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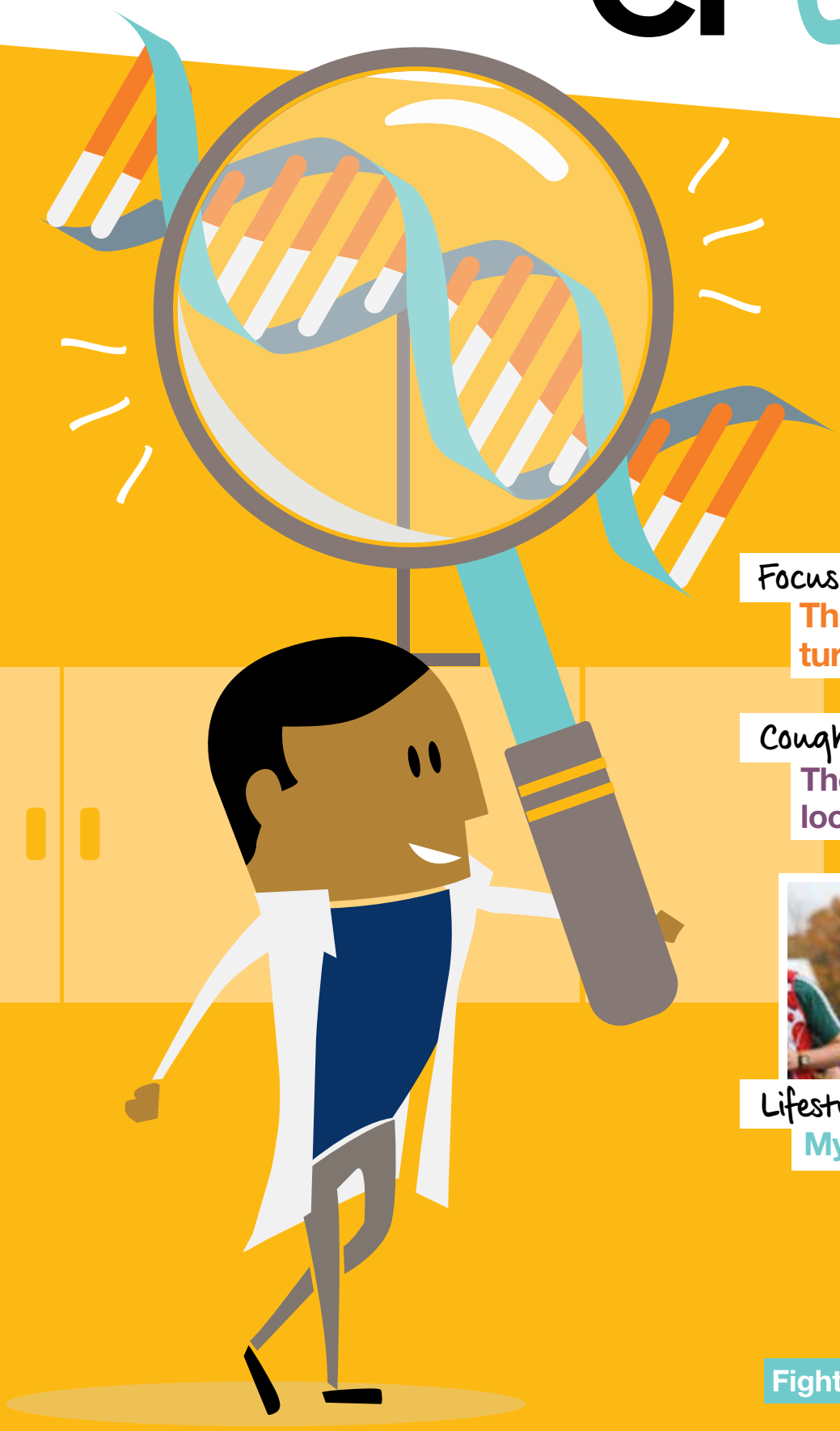
CF Life

Issue 6



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Cystic Fibrosis Trust

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ISSN 2513-8391

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Useful contacts

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

Events and fundraising enquiries
T: 020 3795 2176
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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:

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

magazine@cysticfibrosis.org.uk

Welcome to CF Life

In this edition we celebrate the 30th anniversary of the discovery of the CF gene, a breakthrough both for science and medicine. We look back at what it has meant for CF care and science, reflect on what this journey tells us about the nature of research and look at what the future might hold.

We'll also be exploring mental health and cystic fibrosis.This subject can be as personal and emotive as it is complex. Regardless of whether you are affected by CF or not, your experience of mental health is unique. We hope to start a conversation that will empower people to share their stories, learn from one another's experience and contribute to a deeper understanding.

Your support in the fight for access to medicines has led to some big steps forward in recent months, including a breakthrough in Scotland (p15) and a new Health & Social Care Committee in Westminster dedicated to cystic fibrosis.

Finally, this is the sixth 'CF Life' since you helped us redesign and revamp the magazine. We'd like to thank you for your support and ask you to tell us what you think so far.
Please take a moment to fill out our survey at www.surveymonkey.co.uk/r/CFLife2019.

If you have any specific feedback on this issue, or ideas for future editions, email us at magazine@cysticfibrosis.org.uk.

The CF Life Team

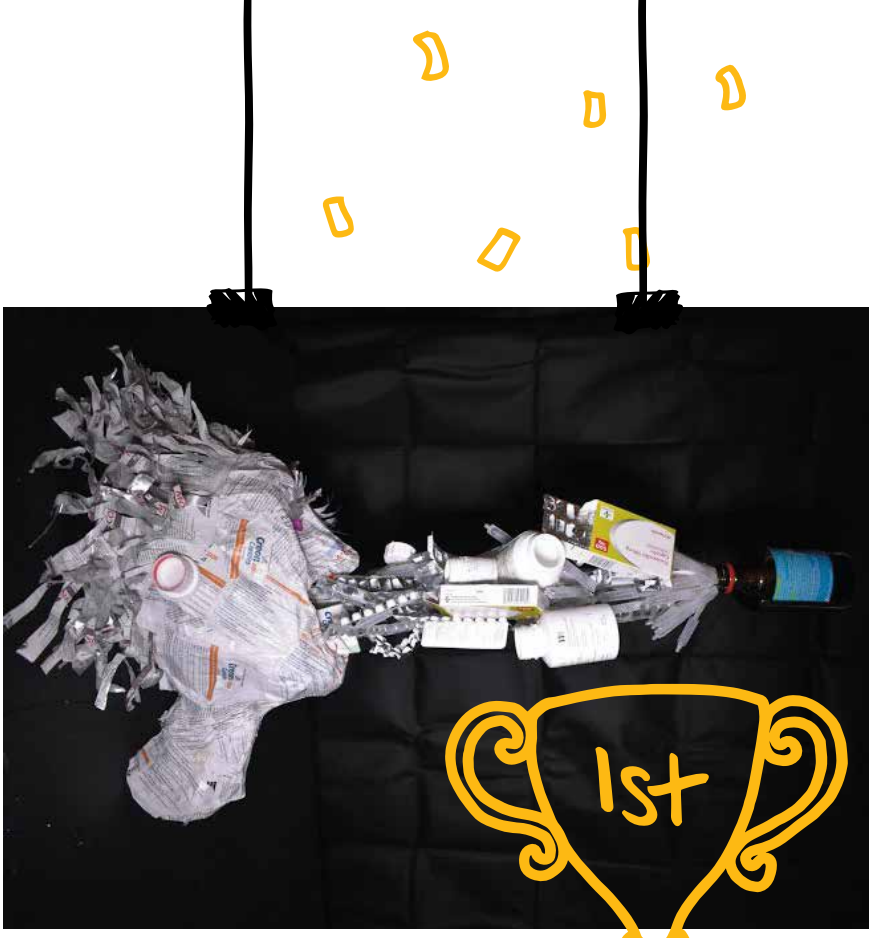
CF Creates

In the last issue of CF Life, we spoke to Robin, a marine biologist with CF who opened the zero-waste shop 'Natural Weigh' in Wales.

Robin told us: "As a household we are constantly trying to reduce the amount of waste going into our non-recycling bin. I often find my thoughts being drawn back to the copious amounts of non-recyclable waste I generate as a result of my medication."

This got us thinking, and we set our community a challenge – to turn some of that empty packaging into something fun, useful or creative, and help us raise awareness of the huge burden of care that medication causes people with cystic fibrosis.

We invited Robin and Laura, co-founder of the luxury ethical womenswear brand Careaux (see p30) to help us judge the results.



