CF News

ISSN 0114-3573 P
ISSN 1179-044X O

SPRING 2021

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CYSTIC FIBROSIS NZ

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ISSN 0114-3573 (Print) – ISSN 1179-044X (Online)

Registered Charity CC10274. Registered office:
64 Grafton Road, Grafton, Auckland.

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Welcome to the CF News Magazine

Kia Ora reader,

Thank you so much for choosing to read the Spring 2021 issue of the CF News magazine!

We are incredibly excited to share interviews with each of the four fantastic 2021 CF Achievers' Award recipients (page 6) and an interview with the wonderful Emma Gawn, the recipient of the 2021 Mark Ashford Scholarship (page 8). It is always such a pleasure to speak to these inspirational young adults and share their story with you.

Since the Autumn 2021 CF News magazine issue we haven't taken our foot off the campaign for Trikafta pedal. High profile news coverage has continued, and major milestones reached such as Vertex submitting application to Pharmac for funding of Trikafta, plus the establishment of a Managed Access Programme in NZ for those in critical need. But we won't stop until Trikafta is funded publicly for every person who is eligible. Read the full update on how the campaign for Trikafta is going on page 23.

The sweatiest month of the year is here…. have you signed up yet?! Last year the Sweatember fundraiser (sweatember.org.nz) raised an incredible $140,000 for CF support and advocacy services, and we're hoping supporters can help "sweat it" to the next level this year! Turn to page 10 to find out more about Sweatember and how to make your sweat count.

Lastly, with rising costs of printing and postage - and being keen to save a few trees too – we'd love for you to join the digital revolution and opt to receive the CF News magazine digitally. Visit www.cfnz.org.nz/sign-up to change your subscription preferences.

Thanks again – and happy reading!

The CF News Team

John Ilott Charitable Trust

perpetual guardian

Thank You

A huge thank you to John Ilott Charitable Trust and the Grumitt Sisters Charitable Trust, proudly managed by Perpetual Guardian, for once again funding this issue of the CF News magazine.

REGULARS

News In Brief

Farewell CFNZ Chief Executive, Jane Bollard

After nearly six years at the helm of Cystic Fibrosis NZ, Jane Bollard has sadly resigned from her position as Chief Executive. Under Jane's leadership we have seen huge waves made in advocacy, CFNZ has grown professionally and grown in strength, enabling the organisation to continue to provide the support to those in need across the country. We can proudly say that the level of public awareness of CF is better than it has ever been.

"The public funding of Trikafta will herald a change in health care for people with CF and a changing role for CFNZ. The organisation is well placed to lead this change bringing us ever closer to lives unlimited for those with CF.

As I step away from my role as Chief Executive, I want to thank people with CF and their families and whanau, donor and supporters, board members and active branch members, staff, volunteers and clinicians for allowing me to be part of your journey. I wish you the best for the years ahead," says Jane.

"The organisation's priorities will not be changing, so the focus will be on finding someone committed to the cause, a strong leader and a great communicator and advocate. We will be doing everything we can to get the right person," says CFNZ Board Chair, Warwick Murray.

We thank Jane for all the time, energy, and passion she has put into CFNZ and wish her all the best for the future.

Christine Perrins, who has generously been volunteering her time as our advocacy advisor for 18 months, has agreed to take on the role of Acting Chief Executive. Christine has had a distinguished career in central and local government as a senior manager and has had heaps of experience in leading teams in complex and sensitive environments. So CFNZ will be in great hands while we recruit our new Chief Executive.

NZ gives a Future for Kiwis with CF

CFNZ took over May with the CF Month "Give a Future" appeal, shining a light on life with CF, access to medications, daily treatment and more. Thanks to the wonderful CFers and their families who put their hand up to be interviewed we managed a phenomenal amount of TV, radio, newspaper, and online coverage, helping to raise awareness and educate the NZ public about CF. You can view the coverage at www.giveafuture.org.nz.

The NZ public responded by donating over $45,000 via the CFNZ website and 'Give a Future' appeal page. T-shirt sales and community fundraisers raised a further $15,000 and continues to grow. Thanks to all involved, bring on May 2022!

Trikafta Managed Access Programme in NZ

At the beginning of July, we were thrilled to confirm that Vertex had put in place a Manage Access Programme in NZ for the triple combination therapy (registered as Trikafta in Australia). The programme can provide access to Trikafta to individuals in urgent need, while regulatory and funding applications are worked through by Vertex with Medsafe and Pharmac. Clinicians in CF centres in NZ are the contact point to discuss this further. This is no substitute for publicly funded access. We will continue to work with all parties to secure Trikafta and Vertex's other CF modulators as soon as possible for our CF community.

Get your SWEAT on!

Our annual appeal SWEATEMBER, calling Kiwis to take on a sweaty challenge for the month of September in support of people with CF, is back and sweatier than ever… Sign up at www.sweatember.org.nz, set a goal, and rope your family and friends into sponsoring you – every sweaty dollar raised with help improve the lives of Kiwis with CF!

Spotlight on our 2021 CF Achievers' Award Recipients

The Cystic Fibrosis Achievers' Awards are awarded annually to people with CF who have achieved excellence in any of four categories; leadership, the arts, sports or education. This year four individuals with CF have been recognised for their optimism, perseverance, and achievement in their chosen category.

A huge thank you to Viatris, makers of Creon, for sponsoring this award.

Gracie McDonald, Education

Eighteen-year-old Gracie is currently studying marketing and communications at The University of Auckland. Gracie and her family moved to Auckland from the UK when she was three. She was diagnosed with cystic fibrosis a year later but has never let it hold her back. Gracie achieved high academic results at college despite the fact she was often in and out of hospital and on IV antibiotics - a part of life with CF that many will understand.

"I was stoked when I found out that I was a CF Achievers' Award recipient; it made me feel like I had been heard - that my challenges were acknowledged by those who truly understand."

While Gracie was at school, she enjoyed history class. Achieving good marks was a big motivator for her, as she says, "it was one thing I had control over."

Gracie is pleased she can take a fraction of the financial burden of some of her medication off her family.

You may have seen Gracie on The Project on TV3 in May, where she shared her story for CF Awareness Month. In the future, she'd like to further her involvement in advocating for the needs of the CF community in New Zealand.

Maisy Millwater, Education

Maisy, who lives in Hawea Flat, Otago, is 16 and our youngest recipient this year.

She's an animal lover with four dogs, lots of chickens, and most recently, has acquired a wild kitten! Maisy has just started a distance Southern Institute of Technology course in pre-health science, with the dream of one day becoming a nurse – something she already knows a lot about.

"Sometimes you have to really push to get to do what you want to do", says Maisy.

The course she is doing usually only accepts applicants over 17 years old, but Maisy found out that SIT do consider people on a case-by-case basis. "With a great supporting letter from a teacher, and the backing of my specialist, I was accepted," she says.

Despite quite a bit of time off school last year, Maisy managed to do very well in art.

It was her art teacher who encouraged her to apply for a CF Achievers' Award, with one of her referees saying "Maisy is a person of enormous character and strength. She is consistently moral, practical, wickedly fun, and fundamentally kind."

Maisy is grateful for the incredible teachers she has had, particularly those who introduced her to the different options and ways to study.

Simon Cowley, Leadership

Last November, nineteen-year-old Simon achieved his Queen Scouts Award. Presented by the Governor General, this award is the highest achievable youth award within Scouting New Zealand, achieved by only 50 people last year.

To qualify Simon had to complete a range of activities in a set period, including community service, multi-day expeditions in a bush environment, and personal and leadership development.

