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The H Word

Welcome to the second edition of Bloodline for 2024. I think the word for this month has to be 'connect'. Why, you ask? At the last board meeting we approved the budget for 2024/25 and we can really see a strong interest from members to connect. The Southern and Central camps are looking to be massive, the Women's Wellness Weekend has strong attendance, next year's family camp planning is well underway, and requests to run events and make those connections keep coming thick and fast. This, of course, is core to what HNZ is here for: care, education, advocacy, and support for all people with inherited (and acquired) bleeding disorders and their whānau.

In this month's news, I share with you my overall impressions of WFH World Congress 2024 and the WFH Global Forum. I was far from the only attendee, so you will be hearing more from others in this edition and subsequently. We will also be coming to a camp or event near you to share what we all learned and gained from attendance.

Our next board meeting is September. The board meeting is not closed in any way, and so if you have an item you would like to bring, please get in touch and we can include it on the next agenda.

I hope you are all keeping as warm as you can and staying healthy over the winter months.

Deon.

WFH 2024 World Congress

The World Federation of Hemophilia 2024 World Congress took place in Spain from 21 - 24 April. HNZ sent five members and staff to attend this fantastic event. This was an important opportunity for our people to learn and share on the world stage. The HNZ group attended as many sessions as they could, and have been hard at work since putting together their reports. You can read the first of these here, and more in the next issue of Bloodline.

Deon York: Chair and Acting CE

This is the first congress in fourteen years I have attended with the support of Haemophilia New Zealand, so I would like to start right up front by thanking HNZ for this amazing opportunity. We had a strong group attending from New Zealand this year. Karl Archibald (Vice Chair), Tineke Maoate (Midland delegate), Connor McCone (youth delegate), and Darian Smith (Northern outreach) attended on behalf of HNZ. A couple of partners were added to the mix and self-funded. Lauren Phillips was invited to speak about intimacy and bleeding disorders for the benefit of all attending. Unsurprisingly, she did a stellar job. Joining us were physiotherapists from all haemophilia treatment centres as well as the wonderful nurses. There was even a haematologist thrown in for good measure, and it was wonderful to spend time with Dr Julia Phillips. Thank you to Roche for the in-kind accommodation contribution and support for the clinicians to attend. This support made this larger attendance possible.

General assembly

For previous conference-goer interest, the General Assembly of the WFH now takes place before the congress starts. This year we had Tineke and Connor) representing us. The General Assembly is the opportunity for all national member organisations (NMO) to hear about the progress of the WFH, vote for the incoming board and future congress location as well as welcoming new members to the WFH family of organisations. The total number of NMOs now sits at 152. I am happy to advise that Ekawat Suwantaroj from Thailand was elected for a four-year term. New Zealand is part of the Asia and Western Pacific region, and so Ekawat is 'our person' around the WFH board table. Congratulations, Ekawat!

NMO training

Karl Archibald attended pre-congress training on our behalf. I was there to facilitate a session on strategic planning. I can see that there has been a large turnover of volunteers globally since the pandemic, and it is encouraging to see a new generation of leaders representing the various inherited bleeding disorder organisations.

Opening ceremony

At the opening ceremony I received the International Frank Schnabel Volunteer Award, and it was a complete surprise! The WFH staff found a way to get me close to the stage, and yet I was still none the wiser. As the WFH president, Cesar Garrido, started describing the recipient he used words like 'New Zealand', 'former WFH board member', 'long-standing volunteer', and so on. The penny dropped just seconds before I was hauled on stage in front of around 3000 people,

completely shocked and speechless which is rare for me. In the distance beyond the blinding stage slights, I could see Lauren, Tineke, and John delivering an impromptu haka. This was incredibly touching to me. Even thinking about the moment some months on, I am still shocked, speechless, and moved.

WFH Congress

What makes a World Congress both inspiring and exhausting at the same time is the amount of activity that has already taken place by the time you get to the conference proper. The days that follow are a blur of people, experiences, information, and sleep-deprivation. With this in mind, it was just as well that we had a troop of Kiwis to cover everything. There's so much in fact that you will all be dining out on this information for a few Bloodlines and events to come.

This year's programme had something for everyone. I cover a summary of the Global Forum which took place in November last year and this focuses on some of the trends and new therapies on the horizon. You can read more about this on page 30.

Other attendees will have different highlights and be sharing these in the coming months. Here are a few from my perspective:

Ageing

The topic is not new, but we are all getting old. As treatment for inherited bleeding disorders improves, it follows that life expectancy increases. For countries with access to treatment, life expectancy is reaching close to that of the general population. So, what does this mean? Some of the new therapies that have led to a greater life expectancy also introduce thrombotic risk (blood clots in your blood vessels). This is completely manageable, but just adds another dimension for our treaters in partnership with you. The other two perennial topics that come with ageing are pain management, mainly musculoskeletal pain and just managing those other challenges of ageing alongside the bleeding disorder. Calls for multidisciplinary teams to manage this are not new, and highlight the importance of a comprehensive care team to work with alongside us.

Women and girls with bleeding disorders

HNZ has long recognised that men and women both 'carry' haemophilia and other related bleeding disorders, they are just carried differently. We have recognised that being a female carrier does not mean a life free of bleeding. While not everyone will have severe symptoms, we know that many of our female members do bleed and have specific needs. That's why we have committed to the period project pilot. You can read more about this on page 24. That being said, there are many places around the world where the focus remains on the men who bleed and the women who make the men suffer.You did read that correctly.

This is a long-standing stigma which has been shown to lead to feelings of guilt and anxiety. As I attended some of these sessions, including Lauren's excellent talk, what it highlighted to me was the need to continue to remember the needs of our women and girls who are part of our community and make sure they have what they need.

Global care realities

I would challenge anyone to leave a WFH event without a greater appreciation of the care realities for so many around the world. Improvements have been made in the identification of people with bleeding disorders, and the WFH Humanitarian Aid programme has made remarkable progress. The two topics above can be focused on by us because we are able to age, and able to broaden our scope beyond severe haemophilia as the sharp end of the business, so-to-speak. It can be tough out there, but in contrast to many realities around the world, we are doing well. This does not mean we should stop being vigilant and let the system know when care is not meeting the standard it should.

HNZ has seen these realities first-hand and been committed to partnering with other countries for many years now, with our first successful twinning being with Cambodia and our last youth twinning being with Nepal. The Nepal twinning feels a little like 'unfinished business' with COVID-19 putting an abrupt end to it among a range of other factors. Right now we are in the middle of an organisation twinning with the Fiji Haemophilia Foundation. You can read more about that in the next edition of Bloodline. There's also a distinct possibility that we will work with Pakistan and complete another youth twinning from 2025.

So, there you have overall impressions of Congress 2024 with more to come from other attendees. Once again, thank you to HNZ for supporting me, and to Roche for supporting HNZ and our clinicians to attend.

Tineke Maoate: HNZ Board member

Firstly, I would like to thank all the HNZ members and Board for giving me the opportunity to attend the WFH Congress in Madrid, Spain in April. It was with great honour that I had the pleasure to hear all the new advances on von Willebrands (vWD) and other platelet disorders. Yes, there was a lot of information on haemophilia, but I tried to focus on the vWD and PD as I felt that we have a good number of our members with these conditions. I also spent a lot of time in sessions covering women and girls with bleeding disorders. One of the most interesting sessions I attended was on intimacy with a bleeding disorder. This has been such a taboo subject for far too long.

There is some amazing medical work being done on vWD with some great results happening. To learn some of the studies that have been concluded has given me faith that we are on to better treatment options soon.

All the sessions and workshops I attended were very insightful. The women and girls with bleeding disorders sessions were especially interesting. All the information we

took away is going to be useful for Lauren Phillips and me to incorporate into our period products pilot.

Two workshops that stood out to me were:

The importance of oral health and quality of life for people with bleeding disorders

I thought this would be more about oral health, but I was wrong. It covered a subject that I never knew existed even with living with a bleeding disorder my whole life. That is that people with bleeding disorders are more prone to having sleep disorders. Wow, that opened a whole new conversation with the members attending Congress, and other members since my return. It was really interesting to hear that so many of them suffered from sleep disorders. Hopefully this information has encouraged our members to have a discussion with their doctors. There are many different methods to treat this. Just be careful that if you decide to use a mouthpiece, that you have it properly fitted by a dentist.

Intimacy for women with bleeding disorders

Now this was a real conversation on intimacy! Something that isn't spoken about enough. This often-taboo subject needs to be given the attention it deserves. An open, honest, and real discussion. I am planning on trying to develop a programme for any willing members. I would love to have our younger members, who are starting their journey, to have a more open and honest understanding of this important aspect of all our lives. I would love to have them comfortable in their bodies, and to be their own biggest advocate. There is also the other side of the coin, in which the partner of a person with a BD could understand how having a BD can impact your intimate lives.

There was so much information provided at this Congress, too much to write about here. But I'm sure you will learn all about it through your outreach worker, and at our Woman's Weekend, Adult Wellness Weekend, and Family Camp.

For me, the highlight of the whole event was Deon receiving his award for his endless and enduring support he has given to the WFH and HNZ over many years. Congratulations Deon, it is well deserved.

Darian Smith: Northern outreach worker

This was my first WFH Congress and I found it an incredibly valuable experience. It was an opportunity to learn about and from experts in the field, people living with bleeding disorders, and those who work with them. It was also a fantastic chance to connect with delegates from our local region and from around the world. I learned a lot about what is being done, what can be done, and what should be done. There are exciting things happening, and even more impressive things coming. It was also an eye-opening experience for me to realise how lucky we are in New Zealand compared to many places around the world.

Here are some of the highlights from some of the sessions I attended.

Psychosocial Day

The pre-congress Psychosocial Day started out with a discussion of ethics. Gaby Golan discussed the importance of ethics and adapting them to specific issues that come with bleeding disorders.

Definition of ethics: Ethics is the philosophical study of morality, encompassing the examination of moral principles, values, and codes of conduct that guide human behaviour and decision making. It explores questions about what is right or wrong, good or bad, just or unjust in various contexts. Ethics seeks to evaluate and resolve moral dilemmas.

The importance of ethics:

- Guidance for individuals
- Foundation for organisational policies
- Influence on societal norms
- Trust and reputation
- Conflict resolution
- Ethics serves as a guiding framework for individuals, organisations, and societies.

There are many ethical concerns related to psychotherapeutic professions:

Confidentiality: This is essential for building trust and rapport. Exceptions involve harm to self or others, legal requirements, supervision and consultation.

Informed consent: Providing patients with relevant information about therapy, its goals, risks, benefits, and potential alternatives. Importance of obtaining voluntary and informed consent from patients before initiating therapy and implementing interventions.

Boundaries and Dual relationships: Maintaining professional boundaries and avoiding dual relationships that may compromise the therapeutic relationship or lead to a conflict of interest. Boundary crossings may include friendships, sexual relationships, business relationships.

Professional competence: Maintaining competence in psychotherapeutic practice through ongoing education, training, and supervision. Ethical responsibilities to seek consultation.

Cultural competence and diversity: Recognising the importance of cultural sensitivity and awareness of diversity. Ethical guidelines for addressing cultural differences, power dynamics, and social justice issues in therapy.

Patient welfare and beneficence: Emphasis on the ethical obligation to prioritise the welfare of patients. Ethical considerations in treatment planning, intervention selection, and evaluation of therapeutic outcomes.

Supervision and consultation: Importance of seeking supervision and consultation to enhance ethical decision making, ensure professional growth and address challenging clinical cases. Discussion of the ethical responsibilities of

supervisors

Group psychotherapy: Considerations for deciding the amount of attention to give each individual participant. Participants are not obliged to any ethics so confidentiality in group sessions can be an issue.

Psychotherapy with children: Confidentiality regarding contents a child shares with you.

Bleeding disorders and ethics

Access to treatment is a major priority of WFH. It is estimated that only one third of expected cases worldwide have been diagnosed. The WFH path for access to care and treatment 2021-2025 includes:

- Advocacy training currently happening in 20 countries
- Onside advocacy and help with national planning
- Global and regional meetings with all stakeholders.

