CF News

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WINTER 2020

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CYSTIC FIBROSIS NZ

USEFUL CONTACTS

General enquiries
T: 09 308 9161
E: admin@cfnz.org.nz

Fundraising enquiries
T: 021 1955 7876
E: laura@cfnz.org.nz

Stories for CF News
T: 09 308 9161
E: comms@cfnz.org.nz

Social
Facebook: /CysticFibrosisNZ
YouTube: 'Cystic Fibrosis NZ
Instagram: @cysticfibrosisnz

All magazine correspondence should be sent to:
CF News Editorial Team
Cystic Fibrosis NZ, PO Box 110 067, Auckland Hospital, Auckland 1148, E: comms@cfnz.org.nz

Further information
For more on our support services, information, advocacy, and research, or to learn about cystic fibrosis, visit cfnz.org.nz.

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WINTER 2020

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Welcome to the CF News Magazine

Dear reader,

Thank you for picking up this issue of the CF News magazine. Your support means everything to us.

Reflecting on the period between the last CF News magazine and this issue, it's quite surreal to think of how much has happened in a relatively short space of time.

Over the last six months we've seen many wonderful success stories of how access to Kalydeco is making difference, we navigated the lockdown life (twice, for some of us!) and uncertainties of Covid-19, launched a truly sweaty fundraising initiative, and celebrated Sir Bob Elliott's knighthood.

We welcomed several new families and whānau to the community, and sadly said farewell to a few too. We were truly humbled by the way people with CF and their families have reached out to each other via social media – asking and answering questions, sharing advice, offering support – all during a challenging time.

We amassed a whopping 34,000 signatures on the 'double the Pharmac budget' petition and got to work advocating for access to more precision medications for Aotearoa.

We hope you enjoy reading this issue and enjoy the lovely new design and layout. Thanks to our design partner, Central Station, for their work on this.

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

A very 2020 CFNZ team photo

Thank You

We'd like to thank Grant Davis for capturing the sweaty front cover photo of the epic Grace and Hunter. (grantdavis.co)

REGULARS

News In Brief

CF Awareness moved to May 2021

We're aligning our annual awareness drive with our counterparts in Australia and America by moving CF Awareness Week to May. Our next street collections will take place 28 and 29 May 2021 and we'll be sharing insights on CF throughout the month. This means that we will not be holding our appeal week this August, and there will be no street collections in 2020.

Creon Micro funded in NZ

June saw Creon Micro, a modified-release granule formulation of pancreatic enzymes, funded in NZ. Prior to this many parents and caregivers were portioning out the amount needed from the funded capsules into a child's food. Creon Micro can be measured with the special scoop prior to mixing with food or fluids, for use in infants and children who are unable to swallow capsules.

Professor Bob Elliott receives Knighthood

Sir Bob Elliott was already royalty in the CF world, notably for his work on the newborn screening method, and his huge passion for research into childhood illnesses. His contribution to CF was formally recognised in the 2020 Queen's Honours list. Congratulations, and thank you for the difference you've made to the lives of not only Kiwis, but people living with CF worldwide.

New fundraising initiative launched

In July we launched our new fundraiser – SWEAT4CF! This year-round initiative challenges Kiwis to get sweaty for CF, and raise money for advocacy, research, and support. It's good for your health, and good for the health of people with CF – it's a win-win! Check it out: www.sweat4cf.org.nz.

Nebulised antibiotics residue study enters phase 2

Cystic Fibrosis NZ – Otago Branch is working with the University of Otago to investigate how people with CF are cleaning antibiotic residue from their nebulisers and how this might impact antibiotic resistance. It's hoped the findings can be shared more widely to help raise awareness of antibiotic resistance. Turn to page 28 to read more.

CFNZ partners with Rewardhub

Help us raise funds for free! All you have to do is sign up via our page, browse the shops, (where you will receive normal prices and exclusive discounts!) and a percentage of your shop will be donated for CFNZ, with no cost to you. It's an easy place to shop with popular stores such as TheMarket NZ, MightyApe and Cotton On. Sign up and get shopping, here: rewardhub.co.nz/cystic-fibrosis-nz.

UK to be amongst first to get triple-combo outside of US

Following the European Medical Agency's positive opinion on 26 June 2020, NHS England and NHS Wales have announced an agreement for Kaftrio, the European brand name for Trikafta, for people with CF aged 12 and over. It is expected that the drug will be available as early as September 2020.

Pins proving popular

In March CFNZ created a set of 'compassion pins' aimed to support people with CF and their families to feel more confident, help educate others, and minimise situations where they may be unfairly targeted. These little pins have been flying out the door – we've had to reorder stocks three times! If you'd like us to provide you with badges, please email comms@cfnz.org.nz with your name, address and which badge(s) you'd like.

SWEATEMBER SWEAT FOR CYSTIC FIBROSIS

Push yourself to the limit and help people with CF live a life unlimited

Are you ready to make your sweat count? In early July Cystic Fibrosis NZ launched its new, sweaty, and exciting fundraiser; SWEAT4CF.

As part of this year-round initiative we're asking everyday New Zealanders like you to take on the SWEAT4CF challenge and help people with CF live a life unlimited.

Choose a personal challenge that gets the sweat dripping, set up a fundraising page at sweat4cf.org.nz, and ask your friends, whānau and followers to sponsor you. It's good for your health and good for the health of people with cystic fibrosis - it's a win-win!

To top it off, during the month of September we're challenging Kiwis across the country to go the extra mile by getting super sweaty for SWEATEMBER!

We've got some sweet SWEAT4CF sweat bands for sale to complement your effort, and heaps of resources such as posters, banners, and social copy to help you get the word out.

Get your workplace, school, social club, sports team, or family involved and make your sweat count for CF.

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

The impact of your sweat

Donations raised through SWEAT4CF 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 SWEAT4CF

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.sweat4cf.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.

Need a bit of inspiration?
Meet some of our SWEAT4CF champions

Tear out the SWEAT4CF poster on the next page and hang up at your school, workplace or gym!

Name: Raewyn
Sport: Running
Representing: Auckland

Inspiration: "I started running to relieve stress in my life. My son was diagnosed with CF at age 4. If you are a parent with a child with CF you may be able to imagine what life may have been like for us over those 4 years. I'm sweating for CF to acknowledge the tough times and to celebrate how far we have come. I run to challenge myself to be healthier, to be a model to my son and my daughter to show them that barriers can be knocked down."

Name: Paula
Sport: Karate
Representing: Timaru

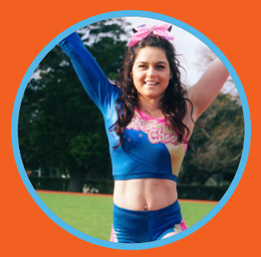
Inspiration: "I've always enjoyed physical activity, but it wasn't until my son was diagnosed with CF at six weeks that I realised how much I needed it. Seido karate plays a huge role in my life helping me to be strong in my mind, body and spirit. The challenges never stop, and you evolve with it in your training to the point of never knowing how far you can be pushed. I sweat for CF because I can. I am lucky to be able to push my lungs to this extent.

Name: Hannah & Chelsey (Sisters), Sport: Endurance
Representing: Otago

Inspiration: "Having a ten-year-old daughter/niece with CF we strive to incorporate exercise into our daily lives, as exercise can benefit us all and is especially important to help keep those with CF healthy. Hills are our favourite - last October we climbed the 343m hill of Puketapu 31 times in 31 days and recently completed Motatapu, our first off road marathon together.

Name: Grace And Hunter
Sport: Gym Workouts
Representing: Whanganui

Inspiration: "My son was diagnosed with CF in the early months of his life. He's nearly 4 now, and doing amazingly. I'm sweating for CF to help my son realise we are stronger than this disease. We will not stop till we have won. Huanter loves to keep active, so we will sweat together for CF to show that no matter what obstacles we come across we will always be together.