Simon took this adventure in his stride, and at the same time gained skills in organising his medication on the trips away with limited resources and learned how to manage his health in less-than-optimal weather conditions.

Simon has also gained knowledge in managing people and groups of all ages. "These skills have given me the resources to go into a career in outdoor education and leadership." Receiving the CF Achievers' Award for leadership has enabled Simon to invest in items he needs for his future career too.

In-line with his Scouting, Simon loves mountain biking, rock climbing, hiking, kayaking and all outdoor pursuits, and looks for any excuse to go camping with his friends. He is currently working towards his diploma in outdoor and adventure education, which will allow him to go into teaching, guiding, and leading groups in the outdoor industry.

Reese Robertson, Sport

Reese is from the Wairarapa and is currently studying a bachelor's degree in health psychology at Victoria University.

Reese has been awarded her CF Achievers' Award for her incredible involvement, leadership and achievements in sport, namely hockey.

"I play for the Dalefield Premier Women's 1 hockey team, which competes in the top Wellington hockey league. I will also be a part of the Wairarapa Women's team later this year in the National competition where there is a tournament held in Tauranga."

Throwing-back to school, Reese was in the Wairarapa College 1st XI hockey team for five years, played over 120 games, and was captain for the final two years. She also captained the Wairarapa Representative teams in U13s, U15s and U18s.

"In year 12 & 13 it was an honour to be presented with the Kim Condon Cup for Best All Rounder Girls' 1st XI hockey."

Reese is keen to continue doing her degree, whilst also playing for the Dalefield Premier Women's 1 team for the next three years, out of passion, but also, she finds playing is a good way to keep fit.

"Sports is something that I have been passionate about my whole life, and I'm very grateful to have been recognised as someone living with CF doing well in sports."

Reese would like to thank her coaches and family for giving her the push she needed to apply for the award.

2021's Mark Ashford Scholarship recipient Emma Gawn

This year's recipient of the $3,000 Mark Ashford Scholarship is Emma Gawn, a 22-year-old marketing professional living in Wellington.

Emma graduated this year from The University of Otago, Dunedin, with a double Communication Degree in Marketing and Economics and currently works for Mountain Buggy and Phil & Teds.

"I'd followed Georgie Northcoat, who won the scholarship previously, so I knew all about it and my mum encouraged me to apply," Emma says. "I read the criteria and thought to myself that I hadn't done enough but mum said I should be proud of what I've achieved despite the extra challenges that CF brought."

It was a big surprise when Emma found out that she'd been chosen as this year's recipient. "When I got the phone call, I'd completely forgotten about it so I was pleasantly shocked. It was so exciting. It turned out to be perfect timing as my laptop had just broken so I was able to buy a new one and have some money left over to put into my savings".

Twinning with CF

Diagnosed at birth alongside her twin brother, Emma says their mum was glad that they had each other to share their CF experiences - something that most never get to experience face to face. She recalls the laughs they'd have when being separated at CF clinics despite having travelled to the hospital together in the same car. When asked if she and her brother looked out for each other she said,

"Of course, as kids, there were the usual arguments but it did help to have someone else there going through the same treatments growing up. We're really close friends now. As adults it's odd to not see him every day - this is the first time we've been apart by not living in the same city."

While growing up in Oamaru, her family always supported them in living life to the fullest. "We were always encouraged to be active and played every sport we could. My parents never considered it to be a factor in what we could do, they didn't let any opportunity slip by and we did lots of travel as a family. They wanted us to just be kids. It was often tough keeping our treatments going when we were travelling but we made it work."

This active lifestyle continued into adulthood as she spent most weekends during her uni years at her family bach in Wanaka skiing and enjoying the outdoors,

Overcoming CF challenges

However, she has had challenges with her health over the years. "Up until I was eight I was healthy but then I got Aspergillus and became really sick. This required a big course of steroids which resulted in me getting CF-related diabetes."

Though she's never let CF rule her life, she reflects on the challenges she's faced compared to her peers,

"In my last three years at university, I had IV antibiotics three times a year. The uni was pretty good and offered extensions for assignments and I was lucky to have a great group of friends. When I was little I didn't tell people about my CF but once you live with people it doesn't take long for them to realise that there's something wrong. They would help by taking notes for me when I wasn't able to attend class and were very supportive in general."

Hopeful for travel and Trikafta

Looking to the future, Emma would love to travel again. "I'm really enjoying my job right now but once things open up and it's safe to do so I'd love to do some travelling. It's always been in my plans but obviously, the pandemic has put a stop to those for now," she says.

"I hold a UK passport so it would be good to explore over there, even though it is a little far away. I am hopeful that Trikafta will be available here in New Zealand but it's also good to know that I could access it in the UK if I really needed to."

Sharing her story to help others

As someone who previously didn't like to talk about her CF, Emma has come a long way and this year she felt it was time to do her bit for CF Awareness Month. Stepping out of her comfort zone, she recorded a 'Day in the Life Video'.

"I made the video as I've got to the stage where I want to let people in more. It was quite big for me to be able to put something like that out there as I'm not someone who likes to share things, but decided it was about time I did my bit if we do want to get the best CF drugs here. The support and comments were so positive. I'm so glad I did it."

Take the wins!

If you're thinking of applying for the Mark Ashford Scholarship in the future, Emma advises that you go for it and not underestimate what you've achieved,

"I think the biggest thing is, even if you think what you've done isn't a huge achievement you've done it while dealing with a lot of extra pressures and health worries. When I was working and studying at the same time people didn't know what was going on in the background. There were things like coughing all night but still making it to uni each day, taking the time for treatments or having time out for hospitalisations. It means it's totally amazing that you have achieved a degree when dealing with CF. Take the wins!!"

The 2021 Mark Ashford Scholarship was made possible through a generous anonymous donation. Thank you for supporting this wonderful award and for allowing individuals such as Emma follow their dreams.

The 2022 Mark Ashford Scholarship opens for applications 1 November 2021 and closes on 28 February 2022. Visit www.cfnz.org.nz/get-support/financial-assistance-and-awards to apply, or get in touch for a paper application.

SWEATEMBER SWEAT FOR CYSTIC FIBROSIS

Ready, Set, SWEAT….
Sweatember 2021 is here!

Join hundreds of New Zealanders as they take on a sweaty challenge of their choice this SWEATEMBER and raise money for a better future for Kiwis with cystic fibrosis.

Choose a personal challenge that gets the sweat dripping, set up a free fundraising page at sweatember.org.nz, and ask your friends, whānau and followers to sponsor you.

Whether you run a marathon, create an office "step" challenge, do 100 squats every day for a month… the money you raise will help provide critical information, support and advocacy services for your fellow Kiwis who are living with cystic fibrosis.

It's good for your health and good for the health of people with cystic fibrosis - it's a win-win!

Last year's amazing sweaters raised over $140,000 for support services for New Zealanders with cystic fibrosis. Can you help sweat it to the next level in 2021?!

- ✓ Make your sweat count
- ✓ Raise money
- ✓ Support Kiwis with CF
- ✓ Get fit
- ✓ Create awareness
- ✓ Have fun!

The impact of your sweat

Donations raised through SWEATEMBER help bring life-saving medicines such as Trikafta to New Zealand, provide support and information through our CF Social Workers, and fund research for a life unlimited. Every drop of sweat, and every dollar, makes a big difference.

How to sweat for CF this SWEATEMBER

1. Set a SWEATY goal

Going for a walk or run every day, dance classes, train for a marathon, cycling, even climb a mountain every day! You can do a challenge on your own or get a group together.

