SUMMER 2018

CF news

CYSTIC FIBROSIS NZ
Creating Better Tomorrows

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50 Years of Cystic Fibrosis New Zealand

Alana's Adventures, CF on Shortland St, Top Scholars
Research News – Trends in NZ CF Care, Gene Editing in NZ
CF Adult News – "I was lucky to get on transplant list"

CYSTIC FIBROSIS NZ
Creating Better Tomorrows

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COVER IMAGE

CFNZ is thankful to Tayler who is the star of our 50th anniversary cover.

UPDATE FROM THE CHAIR

One tends to reflect at the end of each year and beginning of the next on what has gone before and what is to come. When taking stock with regard to CFNZ I take great pride in many aspects of the organisation.

CFNZ branch committees are the backbone of CFNZ. These hard working dedicated bands of community minded people come together to address the needs of their local CF communities, working with our CFNZ staff to provide support and to fundraise. As a personal benefit many lifelong friendships are formed as a result of being a branch committee member – something I have both observed and experienced.

We have 12 branches throughout NZ, each with their own culture and way of working – within the parameters of our CFNZ policies. Each branch works hard communicating with their community, developing their branch committee as a team, maximizing every opportunity that comes their way and actively heightening CF awareness.

Every month I have the chance to have a phone catch up with the "Chairs" group and am blown away with how much they are doing in their areas. This opportunity to network and share with each other is invaluable and I believe it is a testament to the strength and maturity of CFNZ.

When talking about maturity, CFNZ is 50 this year – a milestone anniversary and one worth acknowledging. We are joined in our 50 year celebration by PARI who are marking their 50th year anniversary of the PARI Boy nebuliser.

Our association with PARI is so strong that CFNZ has been chosen as one of three charities to be recipients of the PARI "Never Short of Breath" Campaign where PARI will use their 50th year celebration to say thanks and motivate people to get active, whilst helping those who perhaps can't breathe freely. Along with many of our branch committee members I have ordered my household allocation of two T-shirts and am ready for my personal challenge of walking up Rangitoto (as I thought this would make an awesome photo to post on the PARI website). For every photo posted (and you can do more than one!) PARI will donate 10 Euro to the organisation of your choice – of course for me that is CFNZ.

My challenge to you is to go get yourself a T-shirt and start getting those photos from NZ up on the PARI website – and encourage everyone you know around the world to do the same.

Something that is also of great importance to CFNZ this coming year is the development of our three year Strategic Plan. The wider CF community will have the opportunity to have input into this process through our upcoming survey and our Chairs and Board have already done some work in this space at our annual Chairpersons' Conference in October last year. This is an exciting opportunity for us all to confirm our vision, priorities, and plan, and I welcome your input.

JANE DRUMM, CHAIRPERSON
Email: **chair@cfnz.org.nz**

CF News is brought to you thanks to...

Blue Waters Community Trust and **John Ilott Trust** both provided grants to enable the publication and printing of this issue of CF News.

Both organisations have supported the CF News in the past and it's wonderful to receive their support once again – Blue Waters Community Trust donated $3000 and the John Ilott Charitable Trust provided an additional $1500. We also received donations from two readers which we are very grateful for.

The CF News celebrates the achievements of people living with the disorder in New Zealand, and features updates about the organisation and latest news. It's thanks to the support of funding agencies that we can continue to publish the newsletter.

To get more immediate updates related to CF in NZ "Like" us on Facebook **@Cystic Fibrosis NZ** or ensure you receive our monthly email Panui updates on the last Thursday of every month – email **comms@cfnz.org.nz.**

UPDATE FROM THE CE

CFNZ is keen to understand what is important to families of young children diagnosed with CF.

Having carried out paeds surveys on alternate years for some time, we took a different tack last year to understand the experiences of families of young children with CF.

CFNZ commissioned Connect+Co to carry out face-to-face interviews with some of our families in Christchurch, Wellington and Auckland to look at how best families could be supported by the organisation.

We very much appreciate the parents who generously gave their time and willingly shared their experiences and stories - some had very young children and were in their first encounters with CF, others had up to four years' experience, some were urban and others were rural.

Key themes from the interviews included:

The need for current, reliable NZ information that parents could access when they needed it. Often told "not to google" parents were left feeling frustrated and found themselves desperately seeking online information from multiple sources not knowing if it was current and correct.

The need for CFNZ to provide sound parenting information and practical support. Parents wanted practical information on subjects including routines, parenting, making their house safe, going out, etc.

Support in managing the transition to childcare/ECE and school. Parents felt the tension of needing to have the role of educator, advocate and parent while also wanting CF not to define their child.

The need for a trusting, supportive relationship with a practitioner. Particularly in the first year and during periods of transition the support of a trusted person who was knowledgeable, available and respected a parent's knowledge of their child was invaluable.

The need for parents and CFNZ to advocate on behalf of their child. The role of the parent as the child's advocate and also the unique role CFNZ has in advocating for children both locally and nationally was key feedback.

CFNZ is currently working through these findings and the ideas presented by parents and Connect+Co.

We have immediately incorporated the feedback into the CFNZ website upgrade with the aim of being the "go to" site for current, reliable information for NZ families. And we have drawn on the need for specific knowledge and practical support in the development of CFNZ's publications of the Newly Diagnosed Guide, the Friends and Family Guide, and the CF at School guide. All guides are being road tested by interested parents to see that they meet the needs.

The role of parents and CFNZ advocating on behalf of children is being incorporated in our advocacy plan which we are currently developing and into our thinking about CFNZ's strategic direction.

This work could not have occurred without the generous support of our CF families and a significant grant from SKIP, Oranga Tamariki. Our thanks to Connect+Co for their skill and expertise in co-design.

JANE BOLLARD, CHIEF EXECUTIVE

SNIPPETS

BNZ Crusaders Pitch in for CF

Charity Cricket 2017 was a high point of summer for CFNZ and one of the most successful tournaments thanks to the sunshine, the supporters, and the exciting match.

The BNZ Crusaders were up against the All Stars Invitational team, led by Andrew Mehrtens, which was able to break the Super Rugby team's five-year winning streak in the 35th annual 20/20 Charity Cricket match.

A lot of people put in a huge amount of effort for CFNZ to raise a total of over $30K from the event and we would like to thank the Crusaders, the Invitational team, Dan Vann for his Cricket Academy which raised $800, The Rock for all the publicity, schools that ran mufti days to support the event, Poster Boy Harry, Jen Duncan for organising, artists with wonderful work on display, and all the amazing volunteers who gave up their time to support CF.

MC Chris Bond (Bondy) from More FM gave a moving talk about his personal experience with CF. "I have a cousin who has CF and have experienced first-hand how important CFNZ are and the amazing work they do," he said.

The power of Mercury

Mercury Energy named CFNZ as one of 10 charities to receive a donation of $10,000 thanks to Katy Scoullar who made a heartfelt plea about cystic fibrosis on behalf of her sister Anna Scoullar-Jones who has a son with CF. Thank you Mercury!

Bags for Good at The Warehouse

Be sure to ask for a token and vote for CF if you buy a carry bag at any of these Warehouse stores – Auckland Atrium, St Lukes, Sylvia Park, Te Awamutu, Palmerston North, Levin and Feilding.

CFNZ GETS STREETWISE

FUND RAISING NEWS

On top of raising thousands during Awareness Week, Streetwise Coffee came up with another cool idea for raising funds – auctioning kids' bikes with helmets signed by famous New Zealanders including one from All Black Dane Coles.

A total of four bikes and helmets – others which sported autographs from Olympic Gold medallist Dame Valerie Adams, Silver Fern Maria Tutaia, and regular coffee cart customer, motocross champ Levi Sherwood, fetched over $2000 on Trade Me – with hundreds of people bidding on them.

Streetwise Coffee Franchise first pledged its support for cystic fibrosis after Kayleigh, teenage daughter of the Porirua franchisees Helen and Kenny Rae, passed away from cystic fibrosis in 2009. Another staff member in Pukekohe has a son living with CF.

"We decided to support cystic fibrosis. There's all these other charities out there that get so much more support so we wanted CFNZ to be the major charity that we support," says Luke Mullinger, Operations Coordinator of Streetwise.

Luke says Streetwise first began supporting CFNZ Wellington selling chocolate fish in 2013. This year they have extended their support nationwide with coffee carts around NZ selling chocolate fish and also having donation boxes on the counter during Awareness Week which attracted lots of donations, bringing the total amount raised (including the bike and helmet auctions) to over $7000.

"It's amazing the conversations that have started at the carts and the awareness of cystic fibrosis. This year people are realising what it is and want to donate. It's been a win-win for CFNZ and for us as well," says Luke. "Your posters this year with the little boy and what he has to do for the condition have helped a lot."

