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National Haemophilia

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2021 Australian Conference

2021 Australian Conference Highlights and reports

VWD personal stories

Simoni and Perry share their experiences

Bleeding Disorders Awareness Week

Going red!

HAEMOPHILIA FOUNDATION AUSTRALIA

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Gavin Finkelstein

President, Haemophilia Foundation Australia

From the President

We know that when our national conference is over, we can expect a quick run to the end of the year. I recently presented my annual report to member foundations at the Annual General Meeting and for the second successive year I acknowledged a second extraordinary year because of the impact of COVID-19. I noted that, while we couldn't have face-to-face activities, we have concentrated on digital communications development work, and I hope you see some of the changes in the communications you receive from HFA.

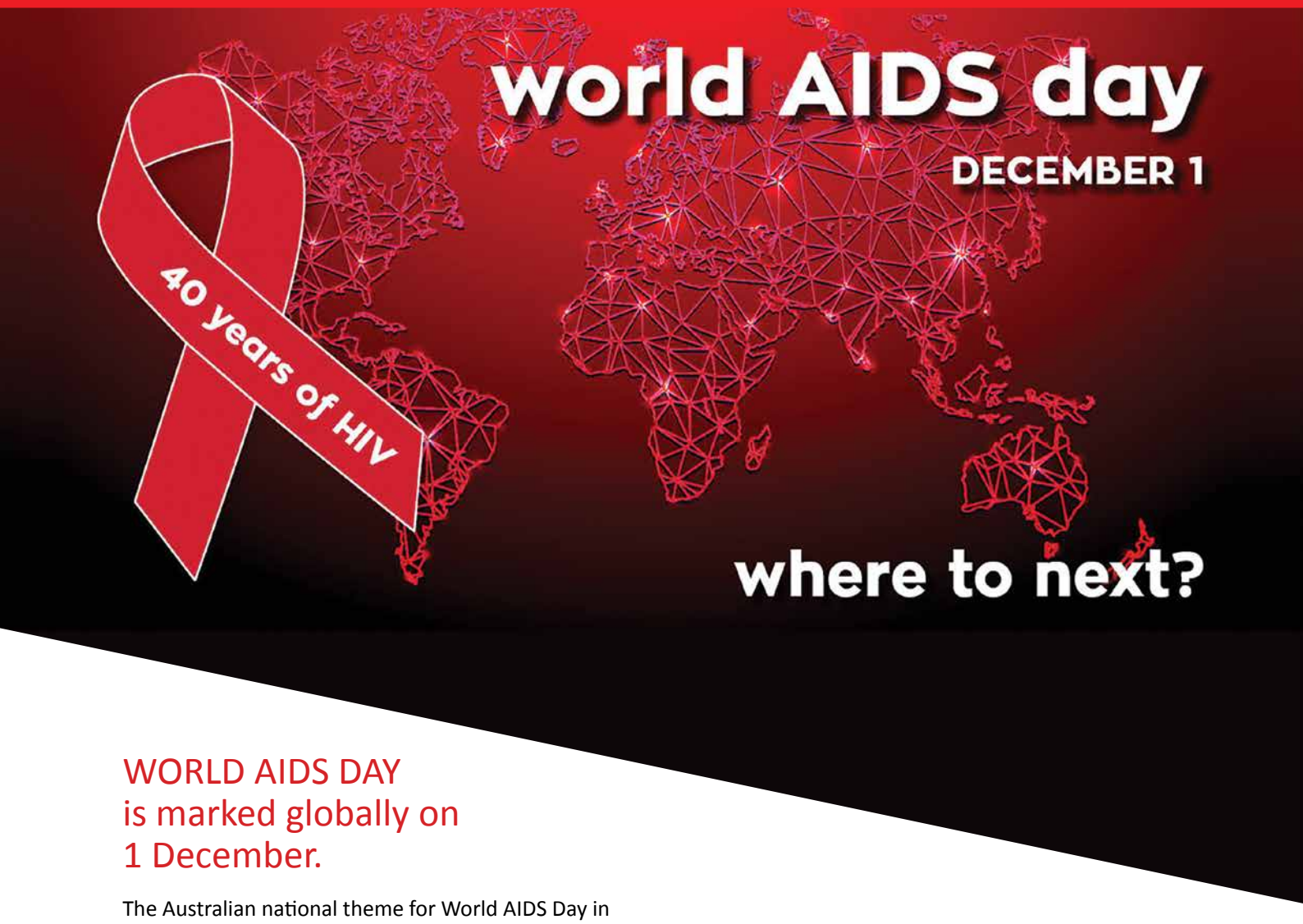
State/territory Foundations have also been having their annual general meetings, so it has been a busy time of reporting all round! I wish them a successful 2022, and hope the COVID-19 situation settles so face-to-face events for all parts of their communities can return to normal. It is great to hear that a group of South Australians met informally in Adelaide in October 2021 and planned another meeting for November.

CONFERENCE

Our first virtual national Conference was quite a learning experience; we learned we can share and learn effectively via Zoom presentations, and that virtual meetings provide opportunities for those who might not otherwise be able to attend. And even better, it's still not too late for you to view the conference sessions 'on demand' in your own time.

I would like to thank HFA staff, the Conference Program Committee and all the speakers and chairs who worked hard to present relevant, up-to-date and high-quality content. The various personal stories and experiences of community members throughout the program added depth and a reality for all delegates regardless of their connection, role or interest in bleeding disorders. I also thank our sponsors who so generously supported the Conference.

There was great interest in emerging therapies at the Conference and this highlights the important discussions needed with governments and other stakeholders. Some will challenge existing assessment and evaluation processes, and obstacles to access will need to be overcome before these therapies become available to those in our community who wish to adopt them for their treatment.



WORLD AIDS DAY is marked globally on 1 December.

The Australian national theme for World AIDS Day in 2021 is ***40 years of HIV – where to next?*** For the bleeding disorders community this is deeply important.

1 December is a day to raise awareness about HIV across the world and in the community.

It is a day when we are mindful of our community members living with HIV, of demonstrating our support for them and commemorating those with HIV who have passed away. Wearing a red ribbon is one way to show that you remember.

But in 2021 1 December is also an opportunity to take stock of the last 40 years and consider the future.

In 1981 AIDS (acquired immune deficiency syndrome), a new health condition, was first reported in Australian newspapers and the first case of AIDS was formally diagnosed in 1982. In the mid-1980s the bleeding disorders community in Australia and internationally was devastated when many people with bleeding disorders were diagnosed with AIDS after acquiring HIV through their plasma-derived clotting factor treatment products. HFA's Getting Older report documented the ongoing impact of this epidemic on our community: the trauma and health challenges for those who were diagnosed with HIV and now live with the consequences, and the grief and sadness experienced by those who lost loved ones and friends to HIV. Not to be overlooked is the emotional impact on the health professionals at the Haemophilia Treatment Centres too, who had cared for their patients with HIV over their lifetime and knew them so well.

Now in Australia HIV infection is usually well-managed with treatment. It is important to recognise the very positive contributions people with bleeding disorders and HIV have made to our community: the inspiring optimism and generosity of individuals with HIV, in spite of all their experiences, speaking out and providing leadership and a way forward into the future.

WHERE TO NEXT?

HFA is marking 40 years of HIV with accounts of the early history. How do we best move into the future? We have asked some community leaders living with HIV for their reflections and thoughts on where next with HIV. We will publish this in the March 2022 issue of *National Haemophilia*.

Visit the HFA website to read more – www.haemophilia.org.au

AHCDO update

Megan Sarson

The Australia Haemophilia Centre Directors' Organisation (AHCDO) held its first virtual Education Seminar and second virtual Annual General Meeting (AGM) in mid-October 2021. These events are usually a time for Haemophilia Treatment Centre (HTC) clinicians to come together with colleagues and discuss issues of particular interest and although, as with everyone, we missed the face-to-face element this year, the online environment did mean that more members than usual were able to attend - and indeed, the clinicians who couldn't attend on the day can access a recording of the Seminar through their member portal to view when convenient.

During the first session of the Education Seminar, we heard all about emicizumab (Hemlibra®). Dr Caroline Dix presented her work on emicizumab assays at the Royal Prince Alfred Hospital, noting the importance of measuring emicizumab levels to identify the development of anti-emicizumab antibodies, unusual bleeding despite good adherence and in preparation for surgery. Next up, Dr Heather Tapp presented on the treatment of Previously Untreated Patients (PUPs) with emicizumab, with a focus on the prevention and management of inhibitors. Dr Tapp noted a number of PUP emicizumab clinical trials currently underway. To close this session, Prof Huyen Tran gave an overview of which patients might be suited to treatment with emicizumab and some of the other novel therapies which are on the horizon.

The second session was led by Dr David Rabbolini who discussed his work with the Sydney Platelet Group, ranging from patient referral, collaborative lab testing to harmonize investigations and the improvement in continuity of care. He then went on to discuss how discoveries made in the laboratory can be translated back into the clinical pathway. Our final speaker of the day was AHCDO's ABDR Senior Research Fellow, Dr Sumit Parikh, who gave an update on two research projects currently being undertaken using data from the Australia Bleeding Disorders Registry (ABDR) – one on the use of Extended Half Life (EHLs) treatment products and the second on the Haemophilia Joint Health Score assessment tool.

Prof Huyen Tran

NEW AHCDO EXEC COUNCIL

Following the seminar, AHCDO held its AGM and announced a new Executive Council. Prof Huyen Tran (The Alfred, Vic) continues as AHCDO Chair and will be assisted by A/Prof Chris Barnes (Royal Children's Hospital, Vic), Dr Simon Brown (Queensland Children's Hospital, Qld), Dr Tina Carter (Perth Children's Hospital, WA), Dr Stephanie P'ng (Fiona Stanley Hospital, WA) and Dr Chee Wee Tan (Royal Adelaide Hospital, SA).

Megan Sarson is Executive Officer at the Australia Haemophilia Centre Directors' Organisation (AHCDO)

Bleeding Disorders AWARENESS WEEK

10-16 OCTOBER 2021

During the week of **10-16 October 2021** we celebrated Bleeding Disorders Awareness Week. The week is an opportunity for the community, Haemophilia Foundations, Haemophilia Treatment Centres, stakeholders and schools to take part in a campaign and activities to raise awareness about haemophilia, von Willebrand disease and related inherited bleeding disorders throughout Australia.

GO RED FOR BLEEDING DISORDERS

During Bleeding Disorders Awareness Week there were some great activities for everyone to participate in, which could be done virtually and face-to-face.

RED CLASSIC

Freddie leading his family ...

... and grandma riding her horse

Our national virtual Red Classic event saw people participate in all different ways – ride, scoot, walk even horse riding! Thanks to our participants for some great photos.

Sam and Rocky walking the Red Classic

Steph and family trekking the Red Classic

RED CAKE DAY

Our Red Cake Day cupcake packs were very popular.

At Jaylin's childcare the cupcake packs were a special highlight, where they baked and ate as a group.

Remy's daycare class had yummy cupcakes

BLEEDING DISORDERS AWARENESS WEEK IN PICTURES

John Hunter Children's Hospital

Haemophilia Treatment Centre, The Alfred

Thank you to everyone who participated in Bleeding Disorders Awareness Week.

