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CF Life

Issue 10

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Feature

The magic of mucus

Fly on the wall

Project Breathe – digital health

Fighting for a Life Unlimited

Cystic Fibrosis Trust

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Issue 10 – April 2021

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FEATURES

ISSN 2513-8391

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Donations
T: 020 3795 2177
E: supportercare@cysticfibrosis.org.uk

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E: events@cysticfibrosis.org.uk

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:

CF Life Editorial Team, Cystic Fibrosis Trust, 2nd Floor, One Aldgate, London, EC3N 1RE

magazine@cysticfibrosis.org.uk

Welcome to CF Life

The ongoing coronavirus (COVID-19) pandemic has continued to make this a tough time for the cystic fibrosis (CF) community. In this issue, we look at some of the positive ways that CF care and people with CF have adapted in the face of the virus. We also hear from 17-year-old Sophie on how she has adjusted the way she's studying for her A-Levels in these challenging circumstances.

We take a look at how mucus works to keep lungs healthy and how research to understand it better could lead to more effective treatments being developed for cystic fibrosis.

While the licensing of Kaftrio was a massive highlight of 2020, we're still working hard to find treatments that will help everyone with cystic fibrosis. Cicely, who has a rare CF mutation, puts some burning questions on gene editing to Professor Stephen Hart.

In this issue, you'll also hear from yoga teacher Nicole, who shares some exercise tips, and Sarah, who has written about supporting her daughter's transition to adult care. Finally, we take a look behind the scenes of our Helpline and get to know the team.

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

Research

We are thrilled that this year's UK CF Conference (UKCFC) will be going entirely online for the first time on 12 May. Showcasing research you are helping us to fund, the one-day conference will feature themes of CF lung infections, understanding and treating CF symptoms and complications and tackling the underlying cause of cystic fibrosis. While primarily aimed at biomedical and clinical researchers, CF professionals and multidisciplinary teams who work in CF care, all are welcome. UKCFC has always been an opportunity to showcase our research and spark further interaction and collaboration, and we look forward to what this year will bring.

Register today by visiting **cysticfibrosis.org.uk/ukcfc**

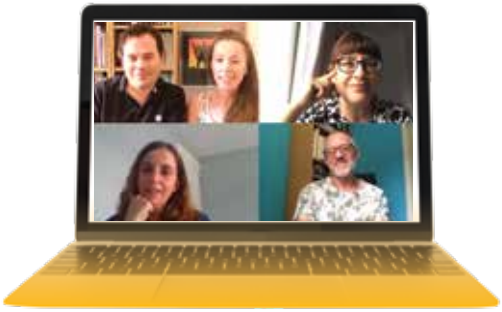
Care

Having access to a qualified social worker with in-depth experience and understanding of your CF can be life-changing. However, a recent survey of UK CF centres by the Quality Improvement team found that just 34% of responding centres have access to CF social workers. In December, we facilitated a meeting between the UK Cystic Fibrosis Medical Association (UKCFMA) and the co-chairs of the UK CF psychosocial group, to better understand this variability. We have committed to supporting CF social worker recruitment, with the development of a business case template and the production of new psychosocial guidelines for later this year.

Find out more about our work to support improvements in CF care at **cysticfibrosis.org.uk/qualityimprovement**

Fundraising

In December, we held the premiere of our first online Carols by Candlelight concert. It was a phenomenal success, raising an incredible £60,000 with over 2,000 people tuning in to watch on the night. The Winchester College Chapel Choir recorded their carols at the legendary Abbey Road Studios, which was then shared via YouTube. We were joined by a chorus of famous faces, including Dame Judi Dench, David Haig, Jenny Agutter OBE, Jane Asher, Damian Lewis and Katie Melua, as well as Emily Wilson from the CF community. On behalf of the Alice Martineau Appeal Committee and the Trust, thank you to everyone who joined us and made this a night to remember!



Campaigning

In October, the Crown Dependencies of Jersey, Guernsey and the Isle of Man joined the four nations of the UK with deals for access to Kaftrio. We are delighted that all those with CF who are eligible for the life-saving drug can benefit from it no matter where they live in the UK. This comes alongside the recent expansion of access to Kalydeco announced in November for children aged over four months. We will continue to work to change and improve access so that these drugs are available for everyone that could benefit from them.

Support

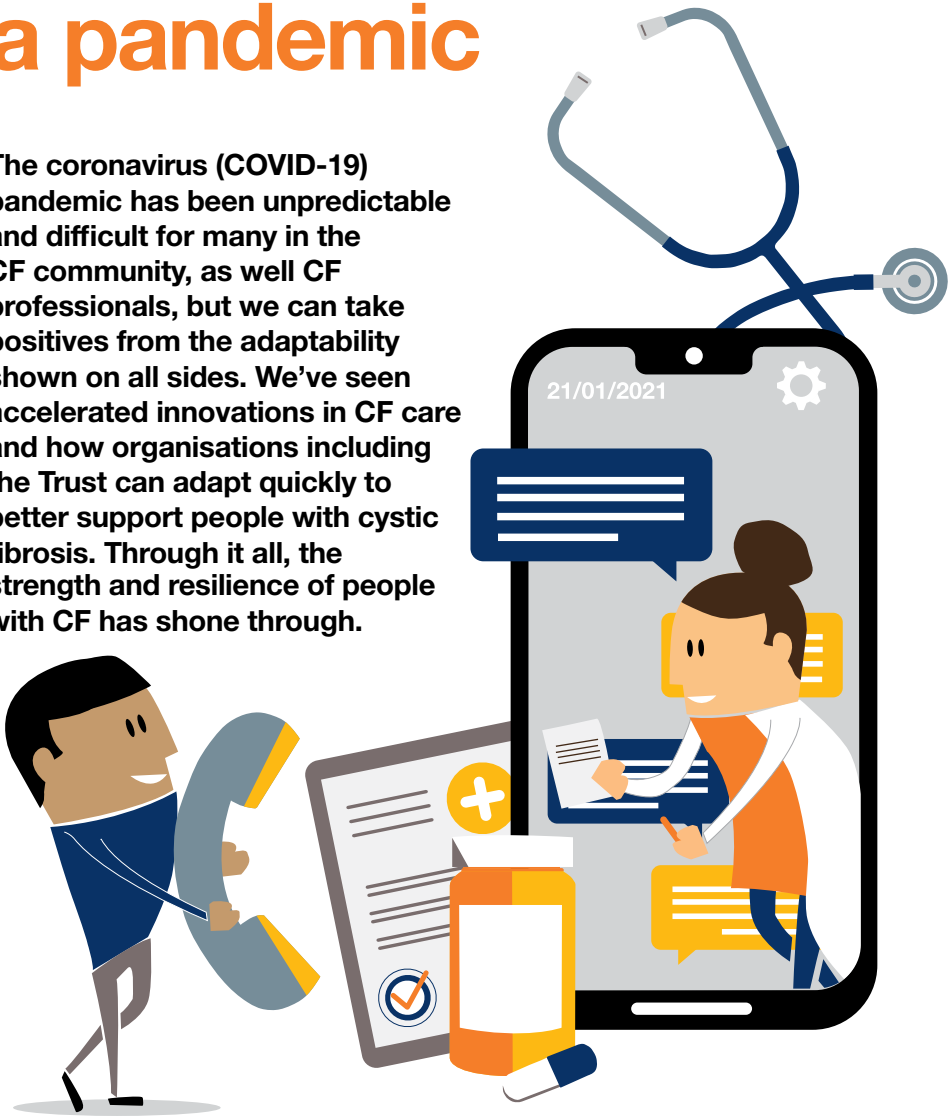
As COVID-19 moved us all into virtual events, we continued to update the community with the latest information and updates from the world of CF through our online CF LIVE series. CF LIVE brings together the CF community, people involved in CF care, researchers and others with an interest in CF, covering topics from Kaftrio, clinical trials and research underway to tackle antimicrobial resistance, to the latest on COVID-19, welfare and employment rights.

