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CF Life

Issue 9

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Cystic Fibrosis Trust

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T: 020 3795 2177
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Cystic Fibrosis Trust Helpline
T: 0300 373 1000
E: helpline@cysticfibrosis.org.uk

Our confidential Helpline offers general advice, support and information on any aspect of cystic fibrosis, including help with financial support.

All magazine correspondence should be sent to:

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Welcome to CF Life

The past few months have been incredibly tough for many, but we have been inspired by the ways the CF community has come together in the face of coronavirus (COVID-19).

As we write, we know that Kaftrio will become available to thousands of people with CF across the UK in the coming months. We look forward to featuring stories of people whose lives will be improved by this medication in future editions. We'll also keep you updated on research to help the people who won't be eligible for Kaftrio.

In this issue we look at the progress being made by our UK CF Innovation Hub at the University of Cambridge, which is working on projects to improve lung health for people with cystic fibrosis. We also hear about a new partnership to tackle antimicrobial resistance in treating CF lung infections.

You'll hear from Polly, who was the first baby in the world diagnosed through the heel-prick test, 40 years ago, and Yasmin, who shares some great tips on supporting a sibling who has cystic fibrosis.

The CF Life team

If you'd like to give us feedback on this issue, or have ideas for what you'd like to see in the magazine, email us at magazine@cysticfibrosis.org.uk

What you might have missed

Campaigning

Shortly after life-saving drug Kaftrio (called 'Trikafta' in the US) was given a positive recommendation for licence by the European Medicines Agency, deals were announced to give access to the drug in all four UK nations. The news brings hope to thousands across the UK, and further reinforces our focus to support those who will not benefit from current treatments.

Marc Cotterill, one of the many who can benefit from Kaftrio

Sammie Read

Research

As the gravity of the COVID-19 pandemic emerged and face-to-face CF clinics were put on hold, developers working on the remote health monitoring study 'Project Breathe' made their app publicly available, to allow anyone with CF to monitor their health from home. The Project Breathe study will help to establish the most effective ways of delivering remote monitoring, as well as use the data collected to improve future care.

Find out more at **cysticfibrosis.org.uk/adaptingcare**


Rebecca Cosgriff

Care

Using data to understand health has never been more important than during the COVID-19 pandemic. The UK CF Registry moved quickly in its response to the crisis, creating a helpline for clinical teams to report data, collaborating with other global Registries to gather data and reporting weekly on statistics of COVID-19 in people with cystic fibrosis. This innovative work demonstrates the power of collaboration and openness to benefit people with cystic fibrosis.

Visit **cysticfibrosis.org.uk/registryresponse** to read more.

Support

The pandemic brought profound new challenges to the CF community, which we saw through significant extra demand for our information and support services. We adapted to keep up with the rapidly evolving situation, increasing staffing on our Helpline, providing up-to-date information and guidance in collaboration with CF healthcare professionals and the UK CF Medical Association and creating a dedicated emergency grant service to support those most in need.

Read the article at **cysticfibrosis.org.uk/adaptingsupport** to find out more.

Fundraising

With the cancellation of hundreds of events, COVID-19 has had a substantial impact on our fundraising income and how we raise money. However, the CF community has inspired us with the creative ways they've taken on virtual events like 24-hour gaming challenges. Gaming is a great way to connect with others, play the games you love and raise vital funds to help support people with cystic fibrosis.

Find out how to organise your own #CFNextLevel gaming challenge at **cysticfibrosis.org.uk/cfnextlevel**

Innovating our way to better lung health

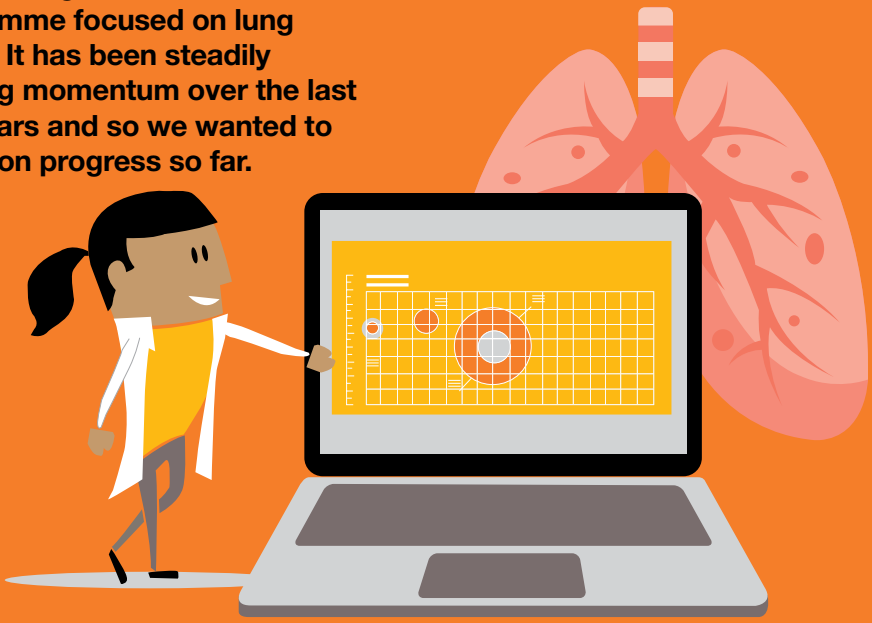
We know that improving lung health could make a big difference to everyone with CF, both on a day-to-day basis and for living a longer, healthier life. Our UK Cystic Fibrosis Innovation Hub at the University of Cambridge is an ambitious programme focused on lung health. It has been steadily building momentum over the last two years and so we wanted to report on progress so far.

