after 2 weeks up on the orthopaedics, I was finally given the green light to go home.

In the end I spent just over 4 months in hospital!

Improved Understaning of VWF Mechanisims

For the first time, researchers have identified the specific mechanism of von Willebrand Factor (vWF) — an essential blood clotting protein — that enables it to bind to platelets and initiate clotting.

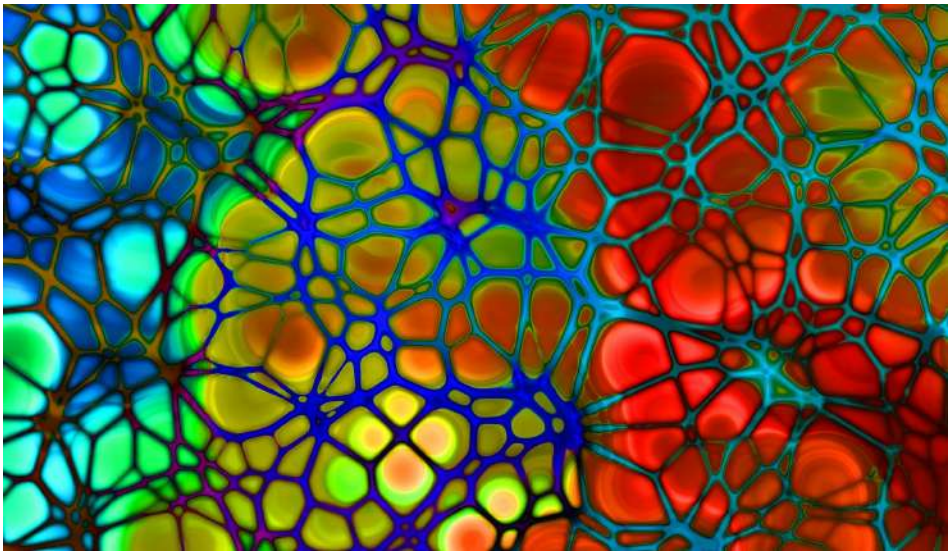
Under normal, healthy circulatory conditions, the von Willebrand Factor (vWF) keeps to itself. The large and mysterious glycoprotein moves through the blood, balled up tightly, its reaction sites unexposed. But when significant bleeding occurs, it springs into action, initiating the clotting process.

According to researchers at Lehigh University, only one drug has been FDA-approved to target vWF and treat thrombosis, or excessive blood clotting disorders, Caplacizumab. It works by binding to vWF and blocking it from binding to platelets. However, no one has understood the specific mechanism behind how it accomplishes this.

Until now. For the first time, researchers have identified the specific structural element of vWF that allows it to bind with platelets and initiate clotting. The team says that the specific unit, which they call the discontinuous auto inhibitory module, or AIM, is a prime site for new drug development. The work is described in an article published last week in Nature Communications, "Activation of von Willebrand factor via mechanical unfolding of its discontinuous auto inhibitory module."

AIM allows the vWF molecule to remain non-reactive in normal circulating blood, and activates the vWF instantly upon bleeding. The researchers identified that Caplacizumab works by binding the AIM region of vWF and enhancing the force threshold to mechanically remove vWF's auto inhibitory structures, opening up a new avenue to the development of antithrombotic drugs targeting the AIM structures."

An essential feature of vWF is that it remains non-reactive towards platelets most of the time in circulation. However, at bleeding sites, vWF can be activated almost instantly to achieve platelet adhesion and blood clot formation. In this research, the researchers identified a structural element, AIM, located around the portion of vWF, called the A1 domain, that is responsible for binding platelets.



In normal circulating blood the AIM wraps around the A1 and prevents the A1 from interacting with platelets. However, at the binding site, the blood flow pattern change leads to enough hydrodynamic force to stretch the AIM and pull it away from the A1, allowing the A1 to grab platelets to the bleeding site.