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

If there's something you'd like us to cover in a CF LIVE event, let us know at **conference@cysticfibrosis.org.uk**

Finding positives in a pandemic

The coronavirus (COVID-19) pandemic has been unpredictable and difficult for many in the CF community, as well CF professionals, but we can take positives from the adaptability shown on all sides. We've seen accelerated innovations in CF care and how organisations including the Trust can adapt quickly to better support people with cystic fibrosis. Through it all, the strength and resilience of people with CF has shone through.



Accelerated changes to CF care

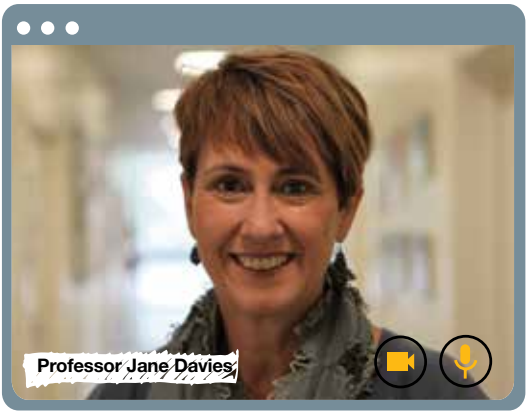
When the pandemic took hold in early 2020, the changes to CF care were rapid: routine clinics were cancelled to prioritise resources for dealing with COVID-19; appointments moved to phone, online or into people's homes; and many people with CF began monitoring more of their own health at home.

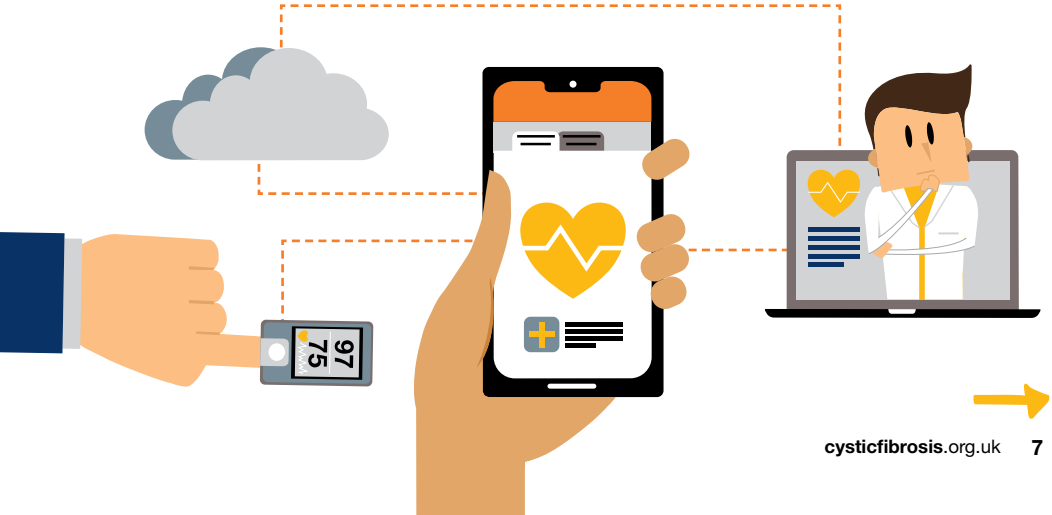
Last year, home-based spirometry was rolled out for thousands of people with CF across the UK. Digital home spirometry kits give people the opportunity to monitor and share clinical data on their lung function with their CF teams without needing to leave home. This initiative builds on Trust-funded digital health research, including the 'Project Breathe' study, which will help us understand whether measuring things like lung function, oxygen levels, activity and weight at home can safely reduce the number of hospital appointments for people with cystic fibrosis.

Developers working on the related smartphone app made it publicly available so that all people with CF could use it to monitor their health from home (see page 17).

Professor Jane Davies, Professor of Paediatric Respiratory Medicine & Experimental Medicine at Imperial College London, explains how the move to home monitoring has been welcome during the pandemic but urges some caution:

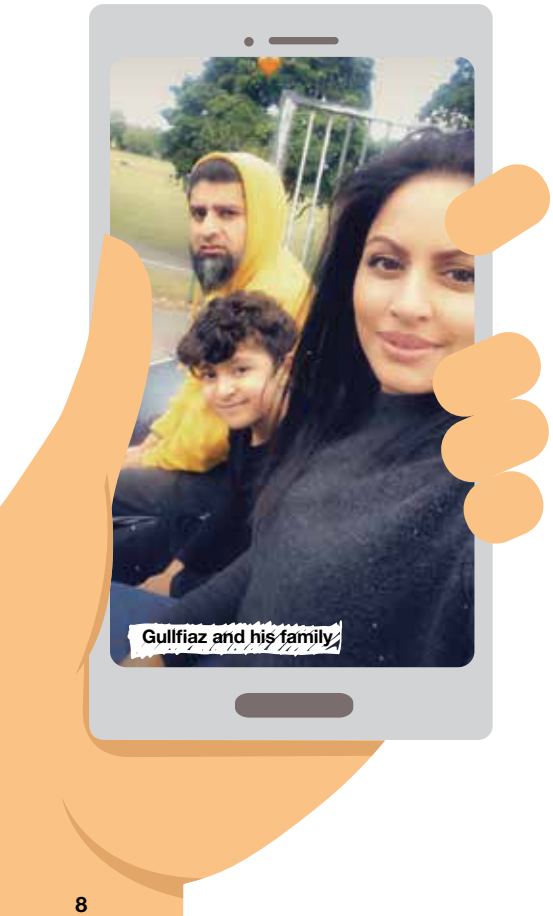
"The adaptability of both the CF community and caregivers has been very impressive but use of home monitoring approaches is still at an early stage and there is more research to be done to be sure of its long-term effectiveness."





Professor Davies notes in particular the difference in accuracy between lung-function measurements taken at home versus in the clinic, variations in equipment, the challenges of monitoring infections (particularly in people unable to cough up sputum), and the potential impact on mental health for people missing out on a face-to-face contact.

Gullfiaz, who has CF, echoes this last concern, saying: "It was great when I was able to go back into hospital for my appointments, not just for my physical health but my mental health too."


Gullfiaz and his family

"It was great when I was able to go back into hospital for my appointments, not just for my physical health but my mental health too." **– Gullfiaz**

Home monitoring isn't new, but it hasn't been so widespread before. Trust-funded CF researchers have been exploring digital technologies for home monitoring since 2016. Other home monitoring systems are also in development across the UK, such as HealthHub. While analysis of the data from the digital health studies is ongoing, there is clearly huge potential for home monitoring to mean fewer face-to-face clinic appointments, faster diagnosis and more effective treatments when health problems develop.

While COVID-19 caused some clinical trials to be postponed, some trials have moved to remote appointments and monitoring rather than people with CF needing to attend hospital sites. This has had a positive impact on future trial design, making trials safer and less time consuming for participants. The Trust has been looking at how to make trial participation more convenient for people and families trying to get on with their lives with CF prior to COVID-19, but the pandemic has forced the issue and shown it's a possibility in some instances.

