WINTER 2019

CF news

CYSTIC FIBROSIS NZ

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CF in the workplace, updates on advocacy and research, meet the latest CF Super Stars, plus all the latest news

Welcome to the CF News Magazine

In this edition we'll be focusing on two major areas in CF; research and advocacy.

We'll look at how our campaign for access to medications and better care is tracking, and share with you what we're doing to advance CF research in New Zealand.

Join us in celebrating the achievements of our latest CF Super Stars, and read about Mark, who was awarded the Mark Ashford Scholarship in 1998!

We'll also be exploring the world of work with cystic fibrosis, with a fabulous interview with Jo, a dispensary technician in Christchurch, plus an overview of our new CF at Work guides.

Finally, this magazine is for the CF community and our wonderful supporters. We'd like to thank you for your support and ask you to tell us what you think of this publication.

Please take a moment to fill out our survey at *www.surveymonkey.com/r/G5KN29B*

If you have any specific feedback on this issue, or ideas for future editions, email us at comms@cfnz.org.nz.

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What's inside

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A big thank you

A sincere thank you to the Blue Waters Community Trust and John Ilott Trust for generously funding the printing and postage costs of the Winter CF News magazine.

We'd also like to thank the many readers that have sent in donations to assist with the costs of researching and writing the CF News magazine. We greatly appreciate the support

Lastly, thank you to everyone who shared their stories, provided advice, guidance and offered expertise for the CF News magazine.

Further information

For more on our support services, information, advocacy, and research, or to learn about cystic fibrosis, visit **cfnz.org.nz**. You can also sign up to receive our monthly CF Panui e-newsletter through the website.

Your feedback is valuable. We'd love to hear your comments.

First nurse prescriber

Viv Isles, a clinical nurse specialist in Canterbury, has become the first nurse qualified in writing prescriptions for cystic fibrosis.

Viv recently completed a post-graduate diploma including papers in pharmacology and prescribing. She can now prescribe medications off a set list, within her scope of practice.

As a member of the Children's Outreach Nursing Service at Christchurch Hospital, Viv provides nursing care, ongoing education and support for children with chronic respiratory conditions. When she visits families at home, she can now write prescriptions on the spot. This saves time on trips to the GP or hospital for a script, and saves money, as faxed scripts can cost up to $5.

One parent of a 9-year-old said, "My son used to be scared of hospital and would spend a third of his time there. Now that Viv administers many of his tests and monitors his condition at home, it means that as a family we live a reasonably normal life."

New resources available for our community

As part of our ongoing work to develop New Zealand specific resources for our CF community, we've published online and in print, a range of new guides and brochures.

The development of these guides included a comprehensive review of all our current information available, interviews with people with cystic fibrosis and their families and ongoing collaboration with our community to ensure the publications met the needs of our community.

Thanks to many generous grants and collaboration with people with cystic fibrosis we now have available:

Guides

- A guide for parents and caregivers of children diagnosed with cystic fibrosis
- A guide to cystic fibrosis for family, whanau and friends
- Starting School – A guide for parents and caregivers of children with cystic fibrosis
- Starting School – A guide to cystic fibrosis for primary schools and teachers
- A guide to cystic fibrosis for employers

Brochures

- Understanding cystic fibrosis
- Support available for people with cystic fibrosis and their families
- Where to from here?

Since the launch of our new website, new logo and new branding last year, these resources have continued to build on increasing awareness of cystic fibrosis in New Zealand and supporting people with cystic fibrosis.

You can download the resources from our website or ask for a printed copy from your fieldworker or the CFNZ office.

Fourth CF fieldworker to be appointed

CFNZ is very grateful to The Lighthouse Foundation for providing funding for a fourth fieldworker, yet to be appointed. The position will be funded for three years from the Foundation, a private charity administered by Goldman Sachs.

Chief Executive Jane Bollard said that, at present, the three fieldworkers have equal caseloads but differing amounts of travel. One works across the South Island, one covers the southern half of the North Island and the third covers Northland, Auckland, Bay of Plenty, and Waikato regions.

"The CF Insight Survey identified the need to make it easier for people with CF and their familes to access CFNZ support services. Cities have the largest number of people with cystic fibrosis, but we know that CF has a greater impact on those living regionally who spend more time off school and work as they need to travel for treatment and care. Parents and caregivers also need a lot of support in the first few years after a new diagnosis.

"We are delighted to have confirmation of this funding, and are currently assessing how best to use this additional resource to meet the needs of the CF community," she said.

CF fieldworkers are skilled, qualified social workers who provide emotional and practical support to individuals and families living with CF. For further information visit **cfnz.org.nz/get-support/personal-and-family-support.**

Turn to page 17 to get to know our three CF fieldworkers!

New exercise gear for the Auckland City Hospital gym

Thanks to a generous grant from the Lion Foundation, the Auckland City Hospital Level 7 physio gym is packed with new exercise equipment.

The physios (pictured) told us they had to try the equipment out first to make sure it was safe for everyone to use, but we're pretty sure that was just an excuse to be the first ones to use it!

We hope our CF community enjoys the benefits of the new equipment.

Auckland Hospital gifted portable oxygen concentrators

In May, three portable oxygen concentrators were donated to Auckland Hospital for use by people with CF.

The donation was made possible by a generous grant from the JM Thompson Charitable Trust and the fundraising efforts of the 2018 Willis Tower Watson Auckland Marathon team.

The oxygen concentrators allow people who need oxygen therapy to have the flexibility to get out and about without the noise and weight of a regular oxygen cylinder.

Photo: Kath from Auckland Branch gifting the concentrators to Auckland adult CF nurse specialist Cath Lamont

CF Conference 2019

This year's Australasian CF Conference will be held in Perth from 3 to 6 August. The biennial conference brings together health professionals and members of the CF community from around the world to discuss advances in CF research, care, and drug development, and to exchange ideas about ways to improve the health and quality of life for people with CF.

Ten high-calibre, international experts will be presenting at the conference, along with numerous local speakers. CFNZ Chair, Jane Drumm, has joined the committee organising the lay conference programme. The CE, plus fieldworkers and branch chairpersons will represent CFNZ, along with some parents who have been funded to attend.

Five Feet Apart raises awareness of CF

The perils of cross-infection for people with CF were brought to public awareness with the release of the movie Five Feet Apart at the end of March.

The film tells the story of two teenagers with CF who meet in hospital and fall in love. The risk of cross infection means they must always stay a safe distance apart. As their attraction to one another grows, however, they are tempted to ignore the rules and take their chances on life and love.

While some critics panned the movie as a less than realistic tearjerker, others defended it as an entertaining teen romance, saying it was not meant to be a documentary about CF.

Audiences were generally more enthusiastic – giving it a score of 79 percent on Rotten Tomatoes compared with 54 percent positive reviews from critics.

In New Zealand, people with CF commented favourably on Facebook, even if Five Feet Apart was not their favourite kind of movie.

"If nothing else, it raises some much-needed awareness," some said.

Many are looking forward to viewing the DVD which was released in New Zealand at the end of June – even though they know it will make them cry!

Note: Some of the treatments portrayed in Five Feet Apart differ from what's offered in New Zealand. If you have any questions about the movie or would like to discuss any of the issues it raises, please contact your clinical team, CF fieldworker, or visit our website, ***www.cfnz.org.nz***

Communications hire a talking point

After searching high and low to fill the Communications Coordinator role, we are thrilled to introduce **Lizzie McKay**. Lizzie joins the marketing and fundraising team at the national office in Auckland, and will be responsible CFNZ's communication activities across a range of print, digital and social media.

"I'm delighted to be joining the CFNZ team as the Communications Coordinator. I've worked in digital agency land in Auckland for the last five years, most recently as a Marketing Project Coordinator," Lizzie said. "My main focus was around creating, managing and executing social content for a variety of clients. It's been a goal to be involved in a not for profit, and I'm passionate about CF and the work CFNZ does, so naturally, this feels like a pretty perfect fit! I'm so looking forward to learning the ropes, and building some great connections and relationships within the community."