Name: Nikki
Sport: Cheerleading
Representing: Tauranga

Inspiration: "Four years ago I was given a new lease on life thanks to a lung transplant. Cheerleading is my favourite sweaty activity as it has dance, stunts, tumble, and jumps. There is no barrier on what you can do - I love to be thrown as high as I can! I choose to sweat for CF because it reminds me how lucky I am to be here."

Name: Chantelle
Sport: Bush Walking
Representing: Wellington

Inspiration: "I love keeping active, especially with my family. We enjoy bike rides, swimming and especially bush walks. My family know how important it is for me to keep fit and make sure I stay motivated. I choose to sweat for CF to keep fit and healthy for my daughters, to show them I will not stop fighting, and that I will not let CF win."

A huge thanks to Mylan, Creon brand owner, for sponsoring Sweatember

Mylan

SWEAT
FOR CYSTIC FIBROSIS

Support me as I take on the SWEAT4CF challenge!

Cystic fibrosis is an incurable genetic condition that causes lung infections and other organ complications.

We can help improve the lives of people with CF and aim to find a cure.

DONATE or REGISTER
sweat4cf.org.nz

CYSTIC FIBROSIS NZ

Innovative start to 2020

This year's Mark Ashford scholarship winner is Georgie Northcoat from Christchurch. We last caught up with Georgie in 2019, after she received a CF Achievers' Award for Leadership.

Since then, the 21-year-old has graduated with a Bachelor of Applied Science from Otago University with a double major in Textile Science and Marketing. This year she's continuing with full-time study and is working towards a Graduate Diploma in Information and Communication Technology (ICT), majoring in Information Systems.

During 2019, Georgie developed a passion for sustainability. In November she travelled to Shenzhen, China, and worked on a project aimed at turning textile waste into insulation. She hoped to continue work on the project this year, combined with full-time study.

Pandemic changes focus

However, with the impact of COVID-19 on offshore manufacturing and maintaining the supply chain, Georgie realised she needed to pivot her entrepreneurial ideas.

"I decided to focus on synthetic textile waste, specifically microplastic pollution from the shedding of synthetic fibres during washing," Georgie says.

"The problem of microplastics in our oceans and soils is well known but there aren't many solutions available which address the issue," she says.

This year Georgie founded Lynk Solution, a company committed to minimising the amount of plastic pollutants entering our wastewater. Georgie developed the Lynk Microfibre Filter, a filter that's fitted to an existing washing machine and filters out 90% of microfibres during each wash.

Georgie and her small team aim to launch the product to market in 2021.

Winning the scholarship

It was third time lucky for Georgie, who also applied for the scholarship in 2018 and 2019.

"I didn't know if I'd be successful this year, so after I applied, I put it to the back of my mind. I thought in due time I'd find out who the recipient was, so I was absolutely stoked when CFNZ called to tell me it was me who'd won," Georgie says.

Georgie plans to use the scholarship money to continue to invest in Lynk Solution. It also relieves a bit of financial pressure so she can focus on her studies and develop her business further.

Getting on with life with CF

Georgie keeps fit by playing competitive hockey, skiing and surfing. She also continues to make the conscious decision to live life to the full.

"Living with CF means sacrifices have to be made to complete treatments and exercise regularly. I like to spread optimism about living a full life, despite this."

Only those in the CF community will appreciate the extra effort put in to look after your health while hitting goals in other aspects of life. Watching Georgie achieve all she does, inspires many, and encourages them to live their best lives too.

You can find out more about Lynk Solution at lynksolution.com.

The 2020 Mark Ashford Scholarship was made possible through a generous anonymous donation. Thank you for supporting this wonderful award and for allowing individuals such as Georgie to follow their dreams.

We are absolutely thrilled to share we have another private sponsor for the Mark Ashford Scholarship, who also wishes to remain anonymous. They have kindly shared a few words for us about why they are supporting this award:

"Since becoming a grandparent of a CF child, I have learned so much about this condition and can see how amazing and determined these young people are to reach for their dreams and succeed. I had goals in my life and the more people who said I wouldn't make them, the more determined I became, and I proved them wrong. CF youngsters have so many extra hurdles to cross in their lives, they must need that same dogged determination to achieve and live life to the full. I'm more than willing to help and love to see someone grab that opportunity".

The 2021 Mark Ashford Scholarship opens for applications 1 November 2020 and closes on 28 February 2021. You can visit www.cfnz.org.nz/get-support/financial-assistance-and-awards to apply online, or get in touch with us for a paper application.

CF Achievers' Award recipients 2020

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, a total of nine individuals with CF have been recognised for their optimism, perseverance, and achievement in their chosen award category. We were delighted with the number and standard of the applicants.
A huge thank you to Mylan, makers of Creon, for sponsoring this award.

Renee Hill, 36 years, Education

Rotorua-born Renee, moved to Wellington to study a degree in alcohol and addiction counseling. Due to the challenges of life with CF, Renee didn't get to finish high school; it is through much hard work and determination, that Renee is achieving what she is today.

"When I found out I was a recipient, I was overwhelmed, happy and very appreciative because times are tough when you are a student earning little to no income."

The next step for Renee is to become an alcohol and addiction counselor, as well as immersing herself into further training, and picking up a te reo Maori paper.

Jessica Scott, 31 years, Leadership

Thirty-one-year-old Jess lives in Auckland with her partner working in TradeMe's marketing team. Jess' friends encouraged her to apply for a CF Achievers' Award, after a tough year losing her much-loved cat, Missy, all while continuing to be a leader and mentor to those she worked with, and delivering on her goals.

Grateful to be one of the 2020 recipients of the Award, Jess said "I found out the first week of going into lockdown where there was a lot of uncertainty for me and my partner, so it was exciting to get a piece of good news."

Jess has a dream of one day reaching Everest Base Camp. "I have agreed with my CF team to complete some high altitude climbs in New Zealand first before I attempt anything, just so we can monitor my health in these situations." We hope to be sharing another success story in the coming years about Jess conquering her greatest goal!

Jake Gawn, 21 years, Sport

Jake has recently completed a Bachelor of Applied Science, majoring in sports technology at the University of Otago, and is planning on starting his honours study next year.

Jake has always shown a keen interest in sport, and competed in the Challenge Wanaka half ironman-length race earlier this year. While he was training, Jake also set up a page to raise awareness and funds for CFNZ. Out of the generosity of his friends and family, Jake ended up raising $7200.

"The award helped with some equipment and a swim membership so I can keep training."

Jake enjoys competing in the half ironman distance races, and hopes to continue to enter these races and see how far he can go with it. "It's good for my health, which is a bonus!"

Christopher Headey, 21 years, Education

Christopher is in his third year studying law at the University of Waikato. He has a passion for public speaking, academia, motoring and music.

For Christopher, 2019 was a particularly challenging year, with it being academically demanding, coupled with tackling new CF complications, ENT surgery and ongoing antibiotic therapies. Despite this, Christopher excelled in his studies. He is delighted and grateful to be a recipient of the CF Achievers' Award, and will use it to support his hobbies and study this year.

"I look forward to pushing myself further, with my sights set on finishing my degree by 2021. I strive for balance in my life, and I am certain there will be no end of fun and adventures in store for me along the path to my goal."

Tayler Payne, 13 years, Sport

Tayler is in year 9 at Botany College, and is a keen go-karter. He's been racing for about three years. They travel all over the North Island to race in competitions. Tayler's dad is his mechanic and he is trained by Josh Bethune from Right Karts.

"Mum suggested I apply for the award so that I could show others how CF doesn't stop me from doing what I really enjoy."

