CF News

ISSN 0114-3573 P
ISSN 1179-044X O

AUTUMN 2021

IN THIS ISSUE:

CYSTIC FIBROSIS NZ

USEFUL CONTACTS

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

Fundraising enquiries
T: 021 195 5787
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.

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

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

AUTUMN 2021

REGULARS

FEATURES

LIFESTYLE

ADVOCACY

RESEARCH

Welcome to the CF News Magazine

Kia Ora reader,

Thank you so much for choosing to read this issue of the CF News!

We hope you enjoy reading the interviews with some of our wonderful CF tamariki and matua. It's always a great privilege to share stories on what the CF community is up to, and how they're living a life unlimited.

You may have seen over the past few months the extraordinary publicity and exciting progress on the campaign for Trikafta – with Patrick Gower truly taking the CF community under his wing.

Whilst CFNZ has been working alongside Vertex and Pharmac to improve access to modulator therapies for many years, having a respected national journalist thrust the need for Trikafta into the spotlight has helped progress immeasurably. Read the full update on how the campaign for Trikafta is going on page 23.

CF Awareness Month is also on the horizon – running from 1 – 31 May. Read the feature on page 15 on how to get involved and a few ideas of how you can be an advocate for CF.

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 funding this issue of the CF News magazine.

REGULARS

News In Brief

Sir Bob tribute

We were saddened to hear of the passing of Sir Bob Elliott in August last year. Sir Bob made an extraordinary, lifelong contribution to CF, notably for his work on the newborn screening test, which he believed added 30 years of life for people with CF. Sir Bob has been an inspiration to many and can be credited as the reason some of our greatest doctors specialised in CF – many were even trained by him. The impact he has made on not only the New Zealand CF community, but worldwide, is a legacy that will live on for many years to come.

A thank you to Dr Mirjana Jaksic

At the end of 2020, the paediatric CF team at Starship farewelled Dr Mirjana Jaksic. Mirjana retired after 16 years at Starship, where she spent 13 of those years looking after many members of our CF community. The team at Starship put together a book with messages from families who had the privilege of being under the care of Mirjana, which showed her gentle nature, and the real impact she had on so many families; "The kindness you showed us far outweighed the duty of a doctor."

CFNZ would like to thank you for your dedication to the CF community, and best wishes for your retirement.

We've moved!

The CFNZ national office has moved across the road – from number 79 Grafton Road to number 64. We now share a space with the Neurological Foundation, which means we can collaborate with another brilliant charity while keeping costs down. Note: due to cross infection please make an appointment if you'd like to visit the office. Our postal address remains the same.

Sinus rinse bottle donation

NeilMed has kindly agreed to an ongoing donation of sinus rinse bottles for CFNZ to distribute for free, to the CF community, and it comes with an expense. We hope this deal makes life a little easier, and more hygienic. Speak to your fieldworker or email admin@cfnz.org.nz to access.

Pharmac review announced

The Government announced on 2 March 2021 that it has commissioned an independent review into Pharmac. CFNZ is pleased that the Government has responded to concerns about access to new medicines and timeliness of decision-making. We will take every opportunity to ensure the independent review panel is aware of the challenges faced by those with CF, and how early access to medicines such as Trikafta is essential to help address the challenges.

SWEAT4CF campaign raises over $137,000

We're in awe of the 347 sweaty individuals and the almost 2,500 people who made a donation to them! We launched Sweat4CF back in July, and ran a promotional month called SWEATEMBER in September, and were thrilled to see how many people got stuck in. You can Sweat4CF any time of the year by signing up at www.sweat4cf.org.nz and of course join in for SWEATEMBER in September 2021!

FEATURE

CF Christmas Tree Festival sparkles bright despite Covid challenges

More than 30 businesses, community groups, and organisations welcomed a better and brighter 2021 in a sparkly Christmas display at Wellington Airport.

The annual Christmas Tree Festival – a joint event between Wellington Airport and Cystic Fibrosis NZ – aims to support businesses and organisations whilst raising funds for support, advocacy, and research for people with CF.

The event has been running for over 12 years.

"The Coronavirus pandemic meant 2020 was a truly difficult year for many people, community groups and businesses in New Zealand," says Cystic Fibrosis NZ Chief Executive Jane Bollard.

"We decided to go with a 'pay what you can' approach for the 2020 event, rather than set sponsorship packages, to encourage as many businesses to be involved as possible," she says.

"The festival ended up being a brilliant way of pulling together, promoting businesses and community groups, and raising vital funds to give Kiwis with CF a brighter future".

Cystic Fibrosis NZ is thankful for all the support it receives from Wellington Airport, businesses, individuals, volunteers and of course, the New Zealand public walking through the decked-out airport.

The festival was formally opened by the Hon Andrew Little, Minister of Health, at the gala ceremony, and adult with CF Ed Lee kindly shared his personal experience of living with CF in a speech that had the audience captivated.

The trees were judged by a local artist, with the first-place winner being awarded a $10,000 advertising package at Wellington Airport.

Karen Murrell, the New Zealand-owned natural lipstick company, took first place with its beautifully decorated tree full of whimsical cream and pink roses and lipstick 'baubles'.

"We are ecstatic at coming first place in the Cystic Fibrosis NZ Christmas tree festival competition," says founder Karen Murrell.

"Cystic fibrosis is a cause very close to my heart after working closely with Make a Wish Foundation to grant the amazing Rachael her wish of designing her very own limited edition lipstick. I strongly believe increased support of important charities such as Cystic Fibrosis NZ is greatly needed to further widespread education, and I'm honoured to be involved with the work you do," she continues.

Visitors to the airport were also given the opportunity to vote for their favourite tree, with the People's Choice Award going to New Zealand commercial construction company Naylor Love.

Cystic Fibrosis NZ is thankful for all the support it receives from Wellington Airport, businesses, individuals, volunteers and of course, the New Zealand public walking through the decked-out airport.

The Christmas Tree Festival committee is already working on the 2021 festival.

Send us an email at cfchristmastreefestival@gmail.com to find out more or reserve your tree!

Did you see the cystic fibrosis trees?

This years' Christmas tree decorated by Cystic Fibrosis NZ features poignant photos and stories of some of the Kiwis with CF who would benefit from the breakthrough treatment Trikafta. This medication is currently not funded by Pharmac, and visitors are asked to scan a QR code to sign the petition and help change this.

There was also a quirky "compassion pin" tree, featuring the super-sized pins that were made available to people with CF and their families over lockdown.

FEATURE

Living her best life thanks to Trikafta

I was alive, but I wasn't living. I was losing my ability to be a person. I couldn't see a life where I had a future anymore.

Thoughts like these may be confronting, but for Aucklander Bella Powell, as her health steadily declined because of cystic fibrosis, it was her reality.

Bella is lucky. Seven months ago, she was gifted a 3-months' supply of Trikafta. And thanks to the generosity of New Zealanders, she's been able to continue taking the 'miracle' drug Trikafta, and to live life again. But Bella knows how incredibly fortunate she is. It's why she shared her story with New Zealand to raise awareness about the urgent need for PHARMAC to fund Trikafta.

Before starting Trikafta, Bella, like many people with CF, spent hours each day doing CF related treatment.

"I was spending at least 4 hours a day doing nebulisers, bolus feeds and other treatments. I was exhausted and barely able to do even simple tasks. I struggled to get out of bed each morning and walking out of my room became a chore that left me out of breath. I dreaded having to move anywhere," Bella says.

