the H FACTOR

Haemophilia Foundation Queensland

SPRING EDITION

Issue 72

OCTOBER 2022 **HFQ MEMBER MAGAZINE**

FROM THE **PRESIDENT**

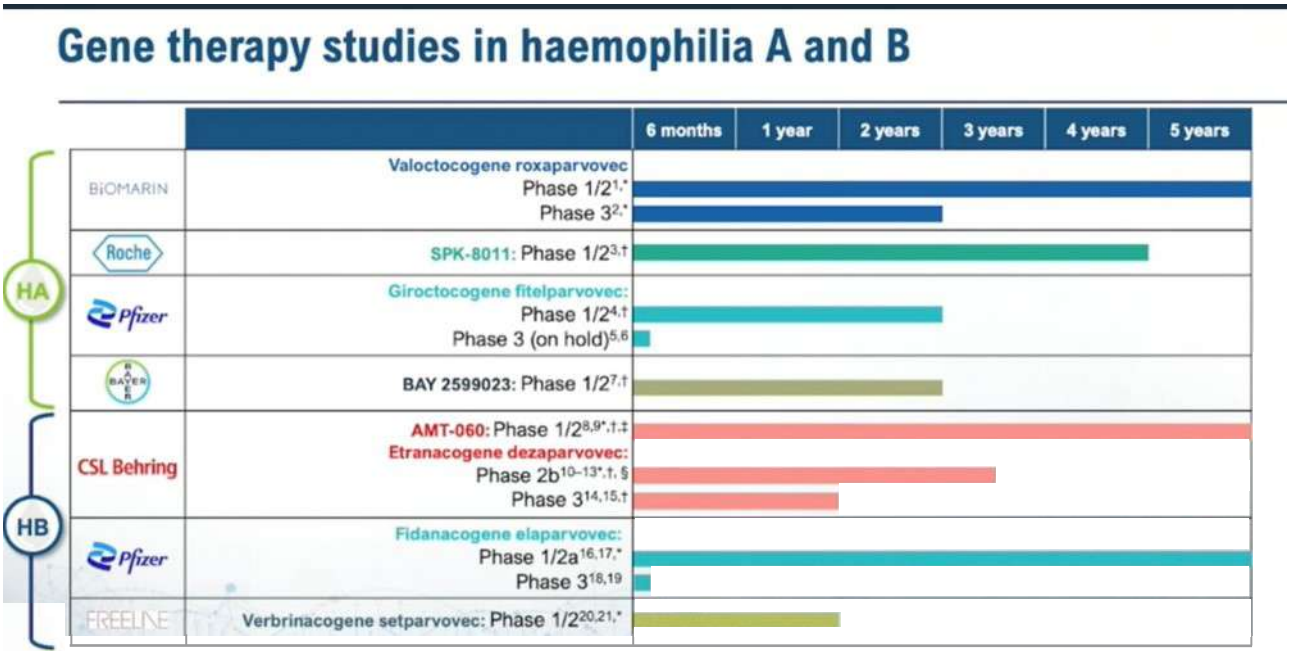
Hello everyone,

I hope this finds you all well and healthy.

As we all know, the clinical and life burden of having a bleeding condition can have a significant effect on people with bleeding disorders - relationships, career, education, income, mobility, where you live… and so on.

It can affect nearly all aspects of life for many, so it's very refreshing to continue to see the life changing treatments of today, where our younger community is experiencing better quality of life with few bleeding impact. Even our older members are significantly benefiting from extended 1/2 life and bypassing treatments – despite existing joint damage and other pre-existing complexities these treatments are improving their daily living.

Gene therapy may be next even though there are still many questions to be answered - long term safety, variability, and durability. Here is a slide from earlier this year which gives you a glimpse of gene therapy timeline progress:



So until next time….

Dave

David Stephenson
President HFQ
president@hfq.org.au

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 AC PSM Governor of Queensland.

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*

QLD HAEMOPHILIA STATE CENTRES

CHILDREN'S CLINIC

PAEDIATRIC CLINIC STAFF (QCH)

Switch: 07-3068 1111 Haemophilia Mobile 0438 792 063

Dr Simon Brown – Haematologist
Dr Nathan Morgan – Haemophilia Fellow
Joanna McCosker – Nurse Practitioner
Tamara Shannen / Salena Griffin – Clinical Nurse
Claire Bennett (Mon, Tues, Wed) - Physiotherapist
Elise Mosey (Thur, Fri) – Physiotherapist
Tiara Tan - Psychologist (Mon 1/2 day, Wed, Thurs)
Lara Nicholson—Social Worker (Mon, Tues, Wed)

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	*On Maternity Leave*
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
Liam Ball - 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

What's On?

MAKE IT YOUR EVENT

Oct to Dec 2022

Some of the HFQ programs and activities already planned

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

OCTOBER

BLEEDING DISORDERS AWARENESS MONTH OCTOBER 2022

HFQ Government House Reception; Honouring our Health Care Workers
6 October.

HFQ BBQ Fundraiser Bunnings Rothwell
8 October

OBE's Monthly meeting
Wednesday 12 October
Norths League Services Club

HFQ ANNUAL GENERAL MEETING
18 October
Nundah Library

HFQ BBQ Fundraiser Bunnings North Toowoomba
22 October

Remembrance Plaque Unveiling
29 October
RBWH Education Centre

NOV

OBE's Monthly meeting
Wednesday 2 November
Venue to be advised

Graham Norton Retires from HFQ

Women's End of Year Lunch
Sunday, 13 November
Tingalpa Hotel

HFQ Board Meeting
15 November
Via Zoom

DEC

Worlds AIDS Day
1st December

OBE's End of Year Meeting
Sunday, 11 December
Venue to be advised

HFQ Board End of Year Meeting
20 December
Via Zoom

HFQ Office Closure
23rd Dec 2022 to
2 January 2023

Dr Simon

Hi I'm Simon or Dr Simon or Dr Brown or Dr Brown Bear, I have many aliases. I'm the director of haemophilia at the Queensland Children's Hospital and I collect Minions.