How the Trust has adapted

People with CF were classified as clinically extremely vulnerable to COVID-19 at the start of the pandemic, adding further challenges to life with cystic fibrosis. From March 2020, we saw a significant increase in demand for the Trust's information and support services, receiving more enquiries to our Helpline, visits to our website and comments on our social media posts than we've ever seen before.

Our response to this demand included: increasing staffing on our Helpline; creating a COVID-19 section on our website and regularly updating the information and advice as things changed; and providing a COVID-19 emergency grant during the first shielding period.

A lot of the Trust's work happened behind the scenes. Early on, we joined with other charities to advocate for people vulnerable to the pandemic. Since the initial outbreak, we have called for improved access to financial support, education, and essential products and services, and for timely and clear guidance from the Government. Back in March 2020, we also called for supermarkets to provide better access to delivery slots for those who were shielding.

More recently the Trust, together with the UK Cystic Fibrosis Medical Association (UKCFMA), wrote to the Joint Committee on Vaccination and Immunisation and Vaccine Minister Jo Churchill MP, calling for clinically extremely vulnerable people to be prioritised for the vaccine. As a result, we were delighted to see that people with CF, along with other conditions, will receive the vaccine sooner than initially proposed. At the time of writing, all adults with CF should have been offered the vaccine.

Like so many around the country, we've had to adapt to working from home (this is the second issue of the magazine brought to you from makeshift desks in living rooms, bedrooms and kitchens!). But going virtual certainly hasn't slowed us down; the Trust has held meetings with the Deputy Chief Medical Officer as well as parliamentarians, clinicians, and civil servants. Since March 2020 we've tabled 16 parliamentary questions via supportive MPs in Westminster, and a further eight elsewhere in the UK to ensure the voices of people with CF continue to be heard during the pandemic.

4,570

enquiries received to our Helpline during March 2020; **40%** relating to financial concerns or support

£130,000

given out through 720 COVID-related grants in 2020

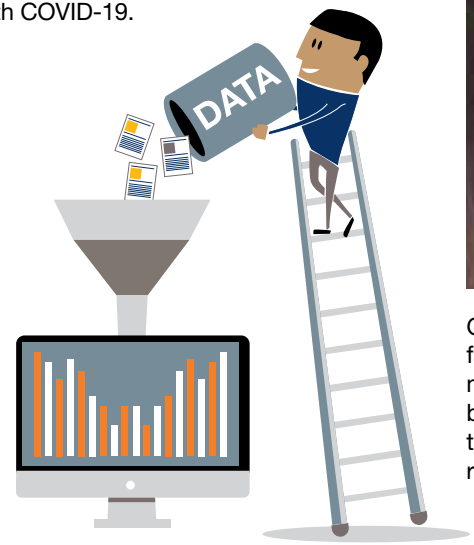
57%

increase in visits to the website in March–April 2020

Collecting COVID-19 data

Throughout the pandemic, the UK CF Registry has showcased the powerful combination of data and global collaboration. At the start of the pandemic, the Registry team created a helpline so that clinical teams could easily report data on cases of COVID-19, which we have shared regularly with the community on our website. With this data, they led the way in a collaboration with other CF registries around the world to build the global picture of COVID-19 and cystic fibrosis. Through this work, they've been able to show that the number of people with CF who have become seriously ill or died from coronavirus is low worldwide.

In further global teamwork, a study supported by the Cystic Fibrosis Trust (as part of the Global Registry Harmonization Group, made up of CF specialists from around the world) revealed that children with CF who do not have pre-existing severe lung damage experience mild or asymptomatic illness when infected with COVID-19.



The strength of the CF community

Through the huge challenges of shielding, changes to care and ongoing uncertainty, people with CF have shown their strength time and again.

Sophie, who has CF, has been able to find positives in a really difficult year, saying: "I think the lockdowns and restrictions have given people more of an insight about what it is like to live with a disability and I hope it will mean people can empathise with us more. COVID-19 has affected my life, but it hasn't really changed it. In fact, COVID has made me more determined to get through the extra adversity we find ourselves in, to get where I want to be in life."

Gullfiaz has found strength through his family: "I'm blessed to have the support of my loving wife and family around me. My boy Isaac, who is six years old, has made this difficult time bearable; he's been my rock who has kept me going."

23-year-old Jessica explains some of the challenges she has faced, saying: “It’s been a tough time, and a very scary one at that. I was in and out of hospital and always had high anxiety when I went in because of not being able to leave the ward, having no visitors and worrying about catching COVID.”

But for Jessica, and many others with CF, the challenges of 2020 came alongside the huge highlight of getting access to the new treatment Kaftrio. “On a positive note, I’ve now started on Kaftrio and I feel like a new woman! I can run up hills without getting breathless, it’s helped me in my areas of delayed puberty and just gave me a whole new lease of life.”

“I was in and out of hospital and always had high anxiety when I went in because of not being able to leave the ward, having no visitors and worrying about catching COVID.”

– Jessica

Despite all the challenges, the CF community has also adapted to new ways to come together and fundraise. Whether getting involved in events like the virtual Running Festival and Ride for Research, or coming up with your own creative ways to raise money, you’ve continued to support people with cystic fibrosis. We’ve loved seeing you all at our online events, from battling it out at the Big Yellow Quiz in CF Week to dancing along at the Christmas party! Our annual Carols by Candlelight is usually attended by around 650 people, but by bringing the event online we were able to share the magical evening with over 2,000 members of our community.

Find all the latest support and advice on COVID-19 at **cysticfibrosis.org.uk/coronavirus**, or contact our Helpline.

Gene genie

Twenty-two year old Cicely Mathews' CF mutations mean that she's one of the people who has the most to gain from a future gene editing therapy. Cicely posted on Instagram to ask the CF community their burning questions on gene editing, and then put them to Professor Stephen Hart, the Principal Investigator of our new Strategic Research Centre.


Cicely

Cicely: How long will it take to get a gene editing therapy to patients?

Steve: In the early '90s, my boss was asked this question at a CF open day in our lab (before the cross-infection rules came into place!). He said, "Well it's going to be about five years." Over 20 years later, despite some encouraging progress, we're not there yet. So, I hope that you'll forgive me if I don't give you an answer to this question. All I'll say is, we're working as fast as we can, we're aware of the enormous need for people with CF to have new treatments and to keep everyone informed on the progress we're making, and the challenges we face.

Cicely: Would it just benefit the lungs?

Steve: We're focusing on the lungs. In the future, the same principle could be applied elsewhere, but the challenge is always how can you get a gene editing therapy to the right place in the body. Gene editing would be a different kind of treatment to a CFTR modulator which is a pill. When you take a tablet, it goes everywhere in the body. When you look at a genetic therapy approach, it is not so easy to get it throughout the body, which is a challenge.

Cicely: What has been the impact of the CFTR modulators on gene editing research?

Steve: Researchers don't see CF as a fixed problem, and the funding agencies don't see it that way either. The modulators are fantastic and to see the impact that they have had is amazing. However, we're aware that not everyone can benefit from them.

Gene editing therapies could benefit everyone with CF, but researchers are focusing first of all on people with CF mutations that can't be treated by the modulators.

Cicely: It seems funding was no barrier in the development of the COVID-19 vaccine; is funding the only thing holding gene therapy back?

Steve: More money would always be a good thing, to have more people working in the field, but would we be a lot further along even if there had been that money? It is impossible to say.

I'm thankful to the Trust for everything that they do and they do a great job. It really is a global effort to develop genetic therapies for cystic fibrosis. We all speak to each other, share results and pick up what they're doing as well. As you'd hope, everyone is working together on this.

