Haemophilia Foundation of New Zealand Inc.
HFNZ

bloodline

Newsletter of the Haemophilia Foundation of New Zealand Inc

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The H Word

The largest international meeting for the global bleeding disorders community takes place in May in Melbourne. This has been a rare opportunity for HFNZ to send a larger group than ever before to a World Federation of Hemophilia Congress! With the regular registration deadline being 14 March, take the opportunity to remind your haematologists, nurses, phyisotherapists, dentists and anyone involved in your care to register and take advantage of this unique opportunity. Never has this meeting been so close to New Zealand! For more information go to www.wfh.org

Deon York
HFNZ President

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Cover photo:
Jayden Taurerewa and Nariko Waru at the 2014 Children's Education Workshop in Tauranga

Terrific Times learning "Tricks and Tips"

2014 Regional Children's Education Workshops

The year got off to a 'magical' beginning as 36 children aged 6 – 10 years, all who have either haemophilia or von Willebrands or who have a parent or sibling with a bleeding disorder, attended the regionally held 2014 Children's Education Workshops.

Each of the four workshops had the same magical theme "Tricks and Tips", followed the same programme but was held at difference locations around the country to make it easier for more children to attend.

- The Northern Workshop was held in Auckland on 14 January;
- The Midland Workshop was held in Tauranga on 16 January;
- The Southern Workshop was held in Christchurch on 21 January; and
- Last but certainly not least, the Central Workshop was held in Palmerston North on 1 February

At the beginning of each workshop the children made and decorated their own magical hat-shaped name tag and got even more creative as they decorated their own 'Bag of Tricks' and made their own magic wand. A pack of cards generously donated by the Christchurch Casino were also packed into the 'Bag of Tricks' in readiness for future magic tricks.

With all their magician gear ready it was time for the local Haemophilia Nurse Specialists to give some age appropriate education about how blood clots, the different bleeding disorders and bleeding. While the children learned that they could not make the bleeding disorder magically disappear, they did learn that there were tricks to make life easier (see page 2).

The Physiotherapists also had some tricks to share – the primary trick being P.R.I.C.E.

The next essential items for the magicians-in-training were juggling balls. Balloons, rice and freezer bags were magically transform into juggling balls, and there was much laughter as everyone tried their hand at juggling. In Central, multi-talented Haemophilia Nurse B.J. Ramsay and Youth Leader Waylan Tomlinson in Southern impressed with their juggling skills.

At the end of the day a real magician visited each workshop for a Magic Show. After performing some amazing tricks the magicians also taught the children a couple of magic tricks they would be able to perform at home to amaze their families.

Education about bleeding disorders is not just a one-day activity such as the Children's Workshops, it is an ongoing process. To help with this the children all went home with a specially designed workbook full of activities in order to continue the bleeding disorder discussions with their parents and families.

The Children's Workshops combined a mix of education and learning with a sense of togetherness by mixing with other children in a similar situation and, as with every HFNZ workshop, a good portion of FUN to make it all work together.

Comments from children and their parents:

"I learnt a lot about haemophilia at the Children's Workshop. It was lots of fun, I enjoyed the magic show. The lunch was yummy too. I hope I get to go to another Children's Workshop one day. Thank you for letting me come."
– Izack Silva (Child with haemophilia)

"Connor had a great time at the Children's Workshop. It gave him a chance to learn more about haemophilia in an environment without his parents. There were also a lot of fun activities and chances to make friends with other kids going through and managing the same things he is."
- Greg Jamieson (Parent).

"It was really cool! I learnt lots about Haemophilia and my favourite bit was receiving a heart balloon and learning how to do lots of magic tricks which I'm now trying out on my school friends. Love from Isla ♥". – Isla Brodie (Sibling)

"Both of the kids really looked forward to going & as with their past "Hemo" events camps etc knew they were in for a good time.

Rorie had a fantastic time & enjoyed catching up with his peers at the Workshop. As usual Rorie is quiet on the day of the event and on the drive home – but once he has thought about it he talked about it non-stop. He loved the magician and of course the food!!

Maisie loved being included & was chuffed to have been invited (& quietly enjoyed the fact that her little sister was not going). Maisie has a lot of empathy towards Rorie & haemophilia in general and I think an event like this just explains it a bit better, which in turn increases her understanding. On the way out I asked Maisie if she had a good day & her reply was – "nope I had a fantastic good day". – James Poff (Parent)

"I loved learning more about Haemophilia and the games and magician. It was my day." – Toby Scott (Sibling)

"We appreciated that this year the siblings were included. While Toby is a sibling he is still affected by haemophilia. We could see he felt important attending about it. And [it was] great for him to have specific education about Haemophilia. Thanks everyone!!" – Lynley Scott (Parent)

HFNZ would like to say a really big thank you to the Haemophilia Nurse Specialists – Mary Brasser, Maureen Hayes, Daryl Pollock, BJ Ramsay and Kathy Fawcett – and the Haemophilia Physiotherapists – Lee Townsend, Amie Ansley and Lisa Weaver – who gave up their time to attend and for their very creative presentations. Thanks also to the Haemophilia Outreach Workers for everything, Sarah Preston, Joy Barrett, Lynne Campbell and Linda Dockrill.

HFNZ would also like to thank Pfizer for their grant towards the Children's Education Workshop – your support makes workshops like these possible.

Terrific Times learning "Tricks and Tips"

Tricks and Tips for living with a bleeding disorder

Learn how to recognise the signs and symptoms of a bleed early

Telling someone straight away if you are having a bleed

Report your bleed to the Haemophilia Centre

Have prophylaxis treatments on time and in the morning

Choose appropriate exercise to keep fit and to keep muscles and joints strong

Eat well and keep weight within a healthy range.

Preparing for School Trips and Camps

School excursions and camps give children the opportunity for new experiences and learning. Although children with bleeding disorders usually require some additional consideration for these activities, with some pre-planning and general guidelines, children with bleeding disorders can enjoy many of the same opportunities as their peers.

Here are a few tips for parents and youngsters on preparing for various types of school activities — from trips and camps to extracurricular activities.

Benefits of participating in school activities and trips

More than just fun, school trips and extracurricular activities offer many great benefits to children.

School trips reinforce learning. Students get to experience their lessons in real-world settings. These outings put lessons in context and are particularly helpful for children who learn by doing. And, they cut down on boredom!

Extracurricular activities are a great way for students to make friends and discover new interests and hobbies. They're ideal for children who are not interested in school team sports, or who may be side-lined from certain physical activities. Intermediate and high school open up a whole new world of opportunities that are more scholarly and artistic, such as the chess club, band, and service clubs. Getting involved in such activities will help your child explore his or her own identity and interests and build bonds with peers who share similar pursuits and passions. Indeed, activities started in intermediate or high school often continue throughout a lifetime.

When it comes to school field trips and extracurricular activities, your child has the right to participate. A child cannot be denied participation because he or she needs medication or treatment, or requires special assistance. If a school trip or camp is planned, the teacher should give you, your child, and the school administration enough time to put together a plan to meet your child's needs. In some cases, you or a school staff member who has been educated about your child's bleeding disorder may want to go along with the class. Work with the school to ensure that your child has access to medication at all school-related activities.

Dealing with anxiety

School camp is often one of the highlights of childhood – chances are you can still remember some of the campfire songs, the smell of damper cooking and the names of the kids you bunked with. However, it's also true that for some children (and their parents) the thought of staying away overnight can create a little anxiety.

It's not uncommon for kids to worry about who they'll room with, 'being left out' of things, whether something might go wrong, feeling homesick and other understandable concerns.

Even the most outgoing and confident child may have a few moments of separation anxiety as the time to leave gets closer. Helping your child feel positive about going to camp is important, as it provides a new opportunity for them to stretch their wings.

In general, once they are there children enjoy the challenges that camps provide – sharing a room, embracing challenging activities, sharing responsibility for things like washing up and getting to know their teachers outside of the classroom.

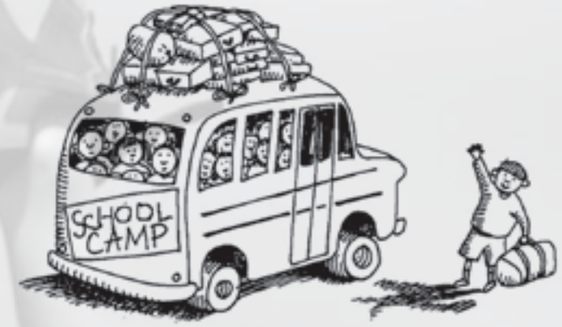
It's worth remembering that every challenge your child overcomes contributes to their resilience and self-confidence when facing more unfamiliar situations later on.