We look after about 300 children with inherited bleeding disorders, of which about 70 have severe haemophilia, which is probably a similar size practice to the Royal Free Hospital UK, so it's quite a big practice and the amazing thing about Queensland is just it's size. The geography and the dispersion of patients.

What do I do?

I'm the director, but I've sort of done a bit of everything from time to time in the health system. But now I'm here to hopefully help everyone and be there to answer the medical questions and talk to the families and the children about medical issues and do those sort of things. But I'm a Jack of all trades, I hope.

How long have I been in this role as director of haemophilia?

As director of haemophilia, I think I started in 2007, it was more by accident than anything. Dr. Bill McWhirter had retired and I was floating around the system and got invited to give a talk and was asked whether I would like to look after the service, so I said yes, and I've been there since and just steering the rudder a bit, hopefully pointing our service in the right direction,

From the perspective of haemophilia, prior to coming here, my first experience with haemophilia was at St. Thomas' Hospital in London in 1992. That's when I started as a registrar in haematology, and that's when I fell in love with haemophilia and bleeding conditions. And I've been involved since then. I did my research at Cardiff Hospital with Peter Collins on von Willebrand disease and then I took a job as a consultant to the Royal Free Hospital prior to coming here.

So on and off, I've been involved with haemophilia for probably nearly 30 years.

How has treatment changed in that time?

Incredibly. I can remember at St. Thomas's, the children that were under our care were still coming in regularly with bleeds. Prophylaxis at that time in the U.K. was really in its infancy because it was just post HIV and the hep C contaminating some of the blood based products we used, so it was difficult getting adequate concentrate.

The centre at St Thomas' Hospital was involved in the early Novoseven recombinant studies as a way of trying to treat and manage inhibiters. At that time it was hugely challenging.

And of course all the centres I've ever worked prior to working here in Queensland were involved in the care of people from birth until their old age. And the thing that I think still resonates is what the community went through in terms of HIV and hepatitis C.

So, treatments have changed for the good. Since coming to Australia I've been impressed how the community as a whole, right from the foundation level, to the families, right through to the Clinical area and on to government, have worked together to try and get the best treatments. I know it's challenging and we all get frustrated at times, but there have been remarkable changes.

We're now on the cusp of a new era with non-factor treatments, like Hemlibra and gene therapy in the wings where we would be going from not just treating bleeds, but actually being able to hopefully normalise life for children that are growing up now.

Do you have a key message for children and families.

It's amazing the changes that have occurred in therapy. And I think what we would try to instil in the families is that we are moving towards better and better treatments, whether we ever get a cure with normal factor levels or a mild to moderate improvement in levels that's long lasting, it's really difficult to tell with gene therapy at the moment, but if boys can keep their joints healthy through meticulous prophylaxis, then they can enjoy themselves, participate in appropriate sports, do what they want to do at school and achieve what they want to achieve.

We've made significanrt progress over the last

50 years with haemophilia and I think really the world is your oyster. We have effective treatments for preventing bleeds and we should be able to maximise joint health.

We should be able to keep boys and girls healthy and fit and allow them to lead a normal life. My wife tells me "normal" is a setting on a washing machine. You can never talk about q normal life, because my normal life is different to your normal life and we all have challenges that we face. But, you know, go out there and achieve things.

Vale Brett Williams

(4 Sept 1976 – 30 September 2022)

Brett Williams, a long-time member and active volunteer of HFQ died on 1 October. Brett was a visible presence at many HFQ events because of his green wheelchair and a large tumour he had over one eye, caused by Neurofibromatosis.

While his death was too soon and somewhat unexpected, since December 2020 Brett had experienced a series of hospitalisations and health setbacks.

Prior to these health issues Brett found time to get married, enjoy music (even travelling overseas to meet his favourite group, Roxette), collect celebrity autographs, barrack for the Canberra Raiders and express himself through paint where he created many works of art. Brett also interviewed people for this magazine and told us parts of his own story as well as attending the OBE's support group.

HFQ extends our heartfelt condolences to his bereaved family. May his soul rest in peace.

Remembrance Spot

Until the 1950's people with haemophilia seldom lived beyond 20 years old because there was no effective treatment available. In the 1960's it was found that precipitate from blood plasma could control bleeding, if used at the time of the bleed, but some people with haemophilia still died prematurely from bleeding incidents. In the mid 80's some batches of a new treatment for haemophilia made from multiple blood donations became contaminated, as some donations harboured HIV and/or Hep C. Many treatment recipients became infected through the very blood that was saving their lives, causing them illness and death.

As a community many of our members and their extended family have lost a loved one with a bleeding disorder and we have arranged for a plaque to be installed in the RBWH remembrance garden so there is a place people can go to, where all those people from our community who have already died are acknowledged and remembered.

This plaque will be unveiled at 11:30am on Saturday 29 October 2022 at the RBWH Education Centre. Everyone is welcome to attend but must RSVP by calling the HFQ office on 0419 706 056 providing refreshments after the ceremony.