"The last few months before starting Trikafta I was beginning to lose hope. I was battling against a body that no longer wanted to work," she says.

"In my mind I was normal. I could do anything anyone else could do, but in reality, my body was giving up and there was nothing I could do about it."

At the lowest point in Bella's health, her FEV1 hit 22%. She'd been waiting on the active donor transplant list for 2 years.

In August 2020, Bella started Trikafta. Within 6 hours, it began to work.

"In only a few hours, I began to feel lighter, as though my lungs weren't weighing me down anymore. I felt like I could breathe normally. It was the most amazing and freeing feeling," Bella says.

"The first few days after starting Trikafta I experienced what is known as the purge. It was like 16 years of 'junk' was finally being cleared. It was incredible – and slightly disgusting – the actual amount of stuff I was clearing from my lungs. I was coughing all the time because the mucus was so loose. It felt similar to when I've had a lung infection and coughing all the time to clear my lungs."

Seven months on, life for Bella couldn't be any different from what she's known for most of her life. She's back at school full time, playing sports, doing aerial acrobatics and doing everything a 17-year-old should be able to do.

"No words can describe how amazing my life is now. I didn't think it was possible to improve so much and live the life I have now. When I was so sick I had to close myself off. I could barely speak a full sentence without becoming out of breath and even laughing was a challenge because it usually caused a coughing fit," Bella says.

"Now I can live and be my whole authentic self without limitations."

Trikafta has also helped Bella gain weight and improved her overall health.

"Probably a lot of 17-year-olds wouldn't get excited about gaining weight, but I'm thrilled! I'm the heaviest and healthiest I've ever been. Before Trikafta, chewing and breathing at the same time was difficult, which meant it was a challenge to gain weight and keep it on. But now I can chew and breathe at the same time and my appetite has returned to what it was before I was really sick," Bella says.

"I also don't cough during the night, so I have loads more energy when I wake up and generally feel so much better."

Starting Trikafta meant Bella wasn't eligible to be on the active transplant list. Making the decision to remove herself from the list was an incredibly hard decision for her and her family.

"Initially I only had access to a 3-month supply of Trikafta. I was so scared that in those 3 months a donor was going to come along, and I would have missed out on my only opportunity for a lung transplant. But the benefits of Trikafta were incredible right from the first day, so I knew I'd made the right decision," Bella says.

"I couldn't travel further than 2 hours from Auckland when I was on the transplant list. Now I can go on road trips with my family and do so many more things I've wanted to do. For the first time ever I feel practically normal, like I don't have CF anymore. It honestly feels like a miracle," she says.

"Trikafta is more than just 3 pills a day that improves the life of a person with CF. It changes the lives of everyone around them. We need to continue to fight and do everything we can to keep Trikafta front and centre in the media and the medical community"

Bella knows how lucky she is to have the opportunity to take Trikafta. While it means life feels pretty normal, she continues to do her daily nebulisers and take other CF medications. Her CF team is monitoring her closely and Bella must have an additional monthly liver function test. Since starting Trikafta, Bella's FEV1 has increased from 22% to over 30%, and continues to improve each month.

"It feels incredible to know my lungs are healing. I used to get a lot of anxiety before my lung function test as I was worried my result would be lower than before. Now I don't worry, and it feels brilliant!"

For Bella's mum Alley, Trikafta has opened up a world for her and her family that they never dreamed possible.

"I don't think I realised how much of an impact Bella's steady decline was having on myself and other people around us. I'd always lived day to day; I'd appreciate the 'good' days, and on the bad days I'd tell myself, "tomorrow is another day so let go of today and see what tomorrow brings," Alley says.

"But now I have the luxury of looking forward to the future. I'd decided to work part time because Bella was home so much. I wanted to be there with her, to look after her and her mental health as she struggled with the reality of her diagnosis and how hard life had become for her. But now I'm back working full time, I'm back at the gym and I've started my Master's in Educational Management. None of this would have been possible without Trikafta," she says.

"Bella has 2 older sisters who struggled with feeling guilty about living their lives while Bella couldn't. But now that guilt has subsided, they feel like they can fly and anything is possible – for both themselves and for Bella!

"We can go out for dinner as a family because Bella isn't exhausted at the end of the day just trying to breathe. She laughs now, I mean really belly laughs, which I'd never heard from her before. Who knew it took so much breath just to laugh! Bella can now set goals for her future and live her life," Alley says.

Alley and Bella are determined the voices of people with CF in New Zealand are heard. They're living a life every family are entitled to live, because of Trikafta.

"Trikafta is more than just 3 pills a day that improves the life of a person with CF. It changes the lives of everyone around them. We need to continue to fight and do everything we can to keep Trikafta front and centre in the media and the medical community," Alley says.

"I honestly didn't think something this life-changing would come along in Bella's lifetime. But it has, and it works. Now is our time to make sure everyone with CF has the same opportunity as Bella and live the best life they can."

Interview by Sarah Cahill

Last chance to provide feedback for our new Adult Wellbeing Guide

Thank you to everyone who has generously given their time to provide valuable feedback and insight into the development of the Adult Wellbeing Guide.

As we get closer to publishing there's still time to review the guide. The guide includes nutrition (including alcohol), fertility, family planning, sexual health, nutrition, exercise and managing daily life with CF. You can choose to review just one topic or review all the content.

All the comments included in the guide will be anonymous. We'd love to have more lived-experience of people who have travelled overseas or started a family so we can share these with other adults with CF, and some tips for managing mental health and wellbeing.

Please email Lizzie: comms@cfnz.org.nz if you'd like to be involved.

We look forward to sharing the guide with you later in the year, and thank you to Te Hiringa Hauora/Health Promotion Agency for a grant to develop the guide.

WTCF! Homegrown CF podcast

Ingrid Grenar and Ian Wright are parents of an 18 month old CFer who was diagnosed at six months old. As seasoned media professionals, they've decided to produce a podcast to give back to and galvanise the amazing CF community they've now found themselves part of.

What the CF! A Cystic Fibrosis Podcast will talk openly and honestly about Cystic Fibrosis from a layman's perspective but with the responsibility of ensuring expertise when it's needed. They want to support individuals and families by providing the answers to their questions in a clear, correct but (hopefully) entertaining way while increasing both the value of our community and the awareness of CF both nationally and internationally.

"When our son was diagnosed with Cystic Fibrosis at six months old we were shocked and devastated. The road to diagnosis took several painful weeks. What started with kisses on salty skin soon became a persistent cough prompting many GP visits and eventually an X-Ray - the results of which opened the gates to the possibility of CF.

From the first moment seeing his foggy little lungs, we knew this was gonna be a rollercoaster and we weren't wrong. Only a week after diagnosis we were chucked into the COVID lockdown left to digest this huge news within our wee family.

The overwhelm was real, and although the team at the hospital were amazing, it was the online community of CFers and their families where we found an abundance of warmth and understanding.

What the CF! A Cystic Fibrosis Podcast was born out of a curiosity to learn more about this disease, support others, tell stories and share experiences while we navigate our own CF journey."

The core goals of the podcast are to SUPPORT, EDUCATE and INCREASE AWARENESS of Cystic Fibrosis. This will be achieved through content that'll cover DIAGNOSIS AND LIFE WITH CF, TREATMENTS and EXPERTS.

The first episode was released on 16 February, and a new episode will come out every fortnight for the duration of each series.