'CF packaging – a little hard to swallow' – Emma, Milton Keynes

"My 14-year-old daughter Maisie has cystic fibrosis. I collected the packaging from her medication over two months, though only used about 2/3 of it in the finished piece. I think it will shock most people to see how much medication a healthy child with CF needs to take. It shocked me to see exactly how much packaging is used, but to keep Maisie healthy there isn't much choice."

Laura says: "It is a really cool, arty piece and it shows how varied the medications we have to take are."

Robin: "This a great little creation making use of all the different types of packaging the medicines come in. It's quite a thought-provoking design as well, showing just how much medication gets poured down patients in order to ward off infection and keep us fighting fit."

Think you can have a go and missed out on the competition? We're always on the lookout for people with CF who are creative to share their work and stories with us for future articles, features and campaigns. So, whether you are an artist or a creative genius, drop us a line at magazine@cysticfibrosis.org.uk.

2nd

‘Medi-cender’ – Lauren Blow and her eight-year-old son, Finley, who has CF, Manchester.

“This is an ender dragon from Minecraft. My son is such a trooper with his medication and he looks forward to playing Minecraft once his nebs and physio have been done each evening. His favourite character is the ender dragon so he helped me make one from his medications.”

The dragon features: 18 days’ worth of muco-clear saline solution vials, two weeks of mucoclear boxes, four weeks’ worth of Creon 10,000, a month of Menadiol, vitamin tablets, a month’s inhalers, and a week’s worth of antibiotics that Finley had to take for a bug that wouldn’t shift.

Robin: “I really love the creativity of this one, the use of the different parts of medication packaging to make up the dragon is great fun and really effective. It shows just how many different forms medicine packaging takes.”

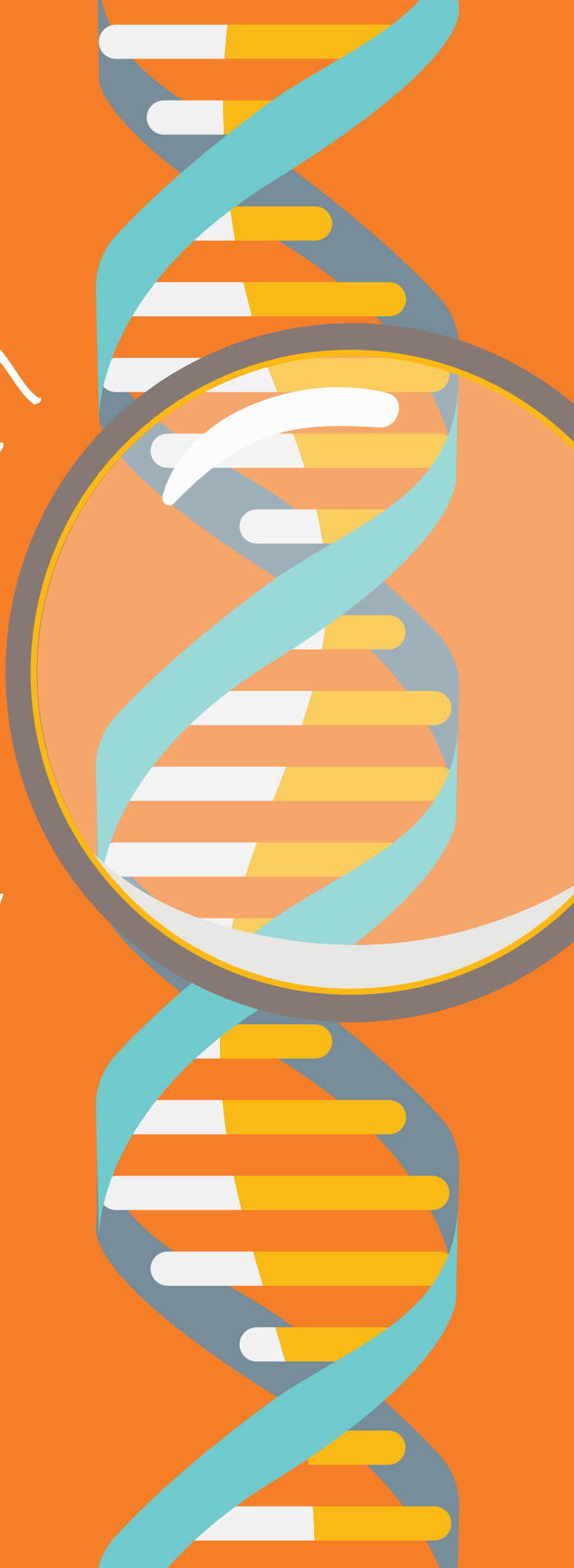
3rd

‘Creon Pot Coffee Table’ – Melissa, Ellis (aged nine, with CF, holding the sign) and sister Hattie.

“It took 5,500 Creon capsules – 137 and a half days of Creon supply – to make the table. We enjoyed entering the competition and Ellis’ sister helped too.“

Laura: “It is a really good and practical creation and it shows how much medication is needed just for one aspect of cystic fibrosis.”

Check out more fantastic entries at cysticfibrosis.org.uk/cfcreates.



The CF gene turns 30: What its discovery means today

Marking three decades since the discovery of the gene that causes CF, we look at the lead up to the incredible breakthrough and examine how the discovery shaped research and what it means for current and future treatments.

When it was revealed at a hurriedly-arranged press conference in 1989 that the gene that causes CF had been pinpointed, it was unlike any research announcement that had come before it.

On one frantic day – 24 August – researchers found themselves being flown by private jet, in almost rock-star-like fashion, to announce at conferences in Toronto and Washington that the search for the gene was over.

"The discovery was not a 'eureka moment'. It was more of a slow jump."

- Dr Lap-Chee Tsui

The discovery was initially going to be made public the following month, with accompanying data in the peer-reviewed journal, 'Science'.

But after the breakthrough was leaked to and published by Reuters two weeks early on 22 August, pandemonium soon followed.

With the journal's press office swamped by reporters, 'Science' broke with tradition by lifting the publishing embargo on the data surrounding the discovery. Patents on the discovery were filed that same day, and press conferences arranged. Meanwhile, the research teams from The Hospital for Sick Children (SickKids) in Toronto and the University of Michigan's Howard Hughes Medical Institute, which jointly discovered the gene, were catapulted into the limelight of the worldwide press.

After years of narrowing down the search, the discovery was not a 'eureka moment', admitted Dr Lap-Chee Tsui, who co-led the team in Toronto. Instead, "It was more of a slow jump."

However, the media frenzy in which the findings were revealed illustrated just how significant the breakthrough really was, not just for CF, but for human genetics itself.

Dr Lap-Chee Tsui

How the gene was found

The human genome – the entire length of our DNA – is made up of approximately 3 billion building blocks (that's 3,000,000,000), so the task of finding the gene that causes CF was always going to be monumental.

Using an analogy, Dr Tsui said researchers first narrowed down the gene's location to somewhere between Halifax, on Canada's east coast and Vancouver, on the west coast, before pinpointing it to Toronto, then honing-in on a particular street, and finally the specific house; the gene.

Numbers game of gene hunting

Researchers in the 1980s knew that somewhere in this massive length of DNA lay a gene that was damaged in people with cystic fibrosis. They found the CFTR gene using a variety of techniques, narrowing down the search by:

- **'Walking' or 'jumping' along the DNA:** systematically analysing the DNA, building block by building block, skipping sections when it became harder to analyse.
- **Finding marker posts for the CF gene:** reducing the distance that they had to 'walk' or 'jump' by only looking between 'marker posts' of DNA that set the boundaries for the area they needed to search.
- **Analysing DNA at the scene:** using data on which segments of DNA were linked to sweat glands; groups of cells known to be affected in cystic fibrosis.

The excitement surrounding the breakthrough was partly down to the potential it had to shape the diagnosis and treatment of cystic fibrosis.

At the time, 'Science' said the discovery would "improve cystic fibrosis diagnosis, including prenatal screening tests for people who carry the defective copy of the gene… It also raises hopes for better CF treatments, perhaps new drugs or even gene therapy to replace the defective gene itself…. No one can predict, however, how long it might take to do this or even if it will prove to be possible."

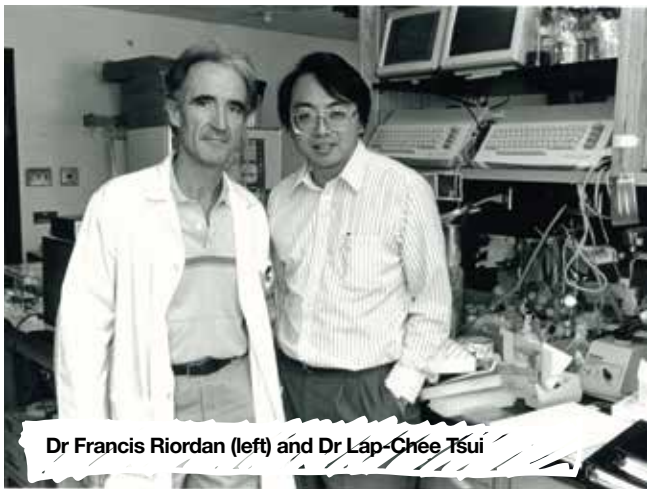
Newborn screening for CF was rolled out across the UK in 2007, following a lengthy campaign by the Cystic Fibrosis Trust, with genetic screening (PGD) available for couples who are carriers of the CF gene.