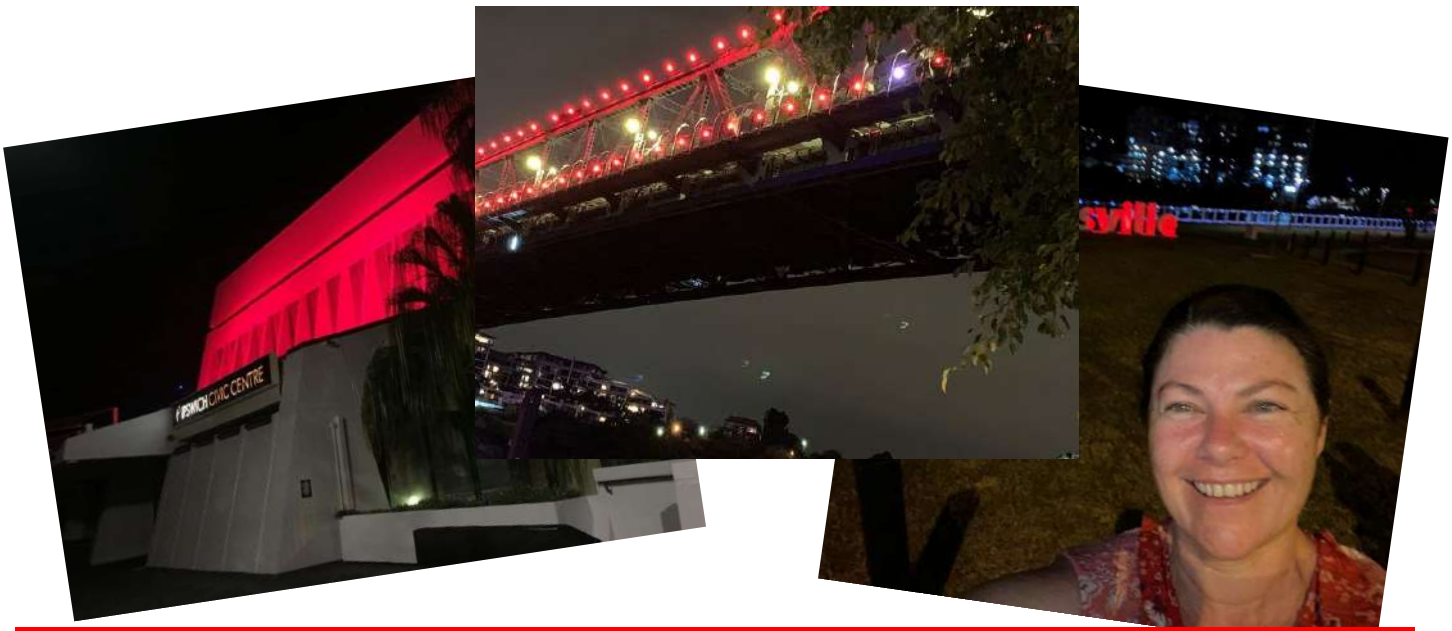
The researches use a specialised tool called optical tweezers, which utilizes a focused laser beam to apply force to objects as small as a single molecule.

These optical tweezers can grab the vWF and at the same time we apply force to see how the protein changes shape, to see how the proteins are activated when there's a mechanical perturbation or a mechanical force.

Edited for size from a paper published in Nature Communications, 2021; 12 (1) DOI: 10.1038/s41467-021-22634-x

World Haemophilia Day

Despite Prince Phillip's funeral being the same day as World Haemophilia Day, which saw many of the structures in Queenland we had booked to go red being turned red white and blue instead, we still had a lot going on in Queensland for World Haemophilia Day in 2021.



Meet Sarah

Sarah has been working as a Counsellor for over 5 years and is about to complete her Master of Social Work degree. Sarah started a social work placement with HFQ from April to June, doing a literature search and key stakeholder interviews on the needs of members for employment and career planning support. Some of you will have spoken to Sarah as she conducted this work and she has given the board a report on each part of the project. These reports support the belief that we should offer member support in this area.

