

CF News

AUTUMN 2022

ISSN 0114-3573 P
ISSN 1179-044X O



IN THIS ISSUE:

Sharing CF life online

A Trikafta Miracle

Moments of Joy

Trikafta Campaign

Advocacy Update

Fundraising Superstars

cf 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

AUTUMN 2022

REGULARS

Welcome 4

FEATURES

Quick-fire with our new CE 5
 Christmas Tree Festival magic 6
 Farmers raise over \$40k for CF 8
 Ashley's Trikafta story 12
 The Sweatiest September yet 14

LIFESTYLE

Adrian's lung transplant story 16
 Moments of joy 18
 Permission to relax – Chani 20

ADVOCACY

A decade of CF modulators 22
 Trikafta campaign update 23
 Review of PHARMAC 24
 Pae Ora Bill 25
 Advocacy timeline 26

SUPPORT

Supporting our CF community 28

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)

Opinions expressed in articles do not necessarily express the official policy of Cystic Fibrosis NZ. The information in this magazine is not intended to take the place of medical advice from your GP or respiratory specialist. Information correct at time of publication.

© Cystic Fibrosis NZ 2021.
 Registered Charity CC10274. Registered office:
 64 Grafton Road, Grafton, Auckland.

Welcome to our Autumn CF News Magazine

Kia ora,

As we bring all the elements together for this edition of CF News, poring over stories, artwork, and layouts, what's happening outside our offices and homes is totally surreal.

COVID cases in the thousands, a protest on our parliament front lawn, and an attack on Ukraine which is heartbreaking and seems senseless.

In our CF world we were also rocked last month by our expectations not being met as the PTAC minutes were published, and we all felt like we took a collective step backwards, but we are holding on to hope together.

Despite all of this, the pages that follow are full of inspiring individuals from our own community, who are choosing to find joy in the small moments and sharing their stories to encourage others.

On the cover is Natalie, at just two years old this image captures true innocence and delight. CF is a part of Natalie, but it does not define her. Like so many in our community the courage, determination, and resilience in the face of adversity is strong. Josh and Kareena were inspired to raise funds for CFNZ. Together with Link Livestock and Bidr, they ran an online auction and raised an incredible \$40,000 for CFNZ. You can read more about their story on pages, 8, 9 and 10.

Anxiety and overwhelm are real, and it is completely okay to feel what you are feeling. Most importantly, be kind to yourselves and do something that makes you feel calm and steady. Know that our CFNZ team has got you and we are always here for whatever you need.

Please stay safe, trust your instincts, make yourself a cuppa and enjoy this Autumn edition.

Happy reading!

The CF News Team.

Thank You

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



The Grumitt Sisters Charitable Trust, proudly managed by Perpetual Guardian.



FEATURE

Quick-fire with our Chief Executive, Lisa



We put a few very important questions to our new CE when she started.

No pressure, Lisa.

If you could convince the world that you came up with one famous quote, which would you choose?

Be the change you wish to see in the world. Mahatma Gandhi.

What's one thing you're deeply grateful for?

The lessons my Nana taught me that led me to a career filled with passion and purpose.

Who has been your greatest source of inspiration?

My son! He has been my greatest teacher! He's taught me the true value of navigating uncertainty, courage and resilience. These are lessons that cannot be taught but come from experience.

What's something that happened that changed how you view the world?

Having a premature baby born at 27 weeks; you can't prepare for something you never see coming and don't understand. It made me realise sometimes you have to take one day at a time, take the good days with the not so good ones, and know you're stronger than you'll ever give yourself credit for.

How do you prefer to start your workday?

With a cup or two of strong coffee!

Who is in your family?

My husband Dan, my son Ben and stepdaughters Heleana, Addy and Ava.

Do you have any pets?

Two dogs, Riley and Ziggy and a few hundred tropical fish!

Are you an early bird or a night owl?

Night owl.

What is the coolest place you have ever visited?

Cuba – it's like stepping back to a place that's been frozen in time.

What energises you at work?

Seeing our team happy, engaged, and inspired by the work we do.

Happiest when...

I see true joy and happiness in my son.

What is your most used emoji?



Do you make your bed everyday?

No but I should. My Nana always said with a clean kitchen and a made bed you've got a tidy house.

What's your favourite way to wind down at the end of a work day?

Embracing the love and excitement of our dogs, so I make time to sit outside with them for cuddles and playtime. It's a re-set for everything.

What songs would make the movie soundtrack of your life?

- These are the days, Van Morrison
- Don't you worry 'bout a thing, Stevie Wonder
- Brighter Days, Hybrid Minds, Charlotte Haining

What drew you to CFNZ?

The chance to make a difference and the privilege of being able to lead an incredible, passionate group of people who want to make life and the future better for people with CF and their families.

What does the future of CFNZ look like?

The future for CFNZ is full of hope and opportunity.

What's one thing you're excited about?

Having conversations about how we continue to evolve and meet the needs of people with CF now and in the future.

Deck the Halls: the magic of the CF Christmas Tree Festival

Every year the creative geniuses at CFNZ Wellington Branch collaborate with Wellington Airport to put on the inspired CF Christmas Tree Festival. Christmas 2021 was no different. Despite the unknowns leading up to holiday season, 47 Christmas trees made the terminal shine.

The magic of Christmas filled the halls of the Wellington Airport thanks to the great mahi of our generous volunteers and sponsors. The trees are placed into groups the night before set-up, and lights are installed in the morning before the sponsors and fairies come in to decorate throughout the afternoon.

The Gala is the official opening night of the Christmas tree exhibits. It was as dreamy as it sounds, guests dressed to impress, and ecstatic chatter filled the room. The night started off with a warm welcome to our special guests, speakers, sponsors, families, and friends of our CF community. Then Steve Tew opened the festivities by introducing Peter Miskimmin. This was followed by a speech from Steve Sanderson, CEO of Wellington Airport. We also heard from Tristan who spoke about living with CF and the new advances in medicine and technology that we are seeing. We are grateful to Tristan for sharing his story so vulnerably and inspiring us all.

As per tradition, the tree decoration competition was one of the main attractions of the night. The creativity and resourcefulness displayed was truly remarkable. The judge for this festival was none other than Lynne Sandri, a local artist from Seatoun. Lynne chose Sam's Art House, as the winner of 2021 CF Christmas Tree Festival for her stunning, bright, and lively tree. The runner-up tree was Naylor Love for their creativity, followed by Bay Plaza for their wonderful feeling of Christmas created with dried oranges and cinnamon.

