Bloodline

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THE H WORD

Looking back on 2021, I write with some certainty that no President's report across the country is complete without a mention of COVID-19. As we continue to find ourselves amid a global pandemic, we have continued to adapt. The importance of connection and community have only become more important at this time, and sustaining a sense of community has been a challenge for many organisations, including HFNZ.

HFNZ was fortunate that one national event could take place in the past year, the National Youth Camp held in Christchurch. It was wonderful to gather young people from around the motu for this important event. It was also great to see the impetus getting together has given the National Youth Committee. Some regional events were also held throughout the year, which was an amazing effort by local branches, particularly among pandemic uncertainty.

Two long-term trustees of the Alan Coster Educational Endowment Trust stepped down through 2021. Thank you to Mike Carnahan and once again to Mike Mapperson for your service to this trust, continuing to provide financial assistance for people with inherited bleeding disorders pursuing training and education.

With more time to be introspective, the HFNZ national council reviewed our mission, vision, and values and while a refresh was in order, the core of care, education, advocacy, and support are enduring activities of HFNZ. We have also refreshed our constitution to keep pace with current realities.

17 April was World Hemophilia Day. Across the globe our community took this day to share stories of resilience during the COVID-19 pandemic. This year's theme was 'Adapting to change: sustaining care in a new world'. Many landmarks were lit up red to celebrate this. On a blustery but clear Wellington evening the Michael Fowler Centre lit up red for all locals to see. This was echoed around the country. While not the same as getting together face-to-face, a nod to our community, nonetheless.

As always, I would like to thank HFNZ staff for supporting our community, and thank all volunteers, including the national council, for remaining motivated during what were sometimes the most demotivating of times.

Appreciation must also be paid to all the donors, benefactors, and trusts who contributed to our mission with special mention of our enduring relationship with Kiwifirst, our key fundraiser. Thank you to everyone who contributed. Our mission is not possible without your support.

Finally, I want to wish you all a Meri Kirihimete and a peaceful, restful New Year.

From the CEO

BY SUE ELLIS

CEO Report

By Sue Ellis

As we say goodbye to 2021 and with summer around the corner, it is a chance to reflect on the challenges of what the pandemic of COVID-19 and the delta variant have given us. It seems last year's lockdown was a preview of what was to come.

However, another year of lockdowns and the country living at different levels has not impeded the care and support we continued to provide for our members. Earlier this year we successfully recruited three amazing Outreach Workers who bring to the Foundation their skills, experience and energy providing us with great ideas and resourcefulness. Welcome to Darian Smith in Northland, Laura-Lee Perawiti in Midland and Rosie Maguire in Southern.

Whilst the Auckland and Waikato regions have been in lockdown the Outreach Workers continued to keep in touch with their members via phone calls, emails, Facebook and, in Auckland zooms. For the Central and Southern regions, living at Level 2 was almost life as usual, albeit with care and concerns on safety as they travelled out to meet with members and attend clinics.

Mindful of the impact the ongoing COVID-19 pandemic can have on people's mental health and wellbeing and knowing downtime can be challenging, we have increased our zoom meetings to more regular weekly or fortnightly get-togethers. We have also taken up the invitations that come through with a focus on building resilience and the importance of social support and connecting and engaging with others. Worksafe New Zealand provided an excellent guide on Staying mentally well when working from home, again raising the importance of maintaining social connections. This is something we all know in the Foundation how important it is to keep connected.

In my report last year, I believed we would emerge from the financial impact stronger than before and this year has proved me right. Despite the initial concerns of a drop in donations, this has not been the case and we ended the financial year in a strong position with significant donations and bequests. This has been due to the hard working teams at Kiwifirst and the wonderful kindness of New Zealanders who have donated to the Foundation. I am also thankful to the pharmaceutical industry that has continued to provide the much-needed grants and support.

Webinars have now become the new normal as more people now work from home. We have taken this challenge up and started a series of well-supported webinars with the first a presentation by Physiotherapist Abi Tikkisetty, on the benefits of physiotherapy for joint health. The second webinar was a Q&A for parents of children with mild bleeding disorders, facilitated by Haemophilia Nurse Specialist Vanessa Minor. Further webinars are in the pipeline. We have also attended webinars offered by the WFH, the WFH Global Forum on Research and Treatment Products and the Global Haemophilia Advocacy Leadership Summit, all in the early hours of the morning.

I, and the four Outreach Workers, attended the virtual

Australian Haemophilia conference over the weekend 8th and 9th October. There were some stimulating presentations, including the changing world of bleeding disorders, managing bleeds under current new treatments, sessions on VWD, sexuality, women and girls and where to from here. You can read the reports on these in this Bloodline.

With an eye on safety concerns around COVID-19, we were really pleased to go ahead with our Youth Camp in April, held at Blue Skies in Kaiapoi, North Canterbury. A total of 39 participants enjoyed the Seven Summits programme, which included a visit to the museum to see how mountaineers dress and venture out into the Antarctic. There was also a visit to the YMCA Adventure Centre for Climbing where we witnessed some serious and scary-looking climbing undertaken by our young people!

However, COVID-19 did interrupt our plans to hold a Women's Weekend in Dunedin in November with some of the women locked down in Auckland and Waikato unable to travel. This event has been postponed to June 2022 and we look forward to reconnecting and experiencing a great programme ready to go.

Planning is progressing well for the long awaited National Family Camp, booked for 19– 22 April 2022 at the Ngāruawāhia Christian Youth Camp. We also have a leadership development day booked in for 18th April. Early indications show a high interest from families wishing to attend.

Other plans in place include a weekend workshop for youth leaders sometime in early 2022.

After 20 months of COVID-19 experience, we know the delta variant will be in our lives for some time to come. As this pandemic continues, and how we live and interact has taken a different shape, we encourage members to be fully vaccinated to attend events. We can continue to be strong and resilient through this ongoing uncertainty and being fully vaccinated will ensure we are managing the risk.

As I close off, remember to wear face covering when required, keep scanning in, and keep you and your whānau safe and well if you are travelling over the Christmas and New Year break.

I wish you all a very Merry Christmas.

Changes to the HFNZ Constitution

At the most recent HFNZ AGM, a special resolution was put to members to revise the HFNZ Constitution. This was the culmination of a long period of work, by a number of people. The revisions were to modernise the constitution, while preserving HFNZ's charitable purpose.

Since the constitution was last updated significantly there have been a number of changes in the world, which have affected the way we do business. Many of the 2021 changes reflect this.

The key changes are as follows:

The HFNZ governing body, formerly the HFNZ National Council, is now the HFNZ Board.

Members will now elect executive members of the Board biannually, rather than annually, and not all members will be elected at the same AGM. This allows for continuity of governance as well as safeguarding HFNZ's assets. Furthermore, each executive member will serve for a four-year term.

The individual member representative groups are now Branches, and more structure has been included around the composition of Branch committees.

The AGM and annual reporting will now take place between 01 October and 31 December each year.

Definitions of key terms have been updated for clarity, accuracy, and contemporary relevance. Where possible, language has been clarified, and plain language is preferred. This is important to enable equitable access to all members.

Areas that are covered by existing policies, or where other documents will suffice, have also been amended. More work on updating policies will be completed this year, and the Board will be working with Piritoto to better reflect HFNZ's obligations under Te Tiriti.

You can view the ratified constitution here: https://www.haemophilia.org.nz/files/council/constitution-and-rules-of-hfnz---12-october-2021.pdf

If you have any questions or concerns about the revised constitution, please contact us at info@haemophilia.org.nz.

National Family Camp 2022

Save the date – April 19–22, 2022, is the National Family Camp for families of children aged 0–10 with a diagnosed bleeding disorder.

This year we're heading to a new venue, the Ngāruawāhia Christian Youth Camp. This spot looks great. There are great facilities, plenty of space, and tons to do. That means we can run a really good camp, confident in the knowledge that we'll be able to keep it rolling no matter the weather.

With the Hamilton Zoo just around the corner, we decided on the theme of Animals for this camp. That means there'll be all sorts of animal related activities, and perhaps the chance to meet some interesting animals in real life!

There are two key components to our family camps. One is making an opportunity for parents and children of families affected by a bleeding disorder to connect and support one another. The other is to supply attendees with up-to-date information and education related to living with a bleeding disorder. We have both of these bases covered at this camp. There will be many opportunities for adults and children to gather socially, in a less structured environment, so that they can get to know one another and share some of their experiences. We'll also have some top-notch speakers and educators. These educational sessions will not necessarily relate directly to everyone who attends, however, we will make sure that all attendees have access to content that is relevant to them.

Family camp is always a good time, even if the weather doesn't cooperate, and is one of the most valuable events HFNZ runs. We're really looking forward to getting going, and reconnecting with our whānau from around the country.