2021 Conference

The **20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders** was held 8-9 October 2021. The event was a great success, with over 30 speakers contributing to a range of stimulating and informative sessions. The Conference is now available to watch on-demand, both for people who registered in advance and to new registrants.

The sessions, virtual exhibition booths, ePosters and Remembrance Service, will be available until April 2022.

EMBRACING A CHANGING WORLD

We have seen substantial change in both the bleeding disorders community and throughout the world since our last conference. COVID-19 and advances in treatment and care have had an impact on everybody. This year's conference took a special focus on these changes, with our theme, **Embracing a changing world**.

With topics across a range of areas and current issues, the 2021 Conference catered to everyone in our community. Sessions were presented by local and international expert health professionals, as well as by community member leaders. Sessions also included personal stories from people with bleeding disorders, giving a vibrant picture of real-life experience and the impact of these changes.

In this issue of *National Haemophilia* we hear from delegates about their impressions of the Conference and we have reports summarising the sessions and their key points.

POST CONFERENCE REGISTRATION IS STILL AVAILABLE

Registration for on-demand access is still open, giving people the opportunity to watch all the plenary and concurrent program sessions until April 2022. Conference satchels with the abstract book can also still be posted on request.

Already registered for the Conference?

Then simply visit the conference portal, log in and click **Program on Demand** to find and watch sessions.

HOW TO REGISTER

Visit the conference website for more information about registration and links to the conference portal: www.haemophilia.org.au/conference21

Conference reflections

A FOUNDATION PERSPECTIVE

Haemophilia Foundation Australia President Gavin Finkelstein spoke with HFA about the Conference and its impact from his perspective.

Gavin Finkelstein, HFA President, opening the virtual conference

What were your overall impressions of the Conference?

The Conference was a great job in difficult circumstances. It was very important to keep the continuity of conferences taking place, even during the COVID-19 epidemic, when we couldn't meet in person.

There is a lot that is happening in the haemophilia space, with the new treatments, and there also needs to be an increased awareness of the issues for women with bleeding disorders. Everything is changing very rapidly and people in the bleeding disorders community are hungry for the latest information. It was great to have information that is practical and delivered in a way that is very accessible.

What stood out for you?

The whole paradigm of accessing information has changed and we need to make use of the platforms available to us. A lot of us prefer to meet face-to-face for a conference, but when we can't, this digital space can still be very dynamic and bring information to people. The short personal stories were invaluable, and they impact on a lot of people because they sit and watch them and think about what it means for themselves.

What was your take-home message?

We just don't know what's going to happen in the future, so it's important to be flexible and go with the flow.

Remembrance service

THE HTC PERSPECTIVE

Haemophilia health professionals reflected on their experience at the conference.

Conference speakers - Managing bleeds under new treatments

The last few years in have been challenging for many in the world, virtual conferences and zoom meetings have become the norm rather than the face-to-face meetings and conferences we had been familiar with. Some of the positives of these virtual forums are less/no travel time, being able to watch at suitable times, re-watch sessions and the ability to watch concurrent streams; negatives would be lack of in person networking and opportunities for discussion around the issues presented.

The HFA conference embraced the changes that have become our healthcare reality in recent times and made the conference even more accessible with most sessions viewable on demand after the event.

Sue Webzell, Haemophilia Clinical Nurse Specialist, Hollywood Private Hospital

The 20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders 2021 was an interactive, informative conference to attend virtually. Even though we could not see in each other in person, the event was thought-provoking and provided great insight into haemophilia, VWD and rare bleeding disorders.

Some sessions in particular resonated with me, providing further understanding and prompts on what patients, families and health professionals can do to improve the care of people with bleeding disorders.

Jaime Chase, Haematology Clinical Nurse Specialist, John Hunter Children's Hospital

WHAT DID THE DELEGATES SAY?

'Overall I can't commend HFA enough for the efforts taken in organizing the conference and making it accessible through the wild circumstances we find ourselves in this year.'

'This conference has provided me with a much greater understanding of living with a bleeding disorder as well as appreciate even more the connectedness this community has with everyone involved.'

'The conference has made me reconsider the way that I approach exercise and active living.'

'Increased knowledge but missed the F2F connections.'

'I would have loved face to face from a very selfish point of view, but I appreciate the opportunity that this has given to participants who usually can't travel easily. I think it also facilitated more questions being asked, for example in the sex, sexuality and intimacy session that I think people would have shied away from.'

'One of the best conferences I have attended. Great variety from all different angles. Thank you!'

The changing treatment experience

Sue Webzell

Plenary 1 - The changing world of bleeding disorders

Chair ~ Dr Ritam Prasad
Short personal stories

Overview of the Australian experience - spectrum of changes in bleeding disorders over the years ~ Prof Huyen Tran

Are we there yet? The innovative future of bleeding disorders treatment ~ Dr David Lillicrap, Canada

An overview of the impact of advances in treatment on quality of life and independence and on clinical practice ~ Dr Liane Khoo

Plenary 4 - Where to from here?

Chair ~ Dr Ritam Prasad
Haemophilia care in 2030 ~ Prof Michael Makris
Where to from here? ~ Dr David Stephensen
My vision for treatment and care into the future ~ Claudio, Alan, Shauna
Bleeding disorders treatment and care into the future ~ Dr Ritam Prasad
Where to from here? Achieving the vision ~ Sharon Caris
Panel discussion/Q&A

The focus on patient experiences around the changes in their treatments throughout the conference was a good reminder of the positive outcomes newer approaches to treatment have had on quality of life. The patients described improvement through less frequent treatments, easier administration routes or, very importantly, reduced bleeding episodes. The less their dependence on factor replacement, the more similarity they experienced to peers with better freedoms and travel opportunities. One patient's experience showed significant improvement from anxiety and dependence using the original treatment to a feeling of freedom and independence on his current treatment.

The evolution of support services and treatment approaches from 1950s to now and into the future was a theme seen throughout many of the sessions. In the last two decades significant increases in scientific knowledge and understanding have led to changes in treatments, with various other treatment options under investigation at the current time.

PROPHYLAXIS TREATMENT

Professor Huyen Tran outlined some of the advances in prophylaxis treatment. It is known there is an increased likelihood of bleeding the longer the factor levels are low. Home prophylaxis treatment with plasma-derived and recombinant clotting factors has been used within the last two decades to reduce the time at low factor levels and therefore reduce bleeding episodes. However, with standard half-life treatments, infusions were frequent and would sometimes result in poor adherence to the treatment regimen.

More recently extended half-life (EHL) therapies have been used in both haemophilia A and B, which remain active for longer periods and reduce the treatment burden for patients and their families by reducing the frequency of infusions. The reduced number of infusions has been seen to increase adherence to the treatment regimen and reduce bleeding episodes in general, with more patients seeing zero bleeds.

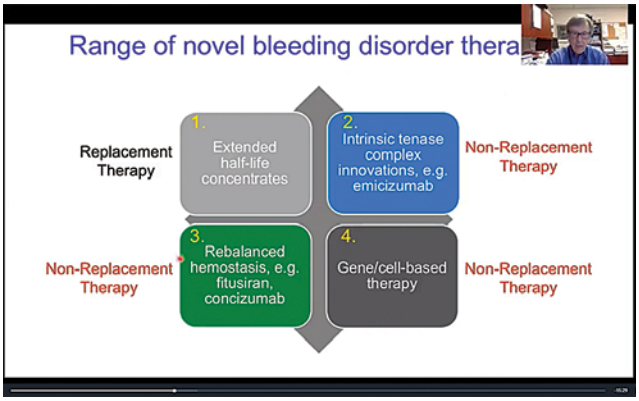
The biggest improvement is in haemophilia B where dosing for some individuals has decreased from 104 infusions a year (3-4 times weekly) to as few as 18-36 infusions a year (every 10-21 days).

In November 2020 emicizumab (Hemlibra®) became available in Australia for treatment of haemophilia A with or without inhibitors, the first of the new non-factor therapies to be publicly funded.

This saw a huge change in treatment from the conventional intravenous factor replacement therapy to a subcutaneous injection of non-factor treatment. These non-factor therapies enable coagulation to occur without the need to replace the missing factor VIII. They allow for a steady state in factor levels without the peaks and troughs seen in conventional replacement factor therapy. Bleed rates in patients having emicizumab were significantly reduced, with reported improvements in quality of life, less bleeds, less sick days and reduced hospitalization.

GENE THERAPY AND OTHER INNOVATIONS

Dr David Lillicrap spoke about the innovations in bleeding disorders treatments in recent years, the treatments in the pipeline and the science behind them. This was revisited by Prof Mike Makris in the final plenary, where he looked at the likely uptake of these treatments in the next decade.



Gene therapy is currently in clinical trial and for some individuals seems to allow for good factor levels for many years. However, it is not expected that it will be widely used, with around 10% of people with severe haemophilia are expected to receive it when it eventually becomes available. The funding process for this treatment would be complex and it is likely to be several years before this may become available. Currently gene therapy for factor IX deficiency seems to have better long-term factor expression.

Other novel non-factor treatments being investigated aim to rebalance coagulation by interfering with those naturally occurring products which would generally prevent thrombosis when present. By reducing the levels of these natural products, it is expected the bleeding risk could be reduced despite a lack of factor.

Prof Mike Makris speculated that in 2030 50% of patients will use non-factor products, 40% factor-based intravenous therapy and 10% gene therapy.

Use of conventional factor products and adjunct therapies for individuals will still be required especially for surgery in non-severe patients.

Dr David Stephensen spoke about the impact of new treatments on quality of life and the role of physiotherapy into the future. Better management with treatments reducing bleeds should in turn reduce joint bleeds and arthropathy and the pain associated with these joint problems. Previously physiotherapy treatment was available for few and focused on limiting impairment by movement avoidance; now treatment for all individuals aims to maximize quality of life by maintaining physical and social wellbeing. Traumatic bleeds may still occur on these treatments, but spontaneous bleeds are significantly less likely on the newer treatments.

THE FUTURE OF TREATMENT

Sessions on virtual health and getting older contributed to the general discussion about where treatment for bleeding disorders is going in the future.

Telehealth and video consultations are here to stay but will be used alongside traditional face-to-face healthcare delivery systems. Seeing a person face-to-face can be more informative to determine a treatment plan but Scott Russell's research study in the Virtual Health session showed that video consultations were a much better option for review than a standard telephone call, with more perceived confidence in the treatment plan when there was visual information alongside the verbal information.

Healthcare priorities should include best practice care and treatments for patients using comprehensive care strategies, with guidance from patient experience and goals for a personalized patient management and treatment plan. Patients with haemophilia are living longer with better treatments so the health conditions of ageing are now becoming a more common experience for the first time. Haemophilia Treatment Centres may need to adapt to better suit these changing patient requirements in the future.

Despite the great improvements in understanding and treatments for haemophilia, there have been limited changes being seen in von Willebrand disease (VWD) and other rare bleeding disorders. Research is being done around the disease biology which in time may lead to improvements in therapies.

Sue Webzell is the Haemophilia Clinical Nurse Specialist at Hollywood Private Hospital, Perth, WA

Where to from here? Achieving the vision

Sharon Caris

This is a transcript of the presentation by Sharon Caris in the final plenary at the 20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders. You can watch Sharon's presentation on the Conference portal at https://haemophilia.delegateconnect.co

A conference like this is just the right time for us to think, plan and to look forward.