There are many ethical concerns with gene therapy. They include:

- Who is eligible?
- Decision process is huge and includes person, family, staff, etc
- Will it work?
- Who pays for it?
- Behavioural restrictions post dosing
- Extensive follow-up requirements
- What do you do about non-adherence?
- Shipping and storage of medicine
- Other treatments:
  - Availability
  - Cost
  - Will they work for an individual person with bleeding disorder
- Similar concerns relate to access to bleeding disorder research:
- Who gets included in clinical trials?
- Who gets informed about clinical trials?
- Are PWBD involved in designing the trial and implementation?
- Practical issues of trial participation
  - Ensuring they understand the risks and requirements
  - Many appointments
  - Transport
  - Childcare
  - Hotel/meals
  - What do you do about non-adherence?

Shared decision making is an important process for helping

patients and clinicians find the right path for the individual. Shared decision making replaces the paternalistic approach to healthcare. It is still evolving and being adopted and is an ongoing dynamic process. There is a wide variance in implementation due to personality factors and cultural factors. The World Federation of Hemophilia has a shared decision-making tool on their website. There are webinars there too.

Most professional organisations have a code of ethics that define standards about multiple relationships with clients/patients. There is a wide variance related to profession, culture, local customs and expectations, and specific situation. Multiple relationships are common in the bleeding disorders community, where community members fill multiple roles, e.g. camps, committees etc. When this arises, first check the policies and code of ethics.

The social media space is particularly problematic with respect to multiple relationships. It can be difficult to know how to respond to requests from patients to follow/friend you in your personal life. You may also be open for social media interactions with therapeutic/home health care reps, or other accidental connections. It is important to think about how exposure to patients' social media posts affects your interactions with them. You are also more able to counter misinformation and disinformation spread through social media.

Susan Cutter: Psychosocial curriculum and resources

Susan Cutter discussed a useful resource that has been developed. Modules include presentations of about 45 mins. The resource includes original content from the psychosocial committee, a trainer's guide, and 16 modules on a wide range of issues.

A self-action plan for participants is available to use to help set goals and achieve them. A psychosocial evaluation toolkit helps tweak the programme.

Modules include:

- Quality of life
- Pain management
- Team approach
- Genetic counselling
- Sexuality
- Life stages
- Coping with hiv or hepatitis
- Women & bleeding disorders
- Support groups
- Coping with join disease
- Coping with inhibitors
- Advocacy
- Disclosure

Additional WFH psychosocial resources

- Medical treatment doesn't automatically translate into better quality of life.
- E-learning platform on the WFH website.
- The resource "Psychosocial Care for People with Haemophilia" covers psychosocial issues encountered by those living with haemophilia, guides healthcare professions and more.
- Also includes some of the psychosocial track of WFH World Congress in 2022.
- New communication strategies for bleeding disorders community.
- Training and career guidance for youth with bleeding disorders.

Advanced techniques in pain management

Fifty percent of people with bleeding disorders report experiencing chronic musculoskeletal pain, and 35-50% report that current treatments for pain are not effective. Developing effective treatments for pain is a research priority and is different to traditional pain services. Pain mitigation demands change depending on life stage and treatment options. This is about preventing trauma and not just minimising or dismissing it.

We are not conscious of our bodies if they are functioning well. When pain disrupts things, we focus on the pain and fix it and it works and we go back to normal. But what if the pain doesn't get fixed? Then what? One option is Acceptant and Commitment Therapy (ACT):

- Be present
  - Be here and now. Mindfulness
  - Stressing about the past or future can lead to anxiety. Focus on now is better.
  - Mind full or mindful? Let it go.
- Open up
  - Make room for thoughts, feelings, sensations
  - Acknowledge, accommodate, and allow these
- Do what matters
  - What actually matters to you? Spend your energy on that.
  - If you can't remove the problem, change the context. Move toward what you want and bring along the issue anyway.

It is important to map the way forward. Ask yourself:

- How close to living how I want am I in every area of my life?
- What actions do I want to take to move towards that?
- What is the smallest and easiest step toward this?

In many cases it's not about people living without pain, it's about living well with pain.

Support groups

A support group is a gathering of people who come together to provide mutual support. Usually led by a professional or trained volunteer

Many people value the benefits of shared experience and emotional support in a safe and non-judgemental environment. These groups often enable better access to educational resources and practical assistance, while increasing advocacy and awareness.

Support groups traditionally meet face-to-face. However, it is becoming more common for these to occur online, or as a hybrid combination of both.

To build impactful support groups for people with bleeding disorders it is essential to first identify the unique needs and challenges of the bleeding disorder community. The goal should always be to facilitate meaningful discussion and encourage active participation by fostering a safe, inclusive, and judgement-free environment. Difficulties with these groups include maintaining momentum and adapting to evolving group dynamics.

You must be able to measure the impact of your support group. This can be done by developing metrics and evaluation methods, including participant satisfaction and engagement, changes in knowledge, skills, and coping strategies, and improvements in physical and mental wellbeing. Another way of showing success is by sharing success stories and highlighting tangible benefits via individual testimonials and case study.

Narrative therapy

We live our lives according to the stories we tell about ourselves, and the stories others tell about us. Life is multi-storied, and we make meaning out of life with the stories we tell. People tend to retell the stories that fit the dominant narrative, so it's good to listen to the ones that are the exceptions to the rule.

There's a lot of power in telling someone else's story and making it the definitive story about them. We can choose to tell stories that make us stronger. Stories about problems can easily become dominant, so we need to get to know the person apart from the problem by allowing the person to tell their story and tell their preferred stories. The person is not the problem, the problem is the problem. Invite people to make a distinction between themselves and the problem.

A draft charter for story telling rights:

- To tell their stories in ways that fits for them, not as defined by others
- The right not to have problems caused by trauma and injustice located internally as if they were a deficit in them
- The right to have their responses to hard times acknowledged
- The right to know and experience that what they have learned through hard times can make a contribution to the lives of others in similar situations

In the Beads of Life workshop participants choose beads to represent stories in their lives and tell their stories and let them be heard

Mental health

Internal family systems

This is not family therapy. It's a therapeutic approach in which the client is the agent of healing.

The mind is composed of various parts. We are often not aware of those parts and not taught to treat them with compassion. Reaching out to the less helpful parts helps brings healing and change. The parts have good intention. Some parts are Exiles, and are thought of as wounded, and others are Protectors, that manage relations with the world.

Exiles are often frozen in time at the point of wounding. They experience trauma over and over and carry experiences and feelings that we don't want to feel. These parts are often locked away by protectors for safety. These are often our most innocent and open intimacy-seeking parts, which can contain joy, playfulness, spontaneity, and creativity.

Protector managers try to keep exiles from activating. They try to keep us stable and functioning, and work on a "never again" promise. They can lead to perfectionism, restriction, and controlling.

Protector firefighters take action when exiles activate. They try to stifle, numb, or distract, and are impulsive. They work to stabilise and balance the system but can lead to substance abuse and other harmful behaviours.

There are no bad parts, they all have good intentions but can be problematic. The 'self' is the executive who directs the parts.

The focus of treatment is the whole system not individual symptoms. Symptoms are the activity of parts. Treatment helps clients to discover and understand what the parts are doing and why, and to show compassion to them. Treatment can involve sand tray, toys, dialogue with parts, visualisation, empty chair, and art, and works towards healing and integrating exiles and reducing protectors' extreme activities.

Trauma-informed care

Trauma refers to an experience that creates a sense of fear and overwhelm. It may be physically or emotionally harmful, have multiple perpetrators, and be influenced by the event and the victim's response to it. This includes neglect. The CDC estimates 60% of adults experienced at least one childhood trauma.

The impact of trauma on health behaviour can include:

- Bing drinking
- Obesity
- Chronic pulmonary disease
- Smoking
- Depression

People who have experienced trauma are 67% more likely to abuse alcohol and drugs, and 12.2 times more likely to commit suicide. It's important health care workers screen for these risks.

It is not uncommon for people who have experienced trauma to ignore advice or leave treatment facilities against medical advice. They often do not comply with treatment, can feel controlled or abandoned, and may be fearful of invasive

exams. In some situations, they may freeze up. We might also see an increased utilisation of healthcare resources related to fearfulness and hypochondria.

There is group trauma associated with the impact of HIV on patients with access to factor replacement products before 1985. Many BD patients developed close friendships during hospitalisation, clinics, camps etc. This includes the parents and whānau of BD patients. The many points of contact and widespread impact of HIV resulted in repeated traumatic exposure. People who remain in the vicinity of trauma may encounter greater challenges in recovery. They may experience a "Who's next?" retriggering over and over.

In trauma-informed care it is important to employ precautions universally since the patient's trauma history isn't initially known. Practitioners should provide a safe environment, ask permission to do any exam and explain what is involved, and build a positive trusting patient/provider relationship.

Strategies to prevent retriggering include screening for trauma, which involves recognising trauma and being aware of how it presents itself, as well as understanding the health effects of trauma. It is important to provide patient-centred communication and care, emphasising emotional safety and avoiding triggers.

An effective practitioner is knowledgeable about helpful treatments, persistent, reliable, and patient. They will use gentle prodding to obtain information and take a nuanced approach, recognising that maladaptive coping methods may be related to trauma. These practitioners provide strengths-based psychosocial counselling to enhance life skills, reduce stress, foster growth, healing, and regulation of emotions via distraction and/or relaxation to reduce the freeze response. They help patients to be kind to themselves and enhance their sense of control and choice.

Other strategies include helping people give themselves permission to move forward, even if it's painful, and encouraging them to actively reflect on the past and work to move through it.

The key principals are:

- Safety
- Trust and transparency
- Peer support
- Collaboration

It is important to be aware of vicarious trauma and the risk to other family members and healthcare workers. You can experience trauma from someone else's trauma.

Mindfulness

Mindfulness is a mental training practice that involves paying attention to the present moment, and acceptance rather than getting caught up in worries about the past or future.

The benefits of mindfulness include:

- Reduced stress and anxiety
- Improved emotional regulation
- Enhanced focus and concentration
- Increased self-awareness.

The key aspects of mindfulness are present moment awareness with no judgement or attachment, self-awareness, and acceptance. For people with bleeding disorders, it is beneficial for stress and pain management and medication adherence, as well as emotional regulation and improved self-care.

Mindfulness practices include:

- Meditation: Formal meditation practices, such as sitting meditating, body scans, and loving-kindness meditation.
- Breathwork: Techniques that focus on conscious, intentional breathing to promote relaxation and awareness.
- Mindful movement: Practices like yoga, tai chi, or

walking meditation that integrate physical and mental awareness.
- Informal mindfulness: Incorporating mindful awareness into daily activities, such as eating, washing dishes, or during conversations.

Digital detox, a period when you abstain from using digital devices, can be helpful in mindfulness practice. The goal is to reduce stress, increase mindfulness, and gain control over your relationship with technology, which can make you more conscious of your attention being fractured by digital devices. Digital detox is good for stress management and improved sleep quality, present moment focus, and increased body awareness.

Tips:

- Eating mindfully: Paying attention to taste, texture, aroma and eating slowly and avoiding distraction
- Drink mindfully: Savour each sip
- Walk mindfully: Notice the surroundings with all your senses
- Set no-screen times of day
- Establish tech-free zones, like the bedroom
- Delete distracting apps or use screen time limits.

By integrating mindfulness and digital detox, individuals with bleeding disorders can cultivate greater self-awareness, stress resilience, and overall well-being to better manage their condition. The two practices complement each other in promoting physical, mental, and emotional health.

Dr Sarah Whitaker: Eye movement desensitisation and reprocessing

EMDR can be useful for people with bleeding disorders to help with trauma, pain, complicated grief, childhood attachment traumas, and phobias. People with bleeding disorders may be at risk of developing post-traumatic stress disorder (PTSD) because of related events. PTSD can lead to reduced adherence and increased risk.

