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CF Life

Issue 7

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Useful contacts

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

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T: 020 3795 2176
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Cystic Fibrosis Trust helpline
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E: helpline@cysticfibrosis.org.uk

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

All magazine correspondence should be sent to:

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Cystic Fibrosis Trust, 2nd Floor,
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magazine@cysticfibrosis.org.uk

Welcome to CF Life

You may have noticed some changes, not least the smaller size. This new format helps us to cut our production costs without reducing the number of people we can send the magazine to.

Thank you to everyone who took part in our recent survey. Readers were overwhelmingly positive about the magazine, and we are glad that it continues to be a valued way of engaging with the Trust and staying up to date with the latest developments in the world of cystic fibrosis. Your feedback has also helped to shape our new design and the articles we run.

In this issue we explore how DNA continues to point the way for breakthroughs in CF treatments and care. We also celebrate 25 years of the CF service at the Norfolk and Norwich, and the unique bond between people with CF and their clinical teams.

Elsewhere we discuss speaking out when the burden of treatment feels too much, hear a personal account of clinical trials and learn how to tell TV producers to 'keep it real' when representing cystic fibrosis.

We hope you like the new issue – let us know and share any ideas for future issues by emailing magazine@cysticfibrosis.org.uk.

The CF Life team

What you might have missed

Care

Keep an eye out for the launch of the UK-wide patient reported experience measures (PREMs) survey, where people with CF and their families will be able to anonymously share their views on the care they receive at their CF centres. The survey was developed after an extensive consultation process with people with CF and parents of people with the condition. 24 paediatric centres will start rolling out the survey this month, with adult centres following next September. **Find out more by visiting cysticfibrosis.org.uk/QualityImprovement.**

Research

We funded three new Strategic Research Centres this year, investigating the fungal infection *Aspergillus fumigatus* (which affects approximately 15% of people with CF), why Pseudomonas is able to thrive in the CF lung, and understanding lung clearance in more detail. All of this research could help to fight infections faster, create more effective treatments and reduce hospital admissions for people with cystic fibrosis.

Campaigning

This August the Scottish Medicine Consortium announced that they would not be able to recommend the routine use of Orkambi and Symkevi by NHS Scotland, citing the cost of the drugs. This news came as a huge disappointment to people with CF and their families across the UK, as it was hoped this recommendation would begin to pave the way for all UK nations to get deals done. The Scottish Government and drug manufacturer Vertex Pharmaceuticals continue to negotiate on a deal.

Fundraising

The biggest fundraising event in our calendars, CF Week, took place in June. We looked at some of the ways your support is helping people with CF and their families, and you rose to the occasion by holding over 220 events across the country, 65 more than last year.

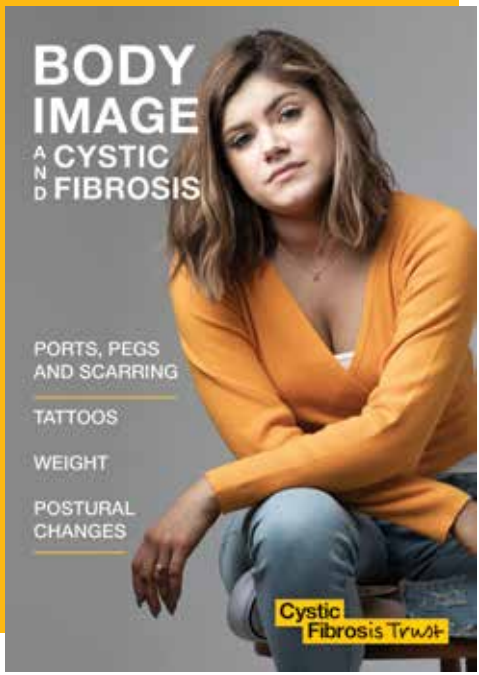
We were joined on Wear Yellow Day by longtime celebrity supporter Jenny Agutter OBE, of 'Call the Midwife' fame, 'Richard and Judy's' Richard Madeley (pictured), actors from the hit shows 'Derry Girls' and 'Sex Education', and The Doctor herself, Jodie Whittaker!