You can follow the podcast on Spotify or wherever you listen to podcasts. Search What The CF! A Cystic Fibrosis Podcast to subscribe.

Visit the website for more info at www.whatthecf.com and follow on Instagram @whatthecfpod and on Facebook / WhatTheCFPod

Charlie and his Chocky Fish Fundraising

Lizzie: Charlie, tell us a bit about yourself and your family

Charlie: My name is Charlie Ford I am 10 years old, and my nick name is Horse! I go to Pyes Pa school, my favourite colour is blue. I love WWE wrestling, Batman, and my all-time favourite thing to do is going to speedway. My favourite thing at home to do is bounce on my trampoline.

My Dads name is Glenn, and my mums name is Kim and I have an 11-year-old brother named Henry. We all love having holidays in our caravan especially when it is going to other speedway tracks out of town.

L: What made you want to start selling chocky fish?

C: I wanted to raise awareness of Cystic Fibrosis and help other families and people with CF. Plus I get to eat the chocolate fish myself, yum yum!

L: Have you enjoyed sharing your story with your classmates, the people buying chocky fish, and being interviewed for the article in The Weekend Sun?

C: Yes, I really enjoyed sharing my story at school so they could all learn a little bit about CF, mum and dad did lots of talking to people at the stands. It was cool being interviewed for the newspaper article and fun getting my picture taken.

L: Do you think raising awareness and talking more about CF has helped you understand the condition and the importance of treatment and taking pills?

C: I already understood quite a bit about CF and how important it is to do my physio and take my tablets, so I don't get sick (I have a book and mum writes down any questions I have, and I ask them at clinic), it is mostly helped me talk to more people and give me confidence.

L: How do you feel about how far-reaching you are fundraising, and awareness efforts have been?

C: I was surprised how much money people were paying to buy a chocolate fish, and people also made big donations and didn't take any fish, so the amount of money coming in was adding up real fast! Then a really nice man with CF called Mark, donated me more fish to sell and gave me a big surprise by sending me a prezzy card for all my hard work! I am still having fun spending it! I never thought things would have got so big, it so cool and exciting!

L: What has been your favourite part about everything that has happened since you started fundraising?

C: My favourite part so far happened after we did the article with The Weekend Sun newspaper - a man named Kerry Remnant saw my story and wanted to help, so he contacted my school to find me. I was so excited to meet him because he is a speedway driver that has super stock and races at the team's champs every year that my family goes to! My Dad took me to meet him at his house and to see his race car, but the best part was his man-cave full of trophies, pictures, and heaps of cool stuff. He was so nice he also gave me flags, posters, and hats.

I am still in contact with him and he has been helping me with raising awareness of CF and posting cool stuff on his face book page, from that people have wanted to buy chocolate fish and donate money. Mum started a Give a Little page so people could make donation to go to CFNZ Bay of Plenty Branch.

L: How can people continue to support your fundraising efforts Charlie?

C: People can contact still contact us for chocolate fish. We are also getting some CFNZ sign written stickers made to go on race cars to help raise awareness - a big thank you to Paige Cook for getting us some stickers (Paige's husband and brother-in-law have them on their race cars already).

L: How has your family been supported?

C: Mum and Dad said right from when I was diagnosed, they had so much support from CFNZ, Fieldworkers, Doctors and Nurses they felt like they weren't alone. Over the years there has been financial help if needed when I was in hospital etc. All the staff at Tauranga Hospital are friendly, caring and Chani our fieldworker now, is a great help to us all, always only a phone call away and regularly at clinic.

I wanted to raise awareness of cystic fibrosis and help other families and people with CF. Plus I get to eat the chocolate fish myself, yum yum!

A note from the Fords

Thank you to all the people in our lives for your support and help. Kerry has been, and is still amazing with Charlie, along with helping raise awareness of CF and willingly contributing his time to fundraising efforts. He is kind-hearted, takes time out for Charlie, he is also like a mentor and teaching Charlie life lessons. Just a genuine nice guy, thank you from the bottom of our hearts.

One of the top pieces of advice we could share, is that seeing a psychologist was one of the best decisions we made, and we still go now! Sometimes it's hard to try and think like a 10 year old and know how to explain things, and our psychologist Karma helps us out with that and loads more! So, a special thank you to Karma, for keeping the Ford family's mental health in check!

Cystic Fibrosis Month – May 2021

Last July Cystic Fibrosis NZ took the decision to align our annual awareness drive with our counterparts in Australia and America by moving CF Awareness Week to May.

We've always felt there are so many aspects to CF that it's hard to pin it to just one week – so we're taking over May with CF Month!

It's the perfect time to advocate for CF – for better access to treatments, better understanding and awareness, and better support during the highs and the lows.

Three ways you can be an advocate during CF Month

1. Tell your CF story

We want to hear what life is like for you now, the ups, the downs, and everything in-between! We'll pitch your story to the media, and together we can educate the public, bust some misconceptions, and promote what YOU want to be raised about CF. Contact comms@cfnz.org.nz to find out more.

2. Use social media to spread the word

Add a frame to your profile picture, share a personal message or photo summarising your connection to CF, or simply repost a news article or interview. Using social media is a great way to raise awareness of CF.

3. Get involved with the campaign for Trikafta

We're seriously hoping that Trikafta will be secured by May, and we can look back on this point and smile. But there's a chance we'll still be fighting – so CF Month is a great time to push the call for public funding of this important medicine. Visit www.cfnz.org.nz/trikafta for ideas.

Get involved with fundraising

It's also an important time to support Cystic Fibrosis NZ – the only charity dedicated to supporting and advocating for people with CF in New Zealand.

As a charitable organisation, we rely almost entirely on the generosity of the New Zealand public to provide services such as our fieldworkers, information guides, and welfare and emergency grants.

Here's five ways you can help us continue to be there for you!

1. Buy a CF Month Raffle ticket

Our 2021 raffle goes live 21 April, with some fantastic prizes to be won. Order a few booklets and entice your friends and family all in support of cystic fibrosis. Keep an eye out on our social media and website for updates.

2. Get sweating for CF

Jump on to www.sweat4cf.org.nz, create a free page, and make your sweat count! You can take on any challenge that breaks a sweat and get your friends and family to sponsor you.

3. Set up your own fundraiser

Get your thinking cap on – your own fundraiser can be as simple or as elaborate as you like. Think bake sales, quiz nights, lemonade stands, ugly jumper parties..!

4. Get in on the street appeal – 28th and 29th May

Many of our regional branches coordinate local street appeals and other fundraising activities like sausage sizzles. Visit www.cfnz.org.nz/volunteer to sign up.

5. Sell chocky fish

You can't go wrong with selling few chocolate fish. Sell them for a $1 each and over 50 cents go to support services. Contact admin@cfnz.org.nz to get started.

It's the perfect time to advocate for CF – for better access to treatments, better understanding and awareness, and better support during the highs and the lows.

Walking with wings

Sibling to take on epic journey in honour of little brother.

In August 2021 Shane Kidby will start an epic trek to circumnavigate the coastline of Aotearoa on foot, traversing coastal sands, scrambling across cliffs, and swimming across turgid seas.

This 15,000km trek of a lifetime won't be an easy feat, expecting to take at least a year to complete. There's only one known person who's done it before, plus Shane had a hip replacement a few years ago as a result of having Perthes disease as a child.

"There will be a lot of challenges physically and mentally. The west coast is always tough and so are New Zealand winters. I know there might be loneliness, potential food issues and I definitely feel that if I don't finish, I might let people down. I'm ready for a range of emotions throughout this adventure - tears, laughter, anger… all of them," Shane says.