An *Update* on Gene Therapy

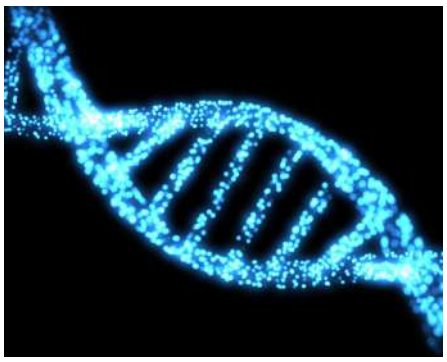
After several decades of research, gene therapy for hemophilia is moving closer to becoming a reality. This one-time treatment may potentially increase a person's factor levels to normal or close to normal, which would enable them to avoid needing factor infusions for a long period of time. Here are some answers to commonly asked questions about gene therapy for hemophilia.

How is gene therapy used to treat hemophilia?

Most of the gene therapies being investigated for the treatment of hemophilia are done using gene transfer which involves putting genetic information into a vector, or carrier, which then carries the working copy of a gene to a person's cells. Hemophilia A and B are caused by a single gene mutation. The gene therapy inserts a functional version of the defective gene into the liver, which triggers clotting factor production. The vector most used to transport the healthy gene to cells in the liver is called an adeno-associated virus (AAV), which is a non-disease-causing virus, modified as a delivery vehicle, and given intravenously. AAV is a popular choice because it especially targets the liver, which is where the clotting factor proteins are made.

What is the status of gene therapies being studied for the treatment of hemophilia?

Currently, there are several gene therapy treatments for hemophilia A and B being studied in clinical trials to determine how well they work and how safe they are.



One of the genetic therapies that is furthest along in the pipeline is valoctocogene roxaparvovec, designed for the treatment of adults with severe hemophilia A. The manufacturers expect to apply for approval to the Food and Drug Administration (FDA) this year, which would be followed by a six-month review by the FDA.

Another investigational gene therapy for hemophilia in the later stages of development is AMT-061 (etranacogene dezaparvovec), which is being studied in patients with severe to moderately severe hemophilia B. The drug manufacturers released the results of a phase 3 clinical trial, in 2021 and said they planned to submit regulatory applications for marketing approval in 2022.

Who can take part in a clinical trial for gene therapy?

In general, individuals who are eligible to participate in gene therapy clinical trials for hemophilia are males 18 and older who:

- Have severe hemophilia (factor levels of less than 1%) without inhibitors
- Have no detectable neutralizing antibodies against the vector
- Have no evidence of hepatitis and if they have HIV, it must be under good control
- Have no evidence of liver dysfunction (such as abnormal liver enzymes or abnormal liver biopsy)
- Are otherwise healthy (no diabetes, history of cancer, heart disease, glaucoma, high blood pressure, etc.)

What are the risks involved with gene therapy for hemophilia?

One of the biggest risks is that there's no guarantee that it will work, and if it doesn't work, you won't get another chance using the same or similar vector. In addition, it isn't clear how long the effects of the gene therapy may last.

Other risks include a strong immune system reaction, the possibility that the vector would travel to other cells in the body outside the liver, and the possibility that the liver would produce too much clotting protein.

If you have concerns about gene Therapies or you are interested in clinical trials for them, or undergoing genetic therapy or you should talk to your Queensland haemophilia Centre staff.

First published by Hemaware in February 2022 in an article called "Answers to common questions about this potentially game-changing treatment" by Donna Behen https://hemaware.org/research-treatment/gene-therapy-for-hemophilia-update

Raising Awareness

Learning, Sharing and Fundraising

At HFQ we have many sources of information that may be helpful to you, your family, health professionals, and the broader community. This magazine for Queensland members and the HFA National Haemophilia Journal are (or can be) delivered to all members and associates of HFQ, please let us know if you are not receiving both magazines via the post or email.

We also have information and resources on our website (www.hfq.org.au); and we post updates to our facebook page (www.facebook.com/HFQLD). We also have an Instagram account (HFQld).

But we have many more resources that you may not be aware of. Among the things we offer is access to the journal; Haemophilia. This is the official journal of the World Federation of Hemophilia and dedicated to the exchange of information regarding the comprehensive care of bleeding disorders. It has peer reviewed scientific papers and research articles you can access through us by joining the contents page distribution list or asking us to research topics of interest.

We also have a library of resources containing books & DVD's for you and your family to better explain aspects of living with a bleeding disorder. They range from cartoon explanations of bleeding disorders for children through to books on managing inhibitors and/or haemophilia as well as autobiographies from people living with bleeding disorders. We also maintain a list of website and on-line resources that can help you better understand or explain your own circumstances. If you'd like to access any of these, please let the office know and we can post them out to you on a loan or keep basis depending on the resources required.

Of course, being informed is important to you if you are managing a bleeding disorder, but so to is getting the public on-side and helping them understand what living with a bleeding disorder is like and we have a great brochure called 'About bleeding disorders' that can be given away to family and friends and explains in simple terms, what a bleeding disorder is. This is a great resource to use at any time when talking to people who don't understand, but it can be especially helpful at times when we are trying to raise public awareness such as World Haemophilia Day (17 April) and Bleeding Disorders Awareness Month (every October in Australia).

Bleeding Disorders Awareness Month originally started as Haemophilia Awareness Week, but it became a month to make it easier for members and supporters to utilise the opportunity to raise awareness during the extended period of a month. It was also rebadged some time ago because we want everyone living with a bleeding disorder to be included and we recognise that HFQ has many members with von Willebrand Disease and several other rarer bleeding disorders. It is vital that the voices of all our members are heard and that the general community know that haemophilia is just two of the many bleeding disorders we represent and support.

If you would like to hold a fundraising event or awareness stall during one of these awareness times (or at any other time) we also have posters, balloons, colouring competitions, branded pens, shopping bags and USB sticks that can be given away for a suitable donation etc.

