Bloodline

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Haemophilia Foundation of New Zealand Inc.
HFNZ

Bloodline

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Disclaimer: The information contained in this magazine is not intended to take the place of medical advice from your GP, haematologist, or specialist. Opinions expressed are not necessarily those of HFNZ.

The purpose of this magazine is to provide a wide range of accurate and timely information on all aspects of haemophilia and related disorders. Haemophilia is a dynamic specialty and therefore opinion may change or be varied from time to time.

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THE H WORD

As I write this, we are in level 4 lockdown across the country. While the level might be the same for everyone, I know the experience can be quite different depending on your circumstances. If your experience is anything like mine, you may be snacking a bit too much or binge watching a bit too much.

HFNZ has long strived to be an organisation, while having a name starting with 'H', that focuses on all people with inherited bleeding disorders and their family. It's fair to say that we can always do more. You may have seen a survey on von Willebrand's which went out in the last couple of months. We are looking at the results of this and how we can do more for everyone whatever the factor deficiency happens to be.

On the haemophilia A front we note that the Pharmacology and Therapeutics Advisory Committee to PHARMAC has recommended funding of emicizumab for the treatment of severe haemophilia A for people without inhibitors (noting that it is now funded for those with inhibitors). This is welcome news and heading in the right direction. We look forward to the swift implementation of this recommendation.

On the activities front, we were lucky to be able to run a face-to-face youth camp. I heard that the museum visit and the Galileo exhibition was a crowd favourite. Stay tuned for the women's workshop weekend scheduled for November and heading to the deep South (Dunedin) for the first time. HFNZ's annual general meeting is scheduled for 30 October at this point. Planning is also underway for the family camp in 2022. All planning is taking place against a backdrop of lockdowns, but I have every confidence that we will prevail!

Wishing you all the best from my bubble to yours.

Deon York

HFNZ President

From the CEO

BY SUE ELLIS

Since my last report to Bloodline, we now have the full contingent of Outreach Workers. Welcome to Darian Smith as the new Outreach Worker for the Northern region. Darian brings to his role experience as a field worker for both the Muscular Dystrophy Foundation and the Arthritis New Zealand, as well as being a registered Counsellor and an author.

The last two months has been focused on getting the Youth Camp planned and underway. We were very pleased that, at long last, the camp was held from 17 to 20 July at Blue Skies in Kaiapoi, Canterbury. Although registrations were low, we still had 19 young members attending, along with 11 leaders including two in training, and two very helpful partners. Everyone got into the activities with enthusiasm and were very happy to be reconnecting as well as making new friends. With the Seven Summits theme of the camp related to mountain climbing, the rock climbing activity was approached by all with serious focus and a joy of accomplishment, despite looking very scary. A visit to the museum to see the Antarctica exhibition also carried a lot of interest by everyone.

I want to highlight the work that Leanne, Phil, and Rosie did to ensure the camp was well organised and well managed. In particular, Phil stepped up to provide direction when needed, and, although Rosie was sick over the weekend, she had organised the activities so well that they ran very smoothly. As the first camp they have attended, our newer Outreach Workers Darian and Laura-Lee confidently ran well-received sessions, getting all the youth involved with their group activities.

We are always mindful as most of the country except Auckland at Level 4, we continue to operate under Level 2 of Covid-19, we continue to plan upcoming events for members. We have Women's Weekend on 12 - 14 November in Dunedin, and the National Family Camp in April 2022 at a venue to be confirmed.

Industry have continued to support our events and other projects with grants from Takeda, Roche, Sanofi, Novo Nordisk, and CSL Behring. Our website has been refreshed and redesigned; we had our first educational webinar offered to members on 25 August, and we are working to get a greater focus to support members with von Willebrands Disorder and other rare bleeding disorders.

Again, a big thank all those very generous people who have continued to support us through their donations and bequests to the Foundation. The Kiwifirst team, as always, have worked hard to keep donations coming in for HFNZ.

With the rest of the world still under great pressure dealing with Covid-19 and its variants, we need to continue to look after each other and ensure we are all safely vaccinated.

HFNZ Youth Camp 2021

BY PHIL CONSTABLE

HFNZ Youth Camp 2021 took place over the weekend of 17 – 20 July, but had been in planning for far longer than that. We first started signposting this event way back in October 2019. Our first date was July 2020, but Covid soon put paid to that. Then we looked at April 2021, but operational issues meant we had to push that back too. We finally settled on July 2021 in Christchurch, and were able to hold the event we'd been talking about for so long.

A winter camp in Christchurch! Sounds cold! However, the Blue Skies venue in Kaiapoi was lovely and toasty warm right through the weekend, and the odd spot of rain didn't get in the way of a great weekend.

We had the leaders come in for some education on the morning the camp started. It was great to have such a diverse group of young people keen to help support the attendees at the camp. The session was very useful, with some excellent contributions from Lauren Phillips and Laura-Lee Perawiti.

Then it was time for the campers to arrive...!

This Youth Camp had the Seven Summits as a theme. This relates to the feats of Chris Bombardier becoming the first person with haemophilia to scale the highest summit on each of the seven continents. It focused on being prepared and resilient in the face of challenges. For our campers that meant learning about some feats of resilience from the past, and practicing meeting challenges themselves.

We watched Chris Bombardier's movie about his exploits, Bombardier Blood, on the first night. That set the scene for our first off-site activity the following morning; rock-climbing at the YMCA Adventure Centre. It was wonderful seeing our young adventurers stepping up to meet this challenge. Not everyone was 100% confident at the start of this activity, but by the end they were all very pleased with themselves.

The outing the following day reinforced the learning they'd already done. We visited the Canterbury Museum, where we were able to experience first-hand the different clothing Antarctica explorers used, explore Hillary's Hut in Antarctica via virtual reality, and tour the Antarctic exhibition. We were also lucky enough to be able to marvel at all the clever things Galileo came up with, in a fabulous interactive exhibition.

These outings were complemented by some excellent educational opportunities back at camp. BJ Ramsay hosted an excellent session on the ins and outs of bleeding disorders, which really got the campers thinking; Kathy Fawcett focused on the benefits of meditation and other relaxation techniques; and the quiz really got them thinking about bleeding disorders and the BD community. Throughout the weekend, campers were also able to learn first-hand about self-infusion and prophylaxis at the morning treatments. It was lovely to see experienced infusers sharing their knowledge with younger, less experienced campers.