The idea of auctioning the bikes and helmets came from a team building exercise at the Streetwise annual conference where franchisees built the bikes. The conference also featured the CFNZ Cystic Sisters video about their lives with CF, which helped the coffee franchisees learn more about the disorder and want to support CF even more.

"Watching the video from the two people with cystic fibrosis really got to them – there were some teary eyes after that," says Luke.

Streetwise approached the agents of Dane Coles, Dame Valerie Adams and Maria Tutaia for their autographs and were able to ask Levi directly because of him being a regular Streetwise customer.

"We didn't have any trouble getting their support," Luke says.

It was Maria's signed helmet and bike that fetched the highest bid of $560.

Streetwise itself started with one coffee cart in Otaki in 2003, franchising in 2006 to currently having 23 outlets throughout NZ.

"The original cart was an old modified Ministry of Works smoko van. Our new carts have evolved since then to having insulated walls and being much bigger because we're getting so busy."

The secret to their success is due to the quality of their coffee and clever marketing, he says.

"We have the Real Trade coffee from Havana Coffee Works – a growing brand in Wellington," Luke explains. "It's coffee on the go – you don't have to get the kids out of the car which mums love. It's quick, you can text your order – it's the whole package."

50 YEARS OF CYSTIC FIBROSIS NEW ZEALAND

Important milestones in the history of Cystic Fibrosis New Zealand and advances in the care of people with CF.

1968

The outlook for children with cystic fibrosis (CF) was bleak – the majority would pass away before the age of 10. There was little support for parents in New Zealand which is what led to the establishment of a support group of 27 people in Wellington and then the formation of Cystic Fibrosis New Zealand, which became a registered charity on 16 December 1968.

1969

Sir James Wattie – founder of the Kiwi food giant Wattie's became CFNZ's first patron. This was the year the organisation's first newsletter was published to keep parents informed. The CF News, still being published today, helps to celebrate the amazing achievements of people living with cystic fibrosis.

1976

Auckland University researcher and current CFNZ patron Professor Bob Elliott made a major worldwide breakthrough in the diagnosis of cystic fibrosis. His research established the blood screening test for newborns to test babies for cystic fibrosis. This method of diagnosis continues to be used throughout the world today.

1981

Newborn blood screening for cystic fibrosis is introduced in New Zealand. There was controversy over this because of research which failed to show any benefit in early detection of CF. This is in direct contrast to research which shows early intervention should start right from birth.

1982

By now Cystic Fibrosis New Zealand had branches in Auckland, Bay of Plenty, Waikato, Hawke's Bay, Wellington, Canterbury and Wellington which greatly improved local support for families – adding to this support was the first Fieldworker employed by CFNZ to provide direct help for families affected by cystic fibrosis. Today CFNZ has 12 branches and employs three Fieldworkers around NZ.

1983

The nutritional supplement, pancreatic enzymes, are approved for use in New Zealand. These enzymes are used to replace the digestive enzymes lacking in CF patients which can lead to poor weight gain and malnutrition. At this stage many children were surviving well into their teenage years but for reasons not understood, girls had a shorter lifespan than boys.

1988

Advances in antibiotic therapy and treatment have led to significant improvements in life expectancy – many people are living into adulthood. In NZ, an adult group of CF patients is established and granted branch membership status. Today, CFNZ has an adult with CF representative on the Board and also an Adult Advisory Group to provide guidance on important issues facing the community.

1989

The faulty cystic fibrosis gene is identified in the human DNA structure which is considered to be one of the most significant breakthroughs in human genetics – this promised to be a major advance towards a cure. Almost 30 years later the world continues to wait for a cure but many researchers around the world are working on a method to correct this gene with gene therapy or using the gene-editing technology, CRISPR.

1992

The Canterbury Rugby team formalise a charity cricket match which had been running for the previous nine years. The annual Charity Cricket match now features the BNZ Crusaders batting against an All Stars invitational team. It starred late rugby legend Jonah Lomu in 1996. Today the event raises over $30,000 for charity.

1993

The biggest advance in cystic fibrosis treatment in 30 years is approved for use in NZ. The drug pulmozyme had a major impact on quality of life but only received public funding in 1997. CFNZ needed to lobby hard to obtain wider access to this medicine.

This was the same year that the first lung transplant took place at Green Lane Hospital. Nowadays eight to ten lung transplants take place each year. Lung transplants can lead to a much longer lifespan but are still not a cure for cystic fibrosis.

1997

The Mark Ashford Scholarship is announced for people with CF who achieve excellence in tertiary education. This scholarship was founded by Fletcher Challenge who employed Mark Ashford – a Kiwi with CF who tragically passed away in 1996. He left an important legacy with his Healthy Houses Book, about how to keep homes safe and warm.

2003

For the first time in its 20 year history, Ironman Taupo announced it would partner with a charity for this event – Cystic Fibrosis New Zealand's Breath4CF. This event raised big totals such as $120,000 in 2004, $145,000 in 2005 and $125,000 in 2006. Tracey Richardson, who was instrumental in establishing CF as the official charity, was awarded the New Zealand Order of Merit for services to cystic fibrosis.

2006

Denis Currie who was awarded the Queen's Service Order (QSO) becomes President of CFNZ as Jane Drumm, who has two children with CF and who has extensive experience in the community sector, is elected as CFNZ Board Chairperson. They both continue to lead CFNZ.

The first reports of cross infection among CF patients are published in the Lancet medical journal. Further research has led to strict hospital protocols for treating patients, and people with CF being discouraged from socialising together.

2011

Cystic Fibrosis New Zealand joins other countries to launch a data registry for cystic fibrosis patients. This registry is known as Port CF in New Zealand and includes information about genetics and treatment patterns.

2012

The FDA approves the first precision medicine that treats the underlying cause of cystic fibrosis. Kalydeco treats a rare genetic type of cystic fibrosis. Another drug called Orkambi that treats a more common genetic type was approved in the US and Europe in 2015.

2018

The future is bright for people with cystic fibrosis and their families. Thanks to new treatments, organ donation and more personalised care, life expectancy has risen to over 40 years in a number of countries around the world. Scientists are constantly working on new therapies and drugs that will continue to prolong life and hopefully lead to a cure.

ALANA'S AMAZING ADVENTURES

In the past six months Alana Taylor has packed more milestones into her life than most people her age have done in a lifetime.

She's been on the trip of a lifetime, celebrated a 30th birthday she'll never forget – and even bought her first home.

Life just seems to keep getting better and better for Alana who underwent a lung transplant in 2015. After the operation she vowed to celebrate her 30th in Las Vegas - giving her friends and family plenty of warning to save for the trip so they could celebrate with her.

There were 28 well wishers at her birthday party in Vegas which was also the start of her seven week-long trip around the US including San Diego, San Francisco's most crooked street in the world, New York, the Grand Canyon, ziplining at Whistler in Canada, and the amazing Wanderlust Festival at Lake Tahoe.

Alana saved for years for the big trip by working hard and was helped by friends and family giving her birthday presents for various activities she wanted to do while on tour. She spent the first leg of her trip on tour with her friends until they parted in New York when Alana took a small group tour for the rest of the trip around Canada.

Attending the music and yoga Wanderlust Festival at Lake Tahoe was a high point of the trip. "I fully recommend it. I've been doing yoga for about seven years and it was amazing."

It was also amazing for one of her friends to compare Alana's health now to a trip to Europe they did when they were 23. She said, "You struggled so much when we went to Europe and now look at you'."

Alana was first up for the thrilling zipline experience at Whistler which involves flying through a forest suspended from a high wire (similar to a flying fox). And because her lung function is now excellent she was able to trek up a high glacier in Canada without too much difficulty.

"The guide offered to help me but I was determined to do it on my own. It's not a goal unless I walk up it myself. But I'm still gaining strength in my body after being unwell for so many years," she explains.

Every time Alana achieved one of her goals on her trip she took a moment to thank her lung donor.

"It's only because of my lungs that I was able to do this. I'd think, 'I'm literally here because of this beautiful woman who gave up her lungs for me – it was a kind of a spiritual thing saying: Good on you – you've done the glacier and you've done ziplining'."

Arriving home in Christchurch could have left Alana feeling flat but instead she achieved another important milestone – becoming a home owner.

"The plan was to buy a house next year but I went to the bank and they said with your savings and Kiwisaver you can afford a house.

"This house just literally came up – it's an on old cottage in Belfast with 3 bedrooms. It was a reasonable price and now I own it!" Alana says.

Alana moved into her house in October and has got a flatmate to help with the mortgage.

