National Haemophilia
Haemophilia Foundation Australia
www.haemophilia.org.au
No. 206, June 2019

LIGHT IT UP RED!

WORLD HAEMOPHILIA DAY 2019

CONTENTS

Haemophilia Foundation Australia
Registered No.: A0012245M
ABN: 89 443 537 189
7 Dene Avenue Malvern East,
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

BLEEDING DISORDERS AWARENESS WEEK 2019

Challenging the status quo

Bleeding Disorders Awareness Week is an opportunity for individuals and families as well as Haemophilia Foundations and other organisations to take part in a campaign and activities to raise awareness about haemophilia, von Willebrand disease and related inherited bleeding disorders throughout Australia during the week of 13-19 October 2019.

This year the theme is Challenging the status quo.

What does challenging the status quo mean to you? Look out for the personal stories from the bleeding disorders community that we will publish during Awareness Week. We invite you to share these stories with other people you know: it's through sharing personal experiences that we can connect and increase understanding in the wider community.

RED CAKE DAY

Start thinking about hosting a Red Cake Day during the week. It is a great way to involve family, friends and workplaces to take part in a special event to help raise funds during the week.

HFA can post out promotional packs for your event or awareness stand, such as stickers, balloons, tattoos, or colouring in sheets. Orders will be open in August.

To go on the Bleeding Disorders Awareness Week/ Red Cake Day mailing list, email donate@haemophilia.org.au or keep an eye on our page www.haemophilia.org.au/BDAW.

Gavin Finkelstein is President, Haemophilia Foundation Australia

FROM THE PRESIDENT

Gavin Finkelstein

OUR FINANCIAL RESOURCES AND FUNDRAISING

When anyone asks us about how HFA funds are used, we are pleased to explain because it is important that our members, supporters and stakeholders understand the work we do and how it is paid for. We aim to be very open and transparent about all our income and expenditure in line with good business practice, to comply with reporting requirements to government bodies and regulatory agencies, and because we want to be accountable to our community.

HFA prepares and submits a range of reports to government agencies, including to the Australian Taxation Office. Most external reporting is done at the end of the financial year, after external auditors have come in to our office to inspect our books and we have prepared annual reports for members at our Annual General Meeting. Annual reports are published on the HFA website. You will find our most recent publication for the 2017-2018 financial year at https://www.haemophilia.org.au/publications/annual-report, and we expect our 2019 annual report to be available in October 2019. We also provide our reports to the Victorian Department of Business Affairs as well as the Australian Charities and Not-For-Profits Commission.

HFA relies on government grants and donations and some sponsorship income to cover the cost of the work we do. Even though government grants may be for two or three years, we report each year according to our agreed proposal and plan about how we intend to use the funds received each year. As there is little opportunity for variation, we take care to align our work plans with the strategy and outcomes identified by HFA Council before we establish agreements about the funds. As the funds are provided incrementally and usually after we have completed the work, we can't afford to deviate from our plan.

The government grants include funding to cover a part of the cost of things you would expect such as rent, printing and stationery, business insurances and other office expenses. It also includes funding for the salaries of our staff who run our office, to make sure we are doing our job as the national peak body for the bleeding disorders community, and for the education and project work we undertake. Staff salaries are included as a part of our administration expenses in our financial reports and we make no apology for this because we do so much of our work 'in-house'.

HFA has not only developed a body of expertise within our staff group, which works with sound planning and goals, but we also have a team of volunteer community members and health professionals who share their expertise and enable us to be more effective than we would be if we outsourced our work. We believe that working with community is the best way of understanding needs, getting feedback and to develop our responses to meet the needs of our community. Our model for employing staff with appropriate expertise to work with the community stands us in good stead to achieve our objectives. A good example of this is our education resources - by working with the community about the information they need about their bleeding disorder we can in turn establish the collaborations needed to produce education resources that our materials relevant, timely and cost effective.

We are only too well aware of the dangers of 'having all our eggs in the one basket!'. A key strategic objective for HFA is to have a specific focus for fundraising, so that we are financially independent and capable of responding to the needs of our community when issues arise. Although we would like to be sustainable without reliance on government grants, this is not yet realistic. We are grateful for Australian Government Department of Health grants which give us the capacity to do some of our work, such as to operate the secretariat. It is a win–win situation. We receive funds from them to run our office and support our State/Territory Foundations, and produce high quality, evidence-based education resources on bleeding disorders for the general community as well as for our own members.

>>

> We are only too well aware of the dangers of 'having all our eggs in the one basket!"

As a relatively small organisation with a very specific focus on rare bleeding disorders, it is easy for our fundraising program to be swamped by other larger charities. I am proud of the way we juggle our income and expenditure to have sufficient funds to achieve the outcomes our community wants.

HFA also raises funds through traditional fundraising activities such as direct mail campaigns, events and sponsorship. Each of these need expertise, and I am proud of the work our staff and volunteers do to raise much-needed funds for our community. We have a group of caring and responsive donors who respond to our calls for support for activities run by HFA or at state/territory level such as camps and peer support workshops and so we can ensure members of local foundations have access to reliable education resources, and the benefits the national office representation and advocacy work. In the last two years we have taken action to increase our number of donors, and to nurture and encourage their support to the Foundation.

There are many challenges and it is important that we only source funds from partners who share our interests and are in line with our ethical approach. It is important that HFA decides how it will spend its money, and that we always act in the interests of our community. Sometimes people ask me how HFA handles income opportunities from pharmaceutical companies which manufacture treatment products and therefore have a strong interest in the sale of their products. HFA addresses this ethically and transparently. Our collaborations can help us raise awareness of bleeding disorders, and their sponsorship for activities such as our national conferences or other education programs are appropriate and beneficial to the community. HFA reaches out to these companies, and to governments who pay for our treatments as part of our advocacy to improve access to best practice treatments and care.

Our Annual Reports always make clear where our income has come from and the partnerships and collaborations we have fostered during each year and I am always happy to discuss this further.

TREATMENT ADVOCACY

The HFA Council has been assured by the National Blood Authority (NBA) that a new tender for treatment products funded by governments will be called in upcoming months. We believe a range of treatment products should be available to meet the needs of the community, including regular and long acting clotting factor products, and we will work with the NBA to ensure they are aware of the needs and preferences of our community. Some of these products were first registered in Australia in 2014 but, other than for an 'expanded access program', which has provided limited access to some extended half-life products, there has been no opportunity for them to be considered for public funding.

As discussed in previous *National Haemophilia* publications, we are entering an era of new treatments for haemophilia, and research and clinical trial experience suggests we are getting closer to a cure!

I believe it is important that Australians with bleeding disorders have funded access to the most effective treatment products. This includes having access to innovative treatments. We have reports from people who have shared their experience in clinical trials and are well aware of the challenges for governments when assessing their value to the Australian bleeding disorders community, but we are seeing and hearing remarkable outcomes.

Currently HFA is advocating to the Medical Services Advisory Committee (MSAC) in support of government funding for emicizumab (Hemlibra®). This is a new non-clotting factor product registered for prophylaxis treatment for people with moderate to severe factor VIII deficiency (haemophilia A) with or without inhibitors. Earlier MSAC consideration of an application from the manufacturer, Roche, for government funding for this treatment to be available for people with inhibitors found it to be cost-effective. Further consideration is now underway for the application in respect of this drug being publicly funded for people with haemophilia A without inhibitors. HFA has been invited to provide further stakeholder input and will do so using feedback provided by our members. Patients and their doctors have reported excellent outcomes with this treatment, including little or no bleeding over several years in people with severe haemophilia.

REDCLASSIC

Leichhardt Oval #3, NSW • Sunday 7 April 2019

Supported by Sanofi Genzyme

The Red Classic was a fun and free event hosted in Sydney on Sunday 7 April 2019 – it was a chance for people to walk, ride, scoot, or run the 4.5km around the bay surrounding Leichhardt to show support for the bleeding disorders community.