Lizzie will play a vital role in developing and delivering engaging content for the CF community, volunteers, donors and supporters, and helping to communicate the important work we do.

Welcome, Lizzie!

CF Awareness Week

12-18 August 2019

Join us on our annual major campaign this August, highlighting how CF affects people, the work we do, and inspire donations. **CF Week is 12-18 August 2019**, with street collections happening on Friday 16 and Saturday 17. Set up a bake sale, sell chocolate fish, organise a mufti day, sausage sizzle, or help shake a bucket all in support of research, advocacy, support and information for Kiwis with CF. Visit the CFNZ website for more information on getting involved.

Winter raffle now open

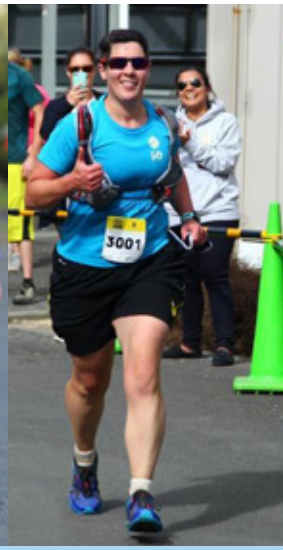
Last winter we saw record numbers of families needing heating assistance, extra nutrition, supplements and emotional and financial support during hospitalisations. Help keep Kiwis with cystic fibrosis healthy this winter by participating in the CF Raffle.

Prizes include an LG 49" Smart 4K TV worth $2,000, three nights at any NZ Choice Hotel, a Harvey Norman gift card for $500, plus heaps more!

Visit *www.cfnz.org.nz/raffle* to get tickets and further information about prizes. Raffle entry closes 31 July 2019.

PARI t-shirt fundraiser raises over $8,000

Last year a special fundraiser was launched to mark the 50th anniversary of both CFNZ and PARI BOY. For every person who wore a PARI BOY/CFNZ T-shirt at a sports challenge or event, PARI donated 10 euros (approximately $16) to CFNZ. In April this year CFNZ was informed that an incredible 289 New Zealanders participated, raising a fantastic 5,000 euros / $8,000 NZD! We cannot thank PARI enough for its support and to the participants who whole-heartedly embraced the initiative.



Cross infection policy updated

After several years work by its Clinical Advisory Panel, CFNZ has released revised guidelines on infection prevention and control in non-healthcare settings.

The position statement is designed to educate people with CF, their caregivers and families about the risks of person-to-person infection from respiratory pathogens of any kind. It recognises that there is no reliable way to completely prevent the risk of cross infection.

Research has now confirmed, for example, that coughing and sneezing can project pathogens into the air further than originally thought. Bacteria in airborne droplets can be 'live' for up to 45 minutes and be detected up to four metres from the source.

People without CF rarely carry the particular bacteria that cause chronic infection in CF, but they may still carry other bacteria or viruses that cause problems for people with CF. Thus, "The concept of 'less threatening' bacteria is no longer accepted and all pathogens should be considered as potentially transmissible and universal precautions should be taken."

The statement outlines general hygiene guidelines for people with CF, as well as particular policies relating to households, schools, workplaces, sports, pools, gardening, pets and animals, and events organised by CFNZ.

CFNZ has updated its cross infection policies to reflect the new global findings indicated in the position statement. The key elements of the new policies are:

- The risk of infection remains for people with CF who have had a lung transplant
- The 'safe' distance between people with CF has been increased from 1.5 metres to 4 metres
- CFNZ will only invite one person with CF to be physically present at an indoor event organised by CFNZ, regional volunteer branches, groups or committees
- CFNZ will not support or promote any event where more than one person with CF is likely to be, for example, Christmas parties, the offer of holiday homes, or CF camps and retreats.
- As there is no way to completely prevent the risk of cross infection at outdoor events, the safest approach is for people with CF not to attend. If they do, they should be separated from others with CF by a distance of 4 metres.

While the position statement and policies are more restrictive than before, other options such as social media, webinars, and online conferences are becoming more available. Several CFNZ branches are now including adults with CF at their meetings via free video conferencing technology.

Sharing the warmth

Winter is set to be warmer for one Christchurch family, thanks to a generous donation from Gavin Lowe Energy.

Kimberley Swaney and her family, which includes Noah and Elijah who both have CF, were this year's recipient of a heat pump, installed in their home on 14 May.

"Gavin Lowe Energy has been amazing and we're so grateful for the gift of warmth and good health for our family," Kimberley says.

"Being able to set a timer for the heat pump to come on in the morning before the children get up for school is fantastic," she says.

CFNZ southern fieldworker Sue Lovelock, Christchurch chairperson Melissa Skene and Kimberley met Mike Little, Business Development Manager of Gavin Lowe Energy at Kimberley's house to formally thank them for their donation.

Mike talked about the challenge of the placement of the heat pump in a narrow hallway ceiling to heat the three bedrooms and choosing the right heat pump to do the job.

"It's been so good to be able to help a family who is affected by cystic fibrosis," Mike says.

A huge thank you to Mike and the team at Gavin Lowe Energy who has donated a heat pump to a Canterbury family with CF for the past three years. Your donation and support are a great example of how businesses can really make a difference to families in our CF community.

Below: Kimberley and Melissa with Mike Little from Gavin Lowe Energy

Building a positive future

Young graduate architect Caleb Skene is "over the moon" about winning the 2019 Mark Ashford Scholarship.

"I got the news when I'd just walked in the door after work one day. It's amazing and I'm very over the moon about it. I feel proud knowing the meaning behind the award and grateful for the possibilities it will give me in developing my knowledge," he says.

Caleb, aged 22, was awarded the scholarship for excellence in his Bachelor of Architectural Studies degree which he completed last year at the Ara Institute of Canterbury. His tutors commended him for his innovative thinking and willingness to push boundaries.

This year, Caleb is working at MC Architecture Studio in Christchurch, the same company that employed him part-time during his studies. One of his former lecturers offered him a job there on completing his degree, so he started fulltime the day after he graduated last November.

Caleb hasn't made up his mind yet how he will use the $3000 scholarship. He may continue his architectural studies with a master's degree, possibly in Melbourne. Or he could put the money towards a trip to Europe to see some of his favourite buildings, such as Frank Gehry's Dancing House in Prague or Peter Zumthor's Bruder Klaus field chapel in Germany.

"I used to think that residential architecture was where I would go in my career, because of the connection you can create with clients building a new home. But I also want to learn more about commercial architecture because there's a wider range of things you can study in that field. I've still got to figure it all out," he says.

Caleb struggles with recurring aspergillosis (a fungal lung infection) and has undergone bowel surgery in the past, but says each spell in hospital "just creates an extra challenge to take on board and overcome". He tries to stay fit and keep on top of his daily medication routine, but admits that working fulltime can be draining compared with life as a student.

"It's annoying when I need time off for hospital admissions, which can sometimes be up to two weeks. It's frustrating to feel I'm not pulling my weight, but I have a very understanding boss who is keen to learn more about CF and what he can do to help."

The immediate future may be unclear but one thing's for certain – Caleb is not letting cystic fibrosis stand in the way of achieving his goals. Receiving the top CFNZ study award is further motivation to keep going.

"I'm very honoured to be commended in this way for my achievements in architecture. I pushed myself hard during my studies and it's great to be acknowledged for my efforts."

A building suspended over ruins was Caleb's entry in EXIT, an exhibition by Ara's third year architectural studies students last November.

“Congratulations, Caleb”

That’s the message from a director of TelferYoung Tauranga, this year’s sponsor of the Mark Ashford Scholarship.

