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CF Life

Issue 8

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Cystic Fibrosis Trust

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Issue 8 – March 2020

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FEATURES

ISSN 2513-8391

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

All magazine correspondence should be sent to:

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magazine@cysticfibrosis.org.uk

Welcome to CF Life

In this issue we hear how one young man with cystic fibrosis (CF) and his mum may have helped to change the benefits system for everyone living with the condition. Turn to page 26 to hear how Tyler and Gaynor worked with the Trust's Welfare and Rights Advisor to appeal an unfair benefits decision.

On page 6, we investigate CF-related diabetes (CFRD), a complication that one in three adults with CF is currently living with. Find out why CFRD is different from other types of diabetes and hear how researchers are turning detective to better understand the condition.

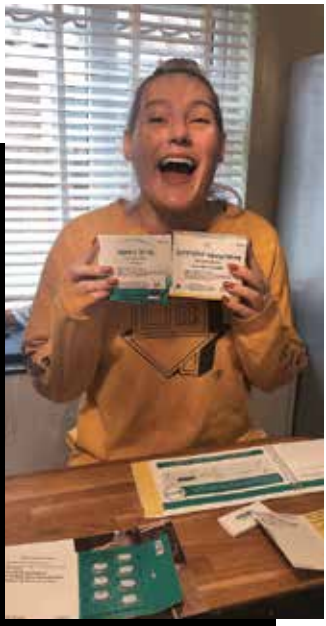
In this issue you'll also hear from Michelle, who wasn't diagnosed with CF until she was 27, and Medical Detection Dog trainer Sarah, who is teaching clever canines to sniff out Pseudomonas!

We're always on the lookout for stories to share in CF Life. So whether you're fundraising, campaigning or just have some exciting news to share, get in touch by emailing **magazine@cysticfibrosis.org.uk**.

The CF Life team

Let us know what you think of our new, smaller format by completing our survey! It won't take you more than a couple of minutes – visit **surveymonkey.co.uk/r/CFLife** to take part.

What you might have missed



Campaigning

Last year the CF community welcomed the news that all four UK nations had secured deals with Vertex for Orkambi and Symkevi, a conclusion to a battle that was sadly too long and too hard fought. The triple combination therapy (known as Trikafta in the United States) is the next drug in the pipeline and could benefit almost 90% of the UK CF population if it is made available on the NHS, the fight for which is the next step in our campaign for access to life-saving drugs. Last year Vertex submitted the drug to the European Medicines Agency (EMA).

Visit **cysticfibrosis.org.uk/lifesavingdrugs** to find out more.

Research

Even with access to these drugs, we know that people with CF are likely to still have a range of complex needs, and 10% of people with the condition in the UK won't benefit from any of these life-saving drugs. We're committed to finding the most effective ways of treating everyone with CF, and researchers around the world are working hard to develop treatments that will work for the 1 in 10 people with rare mutations. This includes other ways to make the CF protein work better, alternative ways to do the same job as the CF protein, and gene editing.

Visit **cysticfibrosis.org.uk/researchforall** to read the full article.

Support

In September, the Trust attended the British Medical Association's Patient Information Awards, where our video on getting involved with clinical trials, secondary school magazine for young people, and nutrition leaflet on leaving home and eating well with CF all received awards. Thank you to the CF professionals and people with CF who helped us create these resources.

You can take a look at all our information resources at **cysticfibrosis.org.uk/information**

Care

In November, the Frimley Park CF centre announced the addition of Dr Alexandra Ewence as their newest CF consultant. Alexandra is no stranger to the CF community, and was part of our Clinical Training Fellowship programme in 2017. She says: "Getting to know people with CF is a rare privilege within hospital-based medicine. I look forward to the opportunity to help shape CF care."

Read her full Q&A at **cysticfibrosis.org.uk/featuringourfellows**

Fundraising

Sybil Edwards, who turned 90 last year, received a British Empire Medal in recognition of a lifetime spent supporting people with CF and their families. Sybil became founding secretary and then chair of the Pembrokeshire branch of the Trust 40 years ago after meeting a family affected by CF, and has helped to raise over £140,000, making a massive contribution to people with CF across the UK.

Looking for answers to some big questions

Developing CF-related diabetes is one of the most common complications of cystic fibrosis. In fact, one in three adults with CF in the UK is currently living with the condition. In this article, we explore how research is helping to provide answers to why CFRD develops, what we know about who develops it and how it can be better managed.

Why do people get CFRD?

Insulin is an important hormone. When the level of sugar in our blood gets above a certain level, an organ in the body called the pancreas releases insulin into the blood to bring the levels back to normal again. Insulin does this by allowing sugar to move into the cells of our bodies.

In type 1 diabetes the insulin-producing cells are damaged, so not enough insulin can be produced. In type 2 diabetes the insulin-producing cells are present, but they don't make enough insulin, and the body responds less well to the insulin that is present.

CFRD is different to both type 1 and type 2 diabetes but shares some features of both. In CFRD, the body can't release enough insulin (as the cells are reduced in number or malfunctioning) and also responds differently to the insulin that is released.

Lucy, who has CFRD, says: "I know what CFRD is but I always find it hard to explain to people what it is and why I'm not a type 1 or type 2 diabetic!" It is complex for researchers too, because currently they don't know enough about what's going wrong in CFRD.

"...if we can spot the very early signs that CFRD is developing, we might be able to stop it in its tracks..."

By understanding what's happening, researchers believe that much more can be done to reduce the impact of CFRD and develop better ways to treat it – as well as providing clearer answers to how it compares to type 1 and type 2 diabetes. For example, if we can spot the very early signs that CFRD is developing, we might be able to stop it in its tracks, or even prevent it from happening at all.