"I'm thankful to the Trust for everything that they do and they do a great job. It really is a global effort to develop genetic therapies for cystic fibrosis."

– Professor Stephen Hart

Professor Stephen Hart

Transitioning care as a carer

One of the biggest changes for a child with CF growing up is the transition from paediatric to adult care. This is a big step for young people, but it can be a big step for their families too. That's why in 2020 we produced a new resource for parents and children. Here, Sarah shares her experiences of managing her daughter's transition to adult care.

Our paediatric CF team started to talk about transition when Grace was approaching 15. At the time I felt it was far too early to be thinking about moving teams. Grace was worried as she thought she would have to take over her own care and talk to the doctors without me there, and she didn't feel ready. We'd been lucky to have the same consultant throughout Grace's care, and it was going to be difficult leaving a fabulous doctor behind!

After all the years I've looked after her, I was also worried about not being involved in Grace's care. I still wanted to be involved and, when the paediatric team started talking about complete independence for Grace, I was worried I wouldn't have the same level of access as I had previously.

Sarah and her daughter, Grace

When Grace turned 16, the transition began. We met with the CF adult team nurses at an annual review. I also went to a transition open day, which was really useful as I got to look around the adult facilities, meet the team, and talk to an inpatient with CF about the care they received and how they juggled everyday challenges while in hospital.

The change in venue has been the most difficult part for us. Before, we would always go to our local centre for regular checkups, which was only 20 minutes from home. It's now an hour and a half away by train, so each clinic appointment now takes up most of a day… not ideal when Grace is still studying!

We also lost our community nursing team, which I found the most worrying as we no longer have a health care professional who can 'pop' out to us if we are concerned about Grace's health. However, we are very fortunate that we still have an outreach community nurse who will come and visit us at home if needed – something that influenced our final decision when choosing our adult care, as no other hospital offered this. We also have mobile numbers and direct emails for the CF nurses, which makes us feel more connected with them even though we are much further away from them distance-wise. As a mum, it's given me immediate peace of mind from day one.

> "We are very fortunate that we still have an outreach community nurse who will come and visit us at home if needed."
> **– Sarah**

Grace

Grace has now been seeing the CF adult team for around a year. The team haven't excluded me as a parent; with Grace still studying, she didn't yet want the full responsibility and still relies on me to manage her medication, appointments, and other bits. It's worked well from both our perspectives and I feel the adult team have been very responsive to our needs.

That being said, I have noticed recently Grace has been far more aware of her cystic fibrosis. She's doing more to manage her care than she was before and I can only imagine this is due to the transition. It's a learning curve for her to understand what to look out for and how to find solutions or seek help. I am pleased she is taking more responsibility for her health, medicines and overall care, but as parents our support will always be there.

"She's doing more to manage her care than she was before and I can only imagine this is due to the transition."
– Sarah

I supported the Trust with the development of its transition resources. I really enjoyed being involved in this project, and I think it will reassure parents of children with CF about the transition process and that it's perhaps not as worrying as they thought.

Take a look at our resources for families and young people with CF at **cysticfibrosis.org.uk/transition**

Project Breathe

When COVID-19 started to affect almost every aspect of our lives, the team collaborating on Project Breathe, a digital health research programme initially part-funded by the Trust and later by the CF Foundation, knew being able to monitor and manage CF from home was vital.

Kirsty Hill, Director of Magic Bullet, was inspired to be involved with Project Breathe because her son has cystic fibrosis. She could see the potential difference health tech and remote monitoring could make, to both his life and others living with cystic fibrosis.

Kirsty says: “Seeing health trends gives new insight and empowers proactive health management and earlier self-intervention.”

When COVID-19 hit, the project and remote monitoring solution were still in evaluation and people were being asked not to attend clinic for safety reasons. The consortium quickly agreed that investing in rapid development and enabling broader rollout had the potential to make a difference to many people at this uncertain time.

Since then the app has been downloaded more than 1,000 times, while the remote monitoring platform now supports four clinics in the UK and there are talks with a team in Canada.

“Feedback from our users, both people with CF using the app and their care teams using the clinician dashboard to support remote consultations, has been overwhelmingly positive,” says Kirsty.

“There are silver linings of the shifts we are all experiencing, maybe a much-needed shake up to the management of chronic health conditions like CF is one of them. I really hope these changes are here to stay.”

Find out more about Project Breathe and the new app at **cysticfibrosis.org.uk/adaptingcare**

Exploring exercise

Fitness instructor and yoga teacher Nicole, who has CF, shares her tips to make exercise an exciting, everyday part of life.

- Get involved with a community that supports you, like Beam CF! Beam offers classes and conversational focus groups specifically for those with cystic fibrosis. There's no awkwardness, and we talk about all things CF from coughing up mucus to IVs and our digestion… all the gross things that we all find normal. There are so many online and virtual communities out there and it's really important to utilise them in the CF community, especially as we can't meet in person.

- I rave about yoga to every single person with cystic fibrosis. It's great for understanding new breathing techniques, ways to help our digestion and learn how our body moves with breath. My lung function performance has definitely improved since learning about 'pranayama' (the practice of breathing exercises and patterns). Yoga isn't just about the body, but the mind and soul.

- Getting a qualification in fitness is a great way to learn how your body works and how to incorporate new ways of exercising into your physiotherapy. I've learnt new ways to work my lungs, understand nutrition and deepen my knowledge of anatomy, just by completing courses that have nothing to do with CF, but where I get to understand my body better.

- Goal setting is great, but not directly about your health. Base your goals on joining an exercise class, walking more, or incorporating breathing exercises into your physio. Set goals that are achievable and realistic, but will push you.

Nicole leads our CF Youth dance and yoga workshops – find out how to get involved by visiting **cysticfibrosis.org.uk/cfyouth**

cysticfibrosis.org.uk

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Cystic Fibrosis Trust

Celebrating a hard-fought campaign

Back in June 2020, NHS England announced the wonderful news that a deal had been agreed for the life-saving drug Kaftrio to be made available from the day of its license, bringing hope to thousands of eligible people with CF across the UK. Similar deals then followed for Wales, Northern Ireland and Scotland, and the formal licence was granted by the European Commission last August.

This breakthrough reflects the hard work and tireless campaigning of the CF community together with the Trust over the past five years: from signing the first petition that was handed in at Number 10 Downing Street in 2016 to sharing your experiences with appraisal bodies such as NICE and the SMC; meeting with health ministers UK wide; and demonstrating outside the Vertex headquarters in 2019. By supporting our work through fundraising as well, you've helped us to keep access to life-saving drugs at the top of the political agenda.

Paul's daughter Niamh has CF and was able to start taking Kaftrio recently:

"When Niamh was born, my dad told me not to worry, that they'd find something for Niamh. He must have had some sort of vision as exactly 12 years later, just as Niamh reached her 12th birthday, Kaftrio became available. The timing couldn't have been any better – it was just meant to be. Her future really is 'unlimited' now.

"For many people with CF, it really is a life-changing drug."

– Paul

Paul and Niamh

"I started fundraising for the Cystic Fibrosis Trust back in 2010, when I got a group of 30 people from work together to conquer Ben Nevis and raised over £3,000. Since then, I've tried to raise at least £1,000 each year. The total now stands at over £14,000, plus a lot of awareness raised for CF too. I have loved playing a part in the future of Niamh's care through fundraising for the Trust, and really feel like I have helped her to get to where we are today."