School camps – extra preparation needed

HFNZ camps are a fantastic way to meet other children with bleeding disorders and learn important skills for growing up with bleeding disorder. The activities are all planned with bleeding disorders in mind. School camps can feel very different but they are a great opportunity for your child to push their boundaries, discover new friends and experience outdoor adventures first hand.

There are many ways you can help prepare your child for school activities or camps, beyond just making sure their treatment is arranged.

- **Talk to your child.** When the opportunity to attend school camp arises, ask your kids how they feel about it. Are they excited by the idea? Do they have any fears? During this conversation you may be able to discern any anxieties or unnecessary mental obstacles, and reassure them with your own positive school camp experiences from your own childhood. Be positive by saying things like "this will be fun, you'll enjoy the challenges". Try to find solutions or strategies for dealing with their fears. For example, if they are frightened that they will be teased because they still sleep with their Teddy or blankie reassure them that Teddy is easily hidden at the bottom of their sleeping bag for them to find when they go to bed at night and no-one else needs to know that Teddy has come to camp!! If they are frightened because they still wet the bed try talking to their teacher about a way to deal with this discretely, such as the mother help coming in and collecting the sleeping bag for washing and replacing it with another one while the children are at breakfast.
- **Reduce the sense of unknown.** Ask the teacher if you can have a rough breakdown of the daily schedule at camp. This way you can help your child envisage all the fun activities and realise that there is a beginning, middle and end to their stay. This can also help you to make plans or arrangements for dealing with activities that might be too physically risky for a child with a bleeding disorder.
- **Practise sleepovers.** Ideally, your child is used to the occasional sleepover at a friend or family members' home. Having a bleeding disorder should not be a reason not to experience sleepovers. If your child refuses sleepovers or regularly calls you in tears at midnight needing to come home, you may need to work with them in the months before camp to reduce their anxiety (or yours).
- **Prepare, make lists, and pack together.** When your child brings home the list of clothes and toiletries they need for camp, get them to help you find (or shop for), then label their things. It's exciting for them to help prepare and pack, but it will also help them be aware of all the things that need to come back home again! Don't forget to include all your treatment supplies.
- **Practise 'independence' skills at home.** Does your child do their hair, remember to clean their teeth, know how to apply sunscreen and turn the shower taps on and off in the right order? There are lots of simple skills they can practise to feel more confident about being away from home.
- **Keep communication with school staff.** Your child's teacher has probably taken hundreds of kids to camp over the years. They understand that children and parents are stepping outside their comfort zones for the first school camp. Don't be afraid to send a note to the teachers who will be supervising your child to raise any concerns you have. They'll appreciate the extra information as makes their time at camp easier too.
- **Make arrangements for treatment.** Speak with teachers about any help your child might need while they are away. Before going tell your Haemophilia Centre so they can contact the nearest hospital or GP in the area of the camp to explain what care you might need while you are there. For example, arrangements can be made for your child can take their treatment product and sterile supplies with them and the GP or a doctor at the hospital can give them their treatment. Contact your Haemophilia Centre if you have any queries and make sure their details are shared with your child's teacher. Often a parent might go on the camp to help out as one of the parent helpers. This way they can be on hand to administer treatment when needed.



- **Learn self-infusion.** When a child starts to self-infuse it is the next stage in them taking control of their haemophilia. They can now take care of themselves and that creates a feeling of independence and confidence.
- **Keep positive.** Telling your child you believe they're ready for school camp and they'll have a great time helps your child believe it too.

A good way to help prepare for school camps is coming to HFNZ Youth Camp. Because of the need for treatment or prophylaxis HFNZ Youth Camp is often the first opportunity for youngsters with bleeding disorders to get away on a camp without mum or dad. They are an excellent place to gain confidence, meet new friends and learn important skills like self-infusion. Because they are planned specifically for children with bleeding disorders all activities are fully inclusive and haemophilia nurses are on site to help with treatment. HFNZ has short videos about Youth Camp that children and their parents can watch so that children can see adventures and parents can see that there is a purpose and that their child will have FUN. A link to the videos can be requested through your Outreach Worker or the National Office or visit the HFNZ page on YouTube https://www.youtube.com/user/haemophiliaNZ/videos (Search for HFNZ or haemophiliaNZ). The next Youth Camp will be in October 2015.

Preparing for School Trips and Activities

Here is a checklist to help you prepare for your child's school trips and activities:

- My child's treatment protocol is current and a copy has been given to the school nurse or administration.
- My child and his/her teacher have a copy of the Sample Letter for Trips and Activities (see page 6), which has been personalised with my child's specific needs.
- The teacher knows to give copies of the Sample Letter for Trips and Activities to all chaperones.
- My child is carrying a copy of his/her treatment protocol and is wearing his/her medical alert jewellery.
- My child has taken the necessary medication/infused prior to the trip or activity (if applicable).
- My child is carrying an Emergency Kit, which includes a cool pack and a traveling dose of factor/medicine.
- I have carefully read the teacher's instructions and researched the location of the trip.

SAMPLE LETTER for Trips and Activities

This is a sample letter. Please work with your Haemophilia Treatment Centre (HTC) and, where applicable, your local school district to create a letter that works best for your child.

____Name of Parent_____

_____ Address _____

(***) *** - **** (mobile)

(***) ***- **** (home)

[Date]

To: _____

Just a reminder for you that _____, ____ years old, has_____.

As you already know, _____ participates in all school activities. In an overnight situation like this, however, you need to keep several things in mind:

1. _____ should not engage in wrestling or contact sports like tackle rugby–all other sports are fine.
2. Although _____ has received a dose of factor, he may need additional treatment if other trauma does occur.
3. _____ is traveling with one dose of his factor in case of an emergency that would warrant his transportation to a hospital.
4. _____ can communicate to you if he is having an internal bleed, and please take him seriously.
5. Treat any cuts, scrapes, or bruises that he may get the same as you would with anyone else.
6. In case of nosebleed use pressure on the nose, a cool compress on the back of the neck, and keep his head forward.
7. _____'s medicine should be kept away from heat and freezing. It can remain out in his room with his stuff at room temperature.
8. Under no circumstances should _____ be given aspirin, ibuprofen (eg, Nurofen), naproxen, or any products containing those items, because they promote bleeding. He may only, and only if needed, take paracetamol products for pain or fever.

In case of emergency, contact _____ at the Haemophilia Centre for further information at (**) ***-****.

If you need to contact me, please call: _____ at (**) ***-**** (cell), or (***) ***-**** (home).

Have fun!

Profile: Reg Fuller

By Chantal Lauzon

When Reginald (Reg) Fuller turned 70 last year he felt it was a real milestone. Considering all the medical issues that Reg has faced, it was certainly an occasion to celebrate.

Reg's earliest memory of any trauma is of being wheeled along a hospital corridor after being hit on the head with a stone thrown by one of the neighbour's children. He remembers sheets soaked in blood and a trip in an ambulance. At the time, treatment was a bandage for his head and transfusions with whole blood.

His family already knew that he was a "little bleeder" but it was a few years before he was officially diagnosed with mild haemophilia B. His cousin was already known to be a bleeder as well. During his teens it was necessary for Reg to have his "wisdom" teeth removed and it seemed that the hospital dentists may not have had an appreciation for the complications of removing four molars at the same time on a patient with a bleeding disorder. The prolonged bleeding led to two weeks in hospital, having his gums packed and stitched without anaesthetic and having transfusions for the blood loss. This, he says, taught him how to deal with physical pain.

Reg married Sue in 1966 and two children followed. After working as a draftsman for the Ministry of Works, Reg trained as a civil engineer and worked for a number of local authorities following jobs around the North Island. His passion was carrying out field work, designing and bringing a project to completion through contract selection and supervision. Many of his structures, such as bridges, roads, pump stations and the like are still in use today.

Reg and Sue moved back to Christchurch from Whangaparoa in 1980 for family reasons and Reg found work at the Waimairi District Council.

Home, Reg believes, has a lasting influence in the shaping one's character. He spent a happy childhood growing up in Christchurch with his parents and four sisters. He remembers home as the centre of the family where his Dad kept a good garden, a garage full of interesting machinery and a place to learn about car repairs. His mother was the one who dispensed wisdom to her offspring and her life skills and values are reflected in all of her children.