Mark Passey knows what it’s like to study while facing the challenge of cystic fibrosis. He was awarded the scholarship himself in 1998, using it to fund his Bachelor of Business degree from Massey University. Now he wants to give back to help those who may not have had the same opportunities he’s had.

“I’ve been really lucky – I’m not as sick as most CFers, and not a lot are as privileged as I am. For me, it’s about making it easier for individuals to follow their dreams,” he says.

Cystic fibrosis has been part of Mark’s life since he was six years old and he takes a pragmatic, matter-of-fact approach to the disease. A couple of years ago, he was thinking about launching his own scholarship programme. When he became the adult representative on the CFNZ board and heard that the Mark Ashford Scholarship needed another sponsor, he decided it was simpler to avoid duplication. TelferYoung will sponsor the scholarship for at least three years.

Mark says there are no particular requirements for how the money is spent – he just wants to make life easier for students with CF.

“Congratulations to Caleb. It sounds like he’ll be using the scholarship for a worthy cause and to help achieve his goals.”

TelferYoung is a home and property valuation company with independent offices nationwide. Mark joined the company in 2006 and covers a range of valuation work across Tauranga and the Western Bay of Plenty.

The Mark Ashford Scholarship is awarded each year to a person with CF who has achieved excellence in tertiary education. It was established 22 years ago in memory of Mark Ashford who had CF and made a huge contribution to home health with his work on the Fletcher Group’s Healthy House insulation book.

TelferYoung
Property Valuers & Advisors

Jayde Knight

CF Achiever, Education

Sixteen-year-old Jayde Knight is the youngest recipient of the CF Achiever Award this year.

Jayde is in Year 11 at Hamilton Girls’ High School where she has an outstanding academic record. She is in extension classes for all her subjects, and last year took Level 1 science subjects a year early, scoring high merit and excellence. This year she is continuing with Level 2 chemistry and physics, and also studies Japanese and Art (painting).

Health issues have been a challenge, with frequent clinic visits and daily treatments impacting school hours and time catching up with friends.

Jayde is living proof that, in her words, “a sick kid does not have to be held back”. She’s passionate about gaining the skills and experience to be able to speak up on behalf of others, and remove the stigma of chronic illness.

“This year I continued with debating and now that I am a senior, my skills are sharpening, to prove to myself and others that being diagnosed with cystic fibrosis should not hold you back, it should empower you to make the most of the life you have. I want to be a voice for my generation and for future youth,” she says.

One thing she’s really looking forward to is a school art history trip to Europe in September, visiting six cities over four weeks. A special highlight will be the new digital “Workshop of Lights” art museum in Paris.

For Jayde, the trip has meant hours spent fundraising, and a course of intravenous antibiotics to make sure she is well enough to go, but she’s confident everything will be OK. The Achievers’ Award has given her a real boost.

“It’s amazing to win this award. The trip is expensive and the fact that others are supporting me to do this is, like – ‘Wow!’” she says.

Briar Lomas

CF Achiever, Arts

Briar is a top student in her Bachelor of Design course at Massey University in Wellington. Now in her fourth (honours) year, she achieved an A+ in all her courses last semester and was nominated as one of only two students to join the Design Institute of New Zealand's Student Council.

Briar says she has always been involved in the arts. At high school she was heavily involved in dramatic theatre, achieving to a high standard in Trinity College exams and the Duke of Edinburgh's Hillary Award. At university she continues to strive for excellence in the graphic arts. Eventually, she hopes to gain experience in a general design studio, and then focus on print design.

"I am extremely passionate about this topic and have loved the three years I have spent studying it so far. Going into my final year, I am feeling hugely motivated to do the absolute best I can," she says.

Briar considers herself "very lucky" that her cystic fibrosis has not hindered her too much in daily life. She makes sure she sticks to her health regime, and stays physically active through social netball and rock-climbing. Being vigilant around other people with colds and infections is important too.

"There have been times when I've got sick but my tutors have been really great about it and my flatmates are fantastic. I think my achievements show that I've been managing it well," she says.

Briar was also a CF Achiever back in 2014 when she was head girl at Rodney College, north of Auckland. She says the 2019 award will be "super-helpful" in paying for fees and a new laptop, allowing her to focus wholeheartedly on her final major design project.

"It's such a lovely validation, very affirming. I've put lots of work into my studies and it's a wonderful incentive to keep achieving the best," she says.

She makes sure she sticks to her health regime, and stays physically active through social netball and rock-climbing. Being vigilant around other people with colds and infections is important too.

Georgie Northcoat

CF Achiever, Leadership

Georgie is only 20 years old but has already demonstrated her flair for leadership and innovation.

Now in her third and final year studying clothing, textile sciences and marketing at Otago University, Georgie's aim in life is to "solve universal problems, challenge myself and create." She is currently interested in social entrepreneurship – developing a micro-credential scheme which would give students experience in charity work and recognition by potential employers.

Georgie has been a class representative and has participated in advisory councils for the Canterbury and Southern District Health Boards. She's also co-managed an Estee Lauder cosmetic counter for the past year. Last summer she was an intern at Fisher & Paykel Appliances where she produced a highly successful project on washing machine performance and was commended for her planning, presentation, and insights.

"She has a wisdom for opportunities and risks that is remarkable, not only for someone of her age and limited work experience, but also for someone ten years older," her supervisor said.

Cystic fibrosis has never held Georgie back. She manages her treatments well and is vigilant about prevention, making sure she has at least eight hours sleep every night. She's also using her marketing skills to build CFNZ's Instagram platform and blog about her experiences.

"It's been part of my life since I was young so I've never known anything different. It's just something you have to fit into your day," she says.

With a Bachelor of Applied Sciences under her belt when she graduates in December, Georgie has several career options open to her. Right now, the idea of starting her own business is the most exciting.

"I would like to upskill and fill the gaps in my personal knowledge. This grant is the first opportunity I will have to invest in myself and pursue entrepreneurship at these early stages of development," she says.

She has a wisdom for opportunities and risks that is remarkable, not only for someone of her age and limited work experience, but also for someone ten years older.

Survey provides insight about living with CF

Last year we conducted our biggest ever insight survey of our CF community. The purpose of the nationwide survey was to gather information about:

- the wellbeing of our CF community
- the extent to which CF impacts on people with CF quality of life
- how helpful our organisation has been
- what we should do in the future to improve the wellbeing and quality of life for people with CF
- set benchmarks and enable our organisation to assess progress made toward achieving our mission and objectives over time.

A literature review was undertaken to find examples of other wellbeing and CF support needs surveys which allowed us to compare our results to other populations of people with CF and the wider New Zealand population.

The survey went to 405 families (of an estimated 505 people with CF in New Zealand). 209 families provided feedback, a response rate of 52% which is exceptionally good and provides very reliable data about the wellbeing, quality of life, our service and aspirations of our CF community.

Results

The results showed that the self-reported health status of both people with CF and parents was lower than the wider New Zealand population. The better the health of people with CF and parents, the more likely they were to report they were satisfied with life. Both people with CF and parents' satisfaction with life was also lower than the wider population. Parents with very young people with CF were most likely to report being less satisfied with life. There were no significant differences between the self-reported health status or satisfaction with life by gender or age for people with CF.

There was a statistically significant difference in the extent to which CF impacts on people's lives by region: people with CF who live outside the main treatment centres (i.e. outside Auckland, Wellington and Canterbury) were absent more often from school, work or other daily activities due to illness or treatments compared with people with CF living in the main centres.

Half the respondents said our services were 'extremely' or 'very' helpful, particularly fieldworkers, grants and information resources.

Fieldworkers were people they know and trust who were connected to the wider CFNZ organisation, CF community and health care services and who provided emotional and practical support. Information resources informed service users about CF, what they could do to help themselves and the different kinds of resources, services and support available. Grants made it possible to purchase or pay for equipment or services that they may have otherwise struggled to afford.