It's been an exciting six months for Alana and for now there are no other big life changes planned but who knows what the future could bring for this inspiring CFer.

CF ON SHORTY

TV's favourite long-running soap did its bit and more for cystic fibrosis when it featured a main character affected by CF, and also hosted Juliet Hubbard from Christchurch as part of her Race4Life Trust adventure.

It was amazing to watch the episode on Shortland Street which featured Sass's love interest, Dr Hawks Logan talking about his own experience as a carrier of the CF gene. He revealed he had a son with another person who carried the CF gene, but their son tragically passed away.

Unfortunately it wasn't an extended storyline but it was featured the week before Cystic Fibrosis Awareness Week which was great for awareness of the disorder and hopefully encouraged more people to donate. It was also CFNZ's most successful Facebook Post, with 283 "Likes".

SELFIES WITH CHRIS AND SASS

But that wasn't Shortland Street's only brush with CF last year. Shortland Street also welcomed Juliet Hubbard of Christchurch on set when she won a Race4Life wish to meet the actors on the show. She had selfies taken with Michael Galvin who plays Chris and Lucy Lovegrove who plays Sass, and also other cast members.

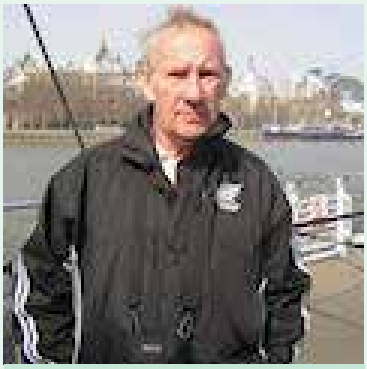
Juliet and her husband Kris were also treated to the Auckland Sky Tower experience and meals at Orbit and Depot in the city. Then they got the fright of their lives with a super scare pass to Spookers.

Other people with CF who have had Race4Life wishes include Sam Churton who was granted a super cool getaway in Queenstown after an operation last year and David Donohue and his wife Tessa got an all-expenses paid trip to Christchurch.

"I would walk 500 miles"

Terry Wilson has walked 800 Kilometres (or 500 miles) as part of a spiritual journey to honour the memory of his granddaughter.

Walking the Camino from France across Spain to the Cathedral at Santiago de Compastella, where legend has it that the remains of Saint James are buried, is also known as a pilgrim's journey. It was one of the most important Christian pilgrimages during the Middle Ages. Even today about 250,000 people walk it each year.



TERRY WILSON

The 35 day trek was something that Terry (68) courageously decided he would do a few days before his granddaughter Maddy passed away from CF in 2016. And he also decided he would walk for a cure – raising funds for Cystic Fibrosis New Zealand and Cure Kids – so far raising over $5000, and with his story featuring twice in the Manawatu Standard.

Terry says the spirit of his granddaughter travelled with him on the journey and since returning home to Palmerston North, Terry has drawn comparisons with his journey and the journey that parents are on from the moment their child is diagnosed with cystic fibrosis.

"Just when a parent gets the news that their child has CF and find they are in for the trip to wherever it takes you, it's the same from the minute you step out on the first day of the Camino – you know you're in for the trip no matter where it takes you.

"Many of my days hurt like hell, just like many of your days. When I rolled my ankle with 500kms to go I was told to take a few days off. Well the timeframe I was working to just did not allow for that. Sounds familiar? Yes, when did you get a break?" Terry asks the CF community.

Terry's journey is not over yet. He is still raising funds for CFNZ and Cure Kids through his Givealittle Page **https://givealittle.co.nz/fundraiser/cysticfibrosis**) and may lace up his hiking boots for another trek for CF – this time a bit closer to home - in New Zealand.

$40K SCHOLARSHIP FOR CF HIGH ACHIEVER

CONGRATULATIONS RACHAEL!

17-year-old Rachael has been honoured with an incredible $40,000 scholarship from the University of Otago for academic excellence at secondary school.

The scholarship is based on her Level 2 NCEA results and a reference written by her school - Logan Park School in Dunedin. Applicants must get a minimum of 80 excellence credits to be considered for the scholarship. Twenty-nine of these scholarships were awarded around NZ and six of these went to students at Logan Park.

Rachael, who also won a CF Achievers' Award in 2017, was happy to answer some questions from CF News about the scholarship.

Q. Why do you believe your entry was successful?

Honestly, I think it was successful due to the sheer amount of excellence credits I received at level 2. I didn't actually get to read the reference that was written about me, so I don't know how much impact this had on my result. I received 106 excellence credits at level 2, which was enough to get the scholarship. For a comparison, my friend who got 95 credits didn't get this scholarship.

Q. How will the scholarship be provided?

The scholarship is over three years of university. It consists of a tuition fee waiver and a fee waiver for residential college (which I won't be attending). For the second and third years ten thousand dollars is payed towards fees and then as a monthly payment or lump sum.

Q. What are you planning to study?

I am planning to study a bachelor of arts majoring in music and a bachelor of science majoring in pharmacology. I decided to do a double degree so I will have a range of options when I leave university. I know what I like but not really what job I want, so no need to limit myself. The science degree also means that I am more likely to get a job when I leave university.

Q. How has CF affected your education in the past?

The biggest impact that CF has had on my education was just the time it takes to maintain my health. Doing physio each night means I don't have this time to study. I also find that I am constantly fatigued, meaning I sleep a lot in the afternoon, and often in class…

Q. Do you think having CF had any impact on winning the scholarship?

I like to think that I got this scholarship just based on my academic ability. There was nowhere in the application where I mentioned that I had CF but I did apply for a disability scholarship so the judges might have seen this and it could have given me an advantage.

Q. Is there anything that you have found to improve your symptoms and help manage the condition?

I do a pulmozyme and saline nebuliser each night as well as PEP and I have done this for as long as I can remember. I think maintaining one's health is really important so I make sure that I am on top of my health by always taking my medicines and doing physio. I also play the clarinet and sing which helps with my breathing techniques so I find that I can get more air in.

Q. Any tips for other high school students with CF?

Always talk to your teachers at the start of the year and make sure they understand the impacts of your condition. Doing this, as well as keeping them updated with how you're feeling will allow a lot more leeway for things like deadlines and will allow your teacher to work with your needs. Some advice I would have for everyone, CF or otherwise, would be making sure you have something to look forward to, short term and long term, that is not related to school or CF. This was really helpful for me because it gave me something to focus on that I enjoyed and didn't feel was tainted or ruined by school or CF. I also enjoy ice skating lessons every week and decided to dye my hair beautiful rainbow mermaid colours once the exams were over!

CALLING ALL CF ACHIEVERS

The CF Achievers' Awards are now open for applications from anyone with CF who has achieved excellence in leadership, the arts, sport or education. The application form is available on the CFNZ website. The Awards are open to all New Zealanders with CF who achieve their goals along with managing the high treatment burden with CF. Applications close **1 April**.

Georgia overcomes obstacles to **win top CFNZ study award**

Georgia Drumm has become the first person to receive the Mark Ashford Scholarship for achieving excellence in an education degree.

An award ceremony for Georgia's scholarship was held in June where Georgia became the 21st recipient of the award for her impressive results during her early childhood education degree at Auckland University.

Her academic record also led to Georgia (21) winning the Aiken Travel Award 2016 – allowing her to visit an early childhood centre in Wales to gain work experience overseas.

Now working at Peacocks Early Learning Centre in St Luke's, Auckland, which is based on the Reggio Emilia educational philosophy, Georgia wants to use her $3000 scholarship to visit Italy and gain further knowledge of early childcare centres overseas.

The Reggio Emilia philosophy focuses on 100 ways of learning through using all human senses such as touch, moving, hearing and observing. The philosophy was developed after World War II by psychologist Loris Malaguzzi and parents in the villages around Reggio Emilia, Italy, and derives its name from the city.

The Mark Ashford Scholarship was founded by the late Mark Ashford's employers, Fletcher Challenge, to honour his contribution to healthy homes with his Healthy House insulation book. Previous to passing away from CF, Mark had hidden his condition from his employers. The award is now sponsored by Tasman Insulation, manufacturers of Pink Batts.

At this year's award presentation at Ferndale House in Mount Albert attended by Mark's widow Kathryn, his daughter Megan and his mother Enid, it was revealed that Mark's mother had previously taught Georgia (21) and both her brothers, Luke (23) and Angus (18, who also has CF), in their early years at St Dominic's Primary School in Blockhouse Bay, Auckland. And in another coincidence, Georgia and Mark's daughter Megan both graduated with education degrees at the same time earlier this year.

Not only did Georgia have to overcome some of the challenges of CF to achieve at school, she also had the obstacle of dyslexia to cope with. She often struggles with spelling and also mishearing words and has to put in double the effort to achieve the same marks as others.