Over the past few months, we've seen lots of great stories of people getting their first Kaftrio prescriptions or sharing the positive impact that it's already having on their health (read about the Kaftrio 'purge' in our mucus feature on page 26). For many people with CF, it really is a life-changing drug, so we're continuing our work to get access for everyone who could benefit. Kaftrio isn't the right treatment for everyone though, and we are working to find alternative treatments to ensure everyone with CF can live a life unlimited by the condition.

I have loved playing a part in the future of Niamh's care through fundraising for the Trust, and really feel like I have helped her to get to where we are today."

– Paul

Kaftrio has brought mixed emotions for those who are eligible for the treatment as well as those who will not be able to benefit from it.

Check out our factsheet on the emotional and social impact of Kaftrio at **cysticfibrosis.org.uk/factsheets**

Paul and fellow fundraisers

Remembering someone special

Since his daughter Louise Jane died in 2011, retired police officer Simon McLean has found comfort and hope in fundraising and remembering Louise with a tribute fund. Simon, whose granddaughter also has CF, shared his story with us.

Louise Jane

The CF gene was identified not long after Louise was born in 1988, and from when she was diagnosed, I was part of the West of Scotland branch of the 'CF Research Trust' as it was then.

Like most children with a life-limiting condition, Louise had an amazing spirit. There seems to be a drive and determination inside such kids, to live life to the full while they can. Louise was the life and soul, her main concern being how her condition was affecting others. Sadly, Louise's mother Maggie passed away in 2016 with a broken heart.

Since losing Louise, I have found some solace in, firstly, the Stars tribute pages, which Louise loved to peruse on occasion as she had lost many friends to cystic fibrosis. The truth is that I find comfort in many things that have become rituals of remembrance: visits to Louise's graveside, visiting her favourite places and talking about her with friends and family.

Part of that is now the tribute fund, which allows anyone, across the globe, to grab a simple, private moment of reflection and to contribute if they want.

My family and friends are forever dreaming up innovative ways to raise money. Kilt Walks, sponsored anything, karaoke competitions and raffles; the list is almost endless but the underlying motives are always the same. To have fun, mix with like-minded folk to raise funds and to remember Louise and all of those whose lives were cut short.

Importantly, there is also hope: that those still with us or yet to be born can lead a full life.

I visit the tribute fund regularly, mostly to 'touch base'. When I light a candle or write a few lines I always share it on social media. This gives me comfort, but I have learned that it also allows others who knew Louise or lost a loved one, to grieve and reflect. This ripple effect is quite profound and satisfying.

Someone only really dies when their name is said out loud for the last time. Louise will always be with me and I can keep her memory alive, hopefully for generations.

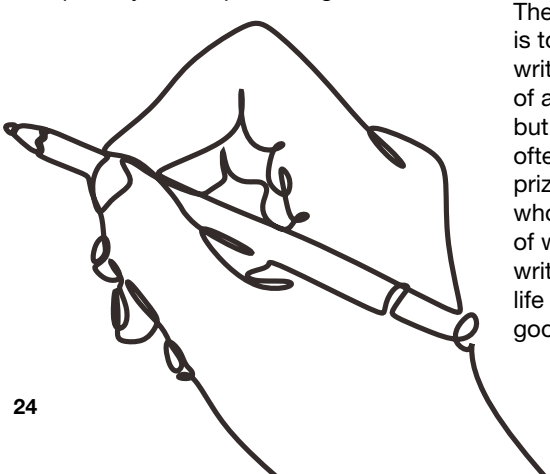
Find out more about tribute funds at cysticfibrosis.org.uk/tributefund or email Daisy Board, In-memory Officer, **Daisy.Board@cysticfibrosis.org.uk**

Changing the narrative

23-year old Jake, who has CF, is the founder and director of The Barbellion Prize – a book prize dedicated to championing ill and disabled voices in writing. Here, he explains why he created the prize.

The idea to create the Barbellion book prize came to me in the process of writing my own memoir, 'Neither Weak Nor Obtuse'. It's not a typical memoir; it's more of a phenomenology (philosophical study) of illness, about how my life-long suffering with CF and various other medical conditions has shaped my view on the world.

In my book I reflect on the wider perception of people with illnesses like mine, or of people with disabilities, and how they are often overlooked and misunderstood. Disabled people around the world do not have a voice that is heard enough, and this is especially true in publishing.



The main purpose of the Barbellion Prize is to further ill and disabled voices in writing, promoting works not in so much of a marketing or advertising kind of way, but championing those authors that aren't often looked at. The other purpose of the prize is as a tribute to W N P Barbellion, whose work is probably the exemplar of what you're looking for in this sort of writing – a very good representation of a life as a disabled person, as well as very good writing.

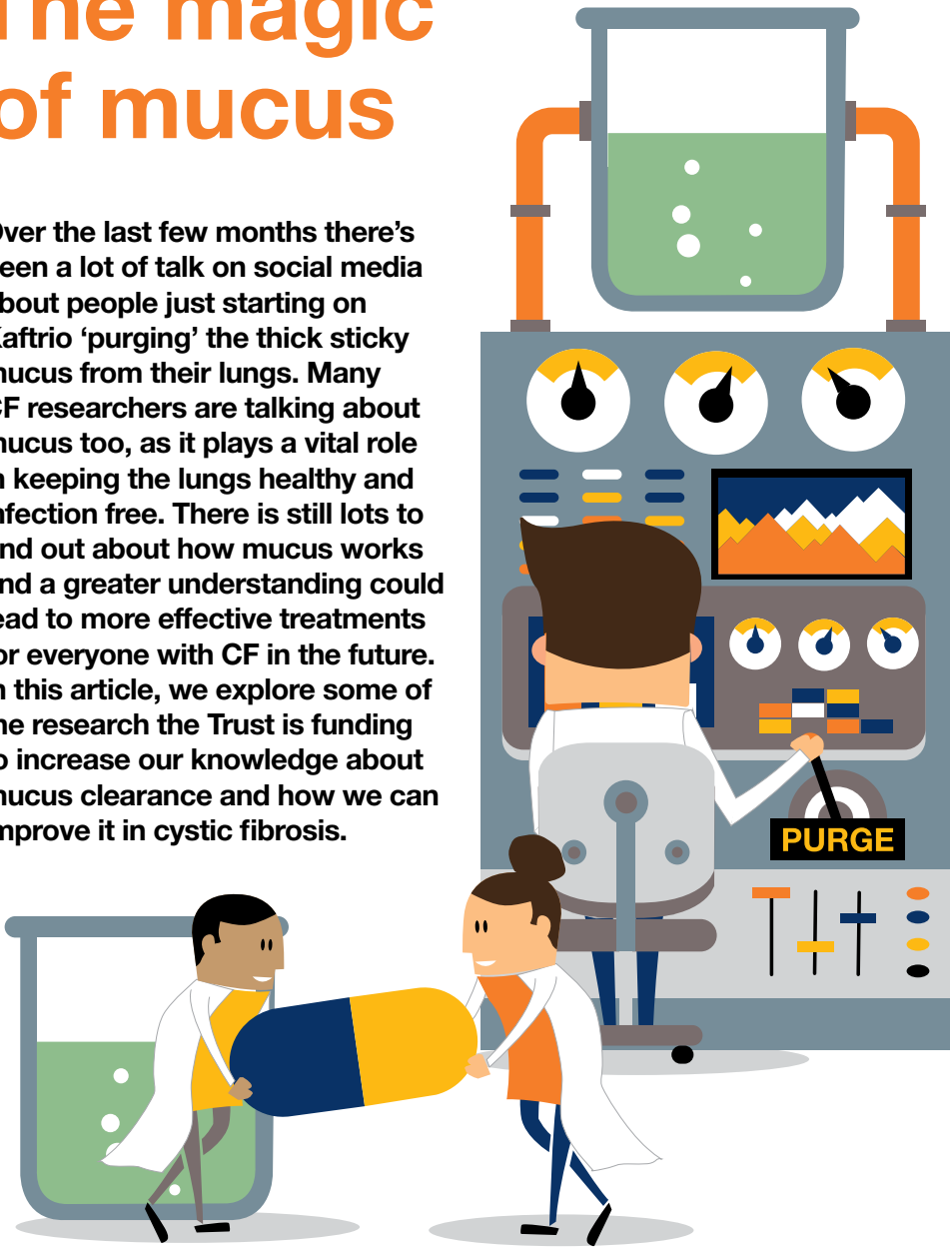
I started the prize in November 2019, so this is the inaugural prize. It's almost completely operated on a shoestring, which is nice and grassroots in a way. We accept submissions from publishers, self-published authors and agents of works from disabled authors. The work need not entirely be about disability but it must be at least represented somewhere, even implicitly or as a secondary consideration. The prize is then awarded to an author whose work has best represented the experience of a chronic illness or disability.