We've covered a range of issues affecting the bleeding disorders community over the last couple of days of the Conference and looked at what's new and the changes we might need to make; but this is all on the back of COVID-19, and I think that alone will likely have implications for health care priorities and costs going forward in Australia for many years, and it will affect all of us. We need to take this into account when we're thinking about treatment requirements, new therapies and what we ask of our governments to supply.

I think what stands out for me today is that we shouldn't put all our eggs in one basket. We don't just need 'this treatment product' or 'that one'. There are many things, at many levels, to be done to achieve our vision for best practice care and treatment for everyone living with a bleeding disorder in Australia, and to support their families and their carers and treating health professionals as well. Nor can we forget that the World Federation of Hemophilia estimates that approximately 70% of the world's bleeding disorders community is either not yet diagnosed or adequately treated.

The first extended half-life product was registered in Australia in 2014, just before we were hosting the World Federation of Hemophilia Congress in Melbourne. We were hearing some of the promising results of clinical trials of other products, and just what could be achieved from these new products. But it has taken a few more years for these important new treatments to be funded and available for everyone who could benefit from them, even though there was evidence of significantly better health outcomes and improved quality of life. And since then, of course, more new treatment products have come into the mix.

There are many people at the Conference who are amazed by what can be achieved now with what we know as 'the new treatments' after such a reliance on conventional therapies for the last 30 or 40 years or so. Of course, standard clotting factor concentrates have been and are still important, but there's a great relief that several new treatment products are now available here.

As a community, I think we do have to decide what we're really aiming for. We've been hearing about other therapies around the corner. Some of these new haemophilia treatments are very close to market, while others are further away, and still need to be adequately tested for their safety and effectiveness. Some might be preferred. Time will tell.

We also need to make sure that von Willebrand disease and other rare factor deficiencies and inherited platelet disorders are addressed as well so that people living with these conditions have the best treatment and care.

It's not a straightforward thing to be able to access a new treatment in Australia and our experience over the past seven years has been frustrating. Some new treatments were approved for use by the Therapeutic Goods Administration for four years or more, so they had been assessed as safe and effective, yet funding and access to them was delayed because of various bureaucratic assessment processes, including necessary funding evaluations.

The principles and processes for approving and funding new treatments could be improved. There are several layers to this, and some changes will be needed to address some of the problems. HFA and other stakeholders are concerned about this and there are opportunities to contribute to solutions at a structural level, and HFA will contribute the experiences of our community. We have already been able to make submissions and participate in stakeholder discussions with governments in the hope that patients as experts, and their treaters, who have

the actual treatment experience are more involved at critical times in decision-making.

So, I do think the processes for assessment and funding can be better informed, so that access is more timely and there are fewer bureaucratic delays. This isn't to under-emphasize, though, the importance of critical assessments about the relative costs, benefits and value for money of these new products against others, that needs to be addressed for all medicines and healthcare services.

There isn't an unlimited pot of money, and we want governments to get the best bang for their buck in our space. We've recently had input to discussions around health technology assessment and how to contribute the experiences of patients and what they think are the most important outcomes to achieve from their treatment. We want to see more patient involvement so patients can demonstrate what is important to them. We have just heard from Shauna, Claude and Alan about their views and hopes for their futures, and just in the space of those three minutes you would agree, this was inspiring and very much of relevance to some of these evaluation processes. Most importantly, we want to see patient involvement so they can explain what is most important and the value of new therapies from their point of view and HFA will be working to ensure this input is taken into account at critical times.

We also want the Australian environment to encourage industry to retain interest and investment in the Australian market and to continue to invest in clinical trials here. So again, at a different level, we want to see barriers removed for industry that will enable them to bring their new treatment products to Australians.

We already have a strong national framework for bleeding disorders in Australia, and this is reflected in the national treatment guidelines for haemophilia. The legislation to establish the National Blood Authority in 2003 created a new system for the purchase and supply of blood and blood products and an agreement by Australian governments to share the cost of funding for blood and blood products. This is very important because so many practical aspects of our treatment and care crosses jurisdictions; treatment for bleeding disorders is more than just clotting factor replacement therapy, it is also about the care and treatment provided by Haemophilia Treatment Centres, and this is in the remit of state/territory governments. Both federal and state/territory governments have a stake in the Australian framework for care.

All governments need to be involved and engaged to make sure that we have the services needed. In addition to clotting factor, Haemophilia Treatment Centres also need to be adequately resourced to provide comprehensive care, which is a critical component of the national framework. It's all there in the framework. We need to make sure that it's understood and embedded in each of the decisions that follow.

I think it is also really important to have the concept of innovation included in the legislation that underpins all of this, so that there is a priority for considering the benefit to all stakeholders of new therapies as these might not only contribute to improved health outcomes for patients but save money as well. We need to embed a priority for innovation and best practice care and treatment as a principle into policy and the decision making that flows from this.

We've heard about potential different models for Haemophilia Treatment Centres, and how care might be provided in the future. The fundamental principles can be built into the 'hub and spoke' approach referred to as one example of a different structure for care to be delivered, and there are probably different concepts that can be considered as well across Australia, including the more recent experience of telehealth. We live in a big country, and comprehensive care doesn't need to be thrown out in any new models for healthcare delivery; it can actually create the very strength and success of new models. But we do need to make sure services are funded adequately so they are able to provide the care and treatment that's needed.

HFA will of course, continue to work with the stakeholder community. We will play our part to represent the community and this includes working closely with our state and territory haemophilia member foundations who are critical to this, as well as with medical and nursing specialists, allied health professionals, governments and industry to achieve our goals.

Most central to success will be how we ensure that the people living with a bleeding disorder are front and centre and that their needs and experience guides all of this.

Sharon Caris is Executive Director, Haemophilia Foundation Australia

Exercise and resistance training

Johanna Newsom

Getting stronger safely

Chair ~ Abi Polus

Research into exercise for boys with haemophilia (DOLPHIN-II Trial) ~ Dr David Stephensen

Less is more - getting the most out of limited time in the gym ~ Dr Mervyn Travers

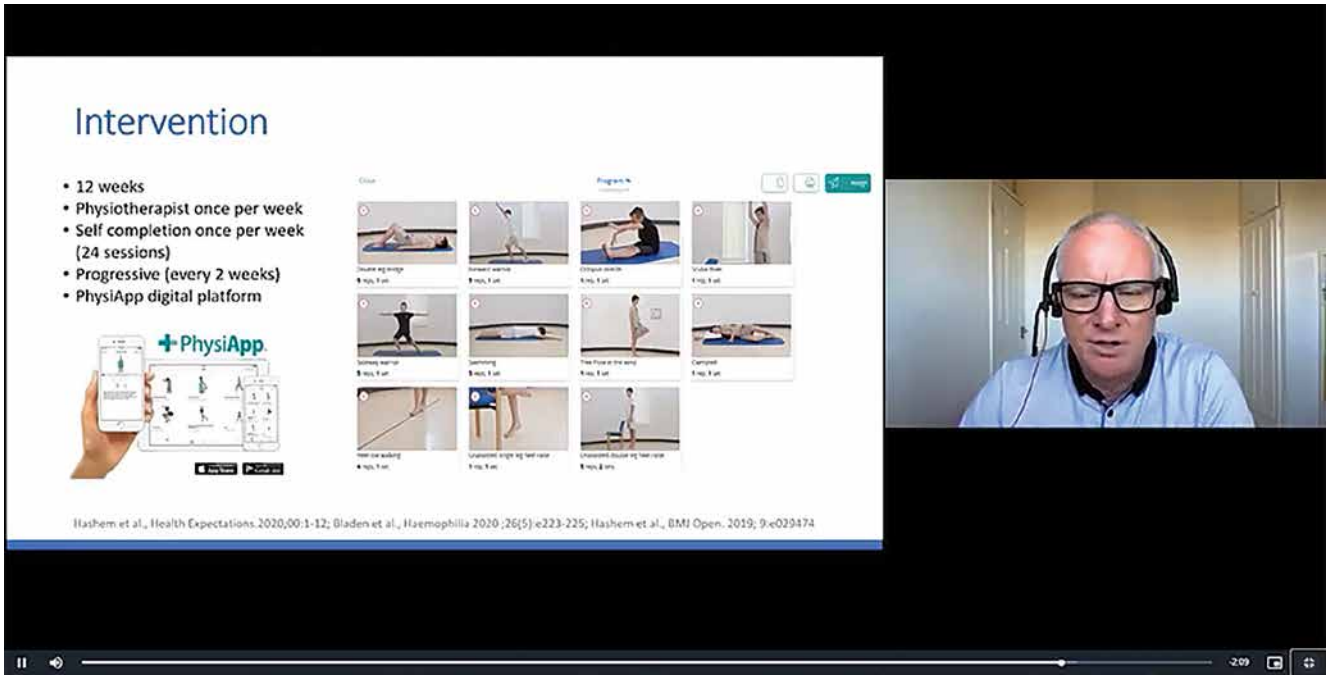
Panel Q&A ~ Abi Polus, Alison Morris, Tim Demos

As a physiotherapist, for me the concurrent session **Getting stronger safely** was a highlight of the conference. The two speakers were engaging and clearly passionate about their areas of expertise. Dr David Stephensen is a very experienced haemophilia physiotherapist working in the UK, and Dr Mervyn Travers has a wealth of knowledge and experience in musculoskeletal physiotherapy, particularly promoting physical activity and safe strength training.

EXERCISE AND BOYS WITH HAEMOPHILIA

David opened the session by telling us about his current research trial (DOLPHIN-II) into exercise for children with haemophilia. It is widely accepted that a well-designed exercise program promotes fitness, strength, agility, balance, and a health body weight, all of which we know is advantageous in people with haemophilia, but astonishingly, there is actual limited published evidence about the benefit of exercise for boys with haemophilia. However, there is good evidence to show the positive impact of exercise in other conditions which cause arthropathies – or joint disease – so it is logical that it would help in this population.

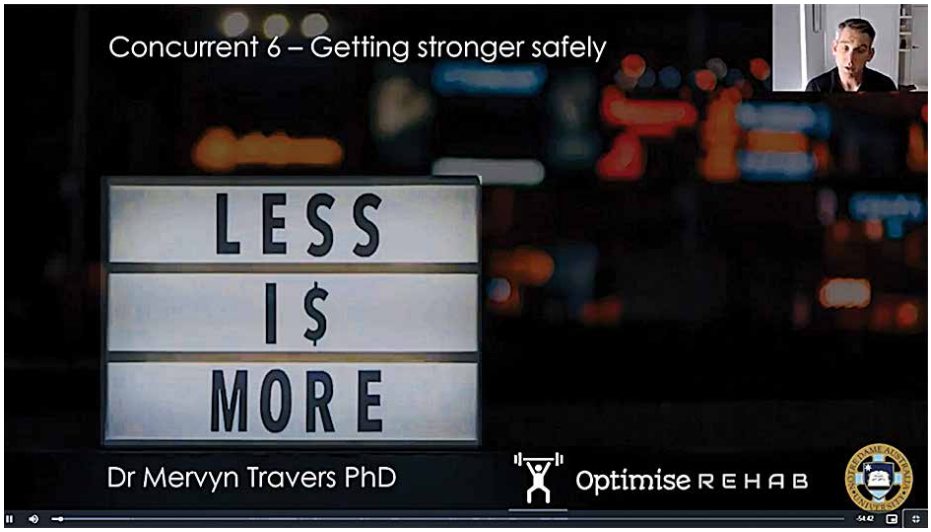
David and his team have designed a long-term study to look at the achievability and acceptance of a regular exercise program, and the effects on strength, participation and quality of life on boys aged 6-12 years with moderate or severe haemophilia. The program is a 12-week progressive design, with regular changes to the exercises to increase challenge (and keep things interesting!). There is a mix of sessions supervised by a physiotherapist, and independent sessions or sessions completed with family members, which are all supported via a digital platform with videos and descriptions of the exercises – very COVID-19 savvy!