"I would mishear things, my spelling, my writing and everything was quite bad. Throughout school and especially in primary I had tutors before school and after school. It did help but it's never going to be fixed. Without the support of family and my tutors I probably wouldn't have gone to Uni and succeeded as well as I have," Georgia says.

The dyslexia is something that's got easier over the years but, like CF, is something Georgia always has to live with.

"Something like a learning story that takes someone else 30 minutes to an hour can take me two hours – maybe more, just to do one," Georgia explains.

Cystic fibrosis will also remain a challenge for Georgia – as it was during her school and university studies.

GEORGIA WITH HER FAMILY FROM LEFT, STEVE, ANGUS, JANE, GEORGIA AND LUKE

"Trying to keep up with my education and the demands of CF – and trying to find a balance. Part of that balance is being active – in a sport or doing something to keep me healthy. I found in school that was a pretty challenging thing to find a balance for. I've only just really found that balance now – I've joined the YMCA and go for an hour after work. Then I come home, do my physio, have dinner and go to bed.

"My health has been good. Working in an early childhood centre, I'm always going to get infections because of the germs. I use hand sanitisers and keep them in my car," she adds.

Her recommendation for other young people with CF is to find a career they are passionate about.

"I've always wanted to be a teacher and I think that's driven me to have success. My mum was a teacher and I used to play teachers with my brother Angus (I'd be the teacher – he'd be the pupil). I like helping people and for me I think teaching is the way I can do it."

HOOK, LINE AND YOUTUBE!

Dunedin Fisherman Dylan Booth is hooked on fishing in more ways than one – he's gone fishing with celebrity fishermen, has his own YouTube fishing channel, reeled in some impressive sponsors, and landed his perfect job at Hunting and Fishing.

Dylan (19) got his first taste of fishing as a preschooler but doesn't believe the sea air has affected his condition much – he always has a bit of a cough and struggles to walk long distances, but he likes to keep as fit as possible.

Dylan was nice enough to share the secrets of his fishing success with CF News:

Where are some of your favourite fishing spots?

Twizel Hydro Canals or Lake Aviemore in Waitaki.

What made you decide to start a YouTube channel?

When I was around 12 I started getting into fishing quite seriously. Most school holidays I'd venture down to the river and land some serious fish. That made me think, "Why not make a fishing channel?" Well, I'm glad I did because now I'm sponsored by two very large companies that are known worldwide! They are called Daiwa and Black Magic Tackle.

Has your YouTube channel Fishing Forever Films been growing in popularity?

Yes, it's starting to take off quite large I'd say. This summer I have some of the biggest fishing YouTubers known worldwide come over to New Zealand to fish with me and film for their channels. I've combined the channels and all together they have just short of two million followers. How nuts is that!

How did the sponsorship come about?

Just usually posting on social media like Facebook, Instagram and YouTube I'd say. That question is hard to answer.

How's your job at Hunting and Fishing in Dunedin been going?

I work four days a week at Hunting & Fishing Dunedin. I love it and the staff are just great. I feel right at home.

What's the biggest fish you've ever caught?

Freshwater would be a 37lb 14oz brown trout which was just shy of the world record.

Saltwater would be a 150lb Seven Gilled Shark off the local wharf where everyone swims...

You received a Make-a-Wish for the fishing show on TV – how did that go?

My Make-a-Wish was unreal! It was awesome to fish with Matt Watson. I learnt a lot of "tips and tricks" that's for sure. We never filmed for TV but it was an epic time.

Any tips for others with CF?

To all the cystic fibrosis people out there I'd just like to say if you have a goal just do step one. Focus on that before all the others, take every step one at a time and slow and steady is the key. Stick to your goal and one day you'll make it happen! Thank you very much for taking time out to read this.

Yaam's Farm

Out of the many Make-a-Wish Foundation applications one of the most original must be from Yaam of Wairoa.

Instead of asking for a trip to Disneyland or to meet a famous star, 15-year-old Yaam wished for his own farm – in particular a farm full of olive trees, but also some chickens and goats.

Make-a-Wish kindly granted his wish and gave him 106 olive trees to plant on the farm he lives on with his mother and father but the family didn't have to plant them all – the Wairoa community pitched in to help when they heard about Yaam's story on the local radio station.

Yaam offered to share his olives with people in Wairoa, which is one of the reasons Yaam was named a One News Good Sort. When the TV crew visited his farm Yaam was able to explain some of the reasons for his more unusual wish. He loves olive trees because they grow for over 300 years and to him they represent longevity.

Yaam is pictured here, and CFNZ Fieldworker Jude Kelly paid a visit to Yaam's family last year.

2017 Australasian CF Conference Report

I attended the Australasian Conference last year. I have been to many NZ CF Conferences but this was my first one abroad (so grown up). I went for the hot-takes, the best practice and the cutting edge sessions. I brought them home, but I'm also going to share them. Here are my hottest hot-takes:

Recent studies from Canada suggest that long term use of nebulised pulmozyme, which has been available for 20 years in some countries, resulted in measurably less inflammation in the lungs. The presenter noted that even if there was no immediate improvement when treatment started, such as movement of more mucous when doing airway clearance physiotherapy, a reduction of inflammation occurred anyway. ***Hot take*** – don't skip pulmozyme treatments.

Personalised medicine is the buzz around the traps. Targeted, specific gene mutation correctors and potentiates, with no mutation left behind. It used to feel special, rare, kind of interesting that my CF mutation was unidentified, unknown, but now it is starting to feel lonely with this unwanted mutation. It feels exactly like I'm being left behind. I leave the presentation with a renewed vigour to hunt down this rampant defect. ***Hot take*** – seek progress from hospital about our missing mutation.

Clinical trials worldwide need more people to progress new medications and improve patient outcomes. The benefits of taking part in a trial can be long term. If a trial medication improves your health there may be access to "open label" medicine post-trial, even while a medication has not been approved or funded for use in your country. ***Hot take*** – say yes to clinical trials.

Studies have shown that depression can cause inflammation in the body, independent of disease. Inflammation is not a friend of CF – inflamed lung tissue can lead to infection. Anxiety, depression, and your personal health matters. Specialists recommend that all people with CF, their care-givers, partners, mothers, fathers, and siblings, should undertake an annual quality-of-life questionnaire, just a five minute check-in, and follow up with a professional as required. ***Hot take*** – be honest when filling out quality-of-life surveys at annual review.

Burn-out among mothers of teens with CF is proportionally higher than the rest of the population. About 35% of CF mothers experience symptoms. But adherence to medication and physiotherapy by teenagers in this group was better than their peers. This is really difficult, I am in this risk group – I have a teen who is compliant because I continue to make things happen, but I know I need to help him become more independent and not hover and stress about every little gram of fat or every little intake of breath through the nebuliser. ***Hot take*** – encourage independence in my CF teen.

Research shows that if a parent is anxious or depressed there is more chance that a child will be too. What to do? Monitor and check-in with professionals. ***Hot take*** – be honest when filling out quality-of-life surveys at annual review, also continue to seek ways to improve resilience in my family. Note: We live in Auckland. Starship has always undertaken quality-of-life surveys at annual review. This may not be the case in other care centres, this may be something that you need to do independent of usual CF care.

Has anyone else noticed this – CF is awful so we get sad, we work hard fighting this disease which eventually makes us burn out, getting sad and burning out then rubs off on our children so that they end up not only sick but anxious and depressed too. ***Hot take*** - be honest when filling out quality-of-life surveys at annual review, continue to seek ways to improve resilience in my family, perhaps don't go to conferences?

On a more positive note, the dietician was a breath of fresh air. Happy, funny and nice. The nugget (no pun intended) from this presentation was that smaller goals are helpful. Actually, there was a second nugget, dieticians are more interested now in a healthy CF diet, calorific but not rubbish food. ***Hot take*** – every little bit helps. Healthy calories are gaining traction among professionals; we all need to keep educating ourselves about diet and nutrition.

Get active, stay active. We know exercise is important. The three key words were strength, stability and posture. For my teen I have made an effort to try new things this year as our old activities were getting a little staid. ***Hot take*** – keep encouraging physical activity and sports, whatever, wherever, whenever.

The latest physiotherapy device is called an "Aerobika", a small oscillating compressor that nebulises medication while you do your PEP treatment. The device can be used in the vertical or horizontal position, which means while you are lying down clearing the left or right extremities of your lungs. The compressor only releases medication as you inhale, not on the exhale, so less medication is wasted. ***Hot take*** – ask our physiotherapist if this would suit my son. Also check about horizontal PEP – I have not heard of this.

