Haemophilia Foundation of New Zealand Inc.
HFNZ

bloodline

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WFH 2016 WORLD CONGRESS

Orlando, USA · July 24-28

AGM 2016

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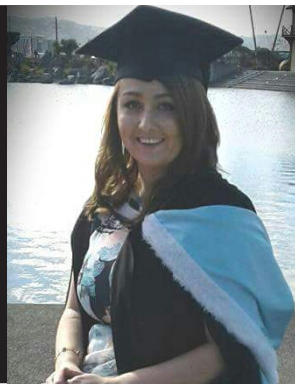

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The Word

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As the weather warms up, and the days get longer, we're all looking forward to a break over the holiday season. At HFNZ we've had another busy year, concluding with the WFH World Congress and the HFNZ AGM.

WFH World Congress is always an inspiring event for the bleeding disorders community. This year we were represented by Te Whainoa Te Wiata and Ashley Taylor-Fowlie. Ashley received the Susan Skinner Memorial Fund Scholarship and also attended the pre-congress training. Both Ashley and Te Whainoa represented us in the usual friendly and unassuming kiwi way and I know they came back with lots of ideas about how to strengthen HFNZ.

We also had staff presenting at Congress. Colleen McKay and Linda Dockrill both led sessions that were very well received. Colleen led a session on Outreach in developed countries, and Linda spoke about building resilience in children with bleeding disorders.

It was great at the AGM to see a number of regional committee members attend the training session that ran before the AGM started. It just shows how committed our people are to being the best they can be, and working to help other HFNZ members. This was reinforced at the end of the AGM, when it was my privilege to present a number of awards to members and supporters:

- The Sir John Staveley Award for Health Professionals to Julia Phillips
- The Elizabeth Berry Exercise Cup to Riley Barnes
- The McKay Trophy for MRG Activities to the Southern Region
- The HFNZ Volunteer of the Year Award to Lyn Steele

We also recognised the long service of one current and one former staff member, and farewelled a staff member who is moving on. Colleen McKay has put in a staggering 20 years as a staff member at HFNZ, and remains our ever-faithful camp mother. Chantal Lauzon, our former Information Coordinator, made it to 10 years; and we are sad to be losing Linda Dockrill, our Southern Outreach Worker, who is moving on to a new role after seven years at HFNZ. We wish you all the best Linda.

Wishing you all a restful and satisfying holiday season.

Dean York
President

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WFH 2016 WORLD CONGRESS
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Cover photo: *2016 WFH World Congress*

WFH World Congress 2016

In July this year HFNZ were represented at the WFH World Congress. This was an opportunity for some of our members and staff to get out into the world, to make connections, and to learn about what's new in the wider bleeding disorders community.

HFNZ were represented by two members, Ashley Taylor-Fowlie and Te Whainoa Te Wiata, along with our President, Deon York; our CEO, Richard Chambers; our 2IC, Colleen McKay; and two Outreach Workers, Linda Dockrill and Joy Barrett. Deon was involved as a WFH council member, while Colleen, Linda, and Ashley all were invited to present at Congress. That these strong HFNZ women were asked to present sessions on the world stage speaks well of the strength of our Foundation.

The ability of our people was also reflected in HFNZ President Deon York's election, in a landslide, to the WFH Board of Directors. This is a great achievement, and well deserved. Deon has worked tirelessly for HFNZ and the WFH over several years, and it's fantastic to see him getting this recognition. It also reinforces the good work we all do down at this end of the world with HFNZ. Deon will serve on the WFH board until the WFH World Congress in Glasgow in 2018.

Another HFNZ member making a name for themselves at Congress was Ashley Taylor-Fowlie, who won the prestigious Susan Skinner Memorial Scholarship. Scholarship recipients demonstrate outstanding leadership potential to improve the care of women with bleeding disorders in their country, and the potential to become future leaders in the bleeding disorder community.

There are only two of these scholarships awarded each year!

On top of the scholarship, Ashley was also asked to present at the National Member Organisation (NMO) training that took place before Congress. This is a real honour, and a sign of the high regard Ashley, and HFNZ, are held in.

It's fantastic that we have such a high level of talent at HFNZ, and that it's recognised internationally.

All those who attended on our behalf made sure they went to as many different presentations as possible, and have prepared the following reports.

WFH President's address

From WFH Congress Daily, July 26 2016

In 1942, The Little Prince author Antoine de Saint Exupery wrote to a friend: "Far from hurting you, being different enriches you."

Nearly 75 years later, it's still important for everyone in the bleeding disorders community to reflect on that truism, said Alain Weill, WFH president, during the Monday morning plenary "Treatment for All: Another Side of the Equation."

While much has been accomplished in bleeding disorders awareness and treatment, Weill said much remains to be done.

Recent accomplishments include the expansion of the WFH Humanitarian Aid Program. Between 1996, when the WFH started the program, and 2011, the yearly average volume of international units (IUs) of clotting factor donated to developing countries was 10.5 million. Between 2012 and 2014, factor donations increased to 25.5 million IUs per year, and they skyrocketed to 116 IUs during the last 12 months.

Along with providing more treatment options, Weill said that another objective is to use the WFH Humanitarian Aid Program as leverage to demonstrate to government officials how proper treatment enables people with haemophilia to live a normal life and actively participate in the social and economic activities of their country.

This is crucial because too many people in the global haemophilia community still face frustration, discrimination and intolerance. "This inequity drives my passion for continued advocacy," Weill said. "We must eliminate the stigma associated with having a bleeding disorder and we should not accept anything else."

Even in developed countries, it can feel like society is at odds with people with bleeding disorders and their families, Weill said. Factor costs can be prohibitive. Employment opportunities can be limited. In some countries, a person with a disability can't get a bank loan. And in numerous countries, it's not unusual for haemophilia patients to be denied services from fearful care providers.

"Along with providing more treatment options, Weill said that another objective is to use the WFH Humanitarian Aid Program as leverage to demonstrate to government officials how proper treatment enables people with haemophilia to live a normal life and actively participate in the social and economic activities of their country."

Children with bleeding disorders continue to face discrimination, Weill said. In a significant amount of countries, when a child is diagnosed with a bleeding disorder, many families experience devastating hardships. In some cases, when a child with haemophilia reaches school age, some misguided teachers believe bleeding disorders are communicable and ban the child from their classroom. "Already having to deal with their clotting deficiency, many children find themselves confronted with anxiety, depression and isolation," said Weill.

But there are steps members of the bleeding disorders community can take to fight this discrimination and intolerance. One way is to educate government and other leaders of existing legislation and regulations protecting people with disabilities. These include the Universal Declaration of Human Rights Article 1, adopted in 1948, and the United Nations Convention's Rights of Persons with Disabilities, which was approved in 2006.

In addition, the WHF has recently committed to the WFH Transform 2016 action plan, which increases the number of regional program managers around the world to assess regional situations and offer solutions.

The WFH will also become a member of Rare Diseases International, which has the prime objective of convincing the United Nations to make rare diseases an international health priority.

Weill urged each member of the bleeding disorders community to also think locally when it comes to education. "As you go through the congress experience, gather the tools that you can take back to your own communities that can help to educate and make a difference," he said.

"For when we educate people to see that being a patient of a rare bleeding disorder does not define who that individual is, and instead see them as an active member of society who enriches their community, we will show that it is indeed true that the highest result of education is tolerance."

WFH VP Medical's address: Preventing bleeds by treatment: A new era for Haemophilia

From WFH Congress Daily, July 26 2016

Haemophilia treatment has entered an exciting era, with new products making diagnosis and treatment available for a far larger population than ever before, said WHF Vice-President Medical Marijke van den Berg during her Tuesday morning VP Medical Plenary address.

The session covered a variety of studies in people with severe haemophilia A that demonstrate how early prophylaxis can prevent bleeding and is key for joint outcome. This replaces episodic therapy, which has been a frequently used haemophilia treatment regimen since the 1970s.

"Modern haemophilia treatment has completely changed the phenotype—but not in countries where early treatment is not available," she said.

> *"Modern haemophilia treatment has completely changed the phenotype—but not in countries where early treatment is not available"*

Ms. van den Berg cited a very large U.S. study of patients with severe haemophilia A, divided into four birth-date cohorts. Even in the age group born in the 1980s, disability was too high, she said, with more than five joint bleeds over six months, despite very high clotting factor consumption.

The large, international Musculoskeletal Function in Haemophilia (MUSFIH) study of children with severe haemophilia A showed that even very high factor dosing resulted in substantial bleeding, van den Berg said. The study also showed that the number of bleeds—but not the dose of episodic treatment—is responsible for joint outcome. This is a key understanding because joint function deteriorates after age 12.

Ms. van den Berg cited a small, randomized study showing that low-dose prophylaxis, rather than episodic treatment, reduces bleeding by 80 percent. Research also shows that early diagnosis is crucial. "Remember, more than 50 percent of those with severe haemophilia A have a negative family history," van den Berg said.

But when and how do you start prophylaxis? van den Berg said research suggests that the key is to start earlier than age 3 because physical examination scores increase with treatment delay. Other research shows that low-dose prophylaxis should be done a minimum of once a week.

There is a correlation between joint scores and dosage of factor replacement. "With a 1,000-1,500 dose, there's a lot to gain," van den Berg said. The good news for people in developing countries, where factor supply is limited, is that data show that lifetime prophylaxis with 1,000 IU per kilo is much more effective than episodic treatment. "You can significantly improve outcome with limited factor consumption," she said.

However, to implement low-dose prophylaxis, comprehensive care centres are crucial, van den Berg said. She recently toured two international haemophilia training centres that are excellent examples of this: the centre in Campinas, Brazil, led by Margareth Castro Ozelo, and the centre in Johannesburg, South Africa, led by Johnny Mahlangu. The Johannesburg centre serves 1,200 patients, with an impressive 35 percent on prophylaxis and home therapy.

Unfortunately, these centres are the exception. Recent data from Africa show that not even 5 percent of haemophilia patients are diagnosed. "The main reason is because limited or no treatment is available," van den Berg said.

The WFH Humanitarian Aid Program will substantially address that deficit. From 2016 to 2020, the program plans to provide a predictable supply of 500 million IUs of factor, van den Berg said. Availability of products will lead to more diagnosis, and that will lead to more training and, in some cases, corrective surgery.

In conclusion, van den Berg said that the evidence shows that only primary prophylaxis can prevent joint disease, and episodic treatment is not an appropriate regimen for severe haemophilia A. After joint bleeds, signs of arthropathy appear even with very high-dose prophylaxis. And signs of loss of joint function are often visible at puberty due to growth spurts.

Ageing with Haemophilia

By Te Whainoa Te Wiata

Joint replacements

The session spoke on current goings on in the bleeding disorder community, specifically our ageing men and women. Our community is getting older due to advancements in medicine, however, now there are other things that are starting to cause concern. There was talk on joint replacements and making sure the rehabilitation stage after these procedures was followed through properly, especially if the replacement joint is too rigid. When a joint replacement procedure has taken place, clinicians are finding that patients often worry too much about regaining strength and aren't worried enough about re-training their balance. The specialist went on to say that most injuries that take place after surgery are due to loss of balance, emphasising the importance of a good rehabilitation regime that focuses on both strength and balance.

Dental Care

Another thing worth mentioning is dental care, and how important it is that we look after our teeth, no matter what age. Andrew Brewer of Queen Elizabeth Hospital Glasgow, spoke on the importance of good dental hygiene. Dry mouth from pain killers can be an issue, as saliva helps to keep teeth strong, so drinking water is a high priority. It is also important to make sure that you use fluoride toothpaste when brushing.

Joint damage in arms is something that can also inhibit caring for your teeth and dentures, however, it is most important for people to find ways of keeping up good oral care. The most important thing mentioned was that bleeding in your mouth is NOT due to a bleeding disorder but rather to bad oral hygiene, plaque can build up and begin to cause damage within 24 hours so flossing and brushing twice a day with fluoride toothpaste is the best medicine.