Registrations are open now! If you and your whānau are keen, get in quick to secure your place, because spaces are limited: https://nfc-2022.lilregie.com/

National Family Camp 2019

Australian Conference on Haemophilia, vWD, and Rare Bleeding Disorders

The 20th Australian Conference on Haemophilia, vWD, and Rare Bleeding Disorders was held virtually 8-9 October 2021. The event was a great success, with over 30 speakers contributing to a range of stimulating and informative sessions. HFNZ staff attended several sessions, and their reports are included below.

BY LAURA-LEE PERAWITI

MANAGING BLEEDS UNDER CURRENT NEW TREATMENTS: THE IMPACT OF NEW HAEMOPHILIA TREATMENTS ON PEOPLE WITH HAEMOPHILIA, PARENTS, AND HEALTH PROFESSIONALS

This report will give an overview into what current new treatment means from a holistic social point of view and what services are moving towards in 2030.

For children's development, this will help them get back on track via injection once a week rather than several times a week, allowing the child to become more independent. The treatment doesn't work for everyone and there is some hesitancy from patients, which is understandable. We need to work out if the process is right for the patient and if the treatment works well this can take significant pressure of patients/consumers and families. Fewer bleeds allows more activities and quality in an individual's life. Clinicians need to support patients to find out about new treatments, and patients should ask a lot of questions to see if this treatment is right for the individual.

Patient experiences:

- Half-life products reduced the number of treatments significantly, which allowed more activities in his life. Prior to this, he depended on his father. In April 2021 he graduated with a bachelor's degree and is in the process of looking for full time employment.
- Mother of two daughters who have haemophilia: The next generation of treatment has reduced bleeds in one daughter. She has had one plaster cast this year, prior there had been several casts a year. This has given her more independence.
- Extended life factor = one treatment a week now, from three times a week prior. Less product to carry when travelling. Bleeds are now well under control. One bleed in the past three years. Positive improvement in his lifestyle.
- Has severe Haemophilia – Uses the product once every 2 weeks.
- Less treatment to travel with, easier to get tasks done. Use to treat 3 times a week, now working towards once a week treatment. Is now confident to be more athletic.
- Fewer spot bleeds now, which affects mood in a positive way. Frequency of administration reduced. Better for children too, less all-consuming, better for parents to try get children to treat less often.

UNDERSTANDING BLEEDS UNDER NEW TREATMENTS: DR JULIE CURTIN:

- Extended half-life FVIII and FIX – In use now for 4 years.
- Factor VIII - Reduce the number of infusions by 30%
- Factor IX - 60% reduction of infusions
- We were asked to do PK studies on patients in order for us to decide how to manage our patients.
- Despite prophylaxis bleeds may still occur

There are a number of choices of products, such as extended half-life factor and standard half-life factor, when treating serious bleeds. We need to consider the mode of delivery of treatment into the patient. Aim to keep factor between 80 – 100% for the first five to seven days and gradually reduce this over the next 10 to 14 days, then ongoing prophylaxis. Continue to monitor levels over time to achieve appropriate haemostatic levels.

Non-factor products: Hemlibra (emicizumab) functions the same as FVIII. This has a very long half-life, given once a week or once every two weeks. It is hard to know the correct FVIII activity. You don't get peaks, unlike prophylaxis with factor.

What about the patients who don't have inhibitors? All factor FVIII products may be used for breakthrough bleeding events. Dosing should follow the same recommendations as when the patient is on FVIII replacement therapy.

Hemlibra can affect some coagulation tests, so they may seem like they don't have a bleeding problem, however they could have a bleed.

PANEL DISCUSSION AND Q&A: PHYSIOTHERAPY – ALISON MORRIS, NURSES – JOANNA MCCOSKER AND BERYL ZEISSINK:

A physiotherapy perspective

There remains no new treatment that will speed up the natural healing process once an injury or bleed has occurred. We continue to learn as we go, and we now need to look at the real-life data rather than the pharma data. The appropriate treatment needs to be a shared decision between patients and clinicians.

Joint bleeds or haematosis: A 2018 study tracked a joint bleed over time, and from day 5 the pain was reduced however there were signs of a bleed up until day 14. Encourage patient to reframe from usage for the 14 days plus.

Muscle bleeds: There are several phases of healing. The risk of re-bleeding is higher in muscle bleeds. The purpose of rehabilitation is to prevent re-injury

Nurse perspective

You should contact your HTC team when you have anything bigger than a blood test, including dental or surgical procedures, or accidents. To treat traumatic injuries, you will need an assessment at hospital, and management of ongoing bleeding. Factor is required to treat the bleed.

Many children don't know what a bleed is like, therefore require ongoing education on what bleeds are. Children should be encouraged to tell parents or care giver when this happens, and report anything that is out of normal. Early and adequate treatment is critical, and it is important the child recognises a bleed.

Preparing for camp? Have product with you. Ask the child what activities are at the camp and let the physio know prior to camp to plan appropriate footwear etc. With the new products, they may not need to take extra product.

People need to be ready to embrace change and update their health literacy. It is a learning curve to trust the new treatment/drug. Communication is vital between consumers/patients and clinicians.

When you have a bleed, you need that peak level and often need more than one dose of treatment. Keep the patient on whatever factor they are on.

We are looking after the patient's whole wellbeing and not just their bleed. Encourage patient to give all things a go.

PLENARY 4. WHERE TO FROM HERE: TOWARDS 2030

Nurses' perspective

There are currently new therapies under evaluation. The current treatment is standard half-life, extended half-life products, new products in the next 2-3 years. BIVV-001 – new treatment and the data shown to date is encouraging. The future is to have extended, replacement therapy with concentrates.

Non-replacement therapies: There are two types, bispecific antibodies and rebalancing agent. Three different groups now, given once a month via injections.

- We will be seeing a new severe haemophilia patient.
- Need a stock of IV factor to keep at home for acute bleeds.
- One possibility is uber nurses who you can phone and can infuse you with factor FVIII rather than you go to the treatment centre.
- Gene therapy 1.0 – Early type of gene therapy.
- Gene addition therapy.

Gene therapy: Currently has issues that need to be sorted out, and at present children are not eligible. There is quite a lot of variability with patients. There are open questions about the long-term safety of this treatment and the cost is 2.5 million dollars per dose.

- No more than 10% of severe haemophilia patients will have received this treatment by 2023 due to not many countries able to afford the $2.5mil per dosage.
- The delivery of care will be via telemedicine, and this be well established in Australia with some video. We need to have live access to medical notes and be able to access these from anywhere.
- The role of the nurse will be much greater. Nurses will be specialists that can view, treat, examine, and prescribe to patients. Not just haemophilia, but a range of patients.
- Multi-Disciplinary Team (MDT) will be normal, all sitting together discussing patients.
- As patients get older there will be a lot more features of old age to treat.
- Comprehensive geriatric assessment (CGA) review patients together.

How do you choose a treatment?

- This will be based on efficacy, price, convenience, and safety.
- There will be many new therapies
- Change from intravenous to subcutaneous
- Gene therapy establishing itself
- We will still need intravenous therapy
- Haemophilia centres will be larger, covering larger populations,
- Relying a lot more on nurses
- Doctors will not just cover haemophilia but all ranges of bleeding disorders.

Physiotherapy perspective

- We move from treatment for none to treatment for all.
- Physiotherapy will continue to be essential in the future.

Key challenges over the next 10 years is that we will need to monitor comorbidities of joint disease. Key features will be milder bleeding disorders where we now know that joint disease exists. Important to respect the patients perspective, and the overall function of the person. Physical literacy will be important and how we monitor this? To have the ability

to measure movements and joints. At home devices such as home ultrasound. Attaching smart watches to parts of the body. We will need new tools, work is currently being undertaken. Managing pain is crucial and PTSD.

Patient perspective

Allan: One of concerns with gene therapy is that we don't know the long-term effects. Gene therapy, as stated only 10% will opt to do this. I was nine or 10 when I did my first IV, and this was quite daunting. Physio, we have been limited because of Covid. I would like to see more patient and physiotherapy interaction. Be more active at home; be a bit more wary of our joints at home. This can decrease joint diseases. E-commerce is the way to go in the future. Would like more treatment choices.

A parent perspective

Over time, you become more comfortable with what you are doing. Treatment has moved along over the years. Hemlibra has made a significant difference to the quality of life for one child. Our child can provide this to himself once a fortnight. What would you like treatment to look like? I would love it to move to an oral tablet and extend this from every two weeks to once a month. Acknowledge all involved for the movements over the past 11 years.

Woman with VWD perspective

Plasma replacement therapy is current treatment. Administered via IV. Haemophilia is paving the way and showing us options. Extended life products would be amazing. Some anxiety when feeling unwell, would be good to have something easier. HTC team to provide more of a holistic care. Access to gynaecologist who are aware of haemophilia. All clinicians communicating and working together to provide a better care support overall.

Summary:

- Multiple gene-therapy trials going on
- Challenge in the next few years is to adapt to these therapies
- Not many will move into gene therapy in the next decade
- Getting new therapies into the country has not been easy – this will continue
- HTC teams will have to evolve with specialist staff and as an MDT team
- Keep up with disease management
- VWD will continue to make significant improvement such as more products etc.
- Treatment options may remain limited vWD
- Challenges remain with rare bleeding disorders
- We haven't covered platelets very well the hope is we continue work in this area
- Many tools to help us deliver better patient care.