Poor lung health in CF is caused by cycles of lung infections and inflammation, and the damage to lung tissue that happens as a result. Research within the Innovation Hub is focusing on three areas of improving lung health:

1) new treatments for infections and chronic inflammation;

2) better ways to diagnose and treat flare-ups of poor lung health; and

3) working towards a long-term aim of developing alternatives to lung transplant.



"The Innovation Hub allows world-leading experts to apply their science in new ways to address issues important for cystic fibrosis."

– Dr Janet Allen, our Director of Strategic Innovation

People with CF can have periods of stable health, and then experience a flare-up of lung symptoms, known as an exacerbation. These can be very disruptive to day-to-day life and can lead to several-week-long stays in hospital and even permanent lung damage.

The exciting news that many people with CF will be able to access the new CFTR modulator Kaftrio could mean big improvements in health for those who are eligible to receive it. We hope that people taking the CFTR modulators will have fewer infections and fewer exacerbations, but we know from data from the first modulator, Kalydeco, that infections are still likely to occur. The modulators will not be able to correct existing lung damage, and so methods of repairing lung damage are still needed. We must also address the needs of those for whom the modulators don't work. That's why research within the Innovation Hub is increasingly important to improve the lung health of everyone with cystic fibrosis.

The Trust and the University of Cambridge are co-funding the Innovation Hub on a partnership basis. It's a five-year programme that began in 2018. The Trust aims to raise £5 million over a five-year period to fund the Innovation Hub, which the University of Cambridge has committed to match pound for pound to £5 million.

"The Innovation Hub arose as a way to capitalise on the extraordinary and dynamic scientific expertise at the University of Cambridge. It allows world-leading experts to apply their science in new ways to address issues important for cystic fibrosis. Research that brings diverse disciplines together has proven time and again to be the most effective way of delivering impact," explains Dr Janet Allen, our Director of Strategic Innovation.

As part of the partnership, researchers working on CF will be based within the new state-of-the-art Heart and Lung Research Institute, which is currently under construction. "The Chief Investigator of the Innovation Hub, Professor Andres Floto, has created an integrated, multidisciplinary team with some of the top experts in the world. This will lead to a better understanding of CF lung health that will benefit people with cystic fibrosis," says Dr Allen.


Dr Janet Allen

Better, more effective treatments to improve lung health

Lung infections are hard to treat due to the unique environment in the lungs of people with cystic fibrosis. The increasing resistance of the bugs to existing antimicrobial drugs also presents a huge issue. The effect of infections on the lungs can be made worse by an overactive immune response leading to inflammation. There are immense challenges in finding effective antimicrobial and anti-inflammatory drugs, and the Innovation Hub team is pursuing novel approaches to address them.

Researchers within the Innovation Hub are focusing on developing new antibiotics to treat *Pseudomonas aeruginosa* and *Mycobacterium abscessus* infections, which have devastating effects in people with cystic fibrosis. They started by identifying new biochemical weak spots where new drugs might have the biggest impact.

Mehro-Nissa Javed

Now they know more about the bacteria, they've begun work to develop the drugs. They are using an innovative process known as 'fragment-based drug design'. Researchers within the Innovation Hub are the pioneers of this drug-design approach, which has led to the licensing of new cancer drugs in the past. By the end of their Innovation Hub funding, they aim to have designed potential drugs ready to be tested as antibiotics in early-stage clinical trials.

"I think the research they are doing is absolutely extraordinary," comments Mehro-Nissa, who has CF and was recently diagnosed with an M abscessus infection.

"For me, a better treatment could have meant spending less time in hospital, having fewer side effects to the IV antibiotics and more time at home with the people I love."

"For me, a better treatment could have meant spending less time in hospital, having fewer side effects to the IV antibiotics and more time at home with the people I love."

– Mehro-Nissa

Predicting a flare-up of lung symptoms

It can be hard to know when a dip in health may be leading to an exacerbation, or whether it is simply part of day-to-day variation. Due to this uncertainty, treatment for exacerbations is often delayed to avoid unnecessary exposure to drug side effects.

Artificial intelligence researchers, who are part of the Innovation Hub, are developing ways to predict the onset and pattern of exacerbations to enable treatments to start sooner. To do this, they're using the data from our digital health research studies such as SmartCare and Project Breathe.

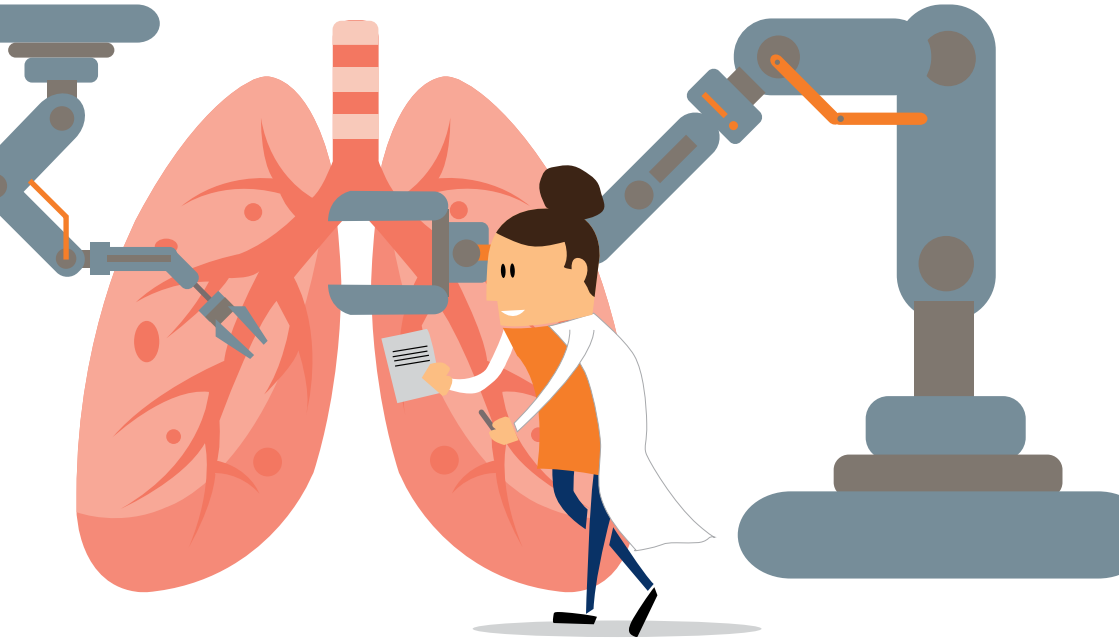
So far, the researchers have been able to find a pattern of measurements that shows someone's health was declining more than a week before the person noticed it. One of the next steps will be to see if they can use this pattern to actually predict whether someone is likely to have a flare-up of symptoms.