Adverse event reporting

By Te Whainoa Te Wiata

Understanding the importance of reporting adverse events is something that still sits in the too hard basket. As a patient that has not been the most diligent at reporting adverse events (AE), my less than desirable actions are generally due to a lack of understanding. The presentations given have cleared up a lot of the mist that surrounds the issue and while presenting some of the success that is being experienced by other NMO's. In addition to that, it is both the healthcare providers and patients that are addressing the issue.

The first question asked was 'what is the difference between an adverse event and medical error?'

This question was addressed by the first presenter, Michael Makris, who is the Director of the Sheffield Haemophilia and Thrombosis centre in the U.K.

- Medical error: If a patient is factor XI patient and is prescribed factor VIII, then this is a medical error.
- Adverse event: Is what happens when a patient has a reaction to some type of product. For example, 10 minutes after having treatment a rash appears or dizziness occurs.

It is important to be able to differentiate between the two. The main thing this session was trying to address is if the healthcare providers don't know that these things are taking

place then they are unable address the issue. Also, any and every AE reporting is important, it not only helps the care providers better accommodate a patient's individual needs, it also provides vital information that is required on a national and global level.

There are many new trials taking place with longer acting treatment and other medical advancements, and the data collection process needs to be plentiful. A lack of reporting can produce many unwanted outcomes, including people suffering for no reason, and a lack of security of safe treatment.

In France, healthcare providers and patients have taken an active role in educating on the importance of AE, and this has had positive outcomes for them. This intervention saw the creation of Mathieu, an online educational cartoon which shows what to do in the case of an AE. It was said that there is still a long way to go, but improvement is happening. There are many different steps being taken all over the world for reporting to ensure safe treatment, but ultimately the take home note from this session was that AE is of high importance and needs to be plentiful to secure safe and usable products for all patients.

Alternative therapies in the management of inhibitors (approach to haemostasis)

By Richard Chambers

The coagulation system is composed of cells, proteins and processes that mediate blood clotting. Damage to a blood vessel

wall triggers the system, initiating interactions between platelets and blood coagulation factors. This leads to the formation of a fibrin-containing clot, which stops bleeding and initiates vessel repair. With a bleeding disorder something is missing that prevents some of those interactions occurring. Doctors have traditionally sought to replace the missing piece, the missing factor, which can at times cause an immune response. Researchers including Professors Midori Shima, Rebecca Kruse-Jarres, and David Lillicrap are now looking at new or novel approaches to managing haemophilia.

Mechanisms of Tolerance (Lillicrap)

- FVIII immunogenicity continues to be a treatment complication
- Standard ITI therapy is inconvenient, costly and unsuccessful in 39% of patients
- Novel tolerance induction protocols have shown promise in animal models
- Consideration is being given to pilot phase clinical studies employing novel primary (preventative) and secondary tolerance induction strategies: eg. Transient anti-inflammatory intervention and FVIII gene therapy
- There is still a lot to learn about FVIII immunogenicity and immune tolerance.

Porcine VIII (Kruse-Jarres)

- Recombinant porcine (rp) FVIII is haemostatic
- Can monitor with FVIII levels
- No allergic reactions, no thrombocytopenia
- Some cross reactivity at baseline – affects recovery and half-life
- rpFVIII is an effective haemostatic agent for patients with inhibitors to FVII
- Effect is dynamic, there is potential for prohibitive cost when compared to current options depending on the protocol used.

FVIIIa mimicking bi-specific anti-body (Shima)

The goal is to remove the necessity of intravenous infusions, using a long acting treatment that makes treatment easy, with no inhibitors.

Three novel therapeutics, FVIII mimicking bispecific antibody (Emicizumab or ACE910), and treatments that seek to rebalance the coagulation system; siRNA targeting anti-thrombin (Fitusiran), anti-TFPI anti-body (Concizumab) are under clinical trials.

So far:

- No severe drug-related Adverse Drug Event has been reported
- Subcutaneously injectable
- Highly effective irrespective of the presence of inhibitor
- Emicizumab has a much improved PK profile when compared to current prophlaxis
- Current testing methods are not all appropriate to these new treatments

These therapeutics may provide new concepts of prophylactic treatment for haemophilia, however more data on safety, efficacy are required.

Bleeding and Women: Time for a paradigm shift

By Linda Dockrill

One aspect of this Congress that was very different from previous events was the focus on women and bleeding. This session's speakers noted that the numbers of women being diagnosed with bleeding disorders has been showing strong growth. In the USA there has been a 300% increase between 1991 and 2007. While those statistics show a growing awareness of bleeding disorders for women, one recent study of women with Von Willebrands disease found that women had waited an average of 16 years before seeking help!

Tatjana Markovic spoke of her experiences in Serbia, where treatment availability is low, and therefore treatment of carriers is also low. The numbers of men with a bleeding disorder show that there should be many more carriers registered than they currently have (7 carriers and 438 men with haemophilia). What is interesting with this information in the background is that women with Von Willebrands have good access to treatment. Tatjana also spoke of the psychosocial aspects of being a carrier, and how the denial, self-blame, shock, frustration, fear, and hopelessness has affected her as a mother of a son with haemophilia. It was powerful to hear how her son's haemophilia has impacted on her quality of life.

Our own Mary Brasser, Clinical Nurse Specialist from Auckland Haemophilia Treatment Centre (HTC), spoke in this session as well. Mary identified that as many as 1% of women may have a bleeding disorder. Many women are unaware of their condition and the "growing body of evidence shows that the quality of life of women with bleeding disorders (vWD in particular) results in increasing limitations in daily activities and time lost from work and school."

Mary discussed why clinics for carriers are not run, saying the main reasons were listed as financial, lack of clinic space and time, lack of haematologist and obstetrician time, and nursing resources. Mary shared that Auckland HTC do not currently do individualized care plans for women/carriers.

Yannick Cole from France identified that a third of carriers with haemophilia have bleeding episodes. They also have to fight against delayed diagnosis and a general lack of information and knowledge in medical personnel. Yannick said "describing their illness as haemophilia would be easier if haemophilia is a shared experience". To help make change, a women's committee has been established in France to provide information to clinical staff and to increase the communication between patients and medical professionals.

To complete the session there was some discussion around some of the challenges in the diagnosis of bleeding in women. These were listed as social stigma (around menorrhagia, postpartum haemorrhage etc.), symptom minimisation, absence of overt

signs, range of severity, inadequate tools to measure bleeding, difficulty accessing testing, and resource allocation issues. The speaker showed how a move to a new model of care, one that placed the patient at the centre and improved patient access to information, education, and tools, needs to happen. One of the tools mentioned was available on **http://letstalkperiod.ca/.**

Colouring outside the lines: WFH 2016

By Te Whainoa Te Wiata

This session was very brief, but informative at the same time. It specifically looked at alternative ways to treat patients with haemophilia, especially those who lack access to treatment.

Pamela Narayan is a physiotherapist from India, who has been treating patients in the bleeding community for around 15 years, and is also an advocate for the importance of physiotherapy, orthotics, and braces. She spoke, from her own personal experience of treating patients with limited access to treatment, on how physiotherapy can help decrease joint bleeds. This includes the use of braces to keep joints supported during the healing process, even using them as a preventative during sleep, as bleeds often reoccur at this time.

Orthotics and shoe inserts is another topic stressed. Through correcting height, and balancing out the weight bearing load, joint damage can be reduced. So too can bleeds into particular joints, especially target joints. The overall outcome is that patients are noticing improvements through physiotherapy and Pamela's methods.

Frederica Cassis is a psychotherapist from Sao Paolo. Frederica presented a few of her projects where she not only treated patients, but also provided support for the family. Like the first speaker, this presentation was focussed around providing care for those with limited access to factor product. The speaker opened with this little slogan:

"No prophylaxis = emotional prophylaxis"

If there is no product then there are other ways to provide care, like teaching the patient about haemophilia through play, drawing, and drama. In addition, the follow on is learning about prophylaxis, and encouraging the patient to think about their future and what that will look like. For many, identity is the starting point, who are you and where do you come from? It is also important to highlight self-esteem, to develop acceptance and improve self-diagnosis. Other methods used are:

- Flash cards of the clotting process, which the patient must put into order
- A play where patients act out the clotting process
- Take home booklets for families
- Breathing techniques for pain and anxiety

Frederica closed by saying that psychosocial-interventions are not new, and should be used in conjunction with treatment (if any), but that the focus should be more about the patient taking ownership of their disorder, giving power to patients and families to accept haemophilia.

Rachel Tiktinsky is a physiotherapist from Israel, whose main topic was hydrotherapy, using water effectively for exercise to improve fitness. As a person is lighter in the water it is easier for most to move, and it is possible to increase strength while in the water due to more movement. Approaches vary from one on one sessions to group sessions, with the one on one sessions generally used post-surgery for conditioning and strength. Achieving and maintaining physical goals post-bleed, she believes, is possible through hydrotherapy, which is a much softer approach to building strength. Most children take part with their parents to help decrease the child's anxiety, if there is any. An added goal is for the child to not see this as treatment, but as something fun to do. An obstacle course is set up in the pool for adults to help with strength and balance, this saw balance outside of the pool improve by 25%. The ultimate intention she said was to improve life through fitness with the use of hydrotherapy.

"If there is no product then there are other ways to provide care, like teaching the patient about haemophilia through play, drawing, and drama."

Tim Ringgold is a music therapist working in Orange County, California, USA. Tim uses music for addiction, stress, and pain. His presentation was about using music for pain management. When music is playing the brain is busy processing the individual parts of the music, which floods the brain and puts it into overdrive. Pain, Tim says, does the exact same thing; it floods the brain. This is what he calls "heavy lifting for the brain". Everything entering the brain, like music or pain, is fighting for position, and the brain has to choose which one to focus on, so it is most likely to choose that which makes it feel good.

The important point raised was, that this requires active listening. Having music playing in the background occupies the background only, which means there is space for the brain to focus on something else. However, active listening occupies the brain and pushes pain into the background. Also, creating music, either on instruments, portable devices, or even with your mouth, can be just as powerful as listening.

In closing the speaker mentions that this type of treatment is complimentary not an alternative, so it is to be done alongside the pain management regime the patient is currently on, not in place of it.

For more information on music therapy for pain management visit the website: music@sonicdivinity.com

There was much more said, and by the sounds of things much more to come, but the ultimate message was to try and think outside of the square. Often, conventional methods don't work, and if they do work it may be only one aspect. All it takes is another view and someone willing to follow it through for the benefit of the patient's welfare.

Communication Issues and Skills

By Ashley Taylor-Fowlie

PRESENTERS:

Diane Standish – *Family and Communication Issues*

Silvina Grana – *Tools for Adolescence*

Anthony Roberts – *Communication with Health Providers*

Mariana Battazza Freire – *Experience from a mother's perspective*

This session focussed on the importance of identifying key communication needs in families who have a member with a bleeding disorder, discussed communication challenges that may become a more common occurrence, outlined some family therapy theories, and looked at some appropriate approaches to communication challenges.

During this session it was discussed that communication between parents/caregivers and children is essential so that children and other members of the family can understand more about bleeding disorders, and so that children are able to communicate to adults when they have injuries, pain, and/or bleeds.

Effective communication between adolescents and health practitioners (haemophilia treatment centres, nurses, doctors, emergency staff, and dentists) is essential in order to form strong therapeutic relationships between the client and their health professional.

Common challenges include children's understanding, sibling's understanding, denial (by a person with bleeding disorder, or family members) and the importance of working with children's teachers in order to minimise future issues.

Controversial Topics in the care of PWH

By Linda Dockrill

Complicated and clinical would be how I would describe this session. With eight different speakers presenting the pros and cons of four topics it required a great deal of concentration. Here is a summary of what we learned.