Tayler plans to spend his award money on a new helmet. "Currently Mum and Dad are my sponsors so we need to find some. :) Thank you CFNZ for choosing me."

Timothy Donkin, 17 years, Leadership

Tim is an exceptional young man who has faced a huge year with resilience and strength. Tim underwent a liver and pancreas transplant, the second of its kind in New Zealand, during Covid-19 Alert Level 4. While waiting for the miracle call for his transplant, he continued to be an excellent leader, being named a Prefect this year. Tim has a passion for public speaking, and has won his school speech award twice, and was chosen to represent his school in the O'Shea Shield Competition in Oratory and debating. Tim has also captained several cricket teams over the years.

"Tim's perseverance and determination he's always more than caught up on anything he misses out on so he still achieved his goals," says his mum, Louise.

Grateful for the CF Achievers Award, Tim will spend it preparing for university next year.

Mylan

Thank you to Mylan for generously sponsoring the CF Achievers' Awards

Layla Collins, 16 years, Sport

Layla lives in Pukekawa with her parents and younger sister who also lives with CF. Layla has been riding horses and has been playing Polocrosse during summer for the last five years. This year, Layla was selected for the NZ Polocrossse junior under 16s team which travels to Australia to compete. They were lucky enough to be able to hold their inbound tour, where the NSW under 16s competed in NZ, but unfortunately COVID-19 meant Layla's team could not get over to Australia for the competition this year.

"I was thrilled to be a recipient of the CF Achievers' Award for sport. I had intended on using the award money to assist with travel costs to Australia, hopefully that will happen next season!"

Layla also won a scholarship to stay with a host family in Australia and play some of the Australian season.

Angus Drumm, 20 years, Leadership

Angus is currently in his third year of his law and art degree at university. Angus has a strong passion for politics, and he leads the University of Auckland's Public Policy Club, and is also involved with the Labour Party. He was campaign manager for a local board election team last year, and this year, is the assistant campaign manager for the Hon Phil Twyford.

Angus also has a love of travel, basketball, reading and nice food!

"I enjoy leading teams, coming up with a strategic direction for my team and then executing plans. I currently hold leadership positions in several organisations and I'm constantly trying to improve my skills and abilities in this area."

Angus has a young leaders conference coming up that he is attending as a member of the U.S Embassy Youth Councillor programme.

Ashlee Sharp-Crowley, 9 years, Sport

At nine years old, Ashlee is our youngest recipient this year. Ashlee lives in Lower Hutt with her mum, 2 year old brother Connor.

In 2019, Ashlee received Riding for Disabled 'Horse Rider of the year' out of 108 other riders. "I had been working really hard over many years to achieve my long term goal of being selected to train with the Special Olympics Riding team to compete at the next summer Special Olympics, which finally came true at the end of 2019."

Ashlee said at times her goals were hard to see, and wondered if they were unrealistic, as the CF challenges are continuous, but receiving a CF Achievers' Award has reminded her that anything is possible if she wants it enough, CF or not!

"I've learnt that I am very stubborn, and when I set my mind to something, I surprise myself at how capable I can be."

The 2021 CF Achievers' Award opens for applications 1 November 2020 and closes on 28 February 2021. You can visit www.cfnz.org.nz/ get-support/ financial-assistance-and-awards to apply online, or get in touch with us for a paper application.

MAKE A WISH BOXES

Sometimes, our gorgeous little ones do not understand the concept of a wish, or may not have dreamed one up yet. This is when the Wish Boxes come in, because children like Quinn here, are totally deserving of a treat!

Once we receive the Wish Application Form and medical eligibility is signed off, we will contact the parent/legal guardian to find out a bit more about the child. For example, their favorite colors, stories, characters etc. A fun box of toys and goodies is then put together and sent to our volunteers who arrange delivery. It feels a bit like Christmas Day! Make-A-Wish will always ensure that a gift for siblings is included.

Once your child is a little older and can have more input in their wish, we would be delighted to look at granting one for them, as long as Make-A-Wish receives their application before their 18th birthday. A new Wish Application Form is required for this.

Please visit our website www.makeawish.org.nz to download an application form or call the Make-A-Wish office of 09 920 4760 and one of the friendly staff will send you out one.

4 year old Quinn being delievered his Wish Box from Batman himself!

Make-A-Wish NEW ZEALAND

A dream adventure trip before lockdown

The Covid-19 pandemic may have scuppered any plans of overseas travel for the foreseeable future, but many of us are already dreaming of travelling or reflecting on past holidays.

One such dreamer is Kelcie, who celebrated her 21st birthday and the end of three-years of studying, with a dream winter holiday not long before lockdown.

"After finishing my Bachelor of Health Science, majoring in Occupational Health and Safety, I was ready to prove to myself that I could take on the world," Kelcie explains. "So, I went to Canada on a solo mission to ski the Rocky Mountains!"

Gretchen (CFNZ Social worker) and Cath (CF nurse) helped Kelcie get set up with what she needed to travel, including getting a battery nebuliser, and helping with lists of meds and medical history in case she needed them at customs.

"I took enough meds for a few days on the flight, and enough for the whole trip and an extra few days' worth in my suitcase.

"I did my nebs in the morning before leaving for the airport and again once I got to my hotel in Vancouver. I wasn't 100% perfect with doing physio throughout the trip - but I told myself that skiing all day was a good substitute," Kelcie muses.

"After arriving in Vancouver, I jumped on a TopDeck tour (an 18-35 tour group) where I got to make some ultimate new snow friends. We went mountain hopping - from Whilster Blackcomb, to Big White, to Lake Louise and Sunshine Village. Spending a few days at each place to hit the slopes.

"I finished up my trip with a week at Panorama Resort, where I met up with a couple of school friends that worked there. It was a very different social experience than heading to the beach on the weekends!"

During Kelcie's trip, the average day was a cosy -8 degrees Celsius, and the coldest day a numbing -18 degrees Celsius.

"I had never experienced weather so cold or been at such high altitudes," Kelcie continues. "But what really took my breath away was the scenery. It's just like something out of a David Attenborough doco with the snow-capped peaks, endless trees, and frozen lakes.

"I got to ski some world-renowned ski resorts, ice skate on Lake Louise, watch an Ice Hockey match, go snowmobiling, go ziplining, have endless snow fights and partake in plenty of road trip karaoke. My body was pushed to its limits and I luckily only came home with minor injuries!

In preparation for the holiday, Kelcie trained for months, doing everything she could to be in good physical shape.

"It was important to me that I made sure I was in peak shape so I could make the most of my trip. And my goodness it paid off, with 10 days of skiing and plenty of partying - it was undoubtedly the best experience of my life.

"It really showed me that I was my only barrier and that all the efforts I'd put into mastering my physio routine and doing all the extra exercise was totally worth it. It motivates me to push harder to go to do more adventure trips.

"I'm so thankful that I just went and did it when I did - with the world in lockdown now. Now is the time for me to keep studying, try stay on top of normal life and keep dreaming up the next trip - for when the time comes..."

Kelcie's top tips for an adventure holiday

1. Master your physio routine and fit in some extra exercise if you can. It won't always go to plan, but ultimately it made a big difference to my ability to make the most of my holiday.

2. High-quality base layers (warm layers) are essential. Getting cold makes it harder to breathe so having proper alpine thermals saved me from burning energy trying to stay warm.

3. Take a compact bag/ fanny pack on all day trips and stock your inhaler, Creon, and dense snacks (nuts, picnic or moro, a protein bar or muesli bar). That way if I felt chest tightness, my feel blood sugars drop, or just want to eat - you are covered from all angles. I kept my bag tucked in under my ski jacket so wherever I was on the mountain I was good.

There's still time to join the Beam team

Grab your spot on a 3-month pilot programme with the online exercise platform Beam.

