National Haemophilia
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ADAPTING TO CHANGE AND RESILIENCE

CONTENTS

Haemophilia Foundation Australia
Registered No.: A0012245M
ABN: 89 443 537 189
7 Dene Avenue Malvern East,
Victoria, Australia 3145
Postal address:
PO Box 1208, Darling,
Victoria, Australia 3145
Tel: +61 3 9885 7800
Freecall: 1800 807 173
Fax: +61 3 9885 1800
hfaust@haemophilia.org.au
www.haemophilia.org.au
Editor: Suzanne O'Callaghan

ARE YOU GETTING YOUR HFA E-NEWS?

HAVE YOU BEEN RECEIVING YOUR HFA E-NEWS OVER THE LAST FEW MONTHS?

If not, check your junk folder!

We have started using a different system for our e-news and we have heard reports that it has been treated as spam by some email services.

How to keep the e-news out of the junk folder

- Go to your junk or spam folder
- Select the e-news and click on 'not junk' or 'not spam'

Other options

- Add news@haemophilia.org.au to your contacts list

Gavin Finkelstein is President, Haemophilia Foundation Australia

FROM THE PRESIDENT

Gavin Finkelstein

COVID-19 VACCINE

As the vaccine for COVID-19 becomes available around the country, the lives of our vulnerable members and health workers at the front line will change considerably and we look forward to the vaccine being rolled out to everyone. We have provided some information on our website following questions from our members, but if you have specific questions about the vaccine and your bleeding disorder discuss this with health professionals at your Haemophilia Treatment Centre (HTC).

ADAPTING TO CHANGE

It has been an extraordinary year, and we have learned much about patience, resilience and of the importance of supporting one another. HFA has been able to adapt and continue its operations by turning possibilities into reality.

We had several parts of our digital communications project awaiting people or financial resources and we are delighted we have been able to bring more of that work to fruition than we might otherwise have done without the pandemic, because it became a priority! Digital communications have become more and more important for people to feel connected and to get their information generally, but we had to re-engineer some of the work our staff would do over the last year because they have mostly been working from their homes.

HFA has suffered on the income side of the equation and we have pulled back hard on some of our planned expenses to make ends meet. However, with the support of some very loyal donors, grants, government business support and absolute diligence we have been able to maintain productivity and manage our situation through COVID-19 without loss of staff or important services to our community. Our staff have adapted to not being able to meet face-to-face and travel but we recognise this has had an effect on our business – we have survived with Zoom meetings, teleconferences and email communications, but it's just not the same as a face-to-face discussion. We all know that, but it's OK for now!

We have been fortunate to have been able to concentrate on our digital communications project work and you will notice website enhancements that will make it easier for you to find information and news, and more personal stories and the experiences shared by people in our community about their journey of living with a bleeding disorder. The extent of the work done on both the main HFA website www.haemophilia.org.au and the youth website www.factoredin.org.au is not completely visible to users, but it involves a lot of work at the backend to upgrade and improve the websites and make them more user-friendly. The HFA website provides the framework for the websites and social media platforms for state/territory foundations as well, so continuous improvement is important for all of us.

Some people in our community have had the opportunity to consider new treatment for haemophilia. The National Blood Authority finalised its tender for extended half-life (EHL) factor VIII and IX and Hemlibra® became available for prophylaxis for people with haemophilia A with and without inhibitors late last year. It is important to discuss options carefully with HTC staff so you understand how these treatments work and what the best treatment is for you.

AUSTRALIAN CONFERENCE

We have our very first virtual national conference coming up in October 2021 - the 20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders - and we look forward to welcoming you to attend. We decided not to hold a face-to-face meeting due to the uncertainty of the pandemic, but we are very excited to be having this virtual meeting and can assure you that nothing will be lost from the conference. We have engaged a professional virtual conferencing service and all sessions will be moderated. Some will be recorded to manage time differences and for speaker convenience. The program will be rich and full, as we will have the very best Australian and international experts bringing you the most up to date information about treatment and care and living well with a bleeding disorder.

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This might be your first opportunity to participate because of distance, travel, time, cost, or simply because sitting in a conference or meeting is not your cup of tea. You will be able to register for the conference and participate in sessions at your computer in the comfort of your own home.

It is a great chance for you and your family to learn more and meet up virtually with other people living with a bleeding disorder, as well as with the broader bleeding disorders community who provide our care and treatment. There will be a registration cost for each delegate who registers, but if you need financial assistance to participate, we will make sure you don't miss out on this great opportunity.

We will be in touch with further details of the conference and financial assistance programs.

SAVE THE DATE

20TH AUSTRALIAN CONFERENCE

8 | 9 October 2021

20TH AUSTRALIAN CONFERENCE
ON HAEMOPHILIA, VWD & RARE BLEEDING DISORDERS

The **20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders** will take place this year from **8-9 October 2021** virtually.

We have decided to hold the conference virtually this year due to the uncertainty of the pandemic. We are very excited to go down this path. Bringing together the different parts of our community with health professionals and stakeholders has always been stimulating and rewarding. This year we expect our virtual conference will attract more delegates and create innovative learning opportunities and discussion for everyone. We are confident that nothing will be missing - in fact it will be enhanced.

Registrations will be available soon.

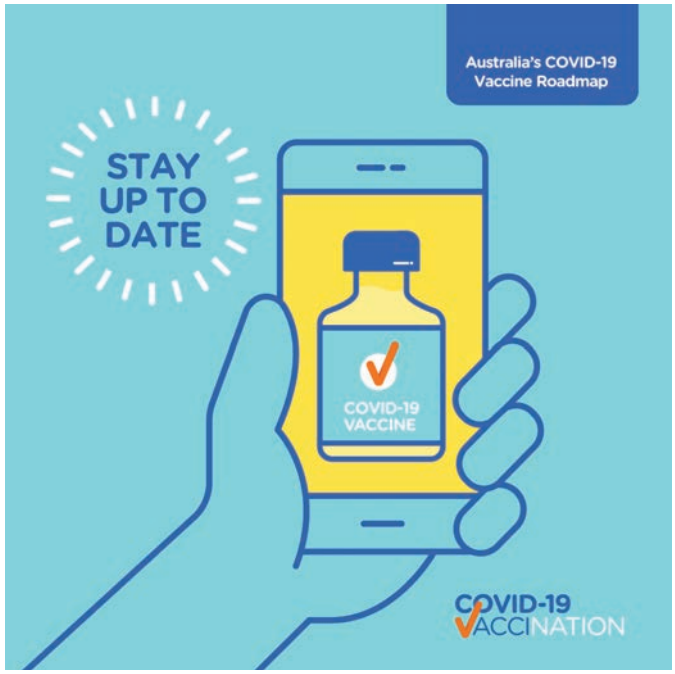
TO BE KEPT UP-TO-DATE

Register for our
Enews www.haemophilia.org.au/Signup

Or visit the conference page
www.haemophilia.org.au/conference21

For any enquiries contact:
Natashia Coco
Director of Development and Relationships
ncoco@haemophilia.org.au

COVID-19 VACCINE FAQS



With the rollout of the COVID-19 vaccine commencing in Australia, members of the community have asked us about how this will impact on people with bleeding disorders.