To tackle these two challenges, researchers are looking at the detail of what's happening inside the pancreas. The pancreas has two jobs to do: creating the digestive juices that break down food in the intestines and producing insulin and other hormones to manage sugar levels in the blood. Both of these jobs go wrong in CF, but are these problems linked and if so, how? We're funding a group of researchers with different expertise based all across Europe as part of our CFRD Strategic Research Centre (SRC) to find out more.

Detective work

Scientists searching for answers to a big research question are like detectives trying to solve a case. Both start by following a number of lines of enquiry. Researchers working on the SRC have been pursuing three:

1. Is there any CFTR protein in the parts of the pancreas where insulin is made, that may be malfunctioning?
2. Is it the action of the CFTR in other parts of the pancreas that is preventing the production and release of insulin?
3. Is there something happening as a consequence of the damage caused to the parts of the pancreas releasing digestive enzymes?

Researchers based in Newcastle, Northern Ireland, Sweden and Hungary have been painstakingly exploring every possibility. Recently, they have made an important discovery: there is no CFTR protein in the parts of the pancreas involved in insulin release. This has eliminated the first line of enquiry from their investigations, and is a call to action to researchers in the future to look at the consequences of what's happening in the rest of the pancreas to truly understand what's causing CFRD.

"It's mainly adding more to my extensive CF medical burden."

– Tim, who has CFRD

What's the deal with diet?

Diet plays an increasingly important role in the management of all types of diabetes. However, there's very little evidence about the best way to manage diet for people with CFRD. Bristol-based research dietitian, Laura Birch, decided to investigate:

"I know that people with type 2 diabetes in particular are advised to have a 'low glycaemic index' diet, so I wondered whether a similar approach would mean that people with CFRD might be able to manage their condition more easily."

A low glycaemic index diet involves eating foods that result in a slower release of sugar into the blood, making blood sugar levels easier to manage. However, whether people with CFRD are on this diet or their usual diet, it is important that their overall calorie levels remain the same.

Laura Birch, research dietitian

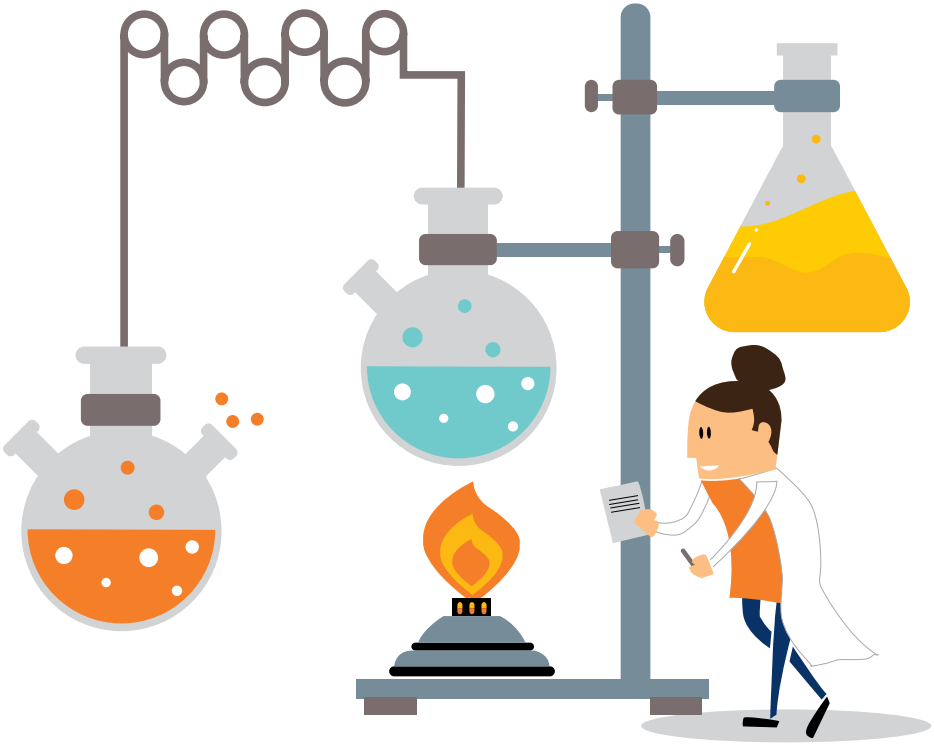
Laura explains: “There are a lot of changes that people have to think about when they have impaired glucose tolerance or are diagnosed with CFRD, so my study is looking at whether taking on information about a new diet would be too much to think about, or one too many tasks to add to their normal regime.”

Tim says of his CFRD: “It’s mainly adding more to my extensive CF medical burden. It definitely reduces my spontaneity as it does involve meticulous planning and coordination to take my blood tests and then give myself the appropriate amount of insulin. This is particularly tricky when at work or out socializing.”

The results of Laura’s study will be available in 2021. This research is funded by the National Institute for Health Research (NIHR) and a Venture and Innovation Award from the Trust.

"My study is looking at whether taking on information about a new diet would be too much to think about..."

– Laura



What about life-saving drugs and CFRD?

The fantastic news last autumn that eligible people with CF in the UK will have access to the CFTR modulators Orkambi and Symkevi will mean changes to the care of many aspects of cystic fibrosis. However, understanding what these benefits are and how life-changing they will be will take time. For example, we don't know yet what effects these drugs might have on CFRD.

The 'managed access agreement' between the NHS and Vertex Pharmaceuticals will be supported by the UK CF Registry, which will monitor the effectiveness of the drugs once people start taking them. This will include monitoring their effect on CFRD, and we hope that in a few years' time we'll know much more.