I'm going to end at the beginning, the first session of the conference. Gathered, sitting up straight, not yet slinking in our seats from being overwhelmed or feeling guilty. The audience waits, shuffling into the room, expectant, eager. Dropping jackets, scarves, handbags and conference packs. We stare at each other. Who has CF? Who is a parent? Who is a doctor? Who is from NZ, Australia?

The first speaker is introduced, a young woman, a lung transplant recipient. But that is not what she talks about. Her mission is to make us think, not about how to improve outcomes in CF patients, not about what drugs we can pin our hopes on, not about which country is doing things differently and better, and not about which CF mutation we have. She doesn't want us to focus on "how" to fix CF – she wants us to focus on the person with CF that we are all here for, why is that person worth fighting for? What makes them special, why do they matter?

The room was enormously vast, the air swirling around the vaulted ceiling, the presenter clinging to the lectern on the wide stage, but at that moment, I thought of my son and my world became very small, my desire to love and protect him was pin-point sharp. It was perfectly clear why I was there.

AMANDA BLAKEY

DISCLAIMER: I am a parent of a person with CF, not a CF doctor (in case you can't tell). This information is what was important to me. My descriptions and comments come from what I heard and how I processed it. I did not take note of Professors' names, study titles or clinical trial information, and I did not attend all sessions as I have yet to master how to be in two places at once.

FOOTNOTE: I had a great time too.

New NZ Data Holds Key to Improved Therapies

New information from New Zealand's Port CF patient data registry is expected to be one of the best weapons in the fight for new medicines and improved treatment.

The 2016 Port CFNZ report will be released in the next few months and Auckland University Paediatrics Associate Professor Cass Byrnes says this quality data, which shows where New Zealand sits in terms of health of persons with CF nationwide, is what is needed to convince health authorities of the importance of new precision medicines that are available in other countries.

"This registry is one of the most important resources we have in cystic fibrosis care. If there are gaps in care, or if we can show how many are eligible for any new treatment that is being considered, then we can argue for new medication or better resources. No decisions are made without data.

"Every time Pharmac is considering a new medication for this population, they will do nothing without facts and numbers to assess cost and effect.

"Furthermore, if we can compare the health and availability of medicines with other countries national registries - and we see gaps - we can fight for what they have that we don't."

Parents need to give their consent to have children's health information included on the registry with these young people re-consenting when they reach 16 years of age. The number of New Zealanders who provide this anonymous data is around 98 percent of all persons with CF in our country.

Dr Byrnes has noted some areas where we could improve access to treatment according to the 2016 figures. "You can see poor uptake of some of the treatment options – for example while many people are appropriately on inhaled hypertonic saline to improve secretion clearance, there are fewer on pulmozyme. And given the numbers that have chronic Pseudomonas infection, we should see more on inhaled antibiotic therapy. Is that individual choice? Or are some people not being offered some medications, as they don't realise they are eligible for them?" Dr Byrnes asks.

"There are other areas in the 2016 report that we should drill into to find out what's happening. An example of that is physiotherapy. A lot of people are ticking 'no physiotherapy' as their primary airway clearance technique – and is that right? A lot of people are ticking sport as their primary airway clearance technique and it would be really nice to drill into that to find out how much exercise they are doing.

"Sport might be appropriate for some who are really, really well and do a lot of activity but we've only got one tick and it could mean anything.

"We have gaps in our treatment compared to places overseas but in addition to that we can probably do better with what we have," she says.

Dr Byrnes believes one of the main reasons for these differences is the number of New Zealanders receiving care in different places throughout the country.

"The total number of New Zealand people with cystic fibrosis would make up one single clinic in the States for example. The same in Australia – they have an outreach programme that is just more rigorous than ours. We do have an outreach programme and we have very good paediatricians and specialists around the country managing children and adults with cystic fibrosis but we don't have enough outreach from our centres sometimes to support them."

Dr Byrnes says the genetic make-up of New Zealanders could also be changing with the country's evolving population which may inform changes to the newborn screening programme and which CF genes are tested for with the heel prick test.

"In future we are going to look at whether we are doing the correct screening," says Dr Byrnes who is also on the Ministry of Health's Newborn Screening Programme's Technical Committee.

"It's important for people to know what their genes are for the new drugs coming through, so anyone who doesn't know their genes should retest, especially if their original test was done some time ago," she advises.

For the 2016 report, there was extra emphasis on getting all the data on all persons with CF around New Zealand into the database with CFNZ engaging Jan Tate to support clinics from around the country to input data. "This 2016 report will be the most accurate snapshot of where all people with cystic fibrosis are. It will be the best one so far to compare to how we're doing with international registries – Australia, Canada, the States, UK, France and Germany." Port CF data suggests that the median age of people with CF in New Zealand is a few years behind other countries which is another reason why it's important to have this information available. She says life expectancy data isn't available because the New Zealand Port CF registry hasn't been going long enough to make this calculation.

There are some positive NZ figures compared to other countries that have emerged from the data.

"Our chronic Pseudomonas rate is actually lower and our lung function at the time of transition to adults is better. And nutrition (weight) seems very good compared to other places as we have a high and early use of nutritional supplements. So we do some things very well."

NEXT GENERATION DRUG APPROVED

The Food and Drug Administration in the US has approved the use of a new therapy called Symdeko to treat the underlying cause of cystic fibrosis.

The therapy uses a new drug, tezacaftor/ivacaftor in combination with ivacaftor (Kalydeco) to treat patients aged 12 and over who have two copies of the F508del gene or at least one CF gene that is responsive to the treatment.

Two research trials of the medicine tezacaftor in combination with ivacaftor show the new combination therapy to have comparable effects to Orkambi (which combines ivacaftor with a drug called lumacaftor) for people with two copies of the F508del gene, and significant improvements for those with one F508del and one "residual function" gene.

NEXT STEP FOR BREATHE EASY

NZ-based research company Breathe Easy is hoping to test a higher dose of its cystic fibrosis medicine Citramel after latest results show that it's safe and well tolerated but was not effective at the dose tested.

Breathe Easy is disappointed that so far the potential therapy has not had the impact that everyone hoped for and is planning more studies with a higher dose. It will also analyse sputum samples to determine the reasons why some dissolved well in Citramel and others did not.

Gene Editing in NZ

Gene editing to cure genetic disorders like cystic fibrosis looks set to become a reality in future but more research is needed to determine the safety of the new technology because so far no children have been born with edited DNA.

Gene editing and CRISPR, which is a technique to repair or replace a faulty gene in a human's genome sequence is being carried out in research using human embryos overseas. There is a moratorium on research using embryos in New Zealand, although some scientists are urging for this to be lifted.

The Royal Society of New Zealand has recently held a series of lectures around the country featuring Otago University graduate Dr Josephine Johnston who is now director of research at The Hastings Center in New York. CF News attended the lecture in Auckland called *Making babies: You, me and gene editing?* This lecture, hosted by radio personality Kim Hill, included three other panellists: Maori Health Research Associate Professor Maui Hudson, Medical Genetics Researcher Professor Andrew Shelling and Dr Mary Birdsall of Fertility Associates.

Dr Birdsall said that preimplantation genetic diagnosis (PGD) is currently used when couples are concerned about having a child with CF because they both carry the CF gene. When a couple has IVF, any embryos that are produced are tested for CF. Embryos that do not test positive can be implanted for the pregnancy.

She believes CRISPR could be helpful in cases where all the embryos in an IVF cycle are affected by cystic fibrosis. "Every parent wants to have healthy children. Some people don't have enough embryos and the fix-it technology might be useful for some," Dr Birdsall said.

However, Dr Johnston pointed out that IVF involves a lot of stress on the body and the couple, so the use of CRISPR could be a much simpler method for conceiving children where there is a high chance of a genetic condition.

"Going through fertility treatment to get pregnant is pretty onerous – it places a burden on someone who may feel a responsibility to procreate that way," Dr Johnston said.

Professor Shelling expects gene editing to become an accepted treatment for a small number of conditions.

"I think it's inevitable that some of these nasty diseases will be treated by germ line gene therapy or CRISPR/CAS," Professor Shelling says.

"Most clinicians would want to be targeting those extremely nasty diseases where there's either no quality of life or early death like Huntington's disease, myotonic dystrophy, or inherited breast or ovarian cancer. They are so unpleasant that society would probably want us to do something about them," Professor Shelling said.

Kim Hill asked if the gene would be edited in the embryo or in the infant and Professor Shelling said the earlier the better. "You do it in the embryo as early as possible – that way the disease never begins to develop."

Dr Birdsall said there is currently no information about the safety of this technology in an offspring.

"None of these embryos are being replaced (transferred to a womb)," she said.