Beam helps people with CF get active through on-demand and live classes, group support and handy resources.

Sign up for free if you're aged 16 and over with CF, or you're a mum with a child with CF aged under 3. Beam offers a mix of exercise disciplines including high-intensity interval training (HIIT), yoga, strength and conditioning and Pilates for all levels of fitness. The classes are led by specialist and qualified instructors who either live with CF or work with people with CF.

You need to complete a quick anonymous survey about your general wellbeing at the start and end of the programme. We'll use this data to report back to the Health Promotion Agency who is generously funding the programme through a grant.

Sign up today

1. Send an email to BeamTeam@cfnz.org.nz to register your interest and receive the sign up survey.

2. Once we've received your survey, you'll be sent an access code to access the Beam website.

3. Go to the Beam website and sign up using the access code.

4. Get involved and start achieving your goals.

Stay active to improve your mental wellbeing

As part of the grant we're developing a new guide for adults. From the initial feedback we've received from adults with CF we know how important mental and emotional wellbeing is for coping with the challenges of CF.

The *Five Ways to Wellbeing* from the Mental Health Foundation are evidence-based actions you can build into your life to help improve your mental.

Connect: Develop and nurture your relationships with people.

- Become involved in groups; join your local craft, sports, choir, hobby or book club and enjoying singing, sewing, playing a card game, visiting gardens or croquet on the lawn together.
- Gather some friends for a DVD evening; ask people to share a film they like.
- Grab some mates and get into the great outdoors – go on a bush walk, go surfing or go mountain bike riding.
- Take time to read your local newspaper or newsletter – find out what's going on in your area, such as music or cultural performances, then organise a group outing.

Give: Give your time and presence

- Take opportunities to support and advocate for groups, friends, family or neighbours in need.
- Organise or promote random acts of kindness days at school, work or when you're out with a group.
- While driving, stop to let a car into the traffic.
- If you have fruit trees pop your excess fruit out on the street with a "help yourself" sign.

Take notice: Remember the simple things that give you joy

- Be mindful of the first mouthful of food you eat. See if you can really pay attention to all the flavours and textures of the food, the act of chewing and the act of swallowing. During the following meal, see if you can be aware of the first two mouthfuls of food, and so on.
- Take the opportunity to sit quietly in a busy place like an airport or a mall and notice the interactions between people.
- Take notice of the night sky. Be aware of what phase the moon is in and how the visible constellations change throughout the year.
- Go for a bush walk, try to identify the different animal and plant species you see and photograph them while taking the time to really notice what you are photographing.

Keep learning: Try something new or rediscover an old interest

- Learn something you don't know about the area in which you live by checking out the local notice boards for interesting talks and events.

- Discover the name of the iwi, hapu, maunga and awa of the place you live.
- Memorise a new word every week. Practice using it among friends and family.
- Put your hand up for a new challenge/training in your workplace to broaden your knowledge.

Be active: Do what you can and improve your mood

- Try a 'Have A Go day' with a local sports group. Look out for what's on offer, as often, free equipment and tuition is provided.
- Participate in a fun run/walk to raise money for charity.
- Try tai chi classes for strength, balance and mental wellbeing.
- Find out the most popular sport among your colleagues and then organise a match or tournament for staff.

Reference: Wellbeing, Mental Health Foundation

Our Adult Wellbeing Guide will build on these actions for people with CF in New Zealand.

Use the Five Ways to Wellbeing as part of daily life with CF

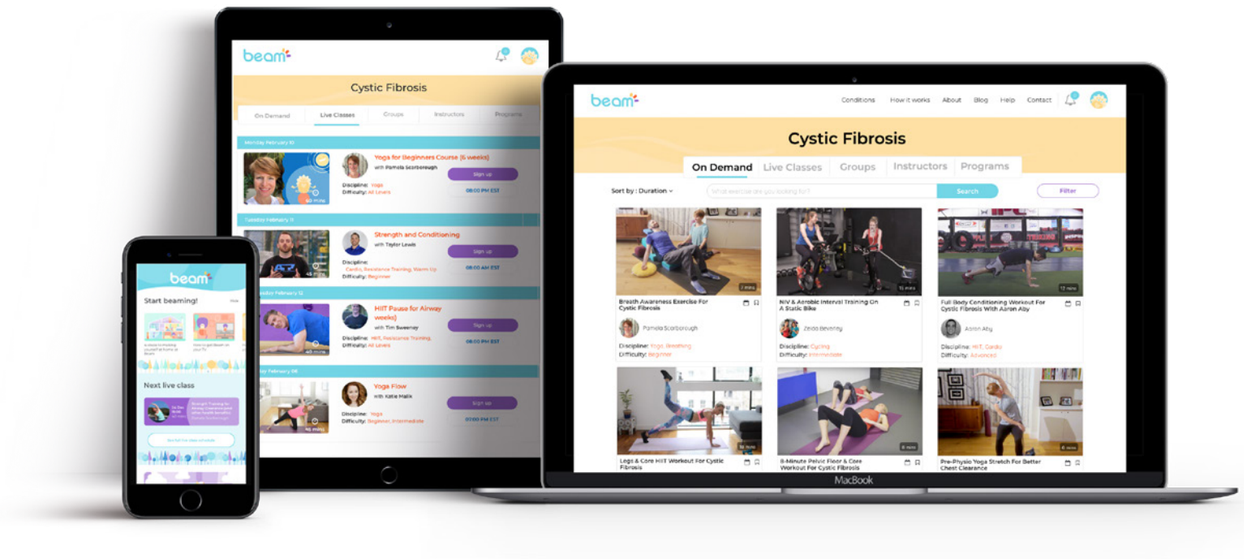
Connect: Sign up to Beam for 3 months and connect with qualified instructions and other people on the programme.

Give: If you're ready to be part of something bigger, sign up for our Sweatember and give back to your CF community.

Take notice: At the start and end of Beam you'll complete a wellbeing survey. Take note of how doing Beam classes regularly makes you feel, both physically and mentally.

Keep learning: Try a new type of exercise – there's plenty to choose from on Beam. Or set yourself a new challenge during Sweatember to try an activity or way of exercising.

Be Active: Set some personal goals you want to achieve using Beam's online programme and make a plan with your physio or fieldworker about how you'll achieve them.



FIVE WAYS TO WELLBEING

INTRODUCE THESE FIVE SIMPLE STRATEGIES INTO YOUR LIFE & YOU WILL FEEL THE BENEFITS

CONNECT

TALK & LISTEN,
BE THERE, FEEL CONNECTED

BE ACTIVE

DO WHAT YOU CAN,
ENJOY WHAT YOU DO,
MOVE YOUR MOOD

Give

YOUR TIME, YOUR WORDS, YOUR PRESENCE

TAKE NOTICE

REMEMBER THE SIMPLE THINGS
THAT GIVE YOU JOY

KEEP LEARNING

EMBRACE NEW EXPERIENCES,
SEE OPPORTUNITIES, SURPRISE YOURSELF

Mental Health Foundation OF NEW ZEALAND
mauri tū, mauri ora
www.mentalhealth.org.nz

Ask the experts

Ask the Experts is the CFNZ expert advice service for people with CF and their families. Questions are answered by a panel of clinicians who specialise in different areas of CF, as well as our very own CF Fieldworkers who provide holistic support to the CF community.

Please note, if you have any queries or concerns about any aspects of CF you should contact your CF team in the first instance.

Submit your questions to comms@cfnz.org.nz. Although your query and the response may appear on the website or in this magazine, your name and personal details will always remain confidential and will not be published.

Q - What can you use for easing stomach pain after not having quite enough Creon?

A - Ensure correct amount of enzymes given for each meal. Discuss with the dietitian for amount needed. If constipation is the problem need correct dose of enzymes and may need a laxative as well - eg Lactulose or Movicol.