Achieving the vision

Covid will have complications moving forward such as costs of therapy and access. New treatments now compared to convention in the past. As a community, we need to decide what we are aiming for. We need to make sure we address vWD and other rare bleeding disorders. Some new treatments have been approved as safe and effective, but funding and access to them has been delayed due to various processes. HFA are concerned about some of these things and there are ways to contribute to some of these solutions. Hope patients are more involved at important times, ones with real life treatment experience. How to contribute the input in patients so they can communicate what is important to them and the value of new therapy to them. We want to encourage the market to invest in research and products etc., share the cost of funding. Treating to be more than clotting factors, all governments need to be involved to have comprehensive care regarding the national framework, make sure this is understood by all and easier to follow. Have included in legislation the concept of innovation. Embed a best patient practice in treatment. How care can be provided such as comprehensive care and how this activity is implemented into practice, adequately funding to follow this through. Ensure the patient is front and centre.

PANEL DISCUSSION: IMPROVING ACCESS TO PHYSIOTHERAPY

The patient guides the physio intervention. Access is difficult. It is all about training. Patients need to ask their clinician or doctor a lot more when they need the care. Embrace more remote working in the future.

Improving therapy and having funding takes time, this can affect new therapy in the future. Advancement in medicine is quite rapid. It takes time for funding authorities to look into funding because it needs to be safe and effective, and needs to be visible. HFA role in this is to ensure there are various treatments, so patients and their clinicians can choose what is right for them. Have better MDT for women with bleeding disorders. More gynaecologists interest in these areas.

It is an exciting space to see all the treatments. It has to be zero joint damage. We have to be aware that they are not creating normal factor to stop bleeds. Important to continue to monitor patients.

BY LYNNE CAMPBELL

YOUTH – CHALLENGES, TABOOS AND MYTHS

Robyn Shoemark: Clinical Nurse Consultant, Haematology/Haemophilia, The Children's Hospital at Westmead, NSW chaired this session on Youth – challenges, taboos and myths.

- Youth represent 18% of the World's population and, for most, this is a spontaneous and adventurous life stage
- For today's youth technology has changed everything
- The journey for every youth is individualised, goals and milestones have to be reset regularly.

Three members who are affected by Bleeding Disorders illustrated the personal perspectives of this journey:

Growing up: Dale's story

Dale is a 33-year-old male from Western Australia with severe haemophilia A.

He had no family history of any bleeding disorder. Dale didn't ever let his diagnosis stop him, learning from his mistakes along the way. Over time, he learned his limits.

Keen on rugby league (his parents let him try) he soon realised he needed to switch sports codes and changed to basketball where he played at State level.

Dale's teenage years were dominated by basketball. His enthusiasm for the sport meant that he was very compliant with treatment. Arthritic ankles ultimately stopped him from playing basketball or running.

He has lots of tattoos but was talked out of having a tongue piercing!

As Dale progressed through his twenties, he found venous access more and more difficult.

Balancing his career was also gradual, and his eventual direction was informed through trying different types of work. He did various jobs related to building and labouring and even worked as a glazier. He is now a qualified electrician and loves it.

Dale chose to always disclose haemophilia to his employer and workmates. He had not experienced discrimination until recently when two potential employers had not recognised that haemophilia is a very well self-managed condition.

Now that Dale is on non–factor therapy, his overall health and joints have felt better than ever.

His advice is to:

- Treat. Treatment is great and provides the opportunity to normalize potential
- Don't let a bleeding disorder limit your choices
- Don't wrap your child in cotton wool.

Mother and Son journey through teenage years: Heidi & Sam

Heidi

Heidi learned early on that having a son with severe Haemophilia was a partnership with her son and with others. She listened when doctors reminded her that Sam is a boy first. She witnessed him trying different sports and allowed him to work out for himself what he could and couldn't do.

For Heidi, the biggest challenge for her as a parent was prophylaxis. Eventually Sam recognised that to be non-compliant toward his treatment would result in a bleed.

Over the years, a lot of time was spent in hospital, so building a strong relationship with the HTC was crucial.

Heidi's advice to other parents:

- Get to know your HTC staff
- Go to camps and get to know other parents and boys with haemophilia.

Sam

For Sam, as a child, the most challenging thing about having haemophilia was that he couldn't really understand what he couldn't do until he found out for himself.

As a teenager, recognizing that haemophilia is a lifelong condition was difficult.

Haemophilia also informed Sam's decisions around study and work. He started out swimming, then became a swimming instructor and did a Master's degree. He deliberately chose teaching as his career as it was less physical.

Sam readily disclosed his condition to friends and always let his managers know, but at work established himself first.

Sam's advice:

- He strongly recommended that parents let the person try what they want to, so that they can work it out for themselves.
- Attend camps. For Sam the best thing about camps was meeting other people with haemophilia and recognising that there are some who have had to deal with a lot worse that he had experienced. He said that camps made him feel less isolated.

PANEL Q AND A

At the end of the session, a range of topics came through on Zoom.

- Piercings and tattoos - plan it first, treat, observe good hygiene, think twice about piercings given bleeding tendency
- Transitioning from the children's to the adults' hospital can require time and support for both parent and young adult, given the move toward the young adult's private and independent relationship with the Adult HTC team
- HTCs need to allow the patient to explain themselves in their own time, a consultation is not a one way street to tell a patient about their bleeding condition
- There is a need to be aware that, even with good treatment, the patient will need to recognise and know what a bleed is and how to treat it. Equally important is for the child, or young adult, to acknowledge when they are in pain
- Prepare for activities by treating ahead of time
- Family dynamics - Ideally a parent doesn't treat the child with haemophilia any differently to the other children
- Parents naturally become more anxious when a young adult is learning to drive
- Important for parents to provide information to the school about their child's bleeding disorder.

RESILIENCE: HOW TO DEAL WITH CYBER BULLYING AND COMMUNICATION.

Looking after your mental health: Monique Craft

Monique is a fitness instructor, and has been a presenter for *Beautiful Minds* in Australia. She has travelled into schools around Australia speaking to girls about:

- mental health
- media and social media
- sexting
- body image
- friendships, bullying.

She demonstrated and talked through a range of interactive strategies to take one's mind off whatever is creating anxiety in order to build resilience.

To uphold harmony in the home, her top tips included:

- body thump to help with anxiety
- face tapping to ground yourself
- straw breathing to ground us and reduce heart rate
- the STOP strategy - **S**top, **T**ake long low breaths out, **O**bserve surroundings, **P**roceed
- the 54321 Activity (5 things you can see, 4 you can feel, 3 you can hear, 2 you can smell, 1 you can taste)
- the ice technique (stops the adrenalin surge as the ice is the only thing you can feel)
- mantra "peace begins with me"
- colour walk – count the colours you can see on a walk around the block
- write and rip - write down your fears then rip up the paper. To release what is holding you back
- the calm palm to create harmony within yourself.

Monique acknowledged cyber bullying is extremely prevalent now that people are online so often.

Essentially her advice is to treat the people you interact with as you would a guest coming into your home.

- select who you interact with
- no response is powerful
- take screenshots to document what is happening
- report what is happening
- block a bully.

Q AND A

- When in public and feeling overwhelmed, use the colour walk technique to slow down your breathing and move yourself to a quiet spot in the room.
- Online buddy mentoring systems: Requires monitoring to ensure it is safe. Perhaps just a register of people willing to chat is all that is required. (In Australia, some states offer Peer Support Programmes).

BY ROSIE MAGUIRE

WOMEN AND GIRLS

Joanna McCosker: Understanding, recording, and reporting bleeding symptoms in girls and women

Covered:

- defining a normal menstrual cycle
- using a bleeding assessment app
- signs of menopause.

When young women get their first periods it's an anxious time for them, their whānau, and for healthcare. 11-14 years of age is when most girls experience this. It's all about breaking the taboo, and communication free from embarrassment. It's important to define terms: What is heavy? What is a clot? What is menarche?

The young person should ask:

- Is what I am experiencing normal? (This question can be defined by the following questions)
- Am I soaking through a tampon or pad per hour?
- Do I produce clots larger than a 10c piece?
- Am I bleeding for longer than 7 days?
- Does having a period emotionally and physically affect me?
- Do I use double tampons or pads at night?
- Am I soaking through the night?

There are apps and tools that may help, including the 'Let's talk periods' self-bleeding assessment tool, which can help identify if there is a problem, or other track your cycle apps.

Managing life with a heavy period:

- Wear the right underwear and change regularly.
- Tell your teacher and have access to the bathroom whenever you need it.
- Have a period toolbox at home and at school with pads, tampons, clean underwear, clean pants, and pain relief.

Menopause symptoms can include dry eyes, irregular menstruation, sleeping disorders, fatigue, and body temperature changes etc. It can occur from age 30-55. You can still become pregnant while going through menopause.