A study on post-traumatic stress disorder symptoms among adults with haemophilia found that 57% of the 178 participants identified a haemophilia related traumatic event. The prevalence of PTSD is 12%, which is three times the average for the general population. Trauma related to medical intervention or interaction with medical providers was identified by 92% of those who identified previous trauma. Eighty-two percent described recurring/chronic traumatic events, while 58% indicated trauma began or occurred in childhood.

The brain is affected by trauma. The creation of memory can be disrupted, and information may be stored in fragments. Often, traumatic memories have not been stored in wider memory networks. These memories can be "triggered" and recalled in chunks with difficult body sensations.

Eye movement desensitization and reprocessing (EMDR) is a recommended treatment for trauma. The aim is to kickstart the brain's natural processes by allowing the thinking part of the brain to engage rather than just the fight/flight part, reducing the stress around the memory and updating it with newer, more helpful information (wise owl part of the brain). Once stored the memory can be accessed without the guard dog (fight/flight part of the brain) getting involved.

Bilateral stimulation (BLS) helps with this. BLS is external stimuli that crosses the brain midline. It encourages the whole brain to be active at the same time so the wise owl can make sense of what's happening while the guard dog is barking. Any activity that uses both sides of the body and therefore both sides of the brain can work e.g. butterfly hug, follow fingers with your eyes, kicking a ball back and forth.

Safety and Treatment

Monday brought a plenary session discussing safety in treatment. We know that prophylaxis does a better job of preventing bleeds, but there needs to be a better treatment than regular factor VIII. While levels of 12% or above are protective, it's best to have them stay at around 20 - 30%, and this requires a significant burden with conventional factor VIII.

History shows us shifts in the risks and in the perception of the acceptability of those risks.

- Pre 1970s: Inhibitors and uncontrolled bleeding
- 1970s: Hepatitis B, as well as nonA and nonB (later named C), were thought to be an acceptable risk
- 1980s: Hepatitis risk not so acceptable and HIV appears
- 1990s: Variant Creutzfeld Jakob disease, rapid brain destruction, bacterial and fungal contamination in manufacturing
- 2000s: All flavours of zoonotic infections, no transmissions in CFCs but that wasn't known at the time and life finds a way
- 2010s-present: Gene therapy an acceptable risk, rebalancing agents, acceptable risks include liver inflammation and thrombosis.

Due to this, safety underlies everything. When new technology comes along, we must review how we think about safety alongside it. The fundamental concepts to keep in mind are informed decision making, shared decision making, and risk/benefit analyses.

Some of the risk issues we are dealing with today include:

WHO medicines list

WHO essential medicines list is a list of priority conditions and minimum medicine needs for the most efficacious safe and cost-effective medicines. In 2023 they placed cryoprecipitate on the core list and factor VIII and IX on the complementary list, which most would consider not to be the most effective, safe, and inexpensive treatment options. Dependence on blood components comes with a risk of contracting a viral infection as a result. The WFH has challenged WHO to move cryoprecipitate to the complementary list and put factor on the core list.

Gene therapy- risks from vectors and trans genes

A vector is the carrier mechanism for the genetic material, usually a modified virus. At higher vector doses, elevated liver enzymes occur in most patients. We often don't understand the mechanisms, but it's a sign of liver damage. Immune responses to the AAV capsids could also occur.

The high vector burden could have an impact on patient health.

The gene can misfold and fail. Pigs make factor VIII that folds well, but there could be an inhibitor developed because the immune system recognises it as porcine.

Treatment choice and comorbidities

With increasing life expectancy, we see increasing comorbidities. Non-replacement therapies excluded a lot of people with comorbidities and the aged from the trials, which means we don't have as good data on how it affects those groups. Treatments are rapidly evolving but not all patients are represented in development and post marketing surveillance will be valuable to get real world data. Shared decision making crucial for best results

What have we learned from EUHASS real life data?

Detection of rare events in rare populations needs to be followed up with a larger number of individuals. Data collected showed events for people with bleeding disorders across 26 countries that included allergic reactions, inhibitors (reduction in these), thrombosis (rare but does occur), liver disease (hep c should be cleared by now but still see liver cancer and liver disease as a result in some patients).

Other EUHASS data reports on poor efficacy, other possible adverse events, and more.

Dr Zikra: Oral health and quality of life – intimacy, pain and self esteem

Oral health is important for overall quality of life and health. Bleeding gums and dental pain can cause issues for everyday activities. Despite being common, most oral diseases are highly preventable. Good dental hygiene and healthy eating practices should be part of everyone's routine.

It's recognised that oral health is important for PWBDs. It impacts social aspects of life, appearance and self-esteem, chewing and speech, pain and discomfort.

Pain: Dental pain spreads through the head, disrupts sleep, impacts concentration, mood, and sense of wellness.

Intimacy: Bad breath, social embarrassment, the potential to make intimate moments uncomfortable, self-consciousness or low confidence in new relationships.

Function – gum disease causes inflammation of gums resulting in bleeding, impact on intimacy and dining and altered taste. Tooth decay causes pain with hot and cold foods

Self-esteem: Bad teeth and bleeding gums affect how we look and how we feel about ourselves and our smile.

Periodontal disease is the most common chronic inflammatory condition in humans and impacts 50% of the population. People with bleeding disorders aren't more susceptible, but it is significantly more noticeable for them. They can be scared to touch the mouth for fear of causing a bleed, but without cleaning off the plaque, the condition gets worse.

Healthy gums don't bleed. Use fluoride toothpaste, bush your teeth regularly, use mouthwash, and see the dentist regularly. The biggest barrier to oral health for people with bleeding disorders is their own fear. Dealing with it will make it better.

How do we measure bleeding?

Using levels to classify is becoming less valuable when we know there are other factors that come in to play to form phenotype. Categories like 'moderate' and 'mild' can be weaponised to dismiss patients' actual experience.

How does categorisation impact patients:

- Interaction with the welfare state?
- Changing meaning and characteristics of disease over time?
- What is the classification system intended to do and who is the vocabulary for?
- Who is excluded?

It could be time for new classification system to be created in consultation with those who live with haemophilia.

The lack of appropriate care that women have received due to the categorisation of "carrier" is a sharp reminder that the names and language we use have a direct impact on the experience of groups of people to whom those classifications apply.

Any changes will need global buy-in. Is it a priority? Who is it a priority for? Those who live in developed worlds are dealing with different issues than those in developing worlds.

For each male with severe haemophilia, there are 1.6 females with the haemophilia gene. Women can be divided into mild, moderate, severe, and asymptomatic carriers and symptomatic carriers.

Women include a wide range of bleeding disorders. Reproductive tract bleeding is significant. They have higher rates of ED visits and hospitalisation for vaginal bleeding, anaemia, blood transfusions, haemorrhagic ovarian cysts, hysterectomies (and at a younger age), endometriosis, and fibroids. But they are not being heard. Women commonly experience lengthy delays in diagnosis and treatment.

Why?

- Diagnosis bias: Symptoms overlooked or dismissed in women and girls

- Hormonal influences: Menstrual cycle variations can complicate diagnosis
- Socio-cultural factors: Stigma and taboos surround menstruation can prevent them speaking up
- Sexism exists

There is a lack of awareness on behalf of health care professionals and the public that leads to challenges. The consequences of untreated bleeding can lead to a significantly lower quality of life.

So, what do we do?

- Pinpoint what constitutes abnormal menstrual bleeding in ways that make sense to regular people.
  - Do they change product every hour or more often?
  - Do they soak through the products and pyjamas at night?
  - Are they passing clots more than an inch in diameter?
- Screen
- Treat!
- Continue creating awareness of the issues.

Subclinical bleeds

Joint bleeds are damaging, and we know this well. The more bleeds you have, the more risk you have of lasting joint damage. With new treatments we need to focus on damage in joints that comes from non-clinically evident joint bleeds. MRI evidence shows that there is iron in the joints of patients without an overt history of joint bleeding. MRI findings in healthy adolescents found some similar issues and we just don't know if it's haemophilia specific or not.

There is evidence for subclinical bleeds or processes that cause joint changes, and we don't know enough about how it happens or what it means. Joint damage progresses and there is a risk of overt bleeding but it's unclear whether changes on imaging are active or inactive.

Physical examinations can't show these kinds of bleeds, only imaging can find it.

How do we treat them?

- Unsure!
- Improving haemostasis but at what level? And will non-factor therapies be enough?
- Are anti-inflammatories or iron chelation more useful?
- There are many questions on this topic and as yet no clear answers
- There is more study to be done

Adherence vs Compliance

Treatments are great, but they need to be taken as prescribed. Adhering to treatment recommendations can be an issue with some patients.

- Adherence is "the extent to which a person's behaviour; taking medication, following a diet, and/or executing lifestyle changes, corresponds with agreed recommendations from a health care provider."
- Compliance is "the extent to which the patient's behaviour matches the prescriber's recommendations."

Adherence is an active choice of the patient. It is taking responsibility. Compliance is passive behaviour.

Adherence to long-term therapies can be a challenge, but medicines won't work if you don't take them. In developed countries only 51% of patients adhere to anti-hypertension treatment, whereas, in developing countries 26-43% of patients adhere to it. Adherence to antidepressants ranges between 40-70%, and adherence to asthma preventative medication is only 28%. Adherence to antiretroviral meds for HIV varies between 37-83%

Five dimensions that affect adherence:

1. Health system/HTC factors

The key to this is the patient/provider relationship, which hinges on the health care provider's communication skills. This helps reduce health belief disparities and can help mitigate a lack of education about the value of adherence and intervention. Other factors include poor access to health care, long wait times, and lack of continuity of care.

2.Social/economic factors

Language barriers, low health literacy, lack of a support network, a burdensome schedule, and other difficult life situations all contribute to low adherence. Economic factors like unstable living conditions and high treatment costs are also significant factors.

3. Condition related factors

Adherence is affected by other chronic conditions, a lack or severity of symptoms, depression, and cognitive issues.

4. Therapy related factors

Complexity of or changes to the treatment regimen can have a negative effect on adherence. This includes admin issues, therapy duration, and interference with patient's lifestyle. Other factors include a lack of immediate effect, stigma around treatment or the disorder itself, and adverse events related to treatment.

5. Patient related factors may include:

- Impairment
- Problems swallowing or fear of needles etc.
- Perceived risk of the disease or treatment
- Knowledge about the disorder
- Attitudes and expectations toward therapy
- Confidence
- Motivation
- Fear of AEs, dependence
- Psychosocial distress, anger, anxiety
- Alcohol or substance abuse

Treatment adherence is important in haemophilia to reduce bleeding frequency and arthropathy, and thereby maintain joint health. It also leads to a decrease hospital or emergency admissions and length of stay, an increased tolerance of

high-level activities, improved work or academic attendance, performance, and accomplishments, and an overall improved quality of life. Adherence also enhances the cost-effectiveness of treatment.

So, what is the definition of treatment adherence, and how do we measure it? The most accepted measure is that you're infusing 75-80% of the prescribed therapy. This is measured by:

- Self-reporting
- Vial count
- Drug assays
- Direct supervision
- Electronic monitoring
- Clinical outcomes

Adherence to prophylaxis decreases with age. Children adhere much better than adults, possibly because of parental intervention. A global survey on the barriers to prophylaxis asked what factors affected patients' ability adhere to their treatment:

- 75% didn't understand the potential benefits
- 67% were in denial about their treatment
- 66% experienced poor venous access
- 63% identified a lack of parental or family commitment
- 62% said it interfered with their lifestyle
- 48% cited teenage rebellion
- 42% lacked the time needed.

Patients also cite forgetfulness, a lack of commitment, the transition into adulthood, and a lack of social support as reasons for low adherence. If they have a mild expression of their disorder, infrequency of bleed events may also contribute. In many countries cost and availability are barriers to regular treatment. There are also perceptions of risk associated with inhibitor development and other complications.