The Toronto research team

Hopes for better CF treatments

Since 1989, research into CF treatments has been split into two main areas: treatments that tackle the complications of CF, and those that target the root cause – the gene itself.

Once the gene was identified, researchers were able to understand more about the CFTR protein; how CFTR works normally, how it is faulty in CF, and how different mutations affect the protein's functionality. Ultimately, this new understanding led to new drugs – 'precision medicines' targeting specific types of damage or specific genotypes; a success that couldn't have been predicted at the time (see page 10 for more).



Dr Francis Riordan (left) and Dr Lap-Chee Tsui

Gene therapy

Within a year of the discovery of the CFTR gene researchers achieved a "proof of concept" that gene therapy was possible in the laboratory, leading many to consider it as an achievable, near-term goal.

However, in practice this proved a far greater challenge. And while developments in medicine have led to better treatments and greater life expectancy for people born with CF today, gene therapy for CF has yet to become a reality.

One of the main stumbling blocks in the development of gene therapies has been how to package up and safely deliver the new genes into the body.

In 1993, the first single-dose CF gene therapy clinical trial took place. It showed that improvements were needed in the efficiency and delivery of the new gene. Since then different types of non-toxic virus, fatty packaging known as liposomes and nanoparticles have been developed as ways of delivering gene therapies.

In 2015 the UK Gene Therapy Consortium, whose work the Trust supports, reported the results of a second gene therapy trial (a phase IIb study). The individuals who inhaled liposomes every month for a year had stabilised lung function. While these were very modest results, researchers had for the first time demonstrated the effectiveness of CF gene therapy in the human body. The study's authors acknowledged that improvements would be needed before the therapy could be rolled out to clinical care.

Building on these initial trials, a new phase of development of gene therapy was announced in 2018. Collaborating with biotech company Oxford Biomedica and the pharmaceutical company Boehringer Ingelheim, the Gene Therapy Consortium is working on the development of a non-toxic viral delivery system.

Thirty years on

So what happened to the researchers who first found the gene? Three decades on, they continue to have an impact on CF research.

Dr Tsui was instrumental in setting up a database detailing mutations found in the CFTR gene. Dr John Riordan continued to lead a CF research group, and in October 2018 researchers in his lab were one of the first groups to publish data on the very detailed structure of the CFTR protein (see page 10 for more). Meanwhile, SickKids continues to conduct research into CF with the goal of working towards improving the quality of life for patients and their families with CF through therapy discovery and implementation.

The first CF precision medicine (ivacaftor) was licenced in 2012 and approved for use in the UK in 2013, treating CFTR gene mutations carried by approximately 5% of people with cystic fibrosis. Drugs are now licenced to treat mutations affecting approximately 50% of people with the condition. Several companies are working to develop drugs that improve the effectiveness of existing precision medicines and modify the CFTR protein in different ways.

In the 30 years since its discovery, the gene has vastly broadened our understanding of CF, and has led to newborn screening, better clinical care and development of precision medicines.

All photos credited to Hospital Archives, The Hospital for Sick Children, Toronto.

How the CF gene helped shape research

Thirty years after the CFTR gene was discovered, we now know more about the protein, its function and ways to correct the effects of the damage. Along the way, CF researchers have led the way and harnessed others' breakthrough techniques to advance our knowledge. Here we explore two of these in more detail.

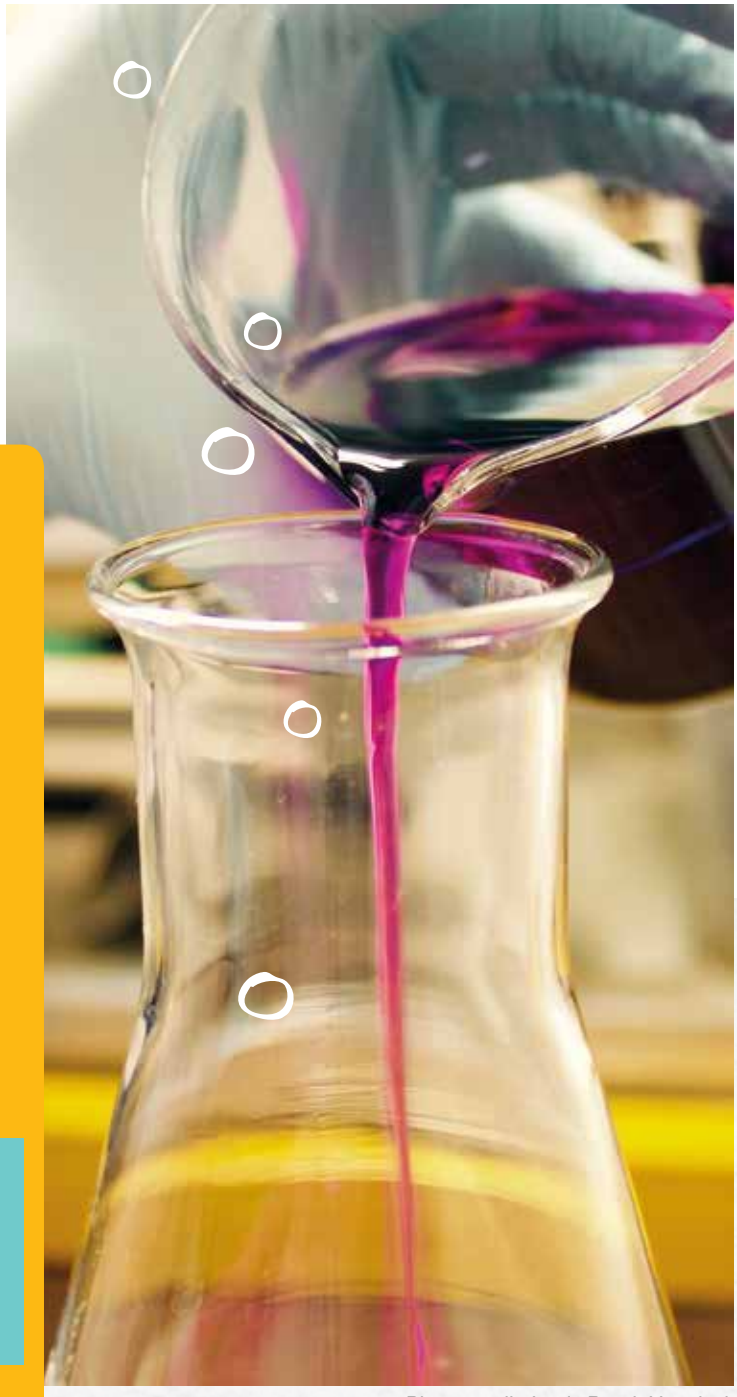


Photo credit: Louis Reed, Unsplash

CFTR drug design created a new mould

In biology, reactions take place within our bodies by one chemical fitting into a very specifically designed receptor or 'docking station' of a protein. When connected to the docking station, the protein either stops or triggers a reaction. Many drugs that we take today, whether it's paracetamol or beta-blockers, work by manipulating that docking station; increasing or decreasing its activity.

In order to correct the CFTR protein, a different approach is needed. In CF, we know that the fault lies in the shape and overall function of CFTR. CFTR modulators are designed to correct the shape of the protein by providing molecular supports (for example, if CFTR were a house then ivacaftor would be scaffolding on the outside). By supporting its shape, the protein can do its normal job again.

CFTR modulators were ground-breaking as they were among the first drugs licenced to correctly adjust the shape and function of proteins, something that the pharmaceutical industry previously didn't believe was possible.

Freezing proteins reveals new targets for drug design

Starting from a line of its building blocks, the final form of a protein is a complex 3D structure with lots of nooks and crannies, and a specific shape. In order to understand how individual changes to the building blocks alter the function of the protein, it's important to view and study the shape of the protein in exquisite detail.

Nobel Prize-winning microscope technology called cryo-electron microscopes (cryo-EMs) have been used to gather new information about how the CFTR protein works. This information can be used to design more precise drugs to correct its function.