Since retiring in 1994 for health reasons, Reg and Sue have built a lovely family home with a beautiful garden on the hills overlooking the city. Their home was gratefully spared from major damage in the earthquakes but the emotional toll still lingers, just as it does with everyone who has experienced this natural disaster. Reg finds it frustrating living in city under so much re-construction. He would dearly love to have been involved in restoring his city to the beautiful place it once was.

Ever optimistic, he knows that the future Christchurch will be a place of beauty once again.

Coming from a family that valued independence, Reg was, by nature, not one to stand back and be intimated. This attitude has served him well; whether it was standing up for himself and others or dealing with the health issues that he has had to cope with throughout his life.

In the 1980s real advances in medical science meant that treatment options available to Reg began to improve dramatically; although Reg admits he still doesn't always treat bleeds as soon as he should, usually adopting the typical "she'll be right" attitude. With the medical advances, however, came unimagined consequences. Reg was treated with Factor IX for a knee injury and surgery in 1983; and in 1985 he tested positive for HIV. It was an indescribably stressful time for his family, but luckily Reg has been effectively treated with a succession of medications as new antiretrovirals became available.

In the meantime, Reg was also diagnosed with ulcerative colitis for which he has needed treatment on and off over the years, including several hospitals stays due to haemorrhages and blood loss. One episode had him undergo cardiac arrest while waiting for treatment at the hospital. Fortunately, he was in the right place at the right time and the appropriate personal were on hand immediately to resuscitate him. Reg hold all his medical carers, most especially the "haemostasis team", in high regard and knows that without them he would not be of this world.

Reg was also diagnosed with hepatitis C in 1991, which was successfully treated with pegylated-interferon in 2005. Sounds undramatic but in reality it was a year of "hell" for him and his family as they not only had to deal with the effects of medication but also a change in Reg's personality. Not for the better he admits.

Unbelievably, that is not where Reg's medical adventures end. In 2008, Reg had a total hip replacement (Aly Inder was there at this bedside when he awoke from the anaesthetic; such is the dedication of his care givers). Then in 2010, he underwent an off-pump coronary artery bypass graft. Amazingly he had no complications from this surgery. According to Reg, "All the medical people that took part in the decision making process throughout New Zealand and those that cared for me with pre- and post-treatment are the true heroes. They all took a huge risk in deciding that it was possible to successfully perform this operation." This type of operation had only been carried out twice before in Australia and never before in New Zealand on someone who had a bleeding disorder as well as HIV.

Always in contact with HFNZ, it has only been in recent years that Reg and Sue have become more active members. They attended the recent Adult's weekend in Christchurch and, for the first time, Sue met others with haemophilia and their partners. They

have since kept in regular contact with others they met there and have also attended other Southern Region events.

When the prospect of a Masters group started up at HFNZ, Reg decided it time to get more involved and give back to the Foundation some of his knowledge and experience living with haemophilia. Having had parents in retirement homes opened Reg and Sue's eyes to the necessity for caregivers to be made aware and be educated about the special needs of people with bleeding disorders as they age. He also thought it would be a good way to keep up camaraderie with his peers across the country.

Always active, growing up Reg experimented with rugby and football until the damage it was doing to his joints made him give them up. Later he played badminton competitively (at Pukekohe and Warkworth) and then golf. When the Masters group started a programme to virtually "Walk to the Congress" Reg decided to make an effort and participate. Essentially, a group of Masters members are trying to walk/bike/swim/work out to the equivalent of the distance between Wellington and Melbourne in time for the Congress in May. Reg started a walking programme but the kilometres weren't adding up fast enough so he has started biking as well to get his distance up. At the last update the members of the group participating had covered just over 1200 km! No mean feat for those with 'dodgy' joints! Updates on the progress can be found on the HFNZ Masters private Facebook group page. Search for HFNZ Masters and ask to be added to the group. Any posts are private to group members.

Other than the walking and biking, Reg keeps himself pretty busy with general hobbies, gardening, playing music, reading and helping Sue with her bead work. The couple regularly take holidays to the North Island or Australia and have visited their son in Sweden where he lives. Everything they do, Reg and Sue approach as team. It is clear that they make a very happy team indeed.

Reg claims to have lived pretty much an ordinary life. I'm sure Sue, and anyone else who meets him, would agree it is instead a pretty extraordinary one.

The article originally appeared in the Summer 2013/14 issue of Talking Hep C magazine, published by the Hepatitis Foundation of New Zealand. If you would like a free subscription contact The Hepatitis Foundation of New Zealand on 0800 33 20 10 or visit www.hepatitisfoundation.org.nz

Cirrhosis vs. fibrosis

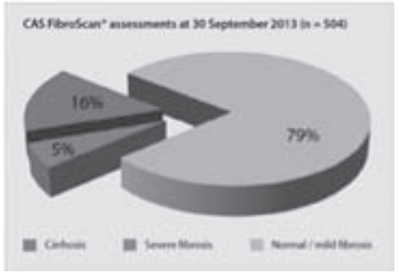
What do I need to know about cirrhosis?

by Professor Ed Gane, Liver Unit, Auckland Hospital

Cirrhosis is a word we are hearing more often. Within the Hepatitis C Pilot, run by The Hepatitis Foundation of New Zealand, we FibroScan® those enrolled in the Community Assessment and Support (CAS) Programme. Out of 504 people, we have diagnosed 107 people with severe fibrosis or cirrhosis.

When the liver becomes damaged, it forms scar tissue. Fibrosis and cirrhosis are levels of scarring and damage. When the liver first becomes scarred, it is known as fibrosis. When the scarring increases so much that the liver can't function properly, this is known as cirrhosis.

Not everyone with chronic hepatitis C will develop cirrhosis. However, if you do develop cirrhosis you should be observed every six months to check for signs of liver failure or liver cancer.



Professor Ed Gane, the Deputy Director of the New Zealand Liver Unit at Auckland Hospital provides an in-depth look into cirrhosis.

What is cirrhosis?

Cirrhosis is a term used to describe a diseased liver that has been badly scarred, usually due to many years of injury. The most common cause of this ongoing injury is heavy and continuous alcohol use. The next common cause in New Zealand is chronic (long-term) viral infection of the liver with either hepatitis B virus (which affects mainly Polynesian and Asian New Zealanders) or hepatitis C virus. The most rapidly increasing cause of cirrhosis in New Zealand is fatty liver caused by obesity or diabetes. Other less common causes include inherited conditions and conditions caused by abnormal immune response (such as autoimmune hepatitis or biliary cirrhosis).

How is cirrhosis diagnosed?

In the past, the "gold standard" test for cirrhosis was a liver biopsy, which is painful and carries risks of bleeding. This has now largely been replaced by the non-invasive FibroScan® machine.

Sometimes, diagnosis of cirrhosis can be made on blood tests or with ultrasound or CT scan of the belly.

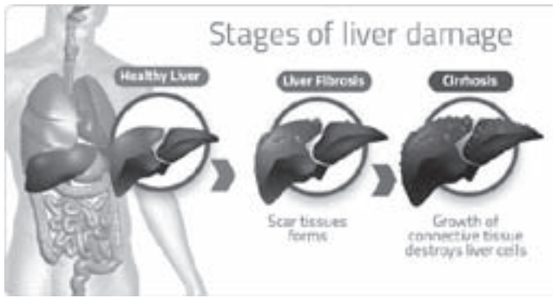
How common is cirrhosis in New Zealand?

There are estimated to be more than 20,000 New Zealanders with cirrhosis – the vast majority of whom feel very well.

However, every year around five per cent (one in 20) will get a serious complication such as liver failure and liver cancer.

What should I do if I have cirrhosis?

1. Avoid all alcohol;
2. Avoid all cannabis;



3. Lose weight if you are obese and improve diabetic control;
4. Optimise your nutrition. Avoid high salt intake and have a high calorie/high protein food intake. See your doctor/dietician about starting nutritional supplements such as Fortisip or Ensure. Avoid exclusion diets – they do not work and can speed up the malnutrition process;
5. Make sure that you have regular blood tests and a medical review at least every six months;
6. Avoid sleeping tablets and codeine as these can cause coma in patients with liver failure.

What are the symptoms of liver cancer?

There are none until the liver cancer grows large enough to stretch the liver capsule or when the cancer has spread outside the liver. By this time, the liver cancer is no longer curable. Small liver cancers have no symptoms, they can only be detected by ultrasounds or CT scans of the liver. There is usually a window of two to three years, when the liver cancer is large enough to be seen by ultrasound, but still small enough to cure. Hence, everyone with cirrhosis is offered six-monthly liver cancer screening.

What are the symptoms of liver failure?