The Australian Haemophilia Centre Directors' Organisation (AHCDO) has advised HFA on answers to some common questions relating to the vaccine and bleeding disorders.

This includes links to the Australian Government information about the vaccine.

Read the **COVID-19 vaccine FAQs** on the HFA website - **https://tinyurl.com/BD-vaccine-FAQs**

The FAQs may be updated as more information becomes known. Please check the HFA website for updates – www.haemophilia.org.au

AHCDO has endorsed the joint **COVID-19 vaccination guidance for people with bleeding disorders,** produced by the World Federation of Hemophilia (WFH), European Association for Haemophilia and Allied Disorders (EAHAD), European Haemophilia Consortium (EHC), and U.S. National Hemophilia Foundation (NHF). This has detailed information and is available on the AHCDO website – https://tinyurl.com/AHCDO-COVID-vaccine

If you have any questions about your bleeding disorder in relation to the COVID-19 vaccine, contact your Haemophilia Treatment Centre or your treating haematologist.

HFA will continue to monitor the situation and provide updates.

ADAPTING TO CHANGE

Sustaining care in a new world

WORLD HEMOPHILIA DAY

"The COVID-19 pandemic has made life challenging for people with a bleeding disorder—but we can't stop striving for Treatment for All. World Hemophilia Day is a platform for showing the world that our community is resilient and we will overcome this new challenge as we have overcome other challenges in the past."

—CESAR GARRIDO, WFH PRESIDENT

APRIL 17, 2021 IS WORLD HEMOPHILIA DAY.

This important event is about bringing the global bleeding disorders community together. With the COVID-19 pandemic having a major impact on people with a bleeding disorder, that objective has never been more important. Our community is made up of a great diversity of people—from patients and their families, to carers, physicians and researchers—each of whom has been affected by the pandemic in a different way. We need to continue providing support to these people now, and in the future once the pandemic has passed. The world has changed greatly over the last year, but one thing hasn't: we are still in this together, and we will always be stronger together as a community in our shared vision of "Treatment for All".

On World Hemophilia Day, the WFH would love to know how you have:

INNOVATED to adapt to change in the face of adversity

DISCOVERED new opportunities to support your community

CONTINUED to advocate on behalf of your community

SUSTAINED care for patients despite the challenges of the COVID-19 pandemic

WFH

WORLD FEDERATION OF HEMOPHILIA
FÉDÉRATION MONDIALE DE L'HÉMOPHILIE
FEDERACIÓN MUNDIAL DE HEMOFILIA

WORLD HAEMOPHILIA DAY 2021

Every year on 17 April World Haemophilia Day is recognised worldwide to increase awareness of haemophilia, von Willebrand disease and other inherited bleeding disorders. This is a critical effort since with increased awareness comes better diagnosis and access to care for the millions who remain without treatment.

World Haemophilia Day was started in 1989 by the World Federation of Hemophilia (WFH), which chose 17 April as the day to bring the community together in honour of WFH founder Frank Schnabel's birthday.

This year the theme is ***Adapting to Change***. Living during a pandemic can pose many challenges, not only for our health, but also for our mental health and wellbeing. In 2021 World Haemophilia Day is a platform across the globe for exploring our community's resilience and how we will overcome this new challenge - as we have overcome other challenges in the past.

ACTIVITIES

HFA will be planning virtual events and activities – to keep up to date visit www.haemophilia.org.au/WHD and follow our social media networks.

Like Haemophilia Foundation Australia on Facebook

Like Haemophilia Foundation Australia on Instagram

Follow HFA @Haemophilia_Au

We will once again participate in **Light it Up Red** and showcase landmarks across the country.

The World Federation of Hemophilia is asking our community to write on their page worldhemophiliaday.org about how you or someone you know has adapted to change in the last year. This page will go live from 1 April 2021.

Nicoletta Crollini is Haemophilia Social Worker at the Royal Prince Alfred Hospital, Sydney

RESILIENCE - BOUNCING BACK AND MANAGING LIFE

Nicoletta Crollini

Considering the impact COVID-19 has had on all our lives over the past year, I thought it would be helpful to focus on the concept of resilience, helping us bounce back and continue on with our lives.

SO, WHAT IS RESILIENCE?

Resilience is recovering or bouncing back from the misfortunes or challenges life throws at us, which are inevitable. These life setbacks come in many forms. A few examples are relationship break-ups, being made redundant, a bleed or even a global pandemic and all the impacts that come along with it. Resilience is building up our mental and emotional strength after something challenging occurs in our life.

HOW WE DEMONSTRATE RESILIENCE

We demonstrate resilience in our everyday lives all the time. Some examples are:

- Developing achievable plans and taking appropriate steps to succeed in completing those plans.
- Having a positive view of ourselves as well as confidence in our strengths and abilities. This is our self-esteem.
- Our skills in communicating, problem-solving and being adaptable in various situations.

‘Resilience is recovering or bouncing back from the misfortunes or challenges life throws at us’

- The ability to self-regulate or manage strong feelings, emotions and impulses.
- Caring about or helping other people in need who are going through their own challenges.
- Maintaining a balanced life of study or employment, engaging in hobbies, social and cultural activities.

HOW TO DEVELOP RESILIENCE

Resilience is not something available only to a few. We are all capable of it and it is something we can build upon in preparation for the next challenge life throws at us. Here are some ways how:

- **Build positive beliefs in yourself.** This can be through achievable goals such as going for daily walks or finding something that motivates you such as cooking new recipes. Resilient people are careful where to focus their attention, so focus on the good in your life, while acknowledging the negative.
- **Other people matter!** Nobody navigates life challenges on their own. Identify your social support network - these are the people you feel comfortable confiding in and who can support you during challenging times.
- **Get some perspective.** Step back and assess your situation as objectively as possible. Is it really as bad as you think? Ask yourself, ‘is the way I am thinking or acting helping or harming me?’
- **Take action in solving a problem** instead of waiting for the problem to solve itself.
- **Be gentle to yourself and practice self-compassion**, which is being there for yourself like you would when a friend is going through a rough time. You won’t always get things right all the time. Failure is part of life but being hard on yourself does not need to be.
- **Practice self-care.** Examples of self-care are doing things like exercise, taking time out to relax, engaging in hobbies, ensuring you eat well and get a good night sleep.