A light microscope (the kind you might use in a school science lesson) can magnify objects by 100 or 200 times, depending on the lens on the microscope. In itself, that's 10 times more than an average camera. The latest, cutting edge cryo-EM microscopes can magnify things by one million times!

In October 2018, 12 months after the pioneers of cryo-EM won their Nobel Prize, CF researchers used this technology to gain a sequence of images of the ion channel of CFTR as it opens and closes, discovering previously unknown folds in the protein.

Cutting to the chase

The latest gene editing techniques offer hope for people with rare CF mutations, for whom current precision medicines won't work.

In this issue 30-year-old Andy Bolton, who has rare CF mutations, puts some questions to gene editing researcher Professor Patrick Harrison, of the University of Cork.

Andy Bolton

What needs to be done to get a gene editing treatment into the clinic?

"A lot of proof-of-principle experiments in animal models of CF need to be done before it can be tried in patients. The animal studies haven't even started yet, so unfortunately there's a long way to go.

"However, there are a lot of data from other diseases that have shown that gene editing is feasible, including one clinical trial of four people. We'll be able to use what they learnt to progress the CF studies faster."

Would the corrected gene take hold indefinitely or would it have to be re-introduced at intervals? Say daily or weekly treatments?

"Studies suggest gene editing could have very long-term effects, from months to years. The unknown is what proportion of cells, or what type of cells, need to be edited to keep the lungs healthy. If a treatment only edits a small proportion, then it may be that multiple treatments are required."

Is there any risk the edited genetic information could also edit another part of the DNA sequence unintentionally?

"These 'off target' effects are a concern, but techniques have improved to both detect and reduce them.

"Of greater concern is the very, very small proportion of cells which may be edited incorrectly and potentially cause tumours. This remains to the best of my knowledge a theoretical risk. However, it's a risk that it would be irresponsible to ignore."

Million-dollar question: is this a treatment that's likely to be available in my lifetime?

"There are many variants of gene editing, and some may be suitable for clinical trial sooner than others. There are three factors to be considered.

"One, we need very strong research to answer the basic science questions relating to CF gene editing over the next three to five years.

"Two, we need really good results from some of the gene editing trials for other diseases over the same period.

"Three, we need to crack the delivery problem. Gene editing medicine is physically large compared to small-molecule drugs. But I know there are lots of groups out there, highly dedicated and motivated, so I hope the answer is yes. Unless, of course, someone finds a quicker, easier treatment that works faster."

Professor Patrick Harrison

Poppy's story

19-year-old Poppy tells us about managing her CF while on her gap year and how she keeps her physical and mental health in peak condition while she's away from her CF team.

One thing I was most worried about was telling people about my cystic fibrosis. When I started my [ski] season here in Austria, I'd known the people I was sharing a room with for around two hours when I had to tell them. In my head it has always been a weakness, so I thought that other people would find out about it and then think less of me, or that I was contagious with some horrible disease. I have actually found that they usually think the opposite – that I am stronger because of it! I am now upfront about it from the beginning. A common misconception with CF is that because I don't look ill, people don't think I'm ill. They don't understand the sheer lengths I have to go to in order to keep myself well, like waking up an extra half an hour to an hour earlier in order to fit in all my medication before work.

Planning and preparation were key parts of my gap year. I have been planning it for around four years, therefore my doctors always knew it was going to happen. However, I only started thinking about the medical side of planning just before I left. Talking through my plans, where I was going and what I was doing, with my CF team was an essential part of planning my gap year and making it my best year yet. They told me what countries would be more of a concern to visit health-wise and what the health systems in the countries I am visiting are like.

In order to ensure my CF team were happy with me leaving for four months without a check-up, I made sure I did everything I could to keep in peak health before coming out on my season. I always find doing sport helps, so I make sure I do some form of exercise every day, no matter where I am. Although there is less oxygen at altitude, my breathing and coughing is actually better high up! Doing my medication every day and staying on top of my health is a priority for me out here, as I still have three months left and want to be able to complete my summer season afterwards as well. I keep in contact with my CF team at home, so I am by no means alone. So far everything is going really well health-wise and, touch wood, I will be able to keep it that way.

> "Talking through my plans,...with my CF team was an essential part of planning my gap year." – Poppy

I find physical strength helps me to manage my condition and stay healthy, but it also helps me to manage my mental health. Even when on IVs I still go to the gym and when in hospital I always pester the physios about doing exercise with me. Generally, if I'm in a bad mood or annoyed about something, whether CF-related things or other issues, a run will help me clear my head. Being in the snowy mountains does make it harder to run so I will often vent to my family or friends instead. I find talking to them helps a lot as they know me better than anyone and people out here haven't known me for all that long. I find a positive mindset goes halfway towards fixing most issues.

At the end of the day, despite all my fears, concerns and reasons not to do it, I had more reasons to go on a gap year and work abroad than not. I would have regretted not going, particularly if it had been worries about my CF holding me back. You only live once and CF will restrict a lot of things in life. Therefore, when I am well, I live the best, fullest life I can.

Visit Poppy's blog to find out more about her travels: stickingtwofingersuptocf.com.

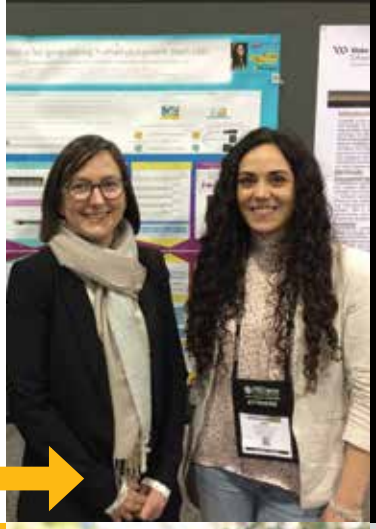
"...I had more reasons to go on a gap year and work abroad than not."

– Poppy

News in brief

Learning what matters at the NACFC

We got the low down on the world's largest CF research conference, the 'NACFC' - from Trust-funded early career researchers Vinciane Saint-Criq and Sara Cuevas Ocaña from Newcastle.



Access to Orkambi and Symkevi in Scotland

In December NHS Procurement and Vertex reached a deal allowing access to Orkambi and Symkevi in Scotland pending an appraisal resubmission of both treatments from Vertex to the Scottish Medicine Consortium.

Fundraising musicians hit the high notes

A team of musicians and adventurers raised £7,500 for the Trust and broke the world record for the 'highest classical piano performance' by taking a piano almost 5,000 metres up the Himalayan mountains for a concert.

Knowing when to talk about transplant

New data to help guide discussions on whether someone should be considered for an organ transplant were generated by Mihaela van der Schaar and her PhD student Ahmed Alaa using machine learning algorithms.

Missing?

Has it gone quiet since May? Not getting the latest news and events from the world of CF and the Trust? Don't miss out - make sure you are opted in to receive emails from the Trust at **preferences.cysticfibrosis.org.uk**.

CF nurse dedicates award to patient who nominated her

Lesley Blaikie dedicated the Nurse Award she received at the annual Scottish Health Awards in November to her patient Ashley, who nominated her and sadly died before Lesley received her award.

Your magazine, your way

CF Life is your magazine and we want to make sure it stays that way. What do you like best? What would you change?

Let us know in our survey at www.surveymonkey.co.uk/r/CFLife2019.

William Marler. Photo credit: Benedict Wilkins

The podcast sharing real voices of CF

William Marler is a man of many talents: animator, filmmaker, stand-up comedian, and the voice behind 'Straight from the Lungs', a podcast about life with cystic fibrosis.

Since the launch of the podcast last September, the response has been incredibly positive. The first four episodes delve into areas such as diagnosis, treatment and school, through Skype discussions with people of different ages and backgrounds.

"CF is obviously an incredibly individual experience," William explains, "Everyone's situation is different and that is something that needs to be represented by the individuals, not by me. So in telling other people's stories it needs to come directly from them - 'straight from their lungs'."

In the first four episodes, we meet Ben, whose CF drove him towards body-building, Martine, whose teenage daughter ran away from home after rebelling against her treatment, and Jerry, who feels his lifestyle is why he's still going strong at 61.

William feels this individuality needs to be reflected. If you Google CF, William explains, you'll get one of two things: "Either the horror stories of people losing their lives… real stories, but totally not the sort of thing for someone to read who's just had their child diagnosed. Alternatively, you might get the incredible, amazing stories - 'Nick Talbot climbs Everest'.

Illustration credit: Vicky Neville

"I wanted to include both sides. So yes, the really, really bad, and the really, really good, but also the really, really normal and not particularly incredible or extravagant. Just two people falling in love, one of whom just so happens to have CF, for example… getting across the normality of what CF can be."

For William, an important characteristic of 'Straight from the Lungs' is inclusivity; it's a dialogue, rather than a broadcast.