But for Shane, this is more than just a physical or mental challenge.

"My brother Daniel was born in 1973 and was soon diagnosed with cystic fibrosis," he says.

"The condition shaped my childhood, and some of my earliest memories are of hospitals, physio, Daniel being hooked up to machines, pills and playing games when he was home and able to play.

"Daniel wasn't at school much in his life but I remember one time where he asked me to turn up to his class at a certain time which I did, only to find that the class were having a show-n-tell. They were presenting their favourite thing to the class, and he wanted to show the class me," Shane reflects.

Daniel sadly passed away at age six, when Shane was nine years old. The trauma of this has had a life-long impact on Shane.

"I absolutely hate hospitals and even now the smells of hospitals bring back sad memories. I promised myself that I would live the life of two people and make the most of every day.

"I've always wanted to do this walk ever since I was a kid, but life gets busy and things happen. I'm doing this for myself and for two causes close to my heart, CF and mental health," he says.

"This challenge is a part of living my life with purpose and helping others find theirs. I want New Zealand, and the world, to understand CF and mental health so that together we can create the change that's needed to improve, and save, lives.

"I also want to acknowledge CF siblings as they are sometimes the invisible heroes. They can often spend a lot of time hanging out unnoticed in hospitals with their CF sibling, they sometimes miss out on school and the fun things in life because their family needs to, understandably, focus on the child with CF. I want CF siblings to know that they are amazing."

Shane will be using his epic trek to raise both awareness and donations for his nominated charities' - Taranaki Retreat and Cystic Fibrosis NZ.

"Supporting CFNZ was a natural one for me because my little brother had CF. I thought of it straight away. So many families living with CF rely on CFNZ and the great work they do with facilitating research, advocacy and providing support. I want to help make sure CFNZ can continue the great work they do," Shane says.

"I have battled with mental health over the years and I feel that a lot of CF family's also go through mental health from time to time. I have chosen Taranaki Retreat, a mental health service in New Plymouth, as they do an amazing job in the local community by creating a space for people to breathe.

"My little brother, my family, my daughters, people living with CF, CF siblings and parents as well as those living with mental health all inspire me. Brando "Wild Boy" Yelavich has also inspired me. He is the first and only person I know of that has successfully walked the entire coastline of New Zealand. His story is incredible," he says.

Shane is currently working with physiotherapist Finn at Greg Brien Physiotherapy, New Plymouth, to get him prepared for the trek.

"Finn is a member of the Alpine Club so he understands hiking the NZ terrain. He has developed a physiotherapy and training program that is suitable to get my health, fitness and hip in the right space to tackle this challenge. It involves gym work and walking a lot of kilometres with my trusty 20kg pack on. The local YMCA fitness Centre Gym has also kindly donated a gym membership to help with training," Shane explains.

Shane is self-funding most of this challenge and is grateful for the NZ public and business community support so far towards making this adventure a reality.

"I've also been busy studying maps to understand the terrain and what sort of equipment I am going to need. So far I am grateful that Swazi, an outdoor clothing company, are providing apparel, Macpac are providing a tent and other items and Oboz are giving me hiking boots. I'm also hoping to organise a GPS tracker so that people can follow my progress as I walk," he says.

Between training, Shane has been busy doing radio interviews, sharing stories on Instagram, and putting a plan together so that when he starts the walk, he can create as much awareness for CF and mental health as possible.

"I would love for people to help me create awareness about CF and mental health by sharing their stories with me, and if they want, the wider Walking with Wings followers. The most powerful tools those living with CF and mental health have is their stories and their voices," Shane says.

"Their stories will help to create awareness and collective understanding, which leads to positive change. Sharing your story and hearing the story of others can be daunting, but it is empowering to know that you are not alone."

You can find out more about Shane's Walking with Wings challenge at www.walkingwithwings.nz or by emailing him on walkingwithwingsnz@gmail.com.

Support Shane's trek

- Make a donation to Cystic Fibrosis NZ (cfnz.org.nz) and Taranaki Retreat (taranakiretreat.org.nz) and let them know it's in support of 'Walking with Wings'.
- Offer a bed for the night, a meal, or a shower along the way.
- Help with a lift across the many harbours or river mouths he'll encounter.
- Contribute to the costs of the trek by donating to his Givealittle page (search: Walking with Wings) or by donating equipment or products.
- Comment, like and share Shane's posts on Instagram 'walkingwithwings' and Facebook 'Walking with Wings NZ' – help him make as much noise as possible!

A word from Alex, Adult Rep on the Board

I am in my second year on the CFNZ Board as the Adult Representative, and I have had just one Adult with CF contact me for help since I introduced myself in the October 2019 Panui. We thought it best that I say hi again, and so that everyone is aware of my role and my availability to you all.

Don't worry, the fact I haven't had many messages does not mean I haven't been busy! The Adult Rep position is a full Board Member role, with the same responsibilities as the rest of the Board. After just a few months in the role, (my first Board role) we went into this new age of COVID-19, and it has been a baptism by fire. I currently Chair the Finance Audit and Risk Committee on the Board (thanks to my background as a Chartered Accountant), and I have been involved across other projects and goings on.

However, importantly I am here to provide the Board with the adult with CF perspective on CFNZ policy decisions and actions, which will affect services provided to, and for, adults with CF.

As I said in October 2019, the CF community is a diverse group of people, spanning age, health, gender, and economic status. I have been a pretty lucky CFer recently, and my health has been very manageable. This can mean that my perspective on CF is not always everyone else's perspective. However, many of you will know Lizzie, who is the Communications Coordinator for CFNZ. She is my twin sister (an important 20 minutes younger), and between us we have seen a lot of the CF spectrum.

Looking forward for 2021, I want to ensure that YOUR 'seat at the table' is as well utilised as possible.

I will try to post on the CFANZ Adults Network Facebook page after each Board meeting if there are any topics / updates which I think are of interest to the Adult Community that I want to get your perspective on. This won't replace the comms channels like the CF News or the Panui though, it'll just be about ensuring that I am the best conduit I can be for you all.

Similarly, I encourage you to reach out if you would like to. I keep a close eye on the CFANZ Adults Network Facebook page, managed by Lisa Borkus, and would love to engage with you there. Or if you want to talk directly to me about something in particular, I'm just an email away at mckayver@me.com. Even if I'm not the right person to be able to help you, I will be able to work out who is.

A little bit more about Alex

I live in Auckland, with my partner Matt, and our pug Dora. I'm an eternally struggling golfer, an avid foodie (although like most of us, I'll eat anything), and always partial to an ice-cold coca-cola.

As I mentioned, my health is pretty good, so I work full-time as the Product Owner of Pet Insurance at Southern Cross Health Society. My role is to help devise and execute on the strategy of the product, making sure that our Members (and pets!) are as well looked after as can be and their experience is as seamless as can be.

Taking care of ourselves

Gretchen Kitching – CFNZ Northern Fieldworker

This last year has brought many challenges. But it's also been an opportunity to reflect on how stress impacts on our lives, how we cope with stress and look after ourselves, and what we prioritise as important to our happiness and wellbeing.

Over this time, I've been reminded of the importance of taking care of ourselves. Taking care of ourselves to be more resilient to the stresses we face, and the complexities that come with both everyday life and living in these somewhat unsettled times.