We had family friendly activities and fun afterwards and it was great to see people of all ages participate on the day.

Thank you to Sanofi Genzyme for sponsoring the event and to all those who attended.

2019 CONFERENCE

The **19th Australian Conference on haemophilia, VWD & rare bleeding disorders** will be held at the Novotel Manly, **Sydney, 10-12 October 2019.**

Our conferences bring together people with bleeding disorders and their families and carers, as well as health professionals, policy makers and industry. It is a great opportunity to learn, discuss and plan for the future.

PROGRAM

Chaired by Dr Liane Khoo, Director, Royal Prince Alfred Hospital in Sydney, NSW, the program committee is developing a multidisciplinary program which will interest everyone.

Keynote speakers

International expert on pharmacokinetics (PK) and measuring treatment outcomes in haemophilia, Prof Alfonso Iorio from the Department of Health Research Methods at McMaster University in Canada will be presenting on his work and contributing to the discussion about where treatments are going in Australia.

We will also have a plenary session with Dr Tim Sharp, AKA Dr Happy. Dr Sharp is an expert in human behaviour, in what makes people tick; but his focus is mostly on the promotion of positive psychology principles. His passion lies in helping individuals, teams and organisations to really thrive and flourish.

Program topics

Program topics will explore the range of what's new in bleeding disorders. Some examples include:

- Novel treatments
- New ways of providing services such as telehealth
- VWD and rare bleeding disorders
- How to live a healthy life with a bleeding disorder
- Bleeding disorders in females over the lifespan
- Managing care for children
- Getting older
- Youth issues.

And much more!

Destination NSW

19th Australian Conference

on haemophilia, VWD & rare bleeding disorders

Challenging the Status Quo

~ 10-12 October 2019 ~

SHOULD I ATTEND?

The Conference is a great opportunity for the bleeding disorders community and people working in the sector to hear the latest information and discuss current and emerging issues together. It is a niche conference, focused on the specific questions relevant to bleeding disorders, and caters for all delegates. We invite the following people to attend:

- People with haemophilia, von Willebrand disease or other bleeding disorders and their families - parents, siblings, partners – all ages welcome from young adults to seniors!
- Health professionals – doctors, nurses, physiotherapists, psychosocial workers and other health care providers
- Treatment product producers, suppliers and service providers
- Policy makers and government officials
- Haemophilia Foundation volunteers and staff.

EARLYBIRD REGISTRATIONS CLOSE 31 JULY 2019

COMMUNITY FUNDING

HFA has allocated funding to assist people living with a bleeding disorder, relatives/partners or carers to attend the Conference for expenses such as flight, registration, accommodation. Part funding applications are encouraged so we increase access and you will generally be expected to contribute towards your costs. Applications will be assessed on their merit – it is in your interests to provide full responses to the questions on the application form. Other funding options may be available from your local haemophilia foundation.

For an application form visit https://tinyurl.com/HFA-conf-funding or call HFA on 1800 807 173.

Applications close 1 July 2019.

MORE INFORMATION AND DETAILS

- Visit www.haemophilia.org.au/conferences and download the registration information and registration brochure
- Or email hfaust@haemophilia.org.au.

The WFH VWD Global Group meeting 2019
Photo: WFH

Suzanne O'Callaghan is Policy Research and Education Manager, Haemophilia Foundation Australia

PUTTING VWD ON THE MAP

Suzanne O'Callaghan

In March 2019 I attended the World Federation of Hemophilia (WFH) VWD Global Group meeting in Amsterdam and was once again impressed by the commitment of my international colleagues to our cause of achieving change for people with von Willebrand disease (VWD).

Last year HFA joined the WFH global VWD call to action. As part of this work HFA was invited to become a member of the WFH VWD Global Group, which is a WFH global working group, dedicated to addressing the unmet needs and improving the quality of life of the VWD community.

In the meeting we discussed the international response to the global VWD call to action and ways to promote it further, barriers that some countries may experience, and strategies to engage other national member organisations of WFH in the call to action.

We also launched some digital stories about peer support from women with VWD on World Haemophilia Day

http://tinyurl.com/connectBD

AWARENESS AND ADVOCACY

We each gave presentations on the work we are doing in our own country to raise awareness of VWD and incorporate VWD into our activities. Each country has their own individual issues and I was very interested to see how my colleagues approach their issues and the strategies they use with advocacy and education. Panama, for example, with limited resources has an inventive and low budget social media awareness campaign with the hashtag #IamVonWillebrand (#YOSOYVW) and educational videos on Instagram and Facebook. Pakistan has very few health care services available for people with bleeding disorders and relies on humanitarian aid for its treatment products. Although they are active in their advocacy and education, a big issue is discrimination against women with VWD, particularly in marriage. In comparison, Canada, like Australia, is well-resourced in health care services and their focus is on diagnosing people with VWD, engaging men as well as women, and supporting self-advocacy to help receive appropriate VWD care in non-HTC health settings, such as emergency departments in hospitals or preparing for surgery. I spoke about Australia's work to be inclusive, in peer support groups and through activities such as the national conference and Bleeding Disorders Awareness Week and the education materials we have developed on VWD, including the new Female Factors resources and upcoming digital stories of women with VWD. The Group was particularly interested in our *Female Factors* magazine for young women with bleeding disorders and its dedicated sections and personal stories about VWD.

WORLD HAEMOPHILIA DAY

On the second day of the meeting we workshopped some social media messages for a special WFH VWD campaign in the lead-up to World Haemophilia Day. You may have seen some of the HFA messages about VWD and the Global VWD Call to Action from this campaign on our Facebook, Instagram and Twitter platforms. We also launched some digital stories about peer support from women with VWD on World Haemophilia Day - http://tinyurl.com/connectBD.

HFA VWD FOCUS GROUP

To help us with our strategic work around VWD, HFA has established the HFA VWD Focus Group. This is a group of community members with VWD who give feedback on specific questions and will review HFA education resources. Recently, for example, they gave suggestions on topics relevant to people with VWD for the 2019 Conference program and how to promote the Conference to people with VWD. They were also invited to participate in the international VWD clinical guidelines survey: supporting the development of the Australian clinical guidelines is HFA's specific commitment to the Global Call to Action, and as the Australian guidelines will be based on the international guidelines, this was an important activity.

If you would like more information about the HFA VWD Focus Group, or would be interested in participating, contact Suzanne at HFA:

E: socallaghan@haemophilia.org.au
T: 03 9885 7800

The real-world PROBE study is now available!

PROBE FOR REAL-WORLD EVIDENCE

WHY IS THE PROBE STUDY IMPORTANT?

What is the impact of haemophilia and treatment on Australians? How can we have access to high quality evidence about this?

With new treatments becoming available this kind of evidence is particularly important. We need to be able to explain what it's like to have haemophilia and the impact of different types of treatments. HFA's advocacy relies on credible data. Without this data we have not had enough strong evidence to use in our advocacy for new treatments.

We also need good evidence to understand the different experiences of living with haemophilia – for example, women with haemophilia or who carry the gene, people with mild, moderate or severe haemophilia or inhibitors, getting older with haemophilia.

The PROBE (Patient Reported Outcomes Burdens and Experiences) study is a great opportunity for our community to give this evidence.

WHAT IS PROBE?

PROBE is a multi-national research study (www.probestudy.org) which allows people with haemophilia to report their haemophilia severity, treatment history and the impact of haemophilia on their daily life. It compares their answers to other people in their community who do not have a bleeding disorder.

You may have done the PROBE survey in the past. This was a test survey. In 2015 Australia joined more than 20 other national haemophilia organisations around the world to successfully test and validate the questionnaire.

The international PROBE team is led by well-respected haemophilia organisation and academic investigators.