The services rated as most important, rather than most helpful, were advocacy, building a strong CFNZ and supporting CF research.

The feedback about what people wanted life for people with CF to be like in five years showed clear trends. They wanted:

- easier access to the best medicines and treatment for children and adults across the country (i.e. outside the main treatment centres)
- the day-to-day management of CF to be less demanding, expensive and time-consuming
- their day-to-day experience and functioning to be less impacted by the disease.

CFNZ's next steps

The results from the survey have provided our organisation for the first time with qualitative data about the health and wellbeing of our community. We have used the survey findings to adapt and improve our service provision and to focus our strategic and annual plan on the areas identified as being important to people with CF.

We continue to incorporate the recommendations to advocate for better care and treatment of people with CF in New Zealand and to ensure we meet the needs of our CF community. A recent example involves the use of the findings in a submission to the Health and Disability Systems Review and presenting the findings at the CF Clinical Forum in Christchurch.

We will repeat the survey every 3 years to see if we've made any improvements in decreasing the impact of CF on people's lives.

CF in the workplace

Life with CF can be a challenge, and no more so than in the workplace. What do you tell your boss and colleagues about your condition? How do you negotiate time off for doctors' appointments or hospital visits?

Interviews last year told us that adults with CF would like ready access to information about the condition to give to their employers. So, we set about producing a new guide that explains what you need from your employer so that you can do your best at work.

The guide points out that adults with CF are experts in managing their own health day-to-day. They've spent their life focused on staying well, eating well and living as normal a life as possible. What they need most from employers is an understanding of CF and how it impacts their life.

The guide provides an overview of cystic fibrosis, and how it is treated and managed. It includes tips for maintaining a healthy work environment and aspects of employment law that employers might need to consider.

It's your choice as to how much to tell your colleagues about CF. Here's how others have dealt with talking about CF in the workplace:

I personally like to go under the radar and only tell people about my condition when necessary.

I like to be treated the same as everyone else in terms of ability, but also have that extra bit of implicit understanding when I need to duck away.

I have been selective about the people I tell about having CF, just because I know that some people do not really understand.

Disclosing CF is something I let happen naturally as people get to know me. I hope people would feel that they can come and ask me about how CF affects me directly if they're curious.

Working with CF

Jo Donaldson considers herself fortunate that she has an understanding employer and has never had a problem managing her cystic fibrosis at work.

In fact, she says she feels "a bit of a fraud" because her CF is not very severe and she hasn't had to have lots of time off due to sickness.

Jo works a 40-hour week as a dispensary technician at a Christchurch pharmacy, as she's done for the past 22 years. She was diagnosed with CF as an adult when bronchiectasis showed up after recurring bouts of sinus trouble and bronchitis.

"When I found out, I decided to take my immune system in hand and get it in top condition. I'm a great believer in large doses of Vitamin C, which absolutely makes a difference," she says.

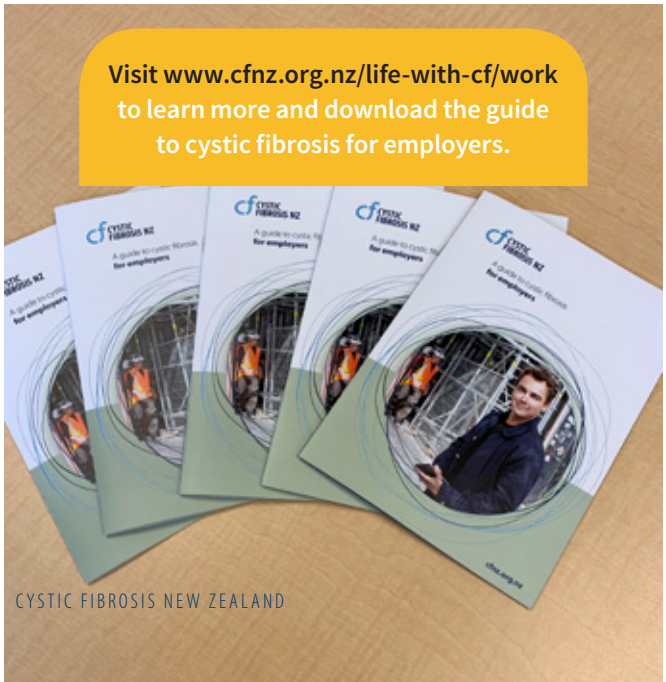
Of course, working in a pharmacy means Jo has access to all the supplements and vitamins she needs, but it also means greater exposure to sick people.

"I don't have to be the one who goes out front to deal with them, though, as I work mostly out the back. My colleagues all know about my CF and are very helpful and understanding. It's never a problem to take the afternoon off every three months for appointments at the respiratory clinic, or if I have to go home early when I'm fighting an infection," she says.

Jo walks in her lunch hour and rinses her sinuses regularly. Occasionally, she's needed a course of intravenous antibiotics but she can manage this herself, even while at work.

Her advice for others coping with CF while working is:

"Talking is the biggest thing – let your employer know how you're feeling. There may be some flexibility but it's only fair to make your needs known. And do your level best to look after yourself and your own immunity – get enough sleep, eat well, exercise and be careful around people with colds or the flu."



Supporting an employee who has CF

Here are some ways employers can help employees with CF stay well and reach their full potential at work:

- Communication is key. Talk with your employee about how CF affects them and any questions you have. Keeping communication channels open benefits both employers and employees.
- Every person with CF is under the care of a specialist CF team. Appointment dates are often known well in advance, allowing employees and employers time to organise workload cover if needed.
- Sometimes people with CF need a course of intravenous antibiotics. Most adults learn how to do these themselves and can manage them around their work schedule after a short time in hospital (usually 3–5 days). The option to work from home, if possible, during this time can be helpful.
- Be open to considering temporary part-time work arrangements, working from home or flexible hours if a person with CF becomes unwell.
- Tiredness or stress can make CF symptoms worse. Encouraging manageable workloads and minimising stress can help reduce the amount of time off work.
- Mutually agree on a plan to cover any time off from work apart from sick leave, such as allowing employees to accrue leave in advance, discretionary leave, or leave without pay.
- CF affects every person differently. An employee with CF will not necessarily be off sick more often than other employees. Most time off is for clinic appointments and is known well in advance.
- People with CF catch 'bugs' more easily than people without CF. A workforce that encourages people not to come to work if they are unwell is beneficial for everyone.
- CF is unlikely to require urgent medical treatment. Most adults with CF are experts at understanding the risk of infection and most risks can be minimised.

Breath of life

A little over a year ago, **Makena Houston** had to fight for every breath. She could hardly walk, had lost a lot of weight and had so little energy that even watching TV was exhausting. An oxygen cylinder was her constant companion, night and day.

If you'd told her then what her life would be like today, she wouldn't have believed you.

Makena has just spent 10 days singing and dancing in a Hawke's Bay production of *Mamma Mia!* She's started her own communications business, goes to the gym, enjoys Pilates and recently climbed Te Mata peak, almost without coughing.

"My life now is incomparable to what it was and that's all thanks to my donor," she says.

Makena had a double lung transplant in April last year after waiting seven and a half months with respiratory failure. Diagnosed with cystic fibrosis before she was born (her older brother also has CF), Makena was reasonably well until the age of nine when open-heart surgery resulted in a permanent loss of lung function. Two years ago, aged 24, her lungs got much worse and she was put on the critical list for a transplant.

The operation saved her life but recovery turned into a rough journey that's not over yet.

Complications after surgery meant Makena was unconscious for two weeks and in intensive care for almost two months. When she woke up, she couldn't hold her head up or move her hands, let alone sit or stand. Her muscles had wasted away. Eating, drinking and talking were out of the question.

Worse still, her kidneys shut down during surgery and didn't kick back into life afterwards, as expected. Makena has been left with diabetes and the need for dialysis three times a week. A kidney transplant may be an option, with her mum as donor, but the risks are high, especially for someone who has already had one major organ transplant.