Early results are indicating that the program is enjoyed by the boys and their families, easy to stick with, and showing improvements in strength and activity participation. This is an exercise program design that could be easily utilised on an individual or group basis, and could work well with our widespread populations across metropolitan, regional and rural areas as well, so we physios are looking forward to hearing the longer term results and seeing how the improvements are maintained over time… and then planning ways to be able to roll out something similar from our treatment centres.

RESISTANCE TRAINING

Merv then gave us an enjoyable and motivating talk about getting stronger safely. He opened with



the concept of the human body being 'anti-fragile', meaning that when a force (like lifting a safe weight) is applied to our bodies, we don't break, we don't just resist the force, but we have the capacity to absorb the force and improve (get stronger) as a result. This is an important idea in haemophilia, where the bleeding disorders community as a whole are still helping the wider population (including teachers and other health care providers) understand that regular exercise and a good activity level is not just safe, but indeed necessary to maintain good musculoskeletal health for people with haemophilia.

An outline of the current recommendations for physical activity broken down by age group then would have had a few of us squirming as we realised we are possibly underdoing our physical activity, especially during our various lockdowns! Merv presented statistics on the average activity levels across Australia and New Zealand, and a massive 30-50% of women, and 20-40% of men are not exercising enough. All exercise should be varied and include some resistance or strength training, balance, and aerobic activity, and be adapted or modified to suit individual ability (your physio or Haemophilia Treatment Centre/HTC can help with that if needed). At a minimum, we should be aiming for:

- 60 minutes per day, every day for the 5-17 year olds
- the 18-64 year olds to reach 5 hours of moderate intensity exercise across the week
- at least 3 sessions a week of moderate-level activity for the over 65s

The subtitle of Merv's talk was 'less is more' and he included lots of tips about how to get the most out of our time spent exercising. He discussed concepts like 'volume-equated training' and 'super-sets' which can help to get more bang for your proverbial buck when going to the gym, and choosing to don your activewear and lace up your trainers. These concepts need to be cautiously applied to anyone with pre-existing injury or joint damage, and as with any new exercise habit, need to be gradually introduced. Again, your physiotherapist can assist with setting up or changing up your exercise program.

The major take home message from this session on exercise and resistance training is the importance of participating in varied and regular activity, at all ages and stages of life. Exercise is safe for people with haemophilia, with and without existing joint damage, and there are ways to make exercise interesting, challenging and time-efficient. Any new program needs to be carefully and gradually introduced, and your haemophilia team can provide advice and support whenever needed.

Johanna Newsom is the haemophilia physiotherapist at The Children's Hospital at Westmead, Sydney NSW

Getting older

Suzanne O'Callaghan

Getting older

Chair ~ Jenny Lees

Personal story ~ Zev

Clinical issues with ageing
~ Professor Michael Makris, UK

Getting Older report – results and implementation
~ Suzanne O'Callaghan

Panel Discussion - What are the issues and challenges ahead? ~ Suzanne O'Callaghan, Frankie Mullen, Kathryn Body, Stephen Matthews

With a lively overview of what it means to him to be growing older with severe haemophilia, Zev set the scene for a multidisciplinary session on getting older that focused strongly on quality of life as well as health issues. Zev spoke about family, friendships and motivation, strategies for keeping fit and for maintaining a positive outlook – the power of the peer group. 'It's so therapeutic to be with someone where you don't have to explain,' was an important aspect for him.

CLINICAL ISSUES WITH AGEING

Professor Mike Makris is an international leader in ageing and haemophilia and outlined some key issues from a clinical perspective. He showed the remarkable change in life expectancy for people with haemophilia now – very close now to the general population. He also pointed out that numbers of people with mild haemophilia are much greater than those with severe.

Zev

Ages of life

He divided issues with ageing into two categories: issues at the third age or retirement and issues at the fourth age or dependency – where a person can no longer live at home independently. By the fourth age, problems are increasingly non-haemophilia related and may involve hospital admissions. He highlighted the increased prevalence of hypertension, musculoskeletal problems and hepatitis C in older people with haemophilia. He recommended that older people with severe haemophilia consider prophylaxis treatment, presenting data showing bleeds reduced dramatically with prophylaxis.

HIV and hepatitis C

Bloodborne viruses remain an issue. Sadly, very high proportions of people with haemophilia who acquired HIV have died, but with new highly effective antiviral treatments, for those who live on with HIV, it is now essentially a chronic disease. New treatments offer the potential for a cure for nearly all people with hepatitis C; however, the risk of liver cancer continues for those with cirrhosis and they need regular monitoring.

Health conditions of ageing

For older Australians with bleeding disorders, weight management is important as risks for cardiovascular disease are increased with obesity and being overweight can be a higher risk for musculoskeletal problems. They also now need to be aware of the risk for thrombosis, such as heart attack, stroke or

atrial fibrillation, which is common in the general population of older people. Usually treated with anticoagulants, it will need careful management in people with haemophilia. Other health conditions of ageing such as cancer or musculoskeletal problems, which would require medical procedures or surgery, need to be managed in liaison with the HTC to prevent bleeding complications. Osteoporosis is also more common in haemophilia. With such a range of problems, an important development in the future will be comprehensive geriatric assessments that include haemophilia specialists in the team.

During the Q&A at the end of his presentation, Prof Makris pointed to a number of current research projects exploring new areas: one in the Netherlands into bleeding-related arthropathy in older people with mild haemophilia and VWD; and some European studies on VWD and rare bleeding disorders, including a EUHASS thrombosis study, a study into whether you can grow out of VWD as you age, and a study to manage specific bleeding complications in some types of VWD.

ISSUES AND CHALLENGES AHEAD

I then had an opportunity to present the results of the HFA Getting Older report. This was a good springboard for the panel discussion, where we looked at some practical responses to questions that came up in the report and the issues and challenges ahead.

As a haemophilia nurse, Steve Matthews highlighted the challenges for HTCs:

- With such new territory, they are learning about the issues as patients present with them.
- Managing problems of ageing, such as heart bypass or stenting for cardiovascular disease, now commonly includes older people with mild haemophilia, who are not familiar with the range of treatments to prevent bleeding and need to learn about prophylaxis and personalised treatment to prevent bleeding, including the careful balancing with anti-coagulants.
- HTCs also need to look into strategies to help manage prophylaxis when older people can no longer self-treat or move into an aged care home, and whether the new sub-cutaneous treatments can be used - clinical trial results for people over 65 years are only now becoming available.

From a psychosocial perspective, Kathryn Body commented:

- Providing care for older people with bleeding disorders was quite time-consuming, which could be difficult for time-poor HTCs, and that it would be important to make the most of co-ordinated multidisciplinary appointments.
- Psychosocial workers may also need to have more of a role in researching for clients to connect them to services and advocating for them as they grow frailer and may not have the ability, the voice or the words.

Issues of ageing are an added complication for physiotherapy management, noted Frankie Mullen. However, this is also an opportunity to collaborate with community physiotherapists so that people can attend local programs for their regular health maintenance.

The panel discussed what makes a good GP for an older person with a bleeding disorder:

- Willingness to communicate and liaise with the HTC and with the patient
- Respecting the urgency of bleeding episodes
- Asking others who live locally about good local GPs.

For community physiotherapists there were similar criteria:

- A good physiotherapist was one who was willing to learn
- Where the patient felt comfortable with them
- And who understood the physical limitations of being older.

Keeping up physical activity as they grew older was particularly important for people with bleeding disorders to develop and maintain bone strength and prevent problems such as osteoporosis.

Self-advocacy can be more difficult as you grow older and more frail or develop dementia. Suggestions from the panel included:

- Taking a trusted person with you to appointments
- Before choosing an aged care home, visit a few and ask them questions about how they would manage someone with a bleeding disorder. If they do not support intravenous treatment, is there a local

community nursing service who could deliver this? Would they be open to education from the HTC on managing a bleeding disorder?

With so many new challenges in this area, the experience of the expert presenters in this session provided some really relevant and practical points, both from the perspective of the patient and the HTC. The session wrapped up with encouragement to view the HFA Getting Older Info Hub, where there is information covering many of these new issues.

My thanks to Jenny Lees from HFACT for chairing the session. Jenny has been a valued member of the HFA Getting Older Project Advisory Group and continues to support its work.

Suzanne O'Callaghan is HFA Policy Research and Education Manager

Pain and haemophilia

Nicola Hamilton

Plenary 3 – Pain

Chair ~ Dr Liane Khoo

Pain in haemophilia ~ Paul McLaughlin, UK

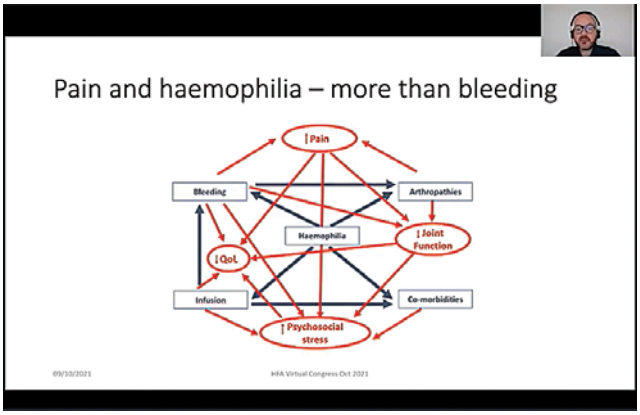
I enjoyed this presentation on pain and haemophilia as Paul McLaughlin, a leading haemophilia physiotherapist at the Royal Free Hospital in London, skilfully took us through an overview of pain and its complexities.

Paul explained that pain is complex and pain in haemophilia is even more complex. Pain can be acute from a joint or muscle bleed or it can be chronic from haemophilic arthropathy. For people with haemophilia, historically pain has been seen as a sign of bleeding and this has been taught from an early age.

Often pain is seen only through a biomedical lens and seeing pain ONLY as a correlation between joint health and joint status could be limiting.

Pain, however, can be distressing and it can interrupt your life and become a real problem.

In haemophilia, pain is more than just bleeding. Pain can be both acute and chronic and both acute and chronic pain can happen at the same time. Pain can be distressing and interfere with daily mobility and being able to attend school and work.



Pain can increase with age and can increase with the number of joints affected by haemophilic arthropathy. It can increase when mental health and wellbeing is affected and it can increase when your quality of life is impacted or activities of daily living become more difficult.

Paul explained that pain is not linear. Yes, it can be caused by an injury or bleed, but it can also be influenced by personal, social, psychological, familial, and medical issues.

And what I though was incredibly important is that pain is always a personal experience, and everyone's experience of pain is different.