Sarah was born in Germany where she also studied social work. In between studying and living in Australia, she has done a lot of travelling, for example to South America. She volunteered for the UNESCO in Argentina and worked with young people in a school setting. Also, Ishe went to India and supported people living with HIV/AIDS. In her free time, she enjoys the outdoors and walking her dog.

We hope that Sarah will come back for youth camp and continue to talk with the participants about their career and employment needs.

Sarah at the World Haemophilia Day Art Exhibition at RBWH

Health Updates

Preventive Hemlibra May Lower Hemophilia A Treatment Costs

Starting or switching to Hemlibra (emicizumab) may lead to a significant reduction in treatment-associated expenses for haemophilia A patients in the U.S., a real-world study, "Real-world cost estimates of initiating emicizumab in US patients with haemophilia A," was published in the journal Haemophilia.

"Importantly," the researchers noted, their study did not factor in key secondary savings seen by children with hemophilia A taking preventive Hemlibra.

The researchers said that consideration of only healthcare costs will also not likely capture all the economic benefits of initiating Hemlibra, such as the time saved by the increased convenience of simplified prophylaxis for people with haemophilia and their families.

The researchers reported data from a real-world study that aimed to estimate the cost of haemostatic therapies, or treatments to stop bleeding.

The team evaluated the costs of treatment in a group of 92 haemophilia A patients, ages 4 to 14, six months before and after the initiation of preventive Hemlibra.

The costs dropped significantly following the start of Hemlibra (by 27.5%). This reduction in median total costs was more evident in patients with inhibitors but once patients were on Hemlibra, there was not a significant difference in total costs of haemostatic therapy between patients with and without inhibitors.

The total costs of treating bleeds also dropped significantly after the start of Hemlibra, decreasing from a mean of $24,441 to $2,542 per patient. For that measure, the researchers considered the type of haemostatics therapy being used, along with the dose, the number of doses, and the number of bleeds taking place during the six months of the study.

The researchers concluded that starting or switching to prophylaxis with Hemlibra results in decreased costs for the treatment of patients with haemophilia A.

The researchers said this result demonstrates that switching prophylaxis to emicizumab is likely more convenient, decreases bleeds and decreases costs.
https://onlinelibrary.wiley.com/doi/abs/10.1111/hae.14347

uniQure cleared to resume hemophilia gene therapy trial

A uniQure gene therapy clinical trial that was halted late last year now has regulatory clearance to resume. The FDA has lifted the clinical hold on etranacogene dezaparvovec, the company's experimental gene therapy for hemophilia B. The hold was placed after one patient who had received the therapy two years earlier was diagnosed with a form of liver cancer.

Cancer is a known risk of gene therapies, which use an engineered virus to ferry a genetic payload into cells. If that virus integrates with a patient's DNA, it can cause cancer. Last month, uniQure reported results of an independent investigation that concluded that the patient had multiple risk factors, and gene therapy was "highly unlikely" to be the cause of the cancer.

The uniQure gene therapy uses adeno-associated virus type 5 (AAV5) to carry a functioning version of the FIX gene to patient's cells. The open-label study gave a single infusion of the one-time treatment to each participant to assess FIX activity 26 weeks after dosing. The secondary goal is to assess annualized bleeding rates at 52 weeks. Preliminary 52-week data are expected shortly.

https://www.globenewswire.com/news-release/2021/04/26/2216691/0/en/uniQure-Announces-FDA-Removes-Clinical-Hold-on-Hemophilia-B-Gene-Therapy-Program.html

1st Patient Dosed with Injectable MarzAA

The first participant has been dosed in a Phase 3 clinical trial testing marzeptacog alfa activated (MarzAA), an experimental under-the-skin therapy for haemophilia A and B patients with inhibitors.

According to Catalyst, the experimental therapy stands out from other approved bypassing agents for combining higher clot-generating activity and longer activity at the site of bleeding with a subcutaneous, or under-the-skin, route of administration.