We also have polo shirts and t- shirts that you can wear to show your support. These are available in assorted sizes and are given away at cost or free depending on your activity.

It takes Guts to Eat Healthily

A lot more goes on in your gut than you might realise. Trillions of microorganisms call your gut home and it's sometimes called the microbiome and when they are in balance, they help keep you healthy.

If the balance is good, you'll have fewer infections and illnesses, and our immune systems will be optimal. When it's not in balance, it can affect everything, from your heart to your mental health.

Start with Your Diet

What you eat plays the biggest part in gut health. Researchers report that a mostly plant-based diet of fiber-rich fruits and vegetables, fish, nuts and seeds supports gut microorganisms that may lower the risk of chronic disease.

Where-as poor diets featuring dairy-based desserts and processed foods, including high-fat meats promoted gut microorganisms linked with high cholesterol, high blood sugar and inflammation. These combine to increase a person's risk of diabetes, heart disease and other chronic diseases. Other research ties gut inflammation to depression.

Adjust to Address Chronic Illness

The influence of gut health on bleeding disorders is not well known, but we do know that one form of vitamin K, known for its role in blood clotting, is produced by gut bacteria. Anyone with a chronic disease, including hemophilia, will benefit from a gut-healthy diet.

You want to eat a variety of foods and nutrients to keep your gut in balance and maintain the diversity of its microorganisms. Eat fiber-rich foods and limited amounts of processed foods.

Many common diets are very focused and can have harmful effects. For example the high-protein meat diet used by body builders can raise peoples bad cholesterol and their risk of fatty liver disease. You are much better off with a more diverse diet of greens, fibre and plant-based protein which are beneficial for your gut.

Consider Probiotics

The gut microbiome also can be maintained with probiotics. These are good bacteria that help balance the disease-causing ones. Probiotics can be found in yoghurt—make sure the label says it contains live and active cultures—as well as fermented foods, such as sauerkraut, kimchi and kombucha.

Probiotics also come in supplement form, but more research is needed to determine their effectiveness and the best strains for the desired clinical outcomes.

For a good article on how to improve your gut health go to https://vichealth.vic.gov.au%2Fbe-healthy%2Fhow-to-improve-your-gut-health&usg=AOvVaw29kxmYuhKYR-LCA5SjOLPi

The information in this article is provided for general information purposes only. Neither HFQ nor the editor of H Factor engage in the practice of medicine or dietetics and under no circumstances recommends and particular approach for specific individuals. For diagnosis or consultation on a specific medical problem, HFQ recommends that you contact your Haemophilia Doctor or Nurse or your local treatment centre in the first instance.

Farewell from Graham

I wanted to let you know that I intend to step down as manager at the end of October and after an extensive recruitment and interview process Lauren Green will succeed me as manager. Sam is staying as Administration and Support Officer and David Stephenson has indicated he will stand again as President, so Lauren will be ably supported by Sam, David and the HFQ board.

Many of you will know Lauren, perhaps by her maiden name Albert as she's been part of the HFQ community since her parents where active on the board and I'm sure she'll introduce herself to you in the next issue of H Factor.

After nine years, it's hard for me to imagine leaving, and in many ways I don't want to retire from HFQ, but it's the right time for me. I leave to have more time for myself and because it seems a good time for the bleeding disorders family. We have new treatments like hemlibra available and more to come in the pipeline. We have a new psychologist and social worker supporting the children's haemophilia clinic and I'm told a replacement for Loretta the adult social worker is not far away either.

HFQ has always worked closely with the treatment centre staff especially the psychosocial staff so this is a good time to hand over to new people who can forge new beginnings in a time of new treatment possibilities. However I reflect back on my time and in that period we had 4 adults (who I came to regard as friends as well as members) and one child, die from conditions that had everything to do with their bleeding disorder and nothing to do with natural ageing. They will be forever in my heart as will the younger families, young adults, couples and individuals I have been privileged to meet.

You may want to ask me what I will miss most about HFQ and I've already hinted at the answer: all of you. The people of the bleeding disorders community have always been our most differentiating strength as a patient advocacy and support group. In past tough times (there's been a crisis or two), you got together to support each other and advocate for solutions and change. In my time as manager we have had better times and I have fed off your excitement and willingness to embrace the future.

I have been honoured to serve you as manager but always knew this day would come. It may be hard for some of you to believe, but I'm told that sometimes I'm not the easiest person to live with. I could not have gone through the ups and downs of the last 9 years without the patience and constant support of David and the HFQ board, Sam my office partner in crime, and Rob my partner in life.

Going forward I hope to devote more time to my family and to pursue my own passions and interests, but I will never do anything that will be as satisfying, nor as much a part of me as working for the bleeding disorders community of Queensland.

NINE YEARS IS L O N G ENOUGH

GRAHAM IS RETIREING FROM HFQ

In a time of new treatments It's time for new blood and new ideas

6 Facts about Women & Haemophilia

1. Women with haemophilia are often undiagnosed or misdiagnosed

Historically haemophilia was thought of as a "man's disease," and for many years the belief was that women could only be carriers, passing the affected gene on to their children. But we now know that some women with the haemophilia gene have low enough factor levels that they also have haemophilia, and that women with haemophilia will often experience similar symptoms and complications as men with haemophilia.. A lack of awareness on the part of some doctors means that many women still go undiagnosed or misdiagnosed.

2. Women are more likely to have mild haemophilia

According to a 2021 USA study nearly one-fifth of patients with mild haemophilia admitted to treatment centres in the US are female. The percentage of women and girls with severe or moderate haemophilia is thought to be somewhere in the range of less than 0.5% to a little more than 1%.