"You hear from a charity like the Trust or from doctors saying that you need to do this, this and this," he says, "but it's an entirely different feeling to having someone with CF talk about the importance of doing treatment. It's more involved. It's sort of saying, we're all in this together, and the aspect of it being a conversation."

William also hopes that the podcast can bridge a knowledge gap for audiences that may be unaware of the condition, such as teachers or employers of people with cystic fibrosis.

After taking a short break from the podcast, he plans to release a further 10 episodes starting this year, focusing on subjects including work, love, transplants and mental health.

"There are amazing stories and amazing ideas that I'm so excited to share."

Hear all published episodes of 'Straight from the Lungs' at lungspodcast.co.uk.

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

The future looks bright

Last Summer, Building Brighter Futures, a programme for 10–18 year olds with cystic fibrosis, held a six-week Art for Wellbeing workshop over Google Hangouts. Hear about the course from Bahar Mustafa, Digital Youth Outreach Officer at the Trust, and the young people who took part, and find out about the workshops coming up this year!

When we put out our feelers on social media to find out what kinds of workshops young people with CF wanted to see, creative writing and arts and crafts seemed very popular, and mental health and wellbeing was clearly an issue that the community cared about.

After meeting Hephzibah, the experienced art therapist who ran the workshop, we were very excited at the prospect of running a programme that would encourage young people with CF to explore creativity as a way to cope with their condition, and allow them to meet other young people like them!

"It taught me lots of new art skills whilst helping me make new friends who also have cystic fibrosis. I loved the 'hand' drawings that we would do as a warm up each week, and one of my favourite pieces of work was drawing my family tree as I was able to incorporate my family in my artwork. A big thank you to Hephzibah for teaching us, and to Bahar and everyone at the Trust for organising the course!"
- Tilly, workshop participant, whose artwork is featured on this page

Octopus

Pot of Gold

Life cycle

Mystical creature

Family tree

"It's the first time I've spoken to someone else with CF and it was good getting to know other people's points of view on having it. Also it was a chance to try doing something I wouldn't normally do. It was a great opportunity and I'm glad I did it."
- George, workshop participant

"She seems to be a bit more accepting of her condition, knowing there are other kids out there just like her." - a parent

What's coming up?

April–May 2019
Policy and Campaigning
Led by our Policy Team, you'll learn skills that will help you to build and run your own campaigns on the issues you care about.

July–August 2019
On Our Radar: CF Storify
You'll work with an external NGO, which specialises in connecting isolated communities through technology, to build and archive your own stories and create illustrations and video content.

Find out more about these courses and sign up at cysticfibrosis.org.uk/brighterfutures.

Spread the word and kick off your fundraising

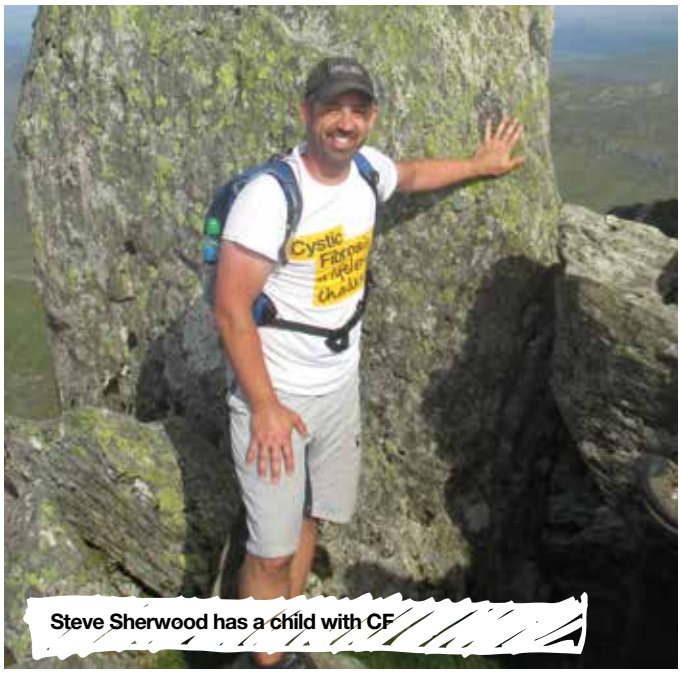
We are pleased to present a new online resource for our supporters. Read on to find out how a supporter sparked the idea for a brand new downloadable resources hub, and how you can use it to help your fundraising!

Fundraiser Steve Sherwood got in touch with the Trust early last year to request support materials to aid him with a series of school assemblies. Armed with our logo and links to a couple of Trust videos, Steve began his mission to raise awareness in his workplace.

A couple of weeks after his school assemblies, Steve reached out to share his experience with the Trust: "I used the logo and created a PowerPoint template for the assemblies, which I was lucky enough to be able to deliver to over 600 students over three days. I used the 'Oli and Nush' video as part of the presentation, which was the right length and explains CF in simple terms. The assemblies were really well received, and I was touched to hear that later in the week many teachers and students were discussing CF in their lessons as part of our moral, social and health curriculum.

"I'm sure there are many people like me who fundraise regularly and would find a standard PowerPoint template very useful. This could also include the logo in the correct format and any copyright and charity number information."

Steve also suggested that a simple downloadable leaflet about CF would be useful to print and hand out at assemblies. Inspired by supporters like Steve, we are thrilled to be launching our new pilot project, a Supporter Resources Hub to help you raise awareness about CF and the Trust in your local area.



Steve Sherwood has a child with CF

Steve took part in the Welsh 3000's Challenge to raise money for the Trust. He successfully summited all of the 15 mountains in Snowdonia that are over 3,000ft high.

Hub how-to – exploring the sections

Spread the word

Here you'll find awareness posters, a screensaver, email banner and more to help you draw attention to CF and the work of the Trust in your community, workplace or school.

Use straight away

This section includes a carefully-selected range of materials including bunting, a fundraising wall chart and a sponsorship form that you can print yourself.

Create your own

This section is home to editable PDFs - think empty-belly posters and invitations! All you need to do is personalise them with your chosen copy, save and print.

The Hub also signposts to other useful resources on our website, like how to guides for planning your own event and information and support materials.

So, what are you waiting for?

Visit the Supporter Resources Hub at **cysticfibrosis.org.uk/resourceshub** and let us know what you think!

Your feedback is vital in helping us to develop the Hub and support you with your fundraising and awareness raising. Tell us what you think by emailing **resourceshub@cysticfibrosis.org.uk**.

What's on your mind? CF and mental health

We all have mental health, good or bad, in the same way that we have physical health, and that means people with CF can experience mental health problems in the same way that people without the condition can.

However, unlike people without CF, there are a number of very specific things that people with the condition and their families can experience throughout their lives, from managing the highs and lows of the transplant process to staying on top of time-consuming treatments.