HOW TO RAISE RESILIENT CHILDREN

Parents, teachers, caregivers and any important adults in children's lives can help children grow up with resilience. Here are a few ideas on how to encourage resilience in children:

- **Encourage your child to make connections and build their own support network.** Remind them that you are there for them when they experience the challenging moments in life. As previously mentioned, healthy social support networks encourage resilience.
- **Allow your child to embrace failure** as well as witnessing your own failings. Learning about failure and experiencing it is not a weakness - it is a part of life.
- **Model positive self-care behaviour for your child to learn.** This can be through the self-care activities mentioned above, such as: healthy eating, exercise, relaxing, engaging in hobbies and maintaining a good sleep routine.
- **Encourage your child's self-efficacy skills in doing things for themselves,** for example, teaching and supporting them to self-infuse.
- **Support your child in setting achievable goals,** which can be accomplished in steps. Achieving goals in steps will allow your child to reflect on what they have accomplished and what they are yet to achieve. Try not to reward your child for each step they complete but cheer them on.
- **Encourage your child to develop 'grit' and persistence.** Developing skills or achieving something in life can take time and practice is key.
- **Remind your child how they have previously demonstrated resilience** through overcoming past challenges and link their past successes to future positive opportunities.
- **Encourage keeping things in perspective** and maintaining a long-term view, especially when your child is focusing on something negative.
- **Teach your child to embrace the inevitability of change in life.** For example, goals that are no longer attainable can be replaced with new or updated goals that are more relevant.

THE TAKE HOME MESSAGE

Resilience is an important trait for us all to have.

Hopefully I have helped you understand that resilience is developed over time and a trait we can all acquire. With resilience, we work through our challenges and rise up to carry on managing our lives

To finish off, a quote by Aija Mayrock which highlights the essence of resilience:

'When you are knocked down, remember it's not what made you fall, it's what makes you get back up.'

REFERENCE

Snyder CR, Lopez SJ, Edwards LM, Marques SC. Eds. The Oxford handbook of positive psychology. 3rd edn. Oxford: Oxford University Press, 2021. DOI: 10.1093/oxfordhb/9780199396511.001.0001

WFH Global Summit on women & girls with inherited bleeding disorders

WFH staged their first ever **Global summit on women and girls with inherited bleeding disorders** in November 2020.

The Summit was an exciting two-day virtual meeting for women with bleeding disorders, patient organisations, doctors, nurses and other healthcare professionals and other supporters from around the world to exchange information and experiences. It covered:

- diagnosis and management
- quality of life
- how women are advocating for better care
- the ways women are getting their voices heard.

Some highlights from the Summit are published in this issue of *National Haemophilia*.

One of the speakers in the Summit was Australia's Susie Couper, who presented on the international VWD (von Willebrand disease) clinical guidelines and how to use them from the perspective of a patient representative. Susie has put together her reflections on the session and her personal involvement in the development of the guidelines - see page 16.

Sessions involved personal stories from women with bleeding disorders around the world – including Sharri Brodie from Perth, Western Australia, who introduced the session on diagnosis and management. The personal stories were thoughtful and compelling and highlighted the reality of the issues for women.

WFH's **Women and Girls Initiative** was also launched at the Summit. HFA Executive Director, Sharon Caris, interviewed key international leaders about what the Initiative will involve. The Initiative will support more education and training, both for health professionals and the community. You can watch Sharon's interviews on the Summit website.

ACCESS THE SUMMIT ON DEMAND

Even though the summit is over, you can still register and watch recordings of the Women and Girls Global Summit sessions on demand at https://na.eventscloud.com/VirtualWGBD/

- click on VIEW RECORDED SESSIONS
- click on NEW REGISTRATION and registerWFH will send you a confirmation email
- On the website, use your email address and your last name to login
- Go to the AGENDA and click on RECORDINGS to watch the session.

Suzanne O'Callaghan is HFA Policy Research and Education Manager

IMPROVING CARE AND QUALITY OF LIFE FOR GIRLS AND WOMEN

Suzanne O'Callaghan

Over the last several years bleeding disorders in women and girls, treatment and care and improving quality of life have taken a front seat in international forums.

There was an impressive panel of key international experts speaking at the WFH Women & Girls Summit. Rather than delving deeply into recent research, they had been invited to give an overview of the current state of play with clinical practice and research and to lay the ground for future directions – although they did refer to some recent research studies to explain why practice is changing.

At the beginning of each session a woman told her personal story of growing up and living with a bleeding disorder. I found these stories compelling. They gave a real sense of the lived experience to the presentations that followed, which often dealt with research tools and data and could be a bit distanced from the human aspects, and I thought were a great way to keep the focus on what is important.

QUALITY OF LIFE (QOL)

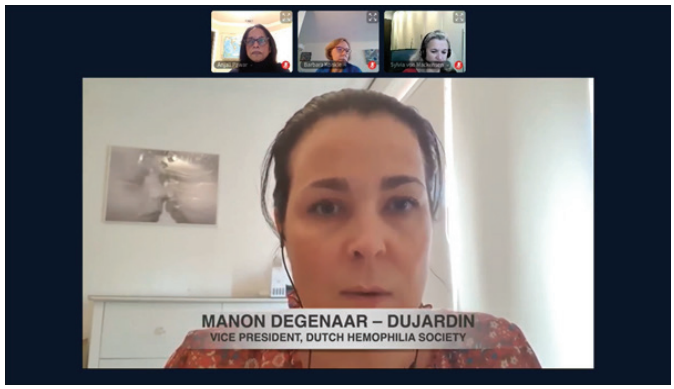
How bleeding disorders affect QoL of women and girls with bleeding disorders

Chair: Prof Barbara Konkle, USA
Physician perspective – Prof Anjali Pawar, USA
Psychosocial perspective – Dr Sylvia von Mackensen

Quality of life in girls and women with bleeding disorders has attracted increased interest internationally in recent years. This session focussed on three aspects:

- What issues have been identified?
- How can they be measured?
- How can the quality of life of girls and women be improved?

A PERSONAL EXPERIENCE



Manon Degenaar-Dujardin from the Netherlands began the session with her personal story, outlining her experiences of growing up and living with a severe bleeding disorder. She talked of the impact of her bleeding issues, having inherited both a Type 2 and a Type 3 VWD mutation from her parents. She was misdiagnosed with haemophilia as a child and grew up with all the bleeds of a severe bleeding disorder, but only blood transfusions for treatment. As an adolescent the bleeding with her first period was very severe. She was hospitalised and re-diagnosed with VWD rather than haemophilia, but accessing appropriate treatment remained a problem.

As a teenager she didn't speak of her bleeding disorder to anyone other than her sister and felt very isolated. This made managing her periods harder – she spent a lot of time in the toilets and feared people would wonder whether she was trying to escape her schoolwork. It wasn't until she went to university in Amsterdam and connected with the local haemophilia society that she learned about the local Haemophilia Treatment Centre and was able to access effective treatment. She talked of the difficulties she experienced in being believed – the common belief that only males have bleeding disorders and that VWD is only ever a mild bleeding disorder.

She spent a lot of time in the toilets and feared people would wonder whether she was trying to escape her schoolwork.