One team discussed the pros and cons of conservative treatment of large haemophilic pseudotumours. The pictures were a graphic reminder of how bad a pseudotumour can become. In Argentina a study of 40 patients used mini invasive techniques to deal with pseudotumours. 95% of the patients were cured using these techniques.

Another team looked at compartment syndrome. The text books write that compartment syndrome follows the laws of the seven P's – Pain at rest, Pain with passive stretch, Pressure, Parasthesia (tingling and numbness), Paresis (weakness), Pulse (diminished pulse in the affected limb), Pallor (loss of normal color). The pressure rises in the closed compartment and the tissues bulge. Conservative management comprises of replacement factor, limb elevation, frequent reassessment, analgesia, and a lot of patience!

According to Michael Heim from Israel, the most common cause of compartment syndrome in America is obesity. Nicolas Goddard from the UK stated that the problem with compartment syndrome is that 85% of bleeding in people with haemophilia (PWH) occurs in the musculoskeletal tissue so compartment syndrome presents as a significant challenge and can be confused with a muscle bleed. Patients often present late (>12 hrs) and normalization of the clotting cascade aims to lower the pressure in the compartment – if this fails a fasciotomy may be required. Nicolas showed that time is of the essence in compartment syndrome and that if a fasciotomy doesn't happen within 12 hours of diagnosis there can be irreversible neurological damage. However, he also added that in the majority of cases compartment syndrome settles with factor dosage.

> *"According to Michael Heim from Israel, the most common cause of compartment syndrome in America is obesity."*

The next topic was Osteoporosis in PWH. Gerry Dolan, Director of the Haemophilia and Thrombosis centre at St Thomas' Hospital, London told us that bone mass reaches a peak in many men in early adulthood. Genetics, exercise, nutrition, vitamin D and a balance of bone maintenance all impact on the development of osteoporosis. Like most of the room I thought osteoporosis was something little old ladies mainly suffered, but he was clear that overall 1 in 5 (20%) of men over the age of 50 will have an osteoporosis related fracture. This is greater than the likelihood of developing prostatic cancer. 20% of vertebral fractures occur in men and 30% of hip fractures occur in men. Sadly, research shows that men only have a 21% chance of living independently after osteoporotic hip fracture. However, age was not the only risk factor, and it appeared that arthropathy, low physical activity, HIV and HCV all increased the risk of developing osteoporosis. His treatment recommendations were mostly familiar – prophylaxis, active lifestyle, vit D and calcium, and bisphosphonates (medications).

Guinluigi Pasta from Italy presented his slides which agreed that studies show that fracture risk in PWH should be higher than in the general population. However, he pointed out that the risk of falling was a bigger issue than osteoporosis.

Dealing with Stigma for persons with bleeding disorders

By Linda Dockrill

This session was part of the Psychosocial Professional Development Day and was presented by Tony Roberts, member of the WFH Psychosocial committee, and a man with severe haemophilia.

Tony began by stating that many people face stigmatization due to their mental health, physical illness, race, colour, gender etc. He identified the process of stigmatization as:

1. Structural inequalities: Laws, religions, and/or institutions are constructed in ways that stigmatise certain groups
2. Stereotypes: Fixed and generalized ideas about groups of people
3. Prejudice: When stereotypes are believed to be true through attitudes and judgements
4. Discrimination: When people are denied equality and treated differently

He summarized this process by stating that "Stigma is stereotyping that leads to prejudice and discrimination".

Tony talked about two particular types of stigma: stigma by association, which is the stigmatisation of those connected to the person who is the victim by others; and affiliate stigma, which refers to the internal feelings of stigmatisation felt by those experiencing stigma by association.

Many people with a bleeding disorder experience affiliate stigma, because, as there are genetic aspects of the disorder, they feel that they may be overlooked for employment opportunities; they may not get selected for sports teams or be able to compete at a level they would like; they have the experience of bringing weakness or ill health to a relationship; and in some traditional communities the family is viewed as cursed or bewitched.

Dealing with stigma as a person with a bleeding disorder is possible in a variety of ways. Some may choose to hide their bleeding disorder from the world, while others may become activists against prejudice, and focus on teaching those around them about bleeding disorders. Self-empowerment is a key concept. Tony noted that attending foundation camps helps provide support to deal with internalized stigma, and programmes such as Parents Empowering Parents enable work on family and affiliate stigma to take place. Self-esteem and social support are key aspects of developing resilience and helping people with bleeding disorders deal with stigma.

Tony finished by saying he is proud to be who he is, he focuses on the positive and gets involved with working with other people with a bleeding disorder, PEP, haemophilia camps and sharing stories with others.

Empowerment through Self Care

By Ashley Taylor-Fowlie

PRESENTERS:
Patrick James Lynch

Patrick talked about his Journey with Haemophilia and how, spurred by the loss of his brother, he started his own journey of self-empowerment. Patrick's younger brother, Adam, died of an intracranial bleed at the age of 18. While trying to discover a reason for his brother passing Patrick found a bag of his brother's factor and suspects his brother stopped taking prophylaxis. "I spent a considerable amount of time thinking about why he fell off his regimen. I finally determined that it was because my brother never identified as having a bleeding disorder. That took him off his regimen."

Patrick began to think on how the community is able to engage and empower young members of the community.

Through the "unique application of my skills for the empowerment of the community" Patrick has created a serious of videos that strengthen and expand awareness around the bleeding disorder community. The videos are designed with humour to discuss serious topics such as self-care, self-infusion, women with bleeding disorders, and the history of haemophilia. Those wishing to watch the videos can at **www.stbhemo.com**

Excessive bleeding in women: Spotlight on Haemophilia Carriers

By Te Whainoa Te Wiata

Bleeding disorders in women has become an important and common theme in recent years. Dr Michelle Sholzberg, a haematologist from St. Michael's Hospital, Toronto, presented her findings and views on female carriers of the haemophilia gene. Joint damage in carriers was probably the most surprising finding, indicating unmonitored excessive internal bleeding into joints. Alongside that were the not so surprising facts like, heavy bleeding during menstrual cycles. The worrying part is many women are potentially experiencing this, but don't know any different and therefore aren't being tested or diagnosed, emphasising the need for awareness and education.

Many barriers are experienced by carriers that are not just physical, but mental and emotional. It has been noted that many mothers experience guilt in passing haemophilia on to their child, and as a result low self-esteem issues begin to surface. This highlights the need to provide those carriers of the haemophilia gene with on-going physical, mental, and emotional support, to allow for as normal a life as possible. Healthcare providers haven't been the best of help either. Research shows that women have been experiencing a massive amount of discomfort and worry and when they go to their doctors. They are being told:

"What you are experiencing is normal…"

This type of reaction has led to Dr. Sholzberg's approach, which is to empower female carriers through education and awareness, as well as increasing the availability of literature to enable care providers to make better judgements. Through this, an individual will be able to self-report, as well as have the ability to access on-going needed support, if it is needed.

As there are still areas that aren't clear, Dr.Sholzberg emphasises the need for further research to collect more data, hence the reason for the Canadian Hemophilia Carrier Study (CHIC study). This study will help draw attention to the fact that it is time to include women, and to broaden the approach towards comprehensive haemophilia care. In summary, Dr Sholzberg notes that:

- There is an increasing awareness that many haemophilia carriers experience abnormal bleeding
- Bleeding is likely to be under reported and underrepresented in haemophilia carriers
- Comprehensive support is needed

Me aro koe ki te hā o Hineahuone

You must pay heed to the dignity of women.
In closing, in any culture, ethnicity or belief, the world revolves around the female element, whether it is acknowledged or not. If it wasn't for women we wouldn't exist. These are results that have come out of addressing previously unaddressed issues, and the research that has taken place is only the beginning. 'treatment for all' is the WFH slogan, and 'all' also includes our women: our grandmothers, mothers, sisters and daughters. Therefore, it is time we acknowledged them for who and what they are, and look further into to this on-going dilemma.

Paimaarire ki a taatou.

Family caregiving and bleeding disorders: Helping parents cope

By Richard Chambers

" Bouncing Back ": How to develop resilience in your child with a bleeding disorder

SPEAKER:
Our very own Linda Dockrill

A highlight of the congress, Linda's presentation was powerful, showing links to the story of a real Kiwi family and a real Kiwi young man – Andrew Scott, who is 16 years old, and has Severe Haemophilia A with inhibitors.

For a child with a bleeding disorder resilience is vitally important. They may have to learn to contend with medical procedures, being poked and prodded with sharp instruments, how their parents deal with learning infusion, missing out on being in school sports teams, bleeds that see them on bed rest for a few days, crutches, wheelchairs, and being different to others. How parents and other significant people in a child's life respond to their fears and anxiety and the hurdles they encounter will set the tone.

> *"There is no single approach to building resilience. What works for one person might not work for another. Build confidence by focussing on the best in a child so that he or she can see those strengths as well."*

There is no single approach to building resilience. What works for one person might not work for another. Build confidence by focussing on the best in a child so that he or she can see those strengths as well.

Andrew's constant bleeds kept him from participating in sport on a regular basis, but he was creative and found he was a good referee of games he couldn't play. He is good at drama and public speaking. his parents signed him up for drama classes and haven't looked back. Recognising and supporting Andrew's strengths and abilities has enabled him to develop his competence, and confidence.

Learning from mistakes is part of growing up. Parents of a child with a bleeding disorder can find this hard, as mistakes can mean bleeds, but mistakes develop the understanding of choices and consequences.

Family connections are strong in Andrew's family. Family holidays are important, and extra strength is gained from the church and the bleeding disorder community. Andrew excels at leadership at HFNZ camps, a skill that has been role modelled by the service of his parents, Lynley and Richard, in HFNZ.

A child is born with their personality, but their character is developed by life experience. Help your child to view himself or herself as a caring person and encourage the development of spirituality, the importance of community, let them see their strengths. Andrew's family life is rich with character-growing experiences.

PWBD know what it is like to feel pain and to persevere – these experiences truly build character. Andrew has been in and out of a wheelchair, used crutches for weeks at times, and spent periods in hospital. Lynley and Richard been there through out and have consistently focused on helping Andrew see that the positives outweigh the negatives in life.

As the adults around a child with a bleeding disorder we need to model positive and effective coping strategies on a consistent basis. Developing a sense of control over life's challenges promotes resilience regardless of the outcome. Children who realize they can control the outcomes of their decisions are more likely to realize that they have the ability to bounce back.

Developing resilience is a personal journey with ups and downs. If your child seems stuck or overwhelmed you may want to consider talking to someone who can help, such as a school counsellor, mentor, or social worker. Your child may also turn to a trusted adult outside of the immediate family for guidance to strengthen their resilience and perseverance during times of stress or trauma and build on the skills developed with their parents.

Genetic Counselling: Understanding the role with patients and families

By Colleen McKay

CHAIR:

Edward Kuebler – *Social Worker, USA*

SPEAKERS:

Jennifer Lemon – *Genetic Counsellor, USA*
Michelle Alabek – *Genetic Counsellor, USA*
Richa Mohan – *Psychologist, India*
Regina Butler – *Haemophilia Nurse, USA*

Genetics is one of the most predictive tools in medicine. A genetic counsellor (GC) is a specifically trained healthcare professional with credentials in areas such as genetic testing, medical genetics, or developmental autonomy. The aim of genetic counselling is patient education for informed decision making.

The GC gathers both clinical and nonclinical information about the client. Family history, charted with standard pedigree forms, helps provide a picture for the counsellor to see the possible risks as well as the psychosocial issues. Using these standard forms allows them to be read across disciplines and providers.

After the actual testing, one of the most critical pieces of counselling is noticing how patients emotionally respond to the information. 'GC's are specially trained to recognise and address the potential psychosocial impact of the news you provide after testing is done' said Jennifer Lemon, noting that both verbal and nonverbal cues should be evaluated.