Most people with cirrhosis have none. However, symptoms often develop when the liver is no longer able to perform its normal functions. The earliest symptoms are increasing tiredness, loss of appetite, and then loss of weight. Occasionally, the patient may develop itchy skin, which does not go away. The patient may find their thinking is not as sharp as it was and, initially, attributes this to age. They may need longer to complete a crossword or remember names. This may become more obvious to family and friends. People with progressive cirrhosis may experience an "inverted sleep pattern", where they cannot sleep at night but need to have a nap during the day.

If cirrhosis progresses and the liver starts to fail, there can be obvious changes to a person's appearance. These changes include:

- Loss of muscle around the neck and shoulders (protein calorie malnutrition).
- Easy bruising of the skin.
- Swelling in the legs (oedema).
- Swelling of the belly because of build-up of fluid. This is associated with weight gain. Occasionally this fluid may move up into the right side of the chest, resulting in shortness of breath and inability to lie flat.
- If the fluid in the belly gets infected, this causes abdominal pain and fever. This is a medical emergency and the patient must go to hospital immediately.
- Darkened urine and yellow tinge to whites of the eyes and skin (jaundice).
- Vomiting blood or passing black, tarry bowel motions caused by burst varicose veins in the gullet. This is a medical emergency and the patient must go to hospital immediately.
- Confusion and drowsiness, known as encephalopathy. This is a medical emergency and the patient must go to hospital immediately.

What will my doctor suggest?

Treatments depend on the cause of cirrhosis and how advanced it is.

1. Treat the cause of the disease:

- If you have hepatitis C cirrhosis and do not have liver failure, you should consider hepatitis C treatment. Current studies demonstrate the new oral antivirals will be safe and more effective in cirrhosis even if you have liver failure. The first polymerase inhibitor, sofosbuvir, will be approved in 2014.
- If you have fatty liver, then weight loss and better control of diabetes will prevent complications.
- If you have alcoholic cirrhosis, then stopping alcohol will help the liver recover, even if you have liver failure.

2. Prevent bleeding from varicose veins in the gullet:

- Cirrhosis can cause the blood vessels around the esophagus to burst and bleed. To prevent that from happening, doctors can either prescribe medicines or do a gastroscopy to place rubber bands around the varicose veins.

3. Decrease the fluid build-up in the belly:

- By placing you on a "low-salt" diet, prescribing medicines that help kidneys pee out the extra fluid (diuretics), draining the fluid from your belly (paracentesis), or inserting a tube through the liver, which reduces the pressure within the scarred liver and prevents the formation of fluid in the belly.

4. Treat or prevent infection of the fluid in the belly:

- Start you on a low-dose of antibiotics to prevent infection, give you the annual flu vaccine, or admit you to hospital for intravenous antibiotics if you get a fever.

5. Improve confusion:

- Start you on twice-daily lactulose syrup to ensure you have two or three soft bowel motions each day, which will prevent the build-up of toxins in your bowel.
- Start you on a special antibiotic called rifaximin. This antibiotic kills the bugs in the bowel, which make the toxins that cause confusion.

6. Liver Transplantation:

- If your liver failure is too advanced to recover with the above treatments or if you have a liver cancer, your doctor may suggest you are considered for liver transplantation. This is a routine treatment with more than 20,000 liver transplants performed every year for liver failure or liver cancer. Outcomes are excellent with more than 90 per cent of people surviving five to 10 years after the operation. If your doctor thinks that you are sick enough to be considered for transplant, you will need to travel to the NZ Liver Transplant Unit in Auckland City Hospital for one week to undergo assessment.

Auckland Bleed Reporting Project

By Ian d'Young

Introduction

Since 2009 the Auckland Regional Haemophilia Treatment Centre (ARHTC) has embarked on a number of performance improvement projects designed to improve the access and delivery of services to our patients and to better and more accountably manage limited financial resources. This work has resulted in significant improvements in the quality of services we are able to offer patients and is associated with a high degree of patient satisfaction.

By carefully analysing our data, however, we were aware that there were still significant opportunities for doing things better. Many adult patients diagnosed with moderate and severe haemophilia living in the Auckland region did not regularly report bleeding episodes within a timeframe that allowed for early intervention, optimal assessment and management. Patients experienced more pain, disability and delayed return to work than was necessary, an increased risk of permanent joint damage and a greater risk of further bleeding episodes at or related to the affected site. Delayed bleed reporting also reduced clinical oversight of the use of expensive clotting factor concentrates and increased the likelihood that factor would be used inappropriately or in greater quantities than was strictly necessary.

Haemophilia treaters in New Zealand are accountable to the National Haemophilia Management Group (NHMG). Cost and volume data on the use of clotting factor concentrates is closely monitored. In a fiscally constrained health environment, poor oversight of the use of clotting factors is a threat to the sustainability of the high standard of care we are currently able to offer New Zealanders with haemophilia.

The future treatment paradigm, for economic and effectiveness reasons, needs to switch from near complete patient autonomy and self-management largely independent of the specialist resources available through the haemophilia treatment centre to a care partnership with patients. With the ageing haemophilia population it is no longer appropriate to regard every symptom as a bleed until proven otherwise. This is especially true of joint pain due to degenerative disease, for which factor concentrate administration is ineffective, wasteful and inappropriate. Rapid reporting and assessment of bleeding episodes and musculoskeletal pain is therefore key to minimising waste in the Haemophilia treatment setting.

Methods

We identified all adult patients (>16 years) living in our catchment area with phenotypically moderate and severe Haemophilia A and B (n = 70). We reviewed patient records and our databases. Based on annual factor consumption we determined that fewer than 50% of the predicted bleeding episodes over the 2011/12 financial year were reported. Of those that were more than 70% were reported outside of a 48-hour time window. Mean time to report a bleeding episode, excluding significant outliers, was in excess of 54 hours.

We then asked adult patients, spouses and family members to recount stories of their experience of our service through patient

forums, focus groups and interviews. We needed to understand the barriers to prompt bleed reporting from our patient's perspective and work with them to develop sustainable solutions. Some of the key barriers our patients identified included:

1. **Expectations and Value.** Patients want clear communication in easy to understand language, outlining the clinical benefits of prompt bleed reporting and the importance of this information for optimally managing product use. They clearly understand the relationship between financial accountability and the long-term sustainability of treatment services in New Zealand. The feedback indicated that we need to repeat this message regularly. Patients indicated a willingness to be involved in developing the communication, content and strategy. How the message is delivered is important. While patients value the support of clinicians, they also feel strongly that their peers have a strong role to play in supporting and educating each other.

   Many of our men aged in their 30s and 40s required reengagement with the treating centre. This age group was involved in the transition from a lifetime of hospital-based services to achieving a far greater degree of independence with the advent of a home supply of recombinant clotting factors. These individuals have negative perceptions about hospitals and perceive increased bleed reporting and engagement as a threat to their independence. Those interviewed also describe a perceived blame culture with bleed reporting. They are keenly aware of the cost of treatment, and reported feelings of guilt associated with the use of clotting factor concentrates. They felt that if they reported each bleeding episode there might be negative consequences, specifically a threat to their home supply of product and therefore their independence. The concept of reporting without blame is therefore critical to patient acceptance of any process change.

2. **Systems.** Detailed process mapping indicated that we need to benchmark, automate and standardise a number of our systems, including our bleed reporting database and product order calculator. Patients emphasised the need for a consistent, documented pathway for managing their compliance, and indicated a willingness to be involved in developing a solution to this issue.

3. **Distance.** As the Auckland Regional Haemophilia Centre covers a large geographic area we predicted that relying on local services or needing to travel significant distances to the centre would negatively influence the likelihood that a bleed would be reported. There were stories of long waits in rural emergency departments and limited knowledge of haemophilia as a condition by local clinicians. Difficulties were described in getting appointments with general practitioners, and travelling several hours in a car to the HTC in Auckland was an option of last resort, reserved for the most severe of bleeding episodes.

4. **Technology.** Improved access and promotion of new technology such as our iPhone app and website was regarded as necessary to facilitate easier patient communication with us. We need to make smart phone technology more available to patients and need to have both iPhone and Android-platform Apps available. In order to see value in prompt reporting, we need to maintain an appropriate speed of response policy for acknowledging patient contact and arranging assessment appointments. The patient group also identified issues associated with previous reporting strategies for prophylaxis (paper forms, spreadsheets) and made a number of suggestions on how this process could be improved.