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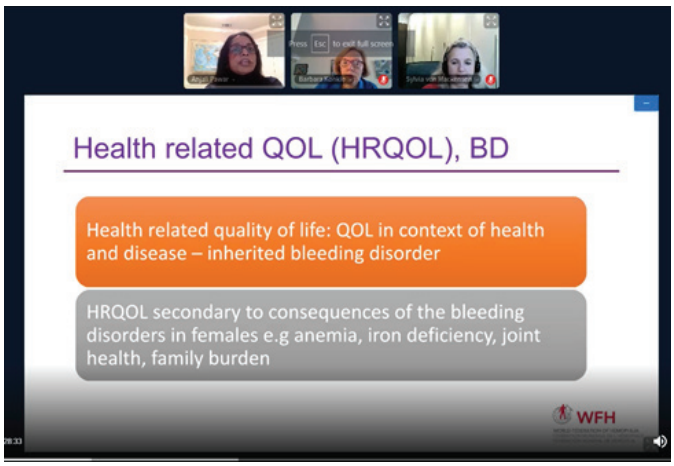
HEALTH-RELATED QUALITY OF LIFE

From the medical perspective, quality of life is a key element of care. Anjali Pawar explained that health-related quality of life involves:

- A patient's specific health care needs
- Assessing the effectiveness of meeting these needs
- Optimising treatment strategies
- Delivering optimal therapies to the patient.



Because they menstruate and can become pregnant and undergo childbirth, females have a different experience of a bleeding disorder to males. Their health-related quality of life reflects this, with issues around anaemia and iron deficiency, as well as joint health and family burden. To understand the outcomes of therapies for females, data needs to be collected on these consequences.

What kinds of measures are useful for girls and women with bleeding disorders? There is a range of standard questionnaires internationally. However, in bleeding disorders there are other significant questions.

Pawar asked the audience to rate their own experience: fatigue, psychological issues, missing school or work, participating in sport, sexual life, medical coverage, feeling embarrassed, being bullied, knowing where to find information.

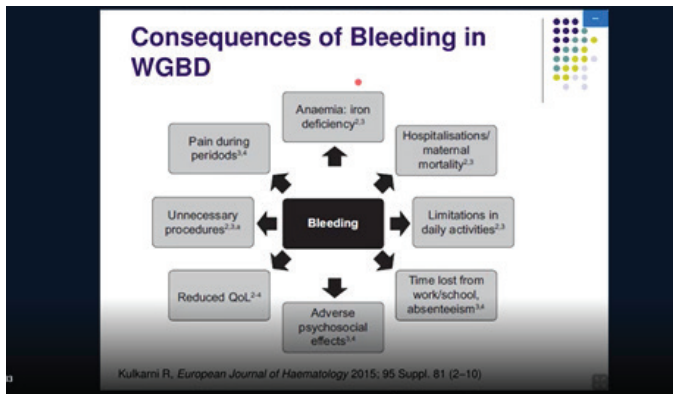
She noted a few issues that have been identified for adolescent girls:

- On average the time to diagnosis with a bleeding disorder is a few years
- Heavy menstrual bleeding is a significant problem
- Lack of education about menstruation
- Bullying at school leading to lower participation in sport.

Regular use of high-quality assessment tools for menstrual bleeding, such as pictorial charts, will help to understand the impact of menstrual bleeding on these young women. She also described the results of studies about the impact of heavy menstrual bleeding on the decision-making of women with bleeding disorders.

PSYCHOLOGICAL ISSUES

Sylvia von Mackensen looked more closely at the psychological issues underlying health-related quality of life and the studies undertaken to measure it in women and girls with bleeding disorders.



She highlighted the areas where women with bleeding disorders can experience a lack of support – from their family, the health care system and the workplace. This can lead to women feeling impaired and isolated from others.

'Regular use of high-quality assessment tools for menstrual bleeding, such as pictorial charts, will help to understand the impact of menstrual bleeding on these young women.'

Heavy menstrual bleeding, sometimes called menorrhagia, is a common symptom of bleeding disorders in females. It has a wide impact on their life – on their bleeding, pain, ability to participate at work/school and at home, in sport and on their social interaction. Embarrassment and taboos about menstrual bleeding mean that women often do not speak about these problems or seek medical help for them.

Girls and women with severe bleeding disorders showed a lower quality of life than those with mild conditions.

Both Anjali Pawar and Sylvia von Mackensen identified a range of areas where quality of life is reduced for women and girls with bleeding disorders. They noted that the challenge ahead of us is to use this information to improve their quality of life. Can it be incorporated into research about the outcomes of treatment? What are other ways to make improvements?

DIAGNOSIS AND MANAGEMENT

Diagnosis & management of women and girls with bleeding disorders - Access to care

Hematologist perspectives:
Chair: Prof Rezan Abdul-Kadir, United Kingdom
How to diagnose a woman/girl and the challenges of making that diagnosis – Assoc Prof Robert Sidonio, USA
Management of women and girls with bleeding disorders – Dr Michelle Lavin, Ireland
Access to care and outreach - Prof Roshni Kulkarni, USA

SHARRI'S STORY

Sharri Brodie, an Australian community member from Perth, started the session with a lively and honest account of her personal experiences. Sharri has mild haemophilia and von Willebrand disease (VWD). She didn't know anything about bleeding disorders until her son was diagnosed with severe haemophilia at 5 years old. After a distressing period dealing with her son's diagnosis, she eventually came to check her own factor levels and was diagnosed as a 'symptomatic carrier' herself. Looking back over her life, her many bleeding issues started to made sense – bruising easily, the black eyes, the heavy periods that she hated, the haemorrhaging with her wisdom teeth. Sharri described her guilt – about her son, for passing on the gene to him, and towards her husband, and wondered if he might have regretted marrying a 'woman with these dodgy genes'. In spite of her fears, her son has never blamed her for his condition.

What has she learned from her experiences? Sharri spoke about the importance of knowing whether you carry the gene and support for family planning, which she has found very helpful. Until she had a hysterectomy in her 40s, her menstrual bleeding was controlled with the Mirena IUD (intra uterine device). She commented on the need for parents to educate their daughters about their bleeding disorder and how to manage their periods - and how, for example, to deal with judgemental responses about being on the contraceptive pill as treatment when it is usually associated with birth control. 'Knowledge is power,' she said.

'Looking back over her life, her many bleeding issues started to made sense bruising easily, the black eyes, the heavy periods that she hated...'