Q - What's the best way to travel and do nebs, such as on a big flight? How should we keep Pulmozyme cold during the flight?

A - Put it in a small chiller bag the night before and keep it in the fridge until you are nearly ready to leave, then place an ice pack/slikka pad in the bag to keep it cool.

Q - What's the best way to sterilise PEP devices? I've been washing it with hot soapy water then sterilising it in an anti-bacterial solution (Milton). My issue with this solution is that it smells like chemicals and I feel like it isn't the best to be breathing in.

A - Check with your physio, some can go in the top shelf of the dishwasher. Rinse after taking out of chemical solution. They can sterilised in a pot of water at a rolling boil for a couple of minutes or you could use an electric or microwave baby bottle steriliser.

Q - My CFer's daycare asked if it's safe to play with clay and playdough. When asked if it would be fresh, they said they change it every few days or once it gets too dirty.

A - Play dough is a tricky one - can the child have her / his own playdough? All of the different children moulding the play-dough each day equals potential for 'bugs' to collect in the dough. If it is homemade the recipe usually uses a lot of salt and this may inhibit the bugs growing. It's best if the child has their own supply of playdough and made up fresh every few days.

Q - I am a person with CF and I'd like to be an organ donor when the time comes. Is this possible?

A - Absolutely, each case will be taken on an individual basis. Lungs and other organs damaged by CF may not be able to be donated but there will still be plenty that can be put to good use.

Q - My child has been on oral antibiotics a lot - pretty much since he was eight weeks old and has been on a prophylactic since July (he's 18 months now). His teeth have started to appear weak and yellow. Is this connected, normal, and what can we do to remedy?

A - Talk to your doctor or dentist about options available.

Q - My CFer has just started on Tobi. They have been breathing on it for 40 minutes and there is still heaps left to go. Is this normal?

A - Depends on how it is given. If the child is using a Pari Sprint and an antibiotic filter attachment (and mouthpiece) should take about 15 minutes. If a younger child using a mask takes a little longer. Shouldn't be taking 40mins - maybe the compressor needs checking for the output - if sluggish output will take a long time. The clinics will check the output and give a dose with the nurse to see how it is given.

Q - I'm looking to get Invisalign or clear braces for my CFer. Which would be a better choice regarding bacterial issues?

A - Discuss with your CF doctor. For any teeth hardware you'll need to clean teeth properly and rinse after sticky foods.

Q - My CFer wants to visit the mud pools in Rotorua. Is this a big no?

A - A visit to mud pools is OK but not swimming in the warm / hot spa pools in Rotorua.

Q - What are your thoughts on a CFer eating camembert cheese, with regards to the bloom/mould on the outside? Is it safe?

A - This is absolutely fine for non-transplanted people with CF.

Q - My CFer refuses to wear the given nose clips because they hurt, which means she is now mucking about during treatment and not getting the full dose. Is there anything else available, which works just as well but doesn't hurt so much?

A - Wearing a nose-peg ensures the person is getting best treatment - persevere!

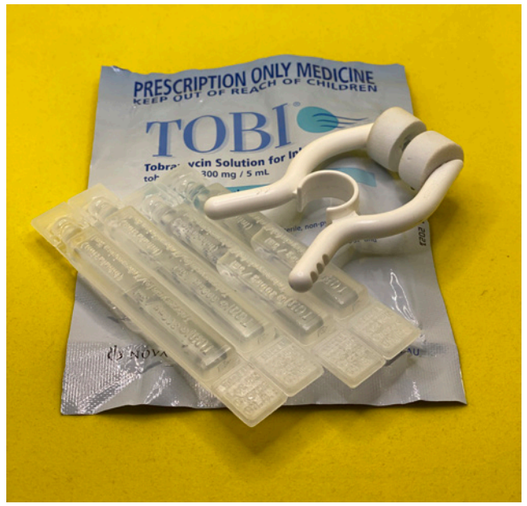
Q - What's the Freestyle Libre blood glucose monitoring system like? And where is the best place to purchase from, and the costs associated?

A - Discuss with your CF team. Sometimes they can supply an initial reader and sensor. Most phones can download the app so a reader is no longer required. The current cost is $85.16 plus $6.50 handling no matter how many you order, and $13.75 gst It is easy to use and cuts out the need for finger prick testing. Each sensor lasts 14 days.

Q - I'm looking to enrol my CFer in a daycare but I am a bit confused about what to expect of the daycare in terms of CF support. I've been told by multiple centres they've never had a child with CF and have asked me how it could work. Any tips?

A - The CF at School resource would be useful for this age group and offers lots of practical tips and experiences. Often your local CF nurse or CF Fieldworker is happy to visit the centre and discuss how it could work too. Visit www.cfnz.org.nz/life-with-cf/school for more information.

A huge thank you to the clinicians and CF fieldworkers who kindly provided answers for this feature.



“It’s been 85 days since I last coughed”

Ed Lee, an adult with cystic fibrosis from Wellington has taken the extraordinary decision to self-fund Trikafta. Here, Ed shares how life-changing the medication has been for him, and why Trikafta is needed for all Kiwis with CF.

“I like to think of my story as the typical cystic fibrosis (CF) story. I am a 37-year-old guy trying to live a normal life, whilst battling CF behind closed doors,” Ed reflects.

“In the past I hadn’t been that open about my CF. I hadn’t spoken to anyone else with CF before. I liked to believe that my health was strong enough without the need of a CF support network. In hindsight, this attitude was detrimental to not only my physical health, but my mental health.

“I was running around, drinking, exploring and generally taking my health for granted. I lived like this up until November 2019 when I finally had the scare I needed to knock me back into reality. I saw my FEV1 fall from my usual 36% to its lowest level ever (28%) and I began to struggle. I remember lying in my hospital bed, with my wife KP by my side and wondering whether my time was nearly up.”

As Ed struggled with what his future might look like, the news broke that Vertex’s triple-combination therapy, known as Trikafta, had received FDA approval.

The FDA granted the medication the status of Priority Review, Fast Track, and gave it the Breakthrough Therapy designation, three programmes designed to expedite the approval process for drugs considered as ‘game-changers’. Trikafta was reviewed and approved within three months’ time, well ahead of the March 2020 goal date.

“Trikafta sounded incredible, demonstrating FEV1 improvements of an average of 10-14%,” Ed explained. “As Trikafta began to be prescribed in the US over the following weeks, stories of how this drug was changing lives flooded social media. You couldn’t help but notice.”

The breakthrough treatment was widely heralded as having the potential to turn CF from a life-threatening condition, to a manageable condition.

Earlier Vertex therapies, such as Kalydeco and Orkambi, only work on very specific genotypes and therefore only effective for a limited number of people. Trikafta has the potential to work for up to 90% of people with CF.

“You see, CFers live a battle that, despite everyone’s genuine intentions, most will never understand what it is like. I sometimes hear that having CF is like breathing through a straw. I don’t know who came up with this analogy, but I find that it simplifies the real struggle,” Ed continued.

“It isn’t just like breathing through a straw, it is constantly living in fear that we won’t live a life beyond 40, let alone 30. It is about living with the constant stress that if we don’t get up each morning and fight then we will end up in hospital or worse.

“It is about a relentless struggle with lung infections, liver problems, pancreatic enzymes and insulin level problems. Not to mention the way this stress profoundly impacts all other areas of your life, or constantly feeling let down by our underfunded health system.

“Don’t even get me started on how poorly funded medicines are here in New Zealand.”

As the success stories increased, Ed took the extraordinary decision to self-fund Trikafta. But as an unfunded drug in New Zealand, it would come at a price tag of over $120,000 NZD for three months’ supply.