As such, MarzAA is expected to be an effective and more convenient approach to prevent or treat bleeds in haemophilia patients with inhibitors and if successful, could fundamentally change patients' lives..

The study's main goal is to assess the proportion of effectively controlled bleeds at 24 hours after initial dosing. Pain severity, the presence of antibodies against MarzAA, the time taken to self-administer treatment, and overall satisfaction will also be assessed.

Edited for size from an article the first appeared in Hemophilia News Today https://hemophilianewstoday.com/2021/05/12/1st-patient-dosed-in-phase-3-trial-of-injectable-marzaa-hemophilia-a-b-inhibitors/

Roctavian Prevents Bleeds Over 5 Years, Data Show

Five-year data from a Phase 1/2 study shows that BioMarins gene therapy Roctavian continues to effectively and safely prevent bleeding episodes and the need for clotting factor VIII replacement therapy in adults with severe haemophilia A,

The therapy, which is administered by a single infusion directly into the bloodstream, provides a healthy copy of the F8 gene, in people with haemophilia A.

Compared to pre-treatment, the mean cumulative annualized bleed rate (ABR) in year five was 0.7 — corresponding to a 95% ABR reduction. There was also a 96% reduction in FVIII use over the course of five years in this group.

Currently, all participants remain off prophylactic FVIII treatment. While FVIII activity levels declined over the study period, they remain within an effective therapeutic range. No patients developed FVIII inhibitors or withdrew from the study.

The most common adverse treatment-related events occurred soon after the therapy's infusion and included short-lived infusion-associated reactions. Brief, asymptomatic, mild to moderate elevations in the levels of certain proteins and enzymes were also reported, with no continued effects.

Researchers said that it is promising that Factor levels continue to remain in a range to provide haemostatic efficacy for the vast majority of patients for a meaningful period of time.

https://hemophilianewstoday.com/2021/05/28/hemophilia-a-gene-therapy-roctavian-prevents-bleeds-over-5-years-data-show/?utm_source=HEM&utm_campaign=5e2bc609ac-RSS_EMAIL_CAMPAIGN_NON-US&utm_medium=email&utm_term=0_ab10fdd11a-5e2bc609ac-71884349

Women and Sports

Do you have any young women with bleeding disorders in your family?

- Ask your HTC for a copy
- Contact HFA to send you copies –
  E: *hfaust@haemophilia.org.au*
  or T: 1800 807 173

Haemophilia Foundation Australia would like to pass on their thanks to all involved in the development: the young women and their parents and the expert health professionals for their comments and advice.

Please let them know that Haemophilia Foundation Australia (HFA) have published a new fact sheet on sport and exercise for girls and young women called; **Sport and exercise for girls and young women with bleeding disorders**.