The other key component of Youth Camp was physical activity. Central to this was 'Action Time with Rau'. On the first day, leader Raukura Riwaka got the campers out exploring their environment in an orienteering activity. Then, each morning Rau help the campers get their blood moving with a short burst of exercise. He also instituted a points system, which continued through camp, rewarding good behaviour with team points. Congrats to the Blue team, which came out on top.

It was lovely to finally get some of our young people together, after over a year of disruption and delays. The campers all enjoyed themselves, and the leaders did a fabulous job with them.

A special thank you must go out to Tineke and John Maoate, camp mum and dad. Tineke really helped to keep things moving, making sure everyone was fed and watered, and that campers took responsibility for ensuring the campsite was in a decent state throughout.

Thank you too to Roche, Sanofi, and Novo Nordisk for contributing funds towards the camp.


HFNZ Youth Camp 2021
Hunter


and Time
"Measure what is measurable, and make measurable what is not"

Remembering Dr Carter

BY DEON YORK, PRESIDENT HFNZ

On 2 June, our community was saddened by the passing of Dr John Carter. Among his many achievements, Dr Carter set up the blood and marrow transplant unit in Wellington Hospital and the regional haemophilia centre. He was instrumental in establishing the New Zealand Blood Service, co-authoring the Carter Marshall Report, with Keith Marshall, recommending the creation of the national service. In the mid-1990s, he was appointed to the clinical lead role for haematology at Wellington Hospital, a position he held until 2018.

Dr Carter looked after me in the cryoprecipitate, freeze-dried plasma, contaminated blood products, pre-prophylaxis era. Every era has its challenges, and these were just some of them at the time. It was the early eighties, and with no previous family history, my parents had just discovered my diagnosis of severe haemophilia A.

At the time, I believe Dr Carter (I can't bring myself to refer to him as 'John', although I don't think he would have minded) had just returned from overseas to take up a position at Wellington hospital. I recall my late parents both referring to what a breath of fresh air he was, and his keen interest in each-and-every patient who came through the door. In the pre-Internet world, I know Dr Carter let my dad in the hospital library to read up on haemophilia while I was at school so he could be better informed. Dr Carter was an early advocate of moving treatment to the home as much as was possible at that time.

In a consultation with Dr Carter, you were always made to feel in that moment that the focus was completely on you (because it was) and your needs as the person with haemophilia. More than that, Dr Carter helped with everything from treatment to career advice and assisted many times as a referee with scholarship applications.

Despite the challenges of the pre-prophylaxis era, I owe much of my health now to the early treatment and care I received from him.

It goes against reason but given that so much of our treaters' role is to work with us to improve our health I forget that out treaters are not invincible. Attending Dr Carter's service on 10 June, almost exactly thirty years to the day that my father's funeral was held, I remember that Dr Carter was present at the time of loss for our family. In the years that followed, Dr Carter referred to Mum and me as the 'team' in every consultation and no doubt this is something we needed to hear. He continued to treat me up until the age of 18. Dr Carter's reassuring manner and his genuine interest continued to be ever-present throughout my childhood as I negotiated living and living with a bleeding disorder.

Over the years I have seen Dr Carter around as an ever-present figure. He always advocated for us and for a comprehensive and sustainable haemophilia service. I recall the two last conversations I had with him: The first was checking in to see if HFNZ had any funds to contribute to a registrar position. The second conversation was when I was in Laos with the World Federation of Hemophilia. I picked up the phone in the forty-degree heat surrounded by patients and treaters. I can't remember the purpose of the call, but I can remember Dr Carter fondly talking about his time in Laos, including many treks, and a keenness to be involved with any future work in the region.

We will all miss you deeply, Dr Carter. To envisage a world with your absence remains unimaginable.

Obituary: Haematologist John Carter, a lifesaver for many

BY NICHOLAS BOYACK

Associate Professor John Carter: haematologist, teacher and researcher; b September 9, 1950; d June 2, 2021

Linda Gray had a simple message for the family of haematologist John Carter.

“Your husband and father saved my life. I remember our first meeting. He projected calm as he explained the treatment plan.

“His steady demeanour helped ground my husband and me in the whirlwind of a life-threatening diagnosis.

“As my treatment progressed, we came to admire how, under great constraints, he led his team with grace.

“I treasured him as a human connection on my treacherous journey. A great doctor, he was a greater human being.”

The funeral of the 70-year-old doctor and associate professor at St John’s in the City attracted a huge crowd and an outpouring of respect for someone who saved many lives.

John Manley Carter was born in St Louis, Missouri, in September 1950 to Patricia (nee Luke) and Cornelius Peter Van Ness.

They had met when Carter’s father was serving in the marines in World War II. After her husband died, Patricia returned to New Zealand and married Hugh Carter, when John was 5.

Educated at Scots College, where he was head prefect, Carter graduated from Otago University with a bachelor of medicine and bachelor of surgery in 1975.

Over the next 45 years he had a distinguished career in a number of high-profile roles, but one thing that everyone – whether it be a friend, colleague or patient – agreed on was that he did everything with a reassuring sense of calmness and a smile.

Recently retired from the role as head of haematology at Wellington Hospital, he suffered a massive heart attack and died in the hospital where his career as a clinician began, many years earlier.

After working as a house surgeon and medical registrar in Wellington Hospital, he undertook a fellowship at the prestigious Fred Hutchinson Cancer Research Centre in Seattle, Washington, honing his skills in the new and developing discipline of bone marrow transplantation.

On returning to New Zealand, he set up the blood and marrow transplant unit in Wellington Hospital and the regional haemophilia centre.

Instrumental in establishing the New Zealand Blood Service, he co-authored the Carter Marshall Report, with Keith Marshall, recommending the creation of the national service.

He then served on the initial board, chairing it from 2002-04. He also helped establish the NZ Bone Marrow Transplant Registry with the Leukaemia and Blood Cancer Board

In the mid-1990s, he was appointed to the clinical lead role for haematology at Wellington Hospital, a position he held until 2018.

Dr Alwyn D’Souza, clinical leader in haematology who now heads the blood and cancer department at Wellington Hospital, said Carter’s contribution to medicine had been immense.

“John has been a much respected and admired member of the haematology community in New Zealand and was considered to be the father figure of the department.”

As well as being an active physician, Carter was also an outstanding teacher and researcher. He published more than 75 research papers and was regularly awarded the best lecturer award at the Wellington clinical school.

D’Souza said his ability to relate to patients, no matter how difficult the circumstances, was outstanding.

“He had an excellent bedside manner and has served as a fantastic role model for conducting difficult conversations with compassion.”