Stress is feelings of emotional, mental, and physical tension. It is a subconscious reaction to a perceived challenge, threat, or demand. How we respond to this is unique to the individual, the context, and the surrounding circumstances. Our bodies experience and react to stress, with the possibility of both positive and negative outcomes. Stress can result in feeling overwhelmed and unable to cope. But it can also promote motivation, awareness and vigilance, building resilience and self-efficacy.

Different aspects of our lives

There are many viewpoints on the different aspects of life that make us who we are, impact on how we see the world, and contribute to our experiences. A perspective that resonates with me is the identification and understanding of ourselves as holistic beings with a focus on mental, spiritual, physical, and family/social health and wellbeing. At the foundation of this is our environment. These four pillars impact each other, meaning we need to nurture and care for each part of ourselves to support our overall selves. If something in our life is challenging the wellbeing of one of these pillars then we have the ability to draw strength from the other four pillars of what makes us who we are.

Maintaining and strengthening these aspects

Our environment, our place of belonging. Take time to enjoy nature. This can be the beach, a mountain, the bush but also the everyday things that surround us. The bees enjoying the summer flowers, the sound the wind makes in the trees or the birds settling in for the night.

Physical wellbeing, a focus amongst the CF community. Sports, gym, walk, swim but also having something physical to do at home. Online classes, trampoline, dance or even a hula hoop.

The key here is finding something enjoyable. Spend time exercising with friends or family, strengthening connections with the people that are important to us. Exercise also lifts our mood and helps us cope with everyday commitments and stressors. Physical wellbeing is also about body awareness, how we respond to the hot sand under our feet or the cool sensations of a crisp spring morning.

Mental and emotional wellbeing, how we feel, communicate, and think. Taking time to do something that makes us happy and something that helps us relax. Listening to music, connecting with someone special, reading in the sun, allow yourself this time. Take a break from your everyday normal, have dinner outside, disconnect from social media and phone for an evening, cook an exotic new recipe, camp on your back lawn, do something you keep postponing.

Spiritual wellbeing, who and what we are, where we have come from, and where we are going. This is different for each of us. Whatever our beliefs are, we will gain from appreciating the beauty around us and taking time to do what makes us happy. Focusing on rediscovering what makes us have hope, awe, and unity. This helps us feel secure with our identity and our beliefs, coping better with stress and challenges, and it also helps to build strong relationships.

Family/social wellbeing, our sense of belonging, who we care about and who we chose to spend our time with. These relationships contribute to our happiness and sense of identity. Spending time with family and friends, doing things for them and together, getting involved gives us a sense of purpose and connection. It benefits us and strengthens the relationships we share. Healthy relationships are a core source to strength, support, security, identity, and wellbeing.

CF no barrier for entrepreneurial Ollie

Oliver Klotz is a 27-year-old Marlborough man, who lives with his partner and daughter.

He's a keen fisherman, he's passionate about anything with a motor, their three dogs, and most of all Ollie loves being at the gym. He's pretty funny too.

"I love winding them up, and then sucking up to them shortly after," Ollie says, when explaining what his family-life is like.

Ollie says that living with CF and holding down a job hasn't always been an easy task. There often aren't enough sick days to cover hospital admissions for IVs, sinus operations and stomach issues. Working fulltime and staying healthy can be difficult to balance.

"Once you wake up at 4.30am, have breakfast, do physio, nebulise, get the kid's lunch packed, go to the gym, get ready for work, work (while somehow eating enough food in 1 hours' worth of breaks) get home at 6pm, sort the kid out, keep the house clean, walk the dogs, do physio and nebulisers and get into bed at 11pm... it leaves you with roughly 4 hours to sleep," he explains.

"My health was slowly declining so enough was enough."

A few months ago, Ollie started a business called Klotz Forestry and Outdoors – a small business that provides equipment to the logging and forestry industry, as well as outdoor equipment for everyday Kiwis.

Starting his own business and being his own boss has been great for Ollie. He's been able to manage his health well, and in his most recent annual review, saw an increase in his lung function. He's also off insulin, crediting this to now to having more time to prepare GOOD foods.

Asked if he's faced any challenges, CF or otherwise, while on his new venture, Ollie explains, "Yes I definitely have, I don't think there could be a worse time to start a business with Covid floating around.

"Freight has been a huge issue and our economy is the worst its been in a long time, so it definitely has been a challenge. But luckily I knew a lot of people in the industry already who have jumped over to us."

Ollie is a big fan of the gym, and he knows how important fitness and keeping active is for people with CF. Since Ollie has started his own business, he's found that he's had much more time to put effort into his training, nutrition, and mindset.

"It's easy to train for an hour a day, but it's the 23 hours that follow which count, and where I was going wrong," he says. "I can now set goals and achieve them. The gym has never really been a thing to 'get jacked' or 'become the strongest man in the world' or even help with my CF if I'm really honest (though it hugely does help). It's more of a therapy session for me, headphones on, only thing you can hear is what's inside your mind, zero other distractions and I guarantee without the gym I simply would not be here today."

Ollie's advice for other people with CF who are budding entrepreneurs, is "Do it!" He says take that first step and everything will start rolling along with it. At the end of the day, the worst thing that can happen is it doesn't work out… But setting goals, and achieving them feels so good, so it's all worth it.

Ollie is a big fan of the gym, and he knows how important fitness and keeping active is for people with CF. Since Ollie has started his own business, he's found that he's had much more time to put effort into his training, nutrition, and mindset.

A massive thank you to my best friend, soul mate and business partner Sera. Without her none of this would be possible. She's been there, driven me to hospital when I wasn't well enough to drive, saved me from multiple insulin hypos and so much more.

Also to my mother, none of this would be possible without her help, time and effort.

Check out Ollie's business here: klotzforestryandoutdoors.com

Access for Aotearoa

Trikafta Campaign – Kiwis with Cystic Fibrosis can't wait any longer.

Working to secure public funding of Trikafta for Kiwis with CF is CFNZ's highest advocacy priority for 2021. We are continuing to work closely with Trikafta for Kiwis to raise awareness of the urgent need for Trikafta in NZ.

Working to secure public funding of Trikafta for Kiwis with CF is CFNZ's highest advocacy priority for 2021. We are continuing to work closely with Trikafta for Kiwis to raise awareness of the urgent need for Trikafta in NZ.

Trikafta needs to be publicly funded in NZ now to ensure that the significant benefits to people with CF, their families and the NZ health system can be realised as soon as possible. Kiwis with cystic fibrosis can't wait any longer.

In December 2020, the Wellington Airport Christmas Tree festival, organised by the Wellington Branch, raised awareness of cystic fibrosis and the need for Trikafta to be made available in New Zealand. The festival was officially opened by the Minister of Health, Hon. Andrew Little.

Ed Lee, a CFer who has been able to privately access Trikafta, spoke at the Festival highlighting the major improvements in his health, and his ability to live a normal life, since taking this break-through medicine. As a result, the Minister was able to hear first-hand from Ed how Trikafta has given him positive hope for the future.

In early February 2021, Patrick Gower again highlighted the benefits of and urgent need for Trikafta through Newshub's Because it Matters series. Following on from the interviews with Bella Powell and Sir Bob Elliott in August 2020, Patrick talked to Bella, Ed Lee and Izaeah Twose.

Over three nights, these CFers bravely shared details of their CF challenges and the life-changing benefits which Trikafta has brought them. Personal stories like this make real the life-long burden that those with CF carry, and bring in to sharp focus how Trikafta would help to lift that burden. We thank them for their willingness to share their stories for the Trikafta campaign.