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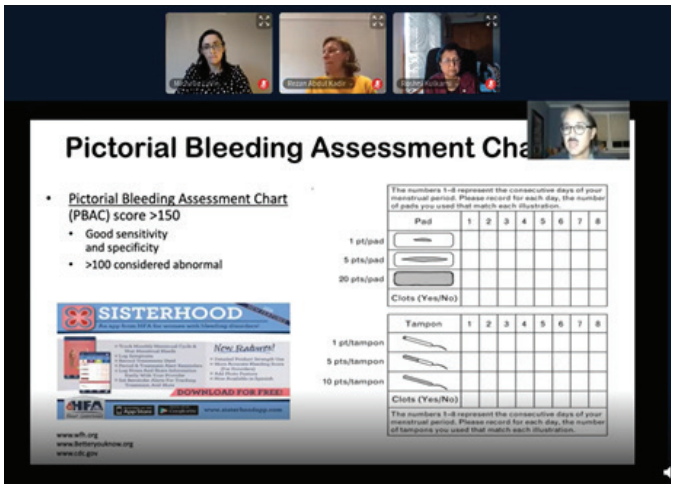
THE CHALLENGES OF DIAGNOSIS

Robert Sidonio outlined the tools that will help a physician to make a diagnosis of a bleeding disorder in a girl or woman.

BATs are bleeding assessment tools – such as the ISTH BAT and the Let's Talk Period BAT. These help to make sure the range of questions are asked to identify the possibility of a bleeding disorder.



PBACs are Pictorial Bleeding Assessment Charts, which help to quantify how much a girl or woman is bleeding during her period.

Other useful assessment tools include the **Menorrhagia-Specific Screening tool,** which looks for any of these criteria:

- Periods lasting 7 days or longer, flooding or impairment of daily activities with most periods
- History of treatment for anaemia
- Family history of a diagnosed bleeding disorder
- Excessive bleeding with tooth extraction, childbirth, miscarriage, surgery.

Sidonio noted that using several assessment and screening tools together is much more likely to identify bleeding disorders in females, particularly adolescent girls.

Other important tools include:

- A specialised coagulation laboratory
- A bleeding disorders haematology expert to interpret the results from the tests, which are very complex. Tests may need to be repeated, particularly in VWD.

Efforts to improve VWD diagnosis over the last 10 years have been paying off. Sidonio showed the increasing numbers of females with VWD being diagnosed and treated by HTCs in the USA and commented that this is an international trend.

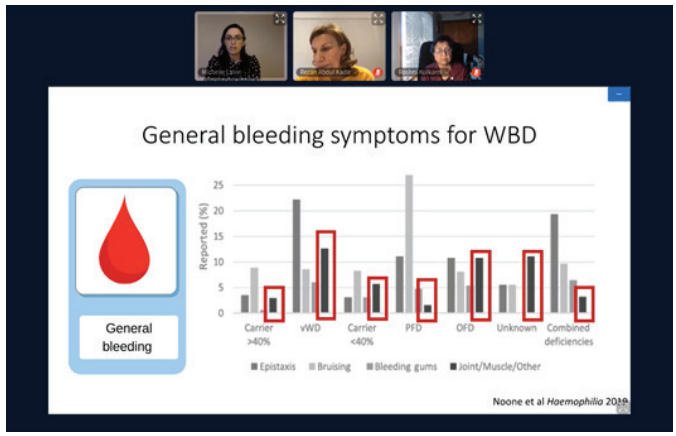
He noted that there is still work to do in the future. Many females with VWD are being diagnosed as adults and it is important to identify them at a younger age. There also needs to be more research on females who carry the gene for haemophilia.

CLINICAL MANAGEMENT

Michelle Lavin made a point of highlighting that although there are overarching principles of care, clinical management of women and girls with bleeding disorders is personalised to the bleeding issues and needs of the individual, and these can vary greatly.

She outlined the results from a recent European Haemophilia Consortium survey of women. It had some surprising results – that women and girls often reported joint and muscle bleeds and general bleeding issues as well has heavy menstrual bleeding. Menstrual problems are the most common symptom of a bleeding disorder for females, both at the beginning of menstruation (the menarche) and at menopause. Women also need support if they are using hormonal contraception as a treatment but need to stop taking it to conceive, during pregnancy and also at childbirth.

Menstrual bleeding may not be recognised as unusually heavy within a family if the women in that family all have heavy menstrual bleeding. This can lead to late diagnosis and management. Like Robert Sidonio, Lavin underlined the need to monitor menstrual bleeding objectively and mentioned a couple of apps as examples – Sisterhood and Flo Wellness & Period Tracker. There is a range of therapies available to help manage menstrual bleeding, include hormonal contraceptives if the woman is not trying to become pregnant. Iron deficiency and anaemia may also be problems and need to be treated proactively and carefully.



The HTC multidisciplinary team will need to provide support to a girl's parents and a woman's partner and family over their lifetime. Because HTCs have a family-centred care approach, it is important to use this as an opportunity to identify other people in the family who may also have the bleeding disorder.

Lavin finished her presentation noting that we are at an exciting time in bleeding disorders management, transitioning to a new paradigm of care for women and girls.

ACCESS TO CARE

Roshni Kulkarni pointed out the need for an outreach approach for women with bleeding disorders: often they are very involved in looking after their family and services and find it difficult to access services. To increase access to services, there needs to be flexibility, including telemedicine, and to consider outreach as an option, particularly for girls and women who live at a considerable distance to the HTC.

Kulkarni described the approach to telemedicine in her paediatric HTC, where a family physician/GP can conduct the consultation in collaboration with the multidisciplinary team at the HTC. Conducting telemedicine with the patient at their home also enables the HTC team to see the home environment and how the patient moves around at home. Genetic counselling can also be undertaken via telemedicine.

Telemedicine has been particularly valuable during the COVID-19 epidemic. Kulkarni noted there were some practical challenges, including communication over poor connections and some patients' lack of access to internet.

There was a clear message from the Summit that much work is still to be done. While treatment and care is personalised to the individual and their needs, we still need a better understanding of the impact of a bleeding disorder on the symptoms of girls and women. Michelle Lavin highlighted that the symptoms and issues of girls and women can vary greatly. It was helpful to hear that women and girls can use tools like Pictorial Bleeding Assessment Charts to give an objective view of their menstrual bleeding. However, data and research into general bleeding symptoms of females as well as menstrual bleeding will be important to support comprehensive care into the future.

'Telemedicine has been particularly valuable during the COVID-19 epidemic.'

Susie Couper is an Australian community member with VWD and a Haemophilia Foundation WA committee member. Susie has been one of the patient representatives on the committee to develop the international VWD clinical guidelines and is now a member of the World Federation of Hemophilia committee for women with inherited bleeding disorders.

INTERNATIONAL VWD CLINICAL GUIDELINES

Susie Couper

At the **WFH Global Summit on women and girls with inherited bleeding disorders** I had the honour of speaking about how patients can use the (now published) 2021 international *Guidelines for the diagnosis and management of von Willebrand disease*.

In the early years, when we were diagnosed with VWD (von Willebrand disease), to me it seemed to be all about the boys and all about haemophilia. Over the last five to ten years I have seen a continued and dedicated shift in people working to draw out the experience and stories of women and people with VWD. I want to contribute to the work being put in internationally to raise awareness and improve health care outcomes for women with inherited bleeding disorders. This is a deeply personal commitment – I was 27 with two sons before I was diagnosed and had improperly treated bleeding; I want to help make things better for others.