ASSESSING PAIN

Paul highlighted that when looking at pain, assessment is important and best addressed in a comprehensive care setting by a multidisciplinary team. Because pain in haemophilia can be complex, it is very important that we try and ascertain which pain we are assessing e.g., is it acute pain from a bleed or an arthritic pain or is it a pain from another cause. A thorough physical assessment is essential and as a physiotherapist, this for me is paramount. Working out what pain we are assessing can be difficult, so an assessment also means listening and talking to the person with haemophilia and trying to gain an understanding of their pain and how it is affecting them and their story.

MANAGING PAIN

Paul made some key points about managing pain and drew on not only the haemophilia world when it comes to pain management but the wider pain community. I think as clinicians we need to get better at looking beyond the haemophilia space, especially when it comes to chronic pain and its complexities.

Managing acute pain in haemophilia can be relatively straightforward. Acute bleeding episodes tend to happen at home and over time the person with haemophilia develops strategies that work for them. In a sense they become the expert in treating their bleeds and what works for them should not be discounted. It is important however that when the pain management strategies being used at home are not working, the person needs to contact their Haemophilia Treatment Centre (HTC) for further help and advice.

Managing chronic pain in haemophilia needs to be a partnership between the person with haemophilia and the comprehensive care team. It should be framed as health maintenance and person-centred, and consider the biological, psychological, and social influences that there may be on that person's chronic pain. It is about finding what matters to the person with haemophilia and working towards it, being goal orientated and potentially using a mix of approaches. People with chronic pain should be encouraged to move every day, even if it is just a little bit. Pain is not always as sign of damage or that things are getting worse - remember exercise is safe for most people with haemophilia.

The take home messages for me from this presentation were:

- Good chronic pain management takes time – it is not a quick fix.
- It is important that the comprehensive care team and the person with haemophilia work in partnership in a person-centred way and that it needs to be a team effort.
- Pain is burdensome and everyone one has their own lived experience with pain and everyone's experience of pain is different.
- We need to be open to looking outside the haemophilia world when it comes to looking at chronic pain management.

Nicola Hamilton is the haemophilia physiotherapist at the Royal Children's Hospital, Melbourne

Sex and intimacy

Jaime Chase

Sex, sexuality and intimacy

Chair ~ Scott Russell

Awkward conversations: talking about sex and intimacy ~ Simone Sheridan

Panel discussion ~ A/Prof Chris Barnes, Alex Coombs, Penny McCarthy, Scott Russell, Simone Sheridan

Simone Sheridan (Sexual Health Nurse Consultant at Austin Health) introduced the session with a fascinating and thought-provoking presentation about the importance of sex and intimacy within a chronic disease environment.

Some takeaway points from this presentation included:

- Normalise the discussion around sex and intimacy.
- Acknowledge the importance of sexual health and wellbeing and know its importance!
- For patients - choose a health professional that you are comfortable with and write your questions down (it's very easy to forget).
- Manage expectations of the 'ideal' sexual experience - think outside the box, if required.
- Sometimes younger people may find it difficult to describe what they mean - they need help from a trusted healthcare provider to find the right words to use.
- Teach body autonomy and the importance of consent early to children as they pass through the healthcare system.

PANEL DISCUSSION

The panel discussion touched on a lot of the fears and queries that people with bleeding disorders have around sex and intimacy. All agreed on the importance of being able to be open around your healthcare providers and be honest with your questions and answers. The staff at your Haemophilia Treatment Centre are not embarrassed by any question that you ask them - so please ask!

The panel discussed common bleeding issues during sex and appropriate ways to mitigate these risks,

A/Prof Chris Barnes in the panel discussion

and how to treat or manage these bleeds if required. An aspect of this was young people's needs when exploring their bodies and the importance of exploring your body in a safe manner to decrease any risks. With the type of treatment that people have access to today, sexual activity is much safer and better covered then it ever has been before.

Disclosing your bleeding disorder was discussed at length - how you are going to bring up the conversation, how to frame it and what to say. A valuable suggestion was to practice it first in front of a mirror and see how you feel about what you are going to say. Planning always helps!

The approach to sexuality has changed immensely in the last 10 to 15 years and the panel reflected on the impact for people with bleeding disorders during these changing times.

Further resources include:
World Federation of Hemophilia - Ask Me Anything: Intimacy and bleeding disorders (video resources, 2021) - https://elearning.wfh.org/resource/ask-me-anything-intimacy-and-bleeding-disorders/

Jaime Chase is the Haematology Clinical Nurse Specialist at the Children's Cancer & Haematology Service, John Hunter Children's Hospital, Newcastle NSW

Virtual health

Suzanne O'Callaghan

Making the most of your health virtually

Chair: Suzanne O'Callaghan

Embracing health care futures: Global trends in consumer health care innovation and where they're taking us ~ Dr Zaana Howard

Future digital healthcare environment in Australia ~ Bettina McMahon

New experiences with diagnostics and programs in bleeding disorders – Australian case studies ~ Scott Russell, Erin Krake

Panel discussion ~ Erin Krake, Julia Ekert, Janine Furmedge, Sharon Danilovic, Helen Dixon, Neil (patient)

With the conference theme of ***Embracing a changing world*** in mind, the session on virtual health took a visionary but pragmatic perspective on the changes in healthcare due to advances in digital technology.

TRENDS IN DIGITAL HEALTH INNOVATION

Dr Zaana Howard, a leader in customer experience and design, introduced the session with an overview of the trends in digital health care innovation. She noted that the COVID-19 has accelerated some shifts that were already taking place in tools and technologies, such as telehealth. With their experience of being connected to simple, accessible and high-quality big brand digital technology with online streaming services and products like smart watches, consumers now have higher expectations of online health care.

She described three major shifts in health care innovation and delivery:

- Virtual care and remote medicine
- Preventative and hyper-personalised health
- Immersive health experiences.

Virtual health care integrates not only telehealth but use of IoT (internet of things) devices that can transmit personal health data remotely, such as wearables: smart watches and medical sensors are common examples. With increased use of telehealth, the point of care is shifting from where the health care provider is to where the patient is. Artificial intelligence can be used to collate large amounts of health data and predict likely outcomes – and provide alerts to patients to prevent poor outcomes. Some programs are now gamifying wellness programs and offer 'healthy challenges' to consumers as an incentive for them to improve their health and wellbeing. She also outlined the use of immersive health experiences in mental health and areas such as surgery, where the user wears a Virtual Reality headset and is immersed within a 3D virtual environment. Where will this lead in the future? Zaana described a new range of innovative health services that provide 'patient-centred ecosystems', with end-to-end care that integrates face-to-face with virtual care.

Bettina McMahon, CEO of HealthDirect Australia, followed with an outline of the future healthcare environment in Australia and the Australian Government digital health strategy. With the massive increase in use of digital and smart technology across Australia, an important priority is a seamless delivery of health and wellbeing services to consumers by integrating virtual healthcare with face-to-face health care services – so that for example, a consumer moves from a website, to a phone call, to a face-to-face appointment as a continuous experience. This involves providing consumers with the information and tools they need to make good decisions about their health and to know when to seek help from health professionals. Bettina underlined that it is particularly relevant to people with chronic health conditions such as bleeding disorders.

VIRTUAL HEALTH IN THE REAL WORLD ENVIRONMENT

How does virtual healthcare work in a 'real world' environment for people with bleeding disorders?

At the Haemophilia Treatment Centre (HTC) at the Royal Brisbane and Women's Hospital, the physiotherapists often conduct telephone consultations with their patients as the service is statewide and patients cannot always visit the HTC for physiotherapy appointments. Scott Russell, haemophilia physiotherapist at the Brisbane HTC, presented research results from their study, which compared video conferencing to telephone calls for physiotherapy consultations. With telephone calls, management and referral is often based on verbal descriptions from patients and there were concerns it could lead to sub-optimal management. His study found that:

- In 40% of the consultations, visual information provided in the video conference changed the clinical management plan made through an initial telephone call
- Both clinicians and patients were much more confident about the management plan from a video conference.
- This visual information was mainly related to the location of the patient complaint, swelling and range of motion.
- There were still some barriers to work through with video conferencing, including technical difficulties and patients' lack of digital equipment and expertise.

The HTC team from the Royal Children's Hospital in Melbourne followed Scott with demonstrations of virtual health in a paediatric environment. Haemophilia nurse Erin Krake showed how she conducted infusion education via telehealth with a young haemophilia patient and his family in country Victoria. Data manager Julia Ekert explained how data from MyABDR, the patient recording app, could be used in a telehealth review.

The question of virtual healthcare into the future continued in the session panel discussion. Erin and Julia were joined by Janine Furmedge, haemophilia nurse from their HTC team, Helen Dixon, haemophilia physiotherapist from New Zealand, Sharon Danilovic, social worker from The Alfred hospital in Melbourne, and Neil, who described himself as a '58-year-old guy with haemophilia A and HIV', to give the patient perspective.

For Neil a valuable part of his relationship with his HTC is the continuity of care with his team. As his health has improved, the frequency of his reliance on the HTC has decreased; but his team is very important when there are problems or when liaising with other specialities. Telehealth has been very helpful to manage distance and traffic.

Janine commented that this was similar for paediatric patients – to be able to pick up the phone and get the care they need when they need it; and to have education for themselves and for their child's school or childcare centre. Although many reviews have been provided via telehealth during the COVID-19 epidemic, there are still occasions when face-to-face consultations are important – especially with new patients, initiating new treatment, or when there are bleeding episodes, or multidisciplinary consultations with a range of specialties.

Helen noted that as a physiotherapist, some things were harder to assess via telehealth. In contrast, Sharon observed that social work clients had appreciated the access to the team via telehealth during the epidemic, the convenience and the opportunities to involve their family members and other professionals, such as a psychiatrist. However, like Scott, she drew attention to the lack of access to good internet technology for some patients.

Discussion moved on to the future. The panel considered the potential for artificial intelligence to work with tools such as MyABDR to prompt patients to connect with their HTC when it detects particular issues, such as unresolved bleeds. Helen also described a study in the UK on the use of ultrasound probes by haemophilia patients in a virtual health environment at home. She was also excited by the possibilities of wearables to share data with an HTC on a patient's activity; Neil was less excited about being monitored constantly! Virtual technology did not remove the need for a specialist HTC. To work effectively, all of these innovations were dependent on skilled patient education and close collaboration between the patient and the specialist HTC team.

Suzanne O'Callaghan is HFA Policy Research and Education Manager

Living with von Willebrand disease (VWD)

Perry's story

Perry spoke about his personal story of living with von Willebrand disease (VWD) at the 20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders. This is an edited version of his presentation. You can watch Perry's full presentation on the Conference portal at https://haemophilia.delegateconnect.co.

My name is Perry. I have von Willebrand disease type 2a. I'm in my 70s, and I didn't get diagnosed until I was in my late 50s. I live with the disease and I would like to talk about that.

When I look back, there were a few hints that von Willebrand disease was in my family, because it's hereditary, as you know. I can remember that my mother had serious nosebleeds at different times. And, of course, she probably had other issues which would have indicated that she might have been a carrier.

I played sport at a high level for a long time, and I noticed I was bruising a lot more seriously than some of the other players that I played with. And that was another hint that I had something a bit different.

BEING DIAGNOSED

In my late 50s, I had a back operation, a laminectomy, and the surgeon mentioned to me after the procedure that he had a hard time stopping the bleeding. He suggested I should see my GP about whether there might be some issue with me.