Although there are textbooks written about how to tell cancer patients bad news, it was a skill that came naturally to Carter.

“Some people just know how to talk to a patient and how to listen, and read visual cues, and John had that in abundance.”

D’Souza remembers his old boss as a “humanist” who was deeply committed to his patients and greatly admired by his colleagues for his willingness to pass on knowledge.

As well as treating the sick, he was also dedicated to finding cures and, as deputy chair of the Malaghan Institute, was involved in cutting-edge research.

Malaghan Institute deputy director Professor Ian Hermans said Carter had an impressive depth of knowledge.

“When asked to comment on anything, his responses were always well considered, clear, and always, always, respectful. There was also the endless enthusiasm – and the grin.”

His advice was particularly valued when it came to complex clinical trials that were not going well.

"When we had issues, I'd often seek out John for advice. Those discussions always ended with a clear plan and a new sense of optimism. I'm sure many of us at the institute have similar memories. He was a mentor to many – we'll miss him a lot."

Malaghan Institute clinical director and fellow haematologist Dr Rob Weinkove described Carter as "a deeply committed and relentlessly cheerful clinician" and an "active and enthusiastic" supporter of research and teaching.

Actively involved in research for decades, his support helped bring the first cellular therapy trials to Wellington and he made a massive contribution to the Malaghan Institute and the wider medical community.

"His commitment to patients, enthusiasm for research, dedication to teaching and development, laconic humour, and sage advice will be greatly missed."

Friend and Scots College principal Graeme Yule said Carter was a much-loved and respected member of the wider school community.

A former head prefect, he served the school in many areas. He was a member of the board from 2001 to 2009 and served as its chair from 2002 to 2009.

After completing his time with the board Carter chaired the Centenary Committee, was a member of the Scots College Society, and served on the Scots Foundation.

Yule said it was a privilege to know Carter and he always valued his advice.

"He was a friend and mentor, John had oodles of common-sense. He was someone you could really talk to in confidence and you would know he understood. He was a bit of a sage who gave sound advice."

Carter would often attend meetings at the school and would say he had to leave the phone on. When he answered a call that was "obviously a life-and-death crisis" and would serve as a reminder to Yule about what was important in life.

Dr Helen Carter says her late husband was a remarkable physician who had the dual focus of trying to find a cure for cancer while providing the highest standard of care for his patients.

She was particularly proud of his pioneering work with haemophiliacs, setting up community-based services to help haemophiliacs cope in their own homes, rather than in hospital.

"He helped them lead a normal life, and got them playing sport and doing the other things they wanted to do."

He was also prepared to speak out for his patients at a political level, including advocating for haemophiliacs who had received contaminated blood.

One of those to benefit from his treatment was Stephen Waring. As a young boy he suffered severe bleeds and was spending up to 10 weeks a year in hospital.

"I was in hospital more than I was at school."

When he and his older brother, Martin, who had the same condition, met Carter it was to prove life-changing.

He taught them how to self-medicate and encouraged them to get on with their lives and not treat haemophilia as an illness.

When Martin was refused a job by the BNZ, Carter wrote to the bank reassuring it that there was nothing to fear.

"The BNZ thought he was going to explode like a grape, and Dr Carter wrote to them and said 'Chill out, he will not die from a paper cut'."

Carter gave the boys his home number and, on one occasion, Stephen had to ring late at night saying he was in a bad way.

Carter responded by saying he would meet him at Wellington Hospital.

"He made a huge difference to our lives and we were like his haemophilia children. It was because of his treatment that I was able to lead a full and normal life."

When not treating patients or overseeing research, Carter enjoyed tramping and skiing.

Never happier than when he was skiing or exploring the Tararua range, he would attend conferences in North America and then head to the ski-field. The Tararuas fascinated him, and he was an active tramper all his life.

"He loved nature and was very interested in conservation and native plants," said Helen.

The Carters had a small orchard near Ōtaki Forks, which they bought to be near the mountain range he loved.

His service to haematology at a national level was recognised when he was made a Member of the New Zealand Order of Merit in 2011.

His funeral was one of the biggest seen in Wellington for some years, with tributes from around New Zealand and overseas.

D'Souza told the audience, which included many leading Wellington doctors, that Carter would have been humbled by such a big crowd.

"Though I think he wouldn't admit it, I hope he feels proud to see how many people cared [about him]."

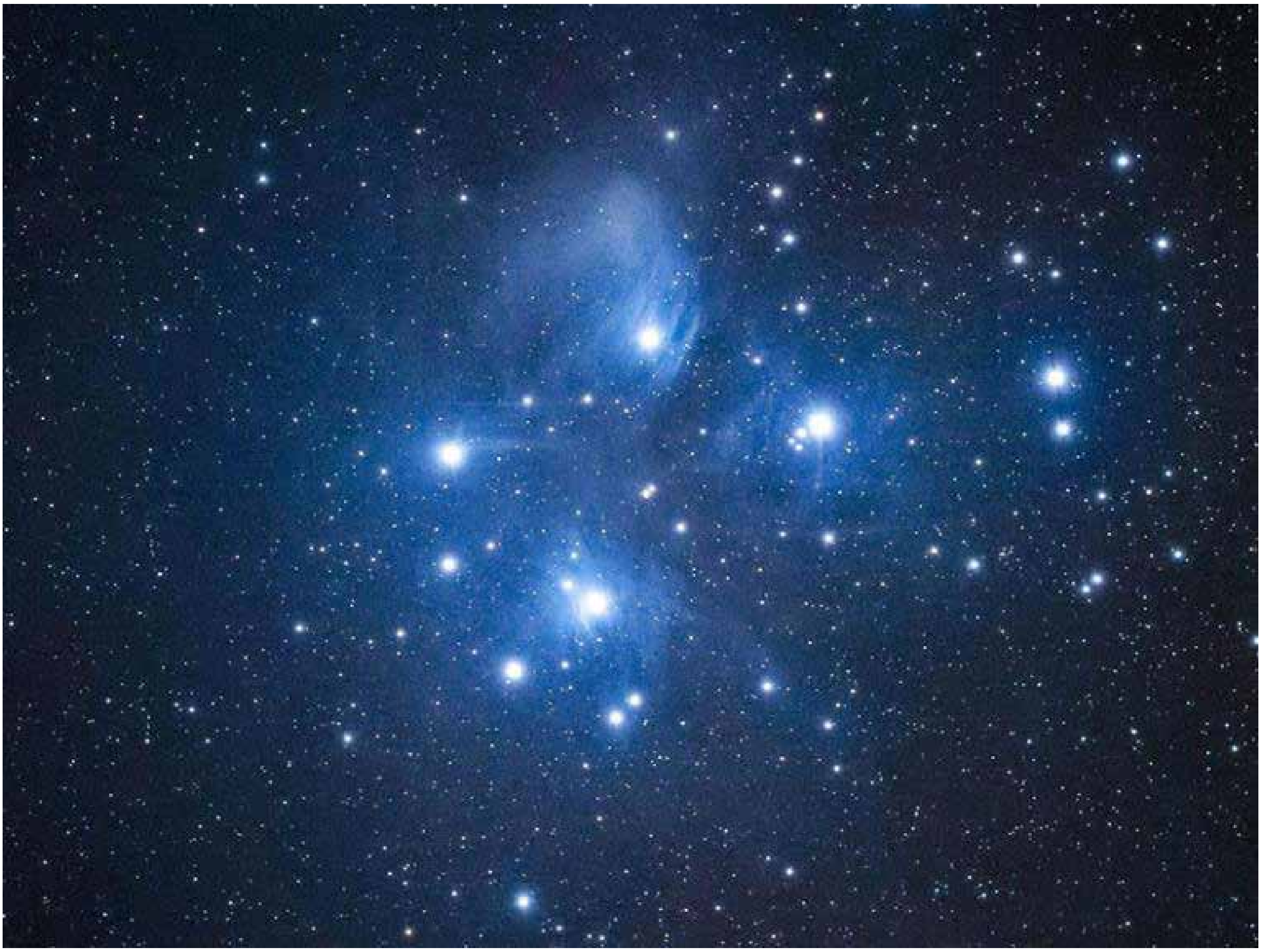
He is survived by Helen and children Rachel, Michael and Peter.