"In the short term, this is the part of the Innovation Hub I'm most excited about," says Michelle Shore, one of our Trustees who also has CF, after receiving a progress update on the Innovation Hub. "This is something that all people with CF could potentially benefit from, regardless of their genotype. Those who won't be offered the life-changing effects of CFTR modulators can be offered the means to improve their life experience and regain some control over their health and the impact it has on their life."

Michelle Shore

"Those who won't be offered the life changing effects of CFTR modulators can be offered the means to improve their life experience, regain some control over their health and the impact it has on their life."

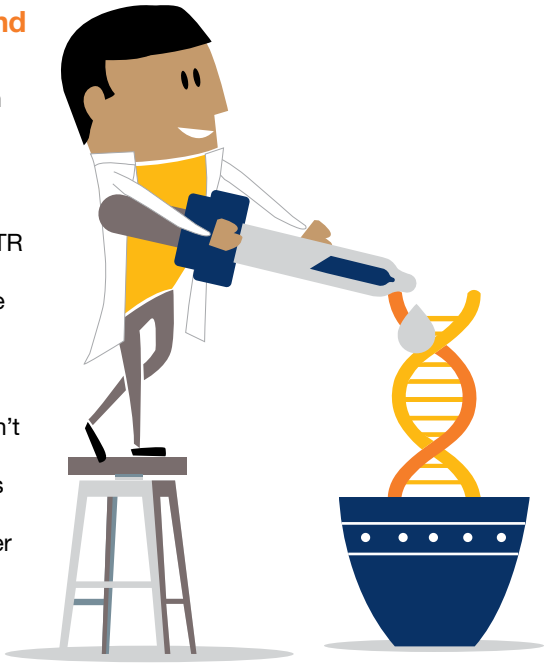
– Michelle Shore, Trustee



Paving the way for lung repair and 'personalised' treatments

An important part of improving lung health is having an in-depth knowledge of all the different types of cells that make up the lining of the lungs. Understanding these differences is important to allow as many people as possible to benefit from the CFTR modulators. It's also key in making steps towards repairing permanent lung damage caused by cystic fibrosis.

The way to gain a better understanding of this is to grow lung cells in the lab. As the current methods of growing lung cells aren't a good comparison to what's happening in the lungs of people with CF, researchers within the Innovation Hub are using stem cell and gene editing technology to pioneer new ways of growing and studying cells.



Amid the excitement of access to Orkambi, Symkevi and Kaftrio is the sobering knowledge that not everyone with CF is eligible for these new drugs, and differences in their effectiveness on an individual basis are emerging. To address this, it is likely that future CF treatments will need to be tailored to each individual with the condition. The stem-cell-derived lung cells, developed by researchers within the Innovation Hub, will become an important tool to develop personalised treatments for people with cystic fibrosis.

Current CFTR modulator treatments reduce the rate of further lung damage but they don't repair any existing damage. Ways of repairing the damaged lungs are urgently needed too. One possible approach is to introduce new lung cells, as a 'graft'. Introducing donor cells would give rise to issues of cell rejection, in the same way that transplanted organs would be rejected without immunosuppressive drugs. A method to avoid these issues would be to manipulate cells from a different tissue of the same person. As part of their studies to understand the cells that line the lungs, researchers within the Innovation Hub are investigating how a lung cell graft could work, and whether stem-cell-derived lung cells from people with CF could be given back to them as a lung cell graft.

Over the next few years, the Innovation Hub is set to make significant improvements to the lung health of people with cystic fibrosis. The advances made as part of the Innovation Hub will also have long-reaching effects to benefit other areas of medical research including antimicrobial resistance, digital health and other lung conditions.

Coming together to beat antimicrobial resistance

Many infections are becoming resistant to the latest drugs, known as antimicrobial resistance (AMR). In September last year, we launched a new partnership between the Trust and an organisation called Medicines Discovery Catapult (MDC) to work together to tackle AMR in treating CF lung infections. The partnership is called the CF Syndicate in AMR. A year on, Candice King, who has CF, spoke to Jessica Lee from MDC and Dr Paula Sommer, our Head of Research, to find out what the Syndicate has achieved.

"The Syndicate is about making the most of investments that the Trust has already made..."

– Jessica Lee

Candice: Why was it the right time to set up the CF Syndicate on antimicrobial resistance?

Paula: The CF Syndicate in AMR aims to get more antimicrobial drugs to people with CF faster, bringing together a range of different people with expertise in AMR. Lots of things came together at the right time – we had identified a need to help pharmaceutical and biotech companies develop new antimicrobial drugs for CF, MDC were looking for new charity partnerships, and the Government had prioritised tackling AMR.