“But we just had to get our hands on it before it was too late. After discussions with my family, and some financial help, a quick trip to San Francisco was planned. We flew over and all of a sudden Trikafta was in my hot little hands. I couldn’t believe it. KP caught that momentous moment on camera and posted the photo onto my social media.

“Before I took my first dose of Trikafta I still believed that it wouldn’t work for me. I believed that I wouldn’t be as lucky as others, and thought that people were simply talking the medicine up, as how could a simple tablet make that much difference?”

It’s been 90 days since Ed started taking Trikafta. “It’s been 85 days since I last coughed. Think about that for second, since taking Trikafta I have not coughed for 85 days - not once! Trikafta has completely and utterly changed my life, all within 24 hours of taking my first tablet.”

Ed described what is often known as ‘the purge’, where he coughed up 37 years of the thick, sticky mucus clogging up his lungs.

“As my lungs were hydrated for the first time, all of the clogged air pockets in my lungs opened up and suddenly I could breathe. That feeling is incredible.”

Since starting Trikafta, Ed has eliminated three infections and his FEV1 is now 48%.

“My ABPA has gone (a hyper-sensitivity to the fungus Aspergillus) and my usual infections of Staphylococcus and Achromobacter are virtually undetectable,” he said.

“We are winning the fight, but after years of lung damage, there is a real risk that I could get an infection again. But Trikafta has given me hope. Hope for the future, hope for your friends and family, and hope that your life will one day be free from hospital admissions, lung infections and the constant fear that you will leave your loved ones behind early. It has enabled me to run again, to keep up with other people my age and to start ‘actually’ living a normal life, without constant stress and fear.

“Saying that I feel extremely fortunate to be able to obtain Trikafta is an understatement. It has saved my life, and I want all people living with CF in New Zealand to be on Trikafta.

“It is simply not good enough to have a government who are trying to lead with compassion to ignore these medical advances and refuse to fund medications which have life-changing clinical results.

“The earlier that people can start on Trikafta, the less permanent damage they will have.”

Read more about advocating for Trikafta in the Advocacy section.

More from Ed

If anyone would like to contact me or follow my story then feel free to reach out to me on Instagram under the username kiwi_cf_warrior where I will be slowly opening up more about my story and to offer support and encouragement to all people living with CF. I am also part of the community advocacy group called ‘Trikafta for Kiwis’ which is working closely with CFNZ to bring this life-saving medicine to New Zealand.

Access for Aotearoa

Campaigning for precision medicines for Kiwis with CF

Trikafta in the spotlight

In Early August Trikafta was thrust into the national spotlight with a public plea for Trikafta by CFNZ Patron Sir Bob Elliott.

Bella Powell, 17, has spent much of her life in hospital and is on the waiting list for a lung transplant. Through their ambassador work with research charity Cure Kids, Sir Bob, Bella and her family have a forged a special relationship.

"I've been treating cystic fibrosis most of my professional life. This is what we've been waiting for and it works," Sir Bob told journalist Patrick Gower, reporting for Newshub.

Knowing that Trikafta might be able to help Bella, he privately paid for a three-month supply of Trikafta at the cost of $100,000 NZD, with "miraculous results".

Sir Bob made a medical breakthrough in the 1970s with the 'heel prick test' that identifies cystic fibrosis at birth. He spent many years trying to find a cure but couldn't.

As an esteemed medical researcher and CF expert, his public endorsement for Trikafta speaks volumes as to its potential impact on the New Zealand CF community.

Trikafta received FDA approval in late 2019 for people with CF aged 12 and over who have at least one copy of the F508del mutation, roughly 90% of people with CF

It is available in the US, and UK and the Republic of Ireland have secured deals with Vertex for Trikafta, in anticipation of its approval in Europe. Trikafta is currently being considered by Australia's regulatory body TGA.

The breakthrough treatment has been widely heralded as having the potential to turn CF from a life-threatening condition to a manageable one.

The exclusive with Newshub generated several follow up items of coverage and piqued the interest of many people across the country.

"We are thrilled to hear that Sir Bob Elliott has paid for a member of the New Zealand CF community to privately receive Trikafta. For Bella and her family this is an incredible gift. But it only exemplifies the inequality of medicine funding in New Zealand, whereby individuals are forced to privately fund modern treatments," says Jane, CE at Cystic Fibrosis NZ.

"We must improve access to modern medications for all New Zealanders who could benefit".

Vertex voices commitment

During the publicity Vertex announced that their plan for Trikafta is to apply to the appropriate government agencies, using their available pathways, to achieve PHARMAC-funded access in the fastest possible way.

They indicated that they are open to discussing further with PHARMAC potential alternative innovative mechanisms to achieve this.

In a mid-August meeting, Vertex reiterated that they are fully committed to exploring access for people with CF in New Zealand to future Vertex medications. As a next step they plan to engage with the appropriate government agencies to gain their feedback into this process.

Here's a statement Vertex provided us:

"At Vertex the patients we serve are at the centre of everything we do, and we have been relentless in our pursuit of innovative therapies for cystic fibrosis for more than 20 years. Our medicines have changed the cystic fibrosis treatment paradigm by treating the underlying cause of the disease.

After a number of years of discussion with Pharmac, we were thrilled when they decided to fund Kalydeco® (ivacaftor) for people with cystic fibrosis who have the G551D mutation or other class III gating mutations in the CFTR gene from March this year. As a result, we now have more than 30 patients who are benefitting from this treatment in New Zealand.

Our global mission is to secure the broadest sustainable access supported by the scientific evidence available, to ensure the greatest value and benefit can be delivered to patients and healthcare systems. We are fully aware that there are many more people with cystic fibrosis in New Zealand that could benefit from our other treatments.

Please be assured that we are fully committed to exploring access for people with CF in New Zealand to future Vertex medications. As a next step we plan to engage with the appropriate government agencies to gain their feedback into this process.

We do however recognize that the process of registration and funding medicines in New Zealand is complex. To support us in this, Vertex has recently appointed a Manager to understand the priorities and needs of stakeholders to prepare for future Medsafe submissions for Vertex medications.

We will keep Cystic Fibrosis NZ updated on progress on an ongoing basis".

Addressing the wider issue

Access for Aotearoa is the name for Cystic Fibrosis NZs ongoing advocacy campaign to secure access to precision medications for all Kiwis with CF. This includes modulators such as Symdeko, Trikafta and any future medications, from both Vertex and other manufacturers as these become available.

The group functions as a collaborative working space for the CF community, supporting groups and anyone interested in improving treatment access for Kiwis with CF. Visit www.facebook.com/AccessforAotearoa to join.

The CFNZ Board and management are committed to accessing Trikafta in the quickest possible timeframe. CFNZ actively seeks the best possible medications and care for all kiwis with CF and advocates to address issues that stop this being achieved.

As with the campaign for Kalydeco, getting Trikafta and other precision medicines is made infinitely more difficult due to the medicines funding model. The model means New Zealand ranks amongst the worst in the OECD for access to modern medicines, and this must change.

Recently CFNZ CE Jane Bollard attended the Medicines NZ dinner hosted by the Minister of Health, Hon. Chris Hipkins, with a panel discussion on Medicines Inequity - the problems & potential solutions.

Medicines NZ advocates for better access for NZ patients to modern medicines through the public health system by supporting:

- An updated to the medicines policy and strategy, which CFNZ participated in the consultation process.
- Collaborative implementation of a rapid access scheme for modern medicines
- A defined separate medicines budget appropriations process leading to better transparency and governance of the budget.

We're continuing to collaborate with Medicines New Zealand, Patient Voice Aotearoa and Rare Disorders NZ, to ensure that the 'access' conversation remains front and central to key stakeholders. Thank you to the thousands of people who supported the 'Reform PHARMAC and Double the PHARMAC budget' petition following recent publicity.