2. Register to SWEAT

Visit www.sweatember.org.nz to register for FREE. Set a fundraising target and personalise your profile page with a photo. Explain why people should support you, and what motivates you to SWEAT!

3. Spread the word

Tell everyone you know about what you're doing, share it on social media, and email your page out to your networks. Get your page off to a great start by making the first donation.

VIATRIS™

A huge thanks to Viatris, makers of Creon®, for sponsoring SWEATEMBER and for helping to make this campaign possible.

Finding the perfect balance

Balance is a tricky thing for most human beings, let alone when you have cystic fibrosis.

We caught up with 28-year-old Vaughan, to chat all things life, work, and university, where he faced his fair share of challenges, with huge resilience at every step.

Tell us about your time at university; what did you study?

I started at the University of Auckland in 2011 studying Biomedical Science and got admitted to Med School/MBChB programme. Due to health issues, and difficulties with the DHBs I was required to leave the programme in the end of 2013 after three years of study. This was a really tough time in my life.

Having worked so hard for something and having it taken away due to circumstances outside of my control was difficult. I knew CF impacted my life in other ways, but this was the first time it really stopped me in my tracks and took away my dream.

It took me a while to figure out what else I wanted to do, and get out of the tailspin it put me in. I definitely have to thank my family and friends for being with me at this time, and for helping to push me on to different things.

I finally strapped up my laces and I managed to cross-credit and graduate in 2015 with First Class Honours in Biomedical Science. I then decided to try my hand at the legal profession and got offered a PhD position in Health Science in 2016. I worked my way through my law degree and PhD for 4.5 years and finally defended my PhD in May 2020!

You really did experience all the ups and downs life with CF can serve! Have you got any tips on managing study and CF?

Try not to develop a coffee/energy drink addiction… haha but in all honesty, once you find something you enjoy and are passionate about, finding the balance becomes easy.

But it is key to be proactive, if you have quiet times/holidays use those to really focus on your routine, or use it to schedule a hospital admission if necessary.

"Once you find something you enjoy and are passionate about, finding the balance becomes easy."

What are you up to now?

I got a law grad role at Simpson Grierson and was admitted to the bar in December 2020. I currently work as a solicitor at the firm in the Intellectual Property, Sports, Sales and Marketing team.

Give us a fun fact about you

I get grief at work though as my email signature sign off is the longest on record - Dr Vaughan Somerville BMedSc (Hons) / LLB / PhD (Health Science).

How you handle or balance CF 'stuff' with work life?

Some weeks are good weeks, but I also acknowledge I have bad weeks.

For me it is really important to stay active and make exercise and physio treatment a priority so I can continue to work and play at a high level. I'm really lucky to have an accepting work environment that allows me to work from home/hospital during my hospital admissions, and also take time off for appointments when needed, but I acknowledge not everyone has this luxury.

Everyone has a different balance, it's a matter of finding what works for you and sticking to it.

About Vaughan

Vaughan is an Aucklander born and bred (and asks us to reserve judgement here – ha!), into Futsal and currently plays for Auckland City FC.

He previously played for the New Zealand University Team who competed in the World Championships in Kazakhstan in 2018. He's currently in the New Zealand Futsal Whites National Training Centre squad.

As well as playing, Vaughan is the goalkeeper coach for the Futsal Ferns and the Auckland City U14, U16 and U19 boys' teams, and assistant coach for the Auckland Women's team. Vaughan also plays football for Onehunga Mangere Club.

With any remaining downtime, Vaughan's admitted his guilty pleasure is binging any reality TV series, with Love Island his current addiction.

FEATURE

Sam Mannering – Cooking up a storm for CF

Chef, Sunday Star food columnist and author of four cookbooks Sam Mannering is releasing a new book *Food Worth Making Vol. II* in October 2021. He's chosen to donate $5 of the sale of each book to Cystic Fibrosis NZ.

His ethos is to make food and cooking accessible for all and says, "there's nothing worse than putting food on a pedestal." After previously participating in other charity work, Sam's motivations to start supporting CFNZ were personal and reflective,

"After covid, things have changed so much and it feels in bad taste just going out there and shouting about my new book. I wanted to give something back and I wanted a charity that I could really help in the long term."

It wasn't an easy decision to make and he didn't make it lightly. After a personal health scare with melanoma, he realised how lucky he had been and how privileged he was to have a platform he could use to help others. Choosing who to support needed to mean something to him and make a difference in people's lives,

"I was trying to decide which cause to align myself with that could be ongoing so I wanted to be careful what I chose. I initially had thought of better-known charities but realised they already had great public awareness. When my mother said, 'what about cystic fibrosis' we realised that most people don't really know what it is. We have some family friends who have CF so it made sense to commit to it."

He continued,

"It's a cause that needs a voice. I wanted to get behind something that would benefit from extra awareness and education as well as money."

Unlike many who fundraise for CF, Sam has no direct family with the disease but amongst those he'd known was a close family friend's brother who sadly passed away in his early 20s. Even with this connection, he knew little about what CF meant for those affected in their day-to-day lives before embarking on this partnership,

"I am using myself as a case study. My knowledge was limited until I decided to work with CFNZ so I am learning as I go and keen to teach others. One goal is to come up with as much financial assistance as I can but I also want to raise awareness and understanding too. The more you know the more you can help. I've seen the way CF affects families and it can be devastating. The naive ignorance that some people have means it can be harder for those with CF to get the support they need. I want to break down those barriers."

A perfectionist and self-confessed control freak, Sam has been dedicated to his craft for 12 years now starting in London, Hong Kong and now home in Aotearoa,

"I'm self-published and do all my own recipes, writing, photography and design. The hardest part is stopping and saying no more. This latest book has been a bit of a battle as covid has resulted in major disruption (it was due to be printed in February 2020). But, I'm really pleased with it and looking forward to sharing it with everyone. "

His relentless schedule of writing and coming up with recipes every week means he has an abundance of content to play with,

"Vol II contains all of the best of my work from my newspaper columns (and there will be a Vol. III coming next year too). I draw inspiration from Nigel Slater OBE and his columns in The Guardian."

Sam is aiming to raise $25k by Christmas. This ambitious figure matches with the man's personal drive and determination and somewhat humble self-awareness,

"I have a platform and my recipes are getting millions of hits online. I worry that I've taken that for granted, so I want to do a bit of good and work alongside the goals of CFNZ. I was shocked by the lack of government funding for PWCF. I'm still staggered by how much support is given privately. If I can help just a little by raising funds and awareness, I'll be pleased."

Food Worth Making Vol. II is out in all NZ bookshops from October 2021 for $65 - or buy Vol. I and II together for $120. $5 from each sale will be donated to Cystic Fibrosis New Zealand.

Food Worth Making Vol. II is out in all NZ bookshops from October 2021 for $65 (with $5 to CFNZ) or head to www.sammannering.com/shop to pre-order.

Josh's fitness journey

Research shows that regular physical activity or exercise provides multiple benefits for people with cystic fibrosis. In this article Josh Chase, a 27-year-old qualified personal trainer with CF, shares his approach to fitness and how he keeps motivated.

A change of mind-set

Living with CF has never been easy - as most of you would know! When I was younger all the extra tasks needed to stay healthy felt like a chore more than anything, especially when friends didn't have to do the same.

I had a big interest in gaming from a young age, and it helped to get me through the daunting physio sessions / hospital admissions. Even though I love gaming, now that I'm older (and I would like to think wiser!) I've reframed my thinking and take pride in looking after my health, and I absolutely love exercise.