The festival was a success due to the support of Wellington Airport, and all the sponsors who put in an enormous amount of effort to create unforgettable trees. CFNZ is immensely grateful for all the support and love shown through the Christmas Tree Festival.

If your business would like to be a part of 2022, please get in touch with Laura Huet (laura@cfnz.org.nz).



1st place:
Sam's Art
house



CFNZ
Compassion
pin tree



People's
Choice
Award: Bay
Plaza Hotel



Runner up:
Naylor Love



Tristan and
his daughter

The Wellington Airport team do a fabulous job of organising this event, lovely food, gorgeous wine supplied by Maude and a great jazz band all added to the atmosphere. But most importantly they invited us to come back next year!

Thank you!

With the support of all our incredible sponsors, and the generous people wandering through the airport, an amazing \$36k was raised!



Tokoroa dairy farmers raise over \$40K for CF causes in 2021

Tokoroa couple Kareena and Josh from Tahau Jerseys have fundraised over \$40k for CF causes over the last 12 months - with \$23.5k of that going to Cystic Fibrosis NZ. The remaining funds were donated to their local CF branch in Waikato.

They have three children and their youngest Natalie was diagnosed with CF in 2019 at 10 days old through the heel prick test. Fast forward a few years and these dairy farmers engaged both the farming community, their contacts and the CF community to collect huge sums in donations for CFNZ.

How a cattle auction for CF became a reality

It all started in early 2021 when Josh had an idea:

"We had some extra stock so I thought we may as well sell them and give the proceeds to Cystic Fibrosis NZ. We had a lot of interest straight away and it spiralled from there."

Spiralled indeed. Josh recalls:

"We sent out an email to all our business contacts to see if anyone would be interested in donating stock or goods for the auction, and the response was huge."



Josh and Kareena ended up working alongside Link Livestock, Bidr and CFNZ to run an online auction that included cattle, prized bull semen, holiday getaways, farming equipment and smaller items like key chains.

"Before we knew it we had 44 lots in the auction" exclaimed Josh, "we didn't expect it to be so big."

Nigel Riddell from Link Livestock donated his time providing the photos for the auction and did all the comms behind the scenes. The CFNZ team were on hand to help with some marketing tips and poster design. Now, all they had to do was get some buyers.

The couple set to work marketing the event via Facebook in local and national farming groups and Kareena used her Instagram account @sassy.and.salty (that she'd started to document Natalie's journey with CF) to spread the word.

Going, going, gone!

When it came to auction night they couldn't believe how it unfolded. Kareena recalls:

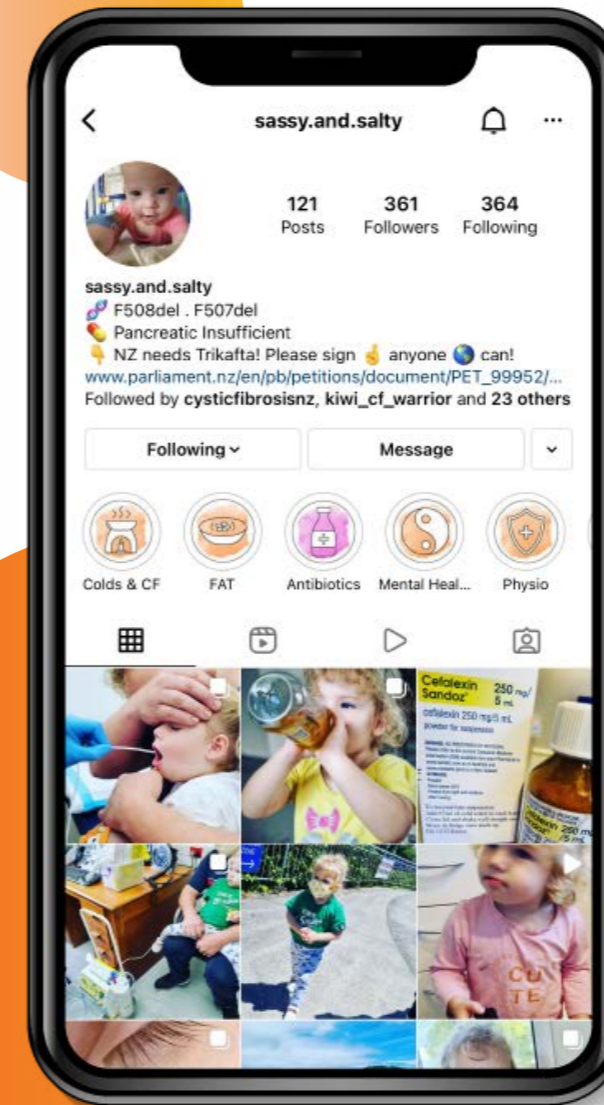
"We had 79 pre-registered buyers and another 120 following the auction. The lots sold quickly and for far more than we were expecting. At the end of the auction, we'd raised \$20,041.50!"

The prices people were willing to pay came as a surprise to both Josh and Kareena:

"We couldn't believe how popular the key chains were. They sold for over twenty bucks each. Afterwards, we were told that people just wanted to donate for CF any way they could," said Josh.

"It's quite a new concept for farmers to bid on stock online and Bidr kindly did it for free too. Going online meant it was nationwide and some livestock sold for over twice their market value on the night. It was also great to see so many CF families purchase lots," he concluded.

After the success, The couple had planned to run this auction biannually but they've had so much interest subsequently that they've decided to run another one to coincide with CF awareness month in May 2022. Josh says:



"It ended up being a bit rushed last year so we'll be smarter and wiser with the second one and we've learned a lot now about how to run it and where to get lots."

Diagnosis and support

Although they'd never hidden Natalie's diagnosis, the work they've done fundraising and Kareena's growing Instagram profile has meant that their friends, family and community are much more aware of what CF is and what it means day today. Josh and Kareena recall getting the life-changing diagnosis:

"The midwife called me to say that CF has shown up on the heel prick test and it all made sense as Natalie had been losing weight, crying a lot and not sleeping. Our midwife has a niece with CF but this was her first positive test in 27 years," said Josh.

"Our midwife has a niece with CF but this was her first positive test in 27 years"

Josh then went into their lounge to break the news to Kareena:

"I bolted upright and asked 'who's died' as his face was so serious and shocked. He explained what the midwife had said and I just burst into tears. But within 30 minutes our midwife was with us and due to her personal connection she was able to tell us positive stories and reassure us that Natalie could have a good life," said Kareena.