‘I view the patient involvement as an expression of deep commitment to patient-centred care and improved health outcomes’

INTERNATIONAL CLINICAL GUIDELINES

The guidelines are a result of a collaborative effort between the American Society of Hematology (ASH), the International Society on Thrombosis and Hemostasis (ISTH), National Hemophilia Foundation (NHF), and the World Federation of Hemophilia (WFH). Patient representatives were actively involved at every stage. This is something everyone can be really proud of: we were brought in early and valued at every stage, with our opinions specifically and frequently sought.

I view the patient involvement as an expression of deep commitment to patient-centred care and improved health outcomes by the collaborating organisations as well as the whole team.

SOME CONSIDERATIONS

That said, there's a few caveats with the Guidelines to be mindful of from a patient perspective. The Guidelines were developed systematically and primarily intended to help clinicians make decisions.

They are not a patient tool like the *Let's Talk Period* Self-BAT (self-administered bleeding assessment tool). If you haven't seen it, I think it is well-worth a look. I personally see it as a gold standard in terms of providing tools for personal use to make decisions. The Self-BAT is a scientifically validated scoring tool developed by Dr Paula James, a Canadian haematologist – some of you will have met her at the 2017 Conference in Melbourne. The test aims to help you better understand whether current, or previous, bleeding episodes are normal or abnormal. You can access it at the Let's Talk Period website - **letstalkperiod.ca.**

The VWD clinical guidelines don't address all treatment situations. There was a limit on the number of questions / variations that could be covered and we were confined to very rigorous methodology in question selection. The guidelines make recommendations on interventions (treatments). These may be strong or weak but that does not mean the treatment is suitable or unsuitable. It simply relates to the ability to make a recommendation based on research evidence availability. It is important to keep in mind that the guidelines are a technical document. There will be more work done on educational resources, both for health care professionals as well as patients.

The guidelines are published in the medical journal *Blood Advances*[1,2]. You might want to view them there but in terms of keeping up with everything, I'd recommend following the World Federation of Hemophilia Facebook group at **https://www.facebook.com/wfhemophilia.**

WHERE NEXT?

So as patients, how can you use them?

These guidelines aren't really ready for patient use and they aren't specific to the Australian setting. I'm sharing this with you now, however, as I want you to know that changes are on their way. I have every faith that the result will be constructive and useful. With planning and proper implementation, we'll have guidelines that bring more certainty to getting diagnosed and managing our care.

'Spread the word, tell your community. Sometimes we need to hear a message more than once to grasp it.'

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I have always wanted to approach the treatment for my and my son's VWD from a shared decision-making perspective. I was so pleased to learn that health care professionals want that too. We even had a specific session for the patient representative on the shared decision making. I'd say it is a beneficial thing to focus on what your desired outcomes are. Speak about what you want, what your priorities are. But also, to say what you don't want. During the process I learned that often our health care providers don't grasp our day-to-day experience and can make assumptions about our priorities that may not apply. There are going to be knowledge gaps - with wide variability and new information, now is a good time to question your own as well as your treating team's assumptions and look at new or different options. Be curious and informed, keep humility with pride. I have been happily surprised to be wrong about treatments we can use and learn how to treat better.

And a final tip - spread the word, tell your community. Sometimes we need to hear a message more than once to grasp it.

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1. Connell NT, Flood VH, Brignardello-Petersen R, et al. ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand disease. Blood Advances 2021; 5(1):301–325. https://doi.org/10.1182/bloodadvances.2020003264

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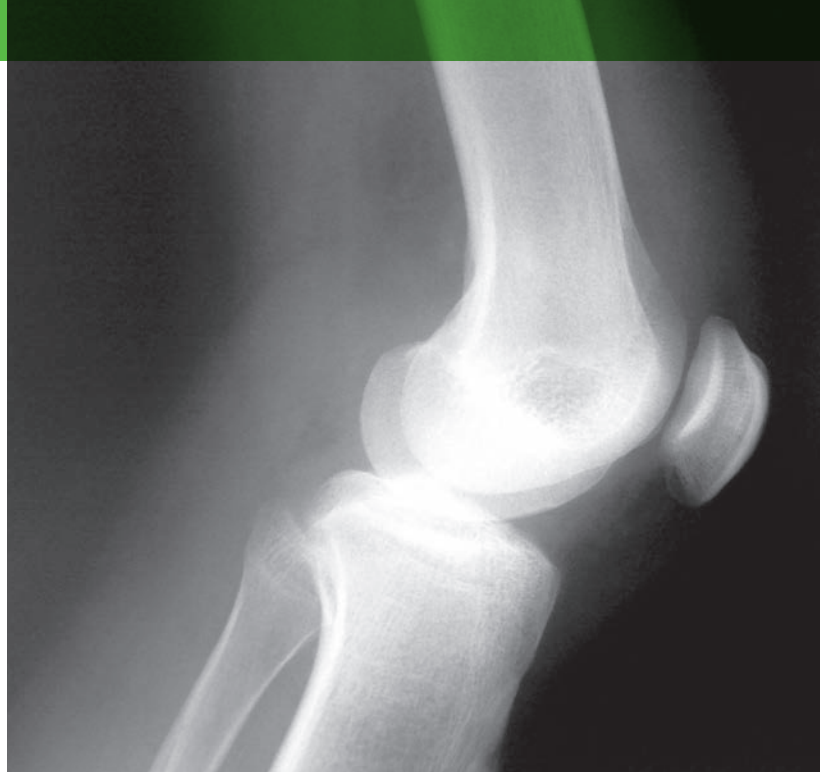
Dr Sumit Parikh is the AHCDO ABDR Senior Research Fellow

MEASURING JOINT HEALTH IN HAEMOPHILIA

Sumit Parikh



How can haemophilia treatment outcomes be measured? The Australian Haemophilia Centre Directors' Organisation (AHCDO) has been exploring the Haemophilia Joint Health Score (HJHS) as a tool to measure joint health over time as one way to show the impact of treatments.

New extended half-life (EHL) clotting factor concentrate and non-factor therapies for haemophilia became available in Australia in 2020. It came as great news for patients that there is now a range of new therapies to improve their current health and treatment outcomes with haemophilia. This looks promising for the future; however, to demonstrate this, we will need to track and monitor treatment outcomes regularly.

Bleeds and joint health are the two core measurable outcomes in the Australian Bleeding Disorder Registry (ABDR) to investigate the effectiveness of an individual's prescribed treatment regimen. Whilst AHCDO has been evaluating self-reported bleeds and uptake of MyABDR, it is also exploring HJHS as a tool to measure treatment outcomes.