SNIPPET

CLIMB EVEREST IN WAIHI

The Waihi Beach community are getting behind a Climb Everest Challenge to raise funds for CFNZ Bay of Plenty and pre-schooler Frankie-Lee. Frankie-Lee (inset) and her family settled in Waihi after travelling around New Zealand looking for the perfect place to live and Waihi won their hearts. The family have been warmly welcomed into the Waihi Beach community with Frankie-Lee's preschool, Beach Kids launching the **Everest Challenge fundraising through Givealittle** after learning all about cystic fibrosis when Frankie-Lee enrolled. The challenge involves walking the Waihi Beach Trig 48 times between 20 February and 21 April.

Givealittle: **https://givealittle.co.nz/fundraiser/breath-for-cf**

"We are certainly not going to do it in New Zealand – we have a moratorium on embryo research. We are in a relatively nice space and can watch the rest of the world," she added.

Maori Health Research Associate Professor Maui Hudson expressed caution about new genetic editing technologies saying it is important to protect Maori's whakapapa (line of ancestry). "The thing people are most interested in maintaining is protecting whakapapa. With gene editing you are still affecting the whakapapa but in some ways you might be strengthening it – it could be seen in a positive way."

Maui predicts that all New Zealanders will have their genome sequenced as part of their medical record within the next 15 to 20 years.

Dr Johnston says it's becoming more acceptable to edit genes to eradicate serious disorders but not so much for enhancing a human being's capability such as for playing sport.

Professor Shelling reported that 10 years ago the Advisory Committee on Assisted Reproductive Technology presented a document to the then health minister that recommended allowing research on embryos in New Zealand. These are embryos that have been stored for 10 years at fertility clinics and need to be discarded.

"Successive ministers have sat on that information and done nothing about it," he said.

Sex selection is also against the law in New Zealand. Dr Birdsall said she gets asked about sex selection from couples at her clinic about 4-5 times a day.

Couples who both carry the CF gene are eligible for publically funded IVF and preimplantation genetic diagnosis (PGD) but Dr Birdsall said there are regulations surrounding it.

"I think here are only about 20 funded PGD cycles across New Zealand each year.

"People can choose to go down this pathway. We are reducing the disease burden on society. If you look at what a child born with cystic fibrosis costs New Zealand – I think it's around $500,000 in health care costs across the average lifetime and that's not including the possibility of heart and lung transplant, I think there is an economic argument for society to offer preimplantation genetic diagnosis for certain conditions."

According to the regulations, Dr Birdsall said couples who qualify for a publically funded IVF/PGD cycle, and who have embryos which all have cystic fibrosis, are unable to have any of them transferred for a pregnancy.

Where couples pay for IVF and PGD themselves ($15,000 - $20,000) they are allowed to transfer an embryo affected by CF but she says in her experience couples in this situation do not go ahead with a pregnancy.

Dr Johnston asked if we were moving to a world where we have voluntary eugenics and added there needed to be support for people who decide not to have PGD.

"I think we are getting more tolerant of disability and at the same time we are gaining powers to stop it.

MORE INFO

CF News asked Professor Stephen Robertson, Cure Kids Professor of Paediatric Genetics at Dunedin School of Medicine for feedback about this gene editing article and he provided the following comments about gene editing - specifically related to cystic fibrosis.

He said gene editing may not be as helpful in the case of recessive genetic conditions such as CF (where both parents carry the faulty CF gene) for the following reasons:

A. Families would still have to use IVF because gene editing needs to be carried out at a very early stage in the embryo to maximise the number of cells that have their CF reverted back to the normal sequence.

B. Many families already use pre-gestational genetic diagnosis (PGD) to have children when both parents carry the CF gene. Gene editing would only be useful when all embryos are affected with CF – which probably doesn't happen very often.

C. **Somatic therapy**, which is a form of gene therapy that aims to edit cells in certain affected organs (e.g. the airway). For instance a virus carrying the gene editing machinery could be administered as an inhaled aerosol. This type of therapy is more likely to be a way by which gene editing is used in the future for conditions like cystic fibrosis, according to Professor Robertson because this would only change the genes in the individual with CF and not be passed on to the next generation. For some organs that are affected by CF prenatally (e.g. the pancreas) this is a less likely proposition.

Note: The Royal Society is now seeking feedback for its discussion paper *The use of Gene Editing in Healthcare* which is available at this link: **https://royalsociety.org.nz/what-we-do/our-expert-advice/all-expert-advice-papers/gene-editing-in-healthcare/**

Farewells

May our loved ones rest in peace and love

Hollie Hale
21/09/2017 – Christchurch

Bridget Hendry
29/10/2017 – Oamaru

Kimberley Bryant
4/1/2018 – Wellington

Paediatric CF Clinical Network Update

New Zealand paediatric CF health professionals are close to completing a set of guidelines to help strengthen children's CF care in New Zealand.

The Paediatric Society Child and Youth Clinical Network for Cystic Fibrosis is one of several different Networks funded by the Ministry of Health. The networks were set up in 2013 to create paediatric guidelines for several different medical conditions including diabetes and dermatology, as well as for the medical care of children with CF.

So far the CF Network has approved 12 sets of guidelines aimed at helping CF centres around New Zealand. The Network is also close to approving guidelines for several others including Gastro-Intestinal and Liver problems in CF, ENT problems, and Palliative Care in CF. They have also ratified the use of the recently published Australasian CF Physiotherapy guidelines, and intend to do the same for the Australasian Nutritional and Dietetic CF guidelines when they are finalised early next year. Once completed, each set of guidelines is published online, via the Starship Hospital Clinical Guidelines web page. The guidelines are accessible to anyone, not just hospital staff. In the last six months the overall number of views for all chapters of the CF guidelines was just under 2000, making them the second most frequently viewed set of guidelines after the Eczema Network.

The Paediatric CF Network is made up of members from all hospitals in NZ that provide Paediatric CF care. It is led by a Clinical Reference Group (CRG) that comprises 10 members: Julian Vyas (Chair, Medical), Jan Tate and Viv Isles (CF Nurse Specialists), Fiona Leighton and Julie Graves (Dietitians) Rebecca Scoones (Physiotherapist) Peter McIlroy and Cass Byrnes (Medical) Rebecca Landreth (Pharmacy), Sue Lovelock (CFNZ fieldworker), Jane Bollard (CFNZ CE). In addition, Mollie Wilson (Paediatric Society CEO) and Karyn Sanson (Project Coordinator for Cystic Fibrosis National Clinical Network) provide operational support. The Network has sought input from many experts in related fields when drafting specific chapters for the guidelines.

Julian Vyas commented: "The aim of the guidelines is to support local CF teams, but also families and whanau to access current best practice thinking to keep a child with CF as well as possible." He says the guidelines should not be seen as dictating the *"official Starship way"* to tackle a clinical problem in CF.

"It is very important to remember that they are guidelines, and not strict rules. Everyone's CF is unique to them, so you cannot be dogmatic and say there is only ever one way of managing a clinical issue in CF. My hope would be that if a particular question arises, the guidelines can help inform a local Paediatric CF team about treatment options, and to inform discussion between the patient, their family or whanau, and their team. But guidelines can only help so far, and Paediatric CF teams across New Zealand know that, whenever necessary, they can always discuss a specific concern with the specialists at Starship."

In 2017 the Ministry of Health decided to continue funding Paediatric Society health networks around New Zealand. And so far the CF Network has been working very well. "It's a big group of people, because CF has so many facets," Julian says. "It is a huge task, given the various ways CF affects people. However the Network Clinical Reference Group has worked hard to bring these various guidelines to completion. The next step we hope to move on to is looking at teaching aids (videos, tutorials, etc.) for patients, parents/carers & staff to help with day-to-day practical things – like doing physiotherapy."

At this stage no similar group exists for developing guidelines for CF Adult care, although this has been highlighted to representatives from the Ministry of Health, by Julian.

He is hopeful the paediatric guidelines will help support provision of care being equitable, no matter how far a patient lives away from a large Paediatric centre, such as Christchurch or Starship. All the guidelines (for CF and other conditions) are accessible to anyone via the Paediatric Society Network web pages – the link is provided below. In this way it is hoped that parents, and teenage patients with CF can empower themselves to understand more about what treatment options are indicated or available. "The people who really need the knowledge here are the patients, or their parents. I hope all the Paediatric Society guidelines will give patients and parents better knowledge to take part in discussions about health choices," Julian says. One of his favourite quotes is from Aneurin Bevan, the Minister who founded the NHS in Britain in 1948. He said "The purpose of getting power is to give it to those who deserve it."

"We welcome comments and suggestions on the guidelines from patients, parents and other carers; as well as ideas for other topics to cover," Dr Vyas adds.