Michelle Alabek, Hemophilia Centre of Western Pennsylvania, acknowledged that GC's may not be available in all areas. 'Ideally the patient will be in contact with a genetic counsellor. However, it may be more realistic that other disciplines are providing genetic counselling at your site' she said.

She suggested the following for sites that do not have a genetic counsellor:

- Assess the skills of existing staff,
- Designate who will be responsible for each aspect of genetic counselling (disease education, risk assessment, testing, psychosocial, reproductive decision-making),
- Provide staff with specific education on genetics,
- Identify genetic counsellors to use for support.

'But when you have a genetic counsellor on your team, the other staff can concentrate on their own areas of expertise', she suggested.

Worldwide, genetic counselling resources may be limited, said Richa Mohan, India. To maximise the impact of what is available, she suggested seeking local, regional, national, and international resources.

Factors that drive genetic counselling needs worldwide include the severity of haemophilia, the incorrect perception that genetic

counselling is directive; healthcare providers in the country; marriage and consanguinity; sexual taboos; as well as religious and cultural beliefs.

Regina Butler, Children's Hospital of Philadelphia, described a programme developed for women held in her region. Topics included an overview of haemophilia in the 2000s, progress since earlier generations, case studies, the difference between genetics testing, and factor levels and resources available to those attending the programme.

Genetics counselling continues to grow in importance and should be part of the patient's comprehensive care plan.

"Genetic counselling provides information without bias; being non-directive is the gold standard. The first step is goal-setting process. Simply ask your patients why they are there"

Jennifer Lomas, University of Texas.

Haemophilia Treatment in 2030

By Richard Chambers

Professor Erick Berntorp challenged the attendees to think about where treatment will be in 2030, and what will have needed to happen before then. Inhibitors, and the fact that a majority of PWBD if diagnosed still do not get appropriate treatment, has prompted the development of long acting concentrates, by-pass therapy, and new treatment approaches such as gene therapy, modification of activation, and control of clotting systems. There is an increasing understanding of clotting factor immunogenicity, and a broad array of tools at our disposal. So what? What is next? We need biomarkers in order to predict and prevent arthropathy in an economically responsible way.

In 2030

- Prophylaxis will be the state of the art treatment for all PWH in wealthy countries
- Next generation products (Extended half-lives) introduced, improving convenience and adherence
- Gene therapy used in selected groups of patients
- Improved prevention and treatment of inhibitors.

However, to achieve this we need registry-based outcome assessment, we need better evaluations of what we are doing.

In high economic countries there will be small incremental improvements for PWH.

For low economic countries PWH will go from having no treatment to robust good health.

There will be much more individualisation of care. Professor Berntorp finished with a warning; never forget the tragedy of the 1980's (56% of treated Swedish PWH were infected with HIV). As a result of contaminated blood products 4000 of the estimated 10000 PWH in the US would eventually die of AIDS.

Hepatitis C Update: Complications and side-effects

By Richard Chambers

Unlike HIV and HBV, hepatitis C (HCV) is not a disease for life. Cure rates have risen from 10% to now over 95% since the virus was discovered in 1991.

Dilip Moonka, from Detroit in the US, took to the stage first in this morning. Looking forward, he said, drugs for HCV will be taken orally, have a cure rate of over 95%, will require less than 12 weeks of therapy and will be well tolerated.

Moonka said that in individuals exposed to the virus:

- 20% will clear the virus
- 75% to 85% will develop chronic infection
- 5% to 20% will develop cirrhosis, which typically it takes around 20 years to develop
- 5% to 20% of patients with cirrhosis will develop liver cancer,

"You have to continue to screen for liver cancer if HCV has been cured if you have cirrhosis," said Moonka. "Persons infected with HCV should refrain from excessive alcohol use and should be vaccinated against HAV and HBV if not immune."

In order to successfully treat the virus, the patient's genotype needs to be known. "The drug Harvoni is effective with genotype 1," he said, warning that this drug should be used with caution if the patient is taking amiodarone and should be avoided if creatinine clearance is less than 30 ml/minute. "The drug AbbVie [offers] is also very well tolerated, even though more pills are taken. Zepatier can be given to those with renal failure and we will still see cure rates close to 100%." Drugs are also available for other genotypes that elicit a cure rate of more than 95%.

Magdy El-Ekiaby, from Egypt, told the audience that Egypt is one of the countries in the world with the highest rate of infections. The HCV burden is great, with the 15 to 59-year age group estimated to be 7% infected. "This is an endemic infection with social, economic and political implications," he said. "On-going transmission is still occurring with up to 200,000 new patients each year."

Until 2006 Egypt did not have a comprehensive national program for control of HCV. At that time the objectives became to track prevalence, implement infection control, expand access to treatment, and ensure high quality scientific research. In the 2011 to 2014 action plan, new drugs were launched and 400,000 patients were treated. "Government scaled up treatment centres nationwide, and they should reach 100 by the end of 2016," said El-Ekiaby. "Treatment guidelines are updated regularly and there is fast track registration of all approved new drugs with special pricing." The government pays for 83% of costs, health insurance pays 9% and 8% is out of pocket.

Real-life results show that therapies have close to a 95% cure rate. "To achieve elimination, we need to increase annually the number of treated patients. Without significantly changing treatment strategies, HCV will remain a highly prevalent problem for the next 20 to 30 years," he said.

Managing Patient Expectations of Extended Half-life (EHL) Products

By Colleen McKay

David Page, a person with Factor IX deficiency and from the Canadian Haemophilia Foundation gave an interesting Presentation highlighting the outcomes that Haemophilia patients and treaters are looking for when considering Extended Half-Life (EHL) Treatment Products.

He explained that patients are looking for increased protection from bleeding, from both major haemarthroses and micro bleeds; less long-term joint damage; an increased ability to lead a full, active life; and fewer infusions, especially in the case of small children.

Meanwhile, treaters and funders are interested in improved health outcomes at equal or lower cost; and improved quality of life as measured by fewer infusions, sometimes called convenience, but not at a cost, even in small children.

Unfortunately, funders may not pay for longer half-life products if the only proven benefit is fewer infusions.

There are a number of factors that influence outcomes, including:

- Access to Factor concentrates
- Support from family and friends
- Knowledge of haemophilia
- Comprehensive care, including medical, nursing, physiotherapy, and psycho-social
- Adherence to a prophylaxis regimen
- The right dose at the right time
- Fitness and wise selection of activities

EXTENDED HALF-LIFE (EHL) CONCENTRATES VS STANDARD CONCENTRATES	
Efficacy to stop bleeding at similar dosage	**Apparently identical.**
Risk of causing inhibitors	**No data to compare EHL products to standard or plasma-derived products.**
Half-life	**Factor VIII: Up to 1.5 times longer Factor IX: 2.5 to 5 times longer**
Cost	**Higher per IU for EHL products**

POTENTIAL BENEFITS VS RISKS / COSTS OF EHL PRODUCTS	
BENEFITS	**RISKS / COSTS**
Opportunity to reduce frequency of infusions (increased quality of life, potential to increase adherence to prophylaxis)	**Fewer peaks, longer time at trough level near 1%**
Opportunity to maintain frequency and dosage and increase trough level (1% is not always enough)	**Higher cost to Funders / Payers**

In conclusion, EHL Factor products have the potential to decrease the number of infusions and increase quality of life, especially in small children. They can also increase the likelihood of adherence to prophylaxis, reduce long-term joint damage, and increase protection from bleeding by raising trough levels, if they're priced competitively and readily accessible! This last benefit, however, can also be achieved with standard half-life products.

I'll take my Bleeding Disorder rare

By Colleen McKay

CHAIR:
Jim Munn - *Haemophilia Nurse Specialist USA.*

SPEAKERS:
Amy Shapiro - *Haematologist USA*
Richa Mohan - *Psychologist India*
Dilli Adhikari *with Factor X deficiency from Nepal*
Evelyn Grimberg *with Glanzmann's from the Netherlands*

Both Dilli and Evelyn provided a patient perspective of life with a rare bleeding disorder.

Haematologist, Amy Shapiro, explained that even defining what should be considered a rare bleeding disorder is difficult and varies from country to country. In the United States a bleeding disorder is considered rare if it affects <1 in 200,000 people whereas in Europe a bleeding disorder is considered rare if it affects <1 in 2000 people. For a range of reasons, the distribution data underestimates the total worldwide affected population. In most countries there is a lack of a systematic national surveillance system. Often there is a lack of recognition or knowledge among healthcare providers, and distribution data underestimates the total worldwide population of rare bleeding disorders. In most countries there is a lack of a systematic national surveillance system. There is often also a lack of recognition or knowledge among healthcare providers, and there may be inadequate access to laboratories with the necessary diagnostic capabilities. There may also be socio-economic barriers and stigma associated the diagnosis of a genetic disorder – this stigma may be based on cultural issues.

The diagnosis of rare Bleeding Disorders can be difficult. It can take many months, or sometimes even years, for a definitive diagnosis, and many factors can contribute to this. There may be inadequate access to laboratories with the required diagnostic capabilities. There is often a lack of knowledge among healthcare providers about the different treatment modalities - at what level bleeding occurs, at what level replacement suppresses bleeding, which patients should receive prophylactic treatment, and at what age to start. There can often be issues related to poorly controlled bleeding episodes, and there can be an inability to fully participate in normal life and work activities. It is important that those with rare bleeding disorders are referred to a Haemophilia Treatment Centre for symptom recognition, an established diagnosis, for determining and access to best therapy in order to manage the full spectrum of the disorder.

Richa Mohan, psychologist went on to explain the psychosocial influences for these patients. It can be a time of great uncertainty, and especially if there is a difficult and delayed diagnosis. During this time, individuals often feel isolated and struggle to obtain adequate information. In some cultures, there can be a great degree of stigma, discrimination, blame, and the associated guilt. Bleeding episodes may cause difficulty in participating in activities, education and employment. There is often fear about what will happen in the future and the ever present feeling of carrying a 'burden'. Richa Mohan went on to outline some Strategies to deal with these issues:

- Knowledge is power, seek more information about the disorder. It helps to understand the situation and deal with it more effectively
- Join or form a support group and take action
- Be economically independent, this gives confidence in dealing with own situation
- Eat well
- Adjust to the 'new normal'
- Practice yoga and/or meditation, and work to develop resilience
- Psychological counselling to help with these issues.

Dilli Adhikari, an amazing woman with Factor X deficiency, described her life in Nepal, living in a remote village and experiencing bleeds from a young age. Her parents took her to neighbouring parts of India for treatment. Dilli was home schooled initially but eventually did attend school. However, she missed a lot of classes due to bleeding episodes. After migrating to Kathmandu, Dilli completed a Bachelor's and Master's degree. Although she finds securing employment difficult – Dilli stresses the importance of a good education.

Another amazing young woman from Germany, Evelyn Grimberg, with Glanzmans Thrombastenia, took us on a journey through her childhood and teenage years. Hers was a journey of sustained bleeding, bruising easily, bleeding after surgery, nosebleeds, and heavy menstrual bleeding. Her treatment includes Tranexamic Acid, Platelet transfusions, recombinant Factor VIIa, and the contraceptive pill, together with a strong relationship with the comprehensive care team at her Haemophilia Treatment Centre. Evelyn spoke about the future, dealing with the insecurities, sharing her story to get attention for rare bleeding disorders, and to get more attention for women with bleeding disorders. Evelyn is an active volunteer at her national member organisation in order to achieve these things and to support other young women in a similar situation.

Those with rare bleeding disorders can feel isolated, and often it is difficult to find support from those with similar experiences within the bleeding disorder community, because many of these disorders are so rare. There is psychological need which needs to be addressed; it is important that support is provided from those at the Haemophilia Treatment Centre by both clinicians and psychosocial health professionals. It is also important that National Member Organisations include those with rare bleeding disorders in their Programmes where appropriate.