The Haemophilia Joint Health Score (HJHS) is a validated, standardized joint assessment tool incorporated into the ABDR to monitor joint health status. AHCDO's study is designed to look at the frequency of HJHS usage and use of the HJHS in routine assessment of people with bleeding disorders such as haemophilia or von Willebrand disease (VWD) in Australia. The study also aims to explore the influence of geographical location as well as individual patient factors that might have an impact on the ability to perform HJHS assessment. Examining the relationships between HJHS and bleed data, including whether a patient has inhibitors, will help us determine the impact of their treatment on long term joint health.

A quick snapshot of HJHS in the ABDR demonstrates that more than 50% of patients with severe haemophilia A and haemophilia B have at least one joint score recorded. The numbers are comparatively lower (less than 30%) for the non-severe population. There were slightly more joint scores performed in the paediatric group compared to the adult group. Investigating further highlighted some factors (i.e,. non-compliance, refusal to consent, time commitment and not showing up) that particularly influenced performing HJHS in the adult group. Most of the routine HJHS assessments were performed on an annual basis, although in certain scenarios HJHS was combined with Haemophilia Early Arthropathy Detection with Ultrasound (HEAD-US) and/or radiology (eg, X-Ray) and MRI to evaluate musculoskeletal health.

The results are still evolving. However, some trends are worth noting. It appears that severe haemophilia A and haemophilia B patients who have been on routine prophylaxis for at least 6 months have a better joint health score than those on on-demand therapy. The data also shows a vicious circle, where patients prescribed routine prophylaxis but who are non-adherent bleed more, and the impact of increased bleeds is reflected in their joint health score.

There are many avenues yet to be explored in terms of joint health for people with bleeding disorders. These preliminary results have been very useful in identifying factors that influence joint health. I am looking forward to coming back with more results as we continue to improve our understanding of these treatment outcomes.

'It came as great news for patients that there is now a range of new therapies to improve their current health'

SOUTH AUSTRALIA UPDATE

We have two activities coming up for the bleeding disorders community in South Australia.

It will be wonderful to see everyone together again, meet old friends and new ones and hear the latest information about bleeding disorders.

Bleeding Disorders Community Information Evening
Tuesday 29 June 2021 (evening)
Women's and Children's Hospital, North Adelaide

Family Day
Sunday 16 May 2021
Bonython Park, Port Road, Adelaide

More information to follow. If you wish to be kept up-to-date, please email your details to hfaust@haemophilia.org.au

WHAT'S NEW ON THE GETTING OLDER INFO HUB?

New content is added to the HFA Getting Older Info Hub regularly.

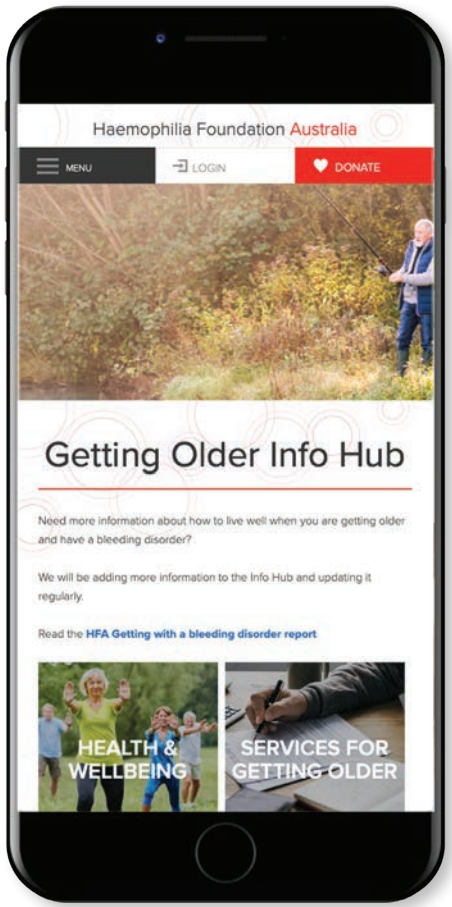
Check it out at
www.haemophilia.org.au/getting-older

WHAT'S NEW?

- HEALTH AND WELLBEING section – lots of great info on getting older and exercise
- WORK AND FINANCES section – concession cards for older people
- CONNECT TO OTHERS section – videos of personal stories

Any feedback or ideas on more topics?

Contact Suzanne at HFA on socallaghan@haemophilia.org.au or phone 1800 807 173.



RARE DISEASE DAY 2021

Rare Disease Day was celebrated globally on 28 February 2021 to raise awareness about the experiences of people with rare diseases.

Many people with rare diseases speak of feeling isolated. Many have never met or heard of another person with their condition. If they are the first in their family with the condition, or have a very rare combination, like two bleeding disorders, it may have taken a long time for them to be diagnosed. Women and girls with haemophilia also talk of not being believed because of the common assumption that only males have haemophilia.

When rare diseases are very rare and numbers are small, this can mean that the development of new and highly effective treatments is slow. There may even be no treatment that specifically targets that condition. For example, while there has been great excitement around the world about the novel therapies developed for haemophilia and the difference they make to reducing bleeds and quality of life, there is not yet a specific clotting factor concentrate that is suitable to treat factor V (5) deficiency and fresh frozen plasma may be used for treatment instead.

Following the 2021 theme of **Rare is many. Rare is strong. Rare is proud**, we shared personal stories from our community members in the weeks leading up to Rare Disease Day. This was an opportunity to acknowledge the challenges for our community members who live with a rare disorder and hear what they have learned.

In this issue of National Haemophilia, Belinda tells her story of living with factor X (10) deficiency.

'Rare is many. Rare is strong. Rare is proud'

YOUTH NEWS

Factor X deficiency

BELINDA SPOKE WITH HFA

about her journey to tackle life independently with factor X (10) deficiency.

It's the goal of many young people to leave home and see the world. But taking charge of your life to achieve this can have its challenges, especially you have a rare clotting factor deficiency.

Belinda was diagnosed with factor X deficiency as a newborn when she had the routine heel prick test and her mother noticed that it didn't stop bleeding. Neither of her two older brothers had a bleeding disorder and it was later found that there was no family history as well.

Factor X deficiency occurs in 1 in a million people and Belinda has the severe form. 'I describe it by saying my blood doesn't clot at all,' she said.

SPEAKING UP

Growing up with a very rare health condition involved watching her mother advocate and learning that she would need to speak up for herself. Even in large cities many health professionals have never had a patient with a rare clotting factor deficiency. When she was younger Belinda lived in a remote outback town. 'Things have changed now but in those days factor X deficiency was sometimes described as a type of haemophilia,' explained Belinda. 'At times the hospital staff didn't believe Mum, because"only boys have haemophilia"!'

TESTING BOUNDARIES

'During my teenage years I felt very isolated and had no connection with anyone else like me. I hated having to have factor treatment and knowing I had to do it all the time. And every time I had a period, I needed a double dose. So I got to a point in my teens where I would push the boundaries with my treatment and then get a bleed.'