This time the PROBE study is collecting real-world evidence. Statistics from Australians who complete the questionnaire will be provided to HFA by the international team for us to use in our advocacy and planning for the future.

HOW CAN YOU HELP?

You are invited to complete the questionnaire if you are **an adult (18 years+) who lives in Australia** and:

- **Have haemophilia or carry the gene**

OR

- **Do NOT have a bleeding disorder.**

You may also like to pass the survey on to your partner/wife/husband or other members of your family or interested friends. If they don't have a bleeding disorder, their answers are also very valuable – the study needs equal numbers of people affected by haemophilia and people without a bleeding disorder.

We need a few hundred Australian participants for good quality results, so the more people who complete the survey, the better!

HOW TO DO THE SURVEY

The questionnaire is available:

- Online at https://plus.mcmaster.ca/PROBE/
- Or ask your local Foundation or HFA for a print survey pack

WHAT HAPPENS TO YOUR DATA?

All responses are anonymous. They are combined for statistical data and will not identify individuals. All responses are treated confidentially. The survey is voluntary – it is up to you if you want to complete it and no one will know if you have or haven't.

TEST SURVEY

You may be aware that in May 2019 Australia was accidentally sent the test website to distribute for the PROBE real-world survey. The international team realised and acted very quickly to fix this. Links on the HFA website and our covering letter have now been corrected to the final website for PROBE - https://plus.mcmaster.ca/PROBE/. If you completed the survey on the test website, don't worry - the international team has transferred your data to the real-world website. People visiting the test website now will receive a pop-up directing them to the real-world survey website. The data security is the same - it's just a different database.

We have had a very enthusiastic response so far from the community. Our thanks to the many people who have already completed the real-world PROBE questionnaire in 2019!

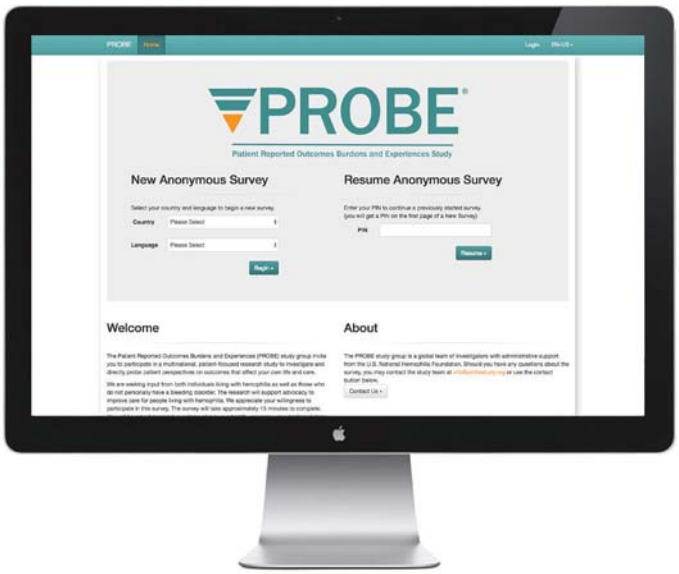
MORE INFORMATION

For more information about the PROBE study in Australia, visit

www.haemophilia.org.au/research

Or contact Suzanne at HFA:

E: socallaghan@haemophilia.org.au
T: 1800 807 173



PROBE

https://plus.mcmaster.ca/PROBE/

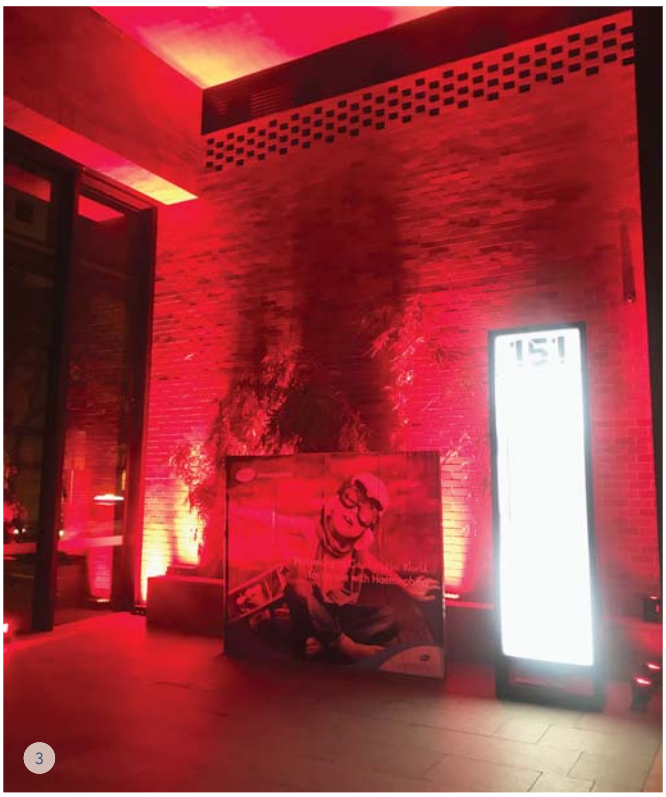
WORLD HAEMOPHILIA DAY

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.

1


2



3

THE THEME FOR 2019 WAS **REACH OUT AND CONNECT TO YOUR COMMUNITY!**

Reach out to others in the bleeding disorders community. Take opportunities to meet others with similar experiences and share your stories and tips. No matter your age or stage, connection throughout life is vital.

On World Haemophilia Day HFA launched a series of digital personal stories of Australian community members sharing their experiences of reaching out to others in the bleeding disorders community - and the value of meeting others in similar situations who understand what it's like. No matter your age or stage, connection throughout life is vital.

TO VIEW THESE STORIES VISIT HTTP://TINYURL.COM/CONNECTBD

LIGHT IT UP RED!

Together with other landmarks from around the world, Australia had landmarks turn red to celebrate World Haemophilia Day.

Thank you to the people who attended the landmarks on the night and shared their photos.

1. Adelaide Oval
2. Matagarup Bridge, Perth
3. Pfizer Australia HQ Sydney
4. Mason at Darwin Convention Centre
5. Brisbane Town Hall
6. Melbourne Observation Wheel - Andrea and Julie, HFV
7. Ballarat Town Hall, VIC
8. The Gonella boys at Mackay City Fountain

Thank you to all who supported World Haemophilia Day.

Kathryn Body is the Haemophilia and Bleeding Disorders Counsellor, Haemophilia Foundation ACT

EXPLORING PAIN

Kathryn Body

Managing chronic pain is complex and is an issue that many of my clients with bleeding disorders deal with daily. I was interested to review the experiences of one of my clients with an acquired bleeding disorder, to provide an example of the individual challenges that each person faces and the range of strategies that made a difference for my client.

David (not his real name) is a young man in his early twenties who was referred to me for counselling by his family 6 months after a bleeding disorder with associated vasculitis, for assistance with managing ongoing chronic pain and associated depressive symptoms.

David's bleeding condition is usually an acquired childhood disorder, and when it occurs in an adult the pain symptoms are known to be severe. David's medical care had been essentially symptom management with steroids and analgesics. His condition was expected to spontaneously resolve in a matter of weeks although there are cases where it has continued unresolved for months.

Until this medical issue David had an unremarkable health history, with robust physical and mental health. He had just completed a degree and was enthusiastic about his future employment plans.

David and his family became very concerned for his future when his pain levels remained high and showed no sign of decreasing in the months after the medical incident.

Counselling contact was made fortnightly, usually over a coffee in a café or a gentle walk for an hour. We talked about the difference between acute pain, chronic pain and its management and the need to educate about what could be happening in David's brain.