Support

This August we launched a brand-new booklet on body image, which explores some of the issues people with CF have struggled with and provides information, support and advice.

The booklet is full of pictures of people with CF and is designed to look like a magazine to challenge the accepted ideas of beauty we see every day. This free booklet can be downloaded or ordered online, or by calling our helpline on 0300 373 1000.

Visit cysticfibrosis.co.uk/bodyimage to find out more and hear from the people featured in the booklet.



What DNA can tell us about cystic fibrosis

In the age of personalised medicine, understanding the impact of genetic variations is increasingly important. Here we explore some of the genetic variations that have been found in CF so far and what needs to be done next.

Since the CFTR gene was discovered 30 years ago – a significant step forward in understanding and treating cystic fibrosis – precision medicines that modify the CF gene have been licensed, newborn screening has been implemented and we know a lot about what the 'CFTR' protein does in the body, both in the lungs and elsewhere.

As we learn more about CF, we see lots of variations in what symptoms develop and how, what bugs people grow and how people respond to treatments. The genetic make-up of someone with CF determines some of this variation, along with socioeconomic and environmental factors.

"There are many different genetic variations … things like different hair and eye colour, or how tall people are … and understanding them is at the heart of the development of personalised medicines."

Everyday genetic variations

Genetic variations affect us every day of our lives without us realising. We can see these genetic variations by comparing people in the queue at the bus stop, our school friends or work colleagues – things like different hair and eye colour, or how tall people are. There are many different genetic variations, and understanding them is at the heart of the development of personalised medicines.

Just as there are genetic variations in our physical appearance, there are lots of variations in how our bodies work; in the hundreds of chemical reactions taking place in your body as you read this. Most of the time they don't make a difference to how you go about your day-to-day life. However, every now and then you might notice things that make you different to your friends and family, and some of these are due to you having a different version of a gene.

For example, perhaps you've noticed an espresso after dinner will keep you awake much longer than it does your friends? If so, you're not alone! Around a third of people in the UK break down caffeine slower than the other two thirds, as they have a slower form of the enzyme that breaks down caffeine (you'll find out why this is important later). In some situations, a different genetic variation can be an advantage and in others it can be a disadvantage.

Variations can change how a condition develops

The known genetic variations in our biochemistry can affect how long drugs stay in our bodies, make us more or less likely to develop a particular condition or disease, or even affect the way diseases develop.

We know which genetic variation affects how caffeine breaks down because the enzyme involved also breaks down some drugs. If a particular drug is broken down by the 'caffeine enzyme', researchers need to know this when working out the safe dosage for the drug. That's because, for those that are kept awake by an evening espresso, the drug will stay in their body longer.

When it comes to genetic variations, however, the subtler the effects they have, the harder they can be for researchers to find.