Inspired by The Barbellion Prize, take a look at some recently published books from members of the CF community!

To find out more about Jake and the Barbellion Prize, visit **thebarbellionprize.com**

The magic of mucus

Over the last few months there's been a lot of talk on social media about people just starting on Kaftrio 'purging' the thick sticky mucus from their lungs. Many CF researchers are talking about mucus too, as it plays a vital role in keeping the lungs healthy and infection free. There is still lots to find out about how mucus works and a greater understanding could lead to more effective treatments for everyone with CF in the future. In this article, we explore some of the research the Trust is funding to increase our knowledge about mucus clearance and how we can improve it in cystic fibrosis.

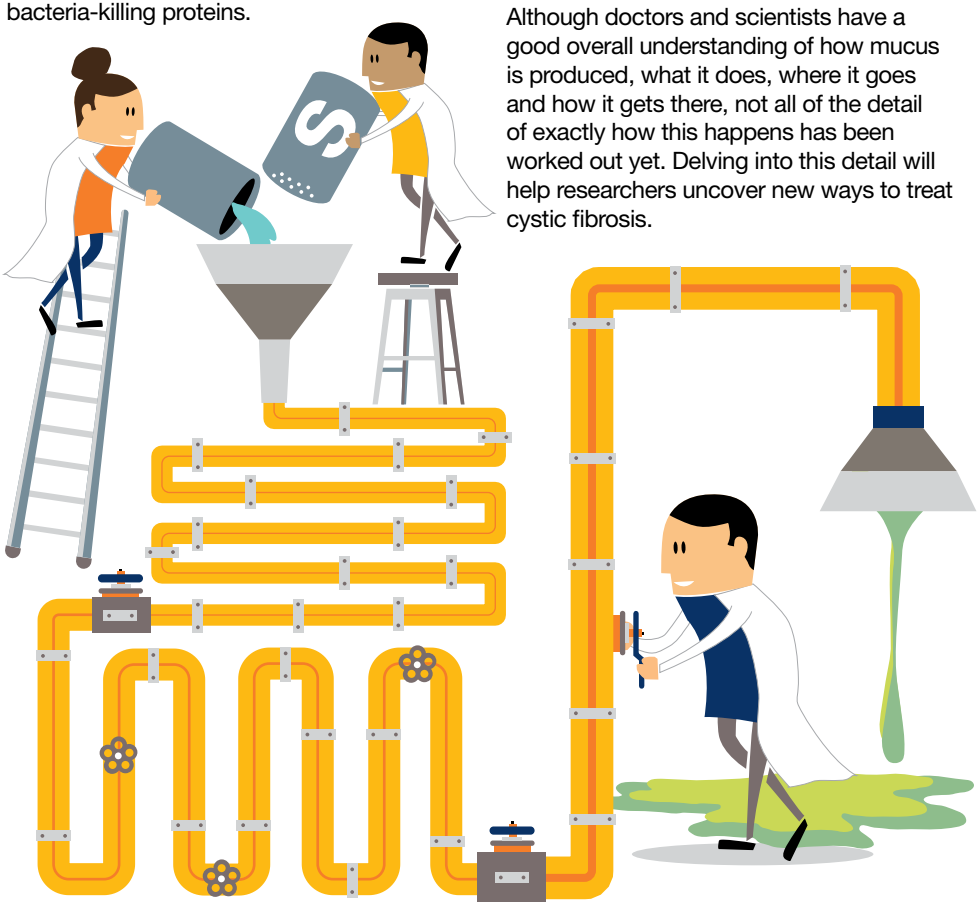


What is mucus and how does mucus clearance work?

Mucus is a liquid that is spread very thinly across the surface of the lungs. It acts as the first line of defence in keeping the lungs healthy and clean. Mucus is made by mixing up a 'recipe' of over 300 different proteins and the right amount of water and salts to keep it the right consistency. The mucus traps bacteria and any tiny debris that has been inhaled, preventing these things from entering the lungs. Mucus also contains bacteria-killing proteins.

Importantly, mucus isn't designed to stay still – once it has trapped the inhaled bugs and debris, it needs to move them out of the lungs quickly. To do this, mucus is carried along the surface of the lungs, up wider and wider airways, until it is carried to the throat where it can be cleared. The mucus is carried on this journey by the beating of tiny hairs called 'cilia' that sit on the surface of specific types of cells lining the lungs. For this to work well, the mucus should be neither too runny nor too thick.

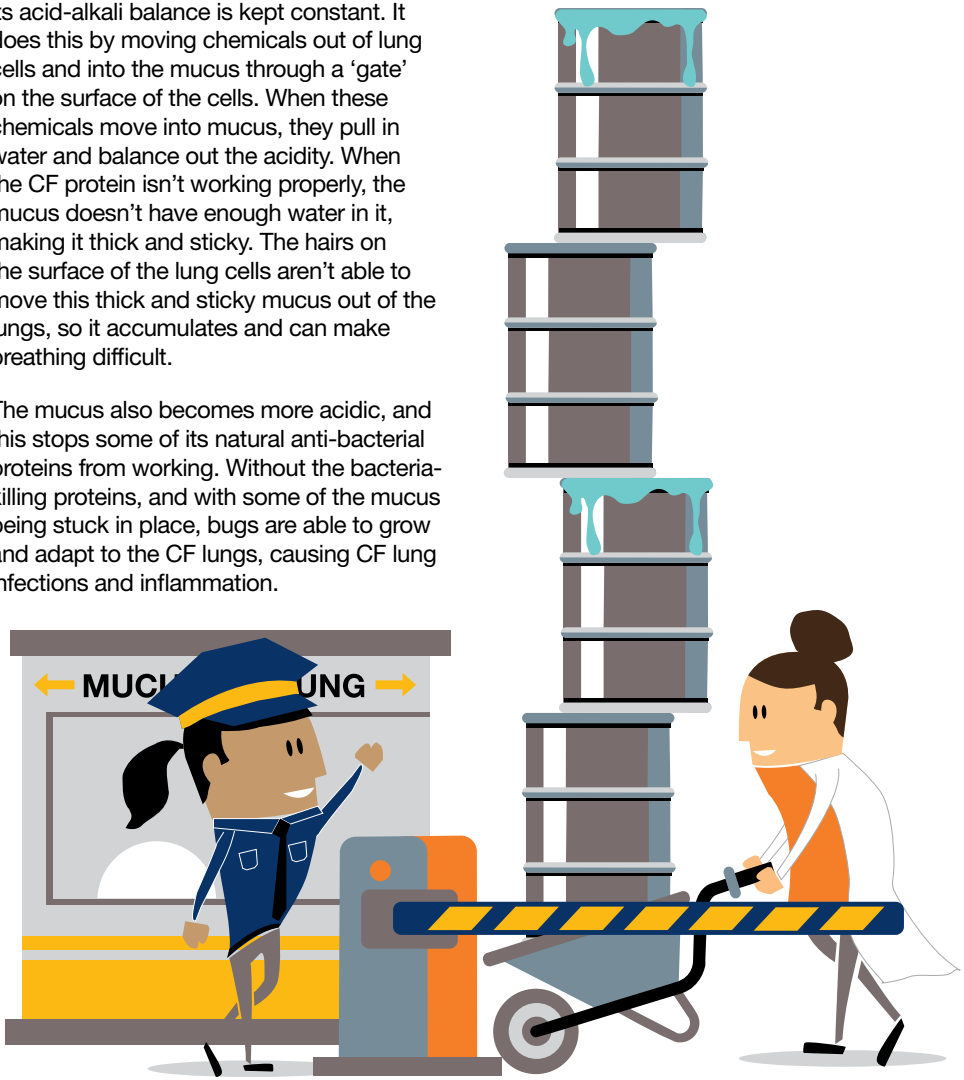
Although doctors and scientists have a good overall understanding of how mucus is produced, what it does, where it goes and how it gets there, not all of the detail of exactly how this happens has been worked out yet. Delving into this detail will help researchers uncover new ways to treat cystic fibrosis.