Jessica: The Syndicate is about making the most of investments that the Trust has already made, accessing the innovative research that is taking place in small companies and also bringing on board people with CF, to make sure that we're focusing on their needs and priorities.

Candice: What challenges have you overcome?

Jessica: Our biggest challenge was to bring together people from different organisations – university-based researchers, people working in small companies and clinicians – and to understand from them what obstacles they were facing. These people have lots of different expertise and they wouldn't normally sit around the table together. Between us we've developed a single research agenda to overcome the common obstacles that everyone is facing, including access to samples from people with CF and a pathway from finding a drug to getting it to patients.


Jessica Lee

"We must also address the needs of those for whom the modulators don't work."

– Dr Paula Sommer

Candice: What have you achieved so far?

Jessica: We're making good progress. Researchers at universities and biotech companies are keen to get clinical samples, as the strains of bugs people with CF grow are different to the strains available in the lab. We're finding out how researchers collect and store samples now and investigating how we can bring them together to start sharing them more widely.

With the help of people from the CF community, we've started to develop 'target product profiles'. These are ideal checklists that companies will use to make sure that the drugs they're developing will meet patient and clinical need. They're still in development but they'll include, for example, that new drugs shouldn't add to someone's burden of care and that they can be taken using the same devices, such as nebulisers, that someone might be using anyway.

Candice: What's next?

Paula: In the longer term we'd like to see a whole range of new antibiotic drugs in development. We hope that people taking the CFTR modulators will have fewer exacerbations and fewer infections, but we know from data from the first modulator, Kalydeco, that infections still occur. We must also address the needs of those for whom the modulators don't work.

To donate to support the work of the CF Syndicate, visit **cysticfibrosis.org.uk/donate**

A lucky life

Polly Crosby was the first baby in the world to be diagnosed with CF through the heel-prick test, in 1980 at Ipswich Hospital. Although told she wouldn't see her teens, today Polly shares the things she has achieved in the last 40 years, thanks to incredible advancements in medicine.


Polly Crosby

'Luck' is a strange word to use about yourself, especially if you have CF, but nevertheless, I consider myself lucky. I was born in 1980: I had a loving family, the NHS, and I had the heel-prick test. If I had been born a hundred years earlier, my illness would not have had a name, let alone a treatment of any kind. My mother would've kissed me and known that I was not long for this world, for every parent knew that a salty kiss on an infant's brow meant the curse of death.

My mum, who was a nurse at the time, was aware something wasn't right. She suspected CF, and the heel-prick test confirmed her suspicions. When I was diagnosed at three-months old, my parents were told I wouldn't have long: possibly 7 to 10 years. They were given books about CF that showed just how awful the condition could be. They had a lot to take in.

I had a very easy childhood. I think knowing that I had CF from birth meant that it was just part of my life. I started having IV antibiotics when I was about 10 to calm all the chest infections that I got, and eventually the hospital stays and home IVs that followed became my normality. At some point, my mum decided my physiotherapy would be a bit easier if we looked at books together. She whacked me and read to me for years, and I think this is where my love of books really began!

In the last 10 years, it feels as if so many groundbreaking treatments have come along. Thanks to them, and with the help of a complex daily routine of physio and nebulisers (and perhaps some good genes somewhere in there too), I was able to have my son, Seb, who is 12 now, and at high school. Incredibly, I'm healthier now than I was when I had him.

"When I was diagnosed at three-months old, my parents were told I wouldn't have long: possibly 7 to 10 years."

Polly and her son Seb

Life was good, but I had one more thing I really wanted to achieve. Those years of reading books had started a lifetime love of writing. I'd tried many times to write a novel, but with the exhaustion that came with CF, I found it hard. Two years ago, I took part in the Kaftrio trial (previously known as Trikafta). In the four weeks I was on the drug, my brain awoke. There's no other way to explain it. My symptoms all but disappeared, and I found myself writing like I'd never written before. I finished my novel, 'The Illustrated Child', and I applied for the MA in Creative Writing at UEA. Incredibly, I received a scholarship from them. I then came runner-up in the Bridport Prize novel-writing competition, and suddenly the whole literary world began to open up. I got an agent, and in one of the strangest weeks of my life, she secured me a publication deal within 48 hours!

"I'd tried many times to write a novel, but with the exhaustion that came with CF, I found it hard."

2020 has been a year of tremendous ups and downs for people with CF, but I'd like to end this article with the word I began with: luck. I am lucky. I've known many people with CF over the years, and very few of them are left now. I've just had my 40th birthday, and I'm riding on the tail wind of that average life expectancy. With everything that I know is coming in the world of CF medicine, I hope to always be chasing it, never overtaking it, and I intend to be here for many, many years to come.

Polly's debut novel, The Illustrated Child, will be published by HQ on 29 October 2020.

If you have a story that you want to share, please email **magazine@cysticfibrosis.org.uk**

Identifying targets for new antibiotics

Professor Julian Parkhill runs a research group within the Department of Veterinary Medicine at the University of Cambridge. Here he explains to us how a statistical theory applied to Allied bombers in the Second World War is now being applied to the dangerous bacteria *Pseudomonas aeruginosa* and *Mycobacterium abscessus*.

Professor Julian Parkhill

I've spent my career investigating how the genetic information in bacteria changes over time. These studies can tell us about the spread of infection and also help us understand what makes them cause such aggressive infections. My recent work in the Innovation Hub involves working with colleagues to develop new, more effective antibiotics.

In the Second World War, statisticians worked out that the most vulnerable parts of bomber planes were where returning planes had no bullet holes – as the planes that were hit in those spots didn't make it back to be inspected. These were the parts of the planes that needed to be reinforced.