David and his family began reading about more recent approaches to pain, such as the research by Lorimer Moseley, David Butler and their colleagues that many of you will be familiar with.[1,2] Moseley, for example, describes how when pain persists after an injury, the system in our body that detects and transmits 'danger' messages becomes more sensitive, and sends danger messages to the brain at a rate that overestimates the true danger level. And then because the pain is (wrongly) interpreted by our body to be a measure of tissue damage, our brain presumes that the tissues are becoming more damaged. So when pain persists, we automatically assume we have persisting tissue damage.[1,2]

These are novel concepts for most of us and the researchers have put together diagrams and artwork to explain them. David commented that he really liked the cartoon-like presentations and the 'almost irreverent' way that pain was explained. The cartoons cut through a lot of words and made immediate sense to him about what he was personally experiencing.

David, his parents and some friends all ended up reading the literature about the pain research and found it really assisted their understanding about what was going on with him. They used it to educate each other and to check out what could be happening to David when he had a severe pain bout.

David was referred to a private Pain Management Clinic and after waiting a couple of months, attended a Pain Management Clinic and was a participant in a Pain Management course for a number of hours each week for 5 weeks. This clinic followed the pain approach that David and his family had been researching.

The principles in this approach are outlined by Lorimer Moseley as:

There's compelling evidence that reconceptualising pain according to its underlying biology is a good thing to do…

- Pain and disability reduce, not by much and not very quickly but they do;
- Activity-based treatments have better effects;
- Flare-ups reduce in their frequency and magnitude;
- Long-term outcomes of activity-based treatments are vast improvements.[1]

12 months after referral David still experiences some pain from his condition. He is optimistic about his future and has commenced further study and has started a part-time job. When we talked about his experiences, he commented that he wouldn't want a repeat of the last 18 months, but said he has 'learnt a lot'. He now has a greater understanding about his condition, about his overall health and how to manage his condition. He has learnt about pacing himself and recognising when his body starts to catastrophise his condition and what to do about it. He no longer 'looks for the magic bullet' but values all the resources he has been able to access.

This has also had a good outcome for David's parents: they explained they feel more in control and are grateful for all the progress their son has made in managing his condition.

From my perspective as a counsellor, this was an interesting journey, given the unique nature of any individual's pain experience. I could see that the literature, courses, learned techniques and counselling support and empowering of the client all contributed to an improved state for David and confidence in his future health.

Thanks to David and his family for their permission to share their story.

REFERENCES

1.Moseley L. Pain really is in the mind, but not in the way you think. The Conversation, 7 August 2012. < http://theconversation.com/pain-really-is-in-the-mind-but-not-in-the-way-you-think-1151 >

2. Butler DS, Moseley GL. Explain pain. 2nd ed. Noigroup: Adelaide, 2013.

This article was originally published in *The Missing Factor*, newsletter of Haemophilia Foundation Victoria (www.hfv.org.au), December 2018 and is adapted with permission.

Abi Polus is Senior Clinical Physiotherapist – Haemophilia at the Ronald Sawers Haemophilia Centre, Alfred Health, Melbourne

JOINT SURGERY

Abi Polus

Joint replacements are the last-line solution to joint management.

A discussion regarding joint surgery is common between people with haemophilia and their medical team.

It is well established that recurrent bleeds into joints can lead to arthritis. The exact mechanism of why the damage occurs is still being researched; however, it has been established that the exposure of joint cartilage (which covers the ends of bones that form a joint) to various chemicals present in blood (i.e., during a joint bleed) causes degradation of the cartilage. Without the cartilage between the bones to buffer impact, the bones may develop osteophytes (excess bony growth), or cysts (areas where bone has been lost). If bleeds are managed well - early factor, rest of the joint during the active bleed (and until pain-free movement has been restored), and rehabilitation of the joint to ensure optimal biomechanics and muscle action has been restored - then the likelihood of the development of arthritis can be minimised. The presence of synovitis (inflammation of the joint lining) and decreased joint function can increase the potential to develop arthritis.

It should be noted that some cartilage thinning and changes are a normal part of aging (like grey hair) and what can be seen on an X-ray has been demonstrated not to have good correlation with the amount of pain or dysfunction someone may experience. This article is not intended to predict everyone with haemophilia will need a joint replacement. Rather it seeks to answer some of the frequently asked questions.

First-line management for arthritis has been extensively researched and clinical guidelines recommend weight management and exercise (both general and specific - as prescribed by physiotherapists). Analgesia that is suggested by and prescribed by a doctor can also aid symptoms. Of note: Non-Steroidal Anti-Inflammatory medications or NSAIDs – for example ibuprofen, Nurofen®, Voltaren® and other medications - are often prescribed in arthritis, but have been found to increase bleeding. These should never be used unless in specific consultation with your haematologist or HTC.

Joint replacements are the last-line solution to joint management. Many questions have arisen regarding this and this article seeks to try and answer some of these.

WHAT IS A JOINT REPLACEMENT?

A joint replacement is an operation where the two ends of a bone are excised (cut away) and a synthetic material replaces the bone that was present.

WHICH JOINTS CAN BE REPLACED?

Hip and knee joint replacements are common and have a good reported success rate. This is because these joints are the most common sites of osteoarthritis in the general population and surgeons have the most experience and development with these joints.

In people with bleeding disorders the elbow and the ankle are two common sites of arthritis.

Currently there are surgeons who perform **ankle replacements,** but this is a relatively new procedure. We do not have the long-term data for this operation and there have been reports in some of the available literature of higher rates of infection and operation failure. At this stage the longer-term data does not show superior outcomes for one over the other; however, given the increased risk associated with ankle arthroplasty (infection/hardware failure), fusion remains the recommended approach. With increased time and studies this may change. We do consider joint fusion in the ankle; this is where the joint is fixed in one position, which usually gives complete relief of pain but may limit movement.

Elbow replacement is not common but can be performed if necessary. Elbow replacement can give relief from pain, but patients are limited to lifting loads of under 5kg, and stability can be an issue afterward. Other joints, for example the shoulder can (rarely) be replaced, which can be an effective procedure for alleviation of pain but typically results in diminished overhead and behind back function.

>>

Acknowledgement: Photo by rawpixel.com from Pexels

WHAT IS THE BEST TIME FOR A JOINT REPLACEMENT?

Joint replacements can technically be performed at any age; however, there are many considerations.

The operation is typically a significant procedure requiring a prolonged period of time under an anaesthetic. A person needs to be medically well enough to tolerate a long anaesthetic; for example, someone with heart or lung issues may experience difficulties with this.

The life of a prosthesis (the term for the new joint that will inserted) is quoted differently in various literature, but currently seems to be around 15 years. This will obviously depend on individual circumstances and could be much longer or much shorter. As an example, an older or less active person is less likely to put strain on the prosthesis than a younger, more active person who engages in repetitive higher loading or twisting activity. This can decrease the length of time it can be comfortably tolerated due to prosthetic loosening.

At present it is usually possible to do revision surgery (a procedure involving replacing part or all of the existing joint replacement) for total joint replacements, but this is a much larger operation and will depend on the bone-stock (how much bone and the quality of the bone) available at the operative site. One revision is usually possible, but it is not usually possible to do multiple revisions because of bone stock. For this reason, if we consider a joint replacement on a 70-year-old: in 15 or so years he may benefit from a revision, in another 15 years it is reasonable to speculate that the joint may not be the overriding issue at 100 years of age. Considering the same timeline for a 40-year-old: in 15 years and then another 15 years, this may have significant implications for life and activity if at 70 years of age we are unable to control the potential pain and loosening of an old prosthesis.

On the other hand, it is reasonable for people to want to live in comfort rather than pain and to be able to perform functionally in the way that they want to. If a person, regardless of age, is unable to perform their usual work and activities due to poorly controlled or escalating analgesic (pain relief) requirements, then this would be a consideration to discuss the possibility of having a joint replacement.

WHERE IS THE BEST PLACE TO HAVE A JOINT REPLACEMENT?