Christine Perrins, who has worked closely with Jane over recent months on strategic advocacy issues including input into a new medicines policy and strategy, will be putting her focus into the development of the Trikafta campaign strategy and to helping CFNZ address some of the wider issues that hamper access to medications for kiwis with CF.

Trikafta for Kiwis group formed

CFNZ is pleased to be working with the new Trikafta for Kiwis group started by Carmen Shanks.

Eddie Porter, who led the Kalydeco for Kiwis campaign, has also joined, together with other adults with CF and some partners.

CFNZ and Trikafta for Kiwis are working together with the objective of securing public funding for Trikafta for people with CF in New Zealand.

As a person with cystic fibrosis, Carmen's motivation to advocate for access to the latest medications, for New Zealanders also comes from knowing life-changing medications are available, but inaccessible for most Kiwis.

"After seeing the success of the Kalydeco for Kiwis campaign, and also seeing the amazing results patients overseas have with CFTR modulators, I got to the point where enough was enough and I wanted to do my bit," Carmen says.

"The turning point for me was a particularly rough hospital admission. I was missing my son Lachie immensely. Lachie was very upset and couldn't understand why I wasn't home and my husband Chris was also having a hard time juggling childcare and continuing to work" she says.

"I knew there was a medication out there which could help improve my quality of life and yet, it was simply out of reach because I live in New Zealand."

Carmen has launched a petition to urge the government to publicly fund Trikafta. Sign it here: https://www.parliament.nz/en/pb/petitions/document/PET_99952/petition-of-carmen-shanks-publicly-fund-trikafta-medication

You can access their updates by searching for 'Trikafta for Kiwis' on Facebook and Instagram, or by messaging trikaftaforkiwis@gmail.com.

The Shanks Family

What exactly is Trikafta?

Trikafta is a prescription medicine used in the treatment of cystic fibrosis in patients 12 years and older who have at least one copy of the F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) protein. As a result of these defects, the CFTR proteins don't work the way they should. Trikafta adds Elexacaftor to Tezacaftor and Ivacaftor to target the CFTR protein defects caused by the F508del mutation. By binding to different places on F508del-CFTR proteins, Elexacaftor and Tezacaftor work together to help more proteins reach the cell surface. Ivacaftor helps the CFTR proteins stay open longer at the cell surface.

What about under 12s, and the other 10%?

Vertex is working to get Trikafta to younger children and continuing to work on genetic approaches to help those who need something other than a small molecule to treat their condition. It's study of Trikafta for those aged six to 11 is ongoing, and it is investigating gene therapy as an option to treat the last 10% of people with CF.

ADVOCACY

Other Advocacy News

Funding approved for Creon Micro

From 1 June 2020 PHARMAC began funding Creon Micro. We first submitted an application for its funding in late 2015 and submitted a letter of support in April 2020 during PHARMAC's open consultation period, so we were delighted when funding was approved.

Creon Micro is a modified-release granule formulation of pancreatic enzymes. This formulation allows for smaller doses of the medicine to be administered. Until now, many parents and caregivers were portioning out the amount needed from the funded capsules into a child's food. This led to wastage of the leftover medicine and possible risk of giving the child the wrong amount.

A special scoop is used to measure out the right amount of Creon Micro for babies and children who can't swallow capsules.

PHARMAC estimates about 95 people with CF every year will benefit from Creon Micro.

TOBI Podhaler

In February 2018, we submitted an application to PHARMAC for TOBI Podhaler to be listed on the Pharmaceutical Schedule.

The application was reviewed at the Pharmacology and Therapeutics Advisory (PTAC) meeting in February 2020.

The recommendations from the February PTAC meeting are:

- The Committee recommended that TOBI Podhaler be listed as cost neutral to the TOBI inhalation solution, accounting for any offsets to the health sector.
- In making this recommendation, the Committee considered that improvements in quality of life associated with the TOBI Podhaler compared with the nebulised tobramycin solution were primarily due to reduced treatment burden, and increased treatment satisfaction and patient preference. The Committee considered however that there is a lack of evidence to support substantial improvements in adherence and other important health outcomes.

The application now moves into the under-assessment stage. We'll continue to advocate for its funding throughout the application process.

Our work with Patient Voice Aotearoa

CFNZ continues to support the work and vision of Patient Voice Aotearoa (PVA). PVA was formed as a collective of patients, caregivers, whanau, advocates and charitable organisations, campaigning together for the rights of New Zealanders. In late March, Jane Bollard facilitated a planning day with PVA and other support charities to develop a strategy and action plan focused on the Double the PHARMAC budget petition. We know it's important to harness the collaborative voice of all charities seeking medications and to be clear about what the 35 agencies supporting PVA could action to get results.

Immediately after this meeting we went into COVID-19 lockdown and PVA developed its open letter to Jacinda Ardern for increased funding of medications for people with pre-existing health conditions across a range of rare disorders, chronic illnesses and cancers.

During April, Jane met with PVA representatives again to discuss progress following the open letter. PVA reported good success with getting stories in the media. Our advocate at the time Lisa Woods continued to work with PVA to develop a communications plan, including charities talking with local media about the impact of COVID-19 for vulnerable people.

PVA is keeping a watching brief on the impact of Covid-19 on PHARMAC's funding of medications and we'll support any collective action needed.

Support Rare Disorders NZ petition

A new petition needs more signatures to help ensure New Zealanders with a rare disorder have access to the best healthcare.

The Petition of Sue Haldane for Rare Disorders NZ: Develop a National Rare Disorder Framework, urges the Government to acknowledge the universal challenges faced by people living with a rare disease, and the unfairness within the current system, by committing to the development of a New Zealand National Rare Disorder Framework.

Please take a minute to sign the petition and share with your networks before it closes on 1 April 2021.

https://www.parliament.nz/en/pb/petitions/document/PET_94998/petition-of-sue-haldane-for-rare-disorders-nz-develop

Research in New Zealand

Nebulised antibiotics residue and resistance research study

Last year, Cystic Fibrosis Otago began working with the University of Otago to investigate how people with CF are cleaning antibiotic residue from their nebulisers and how this might impact antibiotic resistance.

Phase 1 of the research study has been completed and funding has been confirmed for Phase 2.

The objectives of phase 1 of the project were to:

- determine the types of nebulisers and inhaled antibiotics being used in the home by the CF community
- investigate the methods PWCF or their carers use to clean nebulisers and dispose of residual antibiotic solution after treatment with a nebuliser in the home setting
- develop a test to measure the amount of antibiotic remaining in different nebuliser systems after treatment, and the concentration of that residual antibiotic solution in wash water after nebuliser systems are rinsed in wash buckets after use.

Researchers' findings from a telephone survey of 9 Otago-based caregivers of PWCF who've used nebulised antibiotics found that tobramycin was the most common nebulised antibiotic. In most cases, antibiotic residue was poured down household drains.

A national-based survey followed, with similar findings. Antibiotic residue was either drained directly into the sink, rinsed with water then poured into the sink or rinsed with detergent and poured into the sink, before sterilising.

A Liquid Chromatography Mass Spectrometry (LCMS) assay to detect tobramycin at very low concentrations in nebuliser wash water was developed by University of Otago scientists.

To date, this assay has found approximately 20% of a standard 300mg tobramycin dose was found in the wash water from a rinsed nebuliser after phantom aerosolisation

(i.e. the tobramycin dose was nebulised into a laboratory fume cupboard, rather than inhaled by a PWCF).

Phase 2 will focus on developing a practical nebuliser cleaning strategy (or strategies) to help reduce the risk of antibiotic resistance due to the variable disposal practices that were found in phase 1.

Researchers will share their findings with the CF community and work to co-development recommendations for the safe and practical disposal of antibiotic residue from nebulisers. It's hoped the findings can be shared more widely to help raise awareness of antibiotic resistance.