"I have to take more pills post-transplant than I did before and the time on dialysis has just replaced the time I used to spend on physio and nebulising!" she says.

Not that Makena is complaining. She says she would pick dialysis "any day" over her pre-transplant condition.

"It's not the lifestyle I was promised but I'm grateful every day to my donor. I'm doing what I can to stay well and learning to live on my own terms again," she says.

That means continuing to do what she loves – singing and performing in shows and musicals. Makena also hopes to build her business so she is financially free to travel and spend time with her family and friends.

Spotlight on fieldworkers

Gretchen Kitching – Northern

Mobile: 029 773 1398
Email: gretchen@cfnz.org.nz

Gretchen has worked in the health sector ever since she was 16 years old, mainly supporting people with intellectual and physical impairment.

After completing a Bachelor of Social Work degree, she became a CF fieldworker just over two years ago, working with more than 200 CF patients in Auckland, Northland, Bay of Plenty and Waikato. Hospital clinics and home visits take up most of her time, with trips to the regions every few months.

She likes the variety of her work and the opportunity to form special relationships with those she supports.

"I enjoy getting to know individuals and families and witnessing the huge amounts of strength and resilience people possess. My role is flexible so I'm available to see people in the community – in their homes or out for coffee or a meal. Spending time with people outside the hospital environment encourages a different relationship that is not dominated by the medical realm. This gives me the opportunity to learn about other aspects of people's lives apart from CF," she says.

Gretchen also has special responsibility for lung transplant patients, letting them know how to access CFNZ grants and other assistance, or simply being there for emotional support. The need for more organ donors is at the top of her wish list as she's seen first-hand what a difference this gift can make to a person with CF, and their family.

Jude Kelly – Central

Mobile: 021 1926 234
Email: jude@cfnz.org.nz

Jude's message to people with CF and their families is, "Have hope for the future but enjoy today." Based in Wellington, she cares for the CF community in the capital as well as Hawke's Bay, Central Districts and Taranaki.

About half her time is spent travelling to visit patients, some of whom live in quite remote areas – and she loves it.

"Visiting people in their own environment is so different from seeing them in the hospital, which is often an anxious time. In their own home you get to know people a lot better. It can be quite a lot of fun," she says.

As a registered social worker and counsellor, Jude has experience in hospitals, schools, and a variety of support services, both here and overseas. She's also been an army officer and spent some time in the ready reaction force after the Christchurch earthquakes.

Jude works with welfare agencies and schools to support families and educate teachers about CF. She thinks New Zealanders with CF are well-served by the medical profession and by CFNZ itself.

"There is a lot of good things happening in the CF community. In New Zealand we do really well at picking up on CF early through the heel-prick test, so children get better treatment and have a chance of having a really good life. I encourage people to do whatever is on their bucket list – get out there and make the most of life," she says.

Sue Lovelock – Southern

Mobile: 021 0222 1203
Email: susan@cfnz.org.nz

Sue Lovelock has been a fieldworker with CFNZ for 13 years, which makes her our longest-standing staff member. Based in Christchurch, she covers the whole of the South Island, travelling to most regions four times a year to visiting about 160 adults and children with cystic fibrosis.

Sue trained as a social worker and describes her role as "walking alongside people in their cystic fibrosis journey". For her, it's about building relationships with patients as well as their medical and support teams. At clinics and annual patient reviews, Sue acts as an advocate for patients, asking questions on their behalf, or helping seek more information about new procedures or medications.

She also helps people access welfare grants and other support services. Sometimes, her job simply means being a reassuring presence for out-of-towners or people visiting government agencies such as WINZ.

"I love working with people and every day is different. I find people with cystic fibrosis so inspiring. Many of them have such a rough deal but they're strong and they never quit. Some seem to have a particular type of stroppiness that keeps them going! They've figured out what works best for them and just get on with it," she says.

A very special 21st birthday

Trudy Hannan had a lung transplant 21 years ago. As far as we know, that's the longest anyone with CF has survived in New Zealand following a transplant. This is her story:

I was diagnosed with CF as a 1-year-old baby. I was a very active child and teenager, competing in athletics, netball and any sport I could. I loved running, which must have helped my lungs stay strong and clear. I can recall only one hospital admission for pneumonia when I was 12 years old.

When I was growing up, my parents never treated me any differently from my sister. I did everything a non-CF child would, including a lot of things I'm sure we aren't supposed to. Sometimes ignorance is bliss and this was so true of my upbringing, for which I am grateful.

When I did get very sick at age 22, I was admitted to hospital with 24 percent lung function, one dead lung and no more than three months to live. I was married and working fulltime then. I knew deep down I was dying but I was too scared to admit it, so I just kept going until I couldn't any longer. I most definitely have the CF stubborn streak!

A double lung transplant was my only option to live. It was a terrifying time – being on oxygen 24 hours a day, using a BiPAP machine at night, and having breathing treatments every four hours to try and keep open the few airways I had left. While waiting for the call, my sister moved in with me and looked after our home. She was amazing – I owe her my life.

I was living in America at the time so my transplant was at Vanderbilt Hospital in Nashville, Tennessee. The date was 8th February 1998.

Life after transplant was like being reborn. To this day, I can still recall taking a breath in hospital – and it just kept going and going. I couldn't believe it. I didn't think my CF was too bad but at that second I realised I had never ever taken a full, deep breath before. It was one of the most profound moments of my life, and I am forever grateful I was able to experience that. After all, how many adults can say they remember their first breath?

Having a transplant has filled my soul with an everlasting gratitude for absolutely everything. Nothing in my day-to-day life goes unappreciated. I feel so lucky to have a second chance and it's changed my entire perspective on all life has to offer.

Of course, a transplant is also hard work, with the rehab and risk of infection due to a suppressed immune system. Two years after transplant I was diagnosed with chronic rejection,

which was terrifying. I recall my team saying, "Don't worry, Trudy, we have drugs for everything." I took that on, relaxed, and gave myself over to the doctors and medicine. Since then I have had several admissions for chest infections and blocked bowels but, thanks to New Zealand's amazing medical team and drugs, I have always bounced back.

Twenty-one years later, I am still here with my transplanted lungs and filled with enough gratitude to last a lifetime. I consider myself incredibly lucky to be alive and so blessed to be supported and loved by everyone close to me and my phenomenal family, immediate and extended. They give me hope and I wouldn't be here without them. There have been times when I have wanted to give up the fight, but they have kept me going and given me reasons to keep on fighting when it gets tough.

My advice to others with CF is:

Live your life to the full. Don't let CF "have" you; instead, you "have" it. Don't let it stop you from doing anything, ever. CF is just a thing, not a life, so don't let it take over yours.

Opposite: Trudy with husband James
Above: Trudy with Zara
Below: Trudy with Isla

At times I have given myself over to the doctors when I knew I needed to let them do what they do. But you are your own best advocate; you know your body better than anyone else, so follow your instincts always. I think people with CF are born with an innate sense of self – we're warriors in disguise!

For years I didn't let those who love me help me when I needed it but now I do and it's so liberating. Being honest opens all the doors for kindness.

I work fulltime and have been open and honest with my employer and colleagues, and they are understanding and kind.

My donor was a 28-year-old woman, the mother of two children, who got shot in the head by her partner. I spoke with her mum about three months after my transplant, and all I could say to her was "thank you". She told me her daughter was able to donate all her organs. What a woman!

Although CF is a really awful card to be dealt, I do consider myself lucky. Every breath I take is a gift and I am grateful for the insights CF has given me. It's a blessing to be alive.

Fighting to stay healthy

Young **Leah-May Elliott** is using an ancient martial art in her fight with cystic fibrosis. In April, the 9-year-old won a silver medal at the Brazilian jiu-jitsu national championships in her grade (grey belt).