There's already a glimmer of hope that these drugs will make a difference to CFRD from a CF registry-based study on Kalydeco, the first precision medicine made available on the NHS in the UK. After three to four years of taking the drug the study found that there were fewer people with CFRD among those taking Kalydeco compared to those who weren't.

While CFRD is the most common complication that people with CF develop, there is still a long way to go to really get to grips with how to better detect and treat it, and a great deal of research is underway in the lab and the clinic, and through the use of UK CF Registry data.

We hope that in the not too distant future, the combined results of these studies will mean that those with CFRD will find the condition has less of an impact on their lives, and that far fewer people will develop CFRD in the first place.

"There's already a glimmer of hope that these drugs will make a difference to CFRD"

The challenges of CFRD

Julie Al-Siaidi, CF Specialist Dietitian at the Bristol adult CF centre, helped us develop our award-winning nutrition leaflets. Here, Julie discusses CF-related diabetes (CFRD) and the challenges of living with and treating the condition.

Julie Al-Siaidi

How do you diagnose CFRD?

Julie: Mostly people are diagnosed at their annual reviews. We also check blood glucose for all our inpatients for three days; if there are any abnormal results, we get them to test at home and if these results continue, we will diagnose them with CFRD.

How does CFRD affect the dietary advice you give to people with CF?

Julie: It's largely the same advice we give to people without diabetes; eating complex carbohydrates like bread, potatoes, and cereal, which are slower to digest and cause a slow rise in blood glucose, and cutting down on sugary drinks and sweets, which are easily digested and can cause a rapid rise in blood glucose. If someone has a low BMI we would advise having sugary drinks and sweets at meal times only.

"People with CF will be given advice from a CF Specialist Dietitan; there is not one diet for all, so if you see someone eating cake or chocolate … that's okay!"

What do you find is the biggest challenge for people with CFRD?

Julie: People don't like pricking their fingers to measure their blood glucose, but it's essential for good diabetes control. There is a new sensor called the Freestyle Libre, which stays on the arm and allows you to measure blood glucose with a reader or a smart phone.

How do you decide the best way for someone to manage their insulin?

Julie: We use blood glucose data from different points throughout the day, a detailed food chart and information about exercise, then work with the CF nurses and the endocrinologist (specialist doctor in diabetes) to decide the best insulin for the individual.

What should friends and family know about CFRD?

Julie: People with CF will be given advice from a CF Specialist Dietitian; there is not one diet for all, so if you see someone eating cake or chocolate, or having a spoon of sugar in their tea or coffee, that's okay!

Living with late diagnosis

Michelle was 27 and just a month away from getting married when she was diagnosed with CF, after a period of serious illness. Now 30, Michelle lives in Kent with husband Shaun and baby Finn, and works as a Human Resources Coordinator. Here she shares her journey from illness to diagnosis.

When I was three months old, I was admitted to hospital with pneumonia and as a child I recall having a lot of chest infections, often needing more than one week's worth of antibiotics.

I used to also suffer with stomach issues like acid reflux. But the two were never considered to be linked, and medical professionals generally put the chest issues down to asthma and the acid reflux down to diet.

People used to say I was faking it when I had time off school, or didn't really understand what was wrong, especially since I rarely looked ill. When I was in secondary school and even in my early 20s, friends would often joke about how it was always me that got ill!

When I was 18, I was taken to A&E when I suddenly had an episode of haemoptysis – coughing up blood. After a few months of this and a CT scan, I was diagnosed with Bronchiectasis, a condition where the airways become abnormally wide, leading to a build-up of mucus and causing lung infections. For years I lived with that diagnosis, seeing a respiratory consultant every six months and taking constant antibiotics.

I became very ill when I was around 26, with chest infections and episodes of haemoptysis. After several weeks I requested a referral to a specialist lung hospital in London to see if there was another treatment. Part of the investigations was a blood test to check for CF, which came back positive.

> "People used to say I was faking it when I had time off school, or didn't really understand what was wrong, especially since I rarely looked ill."

> "...I remember asking if that meant I was going to die soon"

I was shocked, I remember asking if that meant I was going to die soon and if I could still have children. I was getting married a month later, so it felt like my life was about to be turned upside down. I remember learning about CF at school, and my teacher saying it was a genetic condition and the life expectancy was late 20s.

Shaun and Michelle on their wedding day

I didn't actually think I had it for a while – I thought people were diagnosed at birth and often wouldn't reach their late 20s! I really struggled to come to terms with the diagnosis and didn't tell many people for almost a year. It wasn't until my husband ran the London Marathon for the Cystic Fibrosis Trust that I felt comfortable sharing my diagnosis and recognised it was important to raise awareness of the condition.

Shaun, Michelle's husband

Although I had been living with CF all along, suddenly the label felt life-limiting. Even applying for travel insurance was an issue and yet my health and fitness hadn't changed. When I started telling people I had CF, I remember them saying, "well it's good you now know what was going on and you're being cared for", but it was scary to think that I needed to be seen by dietitians, physiotherapists and consultants on a regular basis. To me that meant it was very serious.

Now I have to take all health concerns seriously; a cold is not just a week of Lemsips and tissues anymore, and I can't get away with not taking Creon when eating or ignoring if I am feeling chesty. My diagnosis has made me appreciate how healthy I currently am, and I don't want to take that for granted. Recently I turned 30, which for many may be daunting, however I genuinely felt lucky to have reached three decades.

Knowing I have CF has changed since becoming a mum. I really need to look after myself to ensure I am the healthiest and fittest I can be for my son, and I want to ensure he understands that an illness shouldn't always limit the life people lead.