The first step to help improve adherence is education. This includes improved communication, the promotion of bleed tracking through diaries or phone apps, and increased awareness of important studies. It is also important to address improvements in treatment, including EHL factors and non-replacement therapies, and to promote prophylaxis and individualised therapy to decrease inhibitors and enhance participation in activities. Psychological interventions to decrease anxiety and training for independence help to improve ease of venous access.

So, what are the challenges of the transition to the adult world? It can be difficult to let go of long-standing relationships with paediatricians, and more difficult to access health care as an adult. Beliefs and expectations about adult care can also affect transition. Many people with bleeding disorders have limited knowledge about disorders, medications, and risks, and lack the self-management skills to facilitate a smooth transition. There can also be relationship barriers.

Research informed recommendations to ease this transition include:

- Allow patients interaction with adult providers before transfer
- Create join clinic visits attended by paediatric and adult providers
- Create a structured transition plan
- Connect patients with peers who have already transitioned
- Provide tours of adult clinic
- Begin early transition and education about responsibility
- Use technology to target knowledge and self-management behaviours
- Regularly assess and discuss transition readiness
- Improve coordination of care between medical teams.

Patient centred care helps with all these issues. Patient centred care means that the treatment provider's mission and values are aligned with patient goals. Care is collaborative, coordinated, and accessible, the patient's physical comfort and emotional wellbeing are top priorities, and patient and family viewpoints are respected and valued. That means the patient and their family are always included in decisions, the family are welcome in the care setting, and there is full transparency and fast delivery of information.

Health literacy

Health literacy is about understanding health information and how the system works. It's easy for medical professionals to forget that not everyone works in a health setting and therefore may not understand what may seem basic to someone who does. It may also be that someone has low general literacy and therefore has issues with reading information materials and understanding numbers.

Studies show low health literacy in 60% of Canadians and Australians, 47% of Europeans, and 43% of Americans.

Low health literacy can contribute to:

- Medical complications
- More hospitalisations and ER visits
- Adherence issues
- Less uptake of preventative care
- Poor overall health
- Shorter lifespan
- Financial consequences

Some signs that someone might have low health literacy or low literacy include being reluctant to ask questions and may involve excuses such as they've forgotten their reading glasses. They may. If they have an awareness of their difficulties, they might have a trusted support person there to read for them, otherwise they may just say they will read it later and leave. People with low health literacy will often have treatment adherence difficulties.

It's best for medical providers to take universal precautions with this. Assume everyone has some difficulty understanding and accessing medical information and assess for barriers. Ask how people like to get information, by writing or by talking, and whether they find themselves asking family for help reading medical information. Check in

about whether they are comfortable reading the information, and whether they have any questions.

It is important to keep education sessions brief, talk slowly, repeat and get them to teach back. It is always worth having a family member attend but remember to not just talk to the family member. Patient education materials should give what they need to know not what's good to know. Materials should use pictures that represent the audience, graphics and visual tools, and plenty of white space. Written information should be in the active voice, not passive voice, and be in multiple languages. It should be visually appealing, incorporate bullet points, be conversational, and at a 7th grade reading level.

The CDC has a resource for health literacy on their website.

If you have issues with understanding your medical team or the information they provide, don't be afraid to ask them to be clearer and go over it again. Also feel free to bring your HNZ Outreach Worker to help.

Women with Bleeding Disorders

Women with bleeding disorders are often overlooked and receive delayed diagnosis and therefore delayed or limited access to treatment. Yet they have extra risks associated with menstruation and pregnancy.

Eighteen million women in the world have heavy menstrual bleeding. This can often be a symptom that is overlooked because the patient and/or the care provider don't have a clear understanding of what is going on and how much bleeding is normal.

Signs of Heavy Menstrual Bleeding or Menorrhagia:

- Clots of greater than an inch in diameter
- Low iron levels
- "Flooding" and having to change a tampon or pad hourly
- Losing more than 50ml per menstrual cycle or bleeding for longer than seven days

When it comes to bleeding disorders that are not sex-linked, women will have just as much to deal with as men, if not more.

Christina Voss: Living with Factor 1 deficiency

She says better cooperation between treatment centres and doctors is needed with rare bleeding disorders. It can be hard to find the right treatment when you have a rare disorder. Talking to different treatment centres can be helpful, as you may be the first example they've seen. Meeting other people with the same condition gives people a better idea of treatment. Prophylaxis freed her from having to wear protective gear, and she hasn't had a heavy period because regular treatment deals with it.

After being told "people like you don't get children" she sought out gynaecologist in private practice as a proactive connection to find one who can work with someone with a bleeding disorder. She recommends that gynaecologists work with the treatment centre to ensure good care.

Latifa Lamhene: Patient perspective in developing countries

spoke about having combined FV and FVIII deficiency which is a rare autosomal recessive condition. In Algeria only 20 people are known to have it.

Before the age of 19 she had multiple external bleeds following falls, dental extraction etc. at 19 they discovered the disease after lab investigation.

There was a lot of misunderstanding about the disease and risk. There was an absence of care and treatment because "girls can't have haemophilia."

At 22 a burst ovarian cyst resulted in abdominal bleeding and required an emergency surgery, which was badly done with no factor and infection developed. After recovery she did extensive research to understand this condition and contacted a haematologist for her first consultation in 2001. A second surgery went much more smoothly as there was

factor available and used.

Other complications include constant proctorrhagia and hematuria, hematoma, hemarthrosis, hemopericardia, hemoperitonia caused by ovarian cysts.

Often during bleeding, she only treats with factor VIII, but when necessary frozen blood is used to replace the factor V. She understands there are risks to using frozen plasma, but there isn't an alternative for her. She now uses prophylaxis for factor VIII.

She offers training days for haematologists, gynaecologists, and biologists, girls summer camp, and education groups for girls on World haemophilia Day.

Her recommendations include:

- Implementation of guidelines for rare bleeding disorders
- Emergency protocol for management of abnormal bleeding
- Have specialised centres for women
- Raising awareness of women with bleeding disorders
- Realising factor levels can be misleading for women.

Roshni Kulkarni MD: Empowering women's health - The importance of early diagnosis, registry participation, and the vital role of guidelines.

Early diagnosis leads to education and awareness, registry participation, and self-advocacy, which feeds into the role of guidelines and will help lead to more early diagnoses.

Early and accurate diagnosis is crucial for timely, appropriate, and effective management. It prevents complications and improves quality of life

Undiagnosed and untreated women and girls have medical consequences at any stage of life, such as surgeries, iron deficiency, etc.

Ideally diagnosis during pregnancy and at birth would be great. Genetic diagnosis helps people make informed decisions.

Women are being diagnosed much later than men. The median age in the US is 22.6 years for diagnosis. Some of the challenges to early diagnosis include limited awareness, access to testing, and societal stigma around menstrual health.

Registry participation is vital to provide data to make arguments and advocate for women. The number of diagnosed women has increased.

In the CDC community count 2012-2020:

- 1667 women and girls received care at an HTC and met criteria for haemophilia
- Severe was rare but there were some.

Registry helps with:

- Public health translation
- Surveillance
- Epidemiology
- Lab research

Guidelines help to make care consistent and ensure proper treatment. Many are available on the web, and you can share them with your doctors. A period pocket guide can help to provide information.

Self-advocacy empowers patients to engage in health decision and advocate for their needs.

Dr Caroline Malcolmson: Control and management for women with coagulations disorders - Steps to advance better and more standardised treatment

What is the need for specialist clinics for women with bleeding disorders?

- There are many women with heavy periods and bleeding disorders
- 5% will get a haematologist and get screening bloodwork and when they are screened up to 60% have a bleeding disorder
- It's an under-identified group

Goals: provide multidisciplinary care to reduce the burden of care and number of visits. Do gynaecology and haematology at the same time.

Find mentors and build a team including haematologist, gynaecologist, and social worker.

Taking careful history: Use open questions. What brings you in? Tell me your story. Consultation reviews medical history, surgical history, family history, menstrual history, and medications, etc. Standardised bleeding questionnaires helped identify how bad the menstrual bleeding is. Some women were getting scores that were twice what would be considered heavy.

Diagnose and treat disorders: Her clinic has now treated many women with bleeding disorders. They now have a diagnosis and get appropriate care. They have a Factor First card which helps them advocate for themselves when in the ER. She advocates tracking bleeds, including menstrual bleeding, so it's able to be treated well.

Diagnose and treat heavy menstrual bleeding: Heavy is defined as 80mls but nobody knows if that's accurate. Better questions are "do you change products every 1-2 hours, double up products, leak, pass clots larger than a quarter?"

Treatment:

- Patients' preference for contraception, drug tolerance, and adherence.
- Most patients chose oral contraceptive with or without tranexamic acid to treat. Results show improvement but there is a way to go.
- Look for and treat iron deficiency. 55% of patience's presented with iron deficiency. Iron supplements helped.

Supportive care and resources: A social worker helps them to work with domestic violence, pregnancy, STIs, depression,

etc. connecting them to resources

Continuous improvement: Continuing to review the data and find trends and see if we are serving the patients. Understanding the barriers women face in receiving care.

Intimacy and sexuality

Exploring sexuality and desire across the lifespan

Sexuality is an intrinsic quality of human beings, but we are surrounded by a bombardment of cultural stuff, guilt, and shame, so talking about sex can be difficult.

Studies show 40-50% of bleeding disorders patients say it has negatively affected their sexual life. A satisfying sex life has multiple biopsychosocial benefits. We need to include sexual health in comprehensive care and in future research.

The most common question is "is what I'm experiencing normal?" and the answer is "what you're experiencing is valid. Let's talk about it."

Sexual desire: Freud was one of the first to explore this and then Helen Kaplan, then Levine, Bancroft adding affective, cognitive and social determinants. So, we know biology, psychology, and context all help shape sexual desire.

Biological aspects can include:

- Chronic disease
- Drugs
- Chronic pain can make physical interactions difficult
- Reduced sexual desire
- Discrepancy with their partner's sexual desire
- Aging and a decrease in sexual hormones, sometimes erectile dysfunction which leads to avoidance

Psychological aspects include:

- Mental burden
- Fears of bleeding, pain, STIs
- Stress, anxiety and depression
- These all affect the patient and their partner.
- Feelings of loneliness and inferiority impact the ways to relate. "I always felt I owed something to my partners, as if nothing I did was enough to make up for them choosing me despite my disorder"

Physical changes:

- Differ from images we are presented as desirable
- Impacts body image and self esteem
- Older adults and people with disabilities are denied sexuality

How can we approach the world of desire when reality seems so distant? Recognise that everyone has the right to a free and full sexuality.

The PLISSIT model is one way to help clients dealing with intimate issues.

PLISSIT model:

- Level 1: Permission
  - Granting permission for discussion
  - Normalising conversations
- Level 2: Limited information
  - Providing basic and scientific information
  - Instructing on the use of resources to enhance pleasure
- Level 3: Specific suggestions
  - Based on specific problems
- Level 4: Intensive therapy
  - Refer to experts (psychological, sex therapy, medical approaches)

Final thoughts:

- Sexuality is individual and belongs to each person
- Aging leads to new ways of intimacy
- Sexual health is an opportunity to enhance overall health
- To continue generating knowledge to improve sexual quality of life
- Limiting beliefs stop us exploring who we truly are and aspire to become.

Gregg Blamey: Intimacy through a musculoskeletal lens

Objectives are to review research on communication addressing this topic, acknowledge the muscular and skeletal impact and possible bleeding potential due to sexual activity, and provide information and resources on positions and practices.

Every clinician surveyed recognised the importance of the topic but seven out of ten never raised it with patients and didn't think they would.