Sources: Dr Alwyn D'Souza, Malaghan Institute, David Watt, Graeme Yule, Dr Helen Carter and Stephen Waring.

Source: https://www.stuff.co.nz/national/health/125471167/obituary-haematologist-john-carter-a-lifesaver-for-many

What Matariki means to me

BY ROSALIE REIRI



Whakanuia a Matariki	**Celebrate Matariki**
Matariki te tipua	Matariki the sacred
Matariki te tawhito	Matariki the old
Tau mai te wairua	Welcome the spirit
Mai i ngā ira Atua	Welcome the life force
Ki ngā ira tangata	To us of the human kind
Haumi e, hui e, tāiki e!	Come together, united as one!

Ngā mihi o Matariki ki a tātou!

Greetings of Matariki to everyone!

The morning of 2 July, I found myself up nice and early at 5am, on a very crisp morning indeed, to whakanui, which means to celebrate, the ushering in of a new year. I have taught myself to find the Matariki cluster, so over the years it has become easier for me to identify Matariki in the sky. I usually find what is commonly known as the Pot, which in Māori we call Tautoru. I follow the three dots of the bottom of the pot, then I scan right to find an ever so bright beaming star, also known as Hinetakurua, who is our winter star. From here, my eyes follow a straight line from Hinetakurua back across to Tautoru and keep in the same line across the sky and in the far left, flickering, is the Matariki star cluster. I was so happy to see Matariki again, and in my heart I reflected on what this time means. I thought about whānau and special people in my life who have recently passed since last year. I thought about the things they have taught me, and the memories they have instilled in my life. I reflected also, with a sense of gratitude in my heart, on what I have in this present moment, and I held a sense of hope in my heart with the possibilities for the future.

Matariki means different things, and has different names, in different regions of Aotearoa. For example, up north they celebrate Puanga rather than Matariki. So, it is really important we acknowledge the local knowledge and histories within the places that we live. I would like to share some of my learnings about Matariki over the years. I have come to learn that each star has its own unique super power …

- Matariki itself is the wellbeing star and is also the name of the whole cluster.
- Waipunarangi is the star who represents the clouds and rain.

- Waitī symbolises the animals in freshwater, acknowledging rivers and streams.
- Waitā symbolises the animals in salt water, acknowledging oceans and salt-water lakes.
- Tupu ā-nuku is the whetū who supports all our vegetation planted in the ground.
- Tupu ā-rangi is the star who supports all the fruits and berries grown in trees, including our birds.
- Ururangi is the star who acknowledges the winds and in particular the strong westerly winds
- Hiwa i-te-rangi is the wish upon a star who makes all your dreams and wishes come true.
- Pōhutukawa is the star that collects the souls that have passed over on a daily basis and she places them on te waka o Rangi. When Te Waka o Rangi descends this Matariki so will all those souls who will descend and then ascend in the sky as a star. In Māori, we call this transition the notion 'kua whetūrangitia'. This is significant, and a reminder to us in the physical world to let go of the emotional ties that we have between us and our loved ones. It is ok to grieve, and at this time of the New Year, it is a great reminder to shift out of the grieving space.

You may be wondering 'I thought Matariki was the seven sisters'? Yes, this is true in different places around the world. In Rome they know Matariki as Pleiades, and in Japan they call it Subaru. In different hapū and iwi there have been up to 13 stars seen and recorded in manuscripts. However, in more recent years, in Māori culture, it is becoming common to recognise Matariki as nine stars with male and female combinations.

Some key learning points:

- The moon gives us days
- The sun gives us seasons
- The stars gives us months

In NZ we follow a Gregorian calendar, even though we are in the southern hemisphere. Our sense of time and the time markers we follow in our national calendars are directives from a colonial, traditional, and western world-view. For example, the Christmas and Easter celebrations. In winter we should be resting and slowing down, yet our main holiday times are during summer, when the days are longer. The sun is out for longer at this time, so we should really be utilising the time to prepare for the winter, yet we don't. Generally, our NZ culture is busy, according to our Matariki timetable, I wonder if she sees us as being out of alignment.

I wonder: Are the significant health issues we see in NZ a result of not following a natural sense of time for the southern hemisphere? How does one align ones self with the environment, to live by the stars, the sun, and the moon?

Matariki is a great time to ask yourself some important questions: What are you grateful for? Who are the people in the past year who have passed on - reflect on what they have taught you? What are your short and long-term goals for your health and wellbeing? What are your personal dreams, hopes, and aspirations? What can you do for our environment?

In conclusion, I am so grateful for our national day to celebrate Matariki starting in 2022, I am really enjoying seeing local councils, schools, and communities come together and share kai and celebrations of a national new year. May we have a wonderful year ahead. May we be kind in our interactions with others and be more mindful at this time of year.