It is generally recommended that you have a joint replacement in a hospital that has a haemophilia treatment centre (HTC). This means that you will have best access to specialists who understand haemophilia, can support your factor levels (or other blood levels) and who are aware of what to do if something does go wrong! Every hospital is different and has different policies so it is worth talking to your HTC to discuss this. Typically the hospital stay for joint replacements is around 3-5 days. However, if you have a bleeding disorder, factor cover may be necessary - and with haemophilia (mild, moderate or severe) or VWD it will be. If you need factor cover, you are likely to spend 7-14 days in hospital to ensure that bleeding complications do not occur.

In Victoria the HTC advises that all joint replacements for adults should be performed at the Alfred Hospital, where the adult HTC is located. Private hospitals will not have the same expertise in managing people with bleeding disorders. Even though a private hospital may offer a shorter waiting list, it is safer to have haemophilia expertise available at the hospital where your surgery is taking place. If it is thought that a joint replacement is warranted in a patient with a bleeding disorder, the HTC and rheumatology team can sometimes liaise with the orthopaedic department regarding timing. We recommend you contact your HTC if you have joint issues, so you can be directed to the physiotherapy, rheumatology and haematology teams accordingly, with onward referral to the orthopaedics team if needed. This usually works out to be quicker than being directly referred to the orthopaedic department by your GP. Every state/territory will have a different process and it is advised that you discuss this with your HTC who will be able to best direct you.

PREVENTION IS BETTER THAN CURE!

Of course, prevention is better than cure. All bleeds should be adequately treated in a timely manner with the correct dose of factor replacement. The less time the exposure a joint surface has to blood the better. There is also some emerging evidence that weight-bearing on a joint that is bleeding is more damaging to the cartilage than not putting weight on it. (That's why your physios nag you to use crutches and completely rest the joint!) So, having enough time off the joint that is bleeding is vital. As a general rule of thumb, you should have FULL, PAIN-FREE range of movement before you weight bear on a joint that has been bleeding. However, every bleed is different so please contact your HTC physiotherapist for individual guidance.

Similarly, there is a mountain of evidence of what the best management of osteoarthritis comprises.

In the last 15 years many different treatments have been compared in rigorous scientific studies and it has been found that weight management and exercise are two interventions that can optimise management of pain and symptoms. Building muscles to support a painful joint and to regain normal control, muscle activity and walking has been shown to be extremely effective in managing symptoms. Trials involving people on a joint replacement waiting list who undertook an exercise program found that: about one-third will go on to have a joint replacement, one-third will delay a joint replacement a few years and one-third will not need to have a joint replacement at all.

BEFORE AND AFTER A JOINT REPLACEMENT

If a joint replacement is ultimately performed, it is IMPERATIVE that you complete your rehabilitation. If you have the operation without the full rehabilitation you are unlikely to get the full benefit of the joint replacement, as you may be lacking muscle power, joint control and range of movement. This can usually be performed in the hospital or at a local physiotherapist, public or private. Your hospital physiotherapist may refer you to a local physiotherapist, but if you notice increased pain, heat or loss of range of movement occurs you should contact your HTC immediately. You may need some prophylactic cover (if you do not usually have it) for physiotherapy during your rehabilitation. Talk to your local HTC for information regarding this.

There is also some research to show that an exercise program prior to a joint replacement can get you 'operation ready' and improve your outcome after the operation, as well as speed up the recovery process. Again, liaise with your physiotherapist at the HTC and this may be able to be accessed locally.

Important note - this article is general advice and information and does not take into account specific circumstances. Please discuss with your personal health care team at the HTC for specific advice.

MORE INFORMATION

Arthritis Australia - arthritisaustralia.com.au

REFERENCES

Heim M, Beeton K, Blamey G, Goddard N. Management of the elbow joint. Haemophilia. 2012 Jul;18 (Suppl. 4):101-4. https://onlinelibrary.wiley.com/doi/full/10.1111/j.1365-2516.2012.02833.x

Solimeno LJ, Pasta GL. Knee and ankle arthroplasty in haemophilia. Journal of Clinical Medicine 2017;6(11):107. https://www.mdpi.com/2077-0383/6/11/107

Srivastava A, Brewer K, Mauser-Bunschoten E, Key N, Kitchen S, Llinas A, Ludlam C, Mahlangu N, Mulder K, Poon A, Street A and Treatment Guidelines Working Group on behalf of The World Federation Of Hemophilia. Guidelines for the management of hemophilia. Haemophilia 2013;19(1):e1–e47. https://onlinelibrary.wiley.com/doi/full/10.1111/j.1365-2516.2012.02909.x

The Royal Australian College of General Practitioners. Guideline for the management of knee and hip osteoarthritis. 2nd edn. East Melbourne, Vic: RACGP, 2018. https://www.racgp.org.au/download/Documents/Guidelines/Musculoskeletal/guideline-for-the-management-of-knee-and-hip-oa-2nd-edition.pdf

Tsailas PG, Wiedel JD. Arthrodesis of the ankle and subtalar joints in patients with haemophilic arthropathy. Haemophilia 2010;16(5):822-31. https://doi.org/10.1111/j.1365-2516.2010.02248.x

Weidel J, Stabler S, Geraghty S, Funk S. Joint replacement surgery in hemophilia. Treatment of Hemophilia, no. 50. Montreal: World Federation of Hemophilia, June 2010. http://www1.wfh.org/publication/files/pdf-1210.pdf

Preetha Jayaram is the HFA Getting Older Project Officer

FUTURE PROOFING

An important aspect of the health and wellbeing of bleeding disorders community members?

Preetha Jayaram

I took up the position of Project Officer at HFA in February 2019 and, as a first step in the project, have been looking into the needs of people with bleeding disorders in the future. This has involved consulting with community members, state and territory foundations, medical specialists, haemophilia nurses, psychosocial workers and physiotherapists to explore current issues and how to 'future proof' as people grow older.

Getting Older is a priority project of HFA. The project aims to identify, understand and respond to the range of needs people with bleeding disorders may have as they are getting older and help find appropriate solutions for them and their partner/family or friends/carers.

In the second stage of the project we will look at some solutions to enable people in the bleeding disorders community and their partner/family to manage their health and wellbeing into the future as they grow older. These will be taken from the recommendations in the needs assessment. To reach the community in this digital age, this will include online options for community members to inform themselves and connect with each other. This may involve, for example, expert information about exercise with arthritis or travelling as you get older. It will be important to give a voice to men and women – both people with bleeding disorders and partners/family or carers - so that they can share thoughts about what is needed and the strategies and services they have found useful. It may also involve strengthening current peer support groups.

I am looking forward to speaking with bleeding disorders community members and their partners/family around Australia to hear the issues they see around 'future proofing' their lives.

Preetha Jayaram

PROJECT OFFICER

If you are interested in sharing your thoughts about 'future proofing' and getting older with a bleeding disorder, please contact Preetha Jayaram at HFA to talk about your availability. Partners/family also welcome.

Phone: (03) 9885 7800
Email: PJayaram@haemophilia.org.au

Joanna McCosker is the Nurse Practitioner at the Queensland Haemophilia Centre, Queensland Children's Hospital, Brisbane.

WHAT DO WE KNOW ABOUT CARRIERS?

Joanna McCosker

In haemophilia care over the last two decades there has been a greater recognition and awareness of the significant bleeding complications that women experience, in particular carriers of haemophilia.[29] In haemophilia genetics, a 'carrier' is a female with an altered factor VIII (8) or IX (9) gene that can cause haemophilia if it is passed on to her children. Sometimes she may also have a bleeding tendency herself.