Potential for injury or bleeding:

- Vigorous vs. controlled movement
- Sedentary lifestyles promote weaker, less flexible muscles
- Sexual positions may demand increased muscular activity in less than perfect positions
- Consider the pressure being put on specific joints in certain positions. Elbows and knees can be expected to take a lot of weight and movement. There are positions that put less pressure on these joints.
- Risks include sprains, strains, fractures, aggravation of existing injury, and pain
- Profiling the potential for risk – adolescents, young adults, usually related to growth spurts, those with recent or chronic injuries, those who are more sedentary, muscle bleeders, and those with an established history of psoas bleeds
- Treatment disparities have an impact.

Erectile dysfunction is now viewed as an indicator of cardiovascular pathology. Older men are more likely to accept it as a fact of life, compared to their younger counterparts. Medications can often cause a disruption of normal sexual performance or desire. When you add arthritis to the mix, painful joints can affect libido.

Why do we need specific resources? Because the internet doesn't provide useful info in this space. Online porn is massive but it's not realistic sex.

Sexual health has gained recognition but needs more attention more consistently from health care professionals.

It is important to be aware of the physicality of sex and the pressure points and threats to injuries.

Diane Standish: Building healthy relationships and emotional intimacy

Emotional intimacy is warmth, closeness, sharing, mutual

respect, honesty, self-disclosure, being vulnerable. It involves a gradual peeling way of one's outer layers and is the foundation of a lasting relationship. Sternberg's triangle of love talks about how relationships have intimacy, passion, and commitment at different levels, creating different types of love. The best has all three.

Over the course of a life, emotional intimacy shifts and wanes. Erikson's life stages suggest we have competing priorities at different stages of life, such as intimacy, independence, identity, etc. Romantic love may give way to an affectionate love where there is less anxiety about attachment, sexual attraction may decrease, and familiarity increases. In middle-age priorities are more tied to security and familiarity. Whereas, late adulthood favours consistency and familiarity, as well as intellectual and spiritual intimacy. Caring for a spouse with a chronic disease can put pressure on these aspects.

Practical obstacles to intimacy include work responsibilities, children, changing priorities, and time. There are also psychological obstacles. People have differing needs and expectations, and it's important to understand that intimacy is not a linear process. We can fall in and out of intimacy, and it cannot be rushed or achieved all at once. Some people have a fear of intimacy, because they may get hurt, give up independence, or perhaps have a history of abuse/trauma so are self-protective.

Creating and maintaining intimacy requires self-reflection, mutual effort, and courage. Making asking about sex part of the "activities of daily living" creates the opportunity to talk about it. To cultivate and maintain emotional intimacy it is helpful to ask:

- Do I bring my best self to my interactions with my partner, or do I hold back?
- Does my fear of loss and abandonment cloud my perspective and prevent me from sharing my authentic self?
- Do I feel comfortable asking for what I need?
- Do I possess self-love and expect to be loved and respected?

Gottman's Principles for Making Marriage Work include:

- Nurture fondness and admiration
- Turn toward each other
- Let your partner influence you
- Create shared meaning together.

Intimacy and Oral Health

Dentistry Today published a study done by Dentavox in 2020 that showed oral health has an impact on intimate relationships and good oral health can lead to more kissing, more sex, greater intimacy, and feeling more attractive.

Aging with Bleeding Disorders

Jan Glazewski: Aging with haemophilia. 70 years on

Jan discussed his experience and revealed he didn't think he would be here 70 years later but is grateful for everyone who worked to improve things for people with haemophilia. He says aging haemophiliacs are a relatively new species.

His parents left Poland "in a hurry" in the early days of WW2. Older brother had haemophilia and died age 3. Jan was diagnosed at birth. At age 32 he was diagnosed with HIV because of blood products. Age 59 married his wife.

He's had a number of issues with joints, including both knees replaced in 1990, left ankle fused in 2001, right ankle replaced in 2019, left elbow replaced in 2020, and he's now

contemplating replacing his right elbow.

In terms of blood products, he had whole blood from birth until age five, fresh frozen plasma in his early teenage years, and cryoprecipitate in his later teen years. He finds self-injecting is difficult as an older person as his veins are bad and his motor skills are reducing.

Aging with haemophilia and HIV has led to damaged veins. He also wonders if what he feels is haemophilia pain or age related. He also experiences the ill effects of HIV drugs.

Jan says aging not for the faint hearted but with haemophilia and HIV it's even more difficult.

Comparing women and men

Diana posed the question, what is it about aging that scares us?

For her it is the uncertainty, fertility and pregnancy issues, access to proper haematological treatment, there being not enough studies regarding thrombosis in bleeding disorder patients, the lack of availability of proper cardiovascular treatment, and the worry that she'll be a burden to her family.

When she reached out to other women about aging with a bleeding disorder and asked what they're most afraid of, they talked about restricted mobility, an incapacity to self-administer, being a burden, difficulty staying fit, hormonal changes, and inadequate treatment. The top three were that they were afraid of incapacity to self-administer medication, afraid of possible surgery due to comorbidities, and not being able to get to treatment.

Geographical differences also came through due to systemic situations such as having to travel to get treatment.

Conclusions: Start figuring out what scares you and learn how to prevent it or tackle it when it happens. You are never too young to start thinking about aging. Talk with your health care providers and family about it.

Other discussions about aging with bleeding disorders

Older haemophilia patients now say their peers have caught up and now everyone has a limp.

The important thing is having quality of life and maintaining quality of life. Much of this depends on family and friends.

For bleeding disorders quality of life has a lot to do with joints and muscle strength and there is much that can be done for that. Many issues can be preventable so maintaining healthy habits is important for the long term of life enjoyment.

There are issues you can impact and some you can't. Some things hit you out of the blue – cancer, infection, trauma, etc. But doing what you can in advance is helpful.

We need specialist care and multidisciplinary teams working for this population. Educating young people is valuable to ensure people can stay active in society.

There's not much evidence looking at the differences in aging with men and women and bleeding disorders. There was a study that revealed people have fears about being able to participate and how will aging affect them. Will the health system be able to meet their needs as they age with this condition.

Sometimes we must think outside of the box for what patients' needs are. In the wider population women live longer but have osteoporosis and other issues. Menopause needs more focus too

Physiotherapist perspective

An important area of focus is on physical function and the effects of aging. Many systems can be affected. Encouraging people to stay active is vital. Other chronic diseases can be comorbidities and pain is a big factor in how people function and if they mobilise well. It also can increase the risk of falls. Some forms of arthritis can be more common in women. Fracture risk can be higher.

The fact that we are even considering these perspectives shows just how far treatment has come

Developing countries

There is extra difficulty when aging with a bleeding disorder in a developing country. Oral health is very important, and issues are preventable. Close collaboration between dentist and haematologists is essential. Individuals with bleeding disorders are at increased risk of developing periodontal disease. Poor oral health can lead to systemic issues including cardiac disease. Establishing regular dental check-ups and maintaining oral hygiene is very valuable for maintaining overall health as we age.

Questions from the audience

Biggest fear is ending up in a home at 90 with a bleeding disorder. Why don't we have a gerontologist?

- There is one with haemophilia in Northern Ireland that WFH is connected with.
- Recently had a patient die at 99 in an aged home facility. Nurses and HTC staff would visit him there and community nurses would go to help him infuse although he could still do it himself.
- There are some cognitive issues with aged patients and a gerontologist that could be referred to at least would be useful
- Even at 90 there are things that can help.

The lack of data is an issue for both women and people aging with bleeding disorders. How can we gather the data we need to guide how we treat these groups?

- A data registry is necessary. Trying to find the gaps where we can benefit
- Studies all exclude people over 65. Which is arbitrary

- First step is to, as a community, identify what studies are needed and do them.

What do you think about prophylaxis through menopause?

- Yes, it's a good idea and helps control excessive period bleeding. HRT isn't for everyone so consult with your specialists
- We could tailor the therapy for this short period of life
- Multidisciplinary clinics for women with bleeding disorders is promising for supporting women.

So, what can we do?

- Promote health education on lifestyle and modifiable risk factors
- Enhance protection from risk of comorbidities
- Promote early detection with relevant screening tools and pathways
- Encourage the use of appropriate services
- Develop relationships with primary care services
- Enable and support self-advocacy
- Create individualised care plans
- Support access to treatment
- Share experiences and participate in registries and research that focus on these populations.

Mild haemophilia

BJ Ramsay: A medical perspective

Mild haemophilia accounts for significantly more than a third of patients.

It can be hard to know where to fit in. They may feel like they're not bad enough for camps etc. but not good enough to not worry about it.

Treatment guidelines say people with haemophilia require periodic monitoring and assessment of their condition and circumstances. They should be evaluated at least once a year, but people with mild may need to be seen less often.

People with mild haemophilia rarely bleed, but some do and there are assay discrepancies. They might bleed a long time after surgery, so it's important that you and your HTC plan well enough for this. They may require less frequent monitoring, but it's important to keep track of when you last saw the clinic. It also raises questions about how hard HTCs try to get them in.

Because they don't happen often, there can be issues in recognising a bleed in a person with mild haemophilia. Bleeds can have a slow onset, and it can be difficult to know how bad they are. People are often not sure how to treat a bleed. They may have tranexamic acid at home, but what about an infusion? People are also often unsure about who to tell about their bleed, and whether they need to go to hospital. There is also ongoing concern about how seriously services take bleeds in people with mild haemophilia.

There are issues that can arise from not recognising when a bleed occurs. Because bleeds are less common in people with mild haemophilia, people can be slower to seek help. That means that they might need more intensive or longer treatment and may have more negative outcomes due to longer time bleeding. Worryingly, INSIGHT study data shows a 3.5-fold higher mortality rate from intracranial bleeding in mild haemophilia from the general population.

It is important to remember that inhibitors screening should be performed in any patient regardless of age or disease severity, who is intensively treated, that is for more than 5 consecutive days, at 7 and 15 days, and within 4 weeks of the last infusion.

Women with haemophilia sometimes aren't even considered, and this can lead to delayed diagnosis and delayed access to treatment.

People with Mild Haemophilia have higher mortality rates, so all people with low levels of Factor 8 and Factor 9 should have access to education, diagnosis, consultations and follow up care, and treatment as required.

Dr Sarah Whitaker: A psychosocial perspective

The impact of having mild haemophilia may include having anxiety and depression. This is underreported, and there's a complex interplay between symptoms that are reported by the patient and those seen by clinicians.

People are often accessing fewer services but carrying a heavier burden. They may have a reduced sense of identity with the bleeding disorder community. They may not have had to explain it as much, haven't connected with others who have it, and not had as strong family history. Their burden is often minimised. They may feel that it's "only mild" and should be dismissed, when it does actually have an impact.

- They make more risk assessments day-to-day
- Any decision to treat is more active. Might rather wait and see if the alternative is travelling to the HTC
- Underestimate bleeding and joint damage

Medical services are often allocated based on severity, so you might be seen less often and less well. That has an impact on the relationship you have with your medical professionals. There is also reduced access to variety of treatments, including new treatments. There is also more potential for trauma because you don't know what to expect in any given situation. People may be diagnosed via an injury, and family can be traumatised while this is investigated. Hospitals aren't as familiar, and it may be a bad experience when you go.

People with mild haemophilia often have unmet needs. Their haemophilia knowledge can be reduced because they are not prioritised. They tend to have lower engagement with HTC & community, and reduced access to treatment. This can impact on school and work, relationships and family dynamics, and on activities.

Recommendations from studies include:

- Encourage joining HNZ
- Training for HTC staff

- Audit and update advocacy information, especially about women
- Update existing treatment guidelines
- Make research a priority
- Be proactive in measuring levels in girls/women
- Assess depression and anxiety
- Promote treatment adherence
- Make screening tools available.

Yannick Colle : A patient perspective

Her father had haemophilia and her son has haemophilia, but she didn't know she was a carrier. She was just a woman with symptoms and no explanation for years. As a child she was considered a tomboy who had many bruises and unexplained long nosebleeds. However, there were no explanations.

At around age 11 she needed a tooth extraction, and it bled all night. The dentist didn't have an answer. Her parents were concerned, and she was tested for vWD because "girls can't have haemophilia". Still nothing.