Rare Bleeding Disorder EXPERTS - Dilli and Evelyn have the following suggestions:

- Avoid cuts and injuries as far as possible.
- Build a strong relationship with the comprehensive care team at your Haemophilia Treatment Centre; and be an active member of the Team.
- Join your local Haemophilia Society for support, and work to make a strong national Haemophilia Society which will help in accessing better treatment and rehabilitation.
- Try to get a good education, it will help to develop a better career for the future.

They both say …. **Never give up, go for the gold!!**

Transition Challenges for Young Adults

By Colleen McKay

SPEAKERS:

Dawn von Muyhauser - *Social Worker USA*

Colleen McKay - *Education & Programme Manager, New Zealand*

David Silva Gomez - *Board of Directors Member, World Federation of Haemophilia*

Harshal Kale - *Youth Chairman, Haemophilia Foundation of India*

Clinical Social Worker Dawn von Muyrhauser, from Connecticut in the USA, noted that, in the last 20 years, researchers have begun to look at the decade from ages 18 to 28 years as a distinct developmental stage characterised by a number of milestones. This is the decade when young people are attempting to take responsibility for themselves and make independent decisions. Many are leaving home for the first time, exploring educational and employment opportunities, and hoping to find a mature, loving relationship. The stage of forming one's own identity can be characterised by exploration and instability.

Haemophilia and other bleeding disorders are complicating factors that may make this stage more difficult. In addition to navigating the milestones characteristic to this age, emerging adults with bleeding disorders must transition from paediatric to adult care, requiring that they demonstrate the ability to manage their bleeding disorder independently. They must make decisions about disclosing their diagnosis to potential employers and partners. They might also have to cope with the feelings of their parents who may have anxiety about allowing their children to be responsible for their decisions. In addition, emerging adults with bleeding disorders often must deal with the physical implications of their disorder including arthropathy, physical disability, and pain.

Emerging adults may need help in overcoming these challenges, so it is important that there is a range of support available for successful transition, including:

- Emotional support providing reassurance
- Tangible support by way of direct aid and services
- Informational support encouraging independent problem-solving skills.

It is important that the Haemophilia Treatment Centre starts transition early, and that they provide support to both the patient and their parents, acknowledging the difficulties. Information should be provided in a user-friendly manner. Psycho-social health professionals should watch for mental health issues and ensure effective executive functioning.

Parents of emerging adults should acknowledge their own anxiety and fears, and realise that their emerging adult still needs and wants support. It is a delicate balance to step back, while staying connected. It is important to know that your child still loves you, even though they don't answer your text fast enough. Emerging adulthood is a difficult, but exciting stage, full of possibilities. It is important for emerging adults to embrace the change and understand that they have the strength to cope with whatever comes along, as well as working to build their support system of family, friends, and health professionals, and committing to managing their own bleeding disorder care independently.

Colleen McKay, Manager of Outreach Services for the Haemophilia Foundation of New Zealand, further elaborated on the transition with particular reference to Education and Career Choices. Treatment for bleeding disorders in the developed world has improved immeasurably in recent decades, beginning in the 1960s with the introduction of factor replacement and followed by more refined treatment products. The availability of prophylactic treatment has increased the possibility of a 'normal' life for people with bleeding disorders. Home therapy has also meant that most people with bleeding disorders are generally independent of the hospital. Despite these major improvements in treatment, research has shown that those with bleeding disorders participated less in full-time paid work than the general population.

The presence of a bleeding disorder adds an extra challenge when choosing a career pathway. The right employment offers more than just money. A successful job or career also contributes to a sense of self-worth. Being able to support oneself, and the self-esteem that goes with that, can help increase feelings of true independence. People must still take into account the impact of their bleeding disorder when choosing a course of study and employment. It is also essential that they carefully consider their interests, values, education, strengths, job satisfaction, job security, as well as the physical demands of the job.

With career planning, it is extremely important to begin early and to think long term. The development of a proactive and effective job search strategy begins with the identification of a career direction, rather than a job. Once a career direction is established, this gives a clear indication of the next step – the education and training that will be necessary to achieve the career goal. When applying for jobs it is important to have a strong curriculum vitae and to practice effective interview skills. However, research suggests that only 25% of jobs are found in this way. To get the job you want there is greater value in networking, developing professional connections, volunteering, and mentoring. Leadership and Training opportunities provide great transferable skills, and these may be available through National Member Organisations (NMOs).

David Silva Gomez, a person with haemophilia and a Lay Member of WFH Board of Directors, discussed Global Perspective for Challenges on Young Adults. Living with a chronic disorder shapes the challenges for young adults; it is a complex process. For any young adult here is new status, new roles, and a new self-image to consider. The process is made even more difficult in the presence of a chronic disease. Psychosocial resources are crucial.

Globally the transition to adulthood varies and is impacted by four key elements:

1. Access to comprehensive care: There are differences in access to comprehensive care between developed and developing countries. Comprehensive treatment is more than the provision of Factor; it also requires psychosocial support from professionals. The quality-of-life perception is important.
2. Family & Community Support: The way in which the young adult has been raised is crucial at this point, especially how the bleeding disorder has been managed at home. Community support from NMOs gives the young adult a sense of belonging and connection.
   Peer support is important.
3. The Psychological Resources of the Young Adult: Current level of maturity is crucial, as well as personal and vocational maturity. Psychosocial resources take time to develop, and professional support is very helpful.
4. Social & Cultural Environment: Adulthood doesn't come at the same time in every culture. Local and cultural customs and beliefs, as well as personal relationships, habits, and way of life all have influence on the transition to adulthood.

This transition period is growing in importance in modern society. It is crucial to remember that factor replacement therapy is important, but it is not everything. NMOs have an important role in supporting youth with bleeding disorders as they transition into young adulthood.

Harshal Kale, Youth Chairman of the Haemophilia Federation of India gave his personal experience of transitioning to become a young adult. Harshal was diagnosed with haemophilia as a child and has spent more than half of his life with no factor replacement therapy. Throughout his life Harshal has had a number of life threatening situations. Yet, despite the difficulties of growing up in rural India, this has not stopped Harshal from taking responsibility for himself, from completing a degree in Computer Engineering, or from finding suitable employment. At the same time Harshal has also been an active youth leader in his local haemophilia community. He urges the need for a dedicated global platform for youth.

Susan Skinner Memorial Fund Scholarship & Global NMO Training

By Ashley Taylor-Fowlie

I was very fortunate to receive a 2016 Susan Skinner Memorial Fund Scholarship along with Claudia Pena Villena from Peru. We were joined at NMO training and Congress by the 2015 recipients Michelle Cecil from Michigan and Joelle Palmatier from Alaska.

The Susan Skinner Memorial Fund Scholarship supports training, education and leadership development of young women with a bleeding disorder. Thanks to the Susan Skinner family and WFH I was provided with this amazing opportunity to attend WFH Congress and Global NMO training.

During the Global NMO Training NHF were holding their national AGM. On arrival NHF hosted a youth gathering, where I was able to meet other youth 18-30 years from around the world.

On Thursday the first official day of training began with Plenary One: Opening and sharing of NMO experiences. During this Plenary I presented on New Zealand's Youth Leadership Programmes. The afternoon followed with Colleen McKay's workshop on Outreach. During the evening I was invited to attend NHF's opening ceremony of their annual meeting and had a chance to look through their exhibit hall.

Friday started off with Strategic Planning and Implementation of Projects workshop. At lunch I attended WFH Youth Meeting, followed by plenary two on Haemophilia Treatment.

Saturday consisted of congress registration, a workshop on volunteer development followed by a closing event at Universal City Walk.

I was also invited to attend the Women's Bleeding Disorders Coalition lunch meeting with My Girls Blood. A lunch for women with bleeding disorders worldwide. They ran a women's booth at Congress, and I also received an open invitation to join their committee.

Perceived Wellbeing: Social, Emotional and Psychological

By Te Whainoa Te Wiata

The multi-disciplinary sessions held at WFH Congress are sessions where a number of speakers from different areas of haemophilia care present their research. One particular session I managed to get to had quite amazing speakers, Luca Negri PhD, of the University of Milan spoke on social, emotional, and psychological wellbeing in haemophilia.

Across the board, people with haemophilia (PWH) aren't doing

well physically, and when tested on physical health, scores are often poor for the majority. However, a study was carried out to score not only physical health, but mental health too. One of Negris opening statements was:

"Mental health is the absence of mental illness"

Which was quite possibly one of the simplest and yet coolest sayings I heard throughout the entire Congress. In short, PWH generally score lower on physical wellbeing, and, as a result, show low self-esteem due to the outcomes of joint damage and other things physically associated with haemophilia. As a whole, general health is low when it comes to PWH. On the other hand, research showed that emotional and psychological scores were higher than those physical scores, and, although the social scores were low, resilience and over all positive outlooks on life in general were high. This produced a desire to sit down with PWH to develop care programs on how to deal with chronic illness, which is currently ongoing.

The other sessions and speakers were amazing, but this session was one that caught my attention. Being a PWH I am completely aware of how strong and resilient our whānau are, especially our ageing men. They are still able to smile even with all of the trials they have faced. Many of us know these things, but it was nice to see a study done on it, and actually see research to back up what we have known for a long time. Negri, in closing, did urge caution to those listening, saying that this is no reason to assume that all PWH are mentally okay, it is still an area that needs just as much attention as any other aspect of a patient.

Ngā mihi

Pacific Asia Meeting

By Richard Chambers

There are changes coming to how WFH provides regional support across the globe.

The Asia Pacific region the largest in WFH with the greatest number of people with bleeding disorders yet to be identified. WFH will be appointing two people to work in the Asia Pacific region – based in the region, previously all WFH staff were based in Montreal.

Our region contains a number of countries where identification of people with bleeding disorders is problematic or non-existent. Remember, the region contains the huge populations of India and China. As yet we don't know where the WFH staff will be based, but we do know that a previously under-resourced region will now be getting more focus and support

Surgical infections and joint replacements

By Linda Dockrill

This session looked at the risk factors leading to infection in patients with haemophilia undergoing arthroplasty, the importance of dental care to lower the risk of infection, coatings to use that reduce infection risk, and surgical strategies for the treatment of an infected arthroplasty.

It was surprising to learn the huge impact oral hygiene could have on the development of post-surgical infection in a joint replacement. Patients are encouraged to maintain meticulous oral hygiene before and after joint surgery. Pre-surgical assessment was recommended to identify patients at high risk of oral disease, and any dental extractions need to be done at least one month before surgery. The take home message was "forget the teeth… don't rely on or depend on the antibiotic prophylaxis… it's all about the health of the gums".

Nick Bernthal from the US spoke about a number of antibiotic coatings that are available for use during arthroplasty. These included, vancomycin powder, iodine coated implants, nanosilver coated implants, and antibiotic impregnated cement. Nick recommended that for prevention of infection in the haemophilia population there should be pre-operative antibiotics, improved factor management pre op, and an aseptic technique used.

Finally, Adolfo Llinas from Colombia shared a case study relating to surgical strategies for an infected arthroplasty. The case study was of a 36 yr old man who had had a double total knee replacement several years ago and now had one painful knee. The patient had severe haemophilia B, Type 2 diabetes and active hepatitis C. After blood and culture tests confirmed an infection, and antibiotics had not made any impact, they decided to remove, debride, implant a spacer, and replace the implant. There were a lot of very graphic pictures of this procedure and the outcome was reported as positive for the patient.

> *"The take home message was forget the teeth… don't rely on or depend on the antibiotic prophylaxis… it's all about the health of the gums"*

vWF & FVIII: Working together…

By Colleen McKay

The blood's ability to clot is a complex process involving platelets and proteins called clotting factors. Von Willebrand Factor (vWF) is one of those clotting factors.

vWF: The glue and the chaperone:

vWF is made in the walls of blood vessels and released into the blood. When a blood vessel is injured, vWF promotes adhesion and aggregation of platelets. In other words, vWF acts like the glue to help the platelets stick together and form a blood clot.