Ngā mihi o te wā ki a koutou e te whānau.

Emicizumab Registry

Emicizumab is a relatively new treatment for Haemophilia, which has transformed the lives of many people around the world. Many of you will be aware of the impact it has had on some of your own members in this country. Some of the stories are truly amazing and it is great to have access to this drug. In New Zealand at this stage only people with factor VIII inhibitors are eligible for treatment but hopefully wider access will become available in time.

What do we want to do?

As this is a new drug, there are still things to learn about it. The clinical trials showed really positive results and I am sure it will become part of our standard treatment for Haemophilia A, but there are still some questions. How do we mange acute bleeds in people taking emicizumab and how do we manage surgery? It also produces some challenges for our laboratories as it interferes with our standard clotting tests.

To help answer these questions, the Haemophilia Treaters from all the centres in New Zealand are keen to collect data on how emicizumab is used. We are particularly interested in getting information about how we manage episodes of bleeding even very small bleeds and we believe it would be valuable to collect information on how we manage surgery in people on emcizumab.

We are at the planning stage of this project. There are a number of steps we need to go through before we can start, but we would like to have your help with the design of the study to make sure it is something you are comfortable with and to make it easy to implement.

Once we have a plan in place, the project will need to be approved by the National Ethics Committee. We can only collect information from you with your consent.

What will we collect?

Our plan is to collect information from you about your emicizumab treatment. This will be stored on a secure database and will only be accessible by the doctors or nurses at your own treatment centre. We will analyse the data anonymously.

We propose to record the following

- Each dose of emicizumab you receive
- Details of any bleeding episodes.
- How the bleeds were managed.
- Details of any surgery and how it was managed.

How will we collect data?

You will be able to enter your own data or data can be added by the doctors or nurses at your own treatment centre. We are proposing to collect the information via a website which will be optimized to use on a mobile phone as well as a tablet or your home computer. This will enable you to enter data anywhere.

We can only store data with your consent.

How can you help?

First we would like to know if you are comfortable with the idea of us collecting this data. We are using the information predominantly to help with treatment in the future which may help you as well as other people with Haemophilia. All data will be kept confidential and secure, and will not be shared with anybody else. The data will be analysed anonymously. It may be published in the future in which case no identifiable data would be included in a publication.

Second, is there any other information that you believe would be helpful to collect? We would like to keep this as simple as possible so you should be able to enter each treatment dose with just a couple of clicks on your smart phone or website, but it may be a good opportunity to collect other information.

Third we would welcome any ideas that could help to make this project as easy as possible.

We are aware that Roche have an app (Hemisphere) to help you manage your emicizumab treatment. Our site will be very similar. We have looked at collecting data in collaboration with Roche, but we would prefer to collect data independently as this avoids any conflict.

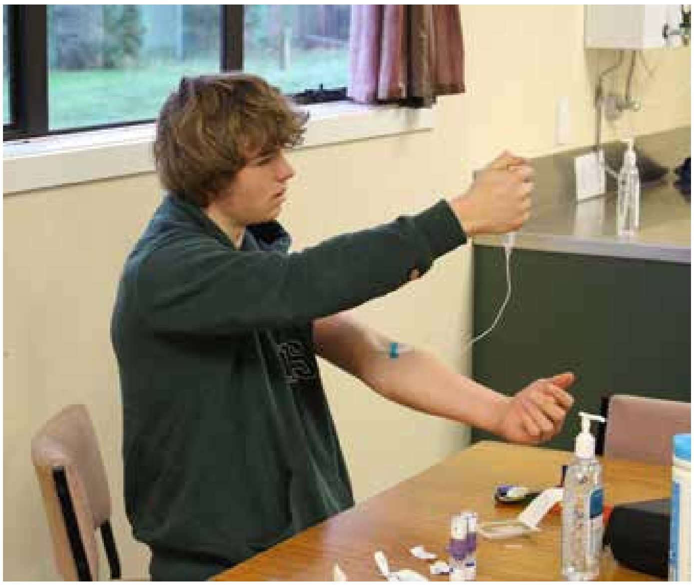
If you have any questions about this project or any suggestions that could help, please contact me. I propose to submit an application to the Ethics committee within the next few weeks.

I have added a few screen shots of a proposed database. If you would like to try the site to give some feedback please let me know.

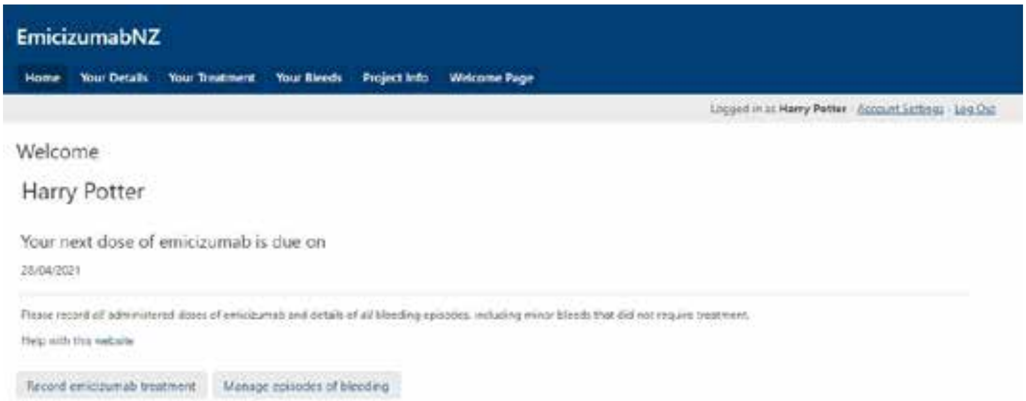
Thanks

Paul Harper, Haematologist, Palmerston North.