Ways to help and combat menopause:

- Dress in layers
- Avoid triggers e.g. spicy foods
- Exercise!
- Moisturise regularly as reduced estrogen levels does lead to drying
- Pay attention to how you feel
- Reach out, talk to others, you are not alone.

Dr Mandy Davis – New clinical approaches in managing women and girls with bleeding disorders across the lifespan

Getting the diagnosis right includes:

- Family history of bleeding
- Investigations looking at clotting
- Factor and platelet function
- Reviewed by hematologist

Correct diagnosis = correct management

VWD

There are six types of vWD so it gets complicated. In the future there are plans for a recombinant vWD treatment, genetic testing in bleeding disorders other than haemophilia, and gene therapy in vWD.

Haemophilia has advanced more quickly than vWD. vWD treatment development is slower, but new therapies will increase over time.

Angela Dunford: Gynaecological issues for women and girls with bleeding disorders

There is ongoing fear and uncertainty around controlling the menstrual cycle with hormones. Contraceptive pills haven't changed for many years. Attitudes have changed, however. Skipping periods can have adverse effects if you start on the pill too early, with bone and breast growth deficiency. Two years post menarche is best.

Breaking the myths:

- Periods on a pill are man-made to mimic the natural cycle. There is no medical reason to bleed on the pill.
- Taking the pill will not affect your fertility. Over 90% of women return to periods within 6 months.

Treatment recommended for heavy bleeders:

A Mirena IUD is the best contraceptive treatment for people with bleeding disorders. Insertion techniques are better now than historically, so pre-pregnant women are fine.

BY DARIAN SMITH

THE CHANGING WORLD OF BLEEDING DISORDERS

Session chaired by Dr Ritam Prasad

There have been huge advances in recent years.

Personal Stories:

Three personal stories were shared.

Leonard – Leonard shared his story of having moderate haemophilia and other conditions due to birth trauma. He was initially treated on demand but then moved to prophylaxis. This proved difficult in his younger years because he couldn't infuse himself due to vision impairment and needle phobia. New subcutaneous treatment products have meant treatments are now only once a week and easier to administer so he can do it himself. The combined effect of this has been empowering and enabled him more freedoms, including graduating from university.

Leah – Leah shared her story as the mother of two obligate carrier daughters. She described a huge change in quality of life for her family when long acting factors became available. They no longer have hugely traumatic situations with every bump and school activity.

Bruce – Bruce has been treating himself by prophylaxis since 2000, and required treatments three times a week. However, three years ago he started extended half-life factor and this cut treatments down to once every six days. This has improved the condition of his veins. He considers another benefit to be the much smaller volume of product that he has to carry around, particularly when travelling. The new extended half-life factor has been a huge improvement to lifestyle and fewer bleeds.

The Australian experience and changes in bleeding disorders over recent years – Professor Tran

Replacing missing factor significantly improves outcomes of haemophilia, and prophylaxis has made a big difference. Short half-life factor works, but requires frequent intravenous injections, which makes adherence to regular treatment difficult and increases the risk of developing inhibitors. Trying to treat inhibitors is expensive and not always effective. Adherence to prophylactic treatment in severe haemophilia is lower than it needs to be for best results due to the difficulties and the impact on quality of life.

New developments include extended half-life treatments, subcutaneous treatments, and gene therapy.

Extending the half-life of factor VIII or IX has enabled them to stay at a higher level in the body for longer, thus reducing the risk of bleeds and allowing a longer time between treatments. Patients using these products showed improved adherence and a reduction in bleeds for both haemophilia A and B.

Hemlibra is a humanized bispecific monoclonal antibody. It is administered as a subcutaneous injection and makes a huge improvement for those with inhibitors, and those without inhibitors. Hemlibra reduces all bleeds. Patients report that they like using this treatment as it's easier to administer and is associated with a better quality of life.

Other novel therapies that look at restoring haemostatic balance are still in clinical trials.

In gene therapy the idea is to use an adenovirus to deliver a genetic payload that enables the body to develop factor VIII or factor IX. Gene therapy works well for a single gene disorder with a clear cause and effect. Replacement therapy is lifelong, inconvenient, and costly, whereas gene therapy has the potential to be effective and to normalise a patient's experience after just one treatment. Factor replacements cause peaks and troughs, but non-factor therapy would provide a more consistent improvement, and gene therapy would provide consistent improvement from one treatment over a long period of time.

How will novel therapies change the multidisciplinary team model? We will need more aged-care, there will be less concern around blood-borne diseases, as these are no longer an issue, and physio will continue.

Therapies for rare disorder patients are being developed.

There are still unmet needs. For example, Factor V deficiency still requires platelet transfusion for severe forms and this has a risk of blood borne viruses.

Are we there yet? The innovative future of treatment for bleeding disorders - Professor David Lilicrap

The clotting cascade uses the various clotting factors plus von Willebrand factor and platelets to create a fibrin clot.

Adjunctive therapies help to mitigate bleeding. For example; Tranexamic Acid to inhibit clot breakdown, oestrogen to increase levels of FVIII, VWF, or Brigogen, or Desmopressin to increase levels of FVIII and vWF. However, there are currently no new adjunctive therapies to help with clotting.

In the early 1960s plasma derived concentrates were used, but from 1985 onwards there have been recombinant concentrates. Since 2011 DNA and antibody based concentrates have become available.

Replacement therapies include extended half-life factors created by pegylation or fusion with other proteins, which enable less frequent replacement of factor because it lasts longer in the body.

Non-replacement therapies include:

- Hemlibra is an antibody therapy that binds to factors IX and X and mimics the effect of Factor VIII. By mimicking factor VIII, but not being factor VIII, it gets around any inhibitors to factor VIII.
- Rebalancing Haemostasis. The balance requires the factors needed for coagulation as well as compounds that prevent thrombosis. One way to balance this in someone who has low factor VIII, for example, is to inhibit anticoagulation compounds like antithrombin or TFPI.
- Gene therapy. One way to do this is an in vivo gene transfer, which is currently undergoing clinical trials for haemophilia. The other kind is ex vivo gene transfer which is when cells are taken from a donor and delivered into the patient where hopefully they produce the missing factor. There is a current clinical trial of FVIII gene therapy via AAV5, which is showing positive results with very little bleeding in the year following gene therapy, and 98.6% reduction in infusion treatments required.

The past two decades have seen an immense impact from innovative science on the treatment of inherited bleeding disorders. We still need to use conventional replacement therapies and adjunctive treatments in some bleeding scenarios. However, non-replacement therapies are now entering the clinic and have the potential to provide innovative treatment options for a wide range of bleeding disorders. In addition, gene therapy has also shown increasing promise in clinical trials and we may see the first licensed product in the next 1-2 years. All these new therapies will require careful and extended follow up to evaluate the potential of adverse effects not yet evident in short term clinical studies.

Overview of the impact of advances in treatment on quality of life and independence and clinical practice – Dr Liane Khoo

Quality of life is defined as an individual's perception of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards, and concerns. How do we measure this? The Haem-A-QoL measures quality of life for patients with haemophilia and covers a range of topics such as feelings, work, relationships, etc.

The type of treatment undertaken has an impact on quality of life measures. Some of this relates to the burden of treatment. For example:

Standard half-life factors – 182 injections a year

Extended half-life factors – 104 injections a year

Potential newer factor replacement – 52 injections a year

Impact of Emicizumab: Studies show fewer missed workdays and a lower number of hospitalisations in those using this treatment. For children, it shows improvement in quality of life for the children and their parents and caregivers.

New therapies are providing improvements in quality of life, but measuring this is challenging. Multiple studies (such as the Probe study) are attempting to capture this.

SEX, SEXUALITY, AND INTIMACY

Presented by Simone Sheridan, Sexual Health Nurse Consultant

It's important to have the awkward conversations about sex and intimacy with patients. Sexuality is part of being human and must be part of a holistic approach to healthcare. Sex is not just about making babies. It's important to talk to patients about how their sexual enjoyment is impacted by their condition. It can be awkward to talk about this outside of a procreation focus.

WHO says everyone should have pleasurable and safe sexual experiences free of coercion, discrimination, or violence.

Humans have a desire for intimacy. This refers to closeness – connection, safety, trust, and love. It can be physical, emotional, or spiritual.

When talking about sex the focus can be very narrow, but it's useful to broaden the definition. It can include any physical contact that brings excitement, pleasure, or connection. Sex is not limited to intercourse and it is not something you "should do" or "have to" do. Keep an open mind about sexual pleasure. The greatest sex organs are the brain and skin so it helps to move away from a genital focus.

The benefits of sexual activity include improved sleep, exercise, intimacy and connection, a sense of wellbeing, fun, improved blood pressure, pain relief, and reduced stress. "If sex was a pill, we'd prescribe it to everyone!"