I exercise every single day by going to the gym, going for a little run, just generally staying active. I also love going on adventures such as exploring waterfalls, hiking tracks or learning to rock climb – if there is an adventure, count me in. My two hobbies are complete opposites, but they make for my perfect balanced lifestyle.

Joining the health and fitness industry

What drove me to take the first steps into fitness was the realisation I needed to better myself, keep myself fit, keep healthy and do what I could to preserve these lungs.

I got a gym membership and didn't look back. Since making the lifestyle adjustment, it has changed how I deal with my CF, and my body loves me for it. I'm gaining weight, breathing better, and it just makes me feel good.

I realise a lot of people might find getting fit a bit harder, so it's doing things such as going for a 10-minute walk after dinner to start with, then increasing it over time to longer walks or even runs, or talk to a gym about a personal trainer for a few months until you feel comfortable doing things at the gym by yourself.

My philosophy is that just because you get dealt a crappy hand doesn't mean you automatically lose - you just have to play it out!

The combination of life with CF and my love of exercise let me to pursue a career in personal training. Last year I started my study through NZIHF; now that I'm qualified, I'd like to continue my studies to learn how I can help more people like us, who have battles with their health.

Something I thought I could never do

On my fitness journey I discovered running makes me feel good, so I tackled the 11km in Auckland's "Run the City" event on Sunday 1 November 2020.

It's something I thought I could never do. In preparation for the event, I added running sessions every couple of days when my lungs felt up to it. As someone with CF, breathing was a big component that I had to manage during the run. I did this by slowing my pace, catching my breath, and to start jogging again.

It was an unforgettable experience, and an awesome achievement to cross that finish line. Now I know what I am capable of, I am hoping to enter a half marathon and a powerlifting competition.

> "Just because you get dealt a crappy hand doesn't mean you automatically lose - you just have to play it out!"

Helping others with CF

Moving forward with my personal training qualification I want to help people be the best versions of themselves. More than anything, I want to help people with CF look after their lungs - I know I can't do it face-to-face, but I have plans, watch this space!

I want to feel and be healthy with my CF and fitness combination, that I can be a role model to others in the CF community, and become someone they can speak to if they want advice.

Shoutout to my Mum – she's the person I look up to the most. She has always gone out of her way to make sure I was the healthiest I could be when I was younger, she is the most selfless and caring person, and if I was anything like her I'd be happy.

Top Tips for your fitness journey – CF Physio, Lucy Goss

1. **Find something you enjoy** - it doesn't matter what it is or whether someone else thinks it's 'exercise'. Do something you like and you will keep it up!
2. **Breathlessness is normal for everyone!** Push yourself to be breathless and become comfortable with recovery.
3. **Strength and cardio** is the best combination - don't avoid one or the other. Mix it up
4. **Learn the correct technique** before initiating any type of exercise. Poor form and execution can result in more damage than good.
5. **Consistency is key!**

Epic 65 Hour Bowls Challenge raises $20k for CF

Jess and Dan Hopkins' daughter Caitlin, who was born seven weeks premature, was diagnosed with CF at 12 weeks old 18 years ago. In December 2020, Dan embarked on an epic 65 hours of bowling at Hampstead Bowling Club, Ashburton fundraising over $20,000 in the process. They wanted to raise funds and awareness of CFNZ but never expected the abundance of love and support that came with it.

Why bowls?

So, why did they choose a bowl-a-thon? Dan had been introduced to the sport at 13 years old alongside his brother Shaun when they stayed with their grandparents during school holidays, "Some of the older members weren't impressed with us." he joked.

His first season was back in 1991 but later taking 10 years off as an adult while he raised his kids before he got back into it. But how could he incorporate the game he truly loves and help people with CF?

"I thought I'd try something different for fundraising and wanted to tie in the 65 roses. I do remember thinking 'I wish CF didn't use the number 65' it's a big number to work with. That's how 65 Hours for Cystic Fibrosis came about." said Dan.

Powering through

Once the logistics of 65 hours of bowling were all worked out, and the many players were signed up to take part, there was no going back. The intense schedule was going to be tough on Dan,

"I decided I'd allow myself a 10-minute break every hour but I could bank it and save it up to allow me to have a sleep if I needed to. It was 24 hours before I took my first break - and that was much easier than I thought to achieve. Towards the end, I had 200 or so minutes leftover of breaks as the adrenaline was keeping me going."

The last hour was saved for the family so Dan, Jess and their son Matthew played together. Daughter Caitlin was working but joined them afterwards for lots of celebrations.

A community united for CF

Dan played alongside a roster of bowlers in two-hour time slots during the day and four-hour time slots at night to reduce the noise and comings and goings for neighbours but they needn't have worried. The event couldn't have gone better.

The whole community rallied around to support the couple in their efforts. People visited Dan on the lawn to hand him cash donations, neighbours passed freshly baked scones and jam over their fence to players, local business Motus Physiotherapy was on-site to provide free massages to Dan to help his body get through the gruelling schedule and friends even brought down their caravan so he could get some sleep during his breaks. Dan was particularly grateful for the support of his boss,

"My employer has been fantastic. He was the first person I spoke to about doing the challenge as I needed him on board so I could actually do it and he was supportive right away. He was there many times during the event and was so generous. I'd put in time for annual leave but when I went to put it in he said he'd decided not to let me take leave and instead to pay me for those hours and donate to our cause. The generosity blew me away."

Challenges and celebrations

Dan completed the intensely gruelling and demanding challenge despite his own physical conditions. He has Autoimmune Arthritis which can cause him a lot of discomfort as well as suffering intense eye pain at the time following multiple surgeries for a detached retina,

"I was in a lot of pain but Jess made sure I took medication when I needed it. I had set my mind to do it and it's amazing what your body can do when you want to do something. The day I finished it I went home for a few hours sleep and then was back at the club for the auction and BBQ. I was the last to leave and got home just before 2 am. I had a big night!"

CF was always on their minds

Daughter Caitlin has always been private about her CF so Jess and Dan wanted to make sure she was on board with the event first. Throughout her life she never wanted it to hold her back so many of her friends didn't know that she had CF as she didn't want to be treated differently. But, she totally supported her dad and has since become more open about her CF. Now 18, she's working, studying and managing her health well. Jess and Dan say,

"She's hard-working and independent so we try not to nag her but just check that she's keeping up with everything. We're here for her when she needs us but mostly she likes to hang out with her friends and get on with life. She has big dreams and goals for the future."

Another local family attended the event with their son who had CF and that visit was a particularly special moment for Dan,

"The last hour was absolute torture so that meeting really spurred me on during that time. But I thoroughly enjoyed the experience of doing the challenge and it pushed me to my limits. Knowing what I was doing was going to help so many people is what got me through. And, I couldn't have done it without Jess."

Smashing fundraising targets

Jess recalls, "We never knew how consuming it would be and our main goal was to get the word out and raise awareness and as many funds as we could.

"We had no idea it would get as big as it did. We would've been happy with $5,000. Before we knew it, we'd got there and we were completely blown away by it. We had TV3 news, papers and local radio supporting us. The Rock did live interviews with us on the day so their listeners got on board too."

She also couldn't believe the community support that came through for them,

"One man came and gave us $100 but said he'd already donated to the campaign but wanted to give something just for our daughter to do something nice," she recalls.

"Another woman did the same and handed me $50 cash so that we could have a meal out once it was all over," added Dan.