Paul.harper@midcentraldhb.govt.nz



Treatment Centre homepage



Welcome

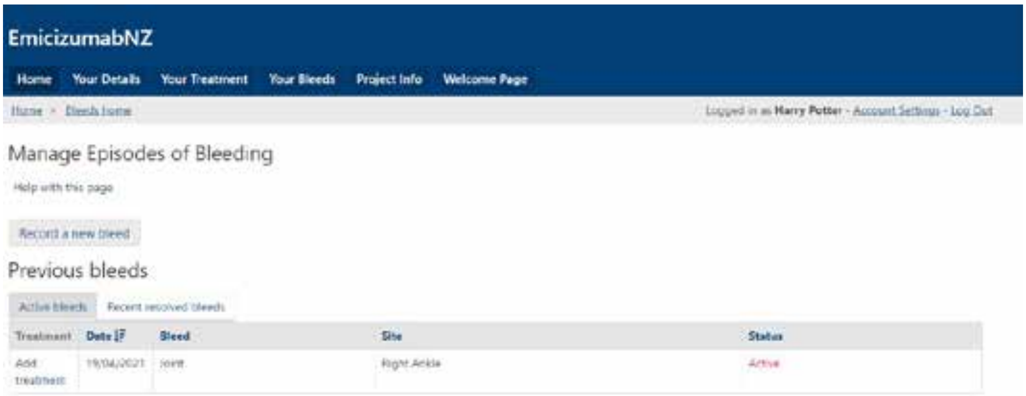
Harry Potter

Your next dose of emicizumab is due on

28/04/2021

Please record all administered doses of emicizumab and details of all bleeding episodes, including minor bleeds that did not require treatment.

Help with this website

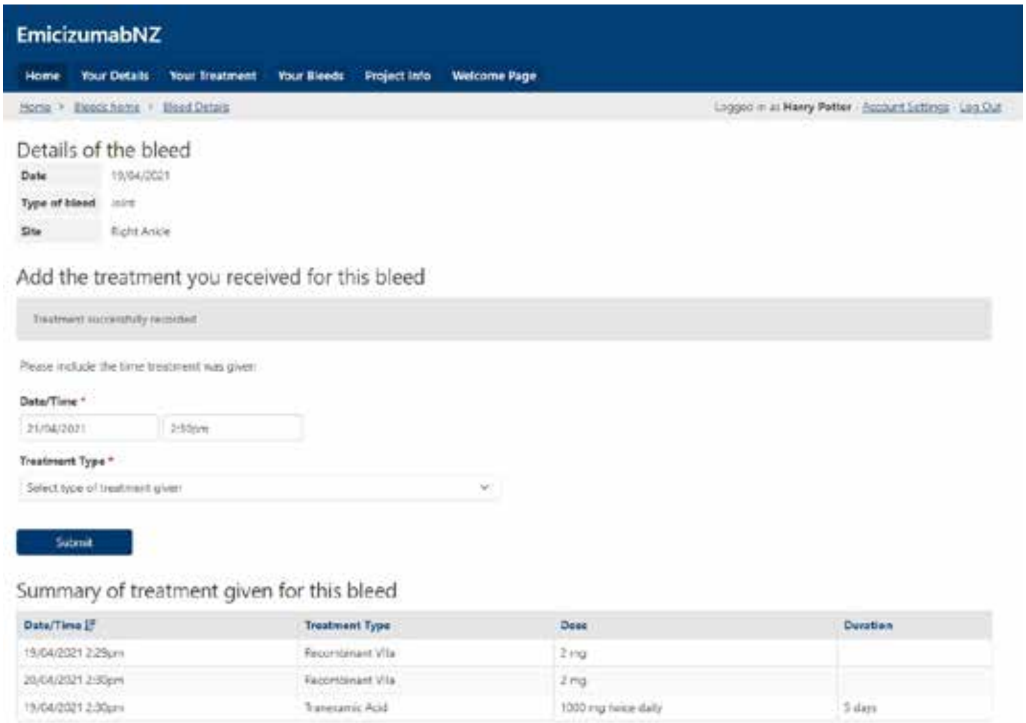


Manage Episodes of Bleeding

Help with this page

Previous bleeds

Treatment	Date	Bleed	Site	Status
Add treatment	19/04/2021	Joint	Right Ankle	Active



Details of the bleed

Date	19/04/2021
Type of bleed	Joint
Site	Right Ankle

Add the treatment you received for this bleed

Treatment successfully recorded

Please include the time treatment was given

Date/Time *

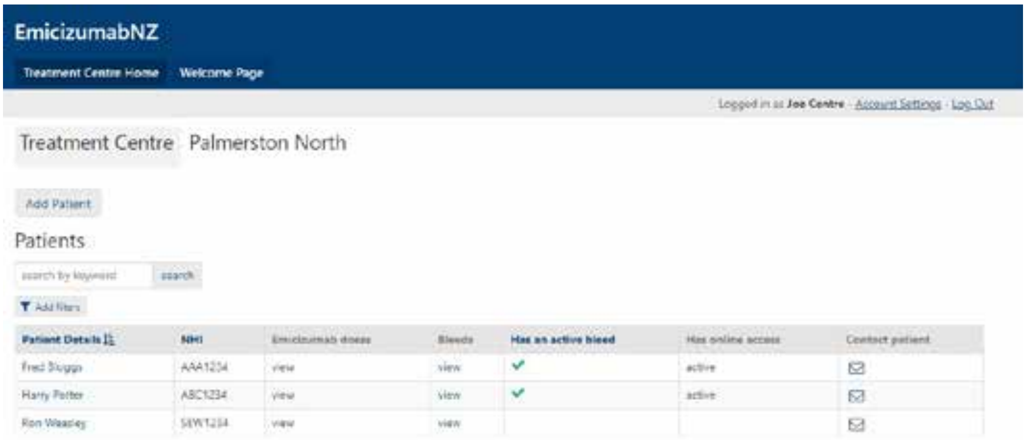
21/04/2021 2:50pm

Treatment Type *

Select type of treatment given

Summary of treatment given for this bleed

Date/Time	Treatment Type	Dose	Duration
19/04/2021 2:29pm	Recombinant VIIa	2 mg	
20/04/2021 2:30pm	Recombinant VIIa	2 mg	
19/04/2021 2:30pm	Tranexamic Acid	1000 mg twice daily	5 days



Treatment Centre Palmerston North

Patients

Patient Details	NHI	Emicizumab doses	Bleeds	Has an active bleed	Has online access	Contact patient
Fred Bloggs	AAA1234	view	view	✓	active	✉
Harry Potter	ABC1234	view	view	✓	active	✉
Ron Weasley	SEW1234	view	view			✉

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 ANA CLULEE

The Haemophillia World Day on Saturday 17 April was celebrated with a lunch for 18 members at Murrayfileds cafe in Levin to end our golden summer.

On 2 June we received some tragic news of the passing of Dr. John Manley Carter- REST IN PEACE. He suffered a massive heart attack and died in Wellington Hospital where his career as a clinician began.

After working as a house surgeon and medical registrar in Wellington Hospital, he undertook a fellowship at the prestigious Fred Hutchinson Cancer Research Centre in Seattle, Washington honing his skills in the new and developing discipline of bone marrow transplantation.