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

Meet the dogtors

When we shared a research story about using Medical Detection Dogs to detect the bug *Pseudomonas aeruginosa* as part of the research we're funding to develop treatments for the bug, it unsurprisingly generated a lot of interest! We asked trainer Sarah from Medical Detection Dogs to answer some of your burning questions.

Jodie, Medical Detection Dog

What does the training involve?

The dogs are trained to sniff a number of stands, each with individual samples in them. When the dog detects one with Pseudomonas, it indicates (sits down). I make an audible 'click', which tells the dog it has done the right thing. They get a reward for getting it right, so they are more likely to find Pseudomonas again in the future.

Pseudomonas is everywhere, how do the dogs tell the difference?

Dogs only learn something in a certain situation, environment or format (known as 'situational learning'). The dogs sit when they smell Pseudomonas in a sample in our training room. They may recognise the same odour when out on a walk or at home, but they have not been trained to sit in that situation, so they're unlikely to perform the behaviour then.

Do other animals have very sensitive noses, or is it just dogs?

Rats, elephants, bears, sharks, snakes, moths and kiwi birds are among many animals with a highly-developed olfactory system.

How, when and where will this be used?

It's early days, but the most likely scenario is that samples would be collected in a clinic or at home and sent to us at Medical Detection Dogs for the dogs to screen, in the same way you'd send other test samples off to a laboratory.

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

Making a splash

11-year-old Edith has CF, and has been a regular water baby since she took her first dip at just eight weeks. Recently, she encouraged her swimming team to take part in a fundraiser for the Cystic Fibrosis Trust.

I joined a competitive swimming team when I was seven years old. I like being able to challenge myself physically and I have also made lots of good friends.

I have been very lucky with my health with no hospital admissions or IV antibiotics. This could be down to my swimming as I always swim at least four times a week. Even if I have a cold or cough, I still go swimming as it always makes me feel better and keeps me going.

My swimming club did a swim night fundraising event, where we sold cakes and made over £60, and challenged swimmers to swim with a straw in their mouths so they could feel what it's like to swim with bad lungs. We also challenged them to put a balloon on the top of their snorkel and try to blow it up! This was really hard. My mum did a little talk about CF and I talked about my daily routine.

As well as swimming, I do a triathlon club, weight lifting and a dance club.

Edith at her local swimming club night

Swimming is a great form of exercise for someone with cystic fibrosis. Sometimes we get asked about whether swimming pools pose an infection risk. As long as the pool is a maintained chlorinated pool it is safe for people with CF to swim in. Hot tubs, jacuzzis and non-maintained pools can pose an infection risk and should be avoided.

Cystic Fibrosis Trust

Looking for information about CF? We have what you need!

Our new catalogue lists all our information resources in one place, so you can browse and find what you need.

We have free, balanced information on all aspects of life with cystic fibrosis. Along with the medical advice from your CF team, our online content and resources can help you make informed decisions about your lifestyle, treatment and care, however you're affected by cystic fibrosis.

To order your copy, contact our Helpline on **0300 373 1000** or by emailing **helpline@cysticfibrosis.org.uk**, or visit **cysticfibrosis.org.uk/information** to download it.

Fighting for a Life Unlimited

Bringing CF-related diabetes research into the spotlight

Dr Catriona Kelly is a co-investigator on our CF-related diabetes Strategic Research Centre. Here, she talks about how she came to work in this area, how CFRD is increasingly recognised as an important research topic and how support from the Trust has shaped her career.

What is your research about?
I'm looking into the causes of CFRD on a cellular level. In my lab we're studying how the cells within the pancreas that produce insulin are affected in cystic fibrosis. We want to know how these cells develop in people with CF and how CF stops them from functioning normally.

"Five years ago, people looked at you like you were crazy when you spoke about CFRD at diabetes conferences."


Dr Catriona Kelly

From left to right: Martin Rolfe, who has CF, Catriona Kelly, Ryan Kelsey who works on the SRC, and Lorna Allen, Patient and Public Involvement Coordinator at the Trust, on a visit to Ulster University

How did you end up working in CFRD research?

I studied diabetes during my PhD, then my first research job was working with Dr Bettina Schock and Professor Stuart Elborn on CF research funded by the Trust. When I got my lectureship, I was keen to put the two parts of my research together – my knowledge of diabetes and my knowledge of CF – so I became interested in CFRD. It took a while to get any funding for CFRD, then the Trust awarded me some funding to recruit a PhD studentship in my lab. After that I got involved in the SRC.

How has being part of the SRC helped your research?

If there's something I can't work out or a technique I can't do, that's when being part of a network of researchers is really helpful. We're continuously discussing and feeding back on each other's work, which means that when you publish the results you have much more confidence in them.

What changes have you seen in CFRD research in the last 10 years?

Five years ago, people looked at you like you were crazy when you spoke about CFRD at diabetes conferences. Now it is much more accepted, both in the UK and internationally. There are nearly 20 researchers who have worked on the SRC in the lifetime of the project, so they are going out to their networks across the world and spreading the word very widely.

What are your hopes for what research can achieve for people with CFRD?

In the long term I hope that research can prevent the development of CFRD in people with cystic fibrosis. In the shorter term, better treatments for CFRD and better ways of predicting its progression are things I hope we can achieve.

To donate to support work like Catriona's, visit **cysticfibrosis.org.uk/donate**

Lending a hand

For Rebecca there's no charity more important than the Cystic Fibrosis Trust, so when her company LendInvest opened charity nominations to staff she quickly put in her application.