An unforgettable 65 hours

After such a huge achievement, they're both still riding high and say it's something they'll remember for the rest of their lives. "We can talk about it for hours," jokes Jess.

They are unlikely to forget, thanks to the body art they now have,

"We both got tattoos to commemorate the event. I got a 65 and a rose. It seemed like a good first tattoo. Jess got the rose and crown as Caitlin has always been our princess. I'm very proud of the memories we have, what we've done and what we did it for."

After completing such a huge challenge, you'd think they'd want a break, they certainly deserve it, but the couple aren't done yet,

"I'd do it all again in a heartbeat for the cause. For sure, without a doubt. We're already trying to think about what our next fundraiser could be."

Watch this space!

"We had no idea it would get as big as it did. We would've been happy with $5,000. Before we knew it, we'd got there and we were completely blown away by it. We had TV3 news, papers and local radio supporting us."

Couple celebrate big, fat, free 'fantastic' wedding

For many young couples, a dream wedding takes years of saving and planning. After gaining 19k votes on Facebook, James Rondell and his wife Alana won theirs through the Marlborough Big Fat Free Wedding competition, and on the first of May 2021 they got married.

A surprise proposal

They met at The Warehouse in Blenheim where they both worked. They'd been dating for two and half years when James decided to surprise Alana with a proposal at her 21st birthday party last December, surrounded by their friends and family. He recalls how he kept it a secret,

"Alana really wanted a pet, but we don't have room in our one bed apartment, so I got her a robot fish for her birthday as a joke. People had thought I'd dropped the ball and got a hard time from her for that. She had no clue what my real present was.

My sister does cake decorating, so I hid the ring on a rose and put it on the cake board so that I could get down on one knee after we presented the cake."

Interfering relatives for the win

They were entered into the Marlborough Big Fat Free Wedding competition thanks to interference from their relatives.

"We didn't know this competition existed, so it was just really great timing. My grandfather had seen the advert and suggested to my mum and aunties that they enter on our behalf. My aunties wrote a few paragraphs about our relationship, my CF and Alana moving away from Christchurch post-earthquake. We had no idea they'd done it and without them interfering we'd still be engaged today."

A few weeks later they were told they were in the top 10. Following an interview with the judging panel, they were delighted to hear that they made the final as one of five couples selected from the 70 plus entrants.

A photo and profile of each finalist were put up on Facebook. To win the big prize, James and Alana needed to get the most likes on their photo. He recalls that "countless hours of sleep were lost" during the two weeks of voting but he needn't have worried.

> "Alana went away and learnt as much as possible by researching online so she actually knew a lot about CF before we even started dating."

"It was amazing. We thought we'd get 500 votes at best, but we won with 19k votes. Some of the work that my friends and family did to get the post out there was amazing. I really want to thank the CF community for their support. I asked Sue Lovelock, my CFNZ fieldworker, to see if she could get it onto the CFNZ Facebook page and it went from there. We couldn't have won without that support."

The morning after the voting closed, James and Alana got the good news,

"They messaged us on a Saturday morning to say we'd won, 'Congratulations, you better start shopping and you've got 10 weeks before your wedding!' We'd set a date for early 2023 to get married giving us time to plan and save and now it was only a few weeks away. We were so excited. There were a lot of tears and celebrations that day. Everyone was so happy for us"

Included in the prize were some dance lessons. In preparation for the wedding, they went along expecting a ballroom lesson to prepare for a first dance,

"The lessons were for salsa dancing which was different than we expected but great fun. We couldn't do it as our first dance due to the wedding dress being too big so we did a quick change and did our salsa dance later on in the night."

It was our dream day

When it came to the big day, it couldn't have been better,

"The day was all just perfect. It was our dream wedding. We got almost everything taken care of from the venue, catering, celebrant, photography, haircuts and styling, cake, DJ the whole lot.

One of my favourite memories was looking and seeing my four groomsmen tearing up. These big staunch guys I've known all my life were all moved to tears with happiness for us. I never knew they were such big softies."

As a rock and metal fan, James wanted to make sure he put his stamp on the wedding music too,

"The DJ asked if I had any requests. I asked if she had a tune from Devilskin. Cue me and my mates at the top table headbanging while everyone else stopped dancing. I even saw someone with their fingers in their ears. It was my wedding and my favourite band so I thought let's go for it. I feel bad for killing the dance floor vibe though," he joked.

Taking CF in his stride

James' life with CF has never been a secret. His mum had worked at The Warehouse for 18 years before he started working there so his colleagues were pretty clued up. He's positive and laid back about it,

"I haven't had too much grief with it. I've stayed relatively healthy throughout my life but my weight was one of my downfalls. When I left high school, I had a PEG tube fitted to help with weight gain. I quickly felt an Increase in energy and the more weight I put on the better my lung function got. I had sinus surgery a few years ago too which also improved my lung function as I get fewer infections and colds."

He generally takes his CF in his stride casually saying, "I have the occasional hospital visit for a two-week tune-up about once a year."

When it came to his relationship with Alana, she took it upon herself to find out everything she could about CF,

"We were friends at work to start with. I mentioned I'd had CF after I'd coughed a few times. She went away and learnt as much as possible by researching online so she actually knew a lot about it before we even started dating. I've always thought there's no point in hiding it. Awareness is half the battle. People are always surprised when I tell them and say, 'I had no idea' and 'you look normal'. I am just open and honest with people about what I can and can't do."

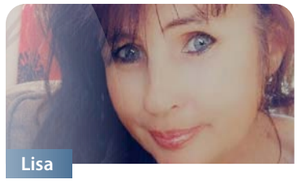
Planning their future

James isn't usually a thinking ahead kind of guy but now that he's married he and Alana can plan for the future.

"I'm not a planner and tend to go with what happens and what will be will be. But now, I'm excited to look to the future. We are hoping to move into a bigger place. We want to buy a house and start a family within the next five years."

As the cherry on top of a perfect wedding, CFNZ corporate supporters Choice Hotels gifted James and Alana a complimentary nights' stay in Christchurch for their honeymoon. Thanks, Choice!

Trikafta Wall of Wishes



Lisa

I want Trikafta so that children being born today with CF live long, fulfilling, happy lives, free from the difficulties of those before them.

Carmen

I need Trikafta so my children do not have to go through the pain of losing their mother.

Ingrid

We wish that all Kiwis with CF will get the best treatment & medicines available. Currently, that's Trikafta. We need this so little one's like our Orson can live a long & happy life.

Kareena

My wish is for Natalie to grow up with a life unlimited.

Lizzie

I want Trikafta for my twin and all Kiwis with CF, so they don't have to have a transplant young like me.

Alaric

I want Trikafta so I can grow up and live a long, normal, healthy life.

Bella

I want Trikafta so I can live to see my future.

Chantelle

I need Trikafta so my children don't grow up without their mum.

Erana

I would love to have Trikafta to enjoy making memories without the struggle.

Dominique

My wish is to get trikafta for my almost 7 month old CFer so she has a better life as she grows up!!

Sophie

I want Trikafta for Poppy so she can live a full life without limits

Rebekah

I would like Trikafta for my son so he can have a much greater quality of life and improved life expectancy. He is worth it!

Trikafta Campaign - Major Progress Made

The last few months have seen major progress in the campaign to secure access to cystic fibrosis (CF) modulator therapies for Kiwis with CF. Vertex has submitted applications for approval of three CF modulator therapies including Trikafta to Medsafe, and made an application to Pharmac for funding of Trikafta. Vertex has also put in place a Managed Access Programme for Trikafta for those in critical need.