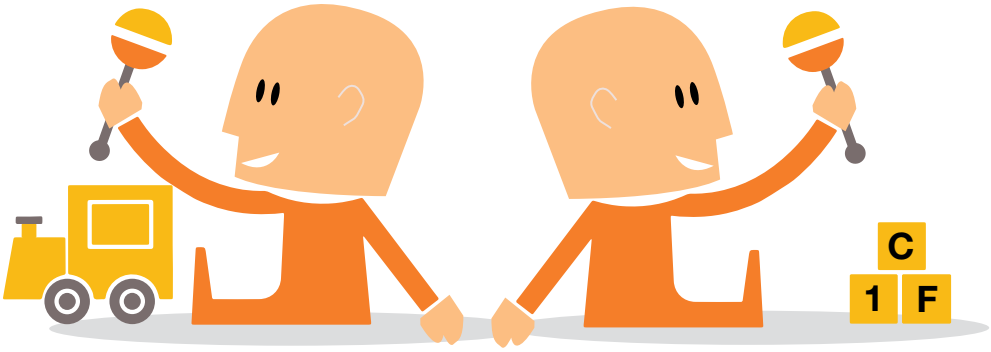
Twins to the rescue

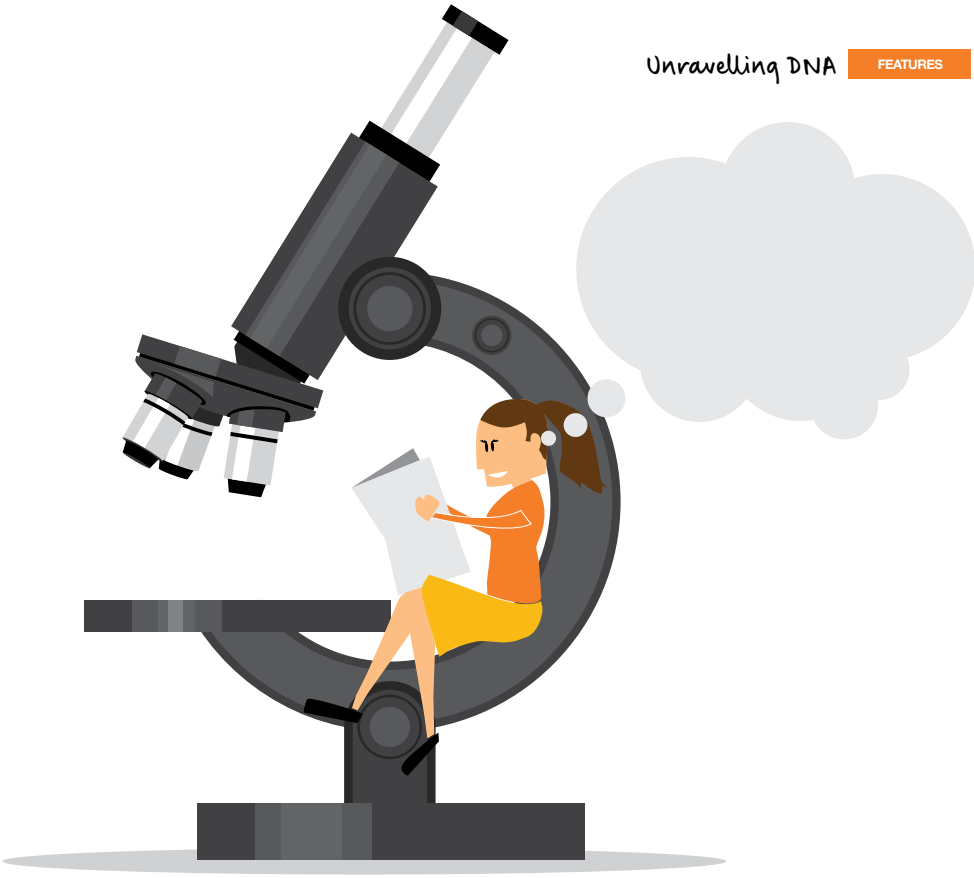
With so many different factors that could contribute to why one person has a different experience of a condition to another, it can be hard to work out whether that variation is due to someone's genetics, or whether it's due to environmental or socioeconomic factors. One way of doing this is to conduct 'twin studies'.

Twin studies work by comparing the symptoms and disease patterns of identical twins with non-identical twins. As they are likely to grow up with the same socioeconomic and environmental factors, it's much easier to spot whether these symptoms and patterns are due to genetic differences or not.

"If it is genetic differences that affect a particular symptom, you would expect the symptoms in the identical twins to match all of the time."

If it is genetic differences that affect a particular symptom, you would expect the symptoms in the identical twins to match all of the time (in the caffeine example, they would either both be kept awake by the espresso, or both wouldn't), but the symptoms in non-identical twins would only match some of the time. If there is no genetic component to a symptom, then the





chances of seeing the symptom in either set of twins should be the same.

Researchers in the USA found that identical twins with CF who also had CF-related diabetes (CFRD) would be more likely to both have the condition than non-identical twins, and so researchers concluded that there was a genetic link to whether people with CF would develop CFRD. They found that CFRD is not entirely due to genetic changes, and that other things contribute to why it develops too. Research is underway to explore the genetic links to CFRD in more detail, and to work out how to apply this new knowledge.