To view the guidelines visit the website: **starship.org.nz/for-health-professionals/new-zealand-child-and-youth-clinical-networks/child-and-youth-cystic-fibrosis-clinical-network/**

Generous Donors Launch New Breathing Therapy

The Auckland Branch has recently received a generous donation from the Hawkesby/Hart family that has enabled us to purchase two new Metaneb machines for adult services at Auckland Hospital.

BRUNO, WHO HAS CF, WITH HIS GOOD MATE DYLAN HAWKESBY WHOSE FAMILY MADE THE GENEROUS DONATION

The Metaneb is designed to maximize treatment by combining three therapies in one—volume expansion, secretion clearance, and nebuliser therapy. The device begins by offering continuous positive expiratory pressure (CPEP) therapy, which helps expand the lungs, and then seamlessly moves on to the secretion clearance part of the treatment. At the same time, the nebuliser is running throughout the entire therapy session.

The therapy cycle takes approximately 10mins and maximises efficiency by combining the three therapies into a single therapy. This means that there is only one machine used for all three therapies.

The new machines have only just been received by Auckland Hospital and should be up and running shortly. Starship Hospital also had a Metaneb machine donated last year and has been seeing really great results.

During the week there were bake sales, mufti days, fundraising soup making, sausage sizzles, chocolate fish and the street collection. Westlake Kindy on the North Shore, asked their children to bring in spare vegetables. The children and teachers then prepared delicious soups with the donated food and sold it back to the families with all proceeds to CF Auckland.

It was great to see Rachel's son Luke in the NZ Herald right on collection day. The article about trialling the new breathing therapy game *BreatheHero* was great for CF awareness and helped with donations as well.

LUKE WAS A BEATHEHERO STAR

Thanks to all the schools and business who supported us through fundraising, selling chocolate fish or helping the street collecting. It always seems like a big task to bring it all together but we are always inspired by new faces that volunteer to help and to those that loyally support our organisation year after year.

THIS GROUP OF MASSEY HIGH STUDENTS SERENADED SHOPPERS WHILE THEY COLLECTED FOR CF.

Awareness Week Action

WESTLAKE KINDY MADE AND SOLD SOUP TO FUNDRAISE FOR CF AUCKLAND FOR AWARENESS WEEK

Auckland Branch had another very successful Awareness Week in August mainly thanks to the energy and hard work of our committee member, Rachel Eliott and her family. Every year we try to get as many schools, businesses and people on board to help with our fundraising. This year we tried a coin trail board that was sent out to schools and preschools.

Auckland Marathon Ran like Clockwork

It was a perfect running day for the Auckland Marathon this year. We were fortunate to have a group of 20 Willis Towers Watson employees who trained/ran and fundraised nearly $7,000 for CF Auckland.

All the runners were greeted at the finish line with bacon and egg butties and a sports massage. Thank you to Niall, from the CF Auckland Committee who recruited the group of runners and facilitated the huge fundraising effort. Also to Mercury Energy for their support of a grant, which enabled us to purchase logo'd t shirts and covered the cost of the gazebo site at the end.

WILLIS WATSON TOWERS RUNNERS

We were fortunate to once again have Larissa and Helen from Ultra Physio (www.ultraphysio.co.nz/) who not only massaged all our runners but also helped with more fundraising by massaging other competitors for a fee. Their energy and enthusiasm was amazing as they massaged hour after hour. Thanks to Jill and John Thorrat on the BBQ and their assistants Leo, Eva and Sean. We are also very grateful to Pak'nSave Glen Innes which generously donated the food for the BBQ.

The Warehouse Have Done it Again!

We feel very fortunate to be chosen again by The Warehouse to be part of the "Bags for Good" Campaign in three stores across Auckland – they are Sylvia Park, St Lukes and the Atrium. If customers choose to purchase a shopping bag they are given a token to put towards one of three charities. We are very grateful to The Warehouse for their ongoing support. Please support CF Auckland if you are shopping in one of these stores.

– Kath Sanderson, Auckland Branch Coordinator

FUND RAISING NEWS

WAIKATO BRANCH NEWS

It's a Wrap in Waikato

Waikato Branch has had a very successful 2017 and gearing up for another year of raising awareness, and of course funds for our families.

The Warehouse has been fantastic for Waikato Branch with the Bags for Good Neighbourhood initiative being chosen for a third round with the Te Awamutu store. Our second round with Matamata, Te Awamutu and Morrinsville have just finished up, and we look forward to hearing the results of those! It's wonderful that Te Awamutu Warehouse has kept CF Waikato in their Bags for Good for the next six months.

We were also asked to be a part of The Warehouse Wrap it for Goodness initiative with one week of wrapping in Te Awamutu and four weeks in Matamata! Te Awamutu made $460.00 in the five days they were there and Matamata made $4200 in the four weeks. We would like to extend our thanks to all our volunteers who helped make this possible.

The money the branch has raised lately has been incredible, although it is even more awesome to see the Cystic Fibrosis name and logo out there and recognised in the Waikato with so many people becoming more aware of who we are and what we do.

ROBYN AND DAMIEN AT THE TE AWAMUTU STORE "WRAPPING IT FOR GOOD". WE ARE SURE DAMIEN WILL NOW BE WRAPPER EXTRAORDINAIRE AT THEIR HOUSE!

Fundraisers Bear Fruit

It is fantastic to see young people out there and raising awareness for CF. Renee, Olivia and Emma were out on their wheels selling Lemons in Morrinsville in the name of CF. They sold them for 10 cents each, although elderly folk got them for free! They made $6 for CF.

These girls and a couple of extras also went out and sold chocolate fish and made $50! They were lucky a kind man bought eight fish and only took three for himself – that kept them going for sure!

– Marieke Latimer, Waikato Branch Secretary

LEMONS FOR SALE!

Flashback to the Swinging 60s

CF Hawke's Bay Casino Night took a trip back in time to the psychedelic 60s for their popular annual fundraising event.

The gambling on blackjack tables and roulette wheels was all in aid of local people with CF - this year raising a record total of $17,000 thanks to some generous auction bids including Cape Kidnappers luxury accommodation that sold for $2000.

The 60s-theme proved popular with everyone dressing up for the occasion. There were hippies, go-go girls, minis, maxis, headbands and high boots. Elvis was also spotted leaving the building.

REAL GROOVY CASINO NIGHT

Christmas Dinner

Hawke's Bay Branch hosted an enjoyable Christmas lunch for members just prior to Christmas.

The lunch, held at the scenic Off the Track restaurant in Hawke's Bay was a small, informal gathering to acknowledge the year's achievements.

This year the Branch decided to invite one person with CF along to each Xmas gathering so committee members can get to know the people that inspire fundraising activities. This year it was Secretary Claire Fisher's daughter Amber who came along with her friend Summer – they are pictured below in front of the Xmas tree.

PAKOWHAI SCHOOL HELD A DRESS IN BLUE DAY FOR CF.

SUMMER AND AMBER AT XMAS LUNCH

FUND RAISING NEWS

Big Billboard at Wellington Airport

The Wellington Xmas Tree Festival was a huge success thanks not only to the beautiful trees which lit up Wellington Airport but also the level of awareness of cystic fibrosis gained from the giant CF billboard donated to the event.

The trees were decorated by local businesses, schools and sports teams, with the Axe furniture tree awarded the best tree because of its inspiring black and white photos of people meeting loved ones at Wellington Airport. It was lovely to hear the sounds of native birds singing from the Kiwibank tree, and also hearing Chamber Music New Zealand and the Symphony Orchestra playing during the Festival.

The XTF has been running for nine years and this time the Airport kindly donated the use of a large indoor billboard to help raise awareness about cystic fibrosis. It highlighted the message that for some families every Christmas is a gift. And with around 1 million people travelling through the Airport over Christmas and January, CFNZ couldn't have asked for a better gift for awareness.

CEO's from Wellington Airport, Air New Zealand, NZ Rugby and CFNZ all spoke at the gala opening in December. The Branch is delighted that Wellington Airport has announced the amazing Xmas Tree Festival has been confirmed for this year as well.

AXE WINNING TREE (LEFT)

CF BILLBOARD

New Children's Hospital

CFNZ Wellington has been impressed by the level of its involvement in the planned new Children's Hospital building. CFNZ and the Branch have been involved from the outset of this project – providing feedback about cross infection and clinical rooms, and has been one of just a few patient groups that have been consulted. The Wellington Branch Committee and CFNZ Chief Executive Jane Bollard were taken on a guided tour of the concept clinical rooms in November. The new hospital is expected to open its doors at the end of 2019.