I had never heard of von Willebrand disease. I knew a little bit about haemophilia, but nothing about von Willebrand disease. My GP did a number of blood tests, and then he sent me to a haematologist, who then did a lot of other tests on me, and I was eventually diagnosed with von Willebrand disease.

MANAGING SURGERY AND PROCEDURES

Initially I thought it was a condition where you can get something that'll fix it, and then you won't have to worry about it anymore. I didn't realise it was an ongoing condition, which I needed to be aware of – and also that I needed to take steps to make certain that I was protected when I had operations or other procedures.

After that point, I decided to make sure that in future I had the proper medical advice and support with

anything that involved bleeding or operations. I was very fortunate to get that at the Haemophilia Treatment Centre. Ever since, all my various procedures, including colonoscopies, a bleed from the bowel and an ankle fusion, have basically turned out quite well from a bleeding perspective.

It's not something that just will go away. I think that needs to be emphasised with people who are diagnosed with von Willebrand disease.

MANAGING BLEEDING

The things that I've learnt to do to live with VWD have been much varied.

Every once in a while, I'll get a nosebleed. Nosebleeds are very serious for me because it takes me a long time to get them to stop. I've had to learn various techniques, including using a paste made from tranexamic acid. I've had some nosebleeds for three days, and it is it is very depressing, as you can imagine. But it's because of von Willebrand disease and I have had to work through it.

I still bruise very seriously, and it bothers me, as I need to be very careful with the physiotherapists I see for treatment. Physiotherapists have to know about my condition and how I can be affected.

Probably one of the things that has really affected me is occasionally I'll bite the inside of my mouth, maybe through eating, and I find that I bleed from the resulting ulcer or sore and that's very, very difficult to stop. I think it must have something to do with the saliva in your mouth as it tends to break down any capacity for the inside of your mouth to clot and to stop the bleeding. I've been fortunate to have been given information about how to stop that bleeding but it does take time.

Another problem I experienced when I had a number of operations where I was losing blood, was that my iron levels would go down. I couldn't work out why I was getting so tired. I was able to get iron supplements and that has enabled me to keep up my strength.

CONTACTING FAMILY

When I was diagnosed, I also decided that I was going to get in touch with my relatives. Originally I came to Australia from America, but that was 50 years ago. I have two sisters and one brother. I let them know that I had von Willebrand disease and that they should all consider getting tested because it's inherited and they come from the same genes as me.

They did. And they all found that they don't have von Willebrand disease. I was the lucky person out of the four who got it.

I have 4 children and 13 grandchildren and they're all aware of the situation. Some have been tested and some will be tested. It may be passed down to them and it's good that they know sooner than later that if they carry or have VWD.

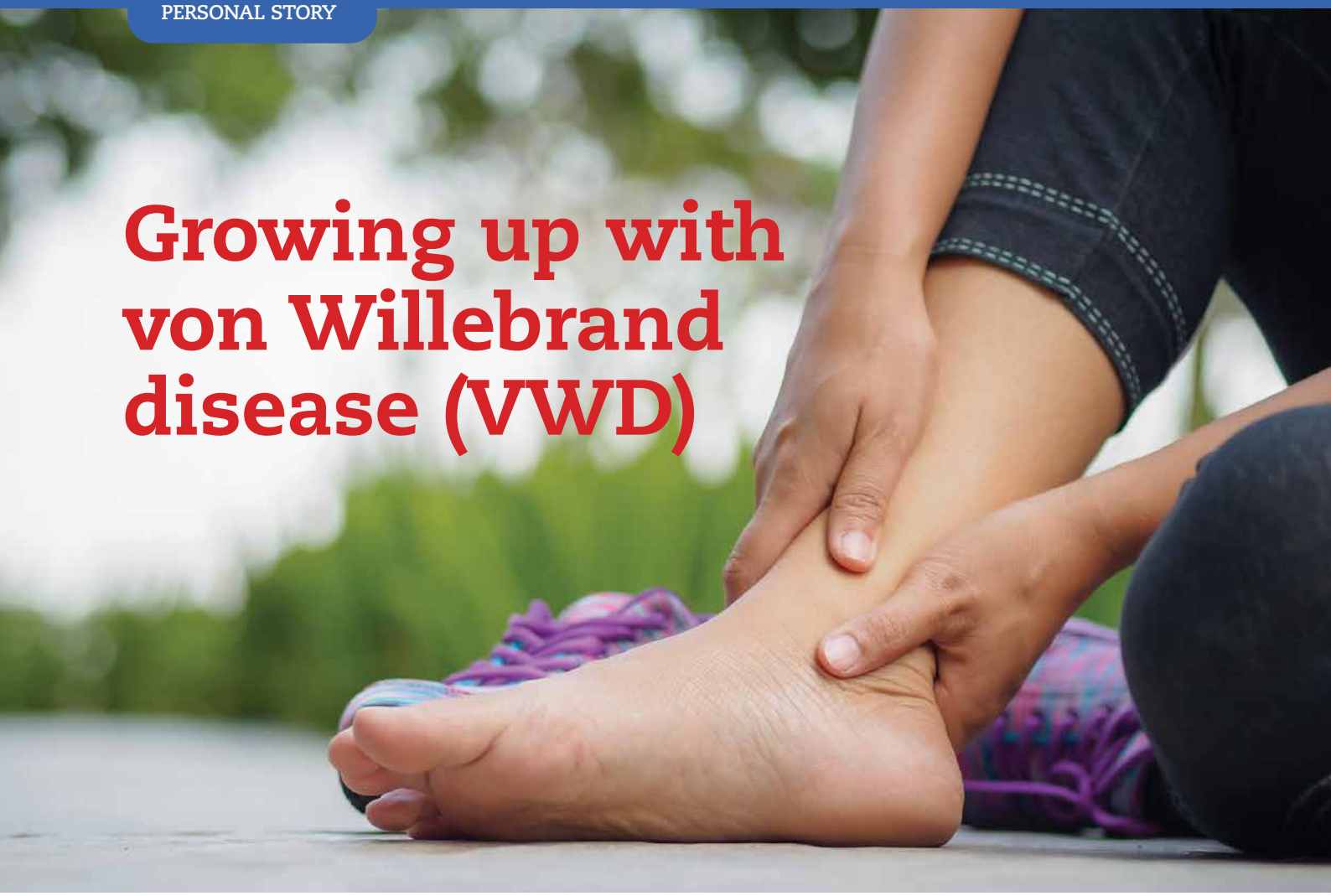
DEALING WITH VWD

My family understands this and certainly my wife is well aware of the various things that occur because of the disease. She deals with it by helping me deal with it.

It doesn't really affect your lifestyle. You just have to be aware of it and how to manage it, so that if certain things happen, you are able to deal with it. I like to walk in the bush and if I have a couple of falls or scrapes and am bleeding, we ensure that we get them bandaged straight away and that they are kept covered until they are able to heal themselves, because if they're open, they continue to bleed and that's not good for anyone.

My message to other people diagnosed with VWD is that they need to be referred to a Haemophilia Treatment Centre, and the Centre will look after them. They will need to go to hospitals that are capable of dealing with VWD, particularly for operations. You might have a favourite doctor, but it's important that they are capable of understanding what's involved in the treatment of people with von Willebrand disease.

You can't pick your parents and you can't pick your previous generations, but you learn to deal with von Willebrand disease. And I just hope that if future generations in my family do have VWD, they will be able to deal with it because of the knowledge that I have been able to pass onto them.



Growing up with von Willebrand disease (VWD)

Simoni's story

This is a transcript of Simoni's presentation at the 20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders. You can watch Simoni's presentation on the Conference portal at https://haemophilia.delegateconnect.co.

Hi, I'm Simoni, and I have type 3 von Willebrand disease (VWD), the most severe and rarest form of the disease. Consequently, I have little to no von Willebrand factor and low levels of factor VIII (8). When I was a child, my parents noticed that I bruise easily from daily activities and was often susceptible to minor cuts, which would bleed for a long time. They then proceeded to have me tested, and it was found that I have von Willebrand's at the age of three years old.

MY TREATMENT

Some of my symptoms include a longer bleeding and clotting time for injuries, easy bruising, heavy nosebleeds, recurring joint bleeds and heavy menstrual bleeding, resulting in anaemia and fatigue. At eleven years old, I was started on prophylactic treatment of intravenously administered clotting factor concentrate every alternate day. The support of my haemophilia nurse was and continues to be instrumental in my treatment from the very beginning.

The factor was first administered by help of my nurse, followed by my parents who administered it regularly until I learned to administer it myself recently. My current treatment also involves taking tranexamic acid tablets during bleeding episodes to supplement my regular treatment, which helps slow down the dissolution of blood clots.

MANAGING INJURIES

In early primary school, when I was not on regular treatment, I was highly vulnerable to serious injuries, particularly in sport classes. I did limit my participation in sports to non- or low-impact activities as a measure of caution, which was not a significant problem as I'm not naturally a sporty person.

In my experience, I found it beneficial to let school personnel and close friends know about my bleeding disorder and inform them of the specifics of managing a serious bleed or injury so that we could act promptly to minimize the severity of any injury.

An event which highlighted the necessity of this was when I was eight years old whilst playing basketball, my ankle was hit by the ball with high impact. As there was no immediately visible injury, I did not pay much attention until I got home that afternoon, by which time my ankle was severely swollen. After further investigation, we found that I had a ligament injury, which developed into a recurrent issue. If we had taken prompt action then, perhaps the long-term impacts would have been eased.

Prior to beginning prophylactic treatments, I would be treated in a children's hospital and would often have a cannula in my arm for a few days to enable daily treatment. Due to von Willebrand's, I have found that my recovery period from major injuries is quite lengthy. For example, about three to four weeks for an ankle injury, sometimes longer, depending on the severity.

Due to recurring injuries, in particular joints, physiotherapy has become a key part of recovery and helps to build muscle strength to prevent further trauma. Also, nosebleeds can be quite heavy and are difficult to stop, for which I use a nasal spray and administer factor treatment as soon as possible.

As a result of having a bleeding disorder, micro bleeds in joint do result in frequent joint pain and sometimes swelling, which I manage through rest and ice compression along with regular treatment.

MANAGING PERIODS

Upon beginning menstruation, I experienced very heavy periods causing severe anaemia and was required to go on hormonal medication to control it. I remained on medication for approximately two and a half years, after which I was able to move forward without medication.

LIVING WITH VWD

Also, when I know I will be participating in strenuous and high-risk activities, it has been essential to ensure that I have had a dose of my prophylactic factor treatment earlier the same day, not only to mitigate the impact of any serious injury, but also to possibly reduce the time which I might need to recover. As a precaution to be able to manage any injuries or bleeding promptly, I keep a kit with pressure wrap bandages, medications and Band-AIDs® close by when traveling anywhere. There have been numerous occasions in which my kit has come in handy, and it has been a key part of living with von Willebrand's.

Prophylaxis has proven to be a very effective method of managing my bleeding disorder, enabling me to participate in daily activities with normality. I do still maintain a degree of caution and try to prevent major injuries or be swift in managing any bleeding events. Prophylaxis is now a regular part of my life and is a monumental component of my experience and living with the disease.