Learning from research into other conditions

Another logical approach to understanding more about the influence of genetic variations in CF is to learn from other conditions. Genetic variations for similar conditions may also affect how the symptoms of CF develop. For example, there are a number of genetic variations that increase a person's risk of developing type 2 diabetes in the general population. Cystic fibrosis researchers wondered whether these risk factors also increased the risk of someone developing CFRD (which has a mix of symptoms related to type 1 and type

2 diabetes). When they looked for the known diabetes genetic variations in people with CF, they found that these genetic variations altered the age of onset of CFRD. If a person with CF had the 'at risk' variations for type 2 diabetes, then they were likely to develop CFRD at a younger age.

The power of large numbers

Comparing DNA samples from thousands of people with CF to thousands of people who don't have the condition is another method for identifying genes that may modify different people's experience of cystic fibrosis. If a genetic modifier is linked to CF, then it will be found more frequently in DNA from people with CF than in people who don't have the condition. The more samples that are analysed, the less likely it is that the findings are 'false positives,' or unreliable. This type of approach is the genetic equivalent of looking for a needle in a haystack. However, new knowledge and modern technology (in terms of lab equipment and number-crunching capacity) has made this much easier than it was 20 years ago – and it is getting quicker, easier and cheaper all the time.

Filling in the genetic gaps

Genetic technology gets better and cheaper with every passing year, meaning that ever more detailed analysis is possible. The level of detail with which it is now possible to analyse our DNA means we can be more accurate than ever before

in pinpointing the DNA change that affects how people with CF have different experiences of their condition.

An important step in this type of genetic analysis has been to work out how the changes in someone's DNA can affect the symptoms of CF they experience. This can be done by collecting information about someone's symptoms at the same time as taking a DNA sample (by taking their blood). Such a link can be done anonymously and is done with the person's knowledge and consent.

"The level of detail with which it is now possible to analyse our DNA means we can be more accurate than ever before in pinpointing the DNA change that affects how people with CF have different experiences of their condition."

A more powerful way of taking this test would be to link a person's DNA sample with information from a CF registry - like the UK CF Registry - as this could help us to understand quickly if many people with the same CF genotype genetic variations experience the same symptoms in the same ways.

Our DNA still holds the answers to many questions we have about why each person's experience of CF is different. The good news is that research to unlock these answers is within our grasp. Together, the worldwide CF research community is working to piece together the complicated jigsaw of genetic variations to improve the symptoms of people with cystic fibrosis.

Helping you access clinical trials

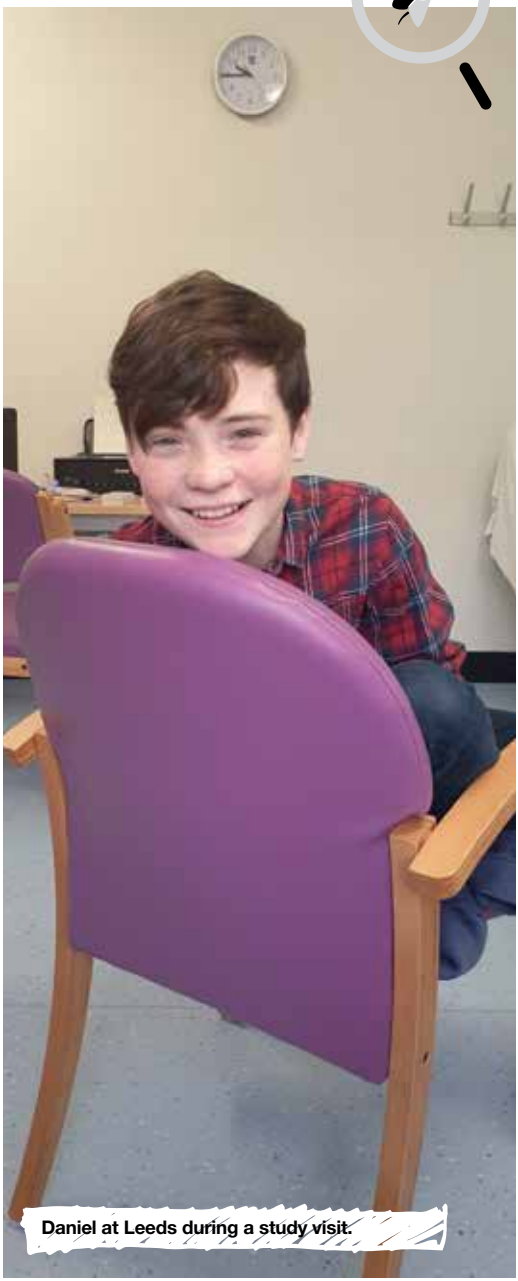
When new treatments tackling the root cause of CF emerged, Sarah used the Trust's Trials Tracker and the network created by the Clinical Trials Accelerator Platform to make sure her 13-year-old son Daniel had the chance to take part.