What happens to mucus in cystic fibrosis?

In CF, the mucus clearance system goes wrong. One of the jobs of CFTR (the protein that is faulty in CF) is to make sure that the mucus layer is kept runny enough and that its acid-alkali balance is kept constant. It does this by moving chemicals out of lung cells and into the mucus through a 'gate' on the surface of the cells. When these chemicals move into mucus, they pull in water and balance out the acidity. When the CF protein isn't working properly, the mucus doesn't have enough water in it, making it thick and sticky. The hairs on the surface of the lung cells aren't able to move this thick and sticky mucus out of the lungs, so it accumulates and can make breathing difficult.

The mucus also becomes more acidic, and this stops some of its natural anti-bacterial proteins from working. Without the bacteria-killing proteins, and with some of the mucus being stuck in place, bugs are able to grow and adapt to the CF lungs, causing CF lung infections and inflammation.



Keeping mucus hydrated for everyone with cystic fibrosis

Kaftrio and other CFTR modulators work by restoring the function of the CFTR protein, re-opening a gate onto the surface of the lungs and allowing chemicals to pass through it. This makes the mucus runny enough to be transported out of the lungs by the hairs on the cells, taking away any trapped bacteria too. The 'purge' people often have when starting Kaftrio is a clearance of the backlog of mucus that has been trapped in the lungs.

"It happened quite suddenly, about nine hours after taking my first dose," explains Holly, who has recently started taking Kaftrio. "Each breath sounded crackly, and I felt a strange bubbling sensation in my upper chest. I then had a very strong need to cough and up came all this dark mucus. It was as if the walls of my chest were melting away, like honeycomb melting into honey. Straight away I could breathe more deeply. It was an incredible experience. Even though it was technically unpleasant, it was such a relief to know Kaftrio was working that I started crying tears of joy."

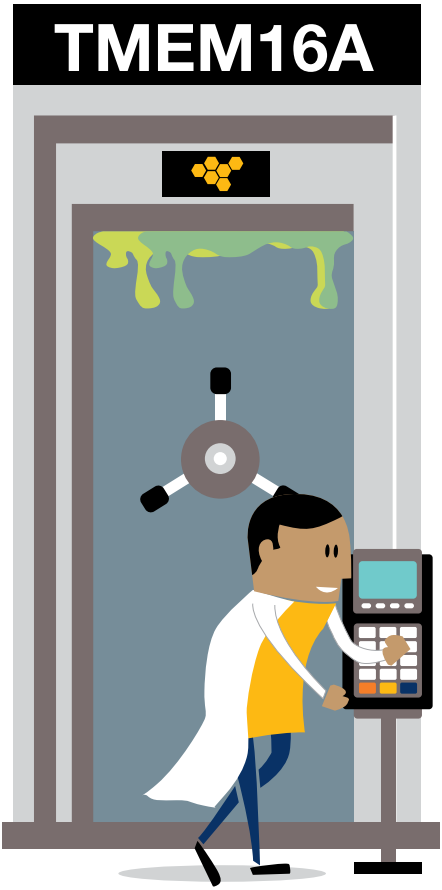
"Even though it was technically unpleasant, it was such a relief to know Kaftrio was working that I started crying tears of joy."

The CFTR protein isn't the only protein responsible for maintaining the right properties of mucus. Dr Mike Gray and colleagues funded by the Trust through a Strategic Research Centre (SRC) grant are investigating several other proteins that work with CFTR to do this. Their goal is to design drugs that will restore mucus clearance for everyone with CF, irrespective of whether or not they're able to take one of the CFTR modulators.

These other proteins that Dr Gray and colleagues are investigating also act as 'gates', to allow chemicals to pass into the mucus and then increase its wateriness and keep its acid-alkali balance. Their idea is that if they can find drugs to boost the properties of alternative gates then it will have the same overall effect of improving how mucus works.

An unsolved puzzle

One gate that Dr Mike Gray and colleagues are particularly interested in is known as 'TMEM16A' – a protein with many different roles in the cell. They have discovered that TMEM16A contributes to excessive mucus production in people with cystic fibrosis. However, it can also improve mucus clearance. So the question is, is it better to block TMEM16A and block the role that makes the situation worse, or is it better to activate TMEM16A to activate its role in improving mucus clearance in the CF lung? Dr Gray is keeping an open mind!



"We're currently considering both options, and will continue to work on the identification of both activators and blockers of TMEM16A. We will examine their effects in as many different lab models of CF as we can and, if possible, in people with CF too. We believe this approach will lead to a much clearer understanding of the role of TMEM16A in healthy lungs and in lung diseases. Ultimately, our studies will benefit and inform future therapeutic strategies that act on this gate to benefit all people with CF, irrespective of their genotype," he says.

The Mexican wave of mucus movement

Re-hydrating the mucus and re-balancing its pH is one way to improve mucus clearance. University of Cambridge-based Professor Pietro Cicuta is working with an international team of researchers to investigate an alternative approach that seeks to better understand how mucus moves, as part of their Trust-funded Strategic Research Centre.

In order to move mucus out of the lungs, the hair-like cilia move in a coordinated way. They take their cue to move from neighbouring cilia, in the same way that spectators co-ordinate the movement of a Mexican wave around a sports stadium. The thick and sticky mucus found in the CF lung disrupts the normal wave movement.

"It's a complicated process to understand; mucus is very stretchy, and that affects how the cilia move it across the lungs," says Prof Cicuta. "If mucus is thick, as it is in CF, the cilia can't move it. But the cilia can't move the mucus if it is too runny either. Easy-to-move mucus is probably somewhere in the middle! Once we work this out, we hope to be able to tailor treatments for a good mucus clearance for every person with cystic fibrosis."


Professor Pietro Cicuta

The mucus clearance system has an important role in keeping our lungs healthy. We hope both Dr Gray and Professor Cicuta's SRC research programmes will make important contributions to restoring the magic of mucus for everyone with cystic fibrosis.

Starting sixth form in a pandemic

17-year-old Sophie, who has CF, shares her experience of choosing A-levels and starting the new school year during the COVID-19 pandemic.