Results

We worked with patients to develop a range of solutions to the issues that were raised. Since the implementation of our solutions in March 2013 mean reporting time from bleed onset to clinician contact improved from 56 to fewer than 15 hours. The mean number of bleeds reported each month increased from 8 to 27. All improvements were significant ($p < 0.05$). Twice as many patients are now using smart phone/ website technology to report bleeding episodes (from 25% to 58% of the project population). Faster bleed reporting means faster access to services for patients, better clinical outcomes and better accountability for clinicians to our national funder. We also noticed that our patients used less clotting factor concentrates (around 15% year-on-year) when there was faster reporting and access to services.

Conclusion

Accountability is essential for sustaining the high standard of care people with haemophilia have access to in New Zealand. We learnt that in order to make meaningful changes we needed to learn from our patients and involve them in every stage of a system redesign process. We all have a role to play in sustaining and developing services for PWH in New Zealand. Patients who have a sense of ownership and engagement with a service, who are active rather than passive participants, experience greater levels of satisfaction with services and improved health outcomes.

Ian d'Young is the former Haemophilia Physiotherapy Practitioner for the Auckland region and national clinical lead. He resigned from this role in December 2013 and now holds a management role within the performance improvement team at the Auckland District Health Board.

Congratulations to Ian d'Young, Laura Young, Mary Brasser, Karen Slavin, Rachel Donegan, Leigh Manson, Paul Ockleford and HFNZ's Sarah Preston, winners of the Chief Executive's Award 2013 for their project on improving timely bleed reporting by adults with haemophilia.

The Auckland District Health Board Healthcare Excellence Awards are an opportunity to showcase excellence and innovation.

They are aimed at those who are publishing cutting-edge healthcare research, enhancing education in healthcare, developing new and improved ways of delivering clinical and non-clinical services or finding ways that enhance the health and wellbeing of Auckland DHB patients and community.

HFNZ offer our sincere congratulations to the project team. It is work like this that truly makes a difference in the care of haemophilia and in our community.

Planning for the next generation

Chances are now or sometime in the future you might begin to think about having children. The challenges for both men and women who have the gene for haemophilia can rise significantly when the possibility of having a child enters the picture. Here are some things you may want to consider when thinking about family planning.

Know your genetics

Understanding the genetics of your bleeding disorder will help you and your spouse or partner determine the best course of action. When thinking about having children, you'll probably need to consider not only the general responsibilities of parenthood but also how your chronic health condition will affect your family and what it will be like to raise a child with a chronic health condition.

Having a child with a chronic health condition such as a bleeding disorder is a shift in what you might have thought to be your normal life, but it can be just as fulfilling as having a child without a chronic condition. You and your partner should discuss the possibility of passing a bleeding disorder on to your children.

It is recommended that people with bleeding disorders and 'carriers' receive formal education about the transmission of haemophilia or related bleeding disorders within their family before starting a pregnancy, if at all possible. If you and/or your partner have a bleeding disorder, talk to the staff at your closest Haemophilia Treatment Centre. They are experts in bleeding disorders and how they are transmitted. They might recommended to having factor levels checked and genetic testing done to identify the genetic mutation for the bleeding disorder that runs in your family. Specialist genetic advice and counselling are also available in many hospitals to inform families of the genetic implications of a diagnosis of haemophilia or related bleeding disorder and to allow families to make informed choices regarding the reproductive options available to them. Genetic counselling sessions can provide families a safe place and designated time to talk. Contact your Haemophilia Treatment Centre for more information.

Haemophilia: The Genetics

Haemophilia is a hereditary bleeding disorder. In terms of genetics, haemophilia is known as a sex-linked disorder because the genes responsible for producing Factor VIII and IX are situated on X chromosome. Here are the possibilities for passing on the haemophilia gene:

- If the mother is a haemophilia carrier and the father does not have haemophilia, there is a 50% (1 in 2) chance that each son or daughter will be a carrier of the haemophilia gene.
- If the father has haemophilia and the mother is not a carrier, sons will not be born with haemophilia, but all daughters will be carriers.
- If the father has haemophilia AND the mother is a carrier, there are four possible outcomes, each with a 25% (1 in 4) chance of occurrence:
  - The child could be a son without haemophilia.
  - The child could be a son with haemophilia.
  - The child could be a daughter with haemophilia.
  - The child could be a daughter who is a carrier.

The Options for Having Children

There are many ways to create a family. Having a bleeding disorder doesn't mean you must forego the joy of having a family. If you are concerned about the chances of having a child who has a bleeding disorder, you may want to consider the following:

For the woman who is a carrier/has a bleeding disorder:

- You may choose not to have your own biological children. Instead, you may choose non-biological options, such as adoption or foster parenting.
- You may choose to try to conceive naturally. You can then choose whether to have tests performed early in a natural pregnancy, such as chorionic villus sampling (CVS) or amniocentesis, to determine if the foetus carries the gene for haemophilia. You can then consider whether to continue or terminate your pregnancy. Check with your doctor for the best time to do these tests.
- You can choose to try to conceive using an assisted reproductive technology method. For example, you can try in vitro fertilization (IVF), using either your own egg(s) or donor egg(s). If you use donor eggs, any child born will not carry the haemophilia gene, assuming the donor is not a carrier of haemophilia.
- If you use your own eggs, then during the IVF procedure, pre-implantation genetic diagnosis (PGD) testing can be performed to determine the presence or absence of the haemophilia gene and the sex of the embryo. This knowledge can be used to choose an embryo without the haemophilia gene to minimise the risk of having a child with a bleeding disorder. Because haemophilia is considered a serious and potential life-threatening condition public funding is available in New Zealand for up to two rounds of IVF with PGD. There is currently a waiting list of approximately 2 years for this service so if you are considering this option it would be best to consult a specialist well in advance and not wait until you are ready to try to conceive.

For the man who has a bleeding disorder:

- You may choose not to have your own biological children. Instead, you may choose non-biological options, such as adoption or foster parenting.
- You may choose to use artificial insemination with donor sperm. If the mother is not a haemophilia carrier, then any child born will not have the haemophilia gene, assuming the donor does not have haemophilia.
- You may choose to try to conceive naturally. Male children will not be affected but all female children will be carriers of haemophilia.
- If the mother has a fertility problem, you may consider trying IVF using donor sperm from a donor who does not have haemophilia. Any foetuses conceived in this manner will not carry the haemophilia gene.

Women and Bleeding Disorders

If you have a bleeding disorder or carry the gene for haemophilia, there are a few additional things you will want to think about when considering sexual relationships and starting a family. Menstruation and pregnancy can sometimes be hard on the body, and even more so if your clotting factor levels are low. This should not stop you from doing the things you want to do in life; you'll just need to be aware of how to best care for yourself.

If you have a bleeding disorder and you and your spouse or partner decide to have your own biological children, you should be aware of how your pregnancy may be affected. Women with bleeding disorders can have successful pregnancies and healthy babies. If you have been diagnosed with a bleeding disorder or if you are a carrier of a gene for a bleeding disorder such as haemophilia, start preparing for your pregnancy before you become pregnant. Being prepared and having the right healthcare team will help ensure that you get the proper care throughout your pregnancy.

During Pregnancy

If you have a bleeding disorder and you are pregnant or plan to become pregnant, have a general health screening about 3-6 months prior to becoming pregnant or early in your pregnancy. This is also a good time to speak with your Haemophilia Treatment Centre and find an obstetrician/gynaecologist and midwife who has experience providing reproductive care for women with bleeding disorders. It is recommended that your pregnancy health team should include a midwife, an obstetrician and a haematologist who all have experience assisting women with bleeding disorders through the pregnancy, birth and aftercare experience.

Know that your clotting factor level will determine your risk of bleeding complications during childbirth. To prepare for potential problems, both you and your medical team should be aware of your carrier status and clotting factor levels. If you are a carrier for haemophilia, have your factor VIII level determined during the third trimester. If you have VWD, have blood tests done in the third trimester to determine levels of von Willebrand factor and factor VIII. This will help your doctors determine if you will need clotting factor or other medications to prevent bleeding during labour and delivery. It is extremely important that the Haemophilia Treatment Centre writes a Protocol for treating you in case of a haemorrhage and Protocols around the delivery of the baby, especially if it might be a boy with haemophilia, which might include recommendations for things such length of pushing time, use of forceps and/or ventouse, and factor replacement for the mother should a Caesarean section become necessary.