The first modulator therapies to emerge weren't suitable for Daniel because of his rarer genotypes, but Sarah lobbied her local MP to try and help others with CF gain access to these life-changing drugs, hoping to set a precedent for future therapies.

Sarah continued to follow developments, and when she spotted news that the next phase of a trial for a triple modulator therapy would be starting shortly, she turned to the Trust's Trials Tracker.

Sarah says: "I waited until the Phase 3 trial appeared on the list and then I contacted Daniel's care team. They referred him to Dr Tim Lee at the CF centre in Leeds, a Trials Accelerator centre where the trial was happening."

Daniel started the six-month trial in 2018: "We don't know whether he was on a placebo or the active treatment.



Daniel at Leeds during a study visit.

Sarah (centre) took on the Great North Run with Team CF.

The visits were quite frequent at first, every two weeks and then once per month, including telephone appointments."

While visits required a 120-mile round trip, Daniel's regular clinic appointments were also carried out at the study visit to save time and all their expenses were covered. If the appointments were early in the day, Daniel was able to get back to school for the afternoons!

Dr Lee adds: "As a clinical trial site appointed to the Trust's Clinical Trials Accelerator Platform, we were delighted to hear from the CF team looking after the family that they would be interested in taking part. It is fantastic to be part of a network where access to clinical trials does not depend on where people live. We have liaised closely with the team looking after Daniel - everything has worked really well."

Daniel has now moved on to the extended two-year study and he and Sarah know that he is on the active treatment. "This started in April and we have already seen the benefits of the trial treatment," says Sarah. Daniel's regular CF care continues with his original CF team.

Some CF centres, like Daniel's, do not have the budget or capacity for clinical trials, but the Trials Accelerator aims to ensure these opportunities are available to everyone. The Trials Tracker empowers people with CF and their families to seek information on trials happening around the UK that they could take part in. The Trials Accelerator funds Trial Coordinators at 32 CF centres across the UK, increasing the number of CF trials centres and extending the opportunity to participate to the wider CF community.

See what opportunities might be available to you at cysticfibrosis.org.uk/clinicaltrials

Carly's tips for parents taking on a DLA appeal

Photos are illustrative and do not depict Carly and her son.

Carly put together these tips for taking on a Disability Living Allowance (DLA) appeal after she won an appeal for her young son. She says: "I would preface everything below by saying this is only my personal experience, and I don't know how reflective this is of the general DLA process. Because the tribunal decided in our favour without a hearing, I'm not sure exactly what swung our case from being awarded no DLA to receiving high-level DLA, but I think these things must have helped!"

Start a calendar

Recording what I do to care for my child each day was the best way to start. I already tracked all of my son's medicines, feeding and treatments on an app on my phone, which made this much easier. It is so easy to forget the little things that go into daily care (the Creon doses, the extra nappies and cleaning), but keeping track makes it much easier to fill in the form and prove the additional care needs.