In addition to acting like glue, vWF also acts like a 'chaperone' or 'carrier' for Factor VIII (FVIII) in plasma. As it's chaperone, vWF performs three important jobs for FVIII:

- The Transporter… vWF carries FVIII safely to the bleeding site where clotting is needed.
- The Bodyguard… vWF may protect FVIII from agents in the blood that try to destroy it, such as inhibitors.
- The Breakdown Blocker… vWF slows the breakdown of FVIII so that it stays in the blood longer.

vWF and FVIII are good travelling companions. In the body, vWF and FVIII travel together.

Nurses Issues

By Linda Dockrill

This debating session featured nurses from Brazil, India, Sweden, and New Zealand (go BJ Ramsay!) Sometimes a nurse was required to argue the opposite from their personal viewpoint, and there were quite a few laughs generated. The first debate was about whether ports were superior to peripheral venous access for children beginning home infusion. In India ports are expensive, there are no specialized nurses, and the patient bears the cost of treatment. This means that learning venous access is easier and safer in that country. In Sweden venous access is the gold standard and parents and children learn venous access from the beginning. Parents are aware and motivated to learn. This nurse thought that ports were mostly suitable for those with inhibitors. BJ argued that ports that are well looked after last a long time and have a low infection rate.

Another debate topic was Short term prophylaxis provides no benefit. Linda Myrin, the nurse from Sweden, argued that the impact on previously untreated adults was nominal, and that all patients should be offered on demand treatment. The nurses from developing countries both argued that quality of life is improved when any treatment is given. B J added to the debate by saying tertiary prophylaxis is like duct tape – high cost treatment but won't repair it for long! He argued that the damage

needs to be stopped early, and we should use money on people when they are younger to prevent damage occurring.

Women with haemophilia – myth or reality?

Sulochana Badagabettu from India argued that you can't label every woman who is a carrier as having haemophilia as in India this causes a lot of stigmatization for that woman and her family. Cost and social acceptance are issues that are heavy to carry and Sulochana argued that we shouldn't diagnose women as having haemophilia as it is a burden for them. Linda Myrin from Sweden argued that women with low levels should be diagnosed as having haemophilia and supported the idea that those with factor levels under 40% should be diagnosed as having mild haemophilia. This received a loud round of applause from the audience.

BJ joined the debate with his memories of the Princess Bride movie, to much hilarity. BJ argued that the word carrier is not used well and that we should "diagnose people rather than label people". He noted that the WFH definition of Haemophilia is having levels lower than 40%. As a further point BJ added that increasing the number of those who could be considered to have a bleeding disorder promoted job security for clinical staff – yes that got applause as well!

The final topic for debate was that neonatal circumcision in boys with haemophilia is child abuse. Elaine Sandoval from Brazil said that with prophylaxis there should be no issue and we should respect other religions. In India circumcision under cover is done

> *"... the word carrier is not used well and that we should "diagnose people rather than label people". He noted that the WFH definition of Haemophilia is having levels lower than 40%. As a further point BJ added that increasing the number of those who could be considered to have a bleeding disorder promoted job security for clinical staff..."*

and the nurse did not consider it child abuse. However, Linda from Sweden argued that the benefits were not sufficient and that, despite the cultural and religious aspects, performing an unnecessary medical procedure on someone with a bleeding disorder was unethical, particularly if that was a child who does not have a voice. She argued that tax money should not be used on this operation. BJ Ramsay had the final say on this topic which he said was "worth discussing" with patients. He argued that 60% of American men are circumcised and that it is the most common surgical procedure in the world. He further added that not fitting in could be considered child abuse – a comment that brought another noisy response from the crowd.

Working with a bleeding disorder

By Richard Chambers

Four very capable speakers Pinsiri Godamunna (President Haemophilia Association of Sri Lanka), Gomes Cavallini (Argentina), Baiba Ziemele (president, Latvia Haemophilia Society), and Shelly Reed a President of the Wisconsin Bleeding Disorders Network). Each speaker brought with them a different perspective on life within a bleeding disorder community.

In each of the four countries, the challenges being faced are different, but do have some similarities. Sri Lanka, Argentina, and Latvia are all working on developing registries of people with bleeding disorders. The employment opportunities for people with severe bleeding are key, when do you disclose your condition, or do you choose not to? In Latvia there is the additional challenge that working will actually mean you could be living on less money, as jobs pay poorly, and state support,

> *"All speakers agreed that there are challenges and limitations in living with a bleeding disorder, and the impact of those challenges varies from country to country."*

which may be higher and more consistent than a wage, ceases if you are well enough to work. Often people with bleeding disorders will undertake volunteer work instead of paid work.

All speakers agreed that there are challenges and limitations in living with a bleeding disorder, and the impact of those challenges varies from country to country. You have to find your own balance, and adjust your goals around your interests, limitations, and how your society works. People want the same thing; to have positive relationships, to develop skills and a purpose, and to be independent.

AGM: 2016

The HFNZ Annual General meeting is a chance for our members to get together and actively participate in the running of the Foundation. The 2016 AGM was held in Christchurch at the International Antarctic Centre. There was a very good turnout for the meeting, and the MRG training that preceded it. It was an opportunity to recognise the achievements of members, supporters, and staff. ***Phil Constable*** *was there…*

This year's HFNZ Annual General Meeting was held at the International Antarctic Centre in Christchurch on September 17th. We had a great turnout for what was a full day.

The day began with a morning of training for committee members of the various representative groups. CEO Richard Chambers led a varied and interesting programme that included small group sessions for Chairs, secretaries, and treasurers, and a communication workshop presented by Colleen McKay.

In the afternoon the AGM proper took place. As well as the routine matters dealt with at every AGM, we were also able to honour those that had passed during the previous year by having Katrina and Tim Miller light the memorial candle. Katrina's son, and Tim's brother, Jeremy, became HFNZ's HIV ambassador after contracting the virus from tainted blood products in the 80s. This year marked 21 years since his untimely passing at the age of just 15.

This AGM saw some changes in the makeup of the National Council. Each year we vote on who are going to be the office holders on Council, and recently that has meant confirming the existing members for another term. This year Grant Hook stood down from his role as Treasurer. After being on Council in various capacities, for several years, we were pleased to be able to recognise Grant's great work for the Foundation, and wish him well for the future.

In Grant's place, Hemi Waretini was elected to be Treasurer, joining President Deon York, and Vice- Presidents Catriona Gordon and Richard Scott on the executive committee.

We have some new regional delegates on National Council this year too. Courtney Stevens comes in as the Youth Delegate, following the end of Hemi's term, and Theresa Stevens replaces James Poff at Southern. James spent several years as a Council Delegate, as well as being a Vice-President and Treasurer. Though he was not at the AGM, we recognised all his good work for HFNZ's members.

We were also privileged to have former Northern Outreach Worker, Sarah Elliott, in as a guest speaker. Sarah spoke about her work with older men with haemophilia as part of her Master's Thesis. This was very interesting, and offered a number of insights that will be very useful to HFNZ as we move forward.

A significant part of this year's AGM was recognising hardworking members, departing staff, and those who had gone above and beyond to help the Foundation during the year. There were a number of people recognised this year, and some awesome work done for HFNZ. They ranged from our volunteer of the year, through to the best regional activity, and included acknowledgement of long-service and of staff who are moving on.

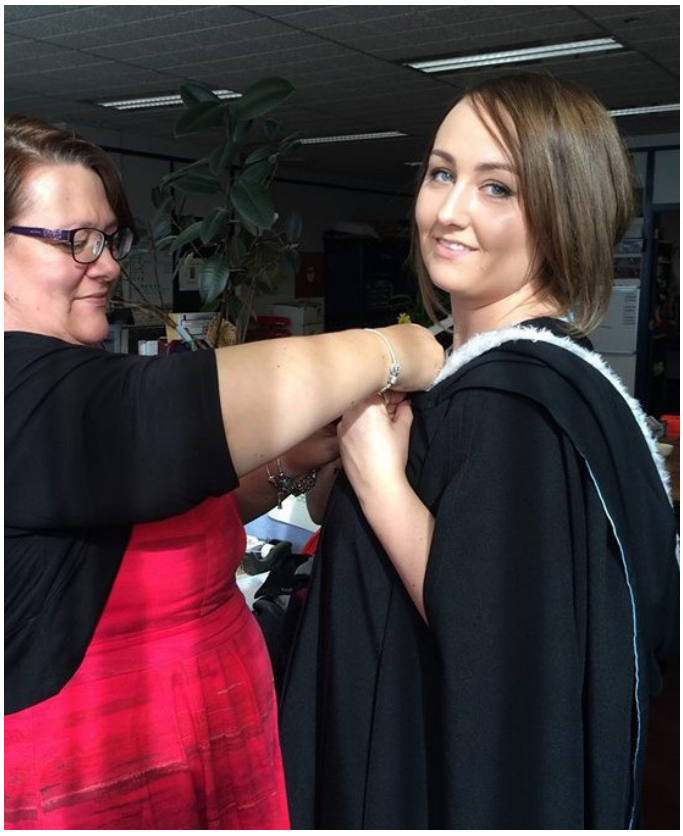
- The HFNZ Volunteer of the Year: Lyn Steele for over 30 years of voluntary service to HFNZ
- The McKay Trophy: The Southern Region for their sterling work on their Play Night, which raised over $1800 for HFNZ
- The Elizabeth Berry Exercise Cup: Riley Barnes for his commitment to exercise and his inspiration of other HFNZ members
- The Sir John Staveley Health Service Professional Award: Julia Phillips for her years of ongoing support and advocacy for HFNZ members in a health setting.
- Certificates of appreciation for long-service: Colleen McKay, 20 years and still going strong, and Chantal Lauzon, former National Information Coordinator, who moved on after 10 years of service to HFNZ.
- Departing staff: Linda Dockrill, Southern Outreach Worker, after 7 years working for the people of the Southern Region, and across the country at camps and workshops.

It was fantastic to be able to acknowledge the contribution of so many dedicated members, staff, and supporters this year. Without people like them we would not be the strong Foundation we are today.

Over-all AGM 2016 was a great success, and planning is underway for 2017…

Look at what ACEET can do for you

The Alan Coster Educational Endowment Trust (ACEET) is one of those organisations that you don't hear too much about, but they make a big difference, even change lives.



Alan Coster was an HFNZ member, and for 19 years was the acting senior dental surgeon and haemophilia dentist in Auckland. During the 1970s and 80s he worked extensively with the team at the Haemophilia Centre to upgrade standards of dental services for people with haemophilia. Upon his death he left a substantial bequest to the existing haemophilia endowment fund, and it was renamed in his honour.

Today ACEET offers grants to help people with haemophilia and other rare bleeding disorders to access vocational training and education. Over the years ACEET has helped many people to change their lives, providing them with the financial support for education that they may not otherwise have been able to access.

The ACEET grants are not just for university study. The ACEET trustees are open to applications for primary, secondary, undergraduate, postgraduate, trades, or vocational training. That means, if you have a bleeding disorder, a dream, and a course of study that will help you to get where you want to be, then an ACEET grant could be just what you need.

Previous recipients have used their grant to complete courses from reading assistance, to business management, to graphic design. If it is going to improve your prospects, then it's worth getting an application in.

One such recipient is Lauren Nyhan. Lauren is the current Chair of HFNZ's National Youth Committee, and has just completed her law degree. She is a smart, committed young woman, and a big part of the future of the Foundation.