Back in New Zealand , he set up the blood and marrow transplant unit in Wellington Hospital, and the regional haemophilia centre.

Dr Carter was particularly proud of his pioneering work with haemophiliacs, setting up community-based services to help them cope in their own homes rather than in hospital. He helped them lead a normal life and got them playing sport and doing other things they wanted to do.

He was also prepared to speak out for his patients at political level, including advocating for haemophiliacs who had received contaminated blood.

One of those to benefit was Stephen Waring. As a young boy, he suffered more bleeds and was spending up to 10 weeks a year in hospital. Steve commented, he was "in hospital more than at school". It was life changing for Steve and his older brother Martin, who had the same condition when they met Dr Carter. He taught them how to self-medicate and encouraged them to get on with their lives. He also advocated for Martin when he was refused a job by the BNZ, by writing to the bank reassuring that there was nothing to fear. He gave the brothers his phone number. Steve said, "he made a huge difference to their lives and they were like his haemophilia children".

Dr Carter's funeral at St John's in the City Church in Wellington attracted a huge crowd and out pouring of respect and aroha for someone who saved many lives.

EVENTS:

HFNZ Women's Weekend - Dunedin Friday 12 to 14 November.

CHRISTMAS PARTY - Palmerston North at Te Manawa Saturday 27 November.

We would like to thank Lorraine and Ross Gordon who served on the committee for a number of years, and have now stepped down.

Last report from me, as I too am stepping down. I wish you all the best.

NORTHERN REPORT

BY GREG JAMIESON

It's been a little quiet for Northern events this year. Our last event over the summer was at the Zoo, which was enjoyed by all. We are currently looking at a family event like rock climbing over the next few months and then there is our Christmas catch-up in December so look out for this.

The big change for Northern is that Amber, our Outreach Worker, has had a great opportunity to build on her skills, and has left HFNZ for a new role in Australia. Our new Outreach Worker is Darian Smith. Darian is an experienced community worker and a published author. He is making an effort to connect with our Northern members so please make him welcome.

I hope you're all keeping safe, warm, and healthy over winter and look forward to catching up soon.

PIRITOTO

BY TUATAHI NIGHTINGALE-PENE

Ko te wehi ki te atua te timatanga o te whakaaro nui.

Ka huri tōku mihi ki tō tātou Kīngi a Kīngi Tūheitia me te whare kāhui ariki nui tonu, rire, rire, hau paimārire.

Otirā ki a rātou kua hemo ki te pō, moe mai, moe mai i roto i te Ariki.

Ko rātou ki a rātou, ko mātou te hunga kanohi ki a mātou anō.

E rau rangatira mā, nei rā te mihi kau atu ki te hunga kai-panui

Hello, Bloodline readers!

First of all, we would like to give a mihi to our newly appointed Outreach workers. To you all, welcome aboard and welcome to the whānau.

We are still feeling the ongoing effects of last year's lockdown as today's climate has made things difficult for us to meet. It seems that we are paying for lost time and everyone is working twice, sometimes three times harder to meet commitments. Generally, for many of us, it is a juggle of work, whānau, iwi / hapū / marae obligations, and many other forms of decisions seem to be today's priorities.

Piritoto would like to give a quick mihi to HFNZ for the many ways the organisation is always focused on improving and sustaining long term care for members. To the wider HFNZ whānau, we would like to give a mihi to everyone, it's great to see the respective MRGs doing their events and activities.

MIDLAND REPORT

BY WENDY CHRISTENSEN

The Midland committee put together a fun evening at the end of June. We had a fun evening playing tenpin bowling with pizza for dinner. It was a nice evening catching up with family and friends and was nice to have a variety of ages and some families travelling quite a distance to attend.

The Midland is looking at holding another get together in September at Taupo. It would be nice to see more new and old faces attend.

We would also like to let everyone from the Midland region know we have a closed Facebook page set up for our members. It's a way to keep up with what's coming up and also if anyone has any ideas of outings etc. they would like to do. You will find it under HFNZ - Midland Region https://www.facebook.com/groups/181254350611346

We look forward to welcoming more members to our FB group. Please note, it is a closed group for Midland region members only.

SOUTHERN REPORT

BY JAMES POFF

The Southern walking group has continued to push on over winter months, with the latest route taking the participants over an unusually flat coastal walk … must mean that Ross and Kylie are planning something a bit more challenging?

As has been previously reported, 2020 was a very challenging year for many of us. One of the consequences of this was that the Southern AGM was postponed, and the incumbent committee was “rolled-over” a period of several months. However, I am pleased to advise that by the time you are reading this the Southern AGM will have been held and a new invigorated MRG will be in place.

New Guidelines Issued to Self-administer Hemlibra

BY MARTA FIGUEIREDO PHD

The National Hemophilia Foundation (NHF), through its Nursing Working Group (NWG), has released new guidelines to help hemophilia A patients self-administer Hemlibra (emicizumab).

In its announcement, the foundation also highlighted the issuance of new guidelines for nurses to ensure safe and effective administration of standard into-the-vein replacement therapies in hemophilia patients.

Both documents, added to the Nurses' Guide to Bleeding Disorders, were developed based on the "clinical expertise, professional experience, and hard work of NHF's Nursing Working Group," the foundation noted.

This 12-member volunteer group was created to develop and promote standards of nursing practice in bleeding disorders, to promote health and education to the bleeding disorders community, and to advocate for the concerns of nurses within such community.

For people with hemophilia A, replacement therapy with FVIII — the blood-clotting protein they are missing — is standard practice. However, about a third of patients develop neutralizing antibodies, or inhibitors, against the delivered FVIII, lowering its effectiveness, and in worst cases rendering it useless.

Hemlibra is an antibody that mimics the activity of FVIII by binding to the activated form of factor IX and X, thereby allowing for proper blood coagulation. Four Phase 3 clinical trials — all part of the HAVEN program — showed the therapy was safe and effective at preventing bleeding episodes in hemophilia A patients with and without inhibitors.

Hemlibra, marketed by Genentech, is approved as a routine preventive, or prophylactic, treatment for hemophilia A patients with FVIII inhibitors in more than 100 countries, and for those without inhibitors in more than 80 countries. Approvals regardless of inhibitor status include the U.S., countries in Europe, and Japan.

Administered as an under-the-skin (subcutaneous) injection, Hemlibra's recommended loading dose is 3 mg/kg once every week for the first month, followed by a maintenance dose of either 1.5 mg/kg once every week, 3 mg/kg every two weeks, or 6 mg/kg every four weeks.