What can cause difficulties? Pain, medications, clinical conditions, fatigue, societal expectations, fear, depression, anxiety, relationship difficulties, aging. With bleeding disorders these can be more specific – pain in joint bleeds,

medications, anaemia, fear of causing a bleed or pain, physical impairments, or lack of knowledge.

Barriers to talking about sex: Both patients and professionals are afraid to ask! There can be many reasons for this, including embarrassment, incorrect assumptions, a lack of confidence, lack of time or privacy, or worries about causing offence.

Tips for healthcare professionals: Normalise the conversation and provide opportunities to talk about it. Acknowledge the importance of sexual health and wellbeing. Maybe have posters or bring it up yourself. Ask questions that normalise it. For example, "Many people with haemophilia have questions about sex and intimacy – is there anything you might like more information about?"

You don't have all the answers. Listen, validate, find good resources, and refer if needed.

Tips for people with bleeding disorders who want to talk to a healthcare provider about raising these issues: Know that this is an important health issue and you have a right to talk about it. Choose a person you feel comfortable with. Flag it as something you'd like to discuss at the start of a session. Write down your questions, either in an email or bring them to your appointment. Seek further advice if you don't get the information you needed.

Communication is key with sexual relationships! When talking to sexual partners talk about:

- What does sex/intimacy mean in your relationship?
- What activities make you feel connected?
- What's pleasurable?

Plan things together. What's the best time of day (considering pain or fatigue levels)? What about the impact of medications? Position preferences to reduce strain on muscles and joints – maybe think about supports etc. What to do in the event of a bleed.

Resources for parents: Ensure your kids have good info. *Talking the Talk* and *Welcome to Consent* are good resources for this.

Three things you can do after today:

- Learn more! Ask questions
- Practice talking about sex and intimacy
- Health professionals – brainstorm ways sexual health might be better acknowledged
- Get in touch with your local sexual health service.

Some issues that came up in the Questions/ Discussion:

Bleeds in the hip muscle can be an issue, and need to be managed well, but we don't want to put people off sex!

Young people who have good information about sex are actually less likely to engage in early sexual activity.

What are the most common embarrassing questions? Patients are hesitant to admit it, but often they will call wanting an urgent appointment and they won't say why. Often it's because of blood in the urine, bowel movement, or ejaculate. Exploring that, and reassuring them that it's okay to talk about it, can help get to the answers. It might be a one-off, or, if they're older, it might be a prostate issue. Can take some probing to help them talk about it. Sometimes they don't have the words to use so it can be helpful to mention things and ask questions so they can say yes or no rather than having to put it into embarrassing words.

Issues around injury during sex: It might be worth discussing what you're doing, positions, and level of force, and considering what you're trying to achieve. Perhaps a gentler approach would actually be more beneficial for everyone involved in terms of enjoyment.

Small tears in the lining of the penis etc. can cause bleeds. Sexual areas of the body are highly vascular and under pressure, so this can happen. It's important to reassure them that this is normal and can happen and reduce shame about whatever they may have been doing to get that injury.

Sometimes basic sexual education is required. Sometimes undoing the influence of pornography is necessary to correct understandings about sex.

It's important to be mindful of culture, religion, and family culture when talking about sexual issues. Try to normalise sex as a part of who you are and your holistic wellbeing. For the older haemophilia population trauma from HIV and Hep C may affect how they engage or don't engage in relationships and sexual behaviour.

Be open to all the diversity of sexuality.

What advice about masturbation? It's normal for kids to explore their bodies. Give guidance on where is safe to do that. Thinking about simple logistics like using lubricant and making sure fingernails aren't sharp etc. will help keep them safer from bleeding. Understanding your own body makes it easier and safer to engage with a partner.

Positions: Think about your joints and comfort. On your side or back is likely safest. Talk to your physio if you're not sure.

Tranexamic acid can be useful, particularly for bleeds in mucosal areas like the mouth etc.

Is Viagra okay for someone with a bleeding disorder? It's important to investigate the cause of impotence as there can be many reasons for this. Talk to your doctor. Viagra is a vasodilator so check with your doctor first. Some painkillers can be associated with impotence. Other options can be useful for erectile dysfunction as well. Maybe even try a vibrator.

How do we disclose about the bleeding disorder with a partner? Start at the base level of comfort and experience and have open communication about what your ideas are about what it means to be sexually active etc. and practice how you want to talk about this and how to frame it. Get comfortable with talking about it yourself before you talk to your partner.

Check out the *Ask Me Anything* section of the World Federation of Haemophilia website.

12th World Federation of Haemophilia Global Forum on Research and Treatment Products for Bleeding Disorders - 4 and 5 November 2021

BY LAURA-LEE PERAWITI

This report will give an overview of safety issues, observations, studies, and impacts regarding plasma and access, from six key speakers who hold various professional roles and are from different countries.

PLASMA AND ACCESS

Facilitators: Glenn Pierce & Mark Skinner

Speaker 1: Brian O'Mahony

Demand for immunoglobulin increased by 8% per annum between 2010 & 2019. This trend is predicted to continue. By 2026 there will be a requirement for an additional 30 million litres of plasma, which would mean all the current countries currently collecting plasma would need to increase to meet the demand.

Currently plasma mostly derives from:

- USA – 67%
- Europe – 14%
- Asia Pacific 18%

We are seeing some impacts of decreased plasma supply due to COVID-19 and US border issues.

In 2011 we tried to bring all the sectors together to come to some sort of agreement on supply of plasma.

- There was a public sector view and a private sector view, which wanted to increase donors from both sectors.
- Europe needs to collect more plasma and increase donors as they're too dependent on the USA.
- Europe collects more plasma by limiting the types of donors!

There are currently different philosophies:

1. Sufficient quantity of factor should be available to each person with haemophilia
2. The amount of treatment provided will be based on the amount of plasma collected.
3. Who should decide? There are a number of ethical debates happening!

Europe to increase donors by learning from EU countries with high plasma collection, invest to allow plasma from paid donors and minimise waste.

WHO guidance March 2021 – new guidance on increasing supplies published:

- Reduce waste of plasma
- Plasma donations must be carefully considered
- Establish programmes for sourced plasma.

Speaker 2: Svetoslav Slavov

Plasma safety and residual pathogen risk with plasma-derived clotting factors.

- Have evolved over time
- We now have mandatory viral testing, specific viral testing.

Due to improvements, we have reduced risks. We also deal with infectious diseases, which emerge and re-emerge to threaten plasma supply.

Emerging viruses transmitted in haemophilia:

- Japanese virus transmitted via blood transfusions

Key message: The residual risks are shaped by emerging viruses, which can contaminate plasma.

How we know which viruses:

- We use a viral metagenomics process by screening samples
- We can now screen all types of different plasma in order to see what type of virus it is, and how we can manage the infection if there is any.

Metagenomics in blood donor plasma. Viral metagenomics in blood donations with post-donations illness reports from Brazil.

It is necessary to test. Further approaches in Haemophilia metagenomics surveillance and residual risk of acquiring viral infections.

How can we reduce the residual viral transmission?

- Prevention includes active surveillance, detection and strategies for avoidance
- Suitable methods for identification
- How to make decisions that balance risks, costs and safety.

Speaker 3: Thomas Kreil (Takeda) – Safety issues and observations

Plasma availability:

- We do not have adequate access to plasma
- We cannot meet the current need
- Restricted by COVID-19

- We need to be clear that the testing comes back normal.

We have come a long way with virus testing. There have been many advancements in science. Previously it has been important to do inventory hold, where you look back and eliminate. We are now testing NAT – virus genome.

Plasma products: Safety and supply. Science-based risk assessment has differentiated policies;

- Blood components for transfusion
- Plasma products manufactured by fractionation, with virus deactivation and removal
- Assessment may differ.

Science based risk assessment:

- Donors deferral for risky sexual practices – times now changed in early 1980s
- Donation testing
- Virus reduction

Tattoos and piercings used to be indicated as risks. These are now classed as the social norm.

Fractionation of UK plasma and Prion risk:

Mad cow caused a new disease in humans. This has now been managed in humans, and various countries have differing opinions on this. A science-based risk assessment is the resolution.

Speaker 4: Steve Pipe - Access issues with DDAVP

There is a gap and it is affecting care.

The Haemophilia Alliance – Nasal spray (Desmopressin Acetate)

Key points in development:

- Nasal desmopressin acetate is a drug product developed in the 1970s.
- After evaluating all major global nasal sprays, a snap-on nasal spray system was chosen.
- Each vial contains 6 sprays of 150mcg.
- Expiration date 90 day after opening to reduce the risk of evaporation being an issue.
- Room temperature storage.

Next steps

- Available November 19th
- First vials shipped 13/9/2021 expiration date 2/12/2021
- DDAVP intranasal next steps is state approvals.
- Alliance member pricing
- Created a mechanism of tracking for this product so they can trace feedback and information.
- Available from STAQ pharma

Patients are currently using this and it is doing what is expected.

Speaker 5: Dan Hart – DDAVP Intranasal shortage and alternatives

Options are Minirin, Octostim, DDAVP, and Stimate nasal sprays.