It is important to identify carriers by finding out who the females are in families with affected males. Drawing family trees or 'genograms' is an important tool to capture the females who are daughter of men with haemophilia (obligate carriers) and other girls and women who may be carriers so they can be involved in discussions and education regarding factor level testing and genetic testing. It is especially important as girls reach adolescence (menarche) and reproductive age (pregnancy), as it has been estimated that for every male with haemophilia there are four to five female carriers.[22]

Little is known about how women and girls in families with haemophilia are informed or discover they are carriers. In 2016 a literature review was undertaken to better understand from the published literature how communication of inheritance or knowledge of genetic information was passed onto females in families with haemophilia.

FINDING OUT YOU ARE A CARRIER

There is a gap in knowledge and understanding of carrier status and the risk of passing on haemophilia in many affected women of reproductive age. The literature suggests that over half of all carriers are unaware of their carrier status by the time they reach reproductive age.[2,23,25] Published studies consistently show that women have poor knowledge of inheritance and are unaware they are carriers at the time of pregnancy.[2,4,5,10,11] In general, studies on communication of genetic risk in families have identified gaps in the communication of genetic information, in particular the inheritance patterns and the risk of passing on the condition. Studies found parents and other family members do share genetic information, however that information is often misunderstood.[7,15,19,27,28]

WHAT IS THE IMPACT OF THIS?

The optimal management of immediate female relatives (mothers, sisters and daughters) of a person with haemophilia is to check their clotting factor levels, especially if bleeding symptoms are present, or before childbirth, invasive medical or dental procedures (where the skin or mucous membranes/inside of mouth, vagina, anus etc are scraped or cut), or tattoos and body piercings as these may result in bleeding complications.[1] However, if female relatives are unaware of their risk of being a carrier then how will they know to seek advice from health professionals? How can optimal health management be achieved if these girls and women are not aware of their health situation or have not been diagnosed?

Many mothers of children with haemophilia, despite having a known family history, were not aware of their carrier status until they had an affected son. Additionally, many of the daughters of males with haemophilia frequently question whether they are carriers, as do their relatives, although genetically they are obligate carriers and will automatically inherit the gene from their father. It is significant to note that over half of all carriers were unaware of their carrier state until their first pregnancy or birth of an affected son.[2,4,10] Carrier status appears to often only become relevant to women when they became pregnant or after the birth of a son.[2,16,17]

An interesting fact is the average age of carrier testing was found to be 30 years of age, but the average age of carriers at first pregnancy was 26 years of age.

>>

Managing bleeding in at-risk females is a common reason for testing of clotting factor levels, as it has been estimated that 30

WHAT IMPACTS ON COMMUNICATION OF CARRIER STATUS?

The family context

Haemophilia is a familial disease and the literature recognises the influence of the family context and its social system on how and to whom information is communicated about carrier status.[9,21,26] Communication can be complex with many factors that can influence the sharing and understanding of carrier information within families.[19,28]

Assumptions of prior knowledge

This may be due to a family history, and a presumed understanding of inheritance and genetic risk information may complicate communication within families.[20,21]

Gender lines

In one study inheritance and the risk of being a carrier were found to be infrequently discussed or not at all, even in families with a father or brother with haemophilia. In general, communication was found to follow gender lines with mostly mothers and sisters discussing genetic information.[26]

Obligate carriers

Communication patterns were found to be significant in how the experience of obligate carriers differed in comparison to possible carriers. The daughters of affected fathers (obligate carriers) appeared to have less understanding of haemophilia and of their carrier status than those who grew up with an affected brother.[9]

Other studies on the psychosocial needs of carriers have also found that knowledge of haemophilia inheritance and the possibility of being a carrier is poor, even in females who have first-degree relatives.[5,24]

WHEN TO TEST: REASONS FOR AND AGAINST

The ideal age of testing for carriers is largely debated in the literature and even in Australia the age may vary between the states and territories. International guidelines recommend that carrier testing should be delayed until minors are deemed competent to understand and participate in their own health decisions OR at the age of 18 years.[3,14]

The ongoing debate about the ideal age to perform carrier testing relates to concerns about whether finding out you are a carrier will cause psychological harm or will be a health benefit to the girl or woman and is a contributing factor to delays in carrier testing.

Reasons for carrier testing in minors:

- For the health benefit of the child – to help predict and manage symptoms.[3,6,12]
- Parents' peace of mind.[27]

Reasons against carrier testing in minors:

- The child is not prepared for that information and it is difficult to explain carrier risk to minors
- Psychological harm
- The burden of disclosure, as it can be difficult for parents to decide whether telling their child is more of a burden or a benefit.[8,7,18]
- Parental distress and anxiety around informing children of their carrier status
- Concern for the impact on the child's self-esteem and social identity. [8,13,18]

On the other hand, testing for clotting factor levels is recommended earlier than 16 years of age. Managing bleeding in at-risk females is a common reason for testing of clotting factor levels,[1] as it has been estimated that 30% of carriers have reduced factor levels. However, this may lead to an incidental finding of carrier status due to a low factor level.[14] Informing children of an incidental finding of carrier status is problematic and the arguments for and against are similar to the debate around the age of carrier testing and focus on psychological harm.[5]

% of carriers have reduced factor levels

WHERE DO WE GO FROM HERE?

More research is required in this area. We need further in-depth study of the experience of carriers, in particular, exploring how genetic information is communicated in families with haemophilia. The review highlighted a lack of communication within families, resulting in a lack of knowledge and awareness of carrier status throughout all life stages, in particular prior to pregnancy. Families and health professionals may find it helpful to discuss what activities or strategies could be developed to better assist families with talking about carrier status. Young girls and women need ongoing education and support to gain a much better understanding of their carrier status.[19]

REFERENCES

1. Australian Haemophilia Centre Directors' Organisation (AHCDO). Guidelines for the management of haemophilia in Australia. Canberra: National Blood Authority, 2016. https://www.blood.gov.au/haemophilia-guidelines

2.Balak DMW, Gouw SC, Plug I, Mauser-Bunschoten EP, Vriends HJT, van Diemen-Homan JEM et al. Prenatal diagnosis for haemophilia: a nationwide survey among female carriers in the Netherlands. Haemophilia 2012;18:584-592. DOI: 10.1111/j.1365-2516.2011.02742.x

3. Borry P, Fryns JP, Schotsmans P, Dierickx K. Carrier testing in minors: a systematic review of guidelines and position papers. European Journal of Human Genetics 2006 Feb;14(2):133-8. https://doi.org/10.1038/sj.ejhg.5201509

4. Cassis FR, Buzzi A, Forsyth A, Gregory M, Nugent D, Garrido C, et al. Haemophilia Experiences, Results and Opportunities (HERO) Study: influence of haemophilia on interpersonal relationships as reported by adults with haemophilia and parents of children with haemophilia. Haemophilia. 2014 Jul;20(4):e287-95. https://doi.org/10.1111/hae.12454

5. Dunn NF1, Miller R, Griffioen A, Lee CA. Carrier testing in haemophilia A and B: adult carriers' and their partners' experiences and their views on the testing of young females. Haemophilia 2008 May;14(3):584-92. https://doi.org/10.1111/j.1365-2516.2007.01649.x

6. Fenwick A. Are guidelines for genetic testing of children necessary? Familial Cancer. 2010 Mar;9(1):23-5. https://doi.org/10.1007/s10689-009-9278-0.

7. Forrest LE, Delatycki MB, Skene L, Aitken M. Communicating genetic information in families--a review of guidelines and position papers. European Journal of Human Genetics 2007 Jun;15(6):612-8. https://www.nature.com/articles/5201822.

8. Fryer A. Inappropriate genetic testing of children. Archives of Disease in Childhood 2000 Oct;83(4):283-5. https://adc.bmj.com/content/83/4/283.

9. Gregory M, Boddington P, Dimond R, Atkinson P, Clarke A, Collins P. Communicating about haemophilia within the family: the importance of context and of experience. Haemophilia 2007 Mar;13(2):189-98. https://doi.org/10.1111/j.1365-2516.2006.01417.x.