The new guidelines, called "Emicizumab Subcutaneous Injection Guidelines," were created mostly to help educate patients about how to properly self-administer Hemlibra.

"It is important to teach our patients correct subcutaneous injection technique, as incorrect technique can lead to poorly absorbed medication," the NWG wrote in the document.

Patients are advised to administer Hemlibra's loading doses under medical supervision, and if maintenance doses are different, "patients should be educated specifically about the maintenance dose and how to prepare it," they added.

The guideline document describes the equipment or supplies needed to administer Hemlibra and states that all should be at hand before starting the procedure.

Of note, patients should allow the therapy vials to come to room temperature before injection, which typically takes 15 minutes, and choose a small, short needle to ensure the medication is given subcutaneously.

The NGW also provides detailed descriptions of the several pre- and post-injection steps, as well as key guidance about choosing an injection site.

Patients are advised to check that they have drawn up the correct dose of the therapy — determined by the patient's healthcare provider based on their body weight — before removing the needle from the vial.

It also should be kept in mind that the therapy's vials come in different concentration strengths, not just different doses, and patients should not combine vials of different concentrations in a single injection.

When choosing an injection site, patients should avoid scars, moles, skin lesions, bruised, red, tender, or hardened areas, and areas over bony prominences, blood vessels, and nerves.

The most common injection sites include the thighs, umbilical region of abdomen (except for two inches around the belly button), and outer area of upper arms (only if a caregiver is giving the injection).

It is recommended that sites be rotated at least one inch from the prior injection to prevent irritation or scarring.

Before injection, the skin must be cleaned with an alcohol wipe and let dry for about 10 seconds. Hemlibra should be injected slowly and at a 45- to a 90-degree angle, depending on subcutaneous tissue and needle length, using the lifted skin technique. This technique, used to avoid injecting the muscle, involves using the thumb and index or middle finger to gently lift a skin-only fold.

After the injection, patients should avoid rubbing the injection site, monitor for any bleeding or adverse reactions, and apply a bandage or gauze as needed.

The guidelines also recommend that patients document the medication name, dose, date, time, and site, on an injection log, and plan their schedule for additional dosage administration.

Patients should contact their hemophilia treatment center if they experience significant weight changes or increased bleeding symptoms.

Any medication left in the vial must be discarded safely, and needles and syringes should be placed in a sharps disposal container.

The NWG also emphasized that the guidelines "are not meant to represent the sole resource" on Hemlibra administration, "but rather to be a supplement to existing patient education on the therapy."

Vonvendi Under Review for Prophylactic Use in von Willebrand Disease

BY BRIAN PARK, PHARMD

A PDUFA target date of January 28, 2022 has been set for this application. (Credit: Getty Images.)

The Food and Drug Administration (FDA) has accepted for review the supplemental Biologics License Application (sBLA) for Vonvendi® (von Willebrand factor [recombinant]) for prophylactic treatment to prevent or reduce the frequency of bleeding episodes in adults 18 years of age and older with von Willebrand disease (VWD).

Vonvendi, a recombinant von Willebrand factor, is currently indicated for use in adults with VWD for on-demand treatment and control of bleeding episodes, and for perioperative management of bleeding.

The sBLA submission is supported by data from a prospective, open-label, non-randomized, multicenter, phase 3 study (ClinicalTrials.gov Identifier: NCT02973087) that assessed the efficacy and safety of prophylactic treatment with Vonvendi in adults with severe VWD over a period of 12 months. Results from the study will be presented at the International Society of Thrombosis and Haemostasis (ISTH) Congress on July 20, 2021.

A Prescription Drug User Fee Act (PDUFA) target date of January 28, 2022 has been set for this application.

Source: https://www.empr.com/home/news/drugs-in-the-pipeline/vonvendi-under-review-for-prophylactic-use-in-von-willebrand-disease/

Why is Misdiagnosis of von Willebrand Disease Still Prevalent and How Can We Overcome It? A Focus on Clinical Considerations and Recommendations

BY CHANUKYA K COLONNE, BENJAMIN REARDON, JENNIFER CURNOW, AND EMMANUEL J FAVALORO

ABSTRACT

Despite von Willebrand disease (VWD) being the most common inherited bleeding disorder, its accurate diagnosis is frequently shrouded by diagnostic pitfalls. VWD is frequently under-diagnosed, over-diagnosed and misdiagnosed, leading to significant avoidable patient morbidity and health care system burden. At the heart of this dilemma lies the heterogeneity and complexity of von Willebrand factor (VWF) and associated defects, and the necessity of coalescing clinical and laboratory features to obtain an accurate diagnosis. Common pitfalls include poor clinical and scientific understanding and familiarity with VWD, incomplete clinical history and lack of routine use of standardised bleeding assessment tools (BAT), difficulty in accessing a comprehensive repertoire of laboratory tests, significant pre-analytical, analytical and post-analytical issues, and lack of expertise in laboratory testing and interpretation. Errors, resulting in under-diagnosis, over-diagnosis, and misdiagnosis of VWD, are presented and discussed. Strategies to minimise errors include better education of clinicians and laboratory staff on VWD, routine use of validated BAT, utilising a comprehensive gamut of laboratory investigations according to a standardised algorithm, and repeating testing to minimise pre-analytical errors. Recommendations on appropriate patient selection for VWD testing, how VWD should be investigated in the laboratory, and how to ensure test results are accurately interpreted in the correct clinical context are detailed.

INTRODUCTION

von Willebrand disease (VWD) is the most common inherited bleeding disorder. Despite this, VWD is one of the most commonly misdiagnosed or overlooked entities in everyday clinical practice. Of interest, VWD may be both over- and under-diagnosed, as well as misdiagnosed, either as another entity or as a different subtype of the disorder. This review will detail the causes underlying the diagnostic uncertainties overshadowing VWD and provide solutions on how to overcome these.

Full article available from:
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8380198/

Adult Weekend 2020

THE YEAR AHEAD

30 October, 2021

- HFNZ AGM
  Wellington. Venue to be determined.

12–14 November, 2021

- Women’s Weekend
  Scenic Hotel Southern Cross, Dunedin

19–22 April, 2022

- National Family Camp
  Christian Youth Camp, Ngāruawāhia

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

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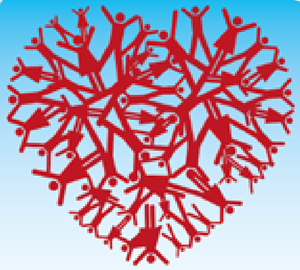

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