And thanks must also go to Patrick Gower and Newshub's Shannon Redstall for their passionate and tireless commitment to improving the lives of those with CF. Thank you, Paddy and Shannon!

At the same time, Jane Bollard, CFNZ Chief Executive, announced "After months of discussions we're pleased to share that we have secured the agreement of the key stakeholders: PHARMAC, Medsafe, and Vertex, to attend a facilitated meeting with CFNZ, and Trikafta for Kiwis, in March 2021 to work together to identify a pathway to public funding of Trikafta in the shortest possible time".

The meeting with key stakeholders will give us the opportunity to make clear the needs of the CF community in NZ, and explore options for progressing the approval of Trikafta as soon as possible. It is a critically important meeting and we are grateful to all parties for confirming their willingness to participate.

Vertex NZ Application

Shortly after Patrick Gower's documentary, Vertex announced its plan to submit its applications to Medsafe in the coming months. The applications will cover three treatments for CF, including Trikafta. Vertex noted its willingness to work in creative and innovative ways to provide access to treatments as soon as possible. Vertex also revealed it is in discussions with PHARMAC in relation to a path forward for Trikafta.

Kiwis with cystic fibrosis can't wait any longer.

Working to secure public funding of Trikafta for Kiwis with CF is CFNZ's highest advocacy priority for 2021. We are continuing to work closely with Trikafta for Kiwis to raise awareness of the urgent need for Trikafta in NZ.

Keeping the pressure on – how you can help

Patrick Gower's documentary received widespread coverage and raised awareness of CF and the urgent need for Trikafta to be publicly funded in New Zealand. The meeting with key stakeholders, together with Vertex's announcement, is a major step forward.

But we need to ensure this progress translates into a decision to approve and fund Trikafta for Kiwis with CF as soon as possible. We need the CF community to help us keep the pressure on.

You can help by:

- Meeting your local Member of Parliament (MP)
- Getting the Trikafta petition out
- Going to your local media
- Posting your story.

To help you do this, we have set up a Trikafta page with material to support advocacy from the CF community. On the page you'll find key messages which summarise why Trikafta is so important to Kiwis with CF and the case for public funding, together with information about contacting your MP, sharing your story and getting signatures on the Trikafta petition.

We know it takes courage to share such personal stories, but having CF is nothing to be ashamed of. These stories make real the life-long burden that those with CF carry, and how public funding of Trikafta would help to lift that burden.

Thank you to all of those who have already contacted their MPs and shared their stories.

www.cfnz.org.nz/trikafta

Scan this QR code using the camera on your phone and it will take you directly to the petition, or follow the link: www.parliament.nz/en/pb/petitions/document/PET_99952/petition-of-carmen-shanks-publicly-fund-trikafta-medication

Trikafta in Australia

On 25 March 2021 Vertex announced that it has received Australian TGA approval for Trikafta to treat people with CF ages 12 and older with at least one F508del mutation. TGA is the Australian equivalent of Medsafe and is responsible for assessing the safety and efficacy of therapeutic goods. The approval from TGA is an important step in the process for Australians with CF to gain access. An application for the funding of Trikafta is being considered by PBAC (equivalent to PHARMAC) with an outcome scheduled for 23 April 2021.

TGA's decision is also a positive step forward for NZ, as approval in Australia can potentially help to shorten Medsafe's process.

CF Australia is running a major advocacy campaign to have Trikafta funded under the banner:

"There is no present like time
There is no time like the present
It's time for Trikafta."

Jane Bollard, CE of CFNZ, is in close touch with CF Australia. We have our fingers crossed for a positive PBAC decision in April 2021. A positive decision in Australia would provide a significant boost to the campaign for Trikafta in NZ.

Trikafta overseas

Trikafta is now available to CF patients in the UK, Republic of Ireland, the USA, Denmark, and Germany, with patients 12 years and over in Switzerland eligible to receive the medicine from 1 February 2021.

Health Canada has accepted an application from Vertex for the approval of Trikafta for those 12 years and older. The application will be considered under priority review and a decision is expected in the first half of 2021.

Vertex has announced that the US Federal Drug Administration has accepted its application to expand the use of Trikafta for those aged 6 to 11 years. The application will receive priority review and a decision is expected in June 2021.

Vertex will also file an application for approval of Trikafta for the 6-11 year age group in the European Union in the first half of 2021, and filings in additional markets, including Canada and Australia, are planned for the coming months.

Trikafta and other CFTR modulators

In December 2020, the US Federal Drug Administration expanded the number of CF related mutations considered responsive to Vertex's three CFTR modulators, Trikafta, Symdeko and Kalydeco. The approval adds between 50 and 170 additional rare mutations to these therapies.

Vertex has provided a page where people with CF can enter their CF gene mutations to determine which CFTR therapy may potentially be an option for them. However, it is important to note that the website is intended for United States residents only in respect of approved treatments.

www.vertextreatments.co

In addition to campaigning for Trikafta, CFNZ is committed to working to secure the widest access to all relevant CF modulators for Kiwis with CF. Vertex's announcement that it will apply for three treatments for CF in New Zealand is very positive news.

Medicines funding

CFNZ has also been campaigning to secure an urgent and substantial increase in public funding for medicines, together with improvements to speed up access to life saving drugs. Current arrangements mean that New Zealand continues to fall further behind other developed countries and now ranks last of 20 OECD countries for market access to modern medicines.

New Zealand's medicines' budget has been significantly underfunded, and is failing to keep pace with population growth and inflation. The waiting list for funding of medicines has grown from 92 in 2015 to 110 in 2019, while the average waiting time for approval has increased from 2.7 years to 4.8 years over the same period. New Zealand has publicly funded only 23 of the 403 medicines launched elsewhere in the OECD between 2011 and 2018.

PHARMAC needs a substantial increase in funding now to enable it to clear the growing backlog of medicines awaiting funding, and to provide for life saving new medicines. Improvements are also needed to speed up access to life-saving medicines.

CFNZ supports the petition from Patient Voice Aotearoa to the Health Select Committee seeking the reform of PHARMAC and the doubling of PHARMAC's budget.

Again, we need as many people as possible to sign the petition which now closes on 31 May 2021.

Scan this QR code using the camera on your phone and it will take you directly to the petition, or follow the link: www.parliament.nz/en/pb/petitions/document/PET_91080/petition-of-malcolm-mulholland-for-patient-voice-aotearoa

Review of PHARMAC

The Government announced on 2 March 2021 that it has commissioned an independent review into PHARMAC.

The review will focus on two areas:

- How well PHARMAC performs against its current objectives and whether and how it its performance against these could be improved.
- Whether PHARMAC's current objectives maximise its potential to improve health outcomes for all New Zealanders as part of the wider health system, and whether and how these outcomes should be changed.

The review will consider a range of factors, including:

- The timeliness of PHARMAC's decision-making (in particular for new medicines)
- The transparency and accessibility of decision-making processes
- Equity, including access to medicines and devices for Maori and Pacific peoples.

The review will be undertaken by an independent panel chaired by consumer advocate Sue Chetwin. It is expected to be completed this year, with an interim report in August and a final report in December.

CFNZ welcomes the review of PHARMAC and is pleased that the Government has responded to concerns about access to new medicines and timeliness of decision-making, in particular.

CFNZ will take every opportunity to ensure the independent review panel is aware of the challenges faced by those with CF, and how early access to medicines such as Trikafta is essential to help address those challenges.