They both felt that the final confirmed diagnosis was a relief as at least they now knew what they were dealing with and how to help Natalie. They're eternally thankful for the amazing support they received from their CF team:

"Our CF nurse came to our house at 8.30 on a Friday night and calmly gave us the information we needed. She was so caring, and I was holding Natalie so close to me as I just needed to protect her. Nothing was rushed and the nurse took the time to answer our questions and it all made sense to me. She'd also brought a care bag with enzymes, apple sauce and everything we needed to start immediately. By 9.30 pm that night we'd given Natalie her first enzymes," said Kareena.

FEATURE

Sharing their CF journey

Kareena's decision to start an Instagram account was a necessary tool to help her to process what was happening in her family.

She says: "I was heading towards rock bottom. I was put in touch with an amazing organisation called TRUE COLOURS via our CF nurse. They work with families whose children have terminal or chronic illnesses. I was assigned a counsellor once a week and I started Instagram as a kind of a blog as a way for me to get out my emotions and feelings. I shared the good days and bad days. I shared what I was learning and experiencing - and there's a lot to learn!"

She feels that having an online profile is an excellent way to educate others and a place she could focus her mind on something:

"I didn't feel lonely or isolated anymore. The wider community would approach me and tell us what they'd learned. My aunty walked up to me at a family gathering and said, "what do I need to do for Natalie? What do I need to know?" It brought tears to my eyes. It blew me away that she'd been reading my posts to understand what we were going through. I find people ask more useful questions as they follow along on Instagram. It helps them learn what to ask."



Bidding for our future

Kareena still believes that the biggest benefit she's gained from her Sassy and Salty account is to her mental health.

"Kareena still believes that the biggest benefit she's gained from her Sassy and Salty account is to her mental health."

"It's a safe place to vent and also put your triumphs. It's a safe place to share with no judgement as everyone's in the same boat. I'm connected with people from the UK and America as well as NZ and Australia and the big difference is Trikafta. It's a double edge sword when you hear of the UK recently getting access to Trikafta for six year olds. I'm so happy for them but so sad for us."

As we now know, Josh and Kareena aren't people to sit and wait for someone else to do something. The busy couple also met with their local MP to discuss NZ's funding of Trikafta and they feel hopeful that it's coming here soon.

When asked if she had any advice for parents facing a new CF diagnosis. Kareena said:

"Don't google! I am thankful that I didn't. I took each day as it came. So please take each piece of information that you're given and get your head around it - Enzymes - get your head around it and move to the next - Percussions - get your head around it - etc - etc. Make sure you get comfortable in your routine and what you're doing."

You can find Kareena on Instagram as @sassy.and.salty and we'll keep you posted about the next Bidr online auction later this year.



bidr



A special thank you to Kareena, Josh, Bidr, and Link Livestock for making this incredible fundraiser happen!

Mark Ashford Scholarship



Each year the Mark Ashford Scholarship is awarded to a person living with CF who has shown excellence in tertiary study and / or shown tenacity to overcome the challenges of CF while studying. This years' scholarship is kindly sponsored by Terra Consultants.

terra consultants



For more information on how to apply scan the QR Code.

Cystic Fibrosis Achievers' Awards



The CF Achievers' Awards also celebrate people with CF who have achieved excellence in any of the four categories: leadership, the arts, sports or education. These awards are kindly sponsored by Viatris, distributors of Creon® products.

VIATRIS Mylan + Upjohn



For more information on how to apply scan the QR Code.

Ashley's Trikafta 'miracle' story

Thinking back to September 2021, my world was very different.

My lungs were functioning at 22%, and day-to-day life was tough. I required oxygen 24 hours a day, walking to the bathroom took lots of energy, and walking to the letterbox wasn't even an option. Some days, just making conversation was hard - and I dared not laugh as I feared it would send me into a coughing fit. Imagine being scared to laugh?

Eating was getting harder by the day and my weight was dropping. At 44kgs I was well below the minimum for my size to be considered for a lung transplant. I needed to put on weight, so I had a peg (feeding tube) inserted to try and help with the struggle, which meant supplemental feeding through the tube overnight too.



"Exercise has become an enjoyable event, not an exhausting chore - I have no issues walking over 10kms now!"

CF took over my life

My average day consisted of waking up in the morning and doing my lung clearance, followed by an array of nebulisers and physio. I'd then struggle my way to the kitchen and force down a couple of Ensures (supplemental feeding) and take all my pills. The rest of my day dragged on as I'd try to eat, rest, and when the afternoon came around, it was time to do my treatment regime all over again for another few hours. CF had well and truly taken over my life.

I found myself in the hospital every two months for three weeks at a time. It was getting to the stage that, even after three weeks of antibiotics, I wasn't feeling that much better. I knew I was running out of time and options, and putting the weight on to be able to get a lung transplant seemed close to impossible.

The 'miracle' pills

Then I got a phone call from my CF nurse. She told me I had been approved for the Managed Access Programme for Trikafta. Two days later I took my first dose and by day three I no longer required oxygen and my appetite was coming back. By week two I did my first small hike. I noticed my weight was going up and my face had started to fill in. I had colour in my cheeks again for the first time in years and I had the energy to really enjoy life.

At week seven, I'd gained 8kg and my lung function (fev1) is up to 33%. I felt it improving each day as I got fitter and I no longer required any of my nebulizers or inhalers. My body changed so much and so fast that I no longer recognised myself. I'd been given a part of myself back. I now laugh with friends and spend quality time with family. And all it took to make this possible was to take three tablets per day. It's an absolute miracle.

It's now been four and half months since I started taking Trikafta and I'm feeling fantastic!! My weight is around 53 kgs but I've also gained a lot of muscle now which has given me a big boost in energy. I've been getting fitter every day and I no longer struggle with day to day activities such as going to the supermarket. Exercise has become an enjoyable event, not an exhausting chore - I have no issues walking over 10kms plus now!

My lung function has stabilised to around 30-32% I'm still off all the nebs and haven't had any IV antibiotics in seven months. Considering I was needing IV antibiotics every six to seven weeks is a huge improvement.



"I now have to start planning for the future which is something I didn't have the privilege of doing before."

A thank you to CFNZ

CFNZ and my Fieldworker supported me immensely. I received supermarket vouchers on multiple occasions to help with food costs while I was in the hospital, I used the breath4cf to get some wet weather gear so I could still exercise during winter last year and I was also helped with transport costs when heading to hospital appointments.