A much more common bleeding disorder in women is von Willebrand disease (VWD), which affects more than 2,300 Australians. About 1 percent of the population, or approximately 1 in every 100 people are believed to have VWD. This inherited bleeding disorder occurs equally in men and women.

3. One of the most common signs of haemophilia in women is heavy menstrual bleeding

Heavy menstrual periods are defined as:

- Bleeding for more than seven days, from the time it began until it stopped
- Flooding or gushing of blood
- Passing clots that are bigger than a $1 coin
- Changing a tampon and/or pad every two hours or less on the heaviest day

Other common signs and symptoms of haemophilia in women include:

- Being told you are "low in iron" or have anaemia
- Heavy bleeding from dental surgery, other surgery or childbirth
- Frequent nosebleeds that last longer than 10 minutes
- Bleeding from cuts lasting longer than five minutes
- Easy bruising (weekly, raised and larger than a quarter)

4. Making the diagnosis can be complicated

There's no single diagnostic test that can confirm if a woman has haemophilia or another type of bleeding disorder, so providers often have to piece together various symptoms, multiple lab results and timing of the lab testing to confirm the diagnosis. One of the challenges for providers is to determine whether heavy periods are related to normal variation or a sign of an underlying bleeding disorder.

5. Pregnant women with haemophilia require special care

Pregnant women with haemophilia are at an increased risk for serious bleeding after delivery because the high levels of factor during pregnancy can fall back to lower levels after delivery. That's why women with a bleeding disorder sometimes need clotting factors or another treatment to prevent excessive bleeding at the time of delivery. All pregnant women with haemophilia should talk to their doctor about appropriate care during their pregnancy.

6. There are supports for women and girls with bleeding disorders

Women and girls with haemophilia can seek care at one of the Queensland Haemophilia Centres who have the expertise to diagnose and treat haemophilia and other bleeding disorders. The QCH Haemophilia centre offers a designated clinic to diagnose and treat girls with bleeding disorders.

To learn more about women and haemophilia, check out the HFA resources on our website at: https://www.hfq.org.au/about-bleeding-disorders/women-with-bleeding-disorders

Bleeding Disorders and Pop Culture

Having a rare disease like haemophilia means you don't often get to see people who share your disease portrayed in movies, TV shows and other popular media. But over the last 20 years, the arts and entertainment industry has featured characters with haemophilia and other bleeding disorders—albeit not always accurately. Here's a roundup of the films, TV shows and plays that we know of that have featured haemophilia or another bleeding disorder in their storylines.

Old. In this 2021 movie by director M. Night Shyamalan, based on the 2010 graphic novel *Sandcastle*, traveler's on a tropical holiday discover that the beach they're visiting is causing them to age rapidly. Aaron Pierre plays an aspiring rapper named Mid-Sized Sedan who has haemophilia.

Rox and Ray. This two-person play, which features a paediatric haematologist and a single father whose twin sons have haemophilia, chronicles the early years of the HIV/AIDS epidemic, when thousands of people with haemophilia died after receiving contaminated factor infusions.

The play premiered in Seattle in 2016 and playwright Karen Hartman says she was inspired to write the play in part because her late father, Gary L. Hartman, MD, was a paediatric haematologist/oncologist in San Diego during the early days of the AIDS crisis.

Royal Pains, was a TV show that aired in the USA from 2009 to 2016, is set in the Hamptons on Long Island, New York, and features a concierge doctor who makes house calls to the rich and famous. Ezra Miller plays the recurring character Tucker Bryant, a teenager with haemophilia. Inspiration for the character came from Enrique Lenchewski, DDS, the father of *Royal Pains* creator and co-executive producer Andrew Lenchewski.

Grey's Anatomy. In 2005 episode 8, of *Grey's Anatomy* (season 1), entitled "Save Me," featured an Orthodox Jewish patient named Devo Friedman who needed a heart valve replacement. She is ineligible for a mechanical heart valve because she has von Willebrand's disease, and her religious beliefs prevent her from accepting a valve from a pig.

Law & Order, Criminal Intent. In 2003 episode 14 of *Law and Order: Criminal Intent (series 3),* entitled "Mis-Labeled," followed the murder investigation of a pharmaceutical sales rep and uncovers a scandal involving clotting factor that's been contaminated with HIV.

The Doe Boy. In this 2001 independent coming-of-age film, James Duval plays Hunter, a half-Cherokee, half-Caucasian boy who has haemophilia.

Older People with Bleeding Disorders and Osteoporosis

Osteoporosis, a disease of low bone mass and strength, is a common concern in aging individuals. People with haemophilia have a higher risk of the disease, compared with other groups. Other bleeding disorders, while less studied, also seem to affect bone health. The reason is not entirely understood. It is likely that there are multiple factors at play at different times.

In male mice, factor VIII deficiency alone leads to abnormal bone formation. At later ages, joint bleeding contributes to increased bone breakdown. Defects in bone metabolism have also been seen in haemophilia B mice.

Possible Causes

In people with haemophilia, the risk factors strongly associated with osteoporosis include: Advanced joint disease, HIV, Low body mass index, and Tobacco use.

As for how factor VIII or factor IX deficiency impacts bone, much of the existing research is conflicting. Studies are ongoing.

One factor commonly thought to cause osteoporosis in people with bleeding disorders is lack of physical activity. There is a common assumption that people with haemophilia develop osteoporosis because they aren't as physically active as others. But physical activity is not as big a contributor as many think, Being active is good, but [being inactive is] not why people get osteoporosis.