The Trikafta campaign has continued to have a high profile with coverage from TV and media, together with meetings with MPs to press for public funding of Trikafta and other CF modulator therapies.

Again, we need to thank all of those in the CF community who've shared their stories, met with their MPs, and raised the profile of CF across New Zealand. It's this commitment, together with the support of Newshub and other media, that has made such rapid progress possible.

Stakeholder meeting held

In late March, CFNZ was able to convene a stakeholder meeting between Vertex, Pharmac and Medsafe to identify a pathway for public funding of Trikafta (elexacaftor/tezacaftor/ivacaftor and ivacaftor) in the shortest possible time. CFNZ was joined at the meeting by representatives of the CFNZ Clinical Advisory Panel and Trikafta for Kiwis.

This meeting provided the opportunity to present information about the current health needs of those with CF in New Zealand, the scope of currently available treatment, and the impact of CF on the lives of those with the condition and their families.

Following this meeting, Pharmac confirmed that it would consider an application for Trikafta in parallel with Vertex's application to Medsafe, enabling the funding assessment process to start earlier than it normally would.

Applications to Medsafe and Pharmac

At the end of April, Vertex took the first step in its New Zealand approval and funding process by submitting an application to Medsafe for approval of Symdeko. This application was the first of the three CF modulator therapies for which Vertex has committed to seeking NZ approval. Symdeko (tezacaftor/ivacaftor and ivacaftor) works for people with two copies of the F508 mutation or one copy of F508 and a mutation that is responsive to the drug.

The application for Symdeko was followed by an application to Medsafe for approval of Orkambi (lumacaftor/ivacaftor), which works for people with CF with two copies of the F508 mutation and has been approved overseas for children with CF two years of age and older.

Vertex's application for Trikafta was submitted in late June, meaning that applications for all three of the CF modulator therapies are now working their way through Medsafe's safety approval process. The application for Trikafta was for people with CF 6 years and older. Medsafe confirmed that it has granted priority status for the Trikafta application, enabling a shorter time for evaluation.

Vertex's application to Pharmac for funding of Trikafta came shortly after, with Pharmac confirming on 9 July that it had received the application and was starting its funding assessment process.

Trikafta Managed Access Programme

Early July also saw the establishment of a Managed Access Programme for Trikafta in New Zealand. Vertex has put the Programme in place to provide free access to Trikafta for those people with CF who are in critical need.

The Programme is part of a worldwide scheme managed globally through Vertex Global Medical Information and is only for Trikafta. CF clinicians in New Zealand are now able to make a formal request to Vertex for patients who meet certain criteria. Patients and their families cannot apply direct to Vertex.

CFNZ is delighted that Vertex has put the Programme in place. It will provide a lifeline to those who not do not have the luxury of waiting while Medsafe and Pharmac going through their assessment processes.

But the Managed Access Programme is only for a small group of people with CF who are in critical need. It is no substitute for public funding of Trikafta, and other CF modulators, for all of those people with CF in New Zealand who would benefit.

Next Steps

Now that Vertex has submitted its application for funding of Trikafta to Pharmac, the funding assessment process is underway. Pharmac has advised that its Respiratory Subcommittee and its Pharmacology and Therapeutics Committee (PTAC) will assess the scientific evidence on this medicine within the next few months. These committees will make recommendations to Pharmac on whether or not Trikafta should be funded. If it is recommended to be funded, Pharmac will carry out an economic assessment and rank it against all of the other medicines awaiting funding.

Following the Stakeholder meeting earlier this year, Pharmac invited CFNZ to provide information to support its assessment of Trikafta. In response to Pharmac's invitation, CFNZ has submitted a comprehensive package of information describing the current health needs of those with CF in New Zealand, the scope of current treatment regimes, and the impact of CF on those with the condition, their families and whānau.

With the help of those who have been taking Kalydeco (ivacaftor) since it was funded in New Zealand last year, and those who have been able to privately access Trikafta, CFNZ has also provided information about the life-changing benefits of CF modulator therapies.

CFNZ will be taking every opportunity to meet with Pharmac during the assessment process, and provide further input and information, to ensure Pharmac understands the life-long burden of living with CF and how Trikafta and other CF modulators can lift that burden.

Trikafta Petition

The Trikafta for Kiwis petition to the Health Select Committee remains a key part of the campaign for public funding of Trikafta. CFNZ will be working with Trikafta for Kiwis to arrange for the presentation of petition to Parliament later this year.

If you have not already done so, you can still sign the petition and encourage everyone you know to do so as well. Almost 36,000 people have already signed – an amazing achievement for a small community. But let's see if we can do even better!

Trikafta Overseas

Trikafta is now funded in 19 countries. Agreements have recently been reached in France and Italy to make Trikafta (Kaftrio in Europe) available for people with CF 12 years and older who have two copies of the F508del mutation, and one copy of the F508del mutation and one minimal function mutation.

The agreements also provide for Symdeko (Symkevi in Europe) to be made available to those with one copy of the F508del mutation and one residual function mutation. In Italy, the agreement also widens access to Orkambi for those with CF who are two years and older, and to Kalydeco for those aged one year and older.

Trikafta received Australian Therapeutic Goods Association (TGA) approval in March 2021, but the Australian Pharmaceutical Benefits Committee (PBAC) announced in April that it had deferred a decision to list Trikafta on its Pharmaceutical Benefits Schedule (PBS) to allow for further negotiation. An update is expected in August.

In addition to funding agreements, there have been a number of approvals to widen access to Trikafta by both mutation type and age group. The European Commission has announced approval of the extension of Trikafta to those people with CF age 12 years and older who have at least one copy of the F508del mutation. Access had previously been limited to those with two copies of the F508del mutation or one copy of F508del and one copy of a minimal function mutation.

The US Federal Drug Administration (FDA) has approved expanded use of Trikafta to include children with CF aged 6 to 11 years who have at least one copy of the F508del mutation or at least one copy of a mutation that is responsive to Trikafta in vitro.

Vertex has announced that it has applied to the European Medicines Agency and the UK Medicines and Healthcare products Regulatory Agency (MHRA) for expanded use of Trikafta for children with CF 6 to 11 years old who have at least one copy of the F508del mutation.

Vertex has also said that it will initiate a Phase 3 development programme in the second half of 2021 for a new once-daily investigational triple combination therapy. Vertex noted that its Phase 2 study data suggest the new triple combination therapy has the potential to restore CFTR function in people with CF to even higher levels than other Vertex CFTR modulators.

Pharmac Funding

CFNZ continues to campaign for a significant increase in the public funding available for medicines, together with improvements to current arrangements to speed up access to life-changing medicines. Without such an increase, CFNZ is concerned that Pharmac will have not have room within its budget to fund Trikafta and other CF modulators.

The need for additional funding was made clear in late April, when Pharmac announced that it has a waiting list of 73 medicines that it would like to fund. Funding these medicines would cost an extra $417 million, an increase of 40% on its current $1 billion annual budget.

To support the campaign for more funding, CFNZ representatives attended the presentation at Parliament of the petition from Patient Voice Aotearoa (PVA) on 12 May. The PVA petition, which was signed by more than 100,000 people, sought a review of Pharmac and the doubling of its budget. CFer Bella Powell was one of the speakers at the event, noting that having access to Trikafta was the only reason she was able to attend the presentation.

In spite of major support for the PVA petition and Pharmac's announcement of its need for additional funding, the Government's May Budget provided only $200m for the next four years, with $40 million for 2022/23.