My CFNZ Fieldworker has been amazing and she goes above and beyond often visiting me in hospital just to see if there was anything I needed and to drop off a bag of snacks. It's a small act but it definitely makes a big difference when you're in the hospital for long periods of time.

Planning for my future and hoping for a future with Trikafta for all Kiwis with CF

I'm hoping to get back into the workforce in the near future and start to get my life back in check. I now have to start planning for the future which is something I didn't have the privilege of doing before.

But now, I find myself frustrated. Frustrated because I have to watch many young people with CF, including my own younger brother, go down the same path as me for absolutely no reason other than money. We desperately need Trikafta funded for all Kiwis with CF.

SWEATEMBER

SWEAT FOR CYSTIC FIBROSIS

The SWEATIEST September you've ever seen.

What a phenomenal SWEATEMBER we had in 2021!

Over the month of September, 324 people committed to a sweaty activity, and together raised over \$166,000 towards support services for people with CF and their families.

All our sweaty superstars, along with our supporters and sponsors truly are our champions. It was so exciting to see the amount raised climb each day, knowing the huge impact it will have on the lives of our people living with CF, and their whānau.

We can guarantee your sweat has never meant so much.



Meet Sam
He formed a team with his mates and together they raised an incredible \$20,700.

"With the help from a few friends, we ran & cycled 6,000 k's during the month of September...the equivalent of a return trip from Cape Regina to the Bluff. More importantly, we raised \$20,700 for a cause a little too close to my heart. I'm 36, was born with CF, had a lung transplant at age 23. Exercise has always been a huge part of my life, despite the obvious physical challenges. My parents got me involved in many sports as a kid helping me develop a deep passion for sports, exercise and generally being active. CF is an illness, that despite best efforts can be largely out of your control, but I've always taken solace in knowing that if I'm exercising regularly in whatever form, then I'm making the most out of the things I can control. The SWEATEMBER campaign is a fantastic initiative that raises awareness & highlights the importance of exercise & activity in the CF community. We've got some big things in the pipeline for SWEATEMBER 2022, watch this space!"



Meet Josie
Josie was our highest individual, bringing in a huge \$13,150.

"Inspired by my good friends Lizzie and Alex, and in memory of the beautiful Bees, I committed to running 150kms in September and raising \$1000 for CFNZ. By the middle of SWEATEMBER, I was challenged by a colleague to up the ante and run 200km... And I am never one to back out of a challenge. I made it to 208km and raised over \$13k thanks to my generous mates. I can't believe the messages and support that I received for CF and running. I'm very proud of the achievement but I'm so very grateful to everyone who supported CF. Lizzie, Alex and the 540 other kiwis that live with CF deserve all the support we can muster and the titles of legend, champ and inspiration. I simply used the privilege of a healthy body, and my dogged bloody determination to use my powers for good."

Why you made your sweat count in 2021

"My partner has CF along with many others! I am playing my part this SWEATEMBER to support the CF community & raise \$\$ to help support him and our fellow CF friends live long and happy lives."

"We're sweating for CF because we want our daughter to get the best possible healthcare, so she can continue to do all the things she loves."

"I'm challenging myself to do something I could've never done before. To support all of those incredible people who have CF in NZ!"

"We are sweating to continue to raise awareness and much needed funds – our boy is 12 and has lived with CF his whole life. There is nothing he loves more than being outside riding his bike, playing soccer, catching up with friends and is super pumped for the next cricket season to begin. CF means there is a whole other element to his wellness and the daily challenges this can bring but he doesn't let it slow him down. The more we can all know and understand about CF and what can be done can only be a good thing."

"Because my beautiful daughter has CF and I want to do what I can to support her and all people living with CF in NZ. I'm going to walk 150km in September!"

Get sweaty in 2022!

Hopefully you're suitably inspired, we'd love you to join us in SWEATEMBER 2022. Joining is simple! First, Set a sweaty goal. It can be a walk or run every day, a dance class or laps in the pool. Second, register to sweat. Visit www.sweatember.org.nz and register for free. Set a fundraising target and personalise your profile page. Add a photo and a small bio about what motivates you. Finally, SHARE! Update your friends and whānau by posting Instagram stories, fill your feed with some sweaty pictures or simply email them your fundraising page.

Healthier, happier, and loving family life six months post lung transplant

Adrian Ashdown is a 45-year-old adult with CF from Helensville, Auckland, who works as a business and process improvement lead at Auckland Council.

Last year he was living with declining health and little hope for the future, when six months ago, he received a lung transplant.

"I was pretty healthy up until about two years ago," Adrian says. "I could count on my hand how many hospital admissions I've had throughout my life. But, over the last few years, I was existing on 30% lung function even though I could still go to the gym and work etc."

Things continued to decline, and Adrian took a turn for the worst:

"In late 2019 early 2020, my lung function dropped to 20%. The hospital team had been talking to me about a lung transplant for five years or so, but I buried my head in the sand. I used to struggle to walk the short distance from the car park to my office at work. Once it got really bad, I was out of options, so I went on the transplant list at the end of 2020."

His health concerns weren't over as he waited for a transplant. It wasn't long before he faced a further challenge:

"In April of 2021, I noticed swelling in my feet but ignored it thinking 'she'll be right', but when I finally went to see the GP they told me to go to the hospital straight away. I was diagnosed with right-side heart failure as my low oxygen levels had meant my heart had to work harder and it had just had enough. At that point, I was put on oxygen full time. This included a BiPap connected to an oxygen concentrator on my mains power which was a little worrying as I live on solar power."

A life-changing day

Adrian then returned to hospital for a more routine appointment that was to become a life-changing day for him and his family:

"I was back in hospital to monitor my oxygen while I slept and the Nurse came in and said the doctor wants to see you. I was told that I would be able to get Trikafta and start immediately. I was thinking 'great I can get 10 - 20% lung function back' but then two hours later I got another phone call saying we've found some lungs for you. I had to decide - did I want to try Trikafta which may or may not give me that extra lung capacity or go for the transplant?"

"I had to decide - did I want to try Trikafta which may or may not give me that extra lung capacity or go for the transplant?"

On the 1st of July 2021, Adrian had a lung transplant.

"I'd accepted it would happen, but I don't think you can ever be ready - my worry was more about what will I do financially not the medical side - my brain goes to the practical side," said Adrian.