Age 14 brought menstruation. This involved very heavy periods, no comparison with friends, no information, and no communication with anyone about it. She went 6 years before getting the contraceptive pill.

Still without a diagnosis, Yannick had a son. No problem during pregnancy or delivery but high bleeding postpartum and returned menstruation. Still no real answer. Eighteen months later, her son had an accident and was diagnosed with haemophilia A, therefore she must be a carrier.

From age 29-46 she restarted the contraceptive pill and had no more issues other than bruising. New recognition started to happen that there are risks for carriers. Finally, she had an answer – diagnosis of being a carrier with low levels of factor 8.

At 48 she stopped the pill, and the heavy periods came back leading to low iron and anaemia. Her gynaecologist proposed hysterectomy and she went ahead with this but is unsure if this was the only solution. Other options were not explored.

At 48 yrs. old, in 2021, the ISTH recognised haemophilia for women and girls. Being recognised makes a difference. It's now easier to get treatment when issues come up. She's learned to be careful with the word 'mild'.

Yannick started volunteering with exchange group, women's committee, and takes actions with and for women.

Her advice: Fight for diagnosis!

Education & schooling considerations for people with bleeding disorders

Peers, relationships, & building healthy networks

Adolescence is a period where individuals develop their identity. Social media has a significant impact on young people.

Camps and workshops with themes including disease knowledge and management, bullying etc. can be very helpful. Especially using an interdisciplinary approach and a variety of resources like video, card games, role play, psychodrama, designing a flag, etc. Including siblings and friends encourages communication.

Issues vary from age group to age group. For example:

- Age 10-14: Knowledge of the disease, management of bleeds, parents' overprotectiveness
- 15-19: Sexuality, haemophilia as a handicap, social violence, world work, insecurity, drugs

Bullying can impact all age groups. Someone can be the harasser, harassed, or a passive witness. Often due to condition, gender, or skin colour, bullying can take a variety of forms, including cyberbullying, ghosting, body shaming, etc. Self-esteem can be impacted by issues stemming from bleeds and body changes as a result.

Asking young people whether they would share good news and with whom, and bad news and with whom, shows who they feel willing to talk to about issues. It also helps to understand that social media may be skewed as people only want to share good things about their lives.

Having a bleeding disorder leaves a mark in different areas of life and has consequences on personal relationships.

Patricia Cabre: Academic and quality of life outcomes for PWBDs

The intention is to take care of the whole person and not just their haemophilia.

In 1994 a home study was done of all minors with bleeding disorders in Catalonia to find out the first-hand reality and needs of the families. Forty-one percent of families were concerned about medical issues, 18% educational issues, 10% psychological and 11% social difficulties. Forty percent of the young people had learning problems, and 26% have repeated a school year. Fifty-three percent of parents were aware of being overprotective.

As a result, they created an education service that can provide a supportive nursing environment enabling young people with bleeding disorders to thrive academically, socially, and emotionally. The premises for this include:

- A holistic understanding of haemophilia
- Individualised care plans
- Access to healthcare professionals
- Empowerment and self-management
- An inclusive environment
- Psychological support
- Educational support
- Emergency preparedness
- Family involvement

In practice these services attend to the educational issues that families raise. They provide information, advice, and

guidance to schools, while undertaking annual monitoring of the educational progress. Young people get individual pedagogical and/or psychological attention, as well as vocational and professional guidance. They also have access to inclusive summer camps.

The services value the input of parent groups, peer groups, and grandparent groups. They have produced publications for families that help guide parents on how to talk to their child about haemophilia through play.

To foster good outcomes for young people with bleeding disorders we need:

- Quality healthcare access
- Education and awareness
- Individualised education plans and accommodations
- Psychological support services
- Peer support and mentoring
- Transition planning
- Fostering collaboration among healthcare providers, educators, families and community organisations.

Jane Portnoy: Finding your best fit for a career

There are some factors that may limit choice and add complexity. There is something to be said for gentle and flexible work, but this should be balanced with the person's passion and what skills and supports they have. The journey to finding fulfilling work can always be difficult. The journey is never wasted though. Experience along the way builds wisdom and helps find unexpected pathways.

A good first step is understanding who you are; quite a challenge when you're young. Teens are often unsure and have lots of pressures on them. The developmental stage leans towards rebellion even without a chronic disorder. Choices can be overwhelming.

Some may try different options or change along the way as they leverage their experiences into new career paths. For example, Max started as an electrician but went back and did a business course and is now production supervisor. He advises that choosing something flexible where you can rest when you need to is good. Paul says careers transform and you can take what you learn along the way. Lenny says it's important to have a strong support network from your employer and team.

Pivoting can take hard work, but determination can make you successful.

Find the intersection of what you love, what you can be paid for, what the world needs, and what skills you have.

Challenges

- Exclusions – such as defence and police
- Stigma
- Attitude of people in the industry/employer
- Needing to take time off
- Education
- Physical factors
    - A job that has flexibility that you can modify to do from home or in a sedentary way
    - A job that doesn't add pain or cause damage
    - Safety, to reduce risk of traumatic injury
- Pain

To make good decisions about their career, people need to have the opportunity to find out, try out, and decide what they are interested in. They may find a mentor or a clear area of interest. They also need clear and pressure free space while they consider their options. Sometimes bad decisions need to be made to allow for growth, and it's important to support them through it.

The benefit of choosing a rewarding career is developing a robust self-esteem, and the satisfaction of feeling fulfilled.

Parenting with purpose – paediatric issues

Parenting a child with a bleeding disorder can be a challenge. It can be difficult to plan anything, and it can feel like you're constantly watching for danger. Children struggling against their treatments just adds to the challenge. A good way to handle this is to empower the child by giving them options. Do you want to have treatment now or five minutes from now? Do you want it in your right arm or your left arm? The child feels they have some power in the situation.

Remaining positive in front of the child is helpful. Don't be negative where they can see or hear it.

When advocating use the emotions of your story covering real-life data. Connect with others and get involved. Learn everything you can.

Your family is your power. Parents are power. You can change the world for your child.

Haemophilia concerns the psyche and impacts the whole family.

There are great challenges in the first years, and it's a process that builds to acceptance. Psychological support is an important part of the comprehensive care for people with haemophilia.

There are questions about how and when to address haemophilia issues. In early childhood people are exploring and gaining motor skills. They have non-verbal reactions and magical thinking. They react to non-verbal cues from those around them. Play is the universal language for children. Symbolic play can allow processing of elaborate traumatic processes and conflicts. You can work with this to help them understand.

Adolescents have very changeable emotions, and they're learning to manage them, so keep that in mind when dealing with them.

Haemophilia impacts the whole family and siblings should be included in counselling, given information, and be allowed to express their emotions about it.

Clinical cases:

- Peter has a song to help him when it's time to treat.
- Yasmin is eight years old and is overprotective of her brother. She wrote a story to convey her feelings.
- Louie is ten years old. He had inhibitors and had to rest lots and got depressed. He was struggling, since he didn't know much about haemophilia. Educating him about it and giving him the chance to explain it to his teachers made him feel more in control and less uncertain about what is happening to him.

For children to accept the condition parents and caregivers need to do so as well. Working through uncertainties and fears through family workshops/camps/therapy/support groups etc. is extremely valuable.

Some ideas to conclude:

- Including children with the condition and their siblings favours the working through process
- Talking to them about bleeding disorders in a way that considers their age and fears is essential
- Motivating them to take a responsible role collaborates in the construction of healthy self-esteem that involves self-acceptance
- Identifying and communicating emotions helps reduce anxiety

Marlene Bijlevelt: Venous access skills in the area of novel therapies

Traditional treatments have always been via intravenous infusions, but novel therapies now aim to address the underlying genetic cause and utilise alternative mechanisms to achieve haemostasis. Subcutaneous injections are less invasive and more convenient than IV.

But if you're not using IV for prophylaxis what happens when you get a bleed? Patients may lose their skills for accessing veins, or not gain them in the first place. When out of practice, it can be easy to get flustered when a situation arises.

So if it's valuable to have the skills when they're needed, how can we help them maintain them?

- Regular assessments
- Practice sessions at camps etc.

Anxiety and pain could be due to fear of needles, lack of trust, environmental factors, etc. So, for less traumatic infusions:

- Take your time
- Empower the patient
- Listen to the patient
- Seek assistance when needed
- Get the patient involved
- Reassure patient
- Praise patient
- Don't diminish fear
- Never use needle as punishment
- Identify triggers
- Prepare
- Calm environment
- Distraction with iPad, comfort talk, music etc.
- Language focus – avoid words that evoke negative feelings like pain and stinging. Use words that focus on what you want to achieve such as comfort and relaxation
- Make use of suggestions aimed at a positive outcome
- Always mention that they did well.
- Nonverbal communication is important
- Changing focus – use a balancing bird on their finger and tell a story
- "How can I help you so it will go faster?"
- "You can help me by keeping your arm still so I can do my work well"
- "You will notice that you start feeling better as you keep breathing in and out"
- Don't say it's going to hurt or counting down to the shot. These just raise stress

If you want to keep IV skills, ask to do your own blood tests when clinic needs labs done.

HNZ Period Project

Haemophilia New Zealand is excited to launch our period project!

In recent years there has been a movement in the bleeding disorders community for stronger recognition of the impacts that bleeding disorders have on those who have periods. This also reflects a global movement to recognise period products as necessities rather than luxuries!

As an organisation, we think that it is unfair that having a period costs so much. Through our surveys and discussions with members we know that having a period is even more expensive for those with bleeding disorders and can impact the way that we work, live, and play. We want to do something about that!

First, a bit of background...

Way back in 2017 an HNZ member asked us whether there was any support available to help pay the additional costs of sanitary items, like tampons and pads, borne by people who are carriers or who have diagnosed bleeding disorders.

For people who experience excessive or prolonged menstrual bleeding, buying sanitary products can be many times more expensive than for those with a cycle that falls more within the normal range. However, it can be difficult to determine how close to 'normal' individual person's bleeding actually is. There is stigma around discussing menstrual issues, and people with bleeding disorders, or who are carriers, often have no idea that they are anything other than normal.

In response to our member's query, HNZ determined to find out more about our people's experiences of menstruation.

The first step was to design a survey that collected the information we needed. The survey was emailed to 218 HNZ members, who identified as women, and were over the age of 16. Young people under 16 were welcome to complete the survey with parental support (if required), or adults could complete the survey on their behalf, with their permission.

We had a very good response rate, with 71 people completing the survey. Of those, 97% had a diagnosed bleeding disorder or carried the haemophilia gene. The results were confronting!

Here's a snapshot:

- 87% reported excessive bleeding (menorrhagia)
- 76% reported they had been affected by menorrhagia for at least a year, with 64% saying they had been affected for more than 3 years
- 75% categorised their bleeding as severe or very severe
- 82% reported using more than one sanitary item at a time
- 60% of these people reported needing to change their sanitary items at least every 2 hours at peak times
- 62% reported that their periods usually lasted longer than eight days each month.
- 46% reported that their higher-than-average use of sanitary products has contributed to financial stress or hardship in their whānau.

Fast forward to today.

HNZ Board members Lauren Phillips and Tineke Maoate have put their heads together to come up with a way to support members with the cost of menstrual products.

Lauren and Tineke are women, mums, and people with

bleeding disorders who have had enough of bloody periods (pun intended). They have set up a pilot project to see how many of their fellow menstruators would benefit from having period products provided for free, so that they can build a case for all HNZ members who have periods having free access to basic needs.

Thirty people have volunteered to participate in the pilot project. They've been surveyed to determine their experiences of menstruation, and whether the data has changed from 2017. They're now able to order from a range of products including period undies, cups, tampons and pads. The products they select are supplied monthly at no cost. Participants track product usage for three out of six cycles, along with how they're feeling etc.

After 6 months, we'll ask the participants to fill out the survey again to see if the products have helped. Lauren and Tineke will also facilitate informal catch ups throughout the project as a focus group, to talk about experiences and challenges and what things may help from HNZ or from their treatment providers.