Getting to the root of the problem

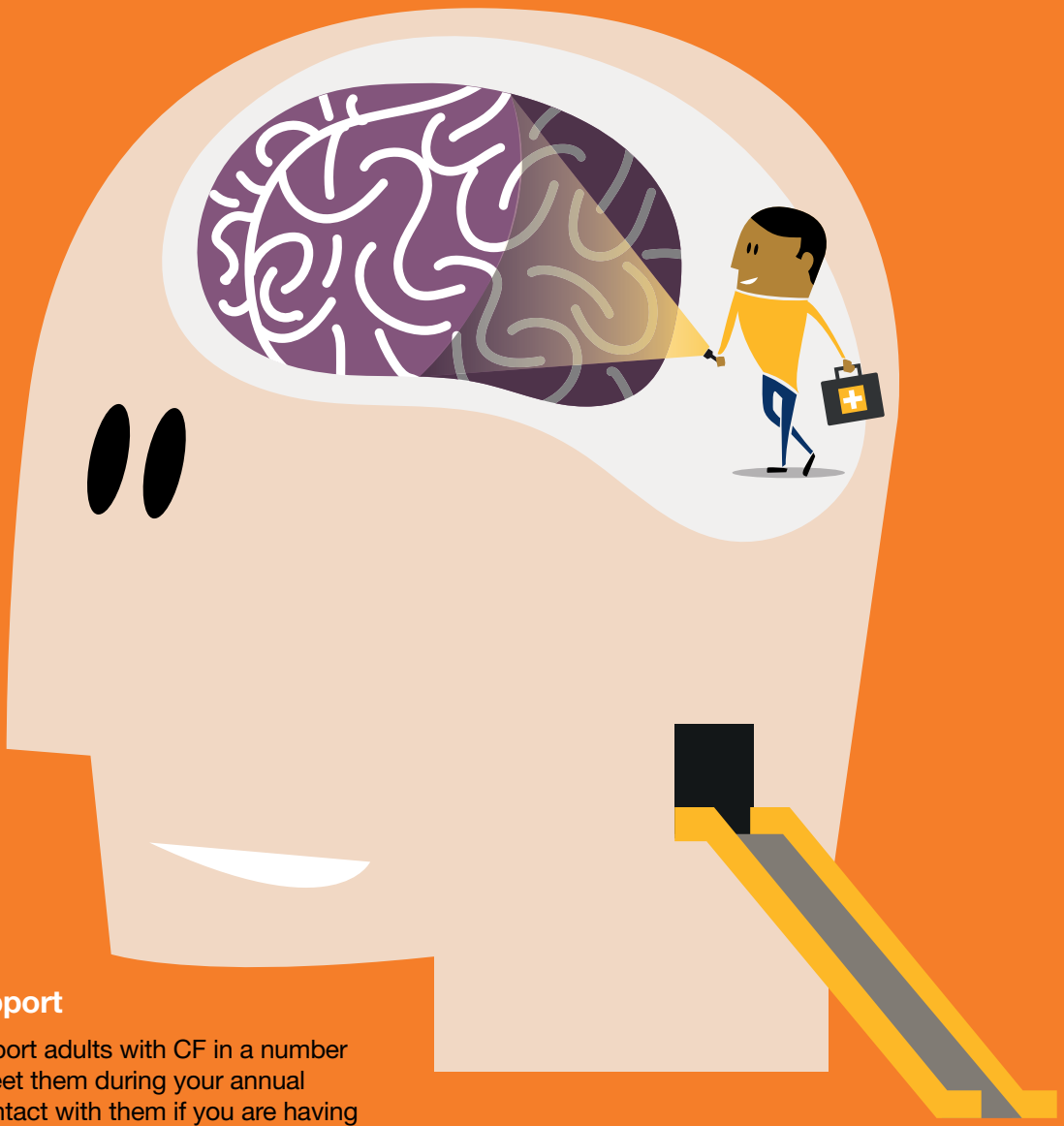
People with CF usually have many years' experience of coping with the demands of their condition, and while some people say that living with its challenges has made them emotionally resilient and helped them to develop impressive coping strategies, for others these huge challenges can feel too great.

Michael found that a drop in his physical health had a knock-on effect on his mental health when he contracted *Mycobacterium abscessus*.

He says: "Having CF never affected me mentally until I was 18 when I was diagnosed with *Mycobacterium abscessus*, which resulted in my lung function dropping from approximately 96% to around 20% in just over a year.

"I spent two years of treatment getting my lung function back above 50%, where I still have it now. I do still get depression as a result of this and the lengthy treatment that I underwent, but I try to be as positive as possible and won't ever let CF define or beat me."

Michael - @mshea1990



Searching for support

CF psychologists support adults with CF in a number of ways; you might meet them during your annual review or be put in contact with them if you are having problems with your mental health.

Adult CF psychologist Helen (see page 26 to find out more about her role) says: "Adult CF teams have their own psychologists because we know that living with CF can be a challenge to emotional health at times. Seeking help at these times is a positive action that can help prevent problems getting worse. CF psychologists can then use a variety of therapy approaches to help you manage life with CF, whether you're experiencing more common issues like low mood or anxiety, or difficulties coping with health issues or treatments."

Often, help can be available within the CF team – many CF centres now have a social worker or psychologist who will have a good understanding of the psychological impact of cystic fibrosis. If a centre does not have these staff, they might put someone with CF in contact with their local GP service, who would be able to offer psychological support.

"I do still get depression as a result of this and the lengthy treatment that I underwent, but I try to be as positive as possible and won't ever let CF define or beat me." **- Michael**

Ellie spoke to her CF team at the Bristol Royal Infirmary, who then helped her to organise counselling sessions nearer to her.

"This really helps, having connections in Bristol and in my home town. The CF team at Bristol have given me coping techniques for when I feel nervous, which has always helped me face leaving hospital. They have also always explained to my employers when I have found it hard to talk about my condition or when employers don't understand me, which triggers my anxiety. I can't thank them enough."

Ellie - @thelifeofellie.blog

Starting early

While we often think of mental health as an 'adult issue', it's also something that can affect young people with CF, though the emotional issues they experience might be very different to those of adults with the condition. One of the biggest difficulties facing children, especially when they reach school age, can be feeling different to their friends, as paediatric CF psychologist Mandy explains

"It is important that children with CF see themselves as 'a normal child who happens to have CF', rather than 'a child with CF who is trying to be normal'."

Unlike their friends, children with CF might need to take time off school for hospital admissions or take regular medication, and worries about these differences can cause them to feel embarrassed by their condition and even to hide it.

Mandy says: "One of the most common emotional issues for young children is to learn their diagnosis, understand how CF affects them and to ensure they have an appropriate perspective on living with cystic fibrosis."

In many cases, however, emotional problems might have nothing to do with CF at all. What makes children and adolescents different from adults, is that they might not always be good at saying how they are feeling, perhaps because they have never experienced these emotions before or because they are too young to put their feelings into words. Having the support of their families, as well as a psychologist or social worker as part of their routine care, can help to identify issues quickly and even prevent them.

"It is important that children with CF see themselves as 'a normal child' who happens to have CF."

- Mandy , paediatric CF psychologist

Family business

Receiving a diagnosis of CF can be an incredibly challenging time for parents, where perfectly natural fears for what the future holds and concerns for their child might arise. Soon after families receive a CF diagnosis, CF psychologists can meet with parents and, if required, other family members, to discuss concerns and offer a listening ear. They can also help parents to think about who they want to tell about their child's diagnosis and how they will manage treatments and, importantly, help them to understand the huge amount of information that they have been given about their child's diagnosis.

Cystic fibrosis psychologists have a wide knowledge of CF and will be able to inform parents about what it's like for a child growing up with CF and how to answer questions their child might have in the future, as well as any questions that other children in the family might have.

"I am so glad I now regularly see a psychologist as it has given me the tools to be able to deal with the good, the bad and the ugly times!"

- Nubi

There's no physical without the mental

Whatever your experiences are of seeking help with your mental health, it is important to remember that this help is not only there for when you feel you are in 'crisis'. Your CF team is there to help you no matter how big or small your problem is, and it is important that you treat your mental health in the same way as you would your physical health; by taking the relevant steps to keep yourself well.

Nubi said: "I am so glad I now regularly see a psychologist as it has given me the tools to be able to deal with the good, the bad and the ugly times! I always say you cannot have the physical without the mental. I would encourage anybody to seek a psychologist if they are wondering about it already. Mental health is a part of CF that often goes unnoticed, so let's do more to get it noticed."

If you have any questions or concerns around mental health, speak to your CF team or GP.

Meet... Helen Oxley

Helen Oxley

"We work with people when coping with their CF is more difficult or when their mental health is an issue, but also work on behaviour change, like improving adherence to CF treatments."

– Helen

Helen is a Consultant Clinical Psychologist at the Manchester Adult Cystic Fibrosis Centre. CF Life spoke to her to find out more about her work and the role of psychology in CF care.

Why is psychology such an important area of CF care?

"Cystic fibrosis can be a challenging condition to live with at times and for this reason all UK CF centres should have psychologists in the team to help people with CF achieve their best possible health and quality of life.

"We work with people when coping with their CF is more difficult or when their mental health is an issue, but we also work on behaviour change, like improving adherence to CF treatments.

"Everything from anxiety and depression to issues like low self-esteem, relationship problems and coping with changing health are common reasons that people might see a CF psychologist."

How do you work with people with CF day to day?

"Most days I will see several people for assessments or therapy sessions. I may see people for a small number of sessions for a straightforward problem, or for longer therapy if required, and we also like to check in with people at certain times in the CF 'journey', for example if they are newly diagnosed as adults or when they are referred for transplant, to help smooth these processes.

"I also attend CF team meetings to give a psychological point of view about a patient and work closely with the CF social workers to make sure our patients are getting all the help they need. I really enjoy my work in CF and it can be very inspiring to see the strength and resilience shown by people I work with."

How do you work with paediatric services when people with CF are moving from child to adult care?

"Our adult CF nurses and social workers meet with young people in the year or two before transfer to adult care. The CF psychologists in paediatric and adult services also liaise closely and will have joint sessions to hand over care with a patient if needed.

"Patients who have needed psychology input in paediatric care can then be offered this straight away when coming to adult care."

My first run

Natalie Crawford was always told she was too ill to exercise. Discovering at age 28 that she had CF spurred Natalie to take up running and fight for her health. As she prepares for her second London Marathon, Natalie tell us about her first tentative steps into running.