Post-transplant positivity

Today, Adrian's life has radically changed, he no longer struggles with the walk from the car park to his office, he can play more with his daughter, and he has a more open and empathetic view of the world. He's very positive about his transplant experience:

"I was only in ICU for 5 days and back home six weeks later. I'm really good with no signs of rejection to date. I've gone from being exhausted taking a shower, to going to the gym five times a week. I'm up from 69kg to 84kg."

Along with generally just having more energy to tackle everyday tasks like housework, the greatest gift his lung transplant has given Adrian has been his ability to be a better parent and do so much more with his 10-year-old daughter:

"She's never known any different and I don't think she knew how bad my health was before my transplant. We're at a stage now where she needs to keep up with me so that's pretty different," joked Adrian.

"We're at a stage now where she needs to keep up with me so that's pretty different."

Getting back to business as usual

Adrian was able to start back at work in January and he says it's business as usual:

"My boss is fair and we had really good discussions about support and workload before I finished up. I didn't know if they would keep my job open, but they kept my role for me (while someone filled in for six months). They've immediately started giving me new projects so they must have confidence in me."



A fresh perspective on life

Having a major procedure like a lung transplant is life-changing in so many ways but for Adrian, it's given more than his health back starting with a new lifestyle with fewer meds and treatments.

"It's weird to cough and nothing coming up and getting home and not needing my nebs or inhalers was weird. I only need to take Creon now for my CF. I'm the healthiest I've ever been in my life. This wouldn't be possible without a lung transplant."

Adrian has reflected on his pre-transplant life and how fiercely independent he was. He wasn't one to accept or ask for help, he says:

"I've previously found it really hard to accept people's help, but I've learnt that you can't live without support - that's a big change in my character."

This means that he's now much more likely to get involved and help others even if it's just a supportive chat.

Adrian is positive for the next generation saying: "Anyone that was born with CF in the last five years will have a much easier journey than we did. The outlooks are constantly shifting. When I was born my parents were told not to expect me to make it through primary school but here I am at 45 and feeling the best I've ever felt."

Advice for others on the transplant list

Now that Adrian feels well into his recovery, he thinks more about people who may be beginning their journey:

"The one thing I wish I'd done was to join support groups. Once you're on the transplant list they have transplant support groups every week via Zoom. I didn't want to go before I had the operation, but I started to go afterwards. I was so annoyed that I didn't go before! It can be scary hearing some of the stuff, but you hear way more success stories than you hear bad. So, I'd say to please go and join groups and share your questions and worries as it really does help. You don't have to face it alone."

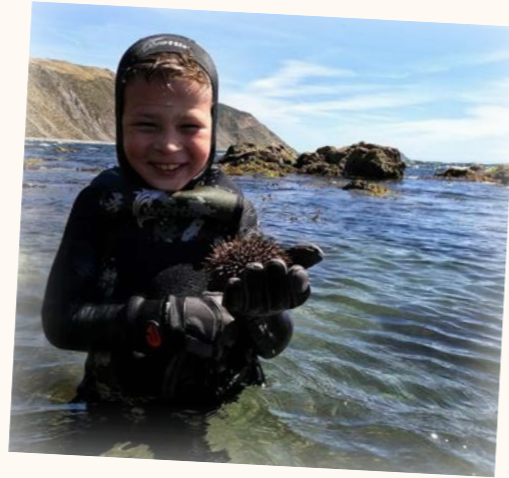
Community Joy Board

Thank you for sharing your moments of joy with us!
We know the whole community loves to see what others are achieving, it's pretty special 💙

Want to be featured in the next issue?
Email Lizzie - comms@cfnz.org.nz



Poppy started at home-based care this week!
She's been at home with me for 19 months, so
this is a big step for us both.



"My very first kina dad"
- Kairo on his 3rd dive mission.



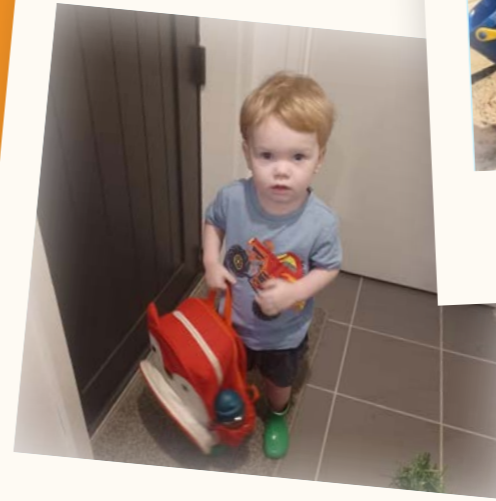
Our son Emerson loves a water fight. The joy
that comes from these times is amazing.



Charlie has explored the north's walking tracks.



Harrison 3 years old has just learnt to ride his
motorbike without trainer wheels!



Alaric was excited to start daycare this week



Liam is excited to start kindy this term
and make some friends!



This is our little man Oscar celebrating being 6
weeks old Thursday. Diagnosed at 4 weeks, we
are just starting our journey.



Max competing in his first horse show.



Dominic just chilling at the skatepark



In September I gave birth to our 3rd child,
a baby boy.

Giving ourselves permission for relaxation...

Lisa Burns

Life is a beautiful resource, seasons change, generations of people come and go. Even though this is a natural cycle of life, it's becoming more and more common in our world today that we don't allow ourselves the time to rest, relax and recharge.

There has been so much happening around us over the past two years, levels of external impact which make it more challenging to manage our mind and our thoughts as we navigate and live through so much change. Our mind can get fixated on worries and fears as we become stressed and try to control what we can't control, struggling with our feelings about how life should be.

Some of us are so fiercely independent, determined to keep going that our greatest strengths can become our greatest weaknesses, taking a long-term toll on our minds and bodies.

Why is it so hard to change the way we think and feel?

Keeping us alive in a time when our lives depended on our physical safety, our primitive brain in today's modern world hasn't evolved. Our brain can become fixated on anything it identifies as a potential threat to our survival and it can't tell if that's a lion or feeling overwhelmed by stress. It will remain fixated on the threat until it's neutralised causing us anxiety and stress that gives us tunnel vision.

Shifting from catastrophising thoughts to counteracting them

"Think positive", or "glass half full", we get told. Reframing our thoughts from negative to positive might feel a bit pollyannaish. It can be a stretch too far for many of us to take an upsetting thought or feeling and attempt to flip it into something positive. Sometimes it's just time, life experience and age that gives the skill and practice to manage our thoughts and even then, it can still be a challenge. Start in the middle, the ground between where you are and where you want to be. It's not a negative thought, and it's not super positive either, it's just a place where you can find some calm and peace.