After proof of concept via this pilot programme, the plan is to roll the project out to all HNZ members who menstruate.

You can read more about the original 2017 HNZ survey here: https://www.haemophilia.org.nz/news-and-research/news/hfnz-womens-experiences-of-menstruation.

In the meantime, if you have any questions, please feel free to email periodproject@haemophilia.org.nz.

All emails and other communications about the Period Project will be kept in strict confidence and won't be shared with identifying details unless your express permission has been given.

We're looking forward to working with members on this bloody awesome project!

Teen & Youth camp 2024

From 1 to 3 March 2024 young people aged 11-17 gathered in Auckland for the 2024 HNZ teen and youth camp (TYC). This event comes around every two years and is a key part of HNZ's psychosocial and educational pathway.

Growing up is exciting, challenging, and often very confusing. When a bleeding disorder is added to the mix the challenges of young adulthood only increase. TYC is an event aimed at for 11-17-year-olds, which uses a combination of education, fun, and peer and mentor support to help young people move to the next stage of managing their condition. The focus is on being well-informed and better able to make decisions about their bleeding disorder treatment, education, work, lifestyle, and relationships. Siblings of people with bleeding disorders are also invited to this camp because living in a household where a person has a bleeding disorder affects everyone.

The 2024 TYC was held at the Sir Peter Blake Marine Education and Recreation Centre (MERC) at Auckland's Long Bay. This is a fantastic venue, with good facilities, which offers a wide range of supported outdoor activities. For this event MERC offered our young people the chance to learn how to get out on the water safely, some tips for survival in the wilderness, and some fun with projectiles. The MERC staff were fun and skilled. They ensured that our young people had a great time and learned a lot.

This was a shorter camp than usual, just two nights. That meant a very full programme indeed.

On day one, everyone arrived at MERC via bus from the airport. Once we arrived, we took the opportunity for some getting-to-know-you activities on the beach. We quickly allocated beds and got organised for dinner. The first big activity of the weekend was to get into our three teams and build our mascots. For this activity each group was given a raw egg and a bunch of junk. Their task was to make a mascot that could be a part of the group, attend all the activities, and keep the egg intact until the 'autopsy' at the end of camp. This is always a popular activity, and we ended up with some innovative creations.

With time up our sleeves, after making our mascots we headed across to the reserve for a game of spotlight in the dark. This was an excellent way to wind up the day and was enjoyed by all.

After not much sleep, and a whole lot of noise, day two dawned bright and clear. This was to be a very full day indeed, which kicked off after breakfast with Darian's big scavenger hunt. Each team was given a list of 15 things to find in the reserve/beach/camp area. This was a fantastic way for the campers to learn a bit about the area, and to bond as a team. It was great to see the mascots going along for the ride too.

Once everyone was all scavenged out, we took to the beach for a quick-fire sandcastle competition, before heading in for the first education session of the day.

HNZ treasurer Hemirau Waretini joined us after morning tea to talk about growing up in HNZ. He recounted his own journey through events like the national family camp and teen & youth camp, and he reflected on the strength of the life-long bonds of friendship he developed. It was valuable for the young attendees to see first-hand the direct pathway from this event to becoming a leader of an organisation like HNZ. Hemi followed up his talk by running a short workshop on events and activities that may be relevant to this age group in the future. This was a very successful session, and we are grateful to Hemi for offering us his time and insight.

After lunch it was time for the most anticipated activity of the weekend. One of the great things about MERC is the access to the beach and the sea. They also supply equipment and experienced instructors to

allow campers to get out on the water. We were very pleased to be able to don lifejackets and wetsuits, and take to the water aboard kayaks, a giant paddleboard, and a trimaran canoe. This was a great time, which was enjoyed by all. Even better, those who were feeling brave were able to do a bit of snorkelling and admire the fish and kina. This was a long afternoon, but everyone returned to shore excited by the experience.

After some quiet time and some dinner, we were very lucky to have haemophilia nurse specialist Vanessa Minor from the Auckland HTC come in to talk to us about all things BD. Vanessa's talk was full of interesting information, and a quick quiz afterwards showed that the young people had taken a lot on board. It was particularly pleasing to see the young men growing an understanding of the challenges facing young women with BDs. We really appreciate Vanessa making the time for us at this event.

As is traditional, the final night of camp is movie night. This year was no exception, and the crew enjoyed the chance to relax, eat popcorn, and wind down after a long day. It was telling that once we all hit bed it got quiet a lot quicker than the night before!

The final day of camp saw another MERC led activity. We were divided into two groups who swapped between a survival activity, and a projectiles activity. For the first, campers were taught to light a campfire using a flint and available natural materials. The payoff for this was marshmallows at the end. The projectiles activity involved contrasting a catapult and launching tennis balls, as well as learning to make and throw flax darts, known as teka. These were excellent activities that really got the kids thinking.

That was it for activities for the weekend, all there was left was to clean up, and reflect on another successful event. At the final assembly we took the opportunity to thank the fantastic leaders, who had volunteered their time and enthusiasm to support our young people. We also recognised the staff and board members who had contributed, especially Tineke and John Moate who were our camp mum and dad once again.

The final act was to give out graduation certificates, and to award the winners of the top team. Throughout the camp we had been giving out points for good behaviour. When the points were tallied, we found that we had a tie! The only correct thing to do was to hold an elimination game of rock, paper, scissors. Congratulations to Cassie who secured the win for the mighty green team in the most exciting and tense game of rock, paper, scissors ever seen.

This camp would not have been possible without the support of Sanofi and Roche, who provided funding support. Without the participation of external funders like these we would struggle to offer the high-quality events we do. Thank you.

World Bleeding Disorders Registry

The term 'bleeding disorder' includes a range of conditions, from haemophilia A and B to von Willebrand's to platelet disorders and more, which all affect the body's ability to form blood clots. Managing these disorders requires precise understanding and comprehensive data tracking. The World Bleeding Disorder Registry (WBDR) offers a centralised platform to collect, analyse, and share crucial information globally.

The WBDR is a collaborative tool, spearheaded by the World Federation of Hemophilia (WFH), aimed at systematically documenting bleeding disorder cases worldwide. It serves as a repository for patient demographics, treatment modalities, genetic profiles, and clinical outcomes. This centralised database not only facilitates epidemiological research, but also empowers healthcare providers with real-time insights into best practices and emerging trends.

The WBDR is the only global registry collecting standardised clinical data on people with haemophilia (PWH) and people with von Willebrand disease (vWD). The WBDR provides a web-based data entry platform to a large network of participating haemophilia treatment centres (HTCs) to collect and manage their data.

For the past five years, the WBDR has been collecting data on PWH. In early 2023, the WFH launched the WBDR vWD module, making it the first global registry to collect data on vWD diagnosis and management, and on the health outcomes of people living with vWD. WBDR data are invaluable for evidence-based advocacy, research, and improving clinical practice.

New Zealand has a robust comprehensive care system. However, like other nations, we continue to grapple with the challenges of managing bleeding disorders effectively. In Aotearoa NZ there is currently no national database of PWBD, and there is a clear need for a comprehensive nation-wide data infrastructure. The WBDR offers a tailored solution to address the unique healthcare landscape of New Zealand in several ways:

1. Comprehensive Data Collection:

The WBDR enables haemophilia treatment centres across New Zealand to input and access comprehensive patient data in a standardised format. From diagnosis to treatment response, every aspect of patient care is meticulously documented, allowing for a holistic understanding of bleeding disorders nationwide.

For this to occur, it's important that all NZ HTCs participate in the WBDR programme. Next time you are at your local HTC, ask whether they're a part of WBDR.

2. Facilitating Research:

By aggregating anonymised patient data, the WBDR becomes a valuable resource for researchers and policymakers in New Zealand. It provides a rich dataset for epidemiological studies, treatment efficacy assessments, and genetic analyses: driving evidence-based decision-making and fostering innovation in bleeding disorder management.

3. Enhancing Clinical Care:

In the realm of bleeding disorders, timely and tailored interventions can be lifesaving. The WBDR equips healthcare providers in New Zealand with real-time access to best practices and treatment guidelines, thereby optimising patient care and improving outcomes. Moreover, the platform facilitates collaboration between multidisciplinary teams, ensuring that patients receive holistic and coordinated care.

4. Monitoring and Detection:

With the WBDR, New Zealand gains the ability to monitor disorder trends and outcomes on a national scale. This proactive approach to data gathering enables early detection of emerging challenges, such as inhibitor development or treatment disparities, empowering healthcare systems to implement targeted interventions and mitigate potential risks effectively.

5. Patient Empowerment:

Beyond its utility for healthcare professionals, the WBDR also empowers patients and their whānau. By providing access to educational resources, support networks, and personalised health information, the platform fosters a sense of belonging and resilience within the bleeding disorder community, enhancing overall well-being and quality of life.

Patient-reported outcome data are playing an increasingly important role in health care decisions at all levels. The value in providing PWBDs with a mechanism to track and monitor their own health outcomes, as well as in supporting regulatory and clinical decisions, is increasingly being recognized.

myWBDR, a mobile application associated with WBDR, was developed to capture patient-reported data for PWBDs participating in the WBDR.

The benefits of myWBDR include:

- Quick and easy entry of bleed and treatment data
- Track changes in health status through EQ-5D-5L and PROBE
- Provides a feedback mechanism directly to PWH to track their own progress in a visually meaningful way
- myWBDR data are stored in the WBDR database, allowing health care providers to track their patient data, including adherence to treatment and severe bleeds
- Functions offline, allowing data to be recorded without an internet connection
- Eliminates the need for paper diaries

While the WBDR holds immense promise for transforming bleeding disorder management in New Zealand, several challenges must be addressed to maximize its impact. These include assurances around data privacy and security, promoting universal participation among healthcare providers, and bridging gaps in data infrastructure and technological literacy. Moreover, ongoing investment in research and innovation is essential to harness the full potential of the WBDR and advance the field of bleeding disorder care.

In an era defined by data-driven healthcare, the World Bleeding Disorder Registry emerges as a key tool for individuals living with bleeding disorders worldwide. Its tailored application within the healthcare landscape of New Zealand promises to enhance patient care, drive research and innovation, and foster collaboration across borders. By embracing the WBDR and leveraging its capabilities, New Zealand stands poised to improve the lives of thousands affected by bleeding disorders and pave the way towards a healthier, more resilient future.

WFH Global Forum 2023

The 13th World Federation of Hemophilia (WFH) Global Forum was held in Montreal, Canada, on November 17 and 18, 2023, and brought healthcare professionals, patient advocates, government officials, and industry members from around the world together. Thanks to the support of Haemophilia New Zealand, Deon attended this forum alongside Catriona Gordon (Vice Chair) and Hemirau Waretini (Treasurer).

The gathering covered the latest scientific developments and challenges related to the treatment of bleeding disorders. This year, the WFH Global Forum and the Gene Therapy Round Table were merged into one event meaning that the experts were together in one place.

There has been an explosion of research and development in haemophilia and other related inherited bleeding disorders since we last connected with the global community. The forum was packed with useful information for our community. Deon shares his take on what we heard:

Challenges and opportunities ahead

While there is much promise and development of many therapies, it is unsurprising that access is inequitable and only reaches some. There are many opportunities for improvement ahead for the community; these include:

Re-classification of haemophilia

Debates on whether re-classification of haemophilia severities is required – This meeting would suggest that the consensus is 'no, not right now'.

Treating rare bleeding disorders

Less than 5% of therapies in development are for rare bleeding disorders – How can research communities be incentivised to focus more on rare?

Sexism in the management of bleeding disorders

Why are women less likely to be included in clinical trials? How does this then impact on the therapies available to women? Why are women living with a bleeding disorder less likely to be recognised by the health system?

Applying gene therapy

Immunosuppression is required. What if you have already been exposed to the viral vector used? Not a current solution for children. How will society pay for it? How long will it last?