Running has become my wonder drug, my favourite treatment, my medicine of choice. Although looking back to when I first learnt to swallow the fitness pill, it sure made me gag!

My first race was probably more of an achievement than the marathon, because it was the stepping stone to bigger, better and healthier. Six years ago I couldn't even run one kilometre, and now I am pushing that finish line to 26.2 miles.

Back in June 2013, at the start line for my first 10km race, I didn't have a clue what I was doing, in all honesty! A single mother of two young children, I used running as my physiotherapy, but also the only 'me time'.

We all fear the unknown, and that was my biggest emotion as I stood there amongst runners who I thought were better than me. This new challenge. To breathe. And believe. I have since learnt that the overwhelming fear on that first race was completely normal, but also that fear can limit what you achieve.

Natalie getting into her stride

The biggest step in getting to that first race, was the want to be well. The need to stay as healthy as I could, and challenge the barriers set upon anyone diagnosed with a chronic, life-limiting illness. The greatest achievement is just to make the switch, to dedicate your all to your health.

Running can create a pathway for you, not only to clear that cement-like mucous, and help unlock that rock-solid bowel, but also to open your mind, wash away anxiety, and give you hope in the fight against cystic fibrosis.

Start small but dream big. Rest and recover, fuel and fight hard. **Cystic fibrosis is why we all fight, but also the reason we can achieve. Never let anyone tell you that your dreams are too big.**

Inspired by Natalie to give it a go? Find the right event for you at cysticfibrosis.org.uk/events.

Is awareness always good? CF in the media

"Hey, what about young people?" That (or words to that effect) was the question that the Youth Advisory Group posed to CF Life. After all, who better to represent the voice of younger people affected by CF than those living that life every day? And so, welcome to our new feature – a space for all young people with CF to talk about what they are passionate about.

Asked what they'd like to discuss first, the group was unanimous – 'Five Feet Apart', a major US film due out in the UK this month, tackling CF, cross-infection and transplant. At the time of going to press only the trailer and a very recent novelisation had been released, so we asked some young people about their hopes and fears for the film, and the depiction of CF in the media in general. This is what they had to say.

Nicole: "Cystic fibrosis has grown in popular awareness since I was diagnosed in 1997. If you told someone that you had CF back in the day, the response was, 'What's that?' Flash forward and the response is often, 'Oh my friend's cousin has that.' Slight improvement. However, CF is still misrepresented in the media."

Niki: "Over the years there have been a few films and television shows depicting a CF storyline, all of which have something in common: gross misrepresentation."

"...what really is the picture of a person with this life-threatening disease?"

Nicole: "From what I've seen on social media and in movies, CF is often associated with outdated information or a tragic death."

Chloe: "How often have you heard about someone over 30 living a healthy life with cystic fibrosis? Of course, relatively speaking, not so healthy: we will likely have annual hospital admissions and so many medications you lose count sometimes. But what really is the picture of a person with this life-threatening disease?"

"As someone who struggled with their mortality throughout my teens, this new film could unnecessarily scare people."

Liv: "This raises the question, is all awareness positive awareness?"

Cicely: "I wanted the awareness to be spread worldwide in a format many would be able to see. But the comments I have already seen have been worrying. Dubbed the next 'The Fault in Our Stars' is not something I want this film to be known for. I don't want it to be turned into #couplegoals."

Jade: "It also reminds me somewhat of a 'Grey's Anatomy' episode featuring a couple with CF that asserts that people with CF could not be loved or understood by anyone other than another CFer."

Niki: "What really struck me about the book is just how much I related to the experiences of Stella and Will [the two leads]. Hyperbole for the sake of a dramatic storyline is something to be discouraged, yet I feel that 'Five Feet Apart' does the opposite by enhancing the storyline through small and sparse dramatizations to hit home to people who know nothing about this disease just how debilitating and dangerous it can be."

> **"The film is no doubt starting a conversation"**

Ellie: "I think that the film will be great for bringing awareness to the general public about what CF is and how it is treated, because as there are only 70,000 people with CF worldwide it is not an illness many people know about."

Liv: "Something positive can always be taken from any kind of awareness. Even knowing one more person may know briefly what CF is and might entail is exciting."

Niki: "The film is no doubt starting a conversation across all segments of society; not just between CF patients but those who have only recently heard of the condition."

Ellie: "I hope that the film will be uplifting, because in reality, many CFers go on to live long, happy lives."

Niki: "The late CF vlogger and activist, Claire Wineland, worked with the cast and crew to make sure that CF was accurately depicted. I think that the film was therefore in great hands."

Chloe: "Of course, we know CF is a challenge and it's not easy, but that's also not to say that we "suffer on a daily basis" (just one of the ways someone has described my life). We all suffer from time to time, CF or not, and some more than others. But there is also so much we can do, so much to be thankful for."

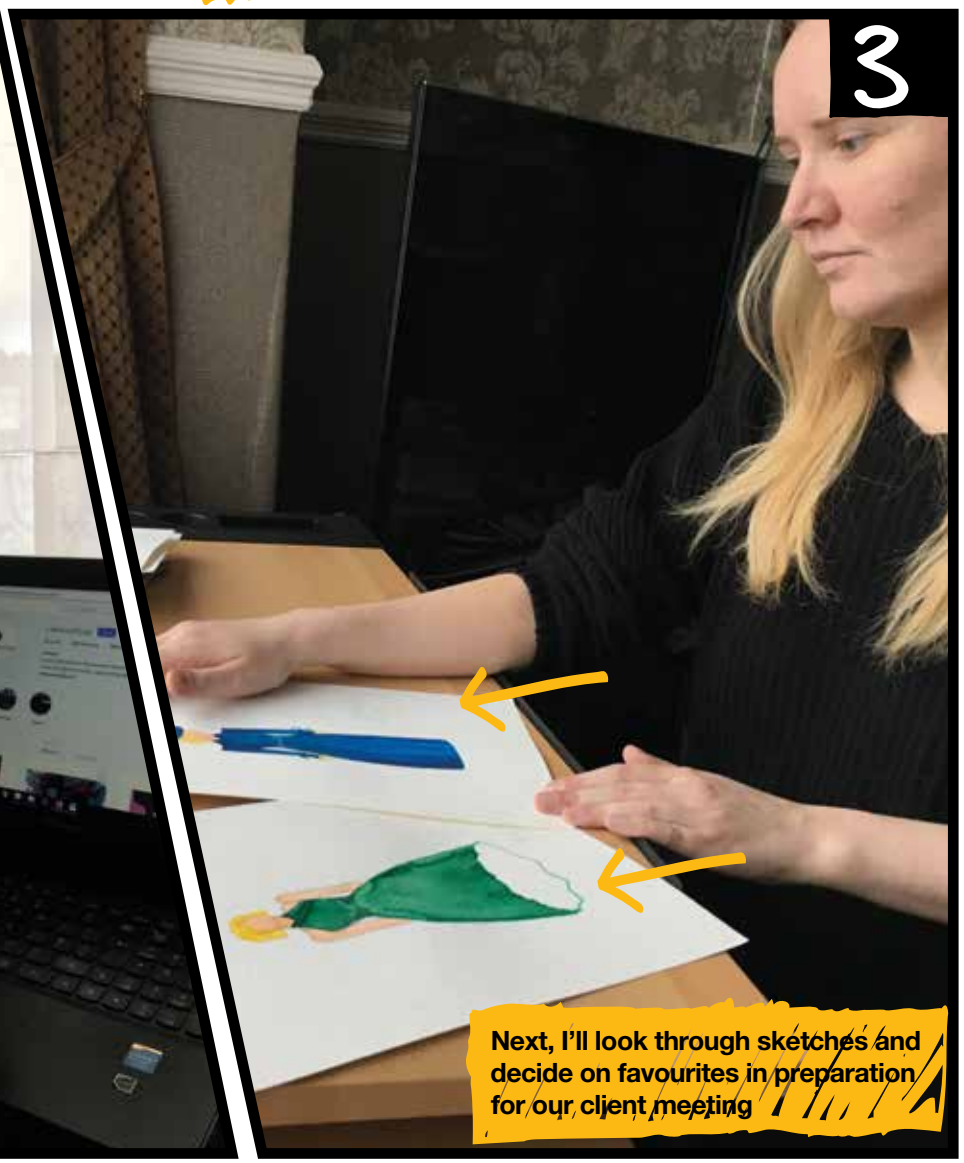
Jade: "All in all, let us be individuals, listen to our stories and appreciate our own individual achievements regardless of our disease."

If you have an idea for the next Young Voices or would like to share your views on 'Five Feet Apart', get in touch on Twitter @cftrustyouth or email Holly-Rae, our Youth Empowerment Officer hollyrae.smith@cysticfibrosis.org.uk.