"My brother Tom was diagnosed with CF at five weeks old," says Rebecca. "The Trust has supported us through some of our most difficult times, so I was incredibly grateful when LendInvest announced them as their chosen charity."

A tangible difference

LendInvest's partnership supports the Cambridge Innovation Hub, our flagship research programme dedicated to improving lung health in people with cystic fibrosis.

Rebecca says: "It's important for us to support a charity where we feel we can really make a difference, and supporting the Innovation Hub definitely gave us this opportunity.

"The Trust is funding cutting-edge research so those with CF can have a 'life unlimited', and it's extremely rewarding to know that LendInvest will have been a part of this!"

The LendInvest team

An action-packed calendar

Over the last year, LendInvest has held a yellow-themed Bake Off, tackled a Tough Mudder, hosted a charity lunch and a Quiz Night, and organised a Horse Racing Night.

Rebecca says: "Everyone has been fantastic. I don't think there's a member of staff that hasn't donated or taken part in an event, which says it all!

"The Race Night was the best event. It went down well with our clients – the bidding wars highlighted just how much they wanted to support the Trust! It raised £13,000 and will definitely make it onto our annual event calendar!"

A partner you can trust

"The Trust have been a great help. They've volunteered to help at every event, filled us with fundraising ideas and educated everyone on what CF is and what the charity does."

When asked why other companies should choose the Trust as a charity partner, Rebecca says: "The Trust is a fantastic charity to support. Their work genuinely makes a difference to those who have CF and their families, something I have experienced first-hand."

A heartfelt thank you

Our partnership with LendInvest has been something special. Their expertise, knowledge and fundraising skills all combine to help us deliver a life unlimited for people with cystic fibrosis. From all of us at the Trust, and the people with CF we support – thank you!

Our corporate partners have the potential to change thousands of lives. Would you like to nominate us as your charity partner? Get in touch at company@cysticfibrosis.org.uk

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

Ready player one

22-year-old Adam Harrison is a lifelong video game fan studying for a degree in Psychology. He hit pause to talk to CF Life about games, his CF Discord server and why online gaming is a blessing for people with the condition like him.

Gaming has become more than just a hobby for me, it's a way to escape the reality of treatments and everything in-between. I've even managed to work gaming into my exercise routine thanks to new evolving technologies like virtual reality.

My deep love for gaming, and the escape it gave me, led me to want more interaction with others with cystic fibrosis. With CF the options are slim due to cross-infection, which is where Discord comes in. It's a 24/7 chatroom where users who otherwise wouldn't interact can do so daily, with instant feedback.

Our server now harbours over 400 members. Not all have CF of course, we welcome everyone as we feel as though educating others about CF is just as important. We also host a variety of other events, such as book clubs, movie clubs and gaming nights.

"Gaming has become more than just a hobby for me, it's a way to escape the reality of treatments and everything in-between."

There's a contrast between what you'd expect people to talk about during these events and what they actually talk about. It becomes less about people with CF and more about friends having fun together, with a bit of CF sprinkled on the side.

Online gaming, or any form of online entertainment really is a blessing in disguise for people with cystic fibrosis. It's an escape from the mundane routines that CF brings. I'd highly recommend someone picks up a controller or hops into the Discord because it already has changed so many people's lives.

Platform of choice?
I am fortunate enough to have all platforms available but if I really had to pick one it'd be PC.

Favourite game of all time?
Halo 3.

Worst game you've ever played?
I spent too long thinking about this, there are too many bad games.

Would you rather own every single video game on one platform or one game on every platform?
Every game on one platform, it'd be so much cheaper!

Join Adam's Discord server at discord.gg/J5mysXj (remember to use the capital letters when typing it into your address bar!) Adam also streams on Twitch at twitch.tv/BlueSensei

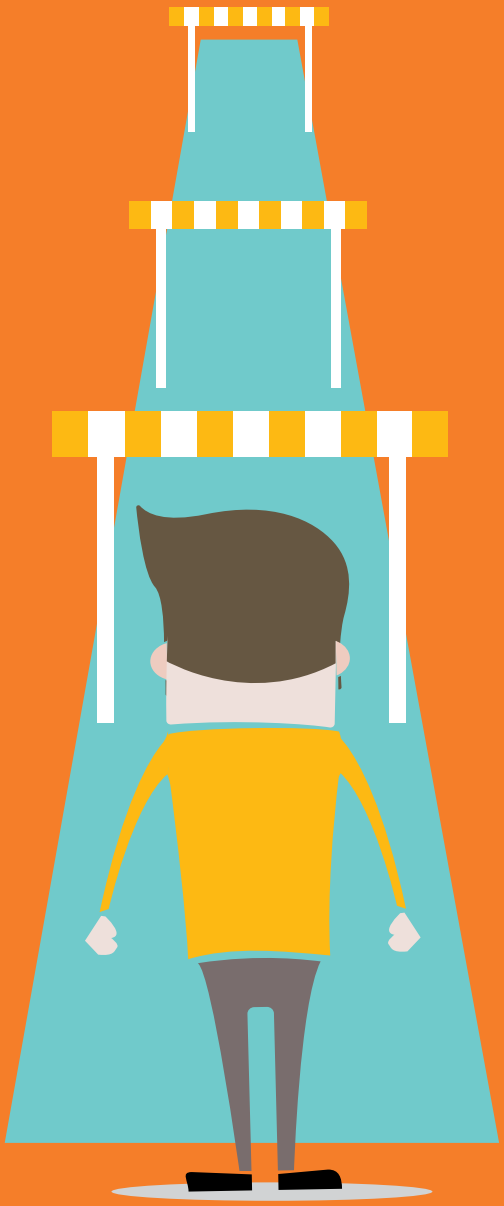
MENU START

Benefits and the burden of care

Find out how one young man and his mum's appeal against the decision on his Personal Independence Payment (PIP) claim may have changed the landscape for people with CF applying for benefits in the future.