Additionally, I am hopeful that a recombinant, long-acting subcutaneous injection will replace intravenous factor in the future, similar to that available for haemophilia treatment, which can be easily and less frequently administered.

Overall, in my experience, living with von Willebrand's does not mean that you should feel restricted or disadvantaged in any way. While it is important to know my limits, learning to manage the disease and symptoms effectively and bringing immediate attention to injuries is even more vital. And with time, I've learned to integrate the symptoms and treatment as a part of daily life.

I think there is a lot of fear and uncertainty that both a person living with the disease and perhaps their friends or family might have. So, I think it's important to cultivate and maintain a support system and try to develop a greater awareness of your condition for those around you.

Thank you.

Von Willebrand disease

Suzanne O'Callaghan

Von Willebrand disease

Chair ~ Susie Couper

VWD personal experience ~ Perry

VWD genetics, diagnosis and classification ~ Dr Simon McRae

VWD personal experience ~ Simoni

VWD treatment and future directions ~ Dr Nathan Connell

The von Willebrand disease session brought together some of the key players in the development of the new international clinical VWD guidelines to discuss the implications for diagnosis, treatment and care into the future.

THE PATIENT PERSPECTIVE

Setting the scene from the perspective of the patient were two compelling personal experiences from Perry and Simoni.

As an older man with type 2A VWD, Perry had not been diagnosed until he was in his 50s. He spoke about the problems he experienced with bleeding episodes over his lifetime before he was diagnosed and had a treatment plan – nose and mouth bleeds, lengthy bleeding after surgery. This bleeding is now much better controlled in liaison with his Haemophilia Treatment Centre.

Simoni is a young woman with type 3 VWD and was diagnosed as a young child after bruising severely. As she has grown up, she has had a number of bleeding problems, including heavy menstrual bleeding and bleeds after sports injuries. For Simoni, starting prophylaxis was a turning point for taking control of her bleeding and her quality of life.

You can read Perry and Simoni's personal stories in this issue of *National Haemophilia*.

What is exciting about the new international guidelines from a patient perspective? As a patient representative in the international panel for the clinical management guidelines, Susie Couper had three reasons to be excited:

1. VWD now has more visibility and as a result there is more understanding
2. The VWD guidelines were produced with patient involvement at every step, acknowledging the differing values of patients
3. The guidelines have opened up research priorities and put a focus on the strength of evidence.

VWD DIAGNOSIS AND CLASSIFICATION

Dr Simon McRae, now based at Launceston General Hospital in Tasmania, was a member of the international panel for the VWD diagnostic guidelines. He explained that there had been some variations in diagnostic criteria in previous guidelines and the new guidelines had been an opportunity to develop some consistency and provide decision-making tools.

The guidelines considered:

- Who should have diagnostic tests performed?
- What is the role of a structured bleeding assessment tool (BAT)?

The circumstances of diagnosis were very relevant.

A BAT would be useful if a patient was being seen at a primary care setting, eg a GP clinic, where the likelihood of them having a bleeding disorder was lower. A validated BAT could identify those who have abnormal bleeding that falls into the type of bleeding seen in a bleeding disorder such as VWD and indicate where further testing is appropriate.

If a patient has been referred to a haematologist or has a strong family history, they are more likely to have a bleeding disorder and a BAT is not needed to decide about testing. However, a BAT can be useful to help define their particular bleeding pattern.

Other areas that the guidelines clarified were:

- The value of newer assays or tests that measure the platelet-binding activity of VWF (von Willebrand factor) more accurately, such as VWF:GPibM, VWF:GPibR etc. Most laboratories in Australia already use these.
- What VWF assay level defines a diagnosis with VWD? The 30-50% VWF level has been contentious in decisions about VWD diagnosis and diagnosis will depend on the person's bleeding history. There may be other factors contributing to bleeding; but it is also important to be wary if a person has not yet had bleeding challenges, eg surgery, particularly males.

The guidelines also considered questions such as differentiating between type 1 and type 2 and when VWD genetic testing is useful (ie, for type 2A, 2B or 2N and for family planning in type 3).

What are the implications of the new diagnostic guidelines for Australia?

Dr McRae pointed out that the guidelines won't change diagnostic practice in most Australian settings and are unlikely to change patient management for individuals. They are likely to lead to a review of current diagnoses in the ABDR and there may be a need to retest patients where insufficient information is available. This will take some time. However, an important outcome for patients will be standardising of the wording for individual diagnoses, including on the ABDR patient card.

He also recommended a national network approach for genetic testing and some functional assays. Genetic testing funding is not always available nationally, making it difficult in some cases to work out how to fund the tests that are required for accurate and timely diagnosis.

VWD TREATMENT AND FUTURE DIRECTIONS

Dr Nathan Connell gave an update on the latest developments in VWD treatment, based on the new international VWD guidelines. Dr Connell was Vice-Chair of the international VWD guidelines Scoping Group and explained that the Group was an international partnership to develop evidence-based clinical management guidelines to promote better health outcomes, quality of life and health equity.

He discussed some of the key clinical management issues covered by the guidelines.

Prophylaxis is recommended for type 3 (severe) VWD, but conditionally – a shared decision-making process between the doctor and the patient:

- Prophylaxis reduces risk of bleeding episodes, hospitalisations, and heavy menstrual bleeding.
- The value of prophylaxis to individual patients will depend on how often they experience bleeding episodes.

The recommendations for heavy menstrual bleeding:

- Different options for women who do not wish to conceive vs women who wish to conceive.
- Focus on hormonal therapy (hormonal contraceptive therapy or levonorgestrel-releasing IUD) or tranexamic acid over desmopressin.
- Women might need to use multiple options at once if their heavy menstrual bleeding is not well controlled with the initial therapy.

- Importance of a multidisciplinary approach, with haematologist, gynaecologist and patient to manage both the bleeding disorder and gynaecological complications, which may or may not be related to VWD.
- Include assessment and treatment of iron deficiency and anaemia.

For people with mild and moderate VWD, what are the 'moments that matter', the triggers to get in contact with their Haemophilia Treatment Centre (HTC)?

- If they are planning any surgery or invasive medical procedures, they should connect with their HTC.
- Managing any bleeding, particularly heavy menstrual bleeding in women.
- Need to plan in advance for pregnancy.

RESEARCH PRIORITIES

Both Simon McRae and Nathan Connell pointed to a real need for evidence-based research to answer a range of questions about VWD, for example:

- Bleeding risk as people with VWD age
- Are there predictors for who will bleed and who will not, particularly those in the 30-50% VWF range
- Evaluating different bleeding assessment tools in males and children
- Research into hormonal contraceptive therapies for women
- Large randomised controlled trials on prophylaxis for VWD
- Use of plasma-derived vs recombinant VWF concentrate for prophylaxis.

We were grateful to all the presenters for sharing their expertise and their personal stories. The expert presentations from Simon McRae and Nathan Connell were immensely valuable and gave a concise and accessible overview of priority issues in VWD diagnosis and treatment. Having the personal perspectives of Susie, Perry and Simoni alongside their presentations grounded the discussion about the guidelines in real-life experience – and highlighted just why the process of developing international guidelines has been so important.

Suzanne O'Callaghan is HFA Policy Research and Education Manager

Women and girls

Jaime Chase

Women and Girls

Chair, introduction and personal story ~ Sharron Inglis

Understanding, recording and reporting bleeding symptoms in girls and women ~ Jaime Chase, Joanna McCosker

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

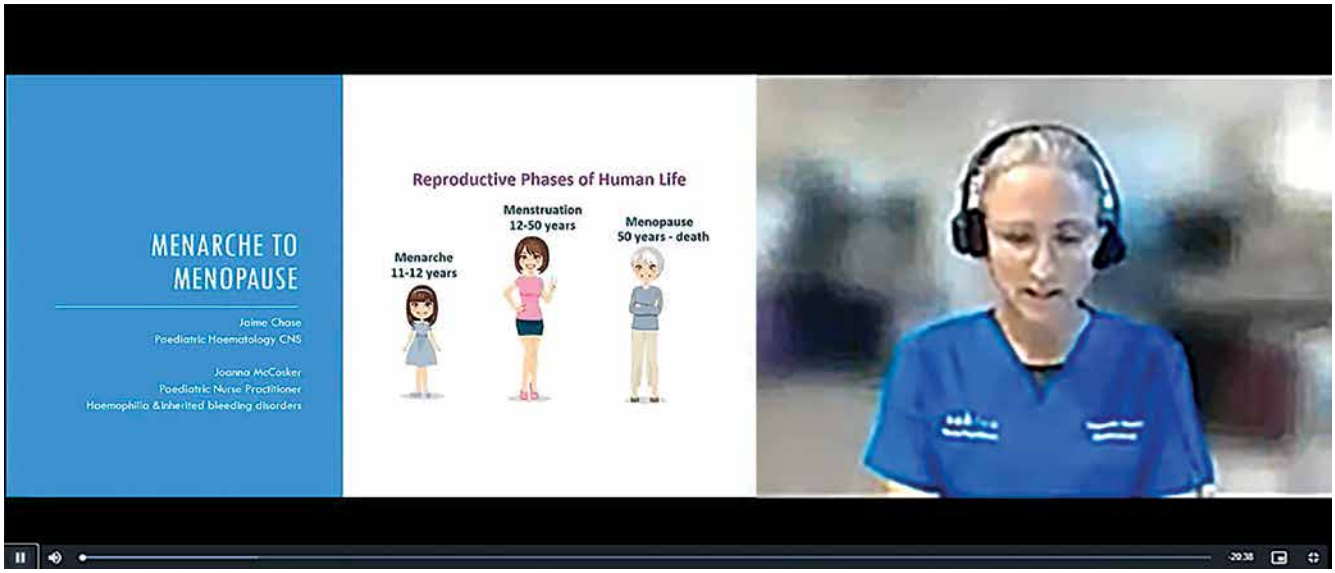
Gynaecological issues for women and girls with bleeding disorders ~ Dr Angela Dunford

Sport and exercise for girls and women ~ Hayley Coulson

The session regarding women and girls commenced with a personal story by Sharron Inglis, an Australian community leader, and raised some interesting questions about society's perspective on women with bleeding disorders. Advocacy, support and education were highlighted as her most important messages.

Heavy periods can negatively impact on physical, emotional and social quality of life and reduce work capacity - so how can we fix this?

Joanna McCosker and Jaime Chase discussed the importance of normalising the conversation around periods as they commence and as a woman moves through her lifespan to menopause.



The session included:

- Define the words you use around periods - be open and honest when you talk to young women.
- What is a normal menstrual cycle and how to identify if you have heavy menstrual bleeding (HMB) - Remember 7, 2, and 1
  - **7** – Over 7 days is too long for a period to last
  - **2** – Soaking a pad or tampon in 2 hours or less is not normal
  - **1** – Passing clots the size of a $1 coin is not normal
- What is a bleeding assessment tool and where to find one
- How to report symptoms and when to seek help
- Period tracking apps and how to prepare for the first period
- Identifying symptoms of menopause.

Further information is located at:

- HFA (Hemophilia Federation of America) Blood Sisterhood App - https://www.sisterhoodapp.com
- Let's talk period - https://letstalkperiod.ca
- Factored In > Girls - https://www.factoredin.org.au/info/girls

Dr Mandy Davis discussed new clinical approaches in managing women and girls with bleeding disorders across the life span, concentrating on the available treatments for bleeding issues, how to treat low iron and diagnostic difficulties that women may face.