Development of therapeutics

'Rebalancing agents', 'monoclonal antibodies', 'novel bypassing agents', 'mimetic therapy', and other terms are now commonplace in any discussion about treating haemophilia A or B, and there are some studies focusing on applying these therapies to the treatment of von Willebrand's disease.

For more than twenty-five years, recombinant and plasma-derived products have been the standard of care in New Zealand. The recent introduction of Emicizumab or Hemlibra for the treatment of haemophilia A marks the first 'factor mimetic' to be introduced to us. Why is it described like this? It is because unlike factor VIII replacement, Hemlibra does not replace this missing factor. Instead, it binds two factors already present in the body (FIXa and FX) together. By bringing them together, FX is activated, and we have a result!

There are a number of these therapies in the pipeline and approved in some places, including studies focusing on lengthening the efficacy of current mimetics or rebalancing agents. Acronyms and names include NXT007, Mim8, Marstacimab, Concizumab, and Fitusiran. Some treat haemophilia A alone; others treat both haemophilia A and B. The exact way these treat haemophilia A and/or B differs in some cases, but the take home message is that refinement and improvement is ongoing.

The therapies in use and those in development have the aim of being used as prophylaxis. Current consensus is that for trauma or surgery, factor concentrates are still required. This area is evolving, as studies are looking at the efficacy of these new therapies for surgical prophylaxis.

World Bleeding Disorders Registry

A presentation about the WFH's progress on this registry highlighted that 119 haemophilia treatment centres are signed up to the registry, equating to 13,120 people with haemophilia in 45 countries. While New Zealand is signed up, this is primarily through the Canterbury and Waikato centres.

Around the branches

HNZ's Branch committees enable all our members to participate in the running of the organisation, and to connect with and support one another. Each Branch runs a number of events through the year, to help educate their local members, to make sure that support goes where it's needed, and to have a little bit of fun. Here's what they've been up to recently.

Southern

Vic had amazing turnouts both down south and in Nelson for at dinners and for home visits as she did her annual roadie. It was great to connect with members from previous visits and others she hadn't met before. It was awesome to be able to introduce long-time members to new members who have the same bleeding disorder as peer support.

Regional camp planning is well underway, and there's a good number of whānau registered. Camp starts on 13 September.

We are due for another committee meeting on Wednesday 24 July. If anyone wants to be added to the email list, contact Vic at vic@haemophilia.org.nz.

Central

Central held a World Haemophilia Day Event at a cafe in Levin. This was well attended, and a great time was had by all.

We have our camp coming up in the Hawkes Bay on 4-6 October. We have over 70 people registered to come. While registrations have officially closed, if you really wanted to get in contact with Ashley ASAP. I am sure I could try to make it. So far we have Deon and Hemi booked to run an education session, our AGM, and a trip to the aquarium and technology museum.

In early December we will have our Christmas event at the Couch House Museum in Feilding, which is home to a collection of tractors, machinery, a cottage, native birds, and trees. There will also be tractor rides, games and activities, and lunch!

We are also seeking people to join our committee so please reach out to Ashley if you're interested: ashley_fowlie@hotmail.co.nz

Midland

Midland is pleased to have a new committee up and running. After a long time with just Tineke and Loren making plans, there's now a committed group working on creating an event schedule for end of 2024/2025.

Midland outreach worker Loren Silva has been working with hospital staff to create bleeding disorder specific resources related to mental health. She has also been connecting with new Waikato HTC nurse Rachelle Fleming, who has taken over from Rosie Davis.

Loren is also keen to see the Piritoto group become more active again. To that end she's interested in hearing from anyone who would like to contribute in that space.

You can contact Loren at loren@haemophilia.org.nz.

Northern

The Northern branch held a lunch at Goode Brothers in New Lynn. It was great to catch up, share some stories, and enjoy a nice meal. We also welcomed Jagjot Dhaliwai to the Northern committee.

We are currently planning our events for the rest of the financial year. All going to plan we are looking at Axe throwing in August, holding our Christmas party at Game Over on 01 December, and having a summer get together early in the New Year.

Axe throwing is evidently a lot of fun so watch out for this invite. Game over was one of the most enjoyed events last year. If you missed this last year, put this date in your diary as it's a great half day of fun.

As always Darian (our outreach worker) is an active and valuable member of our Northern team. Please reach out to him if you need any support, would like to get involved with the Northern committee, or have any ideas on events you would like to see. You can get him on darian@haemophilia.org.nz.

Latest news

Sanofi's ALTUVIIIO shows promise in paediatric haemophilia A treatment trial

The trial met primary and secondary endpoints, indicating no development of inhibitors to factor VIII and ABRs.

July 18, 2024

Sanofi has reported that its ALTUVIIIO [Antihemophilic Factor (Recombinant), Fc-VWF-XTEN Fusion Protein] showed promise in the Phase III XTEND-Kids study in children with severe haemophilia A.

The non-randomised, open-label, interventional trial evaluated the effects of ALTUVIIIO given once a week in previously treated 73 subjects aged below 12 years with severe haemophilia A.

These subjects received ALTUVIIIO prophylaxis over a one-year period.

The incidence of factor VIII inhibitors was the trial's primary endpoint with secondary endpoints examining pharmacokinetics, safety, annualised bleed rates (ABRs) and other health measures.

According to the findings, the trial met these endpoints, indicating no development of inhibitors to factor VIII and ABRs.

Data revealed a median ABR of 0.00, with the estimated mean ABR being 0.61 in trial subjects.

In the trial, 82% of the children receiving ALTUVIIIO experienced no joint bleeds, indicating the treatment's potential to preserve joint health long-term.

ALTUVIIIO was found to be well-tolerated in trial subjects without any adverse events leading to discontinuation of the treatment.

The most common side effects observed were non-serious and included upper respiratory infections and fever.

ALTUVIIIO, also known as efanesoctocog alfa, is approved for use in both adults and children for routine prophylaxis, on-demand bleeding control, and perioperative management.

Sanofi Development global head and chief medical officer Dietmar Berger said: "The XTEND-Kids data validate the connection between high-sustained factor activity levels and improved health outcomes, including joint health. Offering a treatment option that emphasizes effective bleed protection in children with haemophilia can help give families increased peace of mind when their loved ones participate in everyday activities.

"The results are a testament to our scientific expertise and commitment to redefine the standard of care for children living with haemophilia through ALTUVIIIO and our broader portfolio of haemophilia therapies."

Source: https://www.clinicaltrialsarena.com/news/sanofi-altuviiio-paediatric-haemophilia/

ISTH Issues Clinical Practice Guideline for the Treatment of Hemophilia

July 18, 2024

Prophylaxis recommended over episodic treatment of bleeding events for severe and moderately severe hemophilia A and B.

In a clinical practice guideline issued by the International Society on Thrombosis and Haemostasis and published online June 13 in the Journal of Thrombosis and Haemostasis, recommendations are presented for the treatment of congenital hemophilia.

Suely M. Rezende, M.D., Ph.D., from the Universidade Federal de Minas Gerais in Belo Horizonte, Brazil, and colleagues developed an evidence-based clinical practice guideline for hemophilia treatment. Thirteen questions were selected by a multidisciplinary panel: 11 addressed the treatment of hemophilia A and two addressed hemophilia B treatment.

For hemophilia A, the panel addressed questions relating to prophylactic and episodic treatment with factor VIII concentrates, bypassing agents, nonfactor therapy (emicizumab) (with and without inhibitors), as well as immune tolerance induction. Questions on prophylactic and episodic treatment of bleeding events with factor IX concentrates were addressed for hemophilia B. For all 13 recommendations, agreement was reached. For severe and moderately severe hemophilia A and B without inhibitors, prophylaxis is strongly recommended over episodic treatment of bleeding events. Prophylaxis with emicizumab or with factor VIII concentrates is recommended for hemophilia A, and prophylaxis with purified plasma-derived factor IX or standard or extended half-life recombinant factor IX concentrates is recommended for hemophilia B without inhibitors.

"With hemophilia management being complex, our guideline provides clarity, guiding treatment decisions with evidence-based insight, yet highlighting the importance of making treatment decisions consistent with patients' individual risks, values, and preferences," Rezende said in a statement.

Several authors disclosed ties to the pharmaceutical industry.

Abstract/Full Text

Source: https://www.hematologyadvisor.com/news/isth-issues-clinical-practice-guideline-for-the-treatment-of-hemophilia/

RCSI research identifies new method to fight common blood disorder

By Laura Varley

The discovery could pave the way for targeted treatments for von Willebrand disease, which is the most common inherited blood disorder.

The RCSI University of Medicine and Health Sciences has unveiled new research that shows a new medicine, Rondaptivon pegol, could help people living with the common bleeding disorder von Willebrand disease.

The main characteristics of the inherited illness, which are poor blood clotting and excessive bleeding, are caused by underperforming or low levels of von Willebrand factor, a protein that helps blood clot. The symptoms can be serious, for example, during surgery or childbirth, and sufferers often experience heavy blood flow from nose bleeds and menstruation. It is estimated that the disease could affect as many as 1 in every 100 people.

Dr Alain Chan Kwo Chion, a senior postdoctoral fellow at the RCSI School of Pharmacy and Biomolecular Sciences and the Irish Centre for Vascular Biology, explained that currently people with von Willebrand disease treat the condition by increasing von Willebrand factor in their bloodstream, through drug administration, encouraging the release of factor that is stored in cells near blood vessels or by injecting it into their veins.

RCSI's research indicated that Rondaptivon pegol, also known as BT200, makes it harder for immune system cells to bind themselves to von Willebrand factor and remove it from the blood. The end result is that the factor will stay in the individual's bloodstream for a longer amount of time. RCSI's new discovery will clear the way for an increase in targeted treatments aimed at people living with the condition and for other disorders of the blood.

Prof James O'Donnell, from the RCSI School of Pharmacy and Biomolecular Sciences and director of the Irish Centre for Vascular Biology, noted the potential for this discovery to positively impact the lives of people living with blood conditions.

"Our data supports the concept that targeted inhibition of von Willebrand factor clearance pathways represents a novel therapeutic approach for von Willebrand disease and other blood diseases and disorders such as hemophilia A," he said.

"In the case of von Willebrand disease, this novel therapeutic approach may significantly impact the lives of patients who do not respond sufficiently to current treatment or who develop severe side-effects to current treatment."

The research was supported by a Future (FFP) Award and the US National Institutes of Health and involved scientists from BAND Therapeutics, who invented the BT200 drug.

Earlier this year, RCSI partnered with Serosep to find biomarkers that can predict the progression of inflammatory bowel disease to improve treatment options for patients.

Source: https://www.siliconrepublic.com/innovation/rcsi-research-identifies-method-to-fight-common-blood-disorder

Upcoming events

06 - 09 February 2025

National family camp

Ngāruawāhia.

October/November 2025

Adult wellness weekend

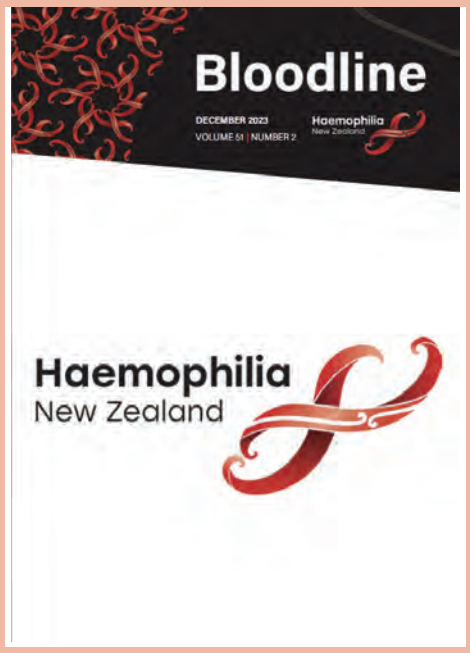
Wellington.

January 2026

Teen & youth camp

Auckland.

Visit www.haemophilia.org.nz for more information on bleeding disorders, HNZ News, and past issues of Bloodline.



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