10. Knol HM, Voskuilen MA, Holterman F, Kluin-Nelemans JC, Meijer K. Reproductive choices and obstetrical experience in Dutch carriers of haemophilia A and B. Haemophilia 2011 Mar;17(2):233-6.

11. Lee CA, Chi C, Pavord SR, Bolton-Maggs PH, Pollard D, Hinchcliffe-Wood A et al. The obstetric and gynaecological management of women with inherited bleeding disorders--review with guidelines produced by a taskforce of UK Haemophilia Centre Doctors' Organization. Haemophilia 2006 Jul;12(4):301-36. https://doi.org/10.1111/j.1365-2516.2006.01314.x

12. Lewis C, Skirton H, Jones R. Can we make assumptions about the psychosocial impact of living as a carrier, based on studies assessing the effects of carrier testing? Journal of Genetic Counseling 2011 Feb;20(1):80-97. https://doi.org/10.1007/s10897-010-9327-8

13. McConkie-Rosell A, DeVellis BM. Threat to Parental Role: A Possible Mechanism of Altered Self-Concept Related to Carrier Knowledge. Journal of Genetic Counseling 2000 Aug;9(4):285-302. https://doi.org/10.1023/A:1009428328837.

14. McConkie-Rosell A, Spiridigliozzi GA. 'Family matters': a conceptual framework for genetic testing in children. Journal of Genetic Counseling 2004 Feb;13(1):9-29. https://doi.org/10.1023/B:JOGC.0000013379.90587.ef

15. Metcalfe A, Coad J, Plumridge GM, Gill P, Farndon P. Family communication between children and their parents about inherited genetic conditions: a meta-synthesis of the research. European Journal of Human Genetics 2008 Oct;16(10):1193-200. https://www.nature.com/articles/ejhg200884

16. Myrin-Westesson L, Baghaei F, Friberg F. The experience of being a female carrier of haemophilia and the mother of a haemophilic child. Haemophilia 2013 Mar;19(2):219-24. https://doi.org/10.1111/hae.12026.

17. Naicker T, Aldous C, Thejpal R. Haemophilia: A disease of women as well. South African Journal of Child Health 2016;10(1):29-32. http://www.sajch.org.za/index.php/SAJCH/article/view/961.

18. Plumridge G, Metcalfe A, Coad J, Gill P. Family communication about genetic risk information: particular issues for Duchenne muscular dystrophy. American Journal of Medical Genetics Part A. 2010 May;152A(5):1225-32. https://doi.org/10.1002/ajmg.a.33364.

19. Plumridge G, Metcalfe A, Coad J, Gill P. Parents' communication with siblings of children affected by an inherited genetic condition. Journal of Genetic Counseling 2011 Aug;20(4):374-83. https://doi.org/10.1007/s10897-011-9361-1 .

20. Ranta S, Lehesjoki AE, Peippo M, Kääriäinen H. Hemophilia A: experiences and attitudes of mothers, sisters, and daughters. Pediatric Hematology and Oncology 1994 Jul-Aug;11(4):387-97. https://www.ncbi.nlm.nih.gov/pubmed/7947011

21. Sorenson JR, Jennings-Grant T, Newman J. Communication about carrier testing within hemophilia A families. American Journal of Medical Genetics: Part C, Seminars in Medical Genetics 2003 May 15;119C(1):3-10. https://doi.org/10.1002/ajmg.c.10001.

22.Street AM, Ljung R, Lavery S. Management of carriers and babies with haemophilia. Haemophilia 2008;14(Suppl. 3):181-187. DOI: 10.1111/j.1365-2516.2008.01721.x

23.Tedgard U. Carrier testing and prenatal diagnosis of haemophilia – utilisation and psychological consequences. Haemophilia 1998;4:365-369. https://doi.org/10.1046/j.1365-2516.1998.440365.x

24. Tedgård U, Ljung R, McNeil TF. Long-term psychological effects of carrier testing and prenatal diagnosis of haemophilia: comparison with a control group. Prenatal Diagnosis 1999 May;19(5):411-7. https://www.ncbi.nlm.nih.gov/pubmed/10360508

25. Varekamp I, Suurmeijer TP, Bröcker-Vriends AH, van Dijck H, Smit C, Rosendaal FR et al. Carrier testing and prenatal diagnosis for hemophilia: experiences and attitudes of 549 potential and obligate carriers. American Journal of Medical Genetics 1990 Sep;37(1):147-54. https://doi.org/10.1002/ajmg.1320370134.

26. Varekamp I, Suurmeijer TP, Rosendaal FR, Bröcker-Vriends AH. The use of preventive health care services: carrier testing for the genetic disorder haemophilia. Social Science & Medicine 1993 Sep;37(5):639-48. https://www.ncbi.nlm.nih.gov/pubmed/8211278.

27. Vears DF, Delany C, Massie J, Gillam L. Why do parents want to know their child's carrier status? a qualitative study. Journal of Genetic Counseling 2016 Dec;25(6):1257-1266. https://doi.org/10.1007/s10897-016-9964-7.

28. Wilson BJ, Forrest K, van Teijlingen ER, McKee L, Haites N, Matthews E et al. Family communication about genetic risk: the little that is known. Community Genetics 2004;7(1):15-24. https://doi.org/10.1159/000080300.

29.Winikoff R, Amesse C, James A, Lee C, Pollard D. The role of haemophilia treatment centres in providing services to women with bleeding disorders. Haemophilia 2004;10(Suppl. 4):196-204. DOI: 10.1111/j.1365-2516.2004.01001.x

Amy Finlayson is the Haemophilia Clinical Nurse at the Queensland Children's Hospital, Brisbane

HAEMOPHILIA SCHOOL RESOURCES

Amy Finlayson

Haemophilia, unlike other well-known conditions such as asthma and eczema, is a rare chronic medical condition. Because it is rare there are also a lot of misconceptions surrounding haemophilia, such as people with haemophilia can bleed more quickly than people without this condition and therefore cannot play sports or run around at lunch time. The problem with these common misconceptions is that the person living with haemophilia and those around them can be excluded from regular school activities.

Haemophilia treatment has come a long way over the past 20 years. In Australia, children with haemophilia are expected to lead normal lives. Participation in play and sporting activities is strongly encouraged in today's generation of children as this creates a broad range of benefits including health and fitness and peer experiences from a young age.

A common concern for teachers who have pupils with haemophilia in their classroom is that physical activities and sports can cause injuries and bleeds. Teachers may then discourage participation in activities such as playtime, football (soccer and touch), running and physical education (PE) classes. These activities, although they do not come without risk of injury for any child are also important for healthy joints and muscles, motor skill development and socialisation in peer groups.

RESOURCES FOR SCHOOLS

School years are an extremely important part of any child's life. The development of school resources at Queensland Children's Hospital was introduced as an informative tool for teachers in the management of injuries and medical emergencies for pupils with haemophilia. These medical action plans and fact sheets are guidelines that have been developed in collaboration with teachers from Education Queensland. These resources also allow for communication between health professionals and teachers and more importantly to integrate these children into school communities.

Our resources refer to the BRuCe bleed risk calculator (www.brucecalc.net), which is used by some patients and families with haemophilia to assess risk of bleeding with physical activities and sports. BRuCe has been produced by researchers at the George Institute for Global Health and Neuroscience Research Australia with funding from AHCDO, the Australian Haemophilia Centre Directors' Organisation. The calculator gives a rating of low, medium or high-risk associated with sports based on the information you input. The calculator will assess the following:

- Number of bleeds you/your child experiences within 12 months
- Average number of hours (per week) doing moderate or high-risk activities/sports (examples given)
- Average number of hours (per week) you expect to participate in moderate or high-risk activities/sports.

It is important to remember that the calculator is an only an ESTIMATE of bleeding risk as everyone is an individual.