Engagement with MPS and Ministers

As part of our engagement with the Government, we have written to all MPs, including the Prime Minister and Minister of Health, describing the challenges faced by those with CF and their families.

We stressed the urgent need for public funding of Trikafta, and for additional funding and faster access to medicines in NZ, and sought their support for making rapid progress towards both of these goals.

We are following up the letter by seeking meetings with the Minister of Health and Health spokespeople.

You can read the letter, and keep up to date with all progress in the Trikafta Campaign here: www.cfnz.org.nz/trikafta

We stressed the urgent need for public funding of Trikafta, and for additional funding and faster access to medicines in NZ, and sought their support for making rapid progress towards both of these goals.

Isaac and Claire with Rt Hon Jacinda Ardern

Other Advocacy News

Changes to access to Pulmozyme

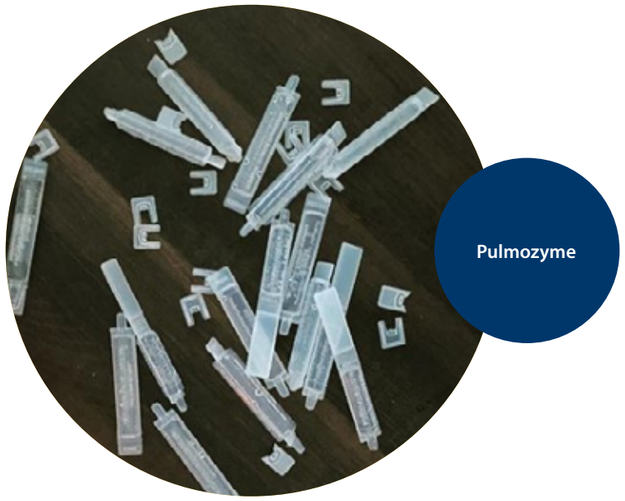
In October 2020, PHARMAC consulted on a proposal to transition the process for obtaining funded access to dornase alfa (Pulmozyme) from the Cystic Fibrosis (CF) Panel to a standard Special Authority.

The change was proposed to provide a more streamlined mechanism for clinicians to apply for funded dornase alfa for their patients, and to improve equity by reducing the burden on applicants and patients when accessing dornase alfa.

CFNZ made a submission in support of the proposal, on the grounds that it would simplify and widen access to a medicine that plays such an important role for CF patients.

The new arrangements took effect on 1 December 2020 and:

- Allow clinicians to apply for dornase alfa funding (Pulmozyme) through the Standard Special Authority application process, rather than applying to the CF Panel
- Remove the requirement for spirometry to commence or renew treatment with dornase alfa for patients older than 5 years
- Focus criteria on exacerbations rather than hospitalisations
- Have one set of criteria for all patients with cystic fibrosis.



Pulmozyme

PHARMAC consultation on advisory committees

In December 2020, PHARMAC announced consultation on improving and expanding how its advisory committees work. Its goal is to improve how these committees provide advice to PHARMAC around medicines and consumer experiences.

These changes are intended to:

- Enable consumers to share their perspectives and advice formally, through the committees
- Enhance the diversity of the committees
- Strengthen health equity knowledge within the committees.

CFNZ has made a submission to PHARMAC on these proposals. We will be seeking changes to ensure that consumer perspectives and advice play a greater role in PHARMAC's decision-making, and that there are opportunities for input throughout the assessment and approval process.

The impact of COVID 19 on the CF community

When COVID 19 hit the world last year among the many anxieties was how it would impact those who were already at greater risk for lung disease such as our CF community.

It was apparent, even prior to our lock down, that families with children with CF were already withdrawing them from school and other crowded events, were already using hand sanitizer which increased further, and had decreased their hospital attendances. In response to the uncertainty, the 'Global Registry Harmonization Group' began regular meetings to capture information about persons with CF infected with COVID 19[1]. As it transpires persons with CF have been affected similarly, not more greatly, to the general population. By mid-June last year, 181 people with CF (32 post-transplant) from 19 countries had been diagnosed with SARS-CoV-2. Of these 11 people were admitted to intensive care (7 post-transplant), and there had been 7 deaths (3 post-transplant). A more severe clinical course is associated with older age, CF-related diabetes, lower lung function in the year prior to infection, and having received an organ transplant[2].

Speculation as to why the pandemic has not hit these communities worldwide as severely as initially feared include that this is a relatively young population, already well-versed in infection control and already adapted to regular hand washing / sanitising behaviour, plus they were conscious of upcoming issues and semi-isolated themselves early. One consequence was the reluctance of persons with CF to access hospital (particularly inpatient) care as needed - meaning, when finally presenting to hospital, individuals had more severe infective exacerbations than they would have usually tolerated, sometimes necessitating an extended hospital stay. One unexpected consequence in the pandemic has been the difficulty in maintaining our usual consistent medical supplies with the occasional medication that is regularly used becoming less, or even not, available.

We are in the process of updating these numbers to December 2020. For my own part I consistently report that (to February 2021) we have had no COVID 19 cases in anyone with CF here, long may it continue. However, we would like to capture any future issues, with COVID 19 remaining on the NZ Paediatric Surveillance Unit reporting systems, and with the our Registry consistently in touch with our adult cystic fibrosis clinics. In the teleconferences, I have much empathy with those who are facing widespread community disease – it is very dispiriting.

There is no reason why vaccination will not work as well in those with CF as in the general population. The Registry group are in the process of reminding various governmental vaccination programme roll-outs that this community remains vulnerable and should be prioritised. Here in New Zealand, our priorities remain a little different with on-going prevention of the entry of COVID 19 to the general community a priority. Therefore immunizing those most at risk of unwittingly being exposed and spreading infection remains a primary focus with border control then front line health care workers being first-up. There is good information regarding the potential roll-put programme depending on the situation we are in as the vaccinations arrive[3].

There are many lessons to be taken out of this pandemic that are only currently being globally explored.

Cass Byrnes

Associate Professor, Department of Paediatrics; Child and Youth Health, Faculty of Health & Medical Sciences, University of Auckland & Paediatric Respiratory Specialist, Starship Children's Health.

References:

1. Cosgriff R et al. A multinational report to characterise SARS-CoV-2 infection in people with cystic fibrosis. Journal of Cystic Fibrosis 2020: 19 355-358

2. McClenaghan E et al. The global impact of SARS-CoV-2 in 181 people with cystic fibrosis. Journal of Cystic Fibrosis 2021, accepted.

3. Ministry of Health, Government of New Zealand. COVID 19 vaccines; Planned roll-out. (https://www.health.govt.nz/our-work/diseases-and-conditions/covid-19-novel-coronavirus/covid-19-vaccines)

COVID 19 and the pivot to telehealth

Aside from the direct effects and anxieties about COVID 19 for our cystic fibrosis community, it has also led to a change in the way we practice medicine.

Across the USA clinical encounters between January-July in 2019 and 2020 were compared; telehealth (phone or video reviews) had increased from 7% to 27%, while face-to-face outpatient reviews had decreased from 91% to 35%, and hospital admissions from 12% to 4%[1] (North American Cystic Fibrosis Conference 2020, NACF). Similarly in the UK, comparing April 2019 to April 2020, inpatient care decreased by 70% for adults and 66% for children, and outpatient care decreased by 27% in adults and 24% in children[2]. The smaller decrease in outpatient visits here was because they continued to count telehealth interactions as outpatient clinic reviews. Given the numbers of individuals involved (31,199 persons with CF in USA, 10,070 in UK) these represent considerable shifts in care. This is not surprising but useful to document, especially to determine if there is an impact on the quality of care provided, and/or whether some of these practices should be adopted further.