CFNZ is deeply disappointed at this limited level of extra funding, being only a tenth of the amount Pharmac says it needs simply to catch up, let along fund new medicines such as Trikafta. CFNZ will continue to lobby for an increased budget for Pharmac to ensure life changing medicines such as Trikafta and other CF modulators can be funded in New Zealand.

Pharmac Review

Earlier this year, the Government announced a review of Pharmac to assess how well it is performing, whether its performance could be improved, and whether its current objectives maximise its potential to improve health outcomes for all New Zealanders.

The review is being undertaken by an independent panel chaired by consumer advocate Sue Chetwin. The review panel will provide an interim report in August and a final report to Government by December 2021.

CFNZ has been able to meet with Sue Chetwin and attend two workshops with the wider review panel and other patient representative organisations. This provided the opportunity to debate key issues of concern over the current arrangements for funding of medicines in New Zealand, and how current arrangements could be improved.

CFNZ has also made an interim submission, responding to questions asked by the review panel. In CFNZ raised a range of concerns, including the lack of transparency over current funding and assessment processes, the lack of opportunities for patients, their families and patient representatives to provide input into the assessment of medicines, and the lack of timelines for decision-making. CFNZ also raised concerns over the lack of provision for those with long term conditions and rare disorders, and the definition of rare disorders used by Pharmac which is far narrower than that used overseas.

While the terms of reference for the review exclude consideration of Pharmac's budget, CFNZ expressed concerns over the lack of funding for medicines, and the model adopted by Pharmac of funding new medicines only from cost savings. Many other patient representative organisations have raised similar concerns to CFNZ.

CFNZ is hopeful that the review panel has heard and understood the concerns of patients, their families and patient organisations, and that the interim report will start the process of identifying how the current system can be substantially improved.

The review panel will provide its interim report to the Minister of Health by 20 August. CFNZ understands that there may be an opportunity for further public submissions to be made prior to the final report being released in December 2021. CFNZ will take all the opportunities it can to encourage the review panel to propose significant improvements to the current system.

Health & Disability Review

In June 2020, the Minister of Health released the final report of the Health and Disability System Review. The review made a number of recommendations to reduce the fragmentation and complexity of the current health system, and improve health outcomes. The report included a recommendation to reduce the number of DHBs from the current 20 to 8-12 within five years.

In April 2021, the Government confirmed the details of the health system reforms in response to the review. The changes go further than the review recommendations and provide for:

- All DHBs to be replaced by one national health organisation, Health New Zealand
- A new Māori health Authority to be established to commission health services, monitor the state of Māori health, and develop policy
- A new Public Health Agency to be created
- Strengthened Ministry of Health to monitor performance and advise Government.

The reforms will be phased in over three years, with enabling legislation to be introduced into Parliament last in 2021, and Health NZ and the Māori Health Authority to be in place by July 2022.

A key issue for CFNZ is how the assessment and funding of medicines, and the recommendations of the Pharmac review panel, will be integrated into the reforms. Ensuring the best standard of care, including access to modern medicines such as Trikafta, is critical to improving the health outcomes of people with CF and must be considered in an integrated way. CFNZ will be seeking opportunities to provide input to the proposed changes to the health system to make sure the needs of people with CF are understood and addressed in the reform process.

What's going (on) down the drain? – More than you think!

Julian Cox, Chair of CFNZ Otago Branch, shares the findings from a collaborative project between the branch and Otago University on nebuliser residue and antibiotic resistance.

The growing issue of antibiotic resistance

People with cystic fibrosis use antibiotics inhaled through a nebuliser to prevent and treat bacterial lung infections. In New Zealand, nebulised tobramycin is recommended for treating or managing infection with Pseudomonas aeruginosa, a major cause of lung infection with people with CF.

Bacteria are very good at adapting to their environment and changing in ways that decrease or eliminate the effectiveness of antibiotics. This antibiotic resistance is a serious and growing health problem.

Knowing that Pseudomonas particularly like living in moist environments (lungs and drains) I've always been a little concerned about what might be happening down our sink drain when we clean up the nebuliser that has just been used for tobramycin. Are all the little P.a's there becoming antibiotic resistant?

There also seemed to be a disconnect between manufacturing advice for cleaning nebulisers (e.g. "rinse and clean to remove residue after use") and national advice for disposing of antibiotics (e.g. "never pour down drains or put in household rubbish").

Curiously, while I'm pretty adept at internet research, I couldn't seem to find any information on this. It turns out that's because, if anyone has thought about it, they haven't formally studied it.

I happened to see an advert in the paper for the Otago Participatory Science Platform, which provides funding to promote citizen science. A few emails and phone calls later (it really was that easy) and CFNZ Otago Branch had almost $20,000 in funding and the University of Otago fully on board to investigate.

The research was led by Karyn Maclennan of the Department of Preventive and Social Medicine with Greg Walker and his research technician, Pummy Krittaphol, of the Pharmacy department responsible for most of the science.

Surveys, spectrometers, and samples

The first task was to confirm that, despite national guidelines not to wash antibiotics down the drain, it wasn't just the Cox household rinsing their neb into the sink. So we set up a phone and online survey which did indeed confirm everyone on inhaled antibiotics who took part in the survey is doing just that.

Next job was to find out just how much tobramycin was going down the drain from a typical neb wash. This is pretty tricky given that tobramycin dissolves in water and you don't know exactly how much is going into and coming out of the lungs. Luckily the Pharmacy department has a very expensive machine call a Liquid Chromatography Mass Spectrometer (LC-MS). Given some pure tobramycin they can develop a test which can then be used to work out how much tobramycin is in the water.

Next, we needed to collect some rinse samples from actual people on inhaled tobramycin. They were given some distilled water and used that to wash the neb into a special bowl from which a sample of the rinse water was taken. Using the LC-MS we could work out the concentration of tobramycin in the water.

Down the drain

When the results came in, I think everyone was a little taken aback - 40% of the contents of that little tube of tobramycin ends up being washed down the sink. That's a lot!

If you're wondering where all that residue is coming from – much of what you inhale gets exhaled out again and then condenses back on the neb (especially if you are using a filter), plus there is always some left over in the nebuliser reservoir that doesn't get aerosolised.

You may also be thinking – woah does that mean I'm not inhaling the right amount of tobramycin? Fortunately, no, it does not. Efficacy studies looking at the appropriate dose of nebulised tobramycin for people with CF account for the dose that actually gets into the lungs.

Survey results

Unfortunately, we still don't know for sure if the amount of tobramycin going down the drain might be causing antibiotic resistance. To work that out we'd need to develop some more cunning tests on the microbes themselves.

"There also seemed to be a disconnect between manufacturing advice for cleaning nebulisers and national advice for disposing of antibiotics"

However, we did rope in some experts as part of a community Hui on the results. Iain Lamont of the Otago University Pseudomonas lab thought that, while many Pseudomonas microbes present may well be killed at that concentration, it was also possible that the exposure could lead to some tobramycin-resistant Pseudomonas.

Safely washing nebs going forward

The study has raised a number of questions that require further investigation. It may be possible to construct a low-cost household filter system which extracts the antibiotic from the rinse water, for example.

In the meantime, disposal of the rinse water down the toilet (with the lid closed) was one key recommendation from this review that no one from our survey was doing. We also recommend weekly bleaching of the key drains. Bleach works through a different mechanism from antibiotics and is likely to be effective on antibiotic resistant Pseudomonas.