We're applying a similar principle to develop drugs against CF bugs. We pepper the bugs with something that damages their DNA and look for the bugs that don't grow, by inspecting the damage in the ones that do. In this way, we can find the genes that are essential for the bug to survive – these are ideal targets for designing new antibiotics.

Our studies of the genetic changes that occur in the evolution of these bacteria will help confirm that we've identified good targets for new antibiotics.

Read more about this research at **cysticfibrosis.org.uk/innovationhub**

Photo: Wellcome Sanger Institute

Bouncing fit!

Exercise is a huge part of 13-year-old Jamie's life, so when shielding meant he couldn't do his normal activities, his mum applied for a grant from the Cystic Fibrosis Trust to buy a trampoline.

I normally do loads of exercise like riding my bike, kicking the ball around with my friends and playing rugby league for Latchford Giants, which means training twice a week and having a game each weekend. This keeps me really fit.

I've been lucky that I haven't had to stay overnight in hospital before but earlier this year I had to stay in hospital for two weeks. I was getting fit again when I had to start isolating because of coronavirus.

With not being able to go out, the trampoline has been a fun and easy way to exercise. I use it three times a day for 10–20 minutes. It's a good workout and helps to keep my lungs clear.

The grant Jamie received was provided as part of our emergency COVID-19 response. You can find details of our other grants, including grants to support health and wellbeing, at **cysticfibrosis.org.uk/grants**

If you're using a trampoline to exercise, Jamie's top tips to keep it interesting are:

- Try something different rather than just bouncing.
- Have people throw balls at you to catch.
- Bounce in time to music or even do a TikTok.

Cystic Fibrosis Trust

Thank you for your support.

Together we are building a better life for all people with cystic fibrosis.

We are currently facing a substantial drop in our fundraising income as we have had to cancel and postpone a number of events and fundraising initiatives.

If you are able, please continue to support us by making a donation now.

Every pound makes a difference and ensures we can continue to invest in research so that no one with CF is left behind.

cysticfibrosis.org.uk/donate

Fighting for a Life Unlimited

Capturing your experiences of care

Patient Reported Experience Measures, known as 'PREMs', are a way of understanding the experience of care, directly from people who use health services. Tracy Turc-Milloy, our Clinical Quality Improvement Manager, talks about how and why we're collecting PREMs for people with cystic fibrosis.

Over the past couple of years, we've worked with people with CF, their families and their care teams, to develop a tailor-made PREMs survey to capture people with CF's experiences of the care they receive. The survey asks about things like satisfaction with your CF team and infection prevention and control at your centre.

With the support of 25 CF centres, we launched the survey in children's CF services back in September 2019. While we had to close the survey early due to COVID-19, we were delighted that the amazing support of people with CF, their carers, and care teams, meant that we still surpassed our target with nearly 700 responses.

We're now busy preparing the formal UK report on people's experiences of paediatric CF services. We'll also provide participating centres with individual reports, allowing them to compare their patients' responses to other CF centres in the form of a UK average.

"The PREMs project is part of our work to drive advances in quality of clinical care. We hope the reports will support centres to develop their own quality improvement projects..."

The PREMs project is part of our work to drive advances in quality of clinical care. We hope the reports will support centres to develop their own quality improvement projects that aim to make care safer, as well as more effective, efficient, person-centred, timely and equitable.

Keep an eye out for a similar PREMs survey that we'll be launching across adult services later this year. As we work to develop the adult survey, we're considering information from a recent survey of UK CF Medical Association doctors that highlights how centres have changed the way they deliver care due to COVID-19. The pandemic has accelerated the move towards access to virtual appointments and home monitoring, so we'll make sure the survey reflects this.

During this time of huge challenge and uncertainty for people with CF and their families, as well as CF teams, we wanted to share a few of the lovely comments CF families had about the care they received.

"Best team of staff I could ask for to look after my child. Always so kind, helpful and supportive."

"We're truly thankful to have such a wonderful team doing their best to keep my son healthy."

"Our CF team has always been amazing! A great team of very special people!"

The 'new normal' for events

Virtual fundraising

Earlier this year, in the midst of the coronavirus outbreak, people across the UK united to support the nation's charities through 'The 2.6 Challenge', an event that promoted the opportunity to participate virtually from anywhere, in any way. Although the challenge was developed under unusual circumstances, the concept of virtual events remains particularly relevant for the CF community. Here, we caught up with some amazing fundraisers on why virtual challenges are important to them, and to explore the future of events.

Aaron, 9, Sunderland. Raised £1,561 riding his bike around his estate for 26 minutes.

"Virtual events are great as they make Aaron feel part of something without any risks," says mum Karen. "We are always careful with cross-infection – it's an aspect of CF that Aaron finds hard but he is mature enough to accept it."

Anita, 50, Wales. Ran 2.6 miles around the garden, did 26 strength and toning exercises and 26 minutes of Turbo bike.

"I knew I would struggle when shielding was announced, as I have always led an active lifestyle. The 2.6 Challenge gave me the incentive to continue training. I loved seeing what other people were doing for their challenges – it was nice to know so many others were doing similar things!"

Dan, 26, Sussex. Cycled 26.2 miles on an exercise bike, inspired by his dad who took part in the 2017 London Marathon.

"Having virtual events like this is a great way of bringing the community together. Smaller and personal challenges are made 100 times bigger by the simple support of the community and increase the awareness of CF, which for me is the most important aspect of these events. Another bonus is it allows more CF warriors to get involved, whether they're too unwell, in hospital for treatment, or living somewhere more remote."