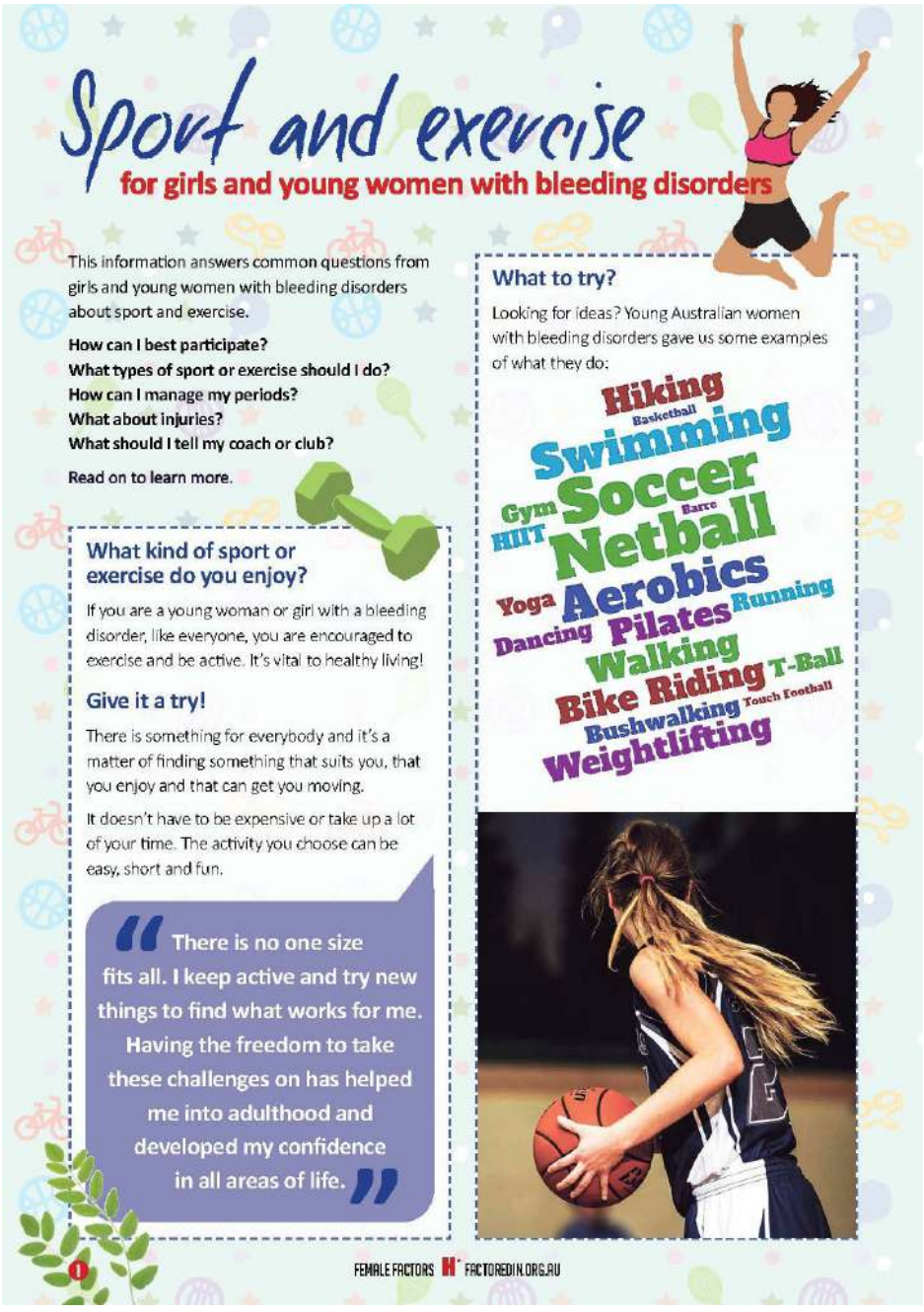
This was developed out of a survey of young women and their parents and HFA's expert health professionals did a great job of putting the answers to their questions together, such as:

- **How can I best participate?**
- **What types of sport or exercise should I do?**
- **How can I manage my periods?**
- **What about injuries?**
- **What should I tell my coach or club?**

Accessing the fact sheet

We are really pleased with the result and hope you will be too!

- You can read it online or download it from Factored In - *https://tinyurl.com/FI-sport-girls*



Joint Surgery Works Well

A RBWH study shows joint surgery works well in patients with inherited bleeding disorders.

The cyclical process of recurrent joint bleeds and synovial proliferation in persons with severe inherited bleeding disorders ultimately leads to cartilage and joint destruction manifesting as pain and dysfunction. Orthopaedic surgery remains the gold standard treatment of end-stage hip and knee arthritis.

Previous studies have reported failure rates as high as 20%. Reported infection rates are higher in patients with inherited bleeding disorders, potentially due to increased bleeding risks.

This retrospective, single-centre study was conducted to evaluate the surgical and rehabilitative outcomes of patients with inherited bleeding disorders undergoing total hip or total knee replacement at the Royal Brisbane & Women's Hospital (RBWH), over a 20-year period.

Patients undergoing joint replacement surgery during 1996-2016 were under the care of a senior orthopaedic surgeon and haemophilia specialist physician.