How can we manage better?

Stress and overwhelm have become part of our everyday lives. Managing CF means living with very real challenges, and on top of that, small things can feel like big things triggering a "fight or flight" response if we don't take the time our bodies and brain need to relax. "Just relax" we get told, easier said than done! When we can relax though, it doesn't just feel good, it's important for our overall health, supporting sleep, mental focus, and our ability to cope. Juggling all our responsibilities can make it hard to make time for yourself but just five or ten minutes a day can make a difference - even if it's those precious few moments of quiet while you're in the bathroom!

When it comes to relaxation, the easier it is, the more likely we'll make time for it. Below we've shared a few tips and ideas that we hope will help.








MASTERING RELAXATION

- 1 Time and place**
Choose a spot that you can be undisturbed and fits in to your daily routine.
- 2 Get comfy**
Wearing something loose and relaxed will allow you to get into a comfortable position inside or outside.
- 3 Start slowly**
Begin with just 3 to 5 minutes, tools for relaxation like the apps suggested below can help you get started.
- 4 Find a motivation**
Recognise why you need to relax, what can help motivate you? Do you need to feel calmer, happier, be less stressed or more focused? Knowing why can help us make a commitment to making ourselves a priority.
- 5 One moment at a time**
Relaxation is a journey, not a sprint. There are abundant resources everywhere from inspirational quotes to sleep stories, guided breathing apps and soundscapes where you can transport yourself to the calming seas, a forest or lake. Try an aromatherapy diffuser, a hot bath, some soft music, or even a gentle walk.
- 6 Stay mindful**
Hold on to the calm; carry the awareness of how you feel and use it to ground you for the rest of your day, evening or use it to get a good night's sleep.

Remember that it's not selfish to take time out. Our brain needs to learn to become less distracted, there is no success or failure, just becoming more aware of how we're feeling and allowing ourselves to relax. If in doubt breathe out.



Download from The App Store or Google Play.

-  Aura
-  Calm
-  Headspace
-  Mentemia
-  Paced Breathing

Trikafta Campaign

Vertex submitted its application to Pharmac for funding of Trikafta in early July 2021. Pharmac’s expert clinical advisors, including those who treat CF, assessed the application and it was considered by the Respiratory Subcommittee in August 2021.

In late October 2021, Pharmac announced that the Subcommittee had recommended Trikafta as a high priority for funding and released its assessment. The recommendation covers people with CF who are six years and older and have one or more copies of the F508 del mutation, or other mutation responsive to Trikafta.

Pharmac also announced that it had undertaken an economic health assessment and had added Trikafta to the list of medicines that it wants to fund – its Options for Investment list. In doing so, it described Trikafta as a “paradigm-shifting” treatment for CF.

Pharmac then took Vertex’s application to its Pharmaceutical and Therapeutics Committee (PTAC) in November 2021 to seek additional advice on how Trikafta should be ranked against the other medicines Pharmac wants to fund.

Pharmac released PTAC’s recommendations on 23 February 2022. After three months of waiting and hoping, the news we received was desperately disappointing. PTAC recommended downgrading the priority for funding Trikafta from high to medium and defer decisions on funding for those aged 6 – 11 years, and for those with genetic mutations responsive to Trikafta in vitro.

CFNZ does not believe that PTAC has undertaken a fair assessment. Investing in Trikafta would bring life-changing benefits to Kiwis with CF by substantially improving their quality of life and life expectancy, and enabling them to live almost normal lives. It would also provide significant benefit to the NZ health system through significant reductions in medical treatment, hospitalisations, and lung transplants.

It is clear what needs to happen now. Pharmac needs to reconfirm that Trikafta is a high priority for funding for those aged 6 years and older. Pharmac also needs to commence negotiations with Vertex to agree a fair and reasonable price for Trikafta. The Government needs to provide Pharmac with



the funds to ensure everyone who would benefit from Trikafta has access as soon as possible. With the support of the CF community, CFNZ will do everything it can to make this happen.

Trikafta Petition

Carmen Shanks’ petition to the House of Representatives seeking public funding of Trikafta is a key part of the current campaign.

The petition closed on 28 February 2022 and achieved an astonishing 42,234 signatures. An amazing feat by Carmen, Trikafta for Kiwis and the CF community for spreading the word far and wide.

CFNZ is working with Carmen and Trikafta for Kiwis to arrange presentation of the petition to make sure that Parliament is fully aware of the urgent need to fund Trikafta in New Zealand. This is even more important now given the PTAC recommendations.

We’re working through the challenge of how to make the presentation in the current COVID-19 Red Traffic light setting and, at the same time, involve as many people with CF as possible without meeting each other face to face. It’s a challenge but business as usual for those with CF.

We’ll provide an update on how you can be involved as soon as the timing and arrangements for the petition are finalised.

A decade of CF modulators

February 2012 was a major turning point in the long journey to improve the lives of people with CF.

Vertex’s CF modulator therapy – Kalydeco - was approved by the US Federal Drug Administration (FDA). The first therapy to treat the cause of CF rather than the symptoms, it changed lives for the better and saved them too.

A decade on and there are now four Vertex CF modulator therapies available – Kalydeco, Orkambi (FDA approved 2015), Symdeko (2018) and Trikafta (2019). Together, these medicines provide the potential to treat more than 90 per cent of people with CF and are funded in many countries around the world.

Unfortunately, New Zealand has to date publicly funded only one – Kalydeco. Finally funded in March 2020, after six years of consideration by Pharmac, it is available to about 40 Kiwis with CF but not to everyone who would benefit.

A very small number of Kiwis with CF have been able to privately fund CF modulator therapies or gain acceptance to Vertex’s Trikafta Managed Access Programme. But for the majority current treatments continue to address only the symptoms of CF and not the cause. This situation cannot go on any longer.

Every Kiwi with CF needs access to a CF modulator therapy that would benefit them to make sure they have a future to look forward to. Pharmac and the Government need to give us a reason to really celebrate a decade of CF modulator therapies. CFNZ is doing all it can to make this happen.

Medsafe gives safety consent

Medsafe gave consent for the use of Trikafta in New Zealand in December 2021, less than six months after Vertex submitted its application. Medsafe had granted the application priority status, meaning that the assessment process was faster than usual.