Here's what she has to say about her ACEET Grant:

> *Having a huge student loan was always going to be a reality for me; but I knew that the benefits outweighed this temporary setback. I had looked at various types of funding and scholarships to assist me throughout my study but never quite met the right criteria. I had heard of the Alan Coster Education Endowment Trust (ACEET) before, but it was my Outreach Worker who encouraged me to apply.*
>
> *Studying law meant expensive books, expensive course fees and, being in Wellington, expensive coffees! ACCET couldn't help me with the last one but it did assist me with course materials - which when you're paying $160 for a 2 cm thick book is very handy! I was successful in obtaining a grant thought the Trust to assist me but perhaps more important than the financial assistance, it felt like a vote of confidence from the trustees. They believed in the work that I was doing and wanted to support that.*
>
> *I graduated from Victoria University of Wellington Law School earlier this year and have recently completed my professional studies as a prerequisite of bar admission. I plan on being admitted to the Bar in March next year. I'm currently working in the Environment Court as a Hearing Manager where I get to travel the country with Judges and ultimately plan to work in resource management litigation. I hope that my legal skills can also be used to benefit the foundation going in to the future.*
>
> *My one piece of advice to anyone studying and thinking of applying for ACEET is DO IT! Compared to other lengthy and convoluted scholarship processes the ACEET process is much more straightforward; and getting that vote of confidence from people in your community is a great feeling.*

Needless to say, Lauren wouldn't have been as successful as she has been if she didn't work hard, and have a good attitude. However, reducing some of the worry that comes with trying to find the right texts at the right price certainly helps.

The ACEET trustees, Elizabeth Berry, Mike Carnahan, and Mike Mapperson, are all Life Members of HFNZ. Elizabeth is the current patron of the Foundation, and both Mikes are former Presidents. They all have the best interests of HFNZ and its members at the centre of their decision making. They consider grant applications three times a year. To check the next application deadline, or apply for a grant, just contact your Outreach Worker.

Bloodline Report

2016 National Family Camp

By Colleen McKay

The final big event of 2016 was the HFNZ National Family Camp. Once again held in Rotorua, this time the weather didn't quite come to the party. Nevertheless, we had a great group who learned a lot and had a fantastic time. With a variety of educational sessions, as well as some great recreational activities, this was another valuable opportunity for our members. ***Colleen McKay*** *reports on this annual favourite.*

On September 30th 98 Campers assembled at Keswick Camp in Rotorua for the 2016 HFNZ National Family Camp. This was another fantastic weekend, even though the weather didn't quite play ball. The kids had a great time fishing, bike riding, and other fun stuff. Meanwhile, the adults learned heaps about bleeding disorders; including Haemophilia A & B, von Willebrand Disorder as well as those with Platelet Function Disorders; and the issues faced by people with bleeding disorders.

There were a number of really valuable workshops

In one session especially for those with newly diagnosed bleeding disorders in the family, Haemophilia Nurse, BJ Ramsay, covered the basics of bleeding disorders. Meanwhile Outreach Worker, Linda Dockrill taught parents about the 7 C's, the building blocks to developing resilience in children – Competence, Confidence, Connection, Character, Contribution, Coping, and Communication.

Haemophilia Nurse Maureen Hayes from Waikato ran a session for those with von Willebrand Disorder and other rare bleeding and platelet function disorders, such as Glanzmann's Thrombocytopenia.

It was great to have Dr Nyree Cole drive from Auckland to speak to parents on some of the complications of haemophilia, like venous access and inhibitors, and to promote a realistic view of the possibilities of future treatments.

A welcome addition to this year's National Family Camp was having a Haemophilia Physiotherapist in residence. Cat Pollard from Auckland, not only presented an education session about Physiotherapy, but also took the time to offer individual consultations for those present.

Outreach Worker, Linda Dockrill's presentation on Constructive Communication gave parents a set of skills for the establishment and maintenance of open communication with their children - skills that will help cope with children's negative feelings, to engage child's willing cooperation, to discipline without hurting or alienating, to develop a positive and realistic self-image, which will foster a family atmosphere of love and respect.

Colleen McKay's presentation, Building Responsible Kids, likened the process of building responsible children to building a house – important to start early and build a strong foundation, upon which the children can then frame or build the house of responsibility. To ensure that the house stays strong, it is essential that parents role-model responsible behaviour themselves.

As always, one of the favourite sessions was 'Ask the Experts', where parents were able to ask young adults about their experiences of growing up with a bleeding disorder.

The Treatment Room was busy each morning with children and parents alike perfecting the art of self-infusion and other treatments. Big ups to Alex Rutherford and Izack Silva for doing their own treatment for the first time, with many other Campers making great strides in their self-management.

Nice work you kids!

While the grownups were doing learning, the kids were divided into three age-based groups and were kept busy with interesting and fun activities:

- In the Crèche the Speedy Construction Workers were kept busy with painting, play dough, and a huge array of toys from the Rotorua Toy Library.

- The 4 – 7 year olds, aka the Pink & Blue Wrecking Crew, and the 8–12 year olds, akathe Architects, undertook a range of activities like rifle shooting, confidence course, as well as woodworking where the children made their own car / truck / construction vehicle.

A highlight for many of the kids was an afternoon out fishing. The day was organised voluntarily by Doug Stevens, an amazing man who has an absolute passion for fishing. Doug explained about Trout Fishing, World Champion Fly Fisher, John Rumpf, demonstrated his skill with a fly fishing rod. There was much excitement along the lakefront when 10-year-old Joseph Esera landed the first trout. Joseph's success was followed closely by Danny Guevara, Andrew Scott, Dylan Christiansen, and R-leeo Maoate making a total of five good sized trout. Back at Camp, the trout were carefully gutted and smoked by HFNZ Vice President Richard Scott, and enjoyed by many.

Despite a lot of rain, the HFNZ Carnival Day went ahead as planned, with an array of activities for families to participate in and enjoy, including Archery, Mountain Biking, Kite Flying, Face Painting, and the novelty Motor Mower Challenge which generated much laughter. To top it all off, the generosity of Jan and Marcus Wilkins of the Rotoiti Explorer saw everyone off on a one-hour tour of the picturesque Lake Rotoiti.

At the Final Assembly we were very pleased to see eight children graduate

from Family Camp. That means they're now ready to attend HFNZ Youth Camps. Well done Logan Turner, Alex Rutherford, Izack Silva, Danny Guevara, Lachlan Fergusson, Sinead Edwards, Joseph Esera, and Connor Daly-Wilson We also took the chance to farewell Southern Outreach Worker Linda Dockrill, as this was her last National Family Camp before taking up a new role.

There are a lot of moving parts that go in to putting together a successful Family Camp. Thanks must go to our huge number of sponsors and those who donated goods and services to make this Camp a success:

- Kiwifirst for their donation of Pads and Pens for parents, High Viz Vests and Drink Bottles for Campers as well as equipment for the Group Leaders
- Doug Stevens from nzfishing.com for organising a wonderful day of fishing
- Honda, who provided caps for the children
- Suzuki, who donated bags for the children
- Kilwell, who supplied fishing rods and other gear for Fishing Day
- Fishermen from the Anglers Association: Thank you to those who came out to assist for the day
- Bunnings, for providing toys for the children
- McConnell Dowell Construction, who donated Construction Straws & Connectors Kits
- Jan and Marcus Wilkins from Rotoiti Adventures for such a wonderful day out on Lake Rotoiti
- Pfizer, for donating back packs for all children and leaders
- Sefton Kowai WI, for donating and making up goodie bags for the children's back packs.
- Thanks to Roger Manson for assisting us with motor(less) mowers for the hilarious Motor Mower Challenge.
- Thanks to Dulux for the donation of Paint for the Woodworking Projects.

Thanks also to Haemophilia Nurses Maureen Hayes, Mary Brasser, and BJ Ramsay, and to Cat Pollard, our resident Haemophilia Physiotherapist. Without the expertise of our Health Professionals, it would not be possible to provide a Camp such as this.

Much appreciation to all the HFNZ Outreach Workers for their support at National Family Camp – for the Sessions and Activities that they ran and their willingness to jump in when things needed doing. Also very much appreciated is all of the support from those back at National Office; Richard Chambers, Phil Constable, and Leanne Pearce.

Once again our Group Leaders – thank you for absolutely everything:

- The Architects: Jonathan Bollman, Amy Waters, Chanelle Spencer, Jean Marsh, Emma Fowkes, and Theodore Maoate
- The Pink & Blue Wrecking Crew: Courtney Stevens, Hannah Manley, Mei Hariki, Andrew Scott, Jethro Worthington, Gabby Hitchcock, Courtney Marjoribanks, R-leeo Maoate, and Dylan Christiansen
- The Speedy Construction Workers in the Creche: Ashley Taylor-Fowlie, Abbey Blackler, and Olivia Ferguson.

These young people worked so hard to ensure that the children in their

care really enjoyed participating in the activities, that they were safe at all times, and that they went home with wonderful positive Camp memories.

And also big thanks to Lynley and Richard Scott, our amazing Camp Resource Parents, for the many and varied activities undertaken to ensure the smooth running of Camp. We appreciate your enthusiasm, your willingness to undertake any task, and that you take time from work to support us all.

So, HFNZ Family Camp was AWESOME! We loved it, and by all accounts our members loved it too. The countdown is on for camp in two year's time...

MRG Reports

HFNZ's Member Representative Groups enable all our members to be involved in the running of the Foundation, and to connect with and support one another. Regional Events meet the social needs of members, with lots of fun, and a little education thrown in too. Here's what they've been up to recently.

Central region

By Stephanie Coulman

Our winter escape to El Rancho Waikanae in August was an enjoyable and active time for those who attended.

The venue was ideal, and there was a fantastic range of activities available; rifle shooting, archery, rock climbing, horse-riding. The most fun was archery-soft where teams shot each other with soft arrows, bringing out the competitive streak in many of us!

We held our AGM as part of the camp and have a new committee line-up. We farewelled Grant Hook, Michael Ho, Ross and Kelsey McCarthy, and Carol Reddie who all stepped down from the committee. In their place we welcomed Ashley Taylor-Fowlie and John Forbes. Ashley is our new Secretary and John joins the committee. It's great to have some new and young faces around the meeting table.

Stephanie Coulman continues as National Council delegate, and Chairperson for one more year but will not be standing as Chairperson at the next AGM. Blair Wightman continues as Treasurer, Kahurangi Carter is the Māori delegate, and joining John on the committee are Judith Dudson, Lorraine Gordon, and Lisa Habershon. The youth delegate position remains vacant; any young members out there may like to think about joining us? There are lots of opportunities for the younger members.

The committee has decided to continue the tradition of an annual regional camp, especially if we can find a cost-effective venue like El Rancho. The committee has other ideas for activities in the coming summer months, and we look forward to bringing those to you.

We congratulate Ashley who was invited by the World Haemophilia Foundation to attend a youth leadership workshop in Spain in November. The organisers were impressed with her youth presentation at the World Congress in Orlando and invited her back to share her ideas in Spain. ¡buen viaje! Ashley!

Our Christmas event at the end of November is Ten Pin bowling. Call us quirky, but we are aiming for concurrent events in Paraparaumu and Palmerston North. By the time you read this it will be done, and we think that it's going to be great fun.

Please note that Central have a new email address: hfnzcentral@gmail.com. Please don't hesitate to drop us a line with your thoughts and idea.

Midland Region

By Linda Mellsop-Anderson

We have had a bit of a spell from local activity due to individual circumstances and the committee regrouping. If you feel you could possibly spare a couple of hours supporting Midland, please don't hesitate to put your hand up – we will enthusiastically welcome you into our midst.

It's great to have wider representation in the organising facet of the group, wide age ranges and genders.

December 4th will be our farewell event for 2016, so mark it in your diary. The committee is organising a harbour cruise with a light lunch in Raglan from 1pm – 3pm.

Our young men R-Leeo and Theo are in the process of organising an event in Hamilton for young members between 12 -17 – Bowling & Pizza. This event or something similar will also be happening in Tauranga and Rotorua in 2017.

We are excited to announce that early February will be an opportunity for our Masters to have a men's boat trip; trout fishing in Taupo.

Also in our planning diary is an event for women next year, which still in the development stages.