Days in the life

Laura Beattie, together with sister Rachel, is the co-founder of luxury ethical British womenswear brand Careaux (who also generously supplied some of the prizes for our CF Creates competition - see p4).

Laura took time out from the couture coalface to share a typical day balancing the demands of CF and the business world.



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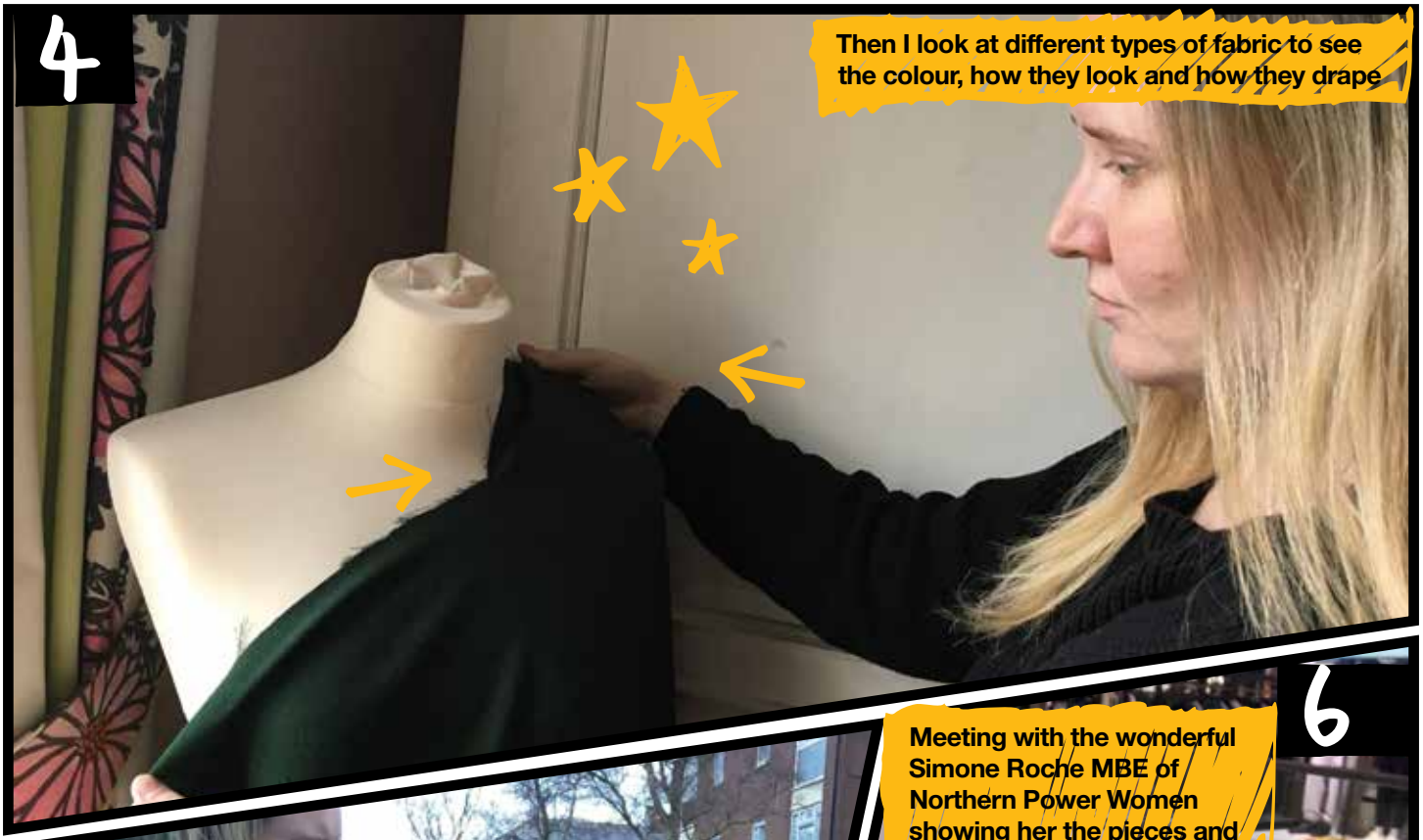


Then I look at different types of fabric to see the colour, how they look and how they drape

5

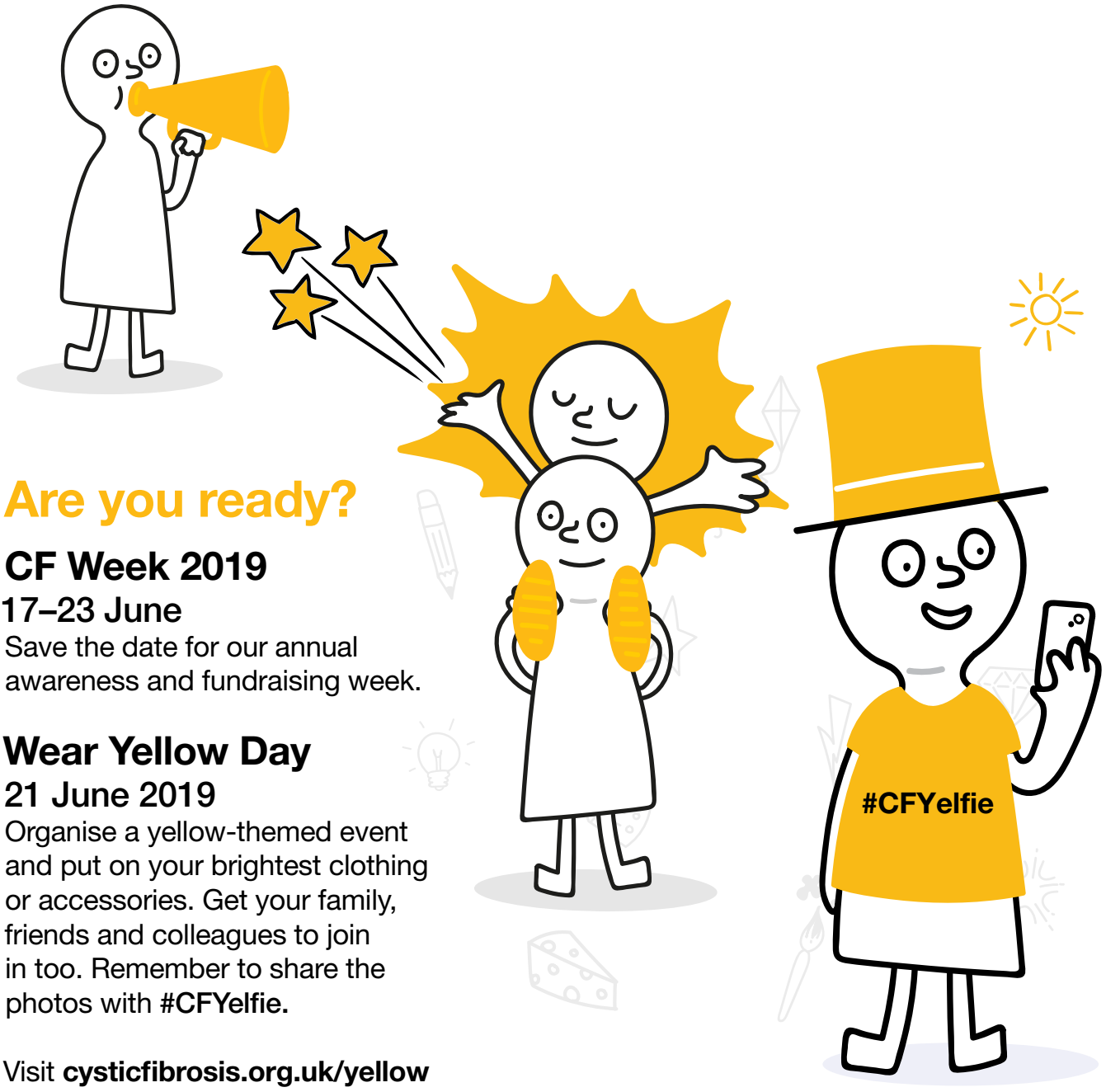
Going to meet our client!

6

Meeting with the wonderful Simone Roche MBE of Northern Power Women showing her the pieces and discussing her favourites!

Find out more about Careaux at careaux.com or follow them on Instagram @careauxofficial.

Cystic Fibrosis our focus



Are you ready?

CF Week 2019

17–23 June

Save the date for our annual awareness and fundraising week.

Wear Yellow Day

21 June 2019

Organise a yellow-themed event and put on your brightest clothing or accessories. Get your family, friends and colleagues to join in too. Remember to share the photos with **#CFYelfie.**

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

Fighting for a Life Unlimited