I knew that the process of choosing and starting my A-levels during a pandemic would be challenging and nerve-wracking, especially alongside living with cystic fibrosis. What made it even harder was that, because of COVID, I couldn't attend open evenings or taster days for the subjects I was interested in studying. My school did host virtual induction days, but it just didn't feel the same as being able to meet the teachers face to face.

Sophie

> "It was scary going back into school in September after months of shielding"

Because of lockdown, I had lots of time to think about my options but ultimately, I had to go with my instincts and choose the courses that would give me the most enjoyment and provide me with exciting career opportunities for the future. I picked English Literature, Psychology and Media Production and I am really enjoying all of them so far, so it's reassuring to know I made the right choices!

It was scary going back into school in September after months of shielding – going from being inside my house every day to now being back at school with lots of people. My sixth form was very supportive though and put plans in place for me to do my study sessions from home, which made me feel more at ease about protecting myself during these uncertain times.

I have always been self-conscious of my CF cough. Going back to school last year, it really worried me knowing that people would see me coughing and panic, thinking it was contagious. I emailed the head of Sixth Form and my subject teachers with some useful information about my CF, which helped them to understand more about me and my medical condition. I felt better knowing that they were aware, and it meant I could leave the classroom for a break if I needed to clear my airways or do some physio. This really helped with my transition back into school life, and my Sixth Form was excellent in making sure that I feel safe.

Although the workload of A-levels can feel overwhelming, it's important to take time to do the things that you enjoy as well, such as a walk through the park or watching your favourite film. For me, this means going on walks with my puppy; not only does it benefit my mental health, but it has also kept me active during the lockdowns. Socialising is also important, and so I really enjoy a video chat with my friends and family to keep me company even when we can't meet in person.

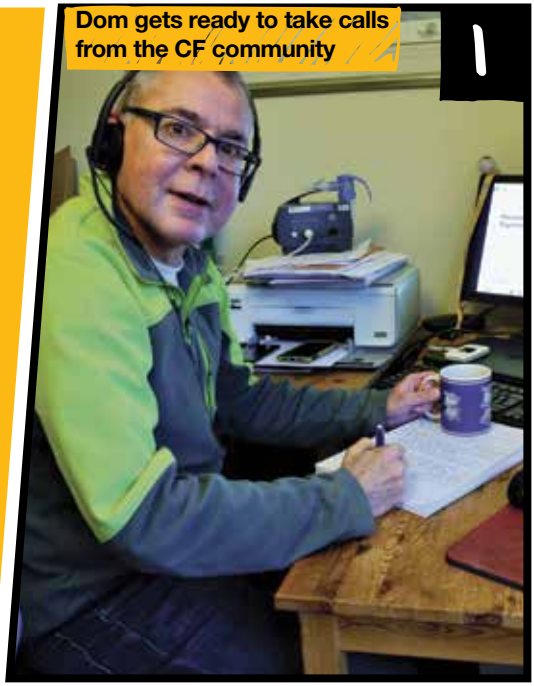
Our secondary school pack includes a new individual healthcare plan called 'My CF Planner', which can help explain your CF to your school.

Find it at **cysticfibrosis.org.uk/secondaryschool**

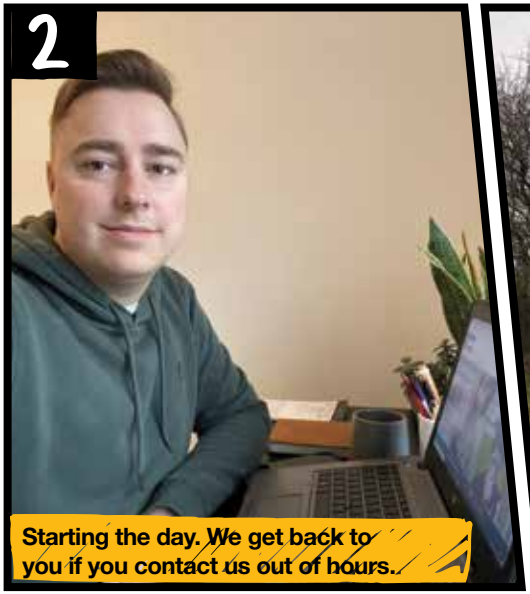
If you have an idea for the next Young Voices or would like to share your views, get in touch on **Twitter @cftrustyouth** or email **cfyouth@cysticfibrosis.org.uk**

Days in the life

Whether by phone, email or even social media, you can count on our Helpline team. Our friendly advisors offer emotional support, give information about CF and can help you access our support services such as Welfare Advice and grants. But who are they, and what does a day in their working life look like? Take a look behind the scenes.

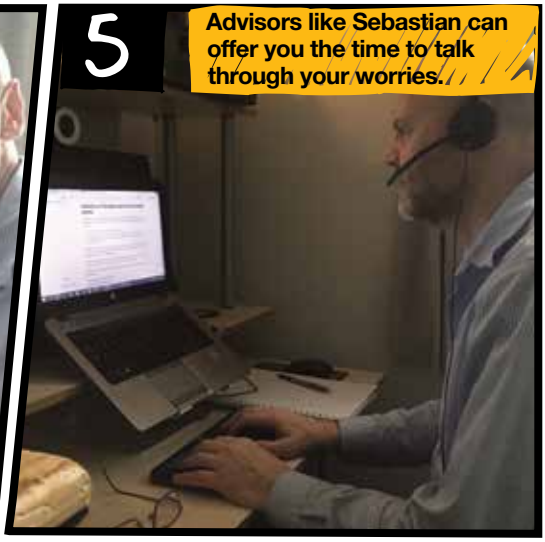


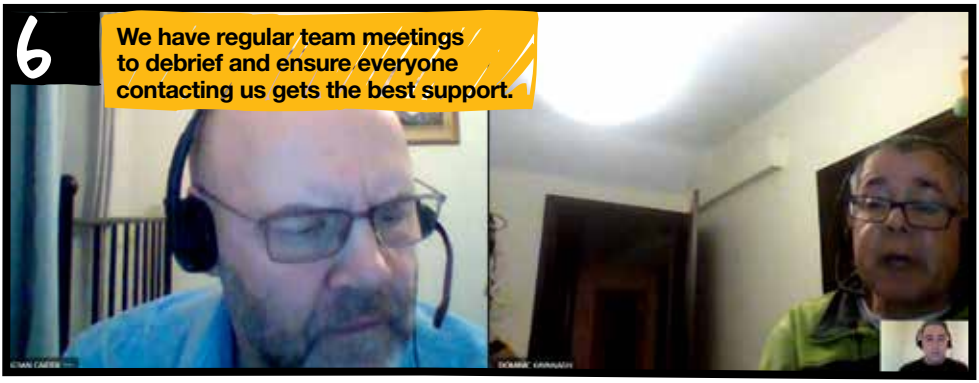
Dom gets ready to take calls from the CF community



Starting the day. We get back to you if you contact us out of hours.

Taking a lunchtime walk gives Matthew (left) time to reflect.





If you need information or support on any aspect of CF, you can call the Helpline on **0300 373 1000** or email **helpline@cysticfibrosis.org.uk**

Cystic Fibrosis our focus

CF Week is coming!

CF Week 2021

14–20 June 2021

Save the date for our annual awareness and fundraising week.

Wear Yellow Day

18 June 2021

Get ready for the boldest and brightest day of the year! We have plenty of hints and tips to help you start planning your yellow-themed event. We look forward to raising vital funds together for people with CF everywhere!

Visit **cysticfibrosis.org.uk/yellow**

Fighting for a Life Unlimited