Delivery and Birth

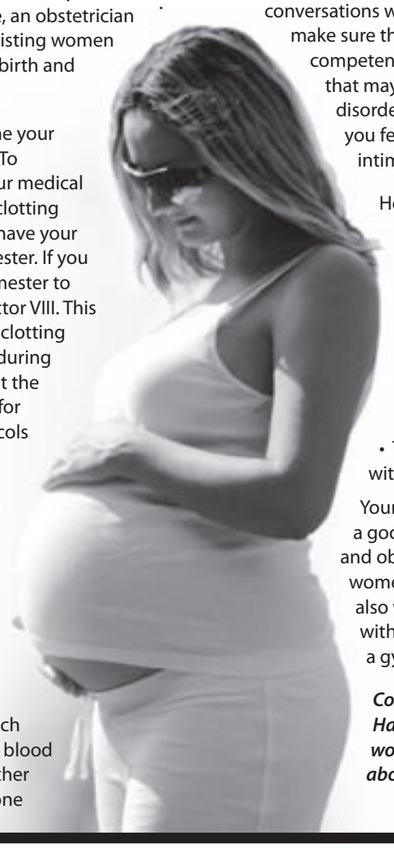
Special testing for haemophilia can be planned before the baby's delivery so that a sample of blood can be drawn from the umbilical cord (which connects the mother and baby before birth). The blood sample can be tested to find out right away whether the baby has haemophilia. Testing can also be done on blood drawn from a vein soon after a male baby is born. It is important to know as soon as possible after birth whether a baby has haemophilia so that special care can be given to prevent bleeding complications.

Women who carry the gene for haemophilia and those with VWD are at risk of serious bleeding after delivering the baby. During pregnancy the body naturally makes higher amounts of factor VIII and VWF; these factors drop back to lower levels after delivery. If a woman has low levels of clotting factor after delivery or Caesarean section, she may experience severe bleeding that lasts a long time. This is called postpartum haemorrhage and is another reason to deliver in a facility affiliated with a Haemophilia Treatment Centre. All women should pay attention for possible bleeding up to a few weeks after delivery and have a plan in place to address heavy bleeding.

If you are considering circumcision for the baby and have bleeding disorders in your family history, you should wait until the test result is provided to determine the circumcision procedure. About half of newborns with haemophilia will develop excessive bleeding following circumcision. Far fewer newborns with haemophilia will sustain a skull bleed, called intracranial haemorrhage, due to squeezing of the head during passage through the birth canal. Look for signs of bleeds, such as swelling and discoloration, so they can be stopped quickly.

Choosing Your Gynaecologist

At some point, most women end up having sensitive conversations with their gynaecologist. You want to make sure that the doctor you choose is not only competent and can address any complications that may arise because of your bleeding disorder but also is someone with whom you feel comfortable asking even the most intimate questions.



Here are some considerations when choosing a gynaecologist:

- How long the gynaecologist has been in practice.
- Where the gynaecologist received training.
- If he or she has cared for other women with bleeding disorders.
- The tests they recommend for someone with your medical history.

Your Haemophilia Treatment Centre may be a good resource for finding gynaecologists and obstetricians with experience caring for women with bleeding disorders. You may also wish to ask other women you know with a bleeding disorder to recommend a gynaecologist.

Contact your HFNZ Outreach Worker or Haemophilia Treatment Centre if you would like to talk to someone further about family planning.

Twinning Connection

Two of the main goals of the HFNZ- Cambodian Hemophilia Association (CHA) Twinning programme was to help CHA operate effectively as an organisation and for CHA to be capable of running educational and social activities for people with haemophilia and their families. Well,3 years on we can report that CHA is succeeding with flying colours on both these counts.

For example, the CHA committee have been busy planning many events for 2014. They have family visits, group meals, and youth events all scheduled, as well as a special awareness event planned for World Haemophilia Day on 17 April. One thing they do at their meetings is go around and discuss how everyone is doing with regards to their bleeding disorder so they can understand and support each better. This sounds like a good idea you might want to adopt at your next committee meeting.

Mr Sem Sokpanha (known as Panha), CHA Vice-President has been chosen to attend the NMO Training and the 2014 WFH World Congress in Melbourne, as well as the General Assembly Meeting on the Friday after the Congress on behalf of CHA through WFH support. In addition, Mr Run Chanthearithy, known as Rithy, has been awarded a 2014 WFH Youth Fellowship Program and so will also be sponsored by WFH to attend the NMO training and WFH World Congress in Melbourne in May. We congratulate Rithy and I am sure many members will look forward to meeting him and Panha at the Congress.

Our Pharmaceutical Friends

Among our valued donors are the companies who manufacture treatment products for haemophilia available in New Zealand. These pharmaceutical companies provide a generous amount of ongoing non-directed funding that goes to support all HFNZ programmes and special donations towards specific educations workshops or meetings.

HFNZ is sincerely appreciative of the commitment of current Sustaining Patrons and other pharmaceutical friends to improving the lives of people with haemophilia and other genetic bleeding disorders, and for the generous additional support they provide HFNZ to help us run educational workshops.

THANK YOU!

Bayer HealthCare
Bayer Schering Pharma

CSL Bioplasma

Pfizer

Baxter

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Upcoming HFNZ Events

HFNZ have many great national events planned for 2014 and beyond! Mark your calendars and keep an eye out for invites to the following plus all the great social activities planned by your local branch.

If you have any questions on any of the events or want to express your interest in attending please contact your local Outreach Worker.

World Haemophilia Day

17 April 2014

This year's World Haemophilia Day will focus on encouraging the global bleeding disorders community to Speak out. Create change. Follow the World Federation of Hemophilia's (WFH) Facebook page, like and share the infographics WFH will be posting leading up to World Haemophilia Day. You can follow the latest global developments on Twitter #WorldHemoDay.

You can also participate in the 1 in 1000 World Haemophilia Day online photo competition and encourage members of your community to do the same.

Don't forget to keep an eye out for local events to celebrate to World Haemophilia Day here in New Zealand.

National Inhibitors Workshop

11-13 July 2014 – Auckland

Inhibitors are a serious medical problem that can occur when a person with haemophilia has an immune reaction to treatment with clotting factor concentrates and is considered one of the biggest challenges in haemophilia care today. This educational workshop will focus specifically on the challenges faced by families with inhibitors and is open to people with inhibitors of all ages and their immediate families. Keep an eye out for your invitation or contact your Outreach Worker for more information.

Parents Empowering Parents (PEP) Programme

1-3 August 2014 – Auckland

PEP is an innovative, peer-to-peer skills programme that introduces parents to tools to better handle the realities of raising a child with a bleeding disorder. PEP programmes are led by an outreach worker, parents and a haemophilia nurse who use classroom discussions, role playing and hands-on experiences to educate parents about the types of skills needed for effective parenting. We have had rave reviews from parents who have participated in previous programmes.

PARENTS EMPOWERING PARENTS

Youth Leadership Weekend

8-10 August 2014 – Christchurch

You've been to the camps, you've helped out at workshops and maybe you've even joined the National Youth Committee. But now is the time to really take a stand. Join HFNZ for a weekend all about you and how you can become a leader in our bleeding disorders community. We'll help you find the tolls and decide on ways you can help the next generation. If you think you can demonstrate your leadership potential within HFNZ check the website for application forms or contact your Outreach Worker (available soon).

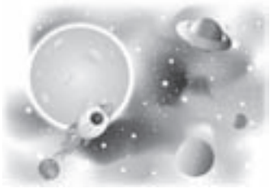
HFNZ National Annual General Meeting

Saturday 13 September 2014 – Auckland

This year's Annual General Meeting is taking place at in Auckland. Because of the change of our financial year to end in June, the National Annual General Meeting will now take place in September. Further details and a copy of the agenda will be available online before the meeting. See the listing on the Events webpage later in the year.

National Families Camp

23-26 January 2015 – Camp Keswick, Rotorua



Held bi-annually, National Families Camps bring together young families affected by haemophilia from all over New Zealand. Parents gain strength through knowledge and understanding, and look to each other for friendship and support. For many children, camp is the first time that they have been around other children and adults with haemophilia. This camaraderie provides campers with a sense of relief that they are not the only ones with haemophilia and with an opportunity to share with others. Children learn about their disorder and enjoy having fun whilst being "just like everyone else". The theme for the 2015 National Family Camp will be Outer Space and the camp will be out of this world!

MRG Reports

HFNZ operates six groups that represent our members, four regional branches (Northern, Mildand, Central and Southern), the National Youth Committee (NYC) and the Piri Toto, which represents Māori members.

Central Region

By Stephanie Coulman

The children's workshop in Palmerston North on 1 February was attended by just a few of our youngsters including Anna and Nicholas Coulman, Isla Brodie and Isaac Nyhan, D'vontae Mackie, Benjamin Mackie, Matia Fricker, Charlize Hanson. Nicholas has started self-infusing which is great breakthrough considering until late last year, he was saying he would 'never do his own factor and that his mother would do it for him and when she died, he would hire someone else.'