Her parents understood that she needed to test the waters and try things out for herself.

'My parents were amazing. They never bubble-wrapped me - the only thing I wasn't allowed to do was ride motorbikes.'

MANAGING TREATMENT

Infusing her treatment was a struggle for Belinda. For most of her life this has involved injecting the treatment into a port, a small device surgically implanted under the skin, usually the chest.

'My veins are pretty shocking and so at a young age I got a port, something I still have today. My mum was infusing me at home, which was great, but it didn't give me any independence as she was the only one that could do it. I never thought I could leave Mum, but I was determined to learn to infuse myself via the port and become independent. By the time I was 18 I could infuse myself. I was able to leave home and move to the big city and travel the world, something I had wanted to do for so long and did do successfully.'

Treatment is still an area where she feels vulnerable.

RARE AND EMBRACING INDEPENDENCE

'By the time I was 18 I could infuse myself. I was able to leave home and move to the big city and travel the world, something I had wanted to do for so long and did do successfully.'

'My biggest fear is losing the port as I cannot infuse into my veins. Last year I got an infection in the port from a wound on my arm. I was able to get rid of the infection and get a new port, but I was very worried during that time.'

RELATIONSHIPS

Like many young people with bleeding disorders, Belinda treats her personal relationships carefully and thinks about when to tell new potential partners and how.

'Meeting new people is always a dilemma. On a first date they might see my scars and ask about them. I live with a wonderful housemate who knows all about my bleeding disorder and understands where I am coming from.' For Belinda, this means being in control of when she tells someone new about her bleeding disorder. 'We have ensured that all my supplies are in a separate fridge and not on show.'

CONNECTING TO OTHERS

Connecting to other people with bleeding disorders is important to Belinda. She started going to Foundation community camps when she was 16 years old and joined the local foundation committee as a young adult.

What are Belinda's tips for other young people with bleeding disorders?

- Reach out for support
- Be part of your local foundation, attend functions and camps
- Work at becoming independent – and you may need to figure out how to do some things yourself
- Don't let your bleeding disorder stop you (within reason!)
- Be capable of more than you think.

CALENDAR

World Haemophilia Day
17th April 2021
www.wfh.org/whd

20th Australian Conference on Haemophilia, VWD and Rare Bleeding Disorders (virtual meeting)
8-9 October 2021
Tel: 03 9885 7800
Email: ncoco@haemophilia.org.au
www.haemophilia.org.au/conference21

Bleeding Disorders Awareness Week
10-16 October 2021
Tel: 03 9885 7800
Fax: 03 9885 1800
Email: hfaust@haemophilia.org.au
www.haemophilia.org.au/BDAW

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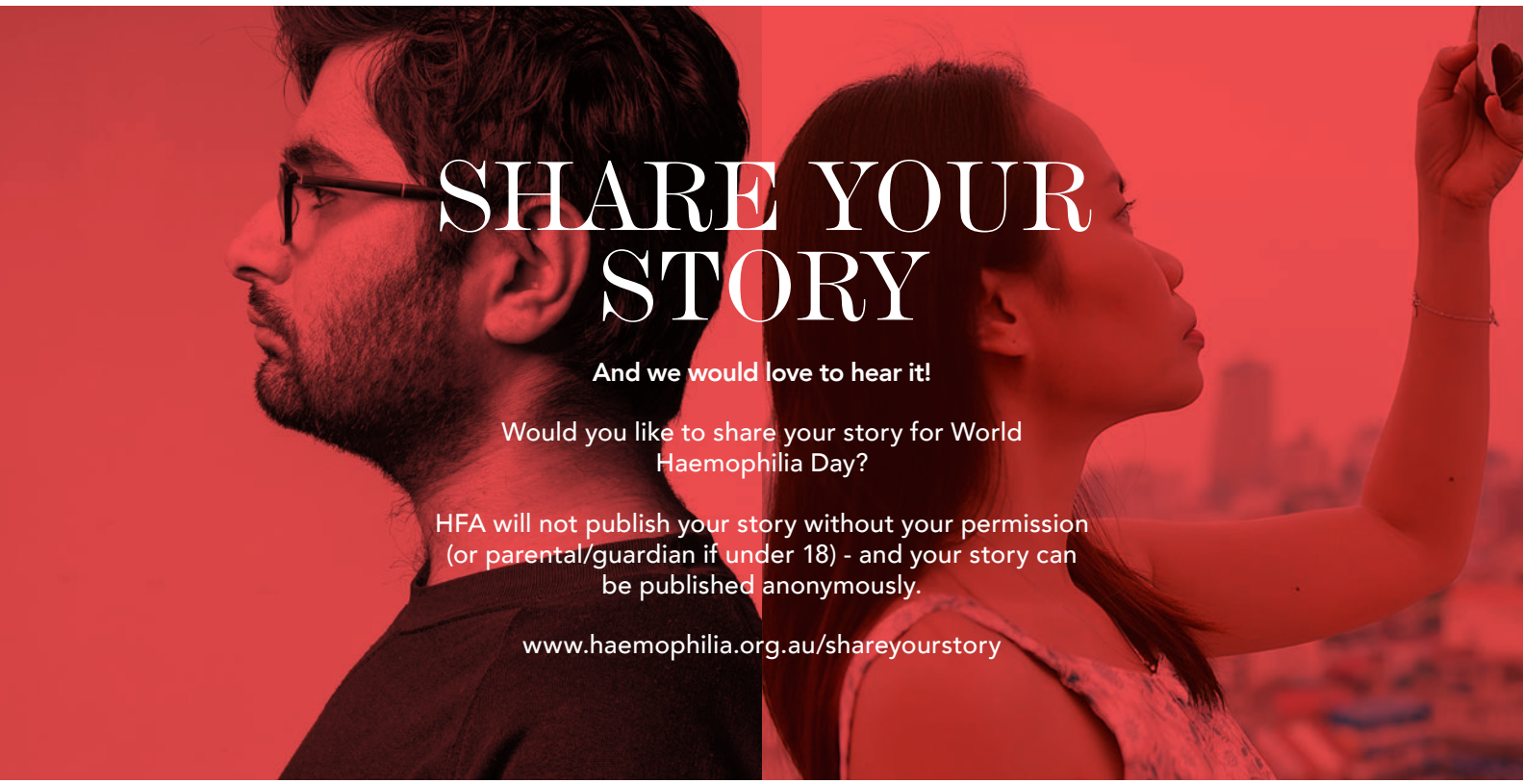
BIOMARIN | CSL BEHRING | PFIZER AUSTRALIA

ROCHE | SANOFI GENOZYME



SHARE YOUR STORY

And we would love to hear it!

Would you like to share your story for World Haemophilia Day?

HFA will not publish your story without your permission (or parental/guardian if under 18) - and your story can be published anonymously.

www.haemophilia.org.au/shareyourstory