It is our intention to host desert evenings in Hamilton, Tauranga and Rotorua. These are just an informal opportunity to chat over a coffee and dessert. Weight Watchers' of course! Dates and venues to be confirmed.

As this is the last Bloodline for 2016. on behalf on your Midland Committee we hope you all have a safe and enjoyable holiday break, however and where ever you celebrate.

Northern Region

By Neil Smith

Hi all,

Our AGM this year was held in Botany, with a few familiar names and some new blood(!) too. Shout out to Lynley Scott, who has served many a year, organised, run, minuted, counselled, and consoled, and leaves the Northern region committee in a fantastic state. Thank you for keeping us going Lynley. AGM formalities were followed by a great afternoon of bowling, gaming, and threading one's self through a laser-maze, mission impossible styles. We have some contortionists among us.

Personally, I got to experience the National AGM in Christchurch where I got to appreciate the level of effort, energy and enthusiasm it takes to run an organisation such as this. Being new to the northern committee it was fantastic to share ideas and lessons learned with people around our other regions, and also hear from Deon and Richard on simplifying the message around what HFNZ is all about.

Family camp in Rotorua was once again a well organised, a kids dream, an enjoyable weekend out for adults, and, as usual, we always learn something new. Really, it's just nice to share a bit of life with fellow bleeders. Sharing ideas and frustrations with people we hadn't met before was a particular highlight where we can see how we can make simple positive changes supporting one another. As a Von Willebrand's family, it was great to have a few focussed conversations around support and understanding in this area.

Volunteering for Armageddon is an annual tradition for Northern members and this year we did it wearing capes, nice blue silky capes, that got in the way whilst you worked. Can't think why super hero's wear them they are just a nuisance. Our duties this year varied from handing out show bags, to queue minding for celebrity signatures, to minding the information desk, wandering the crowds, and checking the weapons.
We had a lot of fun and got to see a few celebrities (Doc Brown from Back to the future, and the Phelps twins from Harry Potter) as we minded their autograph queues.

Thanks to all who volunteered their labour weekend especially those that worked several days, (Hannah-Jayne Rodokal and Ken and Donna MacGregor). Ken did a great job of organising the volunteers and went beyond the call of duty being there each of the three days. We had fun watching all the different people in their weird and wonderful costumes and occasionally checking their weapons to make sure they were safe! We are not sure how much was raised just yet. If you haven't volunteered before have a go next year, it's an experience you won't forget.

Our events coming up are:

- Rainbow's End Christmas Party 10th December (more details to come)
- Northern Camp – Mangawhai, weekend March 3rd, 4th

Please get in touch if there's anything you'd like to see more (or less) of, as we're your committee in Northern representing you.

Thanks

Piritoto

By Kahurangi Carter

Kia ora, I'm Kahurangi Carter (Kahu), the Central Region Takawaenga (Central Representative) on Piritoto. My family live in Ohakune right under Mt Ruapehu. I would love to hear from you and know the kinds of events you would like to see in our district.

Te Whainoa, our Council Representative, and I were invited by Head Office to speak to our staff and Outreach Workers on how best to connect with and support our Maori Whanau who have a bleeding disorder. We started at the beginning with our whakapapa and genealogy and look forward to more sessions to implement a strong health model for the benefit of all in HFNZ.

We are looking forward to 2017 and would love to see you and your Whānau at our next Marae Noho! Watch out for your invite. If you have not had an invite from us, that means you are not on our mailing list... Please contact me on Kahurangicarter@gmail.com so we can keep you up to date with our events! We would love to connect with you and your Whanau and get your feedback. Piritoto are always looking at how to increase our Whanau involvement and we want to hear from you.

Ka Mau Te Wehi!

We look forward to seeing you there next year!

National Youth Committee

By Courtney Stevens

As always, the Youth have been super busy trying to organise and plan events in our regions and nationally as well. On top of that, many Youth members have been involved in helping out at the National Family Camp, and other regional events. Before the Family Camp there was also a training day, which is always a good opportunity to meet up with other Youth members, and to learn some new skills.

The National Youth Committee are planning a winter getaway in 2017 in Queenstown, comprising of both educational and fun elements, for us to connect and grow relationships within our bleeding disorder community. Keep an eye out for invitations and further information to follow soon!

At our last meeting we made some progress with timelines for our planning and have delegated tasks amongst ourselves to help get things over the line.

We are looking at planning some end of year social events regionally for a catch up before the busy Christmas season!

Wishing everyone a safe and happy holiday season.

A Message from Linda

Departing Southern Outreach Worker, ***Linda Dockrill,*** *has been supporting HFNZ members for over seven years. She's been a huge asset to the Foundation, and we're going to miss her. How do you replace the irreplaceable? Well, Josiane McGregor looks like a very good start. She takes over on November 23rd, and you'll all get to meet her as time goes by. In the meantime, here's a final message from Linda…*

It is with a heavy heart that I say goodbye to my role as Southern Outreach Worker and to all of our members of the HFNZ community. I have taken a role at the Aotearoa New Zealand Association of Social Workers (ANZASW) as a national Professional Development Co-ordinator. This is the professional body for Social Workers in New Zealand. It is a very different role compared to Outreach work but it has some big challenges for me and I expect to learn a lot.

My nearly 7 years at HFNZ have been a lesson in life and in resilience. I have loved my time at HFNZ and will miss working with you all. It has been a total privilege to know you and your families so well. As a Social Worker there are very few roles that enable the depth of relationship that I have had with many of you. I am grateful for the opportunity to work with and for a community that has welcomed me, taught me, and allowed me to become a participant in your family life. I have relished the camps and workshops, meeting many of you from outside the South Island, and having fun along with learning. I won't be forgetting laughter yoga, helicopter rides, or laser strike in a hurry! Such fun!

I have grown to know many of you well and have appreciated the level of dedication and support the staff at HFNZ, the clinical teams, and the National council show. Particular mention must go to Colleen McKay who has been my mentor and manager all of these years. Colleen's dedication and commitment to this community is inspirational.

My heart is full of all the aroha and community spirit that I have found working in this diverse and unique community. I will miss you all and sincerely hope that our paths will cross again. Take care out there until then.

Arohanui

Linda Dockrill

News from Around the World...

CSL Behring's Therapy for Severe Hemophilia A Shows Promise in Clinical Study

By Margarida Azevedo

CSL Behring's rVIII-SingleChain, a novel recombinant Factor VIII (rFVIII) treatment candidate, recently demonstrated effectiveness in surgery and in the control of bleeding in patients with severe hemophilia A.

The Phase 1 to Phase 3 clinical trial results, recently published in the journal Blood, also demonstrated low annualized bleeding rates in patients on a medication, and it was well tolerated. The research paper detailing the therapeutic effects is titled "Efficacy and safety of rVIII-SingleChain: results of a phase I/III multicenter clinical trial in severe hemophilia A."

The clinical study of rVIII-SingleChain, "An Open-label Safety, Efficacy and Pharmacokinetic Study of a Recombinant FVIII Compared to Recombinant Human Antihemophilic FVIII in Patients With Severe Hemophilia A," was conducted by Dr. Ingrid Pabinger, MD, professor at the Medical University of Vienna, and her colleagues.

The study's main objective was to assess the therapy's effectiveness for the treatment of hemophilic bleeding episodes and as a routine and surgical medication.

The study included 173 adolescent and adult patients treated with rVIII-SingleChain. Of the total, 146 were assigned the medication as prophylaxis (preventive measure) and 27 received it as on-demand therapy. Researchers observed a total of 848 bleeding events which were treated with rVIII-SingleChain, either as on-demand therapy (590) or prophylaxis (258).

Treatment with the novel therapy controlled 72.2% of the bleeding events with excellent efficacy, and 21.6% of bleeding events showed good efficacy. In the remaining 52 events (6.2%), the therapy demonstrated moderate effectiveness.

Importantly, 43% of patients treated with the product as a preventive measure (prophylaxis) had no bleeding events. Surgical hemostasis was rated excellent to good in all of the 16 surgeries on study, and none of the participants was found to develop FVIII inhibitors.

The most common reported adverse events were headache, arthralgia (joint pain), and nasopharyngitis.

"This study, which was designed to reflect clinical practice, demonstrated with a robust dataset that rVIII-SingleChain is highly efficacious in the treatment of bleeding events, routine prophylaxis, and in controlling hemostasis in a variety of surgical procedures in adolescents and adults with severe hemophilia A," the authors wrote in a news release.

"The study also demonstrated that rVIII-SingleChain has a favorable safety profile and is well tolerated. Very low annualized bleeding rates in patients on individualized prophylaxis hopefully has the potential to translate into prolonged freedom from debilitating joint disease," they wrote.

Source: http://hemophilianewstoday.com/2016/08/24/csl-behring-therapy-severe-hemophilia-a-appears-safe-effective-study/

Genentech's Hemophilia A Data Presented at 2016 WFH

By Andrew Black

Exciting data was presented at the 2016 World Federation of Hemophilia in Orlando, Florida last week on Genentech's hemophilia treatment drug, emicizumab. In the Phase 1/2 trial of emicizumab, the drug continued to demonstrate promising safety and prophylactic efficacy for people with severe hemophilia A, with or without inhibitors to factor VIII.

The Phase 1/2trial assessed emicizumab in an extension of a 21 to 32-month period which included Japanese patients with severe hemophilia A ages 12 to 58 years that were organized in three different dosing groups receiving 0.3 mg, 1mg, or 3mg of emicizumab.

Emicizumab successfully reduced bleeding events by more than 95% in each group that was administered the drug. The successfulness of the Phase 1/2 trial in the treatment of hemophilia showed progression of the drug which excited Genentech.

The next step is a Phase 3 trial that is currently underway. The trial will be an open-label study will evaluate the safety, efficacy and pharmacokinetics of prophylactic emicizumab treatment in patients previously treated with episodic or prophylactic bypassing agents. Phase III will enroll patients 12 years of age or older with hemophilia A with factor VIII inhibitors. Genentech also plans for additional pivotal studies that are planned to commence soon.

Emicizumab

Emicizumab is an investigational humanized, bispecific monoclonal antibody. It is designed to support the interaction between factors IXa and X which are proteins that work together in the blood clotting process, thought to restore clotting function and thereby prevent spontaneous bleeding. Emicizumab can be administered subcutaneously once a week and can stay in the body for four to five weeks, which could allow for weekly or potentially less frequent administration.

Hemophilia A

Hemophilia A is a rare, chronic, genetic disorder that results in impaired clotting mechanisms due to missing or reduced levels of factor VIII. People with hemophilia A experience recurrent and extended bleeding episodes that cause pain and irreversible joint damage. Some of these bleeding episodes can be life threatening. There are an estimated 16,000 – 20,000 people in the United States with hemophilia A (and 142,000 world-wide).

Source: http://www.raredr.com/news/genentech-hemophilia-data-2016

The Year Ahead

More details on all events are available from your local Outreach Worker.

The Holiday Season

The HFNZ head Office will be closed from December 24th until January 4th

Waitangi Weekend: Feb 3rd - 6th

Advanced Leadership Training (Invitation Only)

April 17th

World Haemophilia Day & Buddy Awards

April 20th -23rd

National Youth Camp
Waipara Adventure Centre

July (*TBA*)

National Inhibitor Workshop
Auckland

September 22nd - 24th

Adult Weekend
Venue tba

P.S. Keep an eye out for PEP, coming up on a date yet to be determined in 2017

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

Time to Get Bloodline Online?

Choose to receive your copy of Bloodline by email instead.

Just let us know...
info@haemophilia.org.nz

Honour the memory of a loved one or recognise the unique bond you have formed with HFNZ to make a difference to the quality of life of people with bleeding disorders.

*Information on making a bequest in your will to the HFNZ can be found at **www.haemophilia.org.nz***

give a little

Give a little? Give a lot!

Charitable donations to HFNZ can now be made online at:
www.givealittle.co.nz/org/haemophilia