The sprays we acquire will not be available until 2023.

European discussion and reflection:

- DDAVP is key therapeutic intervention for large cohort of individuals living with a range of non-severe bleeding disorders
- Can be life impacting
- Interruption of supply intranasal preparation is long
- Low cost, poorly reported, and access under-prioritised in some settings
- High-cost interventions – disproportionate attention. Will benefit a minority.

How many are DDAVP eligible?

- UK national haemophilia database has a wide spectrum
- UK NHS should identify individuals who could benefit
- Search criteria process undertaken from registered patients; under half were eligible.

Variability in access:

- Discussions taking place at European level
- It is not clear why this has not been prioritised widely across the continent.

Speaker 6: Magdy El Ekiaby – DDAVP Intranasal shortage and alternatives

DDAVP is for the management of the following:

- Moderate haemophilia
- Type 1 VWD
- Inherited platelet disorders.

Not popular in eastern world, mostly due to not being available.

A survey conducted – survey distribution and findings below:

- DDAVP of inherited bleeding disorders are not registered in their country
- Lack of availability
- Lack of laboratory testing for evaluation.

Q AND A:

Is STAQ pharma only available in the USA?

- If there are interests from other countries, we could explore this.
- Given the need for GT access it should be available to all. It is a cheaper product.
- We should be advocating for this and challenging equality.

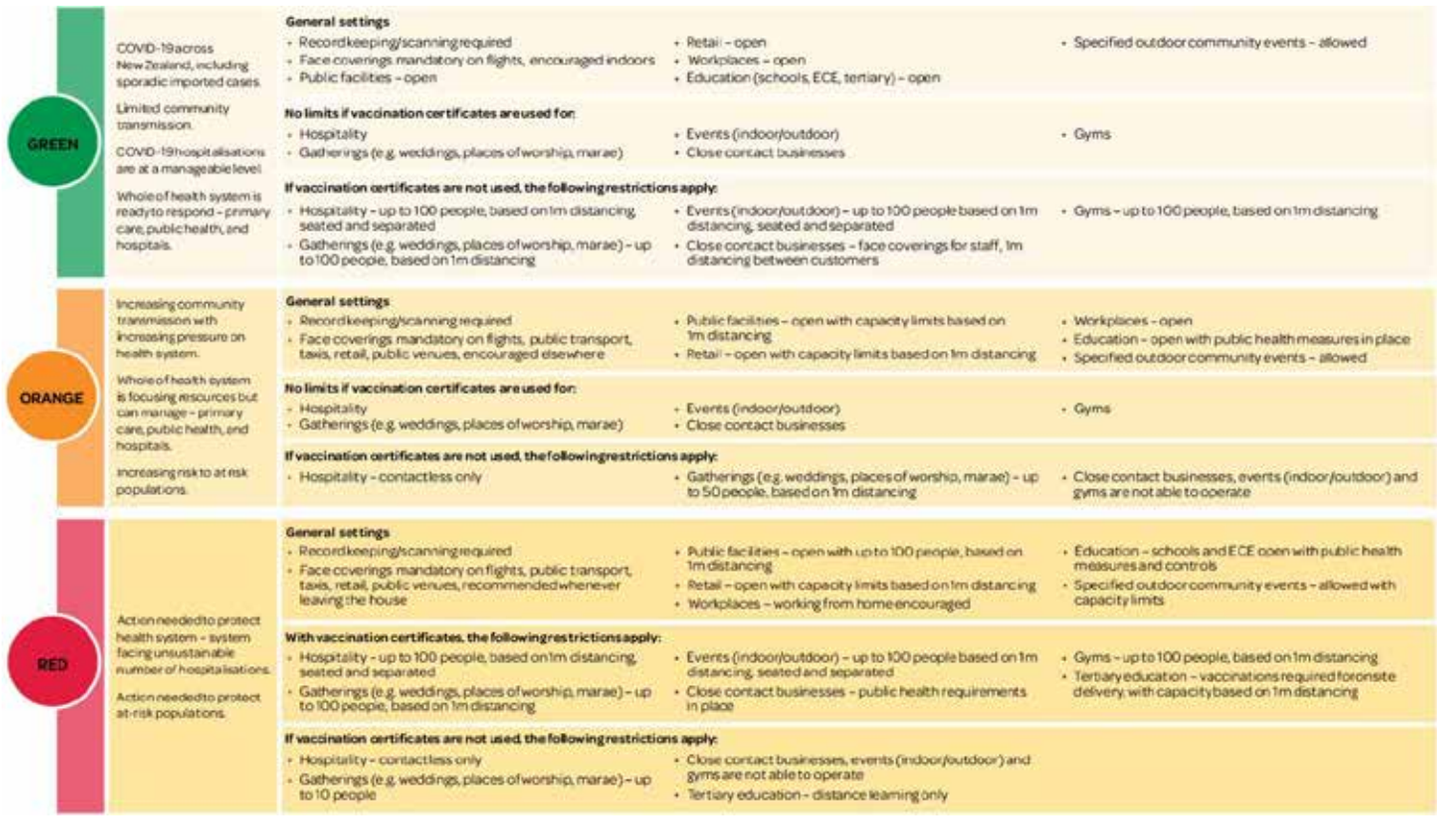
Access issues – can you comment on WFH perspective?

- WFH clearly recognise both sectors are needed (paid and non-paid plasma donors)

Emerging policies and activation techniques: Does this give us confidence in the system?

- We should be expanding the donor pool and carefully monitor the donor variant. We have shown over the past 20 years we have taken care of all infections in donors. We should make use of all donors.

COVID-19 and you



GREEN	COVID-19 across New Zealand, including sporadic imported cases. Limited community transmission. COVID-19 hospitalisations are at a manageable level. Whole of health system is ready to respond – primary care, public health, and hospitals.	**General settings** • Record keeping/scanning required • Face coverings mandatory on flights, encouraged indoors • Public facilities – open	• Retail – open • Workplaces – open • Education (schools, ECE, tertiary) – open	• Specified outdoor community events – allowed
		No limits if vaccination certificates are used for: • Hospitality • Gatherings (e.g. weddings, places of worship, marae)	• Events (indoor/outdoor) • Close contact businesses	• Gyms
		If vaccination certificates are not used, the following restrictions apply: • Hospitality – up to 100 people, based on 1m distancing, seated and separated • Gatherings (e.g. weddings, places of worship, marae) – up to 100 people, based on 1m distancing	• Events (indoor/outdoor) – up to 100 people based on 1m distancing, seated and separated • Close contact businesses – face coverings for staff, 1m distancing between customers	• Gyms – up to 100 people, based on 1m distancing
ORANGE	Increasing community transmission with increasing pressure on health system. Whole of health system is focusing resources but can manage – primary care, public health, and hospitals. Increasing risk to at risk populations.	**General settings** • Record keeping/scanning required • Face coverings mandatory on flights, public transport, taxis, retail, public venues, encouraged elsewhere	• Public facilities – open with capacity limits based on 1m distancing • Retail – open with capacity limits based on 1m distancing	• Workplaces – open • Education – open with public health measures in place • Specified outdoor community events – allowed
		No limits if vaccination certificates are used for: • Hospitality • Gatherings (e.g. weddings, places of worship, marae)	• Events (indoor/outdoor) • Close contact businesses	• Gyms
		If vaccination certificates are not used, the following restrictions apply: • Hospitality – contactless only	• Gatherings (e.g. weddings, places of worship, marae) – up to 50 people, based on 1m distancing	• Close contact businesses, events (indoor/outdoor) and gyms are not able to operate
RED	Action needed to protect health system – system facing unsustainable number of hospitalisations. Action needed to protect at-risk populations.	**General settings** • Record keeping/scanning required • Face coverings mandatory on flights, public transport, taxis, retail, public venues, recommended whenever leaving the house	• Public facilities – open with up to 100 people, based on 1m distancing • Retail – open with capacity limits based on 1m distancing • Workplaces – working from home encouraged	• Education – schools and ECE open with public health measures and controls • Specified outdoor community events – allowed with capacity limits
		With vaccination certificates, the following restrictions apply: • Hospitality – up to 100 people, based on 1m distancing, seated and separated • Gatherings (e.g. weddings, places of worship, marae) – up to 100 people, based on 1m distancing	• Events (indoor/outdoor) – up to 100 people based on 1m distancing, seated and separated • Close contact businesses – public health requirements in place	• Gyms – up to 100 people, based on 1m distancing • Tertiary education – vaccinations required for onsite delivery, with capacity based on 1m distancing
		If vaccination certificates are not used, the following restrictions apply: • Hospitality – contactless only • Gatherings (e.g. weddings, places of worship, marae) – up to 10 people	• Close contact businesses, events (indoor/outdoor) and gyms are not able to operate • Tertiary education – distance learning only	

THE COVID PROTECTION FRAMEWORK

Aotearoa New Zealand has now moved into the COVID-19 Protection Framework (traffic light system) as of Friday 03 December.