Thanks to the researchers, participants and supports of the study. It's been really affirming to see a small question that I had piqued the curiosity of others as well. I was particularly pleased to hear that Iain and Greg have now received significant funding for further research (based partly on the LC-MS test Greg developed for this study) that could lead to more effective treatments of Pseudomonas.

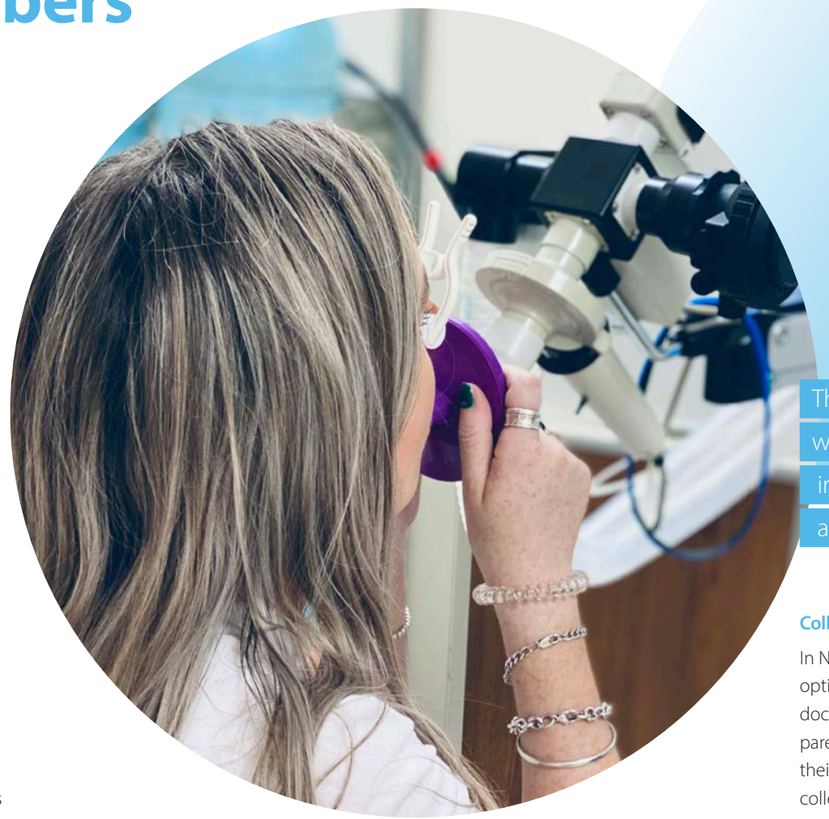
It's important to follow best practice guidelines, such as those in the 2020 Bell et al review: Nebuliser hygiene in cystic fibrosis: evidence-based recommendations.

1. **Wash nebuliser components in warm water and dish washing liquid in a dedicated plastic, glass or metal bowl.**
2. **Dispose of nebuliser wash water down the toilet, closing the lid before flushing.**
3. **Clean toilets and drains with bleach – this may help kill any Pseudomonas bacteria that are present.**

To read the full study results, scan the QR code using your phone camera, or visit www.cfnz.org.nz/nebuliser-study

CF Data Registry: Strength in numbers

Jan Tate, Port CF Project Coordinator, gives an overview of what Port CF is, how it compares globally, and how important the data is for the future of CF healthcare.



Port CF, the name of the New Zealand CF Data Registry, is aweb-based data registry owned by Cystic Fibrosis NZ and uses anonymous patient data to analyse trends in CF care.

Building up a picture of CF in NZ

Port CF was started in Christchurch as a pilot study in 2011, and from 2012 all people with CF in NZ (discussed with parents of babies and children) have been asked to consent for their data to be included.

Prior to 2012, people with CF had data collected and this data was combined with Australia – known as the Australasian Data Registry. Despite best efforts from the healthcare teams adding information to this 'spreadsheet registry', a great deal of data was not included and so didn't give a true 'picture' of CF in NZ.

Around 2005 the CEO of CFNZ investigated world-wide CF data registries and found the USA and UK Trust models best suited the NZ CF population. Following discussions with the CF Foundation, the data registry programme in USA was offered for use in NZ, free of charge. A NZ Port CF committee was formed and CF health professionals from Christchurch and Auckland worked with Andrew Watson from the Christchurch Hospital IT department, to formulate questions for NZ, based on the CF Foundation registry.

Even though the questions asked are slightly different from the US Foundation registry, the data collected in NZ can be compared to the USA and UK CF populations.

Comparing globally

Recently, Dr Cass Byrnes and Jan Tate from Starship were asked to be part of a 'CF Harmonising Committee' from Canada, Australia, France and NZ – a committee formed to standardise the CF registries from these countries.

At present each registry asks similar questions but in different formats or different ways. The outcome of harmonisation will compare results from the same questions asked in the four registries. This will result in comparisons between the countries around CF care and management.

It's a mammoth task as some points are easily approved such as weight and height of a person but other values have had a lot of discussion. We have Zoom meetings which are sometimes tricky with different time zones in each country. This Harmonization Committee is led by Dr Anne Stephenson from St Michaels in Canada and studies are soon to be published and / or presented at CF conferences.

The registry can identify the number of people with a specific CF gene. This has become very important over the past few months with the arrival of the modulator drugs.

Collecting data in NZ

In NZ, within a few months of being diagnosed with CF, the option to be part of the Port CF registry is discussed by the doctor or nurse specialist. Each newly diagnosed person (or parent on behalf of their child) is asked to sign consent for their data to be collected and added to the registry – all data collected is non-identified. Once signed on, initial data is collected, this includes date of birth, gender, CF genotypes, how CF was diagnosed and sweat chloride values.

After the consent and demographic data are obtained, all information from each clinic appointment or hospital (or Home IV) visit is recorded. This includes the results of growth, lung function values, nutrition and enzymes, pulmonary therapies, respiratory microbiology, blood values and complications.

Each year the data from Port CF is collated by a statistician and reports are generated with different end points. A few examples: lung function values with age and gender; types of bacteria in different age groups; growth percentiles with ages and gender; number of people on dietary supplements; types of physiotherapy; number of people on Pulmozyme / Hypertonic Saline, etc.

These endpoints are summarised in an annual report and published on the CFNZ website. Once a person has had a lung transplant their data is no longer collected; their records stay on the registry but they become 'inactive' and not included in the Port CF annual reports.

How is the data used?

At present we have years to 2019 completed (2018 and 2019 almost published) and 2020 yet to be analysed. The data in the registry is dependent on the health professionals putting the data in. Mostly it is the nurses from each clinic that add the data for each clinic or hospital visit, but there are a couple of Paediatricians who have a small number of patients who do this for us. I always say – "a registry is only as good as the correct data put in."

One of the benefits of having an up-to-date data registry, is to confidentially record, how many people with CF we have in NZ. The registry can identify the number of people with a specific CF gene and which combination of CF genes people have. This has become very important over the past few months with the arrival of the modulator drugs.

Ivacaftor is one of the modulator drugs that is now funded in NZ (from March 2020) and is available for those who carry at least one copy of the 'gating' CF gene, G551D.

CFNZ has been working hard with Pharmac officials and the drug company who supply other modulator drugs. From the Port CF registry, we can provide accurate numbers of the different CF genes and which category they fit into.

For example,are the genes minimal function genes, residual function genes, are they gating mutations or do people carry two copies of the most common mutation deltaF508 (now known as c.1521_1523delCTT)? Everyone with cystic fibrosis should be encouraged to know or talk to their medical team to find out which CF genes they carry. The future of cystic fibrosis health care is changing, with the data registry an important tool to provide vital information for decision makers in NZ government and health care.

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