Want to take on your own virtual challenge? Find out more at https://teamcfsuperheroes.cysticfibrosis.org.uk

Isobel, 20, Cardiff. Completed 26 exercises, doing 26 reps, 2.6 times with friends over Zoom.

"Virtual challenges make events available and possible for everyone. It's nice to see people's different interpretations of the challenges – how they do them, who with, where and when. It's also a great support system for people with cystic fibrosis."

Alex, 34, York. Lifted a combined total of 26,000kg!

"The mental health and social benefits of taking on a virtual challenge can be such a help, especially while we've been shielding. The opportunity to do something enjoyable, while connecting with people a bit differently, has really helped."

Coughy break, shining a spotlight on the talented, creative side of the CF community.

Community photo challenge

Keen photographer Shannon, 24, decided to celebrate the CF community by running a photo challenge on Instagram.

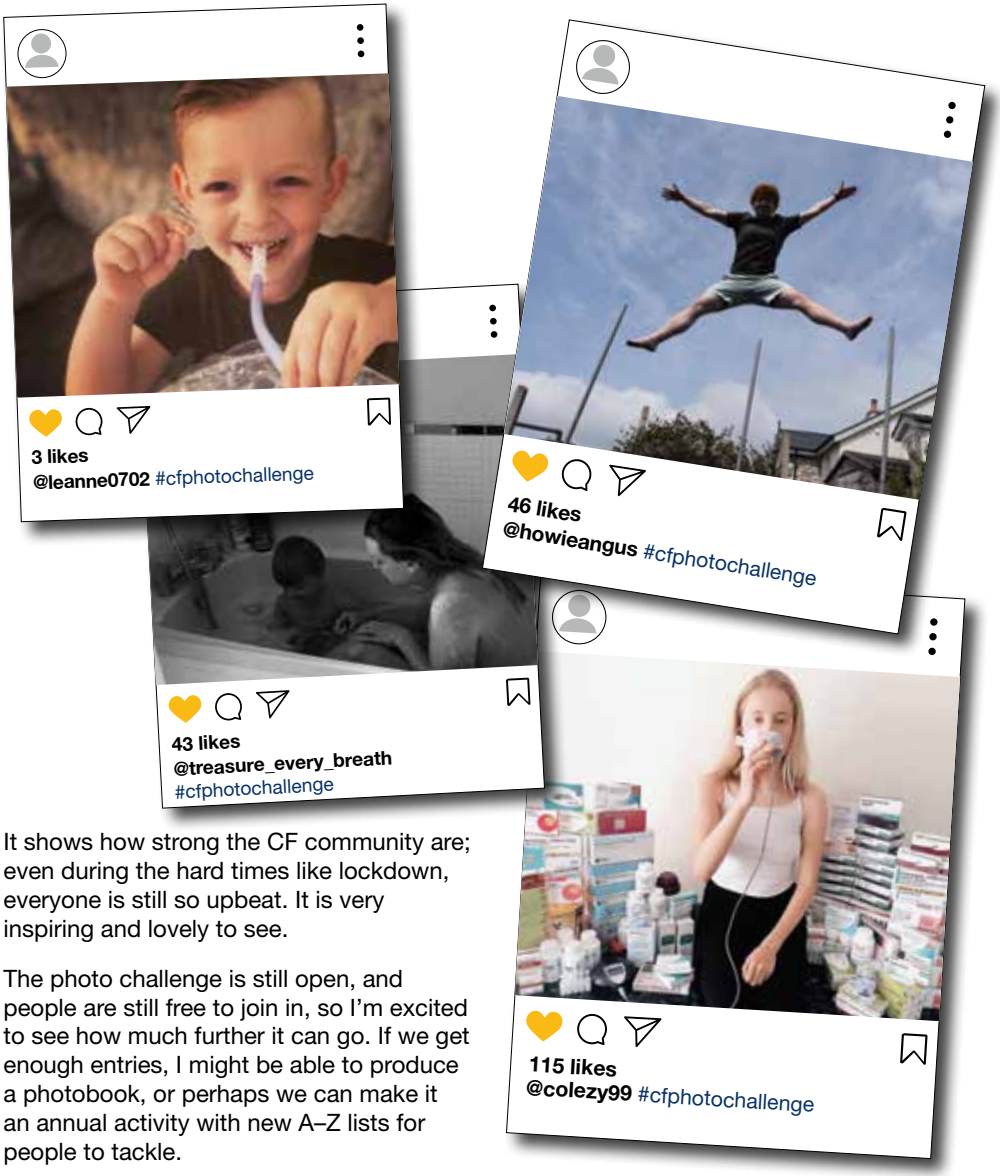
My passion in life is photography and I went to university to study it. I saved up the money and brought my first camera aged eight on a family holiday because I wanted to document the trip. Ever since then, I never put my camera down!

During lockdown, I started the #cfphotochallenge. Like other people with CF, I had to shield, meaning I wasn't allowed to leave our house. I was really missing going out and taking photos. So, I decided to come up with an activity that involves my passion and could involve other people in the CF community too.

The idea of the challenge is to take photos for an A–Z list of themes and objects that you can find around your house. I decided to put the challenge on Instagram because, by using the hashtag #cfphotochallenge, everybody's photos can be easily found in one place.

I have absolutely loved looking through all the photos that have been coming in. What I have particularly noticed is how happy and positive everybody is in their photos. Even if it's a photo of someone doing a treatment, they are having fun doing it.

Shannon @cfphotochallenge



It shows how strong the CF community are; even during the hard times like lockdown, everyone is still so upbeat. It is very inspiring and lovely to see.

The photo challenge is still open, and people are still free to join in, so I'm excited to see how much further it can go. If we get enough entries, I might be able to produce a photobook, or perhaps we can make it an annual activity with new A–Z lists for people to tackle.