The medical action plan (MAP) that we have developed includes:

- Child's details (name, date of birth, school year)
- The condition and severity (also includes the option for port-a-cath)
- Contact details (parent or guardian)
- Description of condition
- Medical action plan date (these plans are valid for 12 months)
- First aid measures (RICE - Rest Ice Compression Elevation)
- Signs and symptoms of a bleed
- Significant injuries (emergency management)
- Haemophilia and port-a-caths.

The Haemophilia fact sheet that we have developed includes:

- Participation in sports and physical activities
- Recommendations for sports (Bruce calculator - www.brucecalc.net)
- Absenteeism
- Returning to school after a bleed
- Helpful tips (access to classrooms when crutches/ wheel chairs are required)
- Female concerns (menstrual cycle)
- Haemophilia treatment (preventative)

In keeping with today's technology, we developed these resources to be user friendly and available online via the Children's Health Queensland website. The online resources allow families in Queensland to access this information easily and is family-centred, allowing families to be autonomous and involved in their child's care.

The feedback from families and teachers in relation to these online school resources has been positive. The online process has made the information easy to access and the forms easy to complete.

MORE INFORMATION

To download the Queensland Children's Hospital factsheets, visit

https://www.childrens.health.qld.gov.au/service-haematology-haemophilia/

For all contact sports such as football (soccer), rugby, boxing etc, speak to your haemophilia team.

The Bruce Calculator (the AHCDO bleeds risk calculator) also includes information on activity types and bleeding risks - www.brucecalc.net.

LOCAL SCHOOL RESOURCES

Each state and territory has its own way of managing health conditions in schools.

Speak to your local Haemophilia Treatment Centre or Foundation about resources and education for schools on haemophilia that is suitable for your child. They may have a school pack that they will tailor for your child. Some may be able to work with you to provide an education session for the teachers and/or students in the school, or to develop an individualised plan in collaboration with you and the school. They may also offer other types of education, such as an annual seminar for teachers.

For more information on sport and exercise for young people with haemophilia, visit the online video set and toolkit *On the move with haemophilia*. Aimed at parents, teachers and coaches, it is available on the HFA website - https://www.haemophilia.org.au/publications/haemophilia/sport

YOUTH NEWS

Andrew climbing at Wye Creek in New Zealand

Australian brothers Andrew, 30 and Scott, 37, are taking on the El Capitan climb at Yosemite National Park in California in September 2019. Andrew is an HFA youth leader. HFA will be following their preparation and adventures on Factored In and our social media.

WHY ROCK CLIMBING?

Andrew and Scott both have severe haemophilia A, with associated degenerative osteoarthritis of the ankles. They discovered rock climbing as an activity that offered a way to experience the outdoors without the physical barriers associated with many mainstream sports.

'The confidence that I've built from knowing that I can deal with the risks and challenges of climbing situations has helped me through plenty of other challenges in life. Climbing has also guided some major life decisions,' said Scott

'For me the most challenging part of climbing has always been about mental self-control,' said Andrew. *'In order to be able to climb well you must control both your body and your mind. At times this has meant enduring adverse weather like being on a climb at Pierces Pass in winter and having ice crystals rain down on top of you, or forcing myself to focus through dehydration and heat stress climbing at Montserrat in Spain, or dealing with terrifying loose rock on screen slopes in the Dolomites of Italy. All of these things are small challenges that must be overcome to continue to improve and fully appreciate the experience of rock climbing.'*

Scott on Cradle Mountain Traverse
Photo: Cameron Semple

ROCK CLIMBING CHALLENGE

Andrew - Pre-climbing factor VIII infusion, Montserrat, Spain

Check out the video of Andrew and Scott's training climb at Ozymandius - https://youtu.be/jDsOmJDYj_4

CLIMBING EL CAPITAN

Yosemite is a world-famous national park located in the Sierra mountain range in California. The best-known part of the park is Yosemite Valley, which features huge vertical granite walls on both sides. Yosemite Valley has been a mecca for rock climbers since the early 1950s. Much of the techniques, equipment and culture of modern climbing evolved in Yosemite. The valley is best known to climbers as the home of 'Big Wall' climbing, which refers to climbing long routes on vertical walls that take more than one day to complete. Nights are spent living on the wall, either on small natural ledges, or in 'portaledges' which are a kind of hanging tent that is suspended from the wall.

While there are many rock formations in Yosemite Valley that lend themselves to big wall style climbing, the biggest and most famous is El Capitan (the Captain). It's roughly a kilometre high, and almost completely vertical from the base to the summit. El Capitan was first climbed in 1958 via a route called 'The Nose' because it follows the huge nose-shaped prow straight up the middle of the highest part of the wall. Since then, there have been a number of other climbing routes established on El Capitan, but the Nose is still the most coveted by climbers from around the world. It's highly technical and involves climbing a series of crack systems, using ropes and gear both to prevent falls and to aid in upward progress. Most climbers take between 3 and 5 days to climb the Nose.

'Climbing El Capitan has been a dream of mine since I started climbing, but its only really become a specific goal in the last five years,' explained Andrew. 'In 2013 I spent a few weeks climbing in Yosemite with my partner Laura. We climbed a lot of the easier routes in the Valley, but at the time we weren't really ready for the bigger walls. One of the climbs we attempted involved spending a night on the wall, but we had to retreat when we realised that we didn't have the technical skills to get up a particularly difficult overhanging section of rock. Since that experience I've spent a lot of time learning the skills necessary to climb big wall routes. In the back of my mind I've considered all the climbing I've done for the last several years to be training for returning to Yosemite to climb El Capitan.'

TRAINING

Over Easter 2019 Andrew and Scott did a training climb at Ozymandius at Mt Buffalo National Park in Victoria. It's located on the north wall of the Mt Buffalo gorge, which is 300 metres high.

'Planning for the training climb and Yosemite, and especially with haemophilia, can be logistically complicated - planning is important,' said Scott. 'Having a plan for managing our factor supplies and infusions will also be critical. I've learnt from personal experience that I need daily infusions of factor VIII to prevent bleeds when I'm doing something physically strenuous like climbing. We'll bring enough supplies with us to ensure that we avoid bleeds, plus extra in case of emergency. Both of us are used to performing intravenous infusions, so we will be capable of treating each other in an emergency.'

FOLLOW THEIR CHALLENGE

We will post updates regularly on

- Factored In - www.factoredin.org.au
- and the Haemophilia Foundation Australia Facebook page - www.facebook.com/HaemophiliaFoundationAustralia

CALENDAR

19th Australian Conference on haemophilia, VWD & rare bleeding disorders
Novotel Manly, Sydney
10-12 October 2019
www.haemophilia.org.au

Bleeding Disorders Awareness Week
13-19 October 2019
Tel: 03 9885 7800
Fax: 03 9885 1800
Email: hfaust@haemophilia.org.au
www.haemophilia.org.au

World Haemophilia Day
17 April 2020
www.wfh.org/whd

WFH World Congress
Kuala Lumpur, Malaysia
14-17 June 2020
www.wfh.org

ACKNOWLEDGEMENTS

Haemophilia Foundation Australia (HFA) acknowledges funding grants received from the Australian Government Department of Health for communications and education activities undertaken by the HFA secretariat.

We thank the individuals, philanthropic trusts and companies which have made donations to support HFA programs, and the following companies that sponsor education programs, conferences or peer support programs run by the Foundation for the bleeding disorders community:

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

South Australia Update

Thank you to Christine for arranging a great get-together for South Australians affected by bleeding disorders in the gardens at Prospect on 5 May. It was lovely to see the children playing together on the excellent playground equipment while their parents kept an eye on them and chatted with one another. Although some people knew each other, it was an opportunity for some to meet for the first time and share their experiences.

A working group was formed during 2018 to consider how a new group can be set up to provide peer support to the bleeding disorders community in South Australia and will be meeting again soon to make further plans.