Medsafe’s consent is a major milestone in providing access to Trikafta and CFNZ is very grateful to Medsafe for undertaking its work so quickly.

Medsafe also granted consent for the use of Symdeko in New Zealand in January 2022, meaning that three of Vertex’s four CF modulator therapies have successfully moved through Medsafe’s process. Only Orkambi remains to be granted consent and, at the time of writing, Medsafe had sought further information from Vertex.

Australia moves closer to Trikafta

In December 2021, Australia’s Pharmaceutical Benefits Committee (PBAC) recommended that Trikafta should be listed on the Pharmaceutical Benefits Schedule. PBAC recommended the listing for people with CF age 12 years and older who have at least one F508 del mutation. PBAC had previously considered Trikafta on a number of occasions, so this positive recommendation was very welcome. However, final price negotiations still need to be completed before people are eligible to receive Trikafta.



Review of PHARMAC

In March 2021, the Government initiated an independent review of Pharmac to assess how well the agency is performing, whether its performance could be improved, and whether its current objectives maximise its potential to improve health outcomes for all New Zealanders.

The Review was undertaken by an independent panel chaired by consumer advocate Sue Chetwin. The Review Panel was expected to submit its Interim Report to the Minister of Health in August 2021, and its Final Report by December 2021. However, the Review Panel's Interim Report was delayed, and released by the Minister of Health in late 2021.

The Interim Report contains no recommendations but makes preliminary observations about Pharmac's performance. While it acknowledges the challenges faced by Pharmac, the report does not paint a positive picture.

The Review Panel concluded that Pharmac is underperforming in helping to remove inequitable health outcomes and that its prioritisation approach appears to disadvantage Māori, Pacific peoples, disabled people, and those with rare disorders. It also concluded that decision-making is opaque and is perceived as slow, and that engagement with consumers and patient advocacy groups needs to be much more meaningful.

The Review Panel also raised concerns that there may be an excessive focus on costs and that Pharmac's cost-saving model may not be the right one to meet future health needs.

The issues and concerns raised in the Interim Report very much reflect those raised by CFNZ in its submission, similar to many other patient groups. CFNZ would like to thank the Review Panel for genuinely listening and for acknowledging the issues and concerns raised.

The Pharmac Review Panel is now preparing its Final Report which will make recommendations on a range of matters, including requirements for integration with the new health and disability system and Pae Ora (Healthy Futures) Bill, governance arrangements, and funding for people with rare disorders. The Panel now expects its Final Report to be delivered to the Minister of Health by 28 February 2022.

CFNZ is keenly awaiting the Pharmac Review Panel's Final Report. We are pushing for the Review Panel's recommendations to be fully addressed as part of restructuring the health and disability system.



Pae Ora (Healthy Futures) Bill

The Government is moving ahead with restructuring the health and disability system, amalgamating all District Health Boards and establishing a new agency – Health New Zealand – to plan, commission and deliver health services alongside a new Māori Health Authority. The proposals also include consideration of the arrangements for Pharmac.

In October 2021, the Minister of Health introduced the Pae Ora (Healthy Futures) Bill into Parliament to make these changes. The Bill was referred to the Pae Ora Legislation Select Committee and submissions invited. More than 4,700 submissions were received. CFNZ took the opportunity to make a written submission on the Bill in December 2021 and, in January 2022, was able to present that submission to the Committee.

CFNZ's submission and presentation focused on the need for people with CF to have access to high quality care wherever they live, ensuring there are appropriately resourced centres with skilled clinicians, and access to up-to-date medicines and equipment. CFNZ also stressed the need for the new health agency to develop national standards of care and treatment guidelines for people with CF to ensure consistency of care across New Zealand.

The arrangements for procurement of medicines were also a key focus, with CFNZ proposing significant change to enable access to modern medicines such as Trikafta and other CF modulator therapies. The importance of taking full account of the Pharmac Review Panel's Final report was also raised.

The Pae Ora Legislation Select Committee is due to report back to Parliament on the Bill on 27 April 2022, and the changes are planned to take effect on 1 July 2022.

Advocacy timeline

Our advocacy timeline summarises our fight for fairer access to life-changing medicines for people with cystic fibrosis.

Access to CF Modulator Therapies

2021

June

- Vertex submits its applicant for regulatory approval of Trikafta to Medsafe. Medsafe accepts the application and grants it priority approval status enabling a shorter time for evaluation. Vertex also submits its application to Medsafe for approval of Orkambi.
- Vertex establishes a Managed Access Programme in New Zealand to provide Trikafta free of charge to Kiwis with CF who are in critical need. The Programme is managed globally by Vertex and access is through CF clinicians. CFNZ is delighted that Vertex has established a Managed Access Programme in New Zealand for the small group of Kiwis with CF in critical need but notes that it is no substitute for public funding of Trikafta for all those who would benefit.



July

- Vertex submits its application for funding of Trikafta to Pharmac, a major step forward in securing access for Kiwis with CF. The application to Pharmac follows Vertex's application to Medsafe in June for approval for the use of Trikafta in New Zealand.
- CFNZ submits a comprehensive package of information to Pharmac to support Vertex's application for Trikafta.



August

- Pharmac's Respiratory Subcommittee considers Vertex's application for Trikafta.

September

- OJ Daniels shares his story of life with CF and why we need Trikafta in a documentary in the Being Me series on Attitude.
- Auckland Branch hosts a Zoom hui on Trikafta, including hearing from Kirsty Parsons on her experience of being on one of the original trials for Trikafta and how she's faring four years on.



October

- CFNZ writes to Pharmac to support the consumer application to widen access to Kalydeco.
- Pharmac confirms that the agenda for its November Pharmaceutical and Therapeutics Committee (PTAC) includes consideration of Trikafta. PTAC's role is to provide objective advice to Pharmac and make recommendations as to whether medicines should be included on the Pharmaceutical Schedule.
- Pharmac confirms that it wants to fund Trikafta and adds it to its Options for Investment List. This follows a recommendation from the Respiratory Subcommittee that it be given high priority for funding and an economic assessment by Pharmac. PTAC's role in considering Trikafta at its November meeting will now be to help Pharmac to determine how Trikafta should be ranked against all the other medicines it wants to fund.
- Patrick Gower interviews Pharmac on TV3 News about the announcement that it wants to fund Trikafta. In the interview, Pharmac's Director of Operations Lisa Williams calls Trikafta a paradigm-shifting treatment for cystic fibrosis.