As many people with CF and their families will know, applying for benefits when you have an 'invisible' condition can present a number of hurdles, most significantly assessors underestimating just how much of an impact it can have on your life.

Tyler is 23 and has CF, but he wasn't diagnosed until he was 10 years old, something he thinks has caused him challenges from the start.



"I think because of this I've always struggled taking meds and doing therapy," Tyler says. "I was just going into secondary school and felt conscious of being different. I relied, and still do, on family and now my girlfriend for support and encouragement."

Tyler received Disability Living Allowance (DLA) but around his 19th birthday in 2015 he was informed that he would have to apply for PIP, which would be replacing DLA. The forms used to apply for PIP were very different to those he had used for DLA, so Tyler and his mum Gaynor contacted the Cystic Fibrosis Trust for help filling them out.

When Tyler received a decision, he found he had been refused points on an area of PIP that has posed particular issues for people with cystic fibrosis. 'Activity 3,' which is covered in question five on the PIP claim form, considers the ability of the applicant to "manage therapy or monitor a health condition," and depending on the applicant they will receive between 0 and 8 points for this activity. In Tyler's case, this could mean the difference between receiving £58.70 per week or no benefit at all. Tyler received 0 points for this activity and said that the assessor felt that "as I was driving a car, I had the cognitive skills to complete these tasks without help from anyone else."

Receiving 0 points on this activity meant that Tyler didn't have enough points to qualify for PIP, so lost his benefits completely. He was supported by the Trust in asking for a mandatory reconsideration, and then an appeal, and eventually received a date to attend a 'First-tier Tribunal', which deals with appeals against benefit decisions.

"I've always struggled taking meds and doing therapy...I rely on family and my girlfriend for support and encouragement."

– Tyler

In preparation for the appeal, Tyler and Gaynor worked with Sangeeta, the Trust's Welfare and Rights Advisor. Tyler says: "She guided us through the next step and gave me an idea of what to expect at the hearing. When attending the hearing we were told that their questions weren't intended to mislead us, but I felt they did. I was asked about my education, when and how I get to work, do I go on holiday… I felt the way they asked wasn't directly related to my CF and treatments."

After the hearing Tyler was awarded four points, two for cooking, and two for managing treatments, but he needed at least eight points to receive PIP.

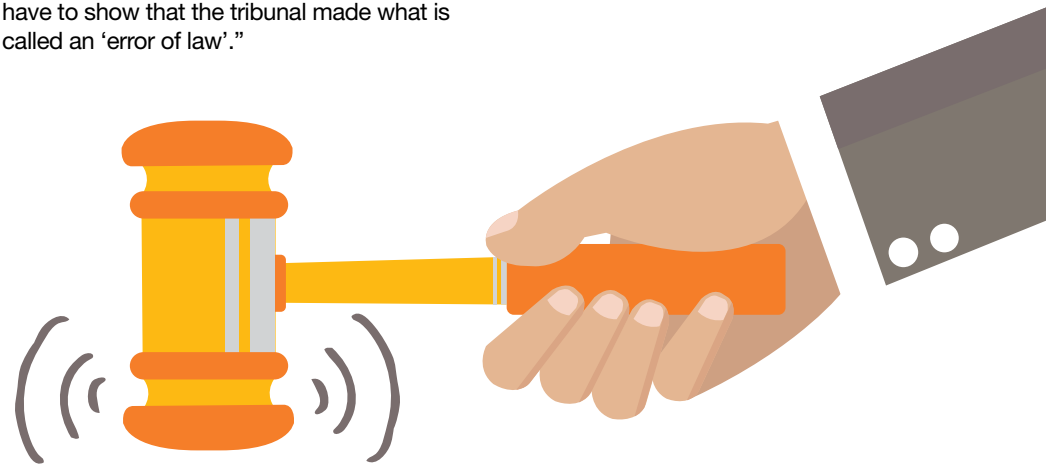
Gaynor got back in touch with Sangeeta, who investigated further and found that Tyler had grounds to appeal to the 'Upper Tribunal,' which deals with appeals against decisions made at First-tier Tribunals.

Sangeeta says: "Appeals to the Upper Tribunal have to be based on points of law. In other words, you have to say more than that you disagree with the decision, you have to show that the tribunal made what is called an 'error of law'."

> "We were told that their questions weren't intended to mislead us, but I felt they did…"
>
> **– Tyler**

Errors of law include things like the Tribunal getting the law wrong or misinterpreting it, failing to take account of relevant facts, or taking into account facts which it should not have, and not properly explaining its reasons for how it arrived at its decision.

Sangeeta believed that the Tribunal did not apply the correct legal test in relation to Activity 1, which is about cooking, and that it failed to give adequate reasons for how it arrived at conclusions in relation to Activity 2, which is about nutrition, and Activity 3.



Tyler had submitted a detailed diary of the help he needed with his therapy to the First-tier Tribunal. Sangeeta says: "I felt that the Tribunal had not shown why certain parts of Tyler's therapy should be managed by himself and why they disregarded the hour's help a day that he received from his mother."

Before the Upper Tribunal even took place, the judge working on the case decided that it was "complex in nature and had potentially wide implications," so called for an oral hearing as well to allow all parties to make their submissions more fully. Tyler was advised to get legal representation, and barrister Tom Royston, from Garden Court North Chambers and Child Poverty Action Group agreed to work on his case for free.