Most of the North Island, and all of the South Island are at Orange. Northland, Auckland, Taupō and Rotorua Lakes Districts, Kawerau, Whakatane, Ōpōtiki Districts, Gisborne District, Wairoa District, Rangitikei, Whanganui and Ruapehu Districts are currently at Red.

Find out what the setting is where you are here: https://covid19.govt.nz/traffic-lights/traffic-light-map/

You can find out more about the traffic light system here: https://covid19.govt.nz/traffic-lights/.

VACCINATION AND VACCINE PASSES

At HFNZ we support the COVID-19 vaccine rollout, and ask all our members to be fully vaccinated. We want to make sure that all of our HFNZ whānau are safe and protected from the most serious effects of the virus.

The best way to make a good decision is with good information. There is a lot of unreliable misinformation out there. To learn more about the vaccine, we recommend the following:

- Unite against COVID-19: https://covid19.govt.nz/covid-19-vaccines/
- Ministry of Health: https://www.health.govt.nz/our-work/diseases-and-conditions/covid-19-novel-coronavirus/covid-19-vaccines
- COVID-19 vaccination: Your questions answered: https://covid19.govt.nz/covid-19-vaccines/get-the-facts-about-covid-19-vaccination/covid-19-vaccination-your-questions-answered/
- Information for Māori communities — Karawhiua: https://karawhiua.nz/
- COVID-19 vaccination information — Ministry for Pacific Peoples: https://www.mpp.govt.nz/covid-19/vaccination/

The sooner we are all vaccinated, the sooner we can get to our new normal.

To ensure the safety of members, HFNZ asks you all to be fully vaccinated when attending HFNZ events, such as the Family Camp in April and Women's Weekend in June 2022. To book your vaccine go to https://bookmyvaccine.covid19.health.nz/

Once you are fully vaccinated you can download My Vaccine Pass. This is an official record of your COVID-19 vaccination status for use within Aotearoa New Zealand. It will allow you to access events and venues operating under the traffic light system. You may not be able to access the venues and/or events that you'd like to over the summer, including upcoming HFNZ events, if you do not have My Vaccine Pass.

Find out more about how you can get yours, and about eligibility and exemptions here: https://covid19.govt.nz/covid-19-vaccines/covid-19-vaccination-certificates/my-vaccine-pass/

Regions and Groups

HFNZ's Regional and Group committees enable all our members to participate in the running of the Foundation, and to connect with and support one another. Due to the Covid situation, events have been limited this year. Here's what's been happening in your area.

CENTRAL REPORT

BY IAN REDDIE

Tēnā koutou,

Let me introduce myself, ko Ian Reddie tōku ingoa. I have severe Haemophilia A and have been in and out of HFNZ since the early 80s. I have recently put my hand up to be the national delegate for the Central branch, and I'm looking forward to sharing ideas and information relevant to all of our different needs and requirements.

COVID-19 has certainly made the past two years very challenging for myself, and I am sure for several of you too. I have found the Central masters physio sessions with Helen Dixon at Kenepuru on Mondays, and Hutt Hospital on Thursdays, very therapeutic, both physically and mentally. These sessions are good for catching up with others and working out under Helen's watchful eye!

If anyone wants to get in touch with me to connect with thoughts or ideas about anything to do with haemophilia I would be happy to bounce ideas around via email imreddie@hotmail.com. I only have 48 years experience of living with haemophilia, and I'm happy to share experiences if they can help even one other member!

With Christmas rapidly approaching I want to wish all our members across the country a fantastic holiday season - hope the sun is shining and that you and yours are safe, happy, and well.

NORTHERN REPORT

BY GREG JAMIESON

This is the second year in a row that it's been quiet for the Northern region.

In between lockdowns, we managed to host our Christmas party at the Auckland Zoo. It was a good turnout and nice

to reconnect after a tough 2020. We had lunch together, the sun was shining and I'm sure the animals appreciated the company after a long period of lockdown.

We had organised a Rock climbing event for September but... as happened last year with our Movie event we went into lockdown so this has had to be postponed. All going well we will aim to host this at a later time.

Planning is underway for a Christmas or Summer catch-up. COVID-19 has had a huge impact on a number of businesses. We were in negotiations with Rainbows End over hosting our party there this year. Unfortunately due to the massive financial impact COVID-19 has had they are unable to offer us the rates we have had in the past which makes this venue not viable so we are looking at other options to get our region together.

Northern has also had a change of Outreach worker. Amber has moved to take on a new challenge and Darian Smith has joined us. Our next event will be a great way to meet Darian if you haven't already. Darian is keen to get involved and will be a great addition to our Northern team so please make him welcome.

PIRITOTO

BY TE WHAINOA TE WIATA

Tū ake nei te tira tukuiho

Ngā hekenga mate ā-toto

Ngā morehu o ngā hē kua taka

He kura tangihia mai i ngēnei e ora ana...

Ko te atua taku piringa, ka puta, ka ora

Whakahonoretia te Kiingi Māori a Tuheitia Pōtatau te Wherowhero te tuawhitu

Ko ngāna nei kupu kia amo ake te iwi, ka puta, ka ora.

Paimaarire ki a ia me tōna whare ariki.

Ka aroha e te iwi ki te tōnga o te rā, kīa nei ko te aroha rere ū ana e

Ko te pae ahiahi ki a koutou, ko te pae awatea ki a mātou ngā morehu.

Kei ngaku rangatira o te hekenga ā-toto, tēnā rā koutou katoa...

So, I was thinking about opening and closing this report with something like...

"See you all next year maybe/hopefully, love from te whānau o Piritoto"

...but I don't think that will be allowed or accepted by anyone, so here we go.

I think most would agree that 2020-2021 are two years that need no explaining. However, I am of the thought that when we do that, we allow ourselves to become numb to the reality of what COVID-19 has actually brought to the world and to us, the whānau of HFNZ. The Piritoto whānau, like countless others, have not been active at all. To put it plainly, we dropped the ball and on behalf of our Piritoto whānau I send our most sincere apology to you all. On the other hand, I want to acknowledge our HFNZ staff and those on the Board for providing us with an example of how to move forward in these times. There are always going to be things that we all can improve on and for us as Piritoto we will take those learnings into the new year. So, 2022 for Piritoto...better than 2021.

I would like to acknowledge our Piritoto whānau and committee. As everyone knows, it's hard enough holding down a job and keeping the whānau sane in such a testing time. Yet, with the iwi, hapū, marae, kaumātua and kohanga commitments that all of you have, I am truly grateful for the time that you do give to our whānau and want to thank you all for your continued persistence in trying to uplift our HFNZ/Piritoto kaupapa...tēnā koutou!

COVID-19 has brought a rise in wellness and wellbeing awareness and, now that it's Christmas, the first present we should be getting each other is rest and time to relax. It would be awesome if we just allowed ourselves to do that. On behalf of Piritoto, please have a good safe Christmas and New Year, be kind to yourselves and each other...I want to close with the words of Kiingi Tāwhiao...

"E kore tēnei whakaoranga e huri ki tua o aku mokopuna"

This way of life will not stretch beyond my grandchildren

Kia kaha tonu mai e te whānau, if not for ourselves then for our mokopuna, Paimaarire

MIDLAND REPORT

BY TINEKE MAOATE

A big Christmas greeting to everyone. There seems to be a tiny light at the end of this year with our borders going to be open very soon. I hope that this will make it possible for us to reconnect again.

Even though we haven't been able to meet up, I'm sure we are all in each other's thoughts. Remember that we are only a phone call or an email away if you ever need us.

Please everyone have a safe and happy Christmas and New Year. Looking forward to seeing you all really soon, and for some normality to set in again.

Take care, from the Midland committee.

SOUTHERN REPORT

BY JAMES POFF

As we come to the end of another year, we find ourselves firmly in the middle of what is often the busiest time for us! It also provides a perfect opportunity to reflect on the past year and to put forward some thoughts for the year ahead.

As previously reported, the Southern branch started the year with an interim committee. However, following an AGM a refreshed committee was elected and started the process of reconnecting with the Southern members, aided by the appointment of a new Southern outreach worker, Rosie Maguire. Rosie has quickly immersed herself in the region and her enthusiasm is great. So, welcome Rosie, and we look forward to continuing the journey.

As with the wider community, we have all had to work, play, and connect with family, whānau and friends within the restrictions that have been a consequence of the COVID-19 pandemic. It is fair to say that, like a lot of groups, we have become somewhat disconnected. A recent example of this for the Southern region was the cancelling of the proposed Christmas party due to lack of numbers.

These are challenges that many groups are facing as the social impact of the lockdown regime affects us. However, there is light at the end of the tunnel (as I work for KiwiRail – I can assure you it is not an oncoming train ☺). The Southern branch, with the help of Rosie, are committed to continuing to provide opportunities to get together and reestablish those ties.

Looking forward to 2022, we're firing up the walking group, and providing more opportunities for catching up with the members.

In the meantime, enjoy the warmer weather, make sure that you take a break and put your feet up, stay safe, connected and be kind to others and most importantly to yourself.