She's been following the sport for about two years, after the family moved to Rotorua and her older brother and sister got involved. Their coach asked Leah-May if she'd like to have a go, and from that moment, she was hooked.

Brazilian jiu-jitsu focuses on grappling and fighting on the ground. At first, Leah-May didn't have the strength to compete and she struggled with tiredness and breathlessness. Through training three nights a week at the Gracie Gym, however, she's gradually got stronger.

Rolling around on the ground is good for her as it helps to loosen mucus, says her mother Janelle Rumney.

"She's a very determined little girl. She just got cleared to go to the nationals only a few days beforehand, after two weeks of intravenous antibiotics for pneumonia. Then on the drive home from her national medal win she told me she was getting sick again at the competition and hadn't wanted to tell me. Two days later she ended up in hospital for three nights."

Apart from jiu-jitsu and mountain-biking with her dad and brother, Leah-May is also interested in penguins – to the point of obsession, according to Janelle. "She's loved penguins ever since she was three years old."

Last year, thanks to the Make A Wish foundation, Leah-May's dream came true when she got to pat a koala and see the little blue penguins at Phillip Island near Melbourne.

"My hopes for Leah-May are that she's happy and lives life to the full. Sometimes she feels different from other children and why does it have to be her, so we talk about that and try to work through it. I tell her that there are lots of kids who have different things, whether it's epilepsy or diabetes or they're in a wheelchair.

"Sometimes it's difficult to stay positive but I want her to have as normal a life as possible. Leah-May talks a lot about her hero Ellie Simmonds the Paralympic swimmer and how she didn't give up trying, so neither will she," says Janelle.

Your story matters

As a charity, we're competing with thousands of other not-for-profits in New Zealand to make our voice, our cause and our people heard. We need your help!

Your story is important – whether you have CF yourself, or are the parent of a child with CF, or another family member. We'd love to hear from anyone who is affected by CF and would like to tell their story for the benefit of others.

Personal stories help put a human face and voice to the work we do. They help raise awareness and encourage others to become involved in improving the quality of life for people with CF. Your voice, your words and your story will support our vision for the future of lives unlimited by cystic fibrosis.

The stories page of our website features five people who have shared their experience of CF and the impact it's had on their own life or the life of someone they know. Why not check out their stories and let us help you tell your own?

Email Sarah at *sarah@sayline.co.nz* if you'd like to share a few words. Thank you.

Kalydeco recommended for funding

As you know, we received news on 8th May that Kalydeco has been recommended for low priority funding for the treatment of cystic fibrosis with the G551D mutation.

This is a major step forward on the path to public funding for this vital medication, so thanks to everyone for your support in the campaign so far. There is still a long way to go however, so while we can celebrate this victory, we need to continue our efforts to gain full access.

The May announcement is good news in one respect – it reverses the 2015 recommendation from PHARMAC's Pharmacology and Therapeutics Committee (PTAC) to decline funding for Kalydeco. But the low priority ranking is disappointing because, earlier this year, the Rare Disorders Subcommittee recommended Kalydeco be funded with a medium priority.

"We have been playing this game a long time and this life-changing medication is high priority – therefore access to it should be a given. Anything less is unacceptable. Our people are high priority, their lives matter and Kalydeco works," says Jane Drumm, CFNZ Board Chair.

What happens now is that Kalydeco joins a list of about 100 other treatments and devices waiting for funding. Unfortunately, the ranking in this list is not made public, nor is there a specific timeframe for medicines to be funded. The priority ranking (low, medium, or high) can also vary, and does not necessarily dictate the order in which the medicines are actually funded. Some approved life-saving medications have sat on this list for years.

We are continuing to talk with PHARMAC and Vertex Pharmaceuticals, which manufactures and supplies Kalydeco, about what the decision means and the next steps to take.

Meanwhile, PHARMAC plans to overhaul its decision-making processes to give greater clarity around which medicines are – and are not – being considered for funding. PHARMAC is chronically underfunded and there have been repeated calls for an inquiry into how the agency operates.

Cystic Fibrosis New Zealand, Kalydeco for Kiwis, and the CF community will continue to put the pressure on until Kalydeco is not only funded, but accessible to all who need it.

Compressors and nebulisers

Since the early 1990s, CFNZ has been buying the compressors that power nebulisers used for inhaling medication, and supplying them to most people with CF in the country. Last year, however, the CFNZ Board decided to stop providing this equipment and transfer this responsibility to local DHBs.

This was because of significant health and safety concerns about CFNZ supplying medical devices, when the organisation deals with CF patients but has no medical employees. The shift means CFNZ can focus on fundraising for other equipment that improves quality of life for people with CF.

We are communicating with all DHBs to make sure they are aware that they are now responsible for supplying essential basic equipment. Some DHBs have now purchased the equipment themselves through EBOS, New Zealand's suppliers of PARI nebulisers and compressors. We will continue to follow up until we are sure that every DHB has organised access to the equipment.

CFNZ's Clinical Advisory Panel has developed guidelines for DHBs, and EBOS and PARI are available to give support, product advice and training to DHB staff.

Still in limbo

PHARMAC's decision to fund Kalydeco hasn't resolved any of the uncertainties for the Porter family, and others like them.

Eddie and Emma's son **Otis**, now aged 2, would benefit greatly from the medication, which would effectively "switch off" the CF gene, possibly adding decades to his life expectancy.

At $350,000 a year, they can't afford to buy Kalydeco themselves. Even though they can assume it will now be publicly-funded, however, there's no knowing when.

"We don't know where it's ranked on the list, what else is on the list, or how long that list is. Being on the list doesn't actually mean it will be funded any time soon. It could be anywhere from 10 days to 10 years."

Emma and Eddie spend three hours a day treating Otis, starting with antibiotics and probiotics, then 20 minutes of physiotherapy, followed by sessions with a PEP mask. Although their boy is generally very well, winter is always "terrifying" and they are very careful about being in public and exposing Otis to infection. Daycare is too risky and Emma, who is carrying their second child, has not been back in paid employment since Otis was born.

If Otis had access to Kalydeco, his quality of life would be very different.

Above: Otis in the park
Below: Porters at home with PEP mask

"People on Kalydeco go to hospital less, they get fewer infections and if they do get infections, they clear them a lot quicker. They don't have the degradation of quality of life, so rather than the slow decline we see with CF normally, it's effectively paused on Kalydeco," Eddie says.

The Porters are grateful that Kalydeco has been recommended for funding but are considering their options, including shifting to Australia where the drug has been publicly-funded for the last five years.

"It's a huge amount of money [to spend on a drug], but the other side of it is that it's a huge amount of quality of life for our child," Eddie says.

For now, though, they're committed to continuing with the Kalydeco for Kiwis campaign, pushing for better outcomes for the 30 or so others who share Otis' CF gene and need access to Kalydeco. Like them, Emma just wants the uncertainty to end. Her message for PHARMAC is:

"Make this next part of the process more transparent so we know what's going on, so we can plan our life."

Meet Lisa Woods

Lisa is our cystic fibrosis 'Advocate', contracted to develop a national advocacy strategy and and with branches identifying local priorities.

Lisa loves working with community and not-for-profit organisations to achieve their vision and aspirations, helping them plot a way forward that will make a difference. Her experience spans roles in communications, policy, community development, advocacy and management. Most recently she worked as communications manager for Community Networks Aotearoa and director of Every Child Counts.

Lisa lives in Wellington with her partner Tim and their son Zach.

Research roadmap

CFNZ is making progress on a national research strategy designed to deliver real benefits for people with CF.

The strategy is the product of a workshop organised by Cure Kids and CFNZ last year for researchers, clinicians and members of the CF community. One of the key findings was the need for an interconnected research, clinical and community strategy, and someone to drive it – hence the joint funding of Dana Felbab as research development manager (see separate article).

The aim is to identify research priorities for our CF community, connect the CF clinical and research communities locally and internationally, and determine how New Zealand can contribute to global scene. Five key elements to achieving this are outlined below.