Just before the oral hearing, the Department for Work and Pensions (DWP) agreed there had been two or three errors of law in the decision made about Tyler's PIP claim. This meant that Tyler's appeal to the Upper Tribunal was successful, but it would now be down to the judge to decide how his PIP entitlement should be decided, as this is a decision usually made by the First-tier Tribunal.

The matter of how the DWP should interpret Activity 3 in future cases still needed to be considered by the judge, so the Upper Tribunal went ahead.

Tyler's social worker, Eileen Reynolds, who works at King's College Hospital, attended the tribunal.

Eileen says: "Everybody acquainted with CF understands how hard treatments are to adhere to, especially when feeling below par. But the DWP pursued a line of thought which said something like, 'these patients are not usually mentally impaired, therefore they can understand what is required to do treatment. They are also not physically impaired, so there is no obstacle to them carrying out treatment'."

At the Upper Tribunal, the only people to speak were the judge and two barristers: Tom representing Tyler and another barrister representing the DWP. Gaynor, members of the Trust and CF professionals from around the country watched in the public gallery.

Eileen was reassured by the judge's assessment of CF from the start: "She said she didn't see how even a healthy person could sustain such a burden of treatment consistently, which seemed promising."

"We would never have been able to do this without the help and support from Sangeeta and the Trust..."

– Gaynor

Gaynor was apprehensive about attending the Upper Tribunal, but was glad that she did: "It wasn't the daunting experience I had expected, but instead very interesting, particularly hearing the arguments from both sides."

At the end of the day, the judge asked that several further statements and counter statements be made in writing before she gave her judgement about Tyler's case and the overall question of Activity 3 and how it relates to people with cystic fibrosis.

Tracey Daniels, a CF physiotherapist at the York and Hull Adult Cystic Fibrosis Centre, was also able to attend the tribunal, and helped Sangeeta to submit a more detailed case about Activity 3 to the judge.

Tracey says: "Physiotherapists are well placed to do this as we tend to be the people who prescribe these treatments and who teach the practicalities and develop routines alongside people with cystic fibrosis. Sangeeta and I were able to clearly describe the need for support with the complex and burdensome treatment routines needed for people with cystic fibrosis."

In February this year, the judge gave her decision. Firstly, she found that the First-tier Tribunal made an error of law in relation to Activity 1. She then went on to deal with Activity 2, which relates to nutrition. Needing a 'special diet' isn't usually considered to be a reason to score points under PIP, but the judge decided that if someone needs to eat a lot repeatedly, due to a medical condition, they may need help to do so, and therefore could claim points under that activity.

She then went on to discuss Activity 3, which relates to how much help you need managing your therapies. Decision-makers often decide that this doesn't apply unless the individual has a mental or cognitive impairment that stops them from doing their treatments. The judge decided after hearing about the condition from CF specialists in this case, that people with CF are usually required to complete burdensome but essential treatments, which they may need help to complete. In her opinion, if they need this help, this should mean they should score points in Activity 3.

From left to right: Tracey Daniels, Gaynor Kelly, Eileen Reynolds, Sue Fenemore (CF social worker), Sangeeta Enright, Claire Oliver (CF social worker) and Becky Kilgariff (Head of Information, Support and Programmes at the Trust)

Martin Williams, a Welfare Rights Worker from Child Poverty Action Group, praised Tyler and Gaynor and the CF professionals that helped make this decision happen: "The presence at the hearing of our client's family members and carers, along with so many CF social workers and other experts visibly demonstrated the importance of this issue to people with cystic fibrosis and their families."

Tyler's lawyer Tom said: "It makes a real difference to the outcome of important test cases like this when expert organisations like the Trust are able to contribute their expertise."

While Tyler will have to have his own case heard again, Sangeeta is hopeful that the outcome is more likely to be positive now that these new interpretations have been made legally binding. Tyler and Gaynor's persistence in taking Tyler's case higher up the courts will also have a huge impact on people with CF applying for PIP now and in the future.

Despite these decisions, in practice it might take some time for decision-makers to apply them, and Sangeeta encourages anyone applying for PIP to speak to their CF social worker or contact the Trust's Helpline for support in making sure they present all the relevant information in their own applications for PIP.

"Unfortunately, the people that do the assessments for PIP, have very little or no knowledge of CF..."

– Gaynor

Gaynor says of their experience: "We would never have been able to do this without the help and support from Sangeeta and the Trust, throughout this whole process.

"Unfortunately, the people that do the assessments for PIP have very little or no knowledge of CF and how it impacts people's lives both physically and mentally, and not everyone is affected in the same way."

Sangeeta is hopeful for the future and encouraged by the help and perseverance of all involved.

"All it takes is a few dedicated people to care and work together, and we can make a big difference," she says. "People with CF should know they have all these people on their side."

Orkambi and Symkevi: when CF becomes big news

24-year-old Nicola lives in Scotland but receives her CF care in England, and has been a member of the Trust's Youth Advisory Group (YAG) since it began in 2016. Here, she talks about the bittersweet feeling of seeing Orkambi and Symkevi made available on the NHS across the UK late last year, while knowing the drugs won't work for her.

I left work on 24 October 2019 – the day that England finally got access to Orkambi and Symkevi – and immediately opened my inbox to floods of messages from family and friends all saying the same thing: "When are you starting it then?" Don't get me wrong, I was thrilled that England had finally been given access to these life-changing medicines and that the hard work and dedication of campaigners had finally paid off. Nevertheless, having to explain to a multitude of people that no, this medicine wasn't a cure, and that even if it were, I wouldn't be eligible for it due to my genotypes, left me feeling slightly bereft.