November

- Ashley from the Bay of Plenty shares his story on how gaining access to Trikafta through Vertex's Managed Access Programme has changed his life.

December

- Medsafe, New Zealand's Medicines and Medical Devices Safety Authority, has given consent for the use of Trikafta in New Zealand. Medsafe assesses medicines to make sure that they meet New Zealand and internationally recognised standards for quality, safety and efficacy before they can be distributed here.
- Australia's Pharmaceutical Benefits Committee (PBAC) recommends Trikafta should be listed on the Pharmaceutical Benefits Schedule. PBAC recommended the listing for people with CF 12 years and older who have at least one F508 del mutation.

2022

January

- Medsafe grants consent for the use of Symdeko in New Zealand – three of Vertex's four CF modulator therapies have now successfully moved through Medsafe's process. Only Orkambi remains to be granted consent.

February

- Pharmac releases the minutes of the meeting of its Pharmaceutical and Therapeutics Committee (PTAC) where Trikafta was considered. Contrary to the Respiratory Subcommittee's recommendation, PTAC recommends only a medium priority for funding Trikafta for those aged 12 years and older and defers decisions on funding for those aged 6 – 11 years, and for those with genetic mutations responsive to Trikafta in vitro.



Follow our progress in advocating for access to medicines and high quality care for New Zealanders with CF

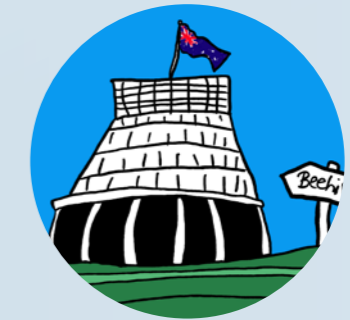
Scan the QR code using your phone camera

Other Advocacy Projects

2021

May

- Government announces \$200m for Pharmac over 4 years, with \$40m for 2021/22. Total funding over 4 years is only half what Pharmac has said it needs to fund the medicines currently on its recommended list. This does not include any new medicines coming through, such as Trikafta.
- Patient Voice Aotearoa's (PVA) petition to double Pharmac's budget and review Pharmac is presented to Parliament. The petition has over 100,000 signatures and CFNZ and the community participate across the country.
- CFNZ meets with Sue Chetwin, Chair of the Independent Panel undertaking the review of Pharmac. The meeting provides an initial opportunity to raise issues over the funding of new medicines, including Trikafta.



October

- The Minister of Health introduces the Pae Ora (Healthy Futures) Bill into Parliament. The Bill proposes the amalgamation of District Health Boards, the establishment of Health New Zealand and a Māori Health Authority, and arrangements for Pharmac.

December

- The Minister of Health releases the Interim Report of the Pharmac Review Panel. The Panel concluded that Pharmac is underperforming in helping to remove inequitable health outcomes and that its prioritisation approach appears to disadvantage Māori, Pacific peoples, disabled people, and those with rare disorders.
- CFNZ makes a written submission on the Pae Ora (Healthy Futures) Bill seeking to ensure that people with CF have access to high quality care wherever they live, including appropriately resourced centres with skilled clinicians, and access to up-to-date medicines and equipment.

2022

January

- CFNZ presents its submission on the Pae Ora (Healthy Futures) Bill to the Pae Ora Legislation Committee.

Supporting our community

We truly appreciate all the amazing people, volunteers, sponsors, friends, donors, funders and supporters. Your kindness and generous contributions make it possible for us to provide

a wealth of support options at every stage of your journey. Here are a few of the support services provided over the last year.



Fieldworkers



Sue

Chani

Jude

Gretchen

CF Fieldworkers can help with:

- **Providing information** to parents about CFNZ services and support available to them.
- **Attending outpatient clinics** and discussing welfare needs with people attending.
- **Meeting with families** in their homes or wherever they are most comfortable.
- **Developing and updating careplans** to ensure treatments regimes are being followed.
- **Providing information** about the **Breath4CF program** which provides grants to facilitate exercise.
- **Sending vouchers** to people for post-transplant help and who are on home intravenous antibiotic treatment.
- **Collecting and distributing** nebulisers, oxygen concentrators and other essential medical equipment.
- **Transporting people to hospital** for procedures as often unable to drive due to anaesthetic, or due to social issues, ensuring children are seen at clinic.
- **Supporting inpatients** during often lengthy hospital stays.
- **Meeting with teachers**, visiting kindergartens, ECE's and schools to inform and educate.
- **Supporting people** at their appointments with WINZ when their health deteriorates so they are no longer able to work.
- **Liaising with Housing NZ** for appropriate accommodation for families.
- **Educating workplaces about CF** and supporting people with CF in new jobs.
- **Working with young adults** as they transition to Adult Services, offering support and advocacy during clinic visits.
- **Supporting people** during the lung transplant process.
- **Support for whole family** during palliative care, ensuring access to services and resources.

Please note face to face support has been limited due to the Covid-19 protection framework.

For more information about support services get in touch with your CF Fieldworker or visit www.cfnz.org.nz.

SUPPORT

Fundraising Superstars

We're forever grateful to the amazing people who go out of their way to raise awareness and funds to support the CF community. Here's a few of the fundraising superstars over the last 12 months.



Dargaville Bake Off
Dargaville



Auckland Ladies Morning Tea
Auckland



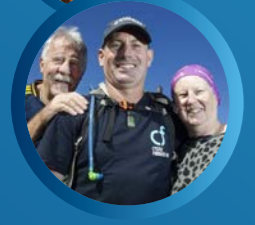
Bread4CF
Auckland



A good wee cookbook - Sam Mannerling
Auckland



Shining Peak 5% project
Taranaki



Walking with Wings
Taranaki



Christmas Tree Festival
Wellington



Bidr Auction Waikato
Waikato



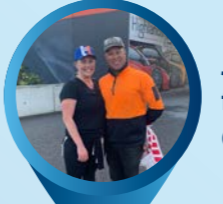
Charlie's Chocolate Fish Fundraiser, sold 1000's of chocolate fish
Tauranga



Duncan "IronDuncs"
Hawke's Bay



Running and cycling the length of NZ - Twice!
New Zealand



Josie running 200km for Sweatember
Cromwell



Breathe easy - Ride4CF - Cape Reinga to Bluff
Bluff



Canterbury Golf Tournament
Canterbury



65 Hours of Bowls
Ashburton