Dr Angela Dunford discussed how challenging menarche can be for a young woman with a bleeding disorder. The causes of abnormal menstrual bleeding can differ greatly across the lifespan and this must be taken into consideration before treating. Hormonal treatments for young women are evolving and there are options available to help control the issue of heavy periods or heavy menstrual bleeding.

Finally, Hayley Coulson discussed the importance of sport and exercise for young women and how to do this safely and in a controlled way. Importantly, she discussed what to do with an injury and when to seek further help.

Jaime Chase is the Haematology Clinical Nurse Specialist at the Children's Cancer & Haematology Service, John Hunter Children's Hospital, Newcastle NSW

Youth – challenges, taboos and myths

Jane Portnoy

Youth – challenges, taboos and myths

Chair ~ Robyn Shoemark

Growing up ~ Dale

Mother and son journey through teenage years ~ Heidi & Sam

Resilience

How to deal with cyber bullying and communication

Looking after your mental health ~ Monique Craft, Beautiful Minds

The patient voice was a strong feature of this virtual conference, and the session on youth was a great example.

Dale and then Sam and his mum Heidi spoke with a frankness, and wisdom which is true for many who have lived with haemophilia. Their presentations and the fabulous questioning from the chair Robyn Shoemark addressed many of the challenges that we see over and over again. How to allow a young person to develop and try new things, how can parents sit back and treat their child as a child first when they are worried about all the things that might happen.

I was in awe of Dale's parents who allowed him to go along and give rugby a try. The decision not to play rugby belonged to Dale and allowed him to learn how to make his own choices. Dale has tested the limits, and tried many things, he has also been a fabulous advocate for himself, and spoke up when he wanted to participate.

Heide and Sam's stories were honest, and so helpful. Thinking ahead in career planning was Sam's strategy, and it has set him up in a career of teaching that will not be limited by any physical challenges.

Heidi and Sam sharing their story and tips

TIPS FOR GROWING UP

A few of the tips from Dale, Heidi and Sam:

'Don't let anyone limit your choices. Try something at least once, and if it doesn't work out, at least you can say you tried. You only live once, and you'd rather not have regrets.'

'Don't wrap your kids in cotton wool, and don't let others do it either. Children need to thrive and try things in their lives.'

'Make treatment a normal part of everyday routine.'

'They supported me. I worked it out for myself.'

The message from all was that kids will find a way, so parental support is worth a great deal. They will also then feel able to tell you about when it doesn't go well.

TIPS FOR MENTAL WELLBEING

After setting the scene so well with the accounts from young men with haemophilia and their parents, we heard from Monique from Beautiful Minds. Her presentation was very interesting and had helpful suggestions with practical tips for young people and their families, accompanied by demonstrations.

Dealing with 'overwhelm' or the build-up of anxiety was the first issue she tackled – in particular for families 'to create harmony in the home'. I have described a few of her strategies as they were great suggestions.

The Body Thump

Stand up.

Put both your arms out in front of you, one with a fist and the other with the palm out flat.

With a gentle fist, start gently patting the opposite hand (which is out flat).

Then move up the arm and across the chest.

Then switch hands and repeat on the opposite side.

Following this with two fists gently tap down the sides of the torso, and then on the chest. Monique mentioned that making a gorilla sound helps to make this exercise even more effective.

Breathing

Monique suggested a variety of breathing exercises, including the straw technique:

Pretend you are breathing through a straw

Inhale through your nose

Exhale through your mouth, with your lips pinched like they're holding a straw

Gradually slow down the breathing, closing the eyes.

This allows the body to relax through the breathing, to get more oxygen and allows us to feel more in control.

The STOP strategy

Stop

Take long slow breaths out

Observe your surroundings

Proceed

Which allows the brain to move on through anxiety and then allow us to proceed.

54321 activity

5 things you can *see*

4 things you can *feel*

3 things you can *hear*

2 things you can *smell*

1 thing you can *taste*

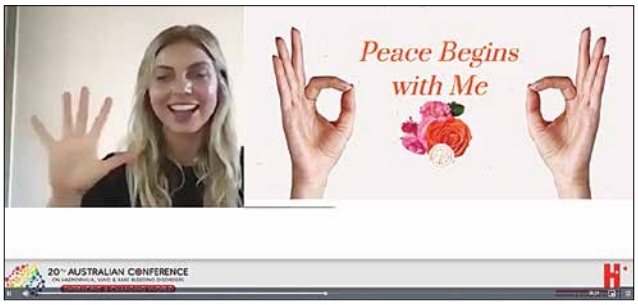
The Ice Technique

This technique is used by celebrities and public speakers.

Grab some ice and hold it. This

1. Lowers your temperature
2. Stops the adrenaline
3. It is the only thing that you can feel

Going through any of these exercises helps you take your mind off whatever is creating your 'overwhelm' or anxiety, and allows your mind to slow down, and have a break and then get back to the things you want to be doing.



CYBERBULLYING

Monique also spoke about cyberbullying, with some very useful tips.

1. Select the guests to your virtual home carefully (ie, choose who you interact with in your social media world and don't be afraid to disconnect people who are disrespectful to you and your friends)
2. The power is in no response
3. Take screen shots (as you will need to have the details and the evidence)
4. Report it
5. Block, block and block

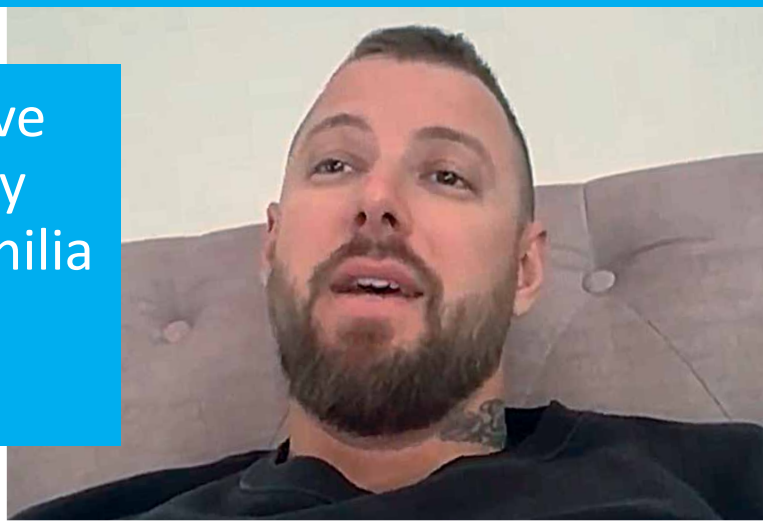
All in all, the youth session was a really thought-provoking look at some of the challenges for young people and their families and to hear tips and advice from young people themselves as well as parents and expert professionals.

Jane Portnoy is Social Worker – Haemophilia at the Ronald Sawers Haemophilia Centre, The Alfred hospital, Melbourne

YOUTH NEWS

Don’t let your bleeding disorder stop you

My name’s Dale and I have severe haemophilia A. My approach to my haemophilia growing up? I never let it stop me doing anything.



Dale spoke to HFA about his approach to haemophilia and dealing with the challenges of growing up, playing sport and working.

You can watch the video of Dale’s interview on the Haemophilia Foundation Australia YouTube channel - https://youtu.be/PdZbLu7emTI

Hey everyone, my name’s Dale. I live in Western Australia and I have severe haemophilia A, which I am treating with a non-factor therapy on a prophylactic basis.

How did you approach your haemophilia as a child?

My approach to my haemophilia, from the age when I understood what was involved with it and how it affects me, was that I basically never let it stop me doing anything.

With no family history, there was obviously a bit of worry and apprehensiveness about my haemophilia in the earlier days, but as I got a bit older, everyone accepted it and my family never stopped me from doing things, even things that I probably shouldn’t have done. But I wanted to give everything a go and they wanted me to learn from my mistakes and see what my limits were.

So, I was very lucky that my parents had that kind of mentality and that was my upbringing.

It was good because it was a choice that I was able to make for myself.

How did you go playing sport?

The first sport I wanted to play was rugby league. My family is very heavily rugby league orientated, which is weird being from WA, because everyone asked, are you from the East states, are you? And I would say, no, I was born and bred in WA.

But I said to my parents I wanted to play, so they said, okay, we’ll take you down for the tryouts. So, I went

Dale's story

down, did a bit of tackling with the bag and stuff like that. And at the end of it, decided it wasn't for me. I was landing pretty heavily on the floor and decided to give up my rugby league dreams pretty early. But it was good because it was a choice that I was able to make for myself.

So, it was basketball for me. I played at a state level. The intensity of the games was pretty full on. The guys I was playing against were very good and really athletic.

I struggled a bit to keep up, my ankles being the way they were, and I'd come away from a lot of games with twisted ankles and jarred fingers. And that was a risk that I knew I was taking. I wouldn't say I regret it, but my ankles definitely are feeling it now.

I'm an electrician now. It's been my dream job since I left school.

How did you balance your work and your bleeding disorder?

My first job was actually building boat canopies and biminis. I've done campervan building, I have done labouring.

My most notorious role was probably when I was a glazier. I did that for a very long time. And every time I told people about it, they were just gobsmacked that someone with a bleeding disorder could work with glass so much. But I enjoyed it. I was good at it. So that's why I kept on with it.

I'm an electrician now. It's been my dream job since I left school and I just got a bit lazy and veered away from it. But I finally came back to it and I'm very happy I did because it's a good job and it pays well.

Does your haemophilia have much impact on your work now?

I'm on non-factor therapy now and my health and my joints have felt better than ever. I have not had an issue with working 12-hour days, being on my feet all day, doing strenuous work.

My body has felt better than ever. I probably feel a little bit fitter and healthier than some of the guys that I work with onsite. So, I'm pretty grateful for that.

What advice would you like to give about having a bleeding disorder?

The best advice I could give someone with a bleeding disorder is don't let your condition limit your choices.

Try something at least once. If it doesn't work out, then at least you can say you tried. You only live once, and you'd rather feel fulfilled in your life than regret something for not giving it a go.

Advice I could give to parents with children with a bleeding disorder is don't wrap your kids in cotton wool. Don't let anyone else do it to them as well.

Children need to thrive and try things in their lives. My parents knew rugby wasn't a good idea, but they allowed me to try it, and I worked out for myself that it wasn't a good idea. But they supported me and that's what was important to me.

Read more

Visit Factored In - www.factoredin.org.au
- read other young people's personal stories
- find out more information about sport and working with a bleeding disorder

CALENDAR

World Haemophilia Day
17 April 2022
www.wfh.org/whd

WFH 2022 World Congress
8 -11 May 2022
https://congress.wfh.org

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BIOMARIN | CSL BEHRING | NOVO NORDISK
PFIZER AUSTRALIA | ROCHE | SANOFI GENZYME
TAKEDA

Season's Greetings

The HFA Council and Staff wish you a happy and safe festive season.

Thank you for your support during 2021 and we look forward to working with you again in 2022.

The HFA office will be closed from COB on **Thursday 23 December 2021** and reopen on **Monday 10 January 2022**.

During that time if you have any queries or need to contact HFA, call 0398857800 or email hfaust@haemophilia.org.au

Messages during that time will be monitored.

Photo by Toni Cuenca from Pexels

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