During CF Week in June, Shannon chose four of her favourite entries to the challenge, pictured above. If you would like to see all the challenge photos, search #cfphotochallenge on Instagram.

Talking trials

With significant advances in CF care and new treatments, this is an exciting time for the CF community, but we know there is lots more to do. Clinical trials are a crucial part of future advances, but a lack of awareness of trials can prevent people with CF having the opportunity to take part. Our Involvement Manager, Lorna, sat down (over Zoom!) with Jane (aged 56) and Bethan (aged 11), who both have CF, Bethan's mum Lisa and Alder Hey Trials Coordinator Vicky, to chat all things clinical trials.

Jane: The first trial I remember taking part in was when I was about 18. It was for the antibiotic ciprofloxacin, which was a huge improvement in my eyes as I could have tablets instead of two weeks of IV antibiotics stuck in hospital. Back then you couldn't do home IVs.

It was a very different time to how it is now. The doctor didn't really explain anything to me; I signed the form and that was about it. Now, if you're thinking about taking part in any trial, you have time to ask questions beforehand. Your care can be even better than normal, because they have to check absolutely everything, and safety checks have been done at all the stages before you get to take it. I think it's hugely worthwhile, and it's also very interesting.

"It was a very different time to how it is now. The doctor didn't really explain anything to me…"

– Jane

The only trial I was involved in that I didn't enjoy was the Alpha-1-antitrypsin, where you had to nebulise another drug. They told me it would take about 10 minutes… well, it must have taken a good half an hour, on top of another hour of doing other drugs, plus physio, and then antibiotics!

Lorna: And that feedback is still really useful. If pharmaceutical companies are considering taking a drug like that through into your daily care regime, they've got to understand that it's not suitable. This is why we encourage them to involve people living with CF when designing treatments and trials.

Jane: Speaking of not suitable, when my baby teeth came through, they were black because of the lovely antibiotics I was given when I was a child! I was told when my adult teeth came through, they would be white… were they hell! I had to have them all replaced! So, they don't give those types of antibiotics to babies or children anymore.

Lorna: That's the kind of thing that clinical trials have contributed to, as research and clinical trials have meant we've moved on… thank goodness! Bethan looked absolutely horrified when black teeth were mentioned!

Bethan, what are your thoughts on what Jane has said about clinical trials?

Bethan: I think it's good because it could benefit your own health but could also help future generations of people with cystic fibrosis. Like you said with the black teeth, we don't have that anymore, probably because you were one of the ones that had a bad reaction.

Jane: And don't forget, this was 51 years ago, so medicine has come a considerably long way since then!

"The first trial I remember taking part in was when I was about 18. It was for the antibiotic ciprofloxacin, which was a huge improvement in my eyes…"

– Jane

Lisa and Bethan

Lorna: It's important to ask questions before starting a trial, which is why in the Trials Accelerator Platform, we have Trial Coordinators like Vicky. They become part of the familiar team, so when you want to ask about trials you feel more comfortable and confident.

Vicky, how would you approach someone for a trial?

Vicky: We'd always check if the patient is eligible first before speaking to them about a specific trial. We've got our own database at Alder Hey and the CF Registry, which also has all patients' information. It's important for my team and I to ensure patients that are eligible have the opportunity to take part in studies, and provide them with the information they need to decide if they want to.

Normally I try to speak to someone face-to-face, so would try and meet them at a clinic appointment. If not, I would give them a call to discuss it over the phone. I'd send over any information about the trial, so that they have time to read over it, make any notes and ask any questions. It's an open and honest conversation: we always let them know everything about the trial, so they have all the information to decide.

"It's important for my team and I to ensure patients that are eligible have the opportunity to take part in studies..."

– Vicky

Bethan, what sort of questions do you think you would ask if you went on a trial?

Bethan: I'd like to learn about it, so I would probably ask a lot of questions: why we are testing it, what it would improve, how long I'd have to take it for? I'd probably ask what type of tests you need to do, because I really don't like blood tests. If I had to have blood tests, I'd be less keen on doing it.

Lisa: I would probably ask if she would have to take some time off school, to find out if it would involve more trips than normal. It probably wouldn't put me off doing it but would be good to know, to be able to tell if the benefits outweigh those factors. It's also important for people to know that any expenses incurred due to taking part in a trial will be covered.

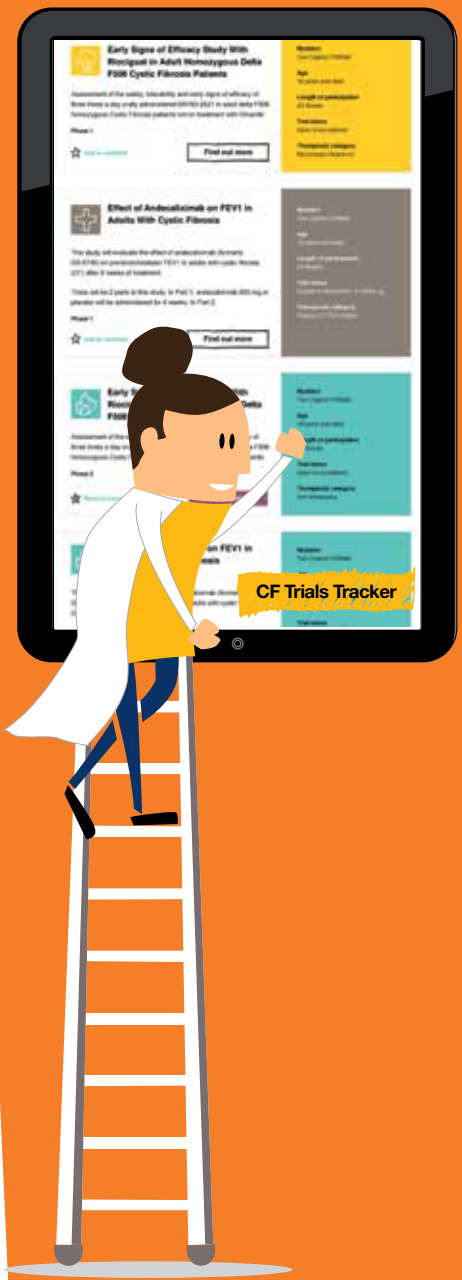
Jane: Yes – there are no worries about extra car parking fees or petrol costs!