> "...keeping track makes it much easier to fill in the form and prove the additional care needs" – Carly

Every second counts

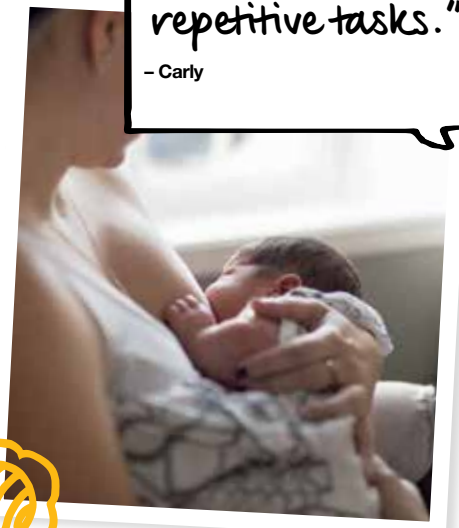
Be very clear in your calendar of the time each step takes, and don't leave out the small, repetitive tasks. It sometimes feels hardly worth mentioning the couple of extra minutes it takes during each feed or meal to give Creon, but these minutes add up throughout the day and can make the difference between receiving a low, middle or high-level threshold. It also gives you something concrete to argue with if (when!) the DLA underestimates the extra care needs. The person who assessed our mandatory reconsideration said we only needed an extra 30 minutes per day, for physiotherapy. I was able to say very clearly in my letter to the tribunal that the assessor had overlooked many other aspects of my son's care.

"...don't leave out the small, repetitive tasks."

– Carly

Explaining breast feeding and CF

Children with CF need a higher calorie intake, and for breastfeeding this translates as longer and more frequent feeding. In my letter of appeal, I said "an average baby feeds for x minutes, x times a day, but my son needs to feed for y minutes, y times a day" (a health visitor would be able to help with the average times for normal babies, depending on their age). If you are still feeding at night, highlight the extra disruption in preparing Creon for night feeds (turning on the lights to measure out Creon wakes my son up and means extra time to get him back to sleep).



Don't be afraid

It seems like a sadly common experience for parents to get rejected for DLA or awarded a lower rate than the guidelines would suggest, and going to tribunal seems to be the only way to get an independent assessment. In our case, it was an easy decision because we hadn't been awarded anything, so didn't have anything to lose! I know that some parents who are awarded a lower rate are afraid that they will have the DLA taken away altogether by another assessment, or by the tribunal. Applying for a tribunal hearing is one way you can see the assessments made by the Department of Work and Pensions. Our assessment was so erroneous that I wondered if the assessor had even read our application. It was infuriating, but also made me realise that I wasn't being unreasonable in appealing.

Be prepared to go the distance

In general, I'd advise parents to prepare for it to be a long process, and not to take a DLA rejection as any reflection on the extra care they give to their child. I was so angry and upset when I read the reports by the DLA assessors, but the tribunal decision felt like a vindication!

If in doubt – ask

I think that asking for advice is incredibly helpful - it is easy to feel isolated, especially if your CF team doesn't have a social worker for support. And while there's plenty of general advice online, having specialised support for the CF perspective was invaluable – for example from my CF team and the Welfare and Rights Advisor at the Cystic Fibrosis Trust.

For information and advice about benefits visit cysticfibrosis.org.uk/benefits or contact our helpline on helpline@cysticfibrosis.org.uk or 0300 373 1000.

Keeping it real in Casualty


Oli

Photos: J C Gellidon and Bill Oxford, Unsplash

Beginning this August, CF hit the small screen in BBC One's 'Casualty', with a storyline featuring 21-year-old Effie, who has cystic fibrosis. We spoke to Oli Lewington, who lives with CF and is Director of Marketing and Communications at the Trust, about his experience working with the 'Casualty' producers to advise on the script and promote a realistic portrayal of CF in the show.

Q: How did you advise on the script?

A: I spent time on the phone, over email and in person helping the researchers understand life with CF and the kinds of things that might make good stories. We didn't have any editorial control, but I was sent all the scripts and was able to provide feedback, some of which they listened to and some they stuck to their guns on! I tried to make sure I minimised the silly, over-the-top elements.

Q: Are there any parts of the script you were concerned about?

A: It's always hard to balance 'real life' with the needs of a TV drama, so reading some of the more far-fetched elements that were there to serve the drama was always a bit concerning. We ironed some of them out, but it's definitely not possible to fake your way onto a clinical trial!