How Osteoporosis Is Managed

Osteoporosis by itself doesn't cause pain or disability, but it can lead to dangerous fractures from relatively minor trauma or falls. Screening is key, if you can identify osteoporosis early you can begin treatment that can lessen fracture risk.

Common, treatments include medications that prevent bone breakdown or encourage bone formation. It is also good to check your levels of vitamin D, calcium, testosterone and parathyroid hormones, and replace with supplements if necessary.

Staying Active with Osteoporosis

Physical activity is good for everyone, and people with osteoporosis are no exception. Experts recommend two and a half hours of moderate physical activity per week. However, joint disease can make it difficult to find exercises that aren't painful.

To lessen the risk of falls (and bone fractures) focus on activities that improve balance, strength and flexibility such as;

- Stretching
- Walking
- Swimming
- Stair climbing
- Weightlifting (free weights, weight machines or floor exercises using your body weight)
- Gardening
- Tai chi

Edited for size from an article "What Older People with Bleeding Disorders Need to Know About Osteoporosis " by Kathryn Anne Stewart that first appeared in Hemaware on 2 May 2022 https:// hemaware.org/mind-body/what-older-people-bleeding-disorders-need-know-about-osteoporosis

5 Ways to Injury-Proof Your Home

The last thing you want to think about is having your child get injured. But there are some hazards in your home, and accidents do happen. More than 5.8 million Australian injuries that happened at home required medical attention, and over 150,000 children go to the emergency room each year because of injuries that happen at home.

And for those in the bleeding disorders community, common injuries can be more severe. Plus, symptoms of haemophilia, such as damaged joints, may increase the risk of falls.

By and large, these accidents around the home are preventable. With the right precautions, you can reduce the risk of falls, poisonings, burns, cuts and other injuries that occur at home.

Eliminate Common Tripping and Fall Hazards at Home

Both Children and older people can be a bit unstable on their feet, so now is the time to remove, replace, or fix these items, which can be a trip hazard including:

- Raised doorway thresholds
- Electrical cords across the floor
- Loose throw rugs
- Loose floorboards
- Cracks in your driveway
- Clutter on the floor, such as toys or boxes
- Poor lighting (replace with brighter bulbs)
- Slippery flooring, such as bathroom tiles (use an anti-slip sealer)

Use Safer Appliances

Common appliances that produce heat, such as ovens, toasters and coffee makers, pose fire and burn risks to unsuspecting or forgetful humans. Plus, children can get shocked by open electrical outlets if they place a metal object, such as a fork, inside them.

To mitigate these burn and electrocution risks, you can install childproof versions of these appliances and your electrical outlets. For example, does your child fiddle with the stove knobs, or do your elderly relatives forget to turn off the hot plates? Get a set of stove knob covers that keep children from turning them but can be replaced when it's time to cook or get an induction hob which stays cold even when on.

Kid specific issues: Sharp Edges and Objects

Some pieces of furniture have sharp edges that can pose a particular risk to children, as kids are always on the move and small enough to run into furniture head-on. Edge and corner guards are easily available on-line and at stores such as K-mart.

If you have sharp objects in your home (and who doesn't?), keep them in a safe and secure place, such as a locked cabinet or buy child safety locks from you local hardware store. Children are naturally curious and could get hold of things like razors, scissors and kitchen knives if left out.

Use Safety Equipment

Toddlers and young children run the risk of falling out of highchairs and changing tables. When using items like these, always fasten your children securely in place to avoid falls.

Keep Household Poisons Out of Reach

Lock any cabinets, closets or storage areas that house substances that are poisonous when consumed, such as cleaning supplies, medicine and detergent.

Edited for size and Australian content from "5 Ways to Injury-Proof Your Home for Your Kids" by Michael Hickey that first appeared in hemaware on 13 August 2021
https://hemaware.org/life/5-ways-injury-proof-your-home-your-kids

Know if you're depressed and do something about it

Signs of deteriorating mental health can be difficult to see. We all have mood changes over time and it can be easy to dismiss as normal, symptoms that might be signs of mental ill health. For example; not eating as much as normal, might be a temporary change, or it might be triggered by poor mental health.

Making an honest self-appraisal, or asking people you trust if they have noticed any changes in you, can help you figure out if these symptoms are temporary or are something that deviates from your regular behaviour patterns.

If you are experiencing any of the following and they are not typical for you, it could be a sign of depression or anxiety:
Apathy: Have you lost interest in activities that used to bring you joy? Has life lost so much meaning that you feel empty?
Helplessness or hopelessness: Do you feel there is little you can do to improve your life? Do you lack motivation for making change?
Changes in habits: Are you sleeping or eating too much or too little? Or are you engaging in high-risk behaviours, such as excessive drinking, drug taking, unsafe sex or cutting.
Persistent fatigue: Do you feel not just tired at the end of the day. But all the time?
Mood swings: Are you easily irritated, extremely impatient or overly self-critical, or if you experience frequent mood swings?
Unending worry: Are you constantly thinking about problems and solutions, and is it affecting your ability to enjoy life?
Wanting to be alone: This is different from enjoying time by yourself. Does it take more energy than usual to interact with others, to the point where you would rather stay home?

Having a couple of these symptoms does not always mean you have depression or anxiety. There may be other causes contributing to these symptoms, or this may be a temporary change in mood that could get better with support.

The Queensland Haemophilia Treatment Centres have psycho/social staff you can talk to. They can help you work through any concerns you might have and provide you with counselling and support, or teach coping skills or new ways to think about your current stressors.

Help for depression and anxiety can come in many forms. Symptoms could be related to underlying medical conditions that can cause excess sadness or worry, so an evaluation from your GP could be helpful.

Creating a plan for support that works for you can feel overwhelming. Your HTC social worker can help you develop a process that works for you.