Jane: I think what's different from back in my day is that if you have any questions or queries, or if you're not sure about it, you can stop at any time, and there's always someone you can talk to about it.

Vicky: That's right. My work phone is always near me – whether it's 8 o'clock at night or a Saturday or Sunday, I'm always here to help. The CF Trials Tracker, which is on the Trust's website, is also a great tool for patients and parents to take a look at what's on offer. It shows that research is certainly something people want to talk about.

Lisa: It's also good to know, as a parent, that you are on the radar and that your CF team will contact you if a suitable trial comes up.

Vicky: A real positive in my role is finding the CF community are always eager to support clinical trials. They're always very engaged and pleased to talk to somebody about what's out there. It's fantastic to be part of – seeing how willing everyone is to help to make things better for people with cystic fibrosis.



With help from people like Jane and Bethan, we have developed resources to help you learn more about clinical trials. We have a detailed booklet on our website, as well as a new comic for younger children and a leaflet for teenagers.

For more information, visit **cysticfibrosis.org.uk/clinicaltrialsinfo**

The unique support a sibling can bring

23-year-old Yasmin shares what she's learned about how to be a supportive sibling to her younger brother, Kareem, who has cystic fibrosis.

It took years for Kareem to be diagnosed, so we've been playing catch up with figuring out how to support him and his new needs in a short amount of time. As a sibling you're in a unique position – one a friend, a cousin, a parent or partner cannot replace. I wanted to share some of the lessons I've learned and ideas from the accelerated experience we've had, as brother and sister, now with CF thrown in the mix.

1. Never assume there is anything that your sibling can't do. This one is most important and is the point he repeated when I bothered him for help with this article. Unless they say, or seem to be really struggling with something, ask first if they need help. Otherwise, it can make them feel like they have another parent, rather than a sibling who's their teammate they can tag in for a boost.

2. Their life is not all about CF – CF is not all of them. It is only a part. Don't treat them as though they are a walking CFTR channel!

> "Their life is not CF – CF is not all of them. Don't treat them as though they are a walking CFTR channel"

Yasmin and Kareem

3. When they're unwell or in hospital, use some time that would've normally been family time to make preparations for them. Do they want you to download some Netflix shows? Could you let them borrow your laptop or toiletries bag? You know all your sibling's little quirks and their favourite pick-me-ups that the world doesn't get to see. A favourite game? Favourite crisps they shove into sandwiches? A drawing from their room? In tougher times they might find it harder to reach out and explain the bigger challenges to their friends. Us siblings see the normal routine, and how it's harder when they're unwell. So step up.

4. Ask them for help too! Ask for their shoulder when you're down, or worried about them. They'll have some great advice, being warriors in their own right, and it means when they're down they'll be more confident you can be there for them too. But remember, just like you, sometimes they might want to be left alone!

5. Since they already need to sit and attack their meds several times a day, another seven reminders won't help them feel your relationship is deeper than cystic fibrosis. Make it a habit to always ask about other important things in their life first, like a project, their friends or a hobby, before asking if they're feeling well that day.

It can be really hard. Sometimes you feel guilty. Was it complete chance, the roll of a four-sided dice of alleles, the hand of divine intervention? However you see it, it feels unfair your sibling has to suffer and not you. But I've learned there's no use in pondering over these huge unanswerable questions. So, I hide behind his door and throw a KitKat at him instead. (I advise you choose their chocolate of choice, and not to aim for the head like I 'accidentally' do!)

If you have an idea for the next Young Voices or would like to share your views, get in touch on Twitter @cftrustyouth or email cfyouth@cysticfibrosis.org.uk.

Days in the life

42-year-old Steve, who has CF, is a sign-language interpreter who lives in Herts with his wife Helen and six-year-old son Austin. With a packed hobbies list that includes running, cycling, reading and spending time with family and friends, staying healthy is paramount. Steve is taking part in the Project Breathe study, using home monitoring to manage his health, which he often fits around his daily routine.

The project is a collaboration between the Cystic Fibrosis Trust, Magic Bullet, Royal Papworth Hospital, Microsoft Research and University of Cambridge.

A blowing test.

Recording my temperature.

Checking my oxygen sats while brewing a cuppa.

4

Balancing the scales and a quick work call.

5

Entering the results into the phone app.

6

Video call with my team at Papworth.

Even if you're not taking part in the research study, you can use the Project Breathe app to help monitor your health at home. Find out more at cysticfibrosis.org.uk/adaptingcare

cysticfibrosis.org.uk

Cystic Fibrosis a fight we must win

Organise your own event – we're here to help

Our supporters have been finding creative ways to raise money despite the challenges of shielding and social distancing.

We've seen head shaves, street bingo, bake sales, quizzes, dancing and garden parties taking part across the country – all raising much needed funds.

Choose an activity that's right for you and help us reach every person with CF and their family when they need us most.

cysticfibrosis.org.uk/myevent

Fighting for a Life Unlimited