Q: What did you like most about the script?

A: I think Effie is a really well-rounded character. She's not just a 'condition', she's an actual person facing real-life dilemmas that I felt I could relate to directly.

Q: Why is it important for people with CF to be involved in representations of the condition?

A: It helps bring the realities of the situation to life for people. You can read up a lot about CF, but when you hear it directly from someone who's been through it, it adds a different dimension.

Ebikes

When 39-year-old John needed a little help to get exercising again, advances in electronic bikes meant he was soon back in the saddle.

Nearly two years ago I had to give up work: being a fulltime Marketing Manager, and trying to take care of myself and live as long as possible for my daughter and family, wasn't really working.

After being referred to the transplant clinic, and with an extra 40 hours free a week, I bought an e-bike. I used to love cycling but as my lungs deteriorated, small hills became mountains.

I had seen grannies riding around on e-bikes but electric mountain bikes also became available and started to look 'cool' in my six-year-old daughter's eyes. Buying one is the best thing I have ever done. It's freedom, fresh air and a great way to clear my lungs.

John on his electric mountain bike.

E-bikes don't do all the work; you still need to peddle. But there are different levels of help, from eco to turbo. When I'm feeling good and I'm on the flat I turn the battery off. When I'm going up a hill, which is common around Bristol, I switch to turbo. When I'm on IVs and feeling rock bottom I can cycle with my daughter to school. It's great fun and easier than getting in the car.

Electric mountain bikes aren't cheap, but we are now a one-car family; we go out on family bike rides and physio has never felt so good.

Our Health and Wellbeing Grants can help with the cost of e-bikes and other exercise equipment. Find out more at cysticfibrosis.org.uk/grants

cysticfibrosis.org.uk

Our Christmas shop is open!

Choose from our gifts and brand new cards, including a beautiful design from last year's card competition winner Lily, and a multipack with designs from our talented runners-up.

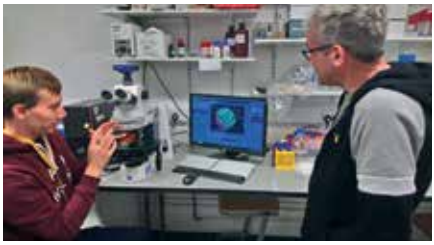
Visit cysticfibrosis.org.uk/cardshop

"My sisters and I enjoy entering competitions and really loved doing the Xmas card competition as my sister Betsy has cystic fibrosis. I chose a cow because our new house has cows in the field at the bottom of the garden!"
- Lily

Cystic Fibrosis Trust

Talking CF-related diabetes

Ryan Kelsey is a PhD student working on our CF-related diabetes (CFRD) Strategic Research Centre. He presented his work at last year's CF's Got Talent! competition, and Martin Rolfe, who has CFRD, helped him to make his presentation understandable to the CF community. We arranged for Martin to visit Ryan's lab in Ulster University, Northern Ireland, so that they could meet in person and talk about CFRD together.



Martin: Did you have an interest in diabetes before you took on your research post?

Ryan: I have always been interested in diabetes, but to be totally honest, I had a very basic understanding of CF when I took on this post, so I was keen to expand my knowledge. I am keen to remain within the area of CF research, as there's a real sense of community between researchers, healthcare professionals and people with the condition.

Ryan: How does your diabetes affect your day-to-day life in addition to managing your other CF symptoms?

Martin: Diabetes is a new time pressure on my already crowded daily treatment regimen. I need to make sure I am leaving enough time to check my blood sugars five minutes before and 1 1/2 hours after eating. Fitting this in around my other treatments is a bit of a mission.

Martin: Would you encourage other scientists to meet up with the people that their research is benefiting?

Ryan: Definitely. Whilst as a scientist I can rhyme off a lot of facts and figures regarding the condition, this doesn't explain the reality of living with CF, and speaking with Martin has given me a fresh insight.

Ryan: What is the biggest challenge of living with CFRD?