For some haemophilia is not the only thing going on in our lives. This is especially so for the Brodies. Since August last year their household has been in upheaval. Three-year old Flynn not only has severe haemophilia, he also has an ultra-rare skin disorder which has caused very serious upper respiratory issues. He has had to have the 'plaques' in his throat trimmed – only the fifth person in the world to have this done.

The Wellington-based family has been in and out of Starship Hospital in August, September, a day stay in October where they thought he was 95% well and realised he was breathing through a pin-hole, and twice in January and again in February for more surgery.

"He should hopefully grow out of these breathing issues but it hasn't been easy. Having haemophilia on top hasn't really been a 'factor' only to make sure the surgeons/anaesthetists follow the correct procedure and don't take extra blood tests," says Jo.

Central region Christmas lunch at Zealandia, Wellington

[Photo courtesy of Fairfax Media]

Jo and Flynn Brodie who, as well as haemophilia, has a rare skin disorder causing serious upper respiratory issues.

2014
HFNZ Inhibitors Workshop
11-13 July
- Auckland -

Open to people with inhibitors and their immediate families.

Register Today
and get ready to take a
STARRING ROLE!

Southern

By James Poff

Hi and welcome to the first Southern MRG Report for 2014.

I hope everyone in the HFNZ family had a great Christmas and New Year including having the opportunity to get away, put your feet up, read a book or relax in some way!

Following on from the Southern 30th celebrations it has been a bit of a quiet period over the region to date; however, there was some exciting activities enjoyed by our members.

Our Christchurch Christmas party was held on the 8th of December, there was a plenty of things to do including a Magician, Bouncy Castle and of course Santa made an appearance.

Harrison Stott proved that even a kid with Haemophilia can survive an encounter with a Magician's guillotine.

Santa "loaned" two of his helpers to operated the BBQ and read a story to the children towards the end of the day.

The weather was perfect for the event, amazingly the forecasted rain held off right until the end of the day, in fact it did not start until the Bouncy Castle was been loaded on the truck to be taken away – brilliant.

On 21 January children from the Southern region with bleeding disorders aged between 6 to 10, including their siblings and children with a parent with a bleeding disorder attended a Children's Education workshop here in Christchurch.

What did the participants think: Well when I picked up my two, I asked Maisie Poff "Did you have a good day?". She replied,"Nope – I had a fantastic good day".

Recently the annual Canterbury Jet Boating club held an open day for community groups, we enjoyed an exhilarating ride up and down the Waimakariri River – next time anyone is talking to Rorie Poff make sure you ask him about what happens when a jet boat gets stuck.

Upcoming events:
The Southern MRG is meeting on Sunday 16th February 2 pm in the meeting room at the MacDonald's on the corner of Gasson St and Washington Way. Part of this meeting will be to hold a planning session for the Southern MRG membership including Otago/Southland.
Again thanks to everyone's hard work in making the Southern MRG events success for our members.

Southern Christmas Party

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

News in Brief

Prophylaxis Tops On Demand Dosing for Haemophilia B

Once or twice weekly prophylactic dosing of recombinant factor IX significantly reduced yearly bleeding rates in patients with haemophilia B in a randomised controlled trial. The study represents the first such effort in more than 30 years to compare prophylaxis with on-demand treatment in patients with haemophilia.

The investigators at the Rush Hemophilia and Thrombophilia Center in Chicago conducted a phase 4 randomised open-label crossover study to evaluate the efficacy and safety of recombinant factor IX nonacog alfa (BeneFIX, Pfizer) as a prophylaxis compared with on-demand administration between May 2007 and October 2010. Pfizer sponsored the study.

Of 47 males aged 6 to 65 years who had moderately severe to severe disease at the start of the study, 41 completed the trial. All participants had 16 weeks of on-demand therapy at the beginning of the trial and then were randomly assigned to receive either 100 IU/kg once weekly or 50 IU/kg twice weekly for 16 weeks. All patients then switched back to on-demand dosing for 8 weeks, followed by 16 weeks of prophylaxis with the dose they had not received during the first 16-week prophylaxis period.

On-demand treatment was allowed during prophylaxis periods if needed. The authors calculated the annual bleeding rate (ABR) as the number of bleeding events divided by the number of days receiving treatment divided by 365.25 days a year.

The researchers found that both prophylaxis regimens significantly reduced ABR compared with on-demand treatment (mean ABR: on demand, 35.1; 100 IU/kg, 4.6; 50 IU/kg, 2.6; $P < .00001$). There was no significant difference between the two prophylactic regimens ($P = .22$).

Most joint and soft-tissue bleeding events resolved with a single infusion, whereas five joint bleeding episodes required more than 4 infusions. One limitation of the study is the relatively short duration, the researchers write.

"The decreased frequency of infusions with once-weekly dosing may present a favourable option for individuals in whom venous access is a concern, potentially improving adherence and offering more convenience to patients and their caregivers compared with more frequent prophylactic dosing regimens," the authors conclude.

Source: Haemophilia. Published online January 13, 2014. DOI: 10.1111/hae.12344

Long-Acting Recombinant Factor VIII Demonstrated Prophylaxis with Less Frequent Infusions in Haemophilia A in Phase III Trial

Bayer HealthCare have announced positive results from the PROTECT VIII trial evaluating the company's investigational long-acting site-specific PEGylated recombinant human factor VIII compound BAY 94-9027. The study met its primary objective of protection from bleeds with fewer infusions. In the study, the site-specific PEGylated factor VIII helped protect against bleeds when used prophylactically every seven days, every five days, and twice per week. The compound was also effective for treatment of acute and breakthrough bleeds with 91 percent of events resolving with one or two infusions.

PROTECT VIII (PROphylaxis in hemophilia A patienTs via directly pEgylated long-aCTing rFVIII) is a multicentre, multinational, partially randomized, open-label trial with four treatment arms evaluating the safety and efficacy of the site-specific PEGylated factor VIII in previously treated adults and adolescents with severe haemophilia A. 134 patients were treated in the study.

It has been reported that it may be another year before the drug is filed for regulatory approval.

Source: http://www.digitaljournal.com/pr/1741536

US Experts Update their Hepatitis C Guidelines

Due to the multitude of new direct-acting antivirals (DAAs) for the treatment of hepatitis C expected to reach the market in the next few years The American Association for the Study of Liver Diseases (AASLD), the Infectious Diseases Society of America (IDSA) and the International Antiviral Society (IAS)–USA have teamed up to create hepatitis C treatment guidance, www.HCVguidelines.org , that will be kept current to help keep clinicians up-to-date on the management of hepatitis C virus.

Of note the new guidelines no longer endorse pegylated-interferon and ribavirin with or without boceprevir or telpaprevir as a treatment option; instead they recommend:

Treatment category	Recommended treatment
Genotype 1: Treatment-naive patients	• Eligible for interferon: SOF + PEG/RBV x 12 weeks • Ineligible for interferon: SOF + SMV ± RBV x 12 weeks
Genotype 1: Retreatment (previous PEG/RBV has failed)	• SOF + SMV ± RBV x 12 weeks
Genotype 2: Treatment-naive patients	• SOF + RBV x 12 weeks
Genotype 2: Retreatment	• SOF + RBV x 12 weeks
Genotype 3: Treatment-naive patients	• SOF + RBV x 24 weeks
Genotype 3: Retreatment	• SOF + RBV x 24 weeks
Genotype 4: Treatment-naive patients	• Eligible for interferon: SOF + PEG/RBV x 12 weeks • Ineligible for interferon: SOF + RBV x 24 weeks
Genotype 4: Retreatment	• SOF x 12 weeks + PEG/RBV 12 weeks; or • SOF + RBV x 24 weeks
Abbreviations: SOF: Sofosbuvir; PEG: Pegylated-Interferon; RBV: Ribavirin; SMV: Simeprevir	