You don't have to go with the first doctor or therapist you meet, it's better to take time to find a doctor, counsellor, therapist or support group that you want to work with. It's OK to try a few options until you find the right fit.

Both Beyond Blue (*www.beyondblue.org.au*) and the Black Dog Institute (*www.blackdoginstitute.org.au*) have good online tool s and let you search for therapists and support groups in your local area as well as other resources.

There are plenty of other ways to boost mood and help curb anxiety that you can try now. These include:

- Exercising regularly—even getting out for a walk around the block can be mood-boosting
- Reaching out to your existing support people
- Journaling
- Using mindfulness or meditation techniques
- Getting adequate sleep
- Trying something new
- Talking things through with a person you trust

We are living in unprecedented times. Prolonged stress experienced by living through the covid pandemic, can seriously affecting mental and physical health. Please know you are not alone, and your QHC staff can help with mental health concerns.

Adapted from "How Do You Know if You're Depressed and What Can You Do About It?" First published in Hemaware June 2021. https://hemaware.org/community-pulse/ask-social-worker-how-do-you-know-if-youre-depressed-and-what-can-you-do-about-it

How Dangerous Is Cutting for someone with a bleeding disorder?

Discovering that a student, friend or someone else you care about self-harms themselves can be extremely upsetting and difficult to understand. You may wonder what you can do to help, or if you should keep their behaviour a secret.

It is a common misconception that self-harming behaviours such as cutting are a suicide attempt; usually, the motivation behind these behaviours is to release painful underlying emotions. While self-injury may bring a momentary sense of peace or release, it is usually followed by guilt, shame and a return to emotional pain. Although suicide may not be the person's intention, with self-harm comes the risk of more serious, even fatal, consequences.

What Is Self-Harm?

Self-harm (or self-injury) means hurting yourself on purpose. One common method is cutting with a sharp object. Hurting yourself - or thinking about hurting yourself - is a sign of emotional distress.

Reasons for Self-Harming

While each person's reason for self-injury is different, generally it is to:

- Signal depression to others
- Cope with psychological pain
- Attempt to feel something when feeling emotionally empty or numb
- Gain a sense of control
- Manage emotions of loneliness, panic, anger or confusion
- Punish for perceived faults
- Process or distract from negative feelings
- Express embarrassing emotions

How Dangerous Is Cutting for Someone with a Bleeding Disorder?

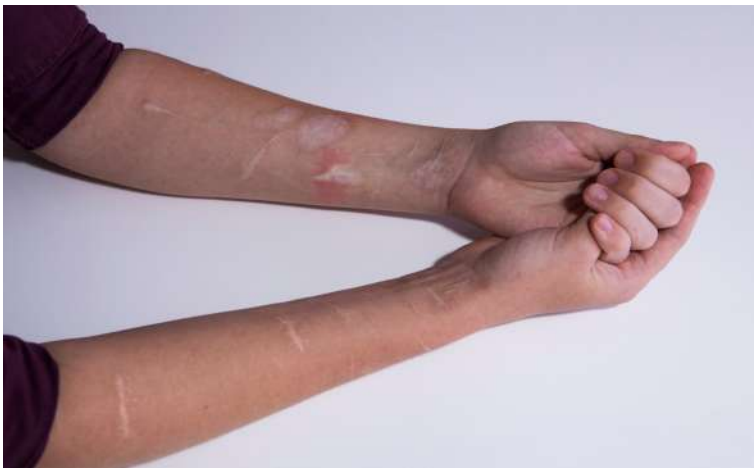
From a haematology perspective, the danger depends on the severity of self-injurious behaviours and the person's diagnosis (such as von Willebrand disease or haemophilia). The risks for people with bleeding disorders include infection, increased oozing, bleeding and bruising. A potential emergency room evaluation for stitches may be necessary.

Depending on the location of the cut, the person may have increased scar tissue around a vein, making it more difficult and painful to access in the future.

Ways You Can Help Someone Who Self-Harms

Ask them how they are doing. Be prepared to listen to the answer, even if it makes you uncomfortable. This may be a hard subject to understand. One of the best things is to tell the person that although you may not fully understand, you will be there to help. Do not dismiss emotions or try to turn it into a joke. Encourage appropriate professional help. Because self-injury is a symptom of an underlying issue, it is important to support the person in finding the appropriate help.

If you or someone you know is struggling with self -harm, reach out to a social worker or physician at your haemophilia treatment centre or Beyond Blue (*www.beyondblue.org.au*) and the Black Dog Institute (*www.blackdoginstitute.org.au*)



Health Updates

Just One Bleed Can Have A Negative Impact

In a letter to the editor, published in the jHaemophilia journal researchers found that the occurrence and severity of bleeding episodes in patients with severe haemophilia A have profound effects on health-related quality of life (HRQoL) and both drug-related and non-drug-related direct costs.

Importantly, according to researchers, the number of bleeds rather than their severity contributed more to a worse HRQoL. This link was stronger for the domains relating to pain/discomfort and anxiety/depression than it was for those pertaining to mobility, self-care, and usual activities.
https://onlinelibrary.wiley.com/doi/10.1111/hae.14616

Positive Early Results of Mim8 Pave Way for Phase 3 Trials

Mim8, an investigational next-generation therapy to prevent bleeding episodes in people with haemophilia and administered as an under-the-skin injection, is safe and well-tolerated at multiple doses, according to data from the Phase 1/2 FRONTIER1 trial presented at this years International Society of Thrombosis and Haemostats Annual Congress, held July 9–13, in London,

The therapy also showed signs of efficacy, with most patients given higher doses experiencing no bleeds during treatment. The company said Mim8 demonstrated clinical proof-of-concept and no safety signals or signs of blood clotting were seen, which supports the further clinical development of Mim8 THROUGH phase 3 clinical trials.
https://www.eventscribe.net/2022/program/fsPopup.asp?Mode=presInfo&PresentationID=1079947

Concizumab Reduces Bleeding in Haemophilia Patients

Concizumab is a once-daily experimental antibody-based therapy designed to block a protein called tissue factor pathway inhibitor (TFPI) that normally halts blood clotting. By doing so, concizumab promotes the production of thrombin, a protein that enhances blood clotting and helps prevent bleeds and has the potential to become a new treatment option for people living with haemophilia A or B with inhibitors.