CF database

The PORT CF registry is vitally important to the future of CF research in New Zealand. This database is internationally regarded due to its high coverage of the New Zealand CF population. It could be improved further to make it more complete, more visible, and more usable by healthcare professionals, the CF community and researchers.

CF community portal

New Zealand's smaller CF population is an advantage for research, and the CF community has a strong track record in driving better care, both nationally and internationally. The creation of a community portal would connect people with CF even more directly with researchers, by:

- enabling individuals to have a say on research priorities
- connecting people with CF together via social media
- creating a strong community voice
- giving individuals a say on how to improve their day-to-day management of CF
- enabling practical, crowd-sourced studies
- supporting individuals who need to travel to participate in CF research projects.

A first step would be to ask people for their top three CF challenges that they would like researchers to focus on.

Research community

Some excellent CF research is happening in New Zealand, but funding applications tend to be driven by individual teams rather than as part of a cohesive plan. CF research would benefit from greater coordination within New Zealand as well as stronger links to clinicians and to international efforts.

Initial steps include plans to:

- audit New Zealand's current CF and related research
- look for existing findings that can be translated into clinical practice
- determine research priorities that include CF community preferences, exploit our research strengths, and explore the unique aspects of CF in New Zealand.

CF clinical and research network

A key goal for CF clinical care is to make sure all New Zealanders have access to the same best practice care, no matter where they live. People with CF would benefit greatly if national or international research was translated into improved clinical practice. We have a well-connected paediatric CF network but the adult CF network needs development.

CF clinicians currently have very little time available for research. With an adequately supported CF clinical network, however, New Zealand could undertake smaller, practical initiatives that deliver better results for patients, as well as benefiting the international community.

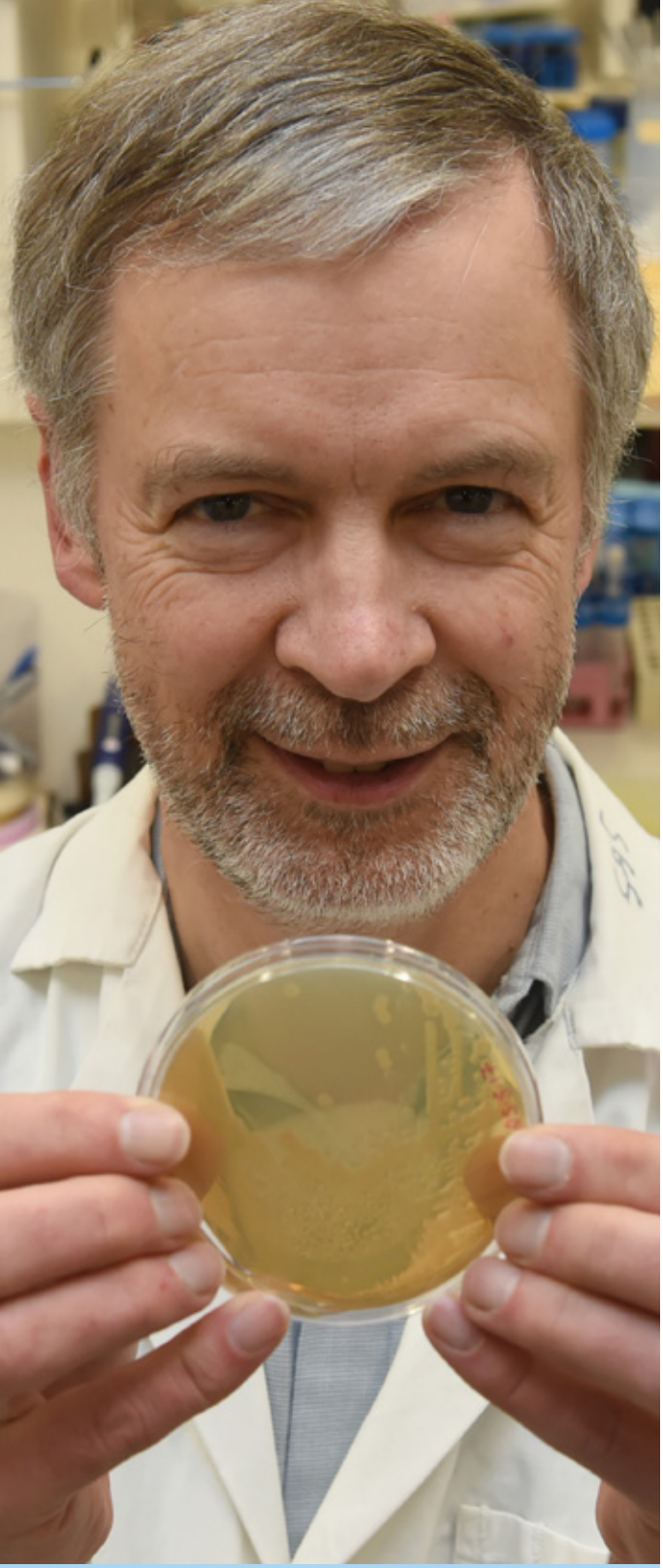
Suggested steps to creating a clinical and research network include:

- training a network of clinicians around New Zealand with an award to recognise innovation in patient care/best practice
- improving patient care by firstly capturing data from the PORT CF registry, then trialling an intervention and measuring impact
- examining CF management, such as looking at day-to-day treatments to determine what can be dropped
- attracting younger people into CF care, e.g. clinical researchers and PhD candidates who are closer in age to people with CF
- creating a dedicated CF research fellowship.

The initial focus could be on creating a telemedicine service for New Zealanders, which would itself be the subject of a research project.

International connections

New Zealand is a great location for clinical trials because many expensive new CF drugs are not used here, creating what is known as a drug-naïve population. Sponsors are reluctant to fund studies here, however, due to low patient numbers. Part of the research strategy involves building stronger connections with funders and overseas pharmaceutical companies to explore how New Zealand patients can be more involved in clinical trials.



Professor Iain Lamont is originally from Scotland, but after a brief spell in Australia, has lived in Dunedin for the last 30 years. He is based in the Department of Biochemistry and has a long-standing interest in how bacteria cause infectious disease.

In collaboration with overseas researchers, he and his team are seeking to uncover the genetic mutations that lead to antibiotic resistance in one of the world's most problematic 'superbugs'.

photocredit: Gregor Richardson

Research projects

Antibiotic resistance in lung bacteria

Antibiotic resistance, a worldwide problem, is of particular interest to people with cystic fibrosis because of the nature of the bacteria that thrive in their lungs, especially *Pseudomonas aeruginosa*.
Professor Iain Lamont from Otago University studies the molecular genetics of bacteria that cause infectious disease. He and his research team are investigating whether *P. aeruginosa* are deprived of oxygen in the lungs and whether this alters the effectiveness of antibiotics used to treat infection.

What is the problem?

The lungs of patients with cystic fibrosis can become blocked with thick sticky mucus which can block off oxygen supply to parts of the lungs. The mucus also creates a rich breeding ground for bacteria like *P. aeruginosa.*

Chronic infection with this bug can greatly reduce lung function, diminish quality of life and lead to premature death. Antibiotics can keep the nastier elements of infection at bay, but increased resistance by the bacteria means this treatment is becoming less and less effective.

Antibiotics are refined and developed in labs – an oxygen-rich environment where they can easily kill *P. aeruginosa.* In oxygen-starved lungs, however, it may be a different story. Professor Lamont and his team suspect that when the bacteria experience oxygen deficiency, antibiotics are less effective.

How was the project conducted?

The team collected sputum samples from CF patients with *P. aeruginosa* infection from four hospitals and a wide range of ages. From these samples, they analysed the levels of bacterium's antibiotic resistance as expressed by two particular genes. They also analysed genes that indicate whether the bacteria are oxygen-starved.