Why this research is important

Antibiotic resistance is a serious and growing health problem and there's concern within our CF community that rinsing residual antibiotic solution from nebulisers down the sink may be contributing.

Disposal of unused antibiotics into waste water or landfills has been reported to lead to high rates of resistant bacteria in the environment. The Ministry of Health, PHARMAC, district health boards and the Pharmaceutical Society of New Zealand recommend unused antibiotics shouldn't be poured down drains or put in household rubbish but instead should be returned to a pharmacist for safe disposal. However, there's disconnect between this advice and how residual antibiotics are currently being disposed of.

Research highlights unmet needs of siblings

Last year we put out a request for siblings of people with CF to participate in a research survey.

This research was led by Katie Armstrong of Massey University and focused on the unmet needs of siblings of children with cancer and serious chronic health conditions (including CF) in New Zealand.

Katie has published her findings which found siblings have very high levels of unmet needs.

The summary of findings also includes:

- Almost all survey respondents endorsed at least one unmet need (98%), 86.3% of respondents endorsed 10 or more unmet needs and 50% of respondents endorsed 32 or more unmet needs (out of a possible 45).
- A difference was found between the average number of unmet needs reported by males and females, with females reporting a higher average percentage than males.
- A comparison of the average percentage of unmet needs between older siblings (>12 years) and younger siblings (≤12 years) at the time of their brother or sister's diagnosis, showed the older age group had a higher average percentage of unmet needs than the younger age group.
- Following a child's diagnosis with a serious chronic health condition, siblings can feel strong negative emotions including fear, anxiety, sadness, resentment and distress. Parental focus on the unwell child can lead siblings to feel 'invisible' and 'forgotten' due to reduced parental attention. These feelings can lead to jealousy of their affected sibling, then guilt for feeling jealous.
- When a sibling's emotional, informational and relational needs are met in this phase, siblings are likely to adjust to their brother or sister's diagnosis and cope well in the long-term with their changed circumstances.

The recommendations from the research point to the need for effective, targeted support for siblings with a focus on:

- age-appropriate information about their sibling's health condition
- guidance on how to support their sibling practically and emotionally
- professional support offered to them close to the time of diagnosis
- support and understanding from peers, family and teachers
- time with parents to feel included and valued
- a safe space where they feel validated and can speak freely
- recreation and 'time out' with other siblings who understand them.

Katie is now doing further research that builds on these findings.

You can read the full summary on our website https://www.cfnz.org.nz/news-and-events/latest-news/research-highlights-unmet-needs-of-siblings/

Thanks to everyone from our CF community who participated in this important research.

Port CF data registry

This year we published our 2017 Port CF report. Key findings from the report include:

- There are 501 patients in the registry which makes up 97-98% of all New Zealanders with CF. There are 225 children (aged 0-16) and 278 adults (aged over 16).
- 49.2% of PWCF have the F508del/F508del genotype.
- The mean FEV1 for each group is: 99.7 children, 79.8 men, 76.6 women.
- 31.4% of adults have CF-related diabetes.
- 53.5% of PWCF are recorded as having Staphylococcus Aureus.
- There were 347 annual reviews recorded.
- There were 5,203 total hospital and home IV days across 161 patients.

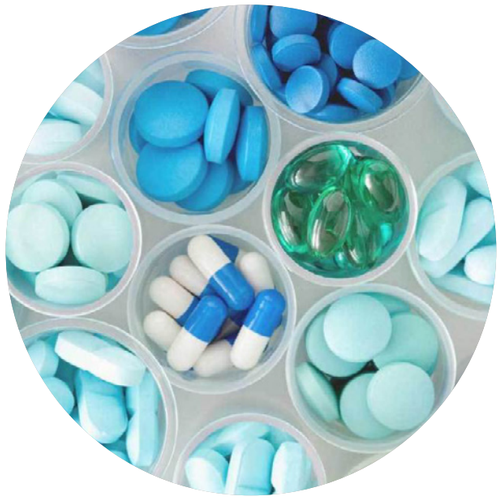
Port CF is a New Zealand data registry owned by CFNZ that uses anonymous patient data to analyse trends in CF care. This, our 7th registry report, provides an ever increasing accurate picture of people with CF in New Zealand and their outcomes.

The database has been further developed over the past years, increasing the amount of data captured, including annual reviews, clinic reviews and hospital admissions.

We're now part of the group working on harmonisation of data registries for cystic fibrosis involving representation from all countries who have a CF registry.

You can read the full report on our Port CF data registry page on our website.

https://www.cfnz.org.nz/what-we-do/port-cf-data-registry/



Study results from Starship research

Results have been published from a study that looked at a urinary protein as an indicator of early kidney damage in children with CF.

A team at Starship Hospital conducted the prospective, single-blind study during 2016 and 2017. Children with CF are at risk of kidney damage because of the use of high dose antibiotics and other medications.

The current test to detect early kidney damage isn't very sensitive and is hard to do in children. The team wanted to find out if the retinol-binding protein (RBP) in urine could be used as an early indication of kidney damage. This protein is released when kidney cells are damaged.

The study included 67 people with CF, aged 1–18, seen at Starship Hospital during their annual review. Healthy siblings of people with CF and orthopaedic patients from fracture clinic with no kidney or other health issues were used as the control group.

Statistical analysis was completed during 2019 and the final report was received late November.

Unfortunately, the conclusion from the study couldn't confirm urinary RBP in children with CF is a good indicator of early kidney damage.

The team's abstract was accepted for presentation at the European Cystic Fibrosis Conference 2020 which was cancelled due to COVID-19. You can read the full abstract on page S131 in the Journal of Cystic Fibrosis, Supplement, June 2020.

CFNZ provided funding for this study and we thank everyone involved.

Research around the world

Gene therapy nearing clinical trials

Scientists have been trying for 30 years to wield gene therapy against CF. In past efforts, the viruses engineered to deliver the working copy of the gene into cells didn't work effectively. Now, thanks to innovations in delivering genetic sequences, gene-replacement therapies are nearing clinical trials, and the field is gaining momentum.

In October 2019, the Cystic Fibrosis Foundation announced US$500 million in funding over the next six years for research into treatments for CF, including gene-therapy approaches.

And in April 2020, Vertex Pharmaceuticals said that it was partnering with biotechnology company Affinia Therapeutics to develop gene therapies for CF.

The gene-therapy company 4D Molecular Therapeutics in California has several therapies in preclinical development. The firm is hoping to test its approach against cystic fibrosis in clinical trials in 2021.

Higher prevalence of oral Candida fungus found in people with CF

People with CF are more frequently infected with oral Candida fungus and carry a higher number of fungi than healthy individuals, a study has found.

The mouth is considered a potential reservoir for opportunistic microorganisms, which can eventually lead to systemic infection. While P. aeruginosa bacteria have been found in saliva and sputum samples isolated from CF patients, little is known about the prevalence of fungal infections such as Candida in the oral cavities of this patient population.

A total of 100 CF patients were enrolled, which included 50 males and 50 females from 3 to 20 years old. As a comparison group, 50 healthy age- and gender-matched individuals were included as controls.

The results revealed that 80% of CF patients with severe disease tested positive for Candida, whereas 68% of those with low disease severity tested positive, compared with 44% of control subjects.

Quantitative analysis showed that CF patients had significantly higher concentrations of oral fungus than controls. However, severe CF patients had similar oral fungal levels compared with those who had low disease severity.

Two Candida species — Candida dubliniensis and Candida tropicalis — were detected only in the CF groups, whereas Trichosporon asahii was found exclusively in patients with severe CF, and Saccharomyces cerevisiae (baker's yeast) was found exclusively in the control group.

All fungal species identified in the study's CF patients were susceptible to currently available anti-fungal medications.

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