Patients meeting inclusion criteria underwent a comprehensive evaluation of validated function and satisfaction questionnaires for each of their affected joint replacements. Patients' satisfaction was measured with respect to pain relief, functional improvement and overall satisfaction with the operation as well as to recall retrospectively, their pain levels just prior to the operation.

A total of 36 patients (34 males; 2 females) representing 55 joints (45 TKR; 10 THR) were initially identified as meeting the inclusion criteria. A further 6 patients (8 joints) were subsequently identified through cross-referencing medical records but all failed to meet the aforementioned inclusion criteria.

Satisfactory factor levels were achieved with our standard protocols in 86% of patients with Haemophilia A, VWD and HB, respectively, on the day of surgery. The vast majority of patients were reportedly "very satisfied" with the reduction in pain as a result of having their surgery while 75% of patients rated their overall satisfaction as the highest score possible.

All patients reported very low current pain levels at the time of assessment.

Since the first TKA for PWH in the 1970s, there have been significant advances in both pharmacological and surgical interventions in this population. Improvements in factor replacement therapy have allowed for an increase in elective surgical procedures.

Within our cohort, patients reported low pain scores and high satisfaction rates with respect to pain and functional outcomes while also reporting high overall satisfaction rates with the procedure. Similar to other groups, we found that patient's (objective) post-operative function is not as favourable as those without inherited bleeding disorders; however, the overall benefits (reduction in pain and disability) still strongly support surgery as a viable procedure for end-stage hip and knee arthritis.

67% of Total Hip Replacements were rated as "good" to "excellent" and 92% of TKR had a KSS in the "good" or "excellent" range.

Reasons for the comparably less favourable functional outcomes are likely to relate to poorer premorbid functioning (due to multi-joint involvement and deconditioning), presence of comorbidities and recurrent bleeding (or perceived risk of bleeding) limiting intensive rehabilitation.

This large Australian cohort provides orthopaedic surgeons and haemophilia clinicians with valuable local data which may assist in counselling their patients regarding realistic post-operative expectations in terms of pain, function and likely overall satisfaction following arthroplasty procedures.

Haemophilic arthropathy is a debilitating condition resulting in progressive joint destruction manifesting as pain and impaired function. Arthroplasty for end-stage arthropathy is a worthwhile procedure with respect to pain and patient-perceived outcomes, despite more modest objective functional improvements compared with persons without haemophilia. These patients are medically and surgically challenging and should be managed in a tertiary haemophilia centre with multidisciplinary care teams encompassing haematology, orthopaedics, physiotherapy and social work. A large collaborative multi-centre prospective study will allow more outcome information to improve patient outcomes.

Edited for size from a paper by Dr Jane Mason et al., published in Haemophilia Journal https://onlinelibrary.wiley.com/doi/abs/10.1111/hae.13559

Important Dates for HFQ Members

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

OBE Men's Meeting
First week of the Month
Wednesday, 7th July
East Leagues Club

Women's Lunch
Saturday, 21st August
Everton Park Hotel

R U OK Day
9th September

Please ask for events and activities happening in your area.

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

8|9 October 2021

20TH AUSTRALIAN CONFERENCE

ON HAEMOPHILIA, VWD & RARE BLEEDING DISORDERS

This years 20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders is a virtual conference due to the uncertainty of the pandemic and at HFQ we are very excited about the opportunities this provides to you, our members. Without the requirement to travel many more people may be able to participate and contribute to the discussions. We expect the virtual conference will attract more delegates than usual and create innovative learning opportunities and discussion for everyone.

The October program will go over Friday and Saturday between 9am – 5pm. There will be keynote presentations for everyone, followed by concurrent sessions for you to choose from, and there will be breaks in between so you can plan your days. If you miss a session, you can playback later that day and all sessions will be as part of your registration for 6 months, you can log in anytime.

If you have ever wanted to attend a conference and couldn't because of time constraints, travel or costs, this is the perfect time to do so!

Website link *www.haemophila.org.au/conference21*

Registration link *www.haemophilia.org.au/registration*

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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