Nicola Torch

> **"I worry that those outside of the community are too quick to put a big red tick in the 'cure CF' box"**

Rebecca Cosgriff, Director of Data and Quality Improvement at the Cystic Fibrosis Trust, (right) and supporter Carlie Pleasant (left) appearing on BBC News. Photo: BBC News

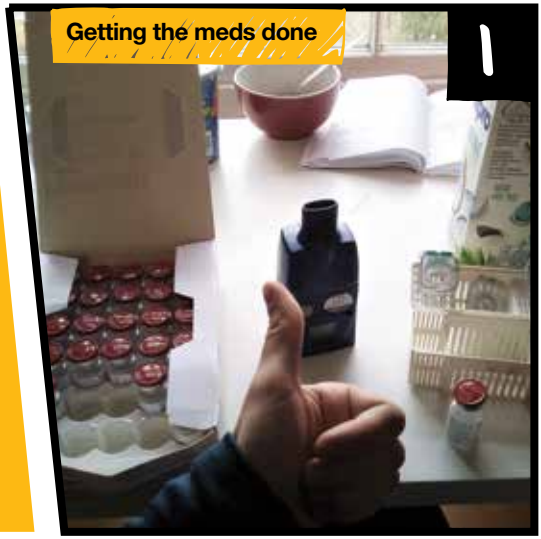
The toughest part of the media attention attached to this decision was those around me assuming that my CF would automatically be fixed. I'd take a pill and poof: my CF struggles would be gone forever. If only it were that easy! While I'm confident these drugs are giving new and incredible leases of life to so many, I worry that those outside of the community are too quick to put a big red tick in the 'cure CF' box. Assuming these drugs are the crowning glory we've all been waiting for not only diminishes the years of struggles which preceded the announcement, but also simplifies the lifelong effects CF will have on us, both physically and mentally.

Despite this, I do believe that the news has done wonders for the morale of CF patients. Despite knowing that the drugs won't personally work for me, logging onto Twitter that evening and seeing the myriad posts detailing how my friends' lives will be changed forever both inspired and comforted me. There's still a long way to go and I'm sure there'll be bumps and setbacks along the way, but for now the CF community is experiencing a period of new and exciting treatments which can only increase. But much more than that: the community finally has some hope.

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

Find out how grants from the Joseph Levy Education Fund are helping Josh to stay fit and healthy during his Philosophy degree by contributing to his food costs and sports memberships, and allowing him to stay in refurbished university accommodation.



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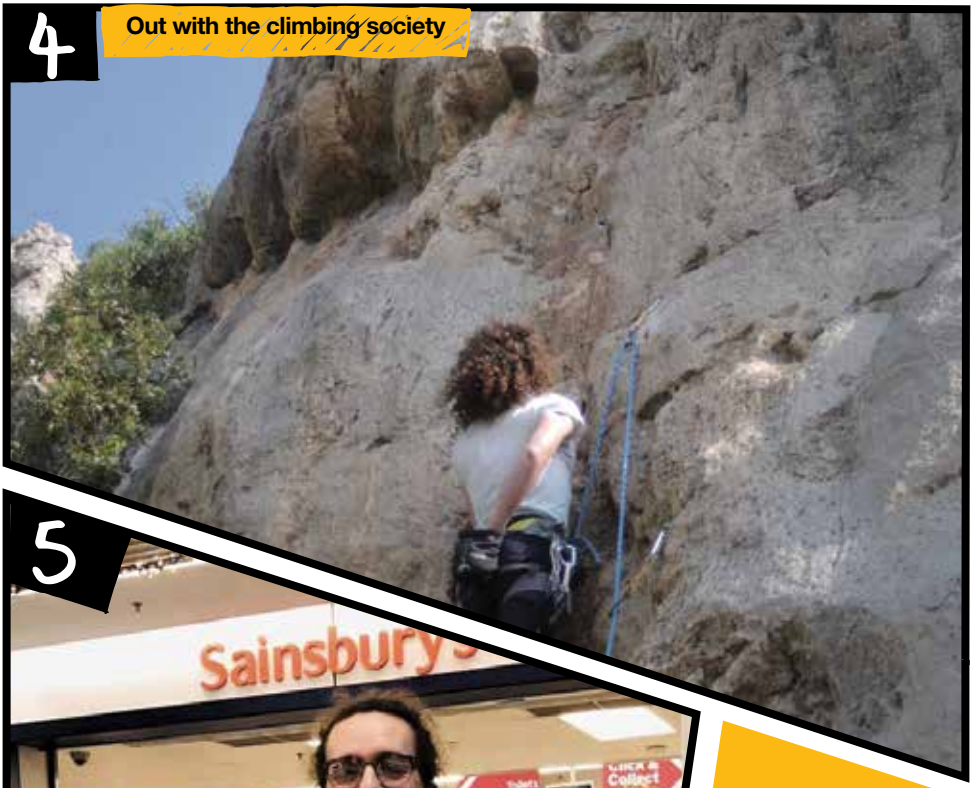

Out with the climbing society

5

Out shopping – getting those extra calories!

Find out about the Joseph Levy Education Fund and how it might be able to support you at jlef.org.uk

Cystic Fibrosis our focus

Are you ready?

CF Week 2020

15–21 June

Save the date for our annual awareness and fundraising week.

Wear Yellow Day

19 June 2020

Organise a yellow-themed event and put on your brightest clothing or accessories. Get your family, friends and colleagues to join in too and help us to raise vital funds for people with CF everywhere!

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

Fighting for a Life Unlimited