New data from the Phase 3 explorer7 trial was presented at the International Society of Thrombosis and Haemostasis Annual Congress earlier this year showed that concizumab significantly reduced bleeding rates in people with both haemophilia A and B with inhibitors. In fact, bleeding rates in patients receiving concizumab in trial decreased by more than 85%.
https://www.eventscribe.net/2022/program/fsPopup.asp?PresentationID=1098884&query=concizumab&mode=presinfo

Factor IX Levels Normal After Gene Therapy for Haemophilia B

Interim data from the first three participants in the B-LIEVE trial of FLT180a, an experimental gene therapy for haemophilia B was presented in a poster presentation at the International Society on Thrombosis and Haemostats (ISTH) Congress, all participants had factor IX (FIX) levels within the normal range more than one month after being treated with FLT180a. The B-LIEVE trial was designed to confirm dosage rates ahead of a planned Phase 3 trial and is still enrolling men between 18-65.
https://www.freeline.life/investors/newsroom/positive-initial-clinical-data-from-the-b-lieve-dose-confirmation-trial-for-flt180a-in-hemophilia-b-presented-at-the-international-society-on-thrombosis-and-haemostasis-congress/

Hepatitis C Raises Liver Cancer Risk in Men With Haemophilia

Hepatitis C virus (HCV) infection remains a major risk factor for hepatocellular carcinoma, the most common type of liver cancer, in men with haemophilia, a U.S. database analysis has found.

In a study published in the journal Haemophilia researchers found that the independent risk factors for hepatocellular carcinoma among men with haemophilia included having the liver condition non-alcoholic steatohepatitis, and having a greater number of co-existing health conditions, or comorbidities.
https://onlinelibrary.wiley.com/doi/10.1111/hae.14607

Hyaluronic Acid Eases Joint Damage of Haemophilia

Adults with haemophilia and ankle damage taking part in a UK pilot study published in the journal Haemophilia reported that two injections of hyaluronic acid (HA), which acts to lubricate the joint, mostly eased ankle pain and improved movement as well as enabling most of the participants to better participate in physical and social activities.

Over the following 12 months, there was a progressive and statistically significant drop in pain scores. From a mean VAS score of 5.6 at the start of the trial to 3.4. Ankle joint function also progressively improved over the course of that year.
https://onlinelibrary.wiley.com/doi/abs/10.1111/hae.14639

Activity of Inflammatory Genes Increased in Patients with Inhibitors

Haemophilia A patients who develop inhibitors have more expression of genes involved in activating the immune system, a small study from China published in Research and Practice in Thrombosis and Haemostasis reported

The results of the study reveal that there is an upregulation of genes involved with activation of the immune system in haemophilia A patients with inhibitors and the findings have the potential to reveal novel therapeutic targets for prevention and treatment of inhibitors.
https://onlinelibrary.wiley.com/doi/10.1002/rth2.12794

Marstacimab Reduces Patients' Bleeds, Even With Inhibitors

Treatment with marstacimab, an experimental antibody-based therapy that binds to and blocks the tissue factor pathway inhibitor (TFPI), reduced the number of bleeds in patients with severe haemophilia regardless of whether patients had hemophilia A or B, or had inhibitors. These results were reported in the British Journal of Haematology.

Two patients experienced side effects that were sufficiently sever that they left the study (one had high blood pressure, the other had a large a drop in their levels of fibrinogen).

Blood tests revealed both TFPI and peak (highest-level) thrombin increased with the treatment, suggesting effective targeting of TFPI which could make Marstacimab a possible alternative to replacement therapy..
https://onlinelibrary.wiley.com/doi/10.1111/bjh.18420

Important Dates for HFQ Members

OBE Men's Meeting
Wednesday, 9th October
North's Leagues Club

Bleeding Disorders Awareness Month
October 2022

Bunnings Rothwell BBQ
Saturday, 8th October

HFQ AGM
Tuesday, 18th October 2022
Nundah Library meeting room

Bunnings Toowoomba North BBQ
Saturday, 22nd October

Remembrance Plaque Unveiling
RBWH Education centre
29 October 2022

OBE Men's Meeting
Wednesday 2 November
Venue tba

Women's Lunch
Sunday, 13th November
Tingalpa Hotel

*Please call HFQ on **0419 706 056** for more info on any of these events and other activities.*

Welcome to Paradise

For our next community camp we've gone up-market. We've booked accommodation at the BIG4 Sandstone Point Holiday Resort. Located halfway between the Sunshine Coast and Brisbane, a stone's throw from the iconic Bribie Island, there is plenty to do with magnificent facilities, and activities planned. Your individual accommodation is air-conditioned with full kitchens and TV's etc. Registrations now open for only $130 per family, $90 for couples and $50 for singles. Call the HFQ office or go on-line at: *www.hfq.org.au/get-involved/events/camp*

About The H' Factor

The 'H' Factor is published four times each year by HFQ by the HFQ manager. 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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