Blood Vibrations: Community Music Project

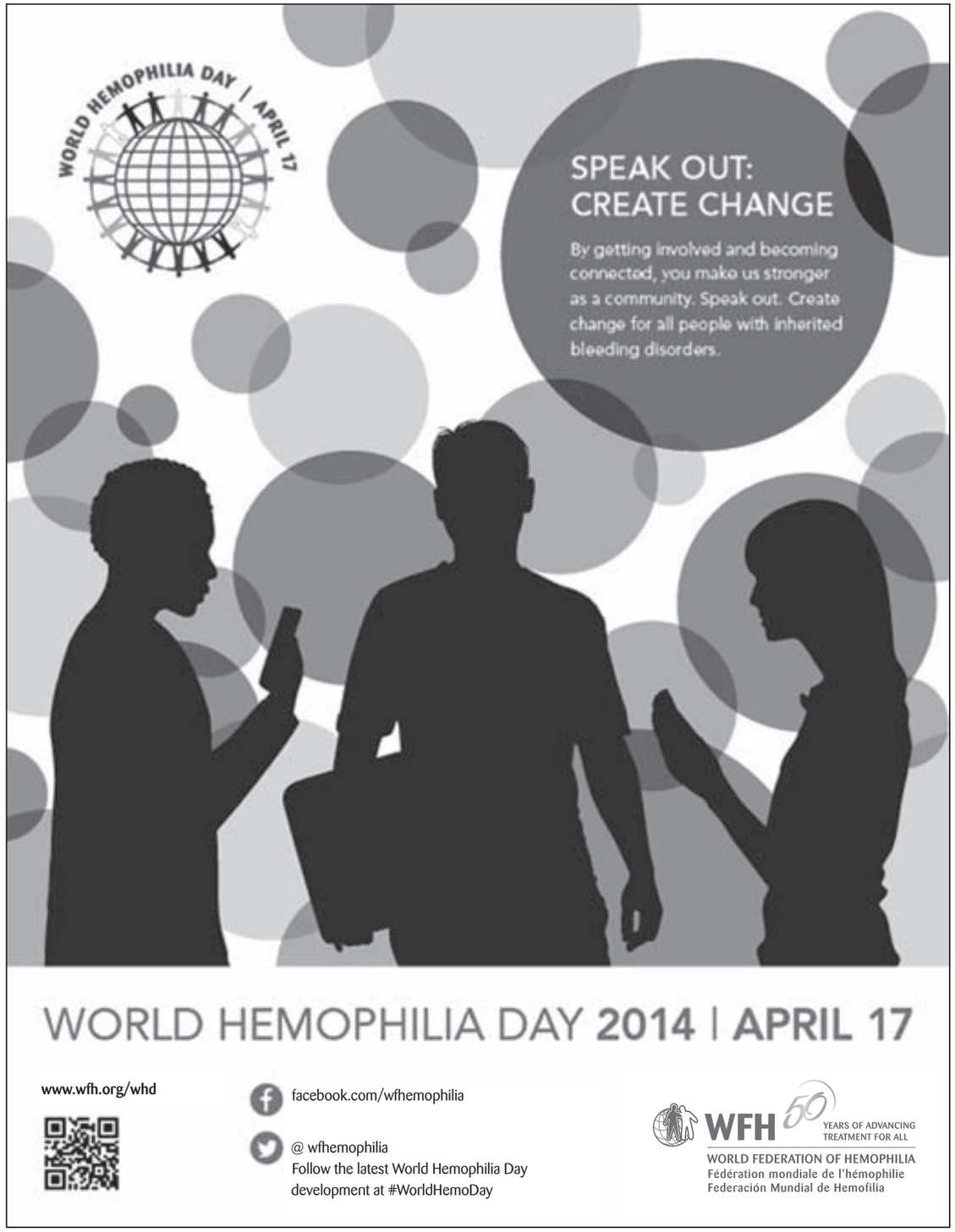
Two guys with haemophilia in the USA, Billy Conde Goldman and Matt Tache recently launched Blood Vibrations, an ongoing project gathering music created by people in the bleeding disorders community. The goal of the project is to provide a forum for creativity, expression and sharing. Anyone with a bleeding disorder (boys and girls, men and women) is welcome to participate in this project. No musical experience required. Blood Vibrations encourages collaborations and group works. Experience the music shared to date and share your own music at www.bloodvibrations.wordpress.com

The Hepatitis C Treatment Outcome Study

If you are considering starting treatment for hepatitis C please consider taking part in The Hepatitis C Treatment Outcome Study being conducted at Bond University, Australia.

The study is open to individuals preparing for hepatitis C treatment and participants are being sought from Australia or New Zealand. Information provided is completely confidential and you have the right to withdraw from the study at any time.

For more information visit http://hcvstudy.bond.edu.au



Upcoming Events

17 April 2014
World Haemophilia Day

11-15 May 2014
WFH World Congress
Melbourne, Australia
www.wfh.org

11-13 July 2014
National Inhibitors Workshop
Auckland

1-3 August 2014
Parents Empowering Parents
(PEP) Programme
Auckland

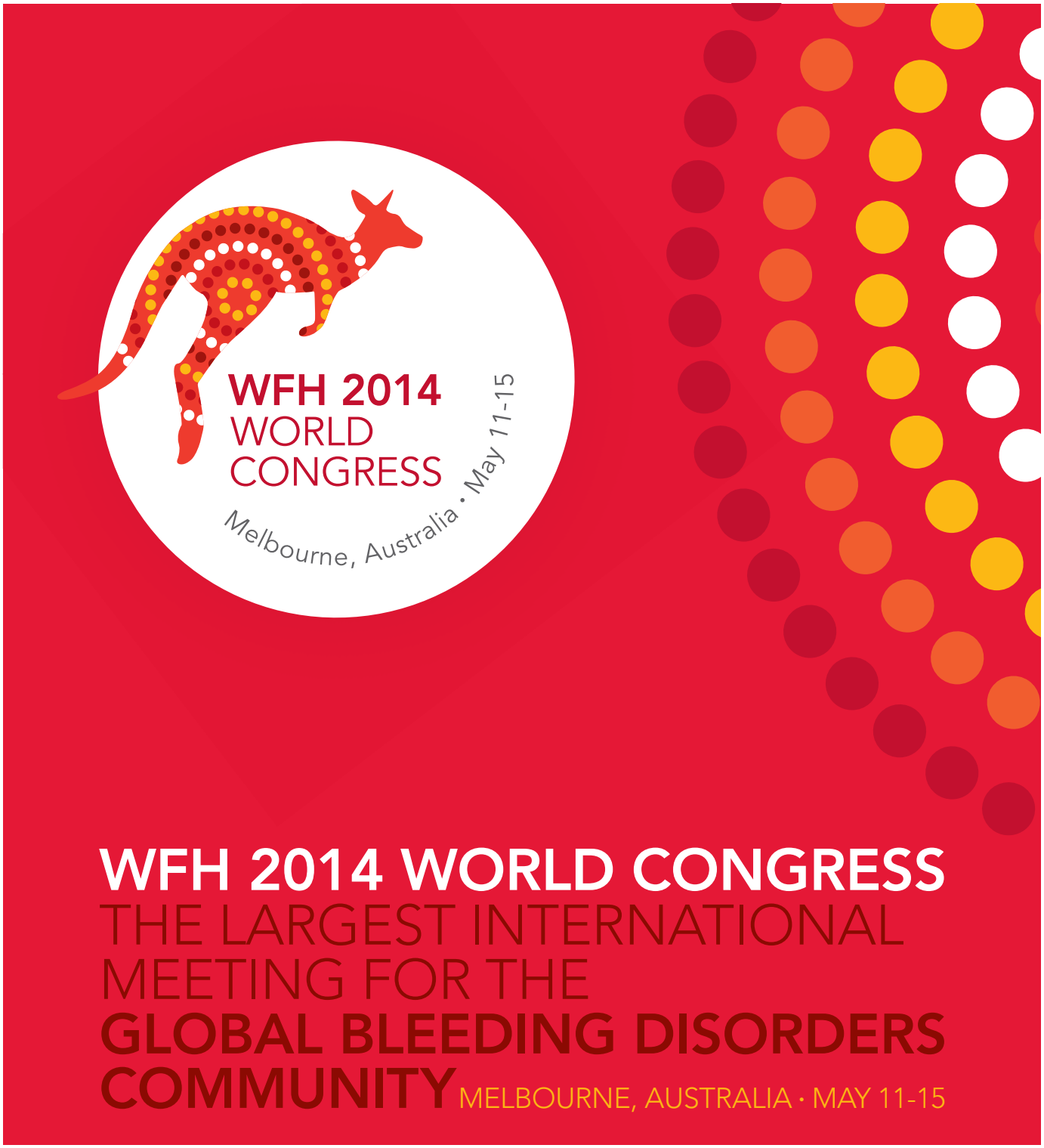
8-10 August 2014
Youth Leadership Weekend
Christchurch

13 September 2014
HFNZ National Annual General Meeting
Auckland

23-26 January 2015
National Families Camp
Camp Keswick, Rotorua

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

Full conference coverage in the June issue of Bloodline



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

Jeff Oliver Print - Whangarei