Hemophilia Carriers Voice Need for Better Pregnancy, Childbirth Care

BY MARGARIDA MAIA PHD | SEPTEMBER 3, 2021

Women who are carriers for hemophilia — meaning they can pass the disease-causing mutation to their children — think a more individually tailored, information-rich healthcare approach could improve the experience of getting or being pregnant, a small study found.

"Healthcare professionals could use these insights to adapt their consultations to meet the needs of these women when they are preparing for having children," its authors suggested.

The study, "A qualitative study on the experiences of haemophilia carriers before, during and after pregnancy," was published in the journal Haemophilia by a team of researchers in the Netherlands.

Women are identified as carriers if they have a hemophilia-causing mutation in a gene (typically, F8 for hemophilia A, and F9 for hemophilia B) on one of their X chromosomes. They may have mild bleeding symptoms due to low levels of some clotting factor proteins needed for proper blood clotting, but they do not have a bleeding disorder.

For women wanting to conceive, doctors consider it important that they know of their carrier status to best ensure a healthy pregnancy and a safe delivery.

Genetic counseling is also recommended for carriers, as they could give birth to a child with hemophilia. This "complicates reproductive decision-making and introduces additional choices, including preimplantation genetic diagnosis, prenatal diagnostics and termination of pregnancy," the researchers wrote, adding that few studies have focused on relationship between hemophilia carrier status and reproductive choices.

Through face-to-face group discussions and phone interviews, researchers collected and evaluated the experiences of hemophilia carriers before, during, and after pregnancy.

Their study included 15 women, all hemophilia carriers who had gone through childbirth within the previous five years. The group's median age was 33. Over the first two months of 2020, 11 participated in three focus group discussions, each lasting about two hours, and four were interviewed.

Five main themes emerged: communication by healthcare professionals, lack of knowledge, insecurity, autonomy, and family experiences with hemophilia.

Most of these women believed that each of the four reproductive phases — preconception, pregnancy, childbirth, and postpartum — could benefit from certain improvements. These included timely access to comprehensive information during each phase, and a healthcare approach tailored to suit a family's experience with hemophilia.

"The perceived severity of haemophilia and the coping of family members strongly influences reproductive choices," the researchers noted.

Women also considered that their various choices, such as learning of the baby's sex, deciding to undergo prenatal screening, or terminating a pregnancy, should be accepted and understood by doctors.

"I felt like I had to defend myself that I did not want to undergo that test. And actually, during each conversation with a doctor I was told, you haven't done that test right? Why not? So during each consultation I had to say that again," one woman said.

To prevent women from being placed in this situation, "these decisions, together with general care plans, should be carefully noted and shared with involved clinicians," the researchers wrote.

A main concern during delivery was the baby's health, including the risk of bleeding in both boys and girls.

For the mother, "a detailed delivery plan including prophylaxis and anaesthetic pain management" should be shared among "all involved clinicians (midwife, gynaecologist, haematologist and anaesthetist)," the researchers wrote.

After birth, women felt that diagnostic testing of their baby was stressful, but should be done as early as possible.

"Future efforts to improve haemophilia care should be aimed at constructing accessible educational strategies and information transfer for involved clinicians and families with haemophilia," the researchers concluded.

Source: https://hemophilianewstoday.com/2021/09/03/hemophilia-carriers-voice-need-better-pregnancy-birth-care/

Promising autologous cell candidates identified as potential delivery vehicles for Hemophilia A treatment

PEER-REVIEWED PUBLICATION

While people with severe Hemophilia A can be treated with Factor VIII replacement products two to three times a week throughout their lifetime, this treatment is short-lived, expensive, and not always available. Wake Forest Institute for Regenerative Medicine (WFIRM) researchers are investigating the possibility of using a patient's own cells as vehicles to deliver long-lasting Factor VIII protein to treat their disease.

This study investigates the use of amniotic fluid-derived, neonatal, and adult cells, and is the first in-depth analysis of any of these cell types for their use to deliver the Factor VIII protein. The research team also conducted the first detailed look at the levels of production of Factor VIII and its carrier protein, vWF, inside each of these cell types. The challenge for the researchers was to identify cells with the capability to create fully functional Factor VIII, which is a large and complex protein.

"We focused our investigation on cell types that we deemed to be most relevant to either prenatal or very early postnatal treatment and that could, ideally, come from the patients themselves," said senior author Graça Almeida-Porada, MD, PhD, a professor at WFIRM. "There is an urgent need for more effective, readily available and affordable treatments that provide long-lasting correction."

This study was recently published in the journal Frontiers in Cell and Developmental Biology.

Hemophilia A is a genetic disorder caused by a deficiency in, or the absence of, coagulation Factor VIII, an essential protein for blood to clot. Hemophilia A is an x-linked genetic disease, and thus almost always affects males, and it occurs in 1 in 5,000 live male births. Roughly 20,000 individuals in the United States suffer from hemophilia A, and it is estimated that more than 400,000 people worldwide have this devastating disease, according to www.hemophilia.org [wfirm.us3.list-manage.com]. Although there are several new products available to treat persons with Hemophilia A, still, the majority of people are treated with infusions of expensive Factor VIII products 2-3 times per week for their entire life.

While these treatments have dramatically improved the life expectancy of people with hemophilia A, they are unavailable to nearly 75% of the world's patients, they cost well over $250,000 a year (per patient), and complications can send the price tag to more than $1 million. Moreover, as many as 30% of patients with the severe form of hemophilia A develop an immune response (inhibitors) to the infused Factor VIII protein, rendering subsequent treatments ineffective and placing the patient at risk of life-threatening bleeding events. In addition, and perhaps most important, these treatments are not curative.

The delivery of Factor VIII through gene and/or cellular platforms has, therefore, emerged as a promising approach to provide long-term correction of hemophilia A. Finding better treatments and potential cures for hemophilia is an ongoing research focus for the Porada labs at WFIRM. Almeida-Porada leads the WFIRM Fetal Research and Therapy Program [wfirm.us3.list-manage.com] which pursues basic and translational research to develop optimal prenatal treatment approaches for genetic disorders and other life-threatening conditions. WFIRM is the only institute or center in North Carolina to currently house such a research program.

Approximately 75% of people affected by Hemophilia A have a family history, and diagnosis can occur as early as seven gestational weeks. Almeida-Porada said that decades of research and data from more than 50 clinical transplants collectively demonstrate the "safety and vast potential of cell-based prenatal therapies." Using cells as vehicles to carry the Factor VIII protein is a potentially safe and clinically acceptable approach to correct the disease prenatally, she added.

The team compared mesenchymal stromal cells from amniotic fluid, umbilical cord, and bone marrow. While all the cell types investigated were found to be viable candidates for use as cellular vehicles, there were marked differences in the levels of Factor VIII produced by similar cell types isolated from different tissues. They found that cells derived from umbilical cord tissue, after transduction with a vector encoding a B domain-deleted human F8 transgene, yield the highest levels of Factor VIII mRNA, and blood clotting activity. These cells far exceeded that of HHSEC, which are cells thought to be the body's primary site of Factor VIII synthesis.

"Our results show that, despite their similar traits, these cells have markedly different abilities to produce Factor VIII. As such, these data lay the groundwork for future studies to gain a better understanding of the cellular and molecular factors, and to harness the pathways to drive high level Factor VIII expression in other clinically viable cell types," said Almeida-Porada.

WFIRM Director Anthony Atala, MD, who is also a co-author of the paper, said this continued cell therapy research is promising. "The goal of this work is to be able to provide a long-lasting and curative treatment option for patients with hemophilia A by using their own cells. We look forward to seeing this work progress."

Co-authors include: Christopher Stem, Christopher Rodman, Ritu M. Ramamurthy, Sunil George, Diane Meares, Andrew Farland, Christopher B. Doering, H. Trent Spencer, and senior co-author Christopher D. Porada.

Source: https://www.eurekalert.org/news-releases/928791

Angus MacDonald

Live Life Long and to the Fullest with Haemophilia

Dad lived life to the fullest … he didn't let things hold him back.

Whether it be losing his brother in a sailing accident as youngsters, losing an eye while building something, cancers, having 7 children or living with Haemophilia …. Dad gave it a go.

Dad's generation built stuff and that's what Dad did all his 93 years of life.

Caravans, boats, skis, houses and driveway, land-yachts and hundreds of model airplanes! (Mum said NO to a micro-light!). He led by example, teaching us we can overcome life's hurdles with planning, care and a can-do attitude. With his faith, a loving wife and quiet determination, that's exactly what he did … so can you!

Thanks to the Haemophilia Family for life-long care.

Youth Camp 2021

THE YEAR AHEAD

Jan/Feb, 2022

- Youth Leadership Training
  Wellington. Date and venue to be determined.

19–22 April, 2022

- National Family Camp
  Venue to be determined.

10–12 June, 2022

- Women's Weekend
  Scenic Hotel Southern Cross, Dunedin

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

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