CF Nurse Locked In

After many years of lobbying, Wellington Branch was thrilled to hear the appointment of Wellington Hospital's CF Nurse Specialist Tricia Martin was made permanent.

Tricia's appointment has made the world of difference to CF families.

Melissa's a Hero

CALEB, MELISSA, PAUL, TORI, AND MAYOR LIANNE DALZIEL

Congratulations to Canterbury Branch Chair Melissa Skene who has been awarded a Kiwibank Local Hero Award, which rewards everyday people doing extraordinary things in their communities.

Melissa stepped in to help run the Canterbury Branch 19 years ago and her award acknowledges her commitment to CF families in the region. She is incredibly supportive to new parents who value talking to someone who has been through what they are facing.

She received her medal at a special Kiwibank Award ceremony in December.

Caleb Conquers Queenstown

CALEB KEEPING FIT

And congratulations to Melissa's son Caleb Skene who achieved his personal goal to run in the Queenstown Half Marathon in November.

Caleb's friends paid for Caleb's entry for his 20th birthday last year, and although he left training for the marathon a bit late, he still managed to meet his target to finish in under 2 hours, with his time of 1.49 mins.

His biggest hurdles were the blisters he got when he decided to see if he could actually run 21km straight with a big training session around Hagley Park the week before. This left him with severe blisters, but wearing jandals and taking good care of his feet meant that he was still able to run on the day.

It's a very impressive result for someone with CF – the only thing Caleb would change next time would be starting to train a good few months beforehand.

Going to the gym four times a week and playing indoor football also helps keep Caleb fit and well.

Ain't No Party Like a Pole-Tober-Fest Party

The Pole-Tober-Fest show in Christchurch produced a record result and a record turnout. The show is the concept of Liv Allchurch, owner of Liberate Exercise and Dance, and Juliet Hubbard, who enrolled in the pole exercise classes to improve her lung function. Juliet reports on the incredible performance which included dancing, acrobatics and burlesque:

"Olivia Allchurch (owner of Liberate Exercise and Dance) once again did an amazing job putting together her annual Canterbury Cystic Fibrosis Liberate show fundraiser.

This was the fourth year and each year it's growing – and by growing I mean with everything – the amount we raised, the awareness that's created, the venue is bigger, we have more performances, more tickets sold, and many more companies donating prizes for raffles, balloon pops and gifts for VIP goodie bags.

Liv is amazing at what she's done and how this event has grown so much. The first year $400 was raised and this year a grand $4,500 was raised! We had many different performances such as pole routines, singers, burlesque performances, belly dancing, even twerking! (Google it if you're not sure on that one ha ha). There was even a horse and a penguin on stage this year! All who performed did so out of their own choice. No one was paid for this, all their time was donated to putting together an amazing show.

I did a wee opening speech to get the night started that I had written the week before with a tiny bit of help from Mr Google and then my own words and I was sorted. It's so good getting as much awareness out there when there're so many people there listening to you. The turnout was fantastic – an estimated 200 people. It was such a fun night with so many laughs and I'm already looking forward to the 2018 CF Pole-Tober-Fest!

– Juliet Hubbard

POLE-TOBER-FEST PARTY / PHOTOS COURTESY OF SUZY T PHOTOGRAPHY

Stevie Wealleans wrote some moving words for the event about what it's like having a friend with CF – this was read out on stage by Liv from Liberate.

"My name is Stevie. I have been friends with Juliet for around 12 years. When they asked if I wanted to say a few words about having a friend with CF I knew I wouldn't be able to stand up in front of everyone and talk about it because it makes me upset that my amazing friend has this lung disorder.

Juliet is a positive and strong person and doesn't let it get her down. We do all the normal things friends do BUT: Juliet needs plenty of time to get ready to go out as it takes her a lot longer than people without CF. She can often get out of breath and needs to take breaks in-between things like showering and doing her hair. She also needs time to take her nebulisers which help her breathe.

On shopping trips I have to remind myself to walk slower so Juliet can keep up or she gets puffed out and has to tell me to slow down. There have also been times when she has had to ask me to carry her handbag as its too heavy with things like her medications - I think her husband Kris has had to do this too – I'm not sure if pink suits him though.

Dinner dates are quite often our mutual favourite of Thai food but many have been spent at Christchurch Hospital and eating while watching Shortland Street.

If any of Juliet's friends get sick with a normal cough or cold we have to make sure to keep away until we are better because if she catches it it could be enough to put her in hospital. This would usually result in antibiotics for a couple of weeks.

Juliet is lucky to have her amazing husband Kris who can help her with some of the household tasks like vacuuming and grocery shopping which are easy for someone with good lungs but can really take it out of her.

Juliet's lung function is currently around 27% and a lung transplant could be an option for her in the future. I have been told having CF is like trying to breathe through a straw, so if any of you have a straw handy, give it a go.

Thanks everyone for coming tonight to support this great cause. These words were mainly written about Juliet but she is just one of the many New Zealanders suffering from cystic fibrosis."

REBEKAH'S SECOND CHANCE FOR LIVING

Hi my name is Rebekah Henderson. I'm 28 and I live in Auckland and I'm incredibly blessed. Like many of you reading this, I suffer from cystic fibrosis, but as of nine months ago, I am now also the recipient of a double lung transplant which is why I am so unbelievably blessed to be here today - healthy and full of life to write this article for you. But I do just want to say that the transplant road is one that you don't want to get onto in a hurry.

Growing up with CF pretty much sucked. All the medications plus physio, fortnightly stays in hospitals, zero energy because your daily coughing fits take it all within the first 45 minutes of your day, yup CF sucks. But it is something that can be managed and we are so blessed that it is a disease that can be kicked on its ass – we just have to put in the hard work to do it. Please trust me when I say that you want to put the transplant option off for as long as you can.

As a child my parents had my routine down pat right up till I was 11-12 years old when my doctor sat me and my parents down and said to them that they needed to start handing the control of my health over to me. Oh boy that didn't go down too well – endless heated conversations with my parents, rebelling and not doing what I needed to – all because I wanted to fit in with my peers, which resulted in me having a slowly dropping lung function, dropping weight and endless counts of admissions. I think one year I got 10 hospital admissions in one year. That's a ton of hours bored out of my mind.

In the end I learnt the only person I was hurting was me and my body was screaming at me to change, but I never listened because I was so stubborn and the want to fit in was so much stronger than what my body so desperately needed. Also the crazy thing is that the people that I wanted to impress so much were never there when I was in hospital and they aren't in my life now, but I am, which is more reason why we need to look after ourselves, CF or not. People come and go in our lives but you have you forever and you need to be your own best friend and look after the lungs and body you have now.

RECIPE ROUND-UP

SULTAN'S DELIGHT SUMMER DIP

INGREDIENTS:

2 garlic cloves
5–6 dates
1 pot 250g cream cheese
1–2 tbsp plain yoghurt
¼ tsp curry powder
(or to taste)
Pinch of cumin

METHOD:

Whiz or chop dates finely, crush garlic and mix with cream cheese, yoghurt and curry powder. Add salt, pepper and a little cumin to taste. Then chill for 2 hours. Serve with crackers or fresh Turkish bread.

Unfortunately, when I finally got that through to myself it was too late, my lungs were shutting down to the point that at 25 I was in respiratory failure and because I had been so slack with my medications and physio it was looking like I might not get a transplant at all because I was told that not everyone gets the chance to get one and that they only pick people who will look after them. So I was given a hospice care booklet. I can easily say it was the worst admission I had ever had but it was the kick up the ass I needed. It just came too late for me to be able to undo any damage so all I could do was make sure I kept my weight up and kept my lung function from dropping any lower.

Thank goodness I put in that hard work because a month later at clinic I was told I was going to be referred to the transplant team to start the assessment process. After weeks of tests I had to wait to see if I was a good candidate for a transplant – and thankfully I was!

The reason why I say it's important to do everything you can do now is that there were no more options for me health wise. They had no more medication that they could give me, they had no more tricks up their sleeves, it was either transplant or nothing at all. And thankfully I was able to get a transplant and I've lived every day since then as fully as I could but not just for me, but for my amazing donor without whom I wouldn't be here today. I am still blown away that I have so much energy that I can laugh, cry, talk, walk, and run without coughing. I can go back to work, hang with my friends, go hiking and go hard at the gym all because of one person's selfless act of kindness and love.

My life now is completely different too, which I'm still getting used to but I am so blessed. But please don't repeat my mistake in life. Look after you and remember that at the end of the day the only person you need to worry about is you, and I know you have it in you because you are amazing and you can fight this disease. If you ever need a friend or just want to ask anything about this article, please feel free to email me on **bekahh72@gmail.com**.

Thank you for reading
Rebekah

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Creating Better Tomorrows

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