Services across New Zealand were rapidly updated to allow these to occur and families increasingly had zoom facilities (rather than phone alone) at their disposal.

The study from the USA looked at the recording of key clinical parameters that would normally be collected as part of routine management and for registry data analysis1. Comparing April 2019 to April 2020, the recording of 'weight' in clinical reviews went from 95% to 40%, 'FEV1' from 75% to 5%, 'respiratory microbiology' from 75% to 5% and 'medications' the individual was taking from 95% to 80%. This data has not yet been reported by the UK study but they did have a greater emphasis on the use of home spirometers (lung function measuring devices)[2]. While some of these measures may not be as important to collect so frequently, increasing the time between recording FEV1 or sputum microbiology could delay recognition of significant change.

These changes reflect global trends in CF care and were certainly mirrored at Starship CF clinic where whole clinics for almost 3 months became virtual reviews. Services across New Zealand were rapidly updated to allow these to occur and families increasingly had zoom facilities (rather than phone alone) at their disposal. The health team also hoped to touch base with more individuals and families per clinic but the clinic reviews took at least the same amount of time.

Additional virtual reviews have also been used to manage anxiety and depression to good effect.

Telehealth has been used in CF care previously. Remote access to glucose monitoring to assess diabetic control has been used for some time. Additional virtual reviews have also been used to manage anxiety and depression to good effect. Similarly, it has been used to encourage airway clearance, engage in exercise programmes and support regular nebulisation use with consistent findings of acceptability to families but not necessarily improved adherence. In the most recent NACF Conference 2020, 11 abstracts presented data investigating telehealth medicine involving between 20-90 patients (predominantly adults) with a 33-63% response rate[3]. The satisfaction was reported at 71% to 95% with one study quantifying the time saving at a median of 180 minutes per clinic and 59% of participants reported avoiding having to take half a day or more of leave from work. Despite this, over 50% recognised there were some limitations to continued care in this way.

The satisfaction was reported at 71% to 95% with one study quantifying the time saving at a median of 180 minutes per clinic and 59% of participants reported avoiding having to take half a day or more of leave from work.

There is definitely a place for ongoing telehealth in the future. It allows decreased travel time and parking costs to the individual/family and it improves infection control with less people physically coming through clinics. It is especially helpful for those being seen frequently where a mix of face-to-face and virtual will be helpful, and similarly for those at distance from the hospital. It is also helpful for some of our outreach clinics nationally where it may allow us to support the local CF teams more often, increasing the efficient use of time by reducing travel to expand the clinic day, especially now when flight timetables continue to vary. It may also facilitate early discharge from hospital admission with ongoing daily interaction.
The disadvantages are the lack of access to spirometry, investigations, and examination. Even in studies that employed home spirometers, results were not as good as those performed in professional laboratories when exhorted to maximum effort by experienced physiologists. Furthermore, the telehealth interface relies on secure high-speed digital access not always available to all, and there is a question about the confidentiality being as secure as the usual hospital services.

However, worldwide CF centres and regional services are working to see where the natural fit of a mix of in-person and telehealth clinical assessments lie with no lapse in quality of care. More to come.

Cass Byrnes

Associate Professor, Department of Paediatrics; Child and Youth Health, Faculty of Health & Medical Sciences, University of Auckland & Paediatric Respiratory Specialist, Starship Children's Health.

References:

1. O'Neil T, Elbert A, Petren KM. Impact of COVID-19 0n CF Foundation Patient Registry Data. Presentation and Poster 140. Pediatric Pulmonology 2020;55(S2):S38-S361

2. Konstant-Hambling R, Carr SB, Madge S, Cosgriff R. Displacement of Cystic Fibrosis Service Utilisation in England during the COVID-19 Pandemic. Presentation and Poster 168. Pediatric Pulmonology 2020;55(S2):S38-S361

3. Poster Session Abstracts. Pediatric Pulmonology 2020;55(S2):S38-S361 (Posters 488, 490, 492, 702, 704, 709, 717, 724, 726, 752, 753).

Know your genes

The future of cystic fibrosis has never been more exciting with the development of the modulator drugs to address the abnormal gene. More than 2,000 mutations in the CFTR gene have now been identified and these mutations grouped into 6 different Classes based on how they affect the protein encoded by the CFTR gene.

As you are all aware Kalydeco (Ivacaftor) was funded and made available in NZ in March last year. This is one good thing that happened in March, 2020; the lockdown - not so wonderful. For those who carry one of the CF genes in Class 3, this was tremendous news as this drug is designed to correct the "gating" abnormality. G551D is one CF gene that is part of the Class 3 CF mutations and those who carry at least one copy of this gene have the option to take this new drug.

The majority of people with CF in NZ have one or two copies of the DeltaF508 gene (now known as c.1521_1523delCTT) – this is a Class 2 CF mutation. Trikafta is a modulator drug which corrects the Class 2 abnormality and is available in other countries but not yet in NZ. The CF community are working hard to lobby Pharmac and the government to have this drug funded in NZ.

It is very important to know which CF genes you carry as you may be eligible for the funded Kalydeco or Trikafta (in the future).

As the coordinator of the NZ Port CF registry I have been looking at the data and identifying all of the CF genes listed in the registry. I have noted that there are quite a few people on the registry who have "unidentified" on the gene question. A few have "Not Known" selected. Most of these people are now in the adult service as years ago when their mutations were "looked for" there were only a small number of CF genes described. Now the Canterbury Genetic Lab can identify most of the CF genes that are found worldwide. To know and understand which CF gene you carry look at the CFTR2 website and find the combination of genes you carry – very informative.

The CF community are working hard to lobby Pharmac and the government to have this drug funded in NZ.

It is very important to know which CF genes you carry as you may be eligible for the funded Kalydeco or Trikafta (in the future).

If you don't know which CF genes you have and would like to know please discuss with your medical team next time you attend the clinic.

Jan Tate

Cystic Fibrosis Nurse Specialist, Starship
Coordinator, Port CF Registry

CYSTIC FIBROSIS NZ

Supporting Kiwis with CF to live a life unlimited

Please add me to the CF News magazine mailing list.

I would like to receive the magazine by: ☐ Post ☐ Email

I am a: ☐ New subscriber ☐ Subscriber updating my details

My Details

First name:

Last name:

Mailing address:

Previous address *(if applicable)*:

Email address:

I want to support Kiwis with cystic fibrosis.

Enclosed is my donation of:

☐ $120 ☐ $80 ☐ $50 ☐ $25 Other

I am paying by: ☐ Visa ☐ Mastercard

Card number: ☐☐☐☐ ☐☐☐☐ ☐☐☐☐ ☐☐☐☐

Expiry date: ☐☐ / ☐☐

Name on card:

Signature:

☐ Online banking, using particulars: (your surname) and reference: 'Donation'. Cystic Fibrosis NZ, 12-3147-0230576-00

☐ Cheque (please make out to 'Cystic Fibrosis NZ')

☐ Please send me information on leaving a bequest to Cystic Fibrosis NZ

Thank You

for your generosity!

Please fill in this slip and post back to us:

Freepost 194668
Cystic Fibrosis NZ
PO Box 110 067
Auckland 1148