They isolated *P. aeruginosa* from the samples, grew it in the lab and exposed it to a range of antibiotics and different levels of oxygen. The results for lab-grown bacteria were then compared with the samples taken directly from infected lungs.

What were the results?

The researchers found a remarkably wide range of antibiotic resistance, both in the sputum samples and the laboratory-cultured bacteria, but the reasons for this variability between patients have still to be determined. It appears that lung bacteria can manifest in many different ways, depending on the severity of infection and the level of oxygen. Decreased amounts of oxygen do reduce the effectiveness of at least one antibiotic used to treat patients with cystic fibrosis, and prolonged antibiotic treatment serves only to increase resistance. It is hoped that further research into exactly how *P. aeruginosa* flourishes in human lungs will lead to more targeted and effective antibiotic treatment.

This research is ongoing and is supported by CFNZ, Cure Kids and the New Zealand Lotteries Board. The team is grateful to the sponsors for allowing such research to be carried out.

A silver bullet?

Silver has been used since ancient times as an antiseptic and purifying agent. It's still used today in wound dressings, to destroy microbes on catheters, and combat "superbugs" that are resistant to traditional antibiotics.

Now, researchers at Victoria University of Wellington have discovered that silver could be a new weapon in the fight against Pseudomonas aeruginosa – a bug well-known to people with cystic fibrosis. This bacterium is resistant to most antibiotic treatments as it forms a protective biofilm in lungs affected by cystic fibrosis.

PhD student **Jennifer Soundy** began her research by looking at small sections of DNA known as aptamers that can be custom-coded to target and kill pathogens.

> "Originally we hoped to bind our DNA aptamers to the Pseudomonas aeruginosa and kill it, but unfortunately that wasn't successful," Jennifer says. "We decided to see if we could bind an antimicrobial to the DNA aptamers instead and then use the aptamers to deliver the antimicrobial straight to the pathogen."

She picked silver as the antimicrobial, attaching it in very small clusters to the aptamers. Because of the way silver attacks multiple cell processes, most bacteria have not developed a resistance to it. The downside is that it's toxic to humans in large doses.

> "We hoped to kill the pathogen without harming the host. Excitingly, we were successful – the pathogen died within ten minutes – and because we could target the pathogen directly we could use very small doses of silver that shouldn't be harmful to the human body," she says.

This discovery has many applications. As well as targeting pathogens directly, the silver could also be combined with other treatments to make them more effective.

> "This could even make pathogens vulnerable to antibiotics that currently have no effect on them. We were able to combine our silver treatment with tobramycin, which is an aerosol treatment currently used to treat Pseudomonas aeruginosa lung infections in cystic fibrosis patients, and initial tests suggest that this combination would be very effective in treating that infection."

Jennifer graduated in May with a PhD in Biotechnology. Her supervisor Dr Darren Day from the School of Biological Sciences, and other researchers, will continue working to improve the effectiveness of aptamers to target different pathogens.

Source: School of Biological Sciences, Victoria University, Wellington.

Introducing Dana

Dana Felbab, Research Development Manager, works one day a week at the CF office in Auckland. CFNZ has co-funded her with Cure Kids to kick-start a national research strategy, following a workshop held last year to explore research priorities and opportunities.

Dana's background is in molecular biology (genetics and pathology). She has experience as a medical laboratory scientist, first in New Zealand and then the United Kingdom. Soon after returning from the UK, she joined Roche Diagnostics NZ as a product specialist and after five years there, moved to Auckland UniServices as a business development manager, securing funding for innovative research at the University of Auckland. Dana is passionate about translating New Zealand research into real world outcomes that can improve people's lives.

Welcome Dana!

The social and economic cost of CF

More funding is needed if CFNZ is to go ahead with a major study on the social and economic impact of cystic fibrosis.

Deloitte New Zealand has been commissioned to do the research, which will cost about $100,000. Almost half of this has already been funded through contributions from CFNZ's research fund and the Wellington and Otago branches.

Cystic fibrosis is New Zealand's most common life-threatening genetic disorder, currently affecting around 500 adults and children, including 1 in 3,500 newborns.

The report will look at health sector and treatment costs as well as productivity losses and the cost of caring for people with CF, both formally and informally. It will also consider more qualitative data such as loss of wellbeing and the impact of premature mortality on families.

The research could have major implications for how cystic fibrosis is managed in the health and education systems, and will be a useful tool in CFNZ's advocacy for new drugs, healthcare provision and support services to improve the lives of everyone with CF.

Chief Executive Jane Bollard said that there is currently no data available on the total cost of CF in New Zealand and this information would be a vital adjunct to the information held in our national CF database, PORT CF.

"The cost of CF is something that affects all of us – the lives of people with CF as well as their carers and family in terms of education, work and social impact. It's a significant and valuable piece of work for us that represents an investment in the future of our community. It's a steep investment for the charity and we're looking for funders and donors to support the project," Jane said.

For further information, please contact Jane at *ceo@cfnz.org.nz*

Did you know?

New Zealanders have a different microbial mix to the typical CF profile. Patients here have fewer pseudomonas and more staphylococcus infections. Research could be focused on how this unique microbial profile impacts treatment options and outcomes.

Positive results for triple-combo therapy

In March, Vertex Pharmaceuticals announced positive results from two clinical trials of treatments that combine three different medications.

Triple-combination therapies represent a significant advance in CF treatment. According to the Cystic Fibrosis Foundation (US), they have the potential to treat the underlying cause of CF for more than 90 percent of people with the condition.

The first study tested VX-445 combined with tezacaftor/ivacaftor (the two drugs that make up Symdeko) in people with cystic fibrosis aged 12 years and older, who have one copy of the F508del mutation and one minimal function mutation. After four weeks, these patients had a 13.8 percent increase in lung function compared to participants taking a placebo.

The second study included people with CF aged 12 years and older who have two copies of the F508del mutation. Participants who were given the triple combination had a 10 percent improvement in lung function after four weeks compared with those who were given only tezacaftor/ivacaftor without VX-445.

The VX-445 triple combination is one of two therapies that have been undergoing late-stage clinical trials by Vertex since 2018. Preliminary results for the other drug combination were released in November.

After completing Phase 3 trials for both triple combination therapies, Vertex will decide which one to put forward to the U.S. Food and Drug Administration (FDA) for approval later this year, with a decision expected in 2020.

How do triple-combo therapies work?

Triple-combination therapies are known as CFTR modulators because they are designed to correct the malfunctioning protein made by the CFTR gene.

The CFTR protein regulates the proper flow of water and chloride in and out of cells lining the lungs and other organs. In people with CF, mutations in the CFTR gene result in either a defective protein being produced or no protein at all. This is what leads to the build-up of thick, sticky mucus in the lungs and other parts of the body.

In people with certain gene mutations, CFTR modulators help the protein form the right shape, move to the cell's surface and function properly once it is there, regulating the amount of water on the cell's surface and how much chloride can get through.

The three main CFTR modulator therapies are ivacaftor (Kalydeco), lumacaftor/ivacaftor (Orkambi), and tezacaftor/ivacaftor (Symdeko). Because different gene mutations cause different defects in the protein, these medications are only effective in people with specific mutations.

In New Zealand, Vertex will not apply to PHARMAC for funding for Orkambi or Symdeko until Kalydeco is funded. The recent recommendation by PHARMAC to fund Kalydeco could open up negotiations for these precision medicines to be made available in New Zealand.

Source: Cystic Fibrosis Foundation (United States)

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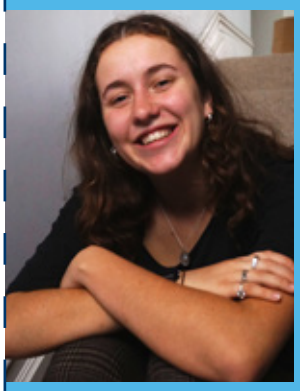

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