Martin: Finding reliable information to help me manage my condition, but also informing people of the differences between CFRD and other forms of diabetes. I have to reassure people that they don't need to 'help me' by reducing the amount of sugar in my coffee. CFRD, although treated like type 1 diabetes, is its own illness – even doctors and nurses at my local GP surgery don't know this!

Martin: How will your research benefit people with CF in the future?

Ryan: Formulating a treatment is an extremely costly and time-consuming process, so it is vitally important to firstly identify the area to direct potential treatments for CFRD, rather than wasting huge amounts of money and resources developing treatments that may have no effect.

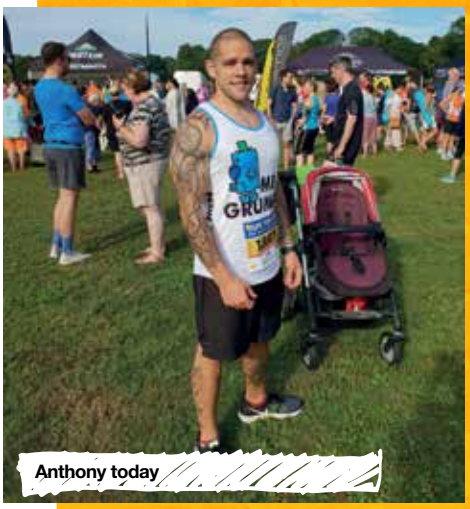
Martin: What unexpected results have come out of your work?

Ryan: In one experiment, we blocked the CFTR channel in healthy mice for eight days, and were shocked by the dramatic reduction in the size of the insulin-secreting parts of their pancreas and the reduction in the levels of insulin (the hormone which controls blood sugar levels) after such a short period of time. This implies that blocking the CFTR channel impairs the development of the insulin-secreting parts of the pancreas.

Martin taking a closer look at CFRD

Peak fitness - Anthony's story

31-year-old father of four Anthony decided to take control after years struggling with his CF and the demands of parenting saw him gain weight. Now 9½ stone lighter, Anthony is planning to climb Mount Kilimanjaro for the Cystic Fibrosis Trust.



Anthony today

Anthony before losing weight

Growing up, Anthony was in and out of hospital, and having a portacath put in at a young age meant he couldn't do contact sports. "Because of this I was never really physically fit," says Anthony.

A rocky start

At 16, weighing around 16 stone, Anthony joined a gym, but it was not a success first time around. "I didn't really have any idea what I was doing, and I fell ill. I stopped the gym after some stern words from the doctors!"

In 2012 Anthony and his wife were considering IVF when "a miracle happened" and Anthony's wife fell pregnant naturally. Anthony was able to find a good balance with family, work and health, but as his family grew, his weight crept up, and at 27 he weighed 21 stone.

A new inspiration

"I realised I wanted to run around with my children, and for them to have great memories with their dad," says Anthony. "I needed to do everything possible to get as many healthy years as I could."

In January 2016 Anthony started a programme at the gym and lost 9½ stone in 10 months.

"My lung function increased, and life didn't feel as much of a struggle. My mental attitude improved too; I was able to start seeing myself walking my daughters down the aisle."

The staff at the gym were so impressed with Anthony's efforts and how he helped others along the way, that they invited him to do a fitness instructor course and work there. Anthony is now a personal trainer too.

Showing the world

"After the weight loss and the change in career, I proved so many people wrong, but I still had people telling me I couldn't do this or that. That's what made me decide to climb Mount Kilimanjaro. I thought, it's time to show the world what someone with CF can do. If I can inspire at least one person to change their lives, I have won.

"My advice to other people with CF looking to make a change would be, make one change at a time. After a few months or even a year you will have changed your life."

If you are struggling with losing or gaining weight, take a look at our new body image booklet at cysticfibrosis.org.uk/bodyimage

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

Art with strings attached

Earlier this year, 35-year-old Colin Milne applied for a Helen Barrett Bright Ideas Award to fund his burgeoning business, 'String-a-Ling-a-Ding-Dong', where he designs chalkboards, string art and carved pieces.

Colin and his string art

Colin's bright idea was born five years ago as his daughter's first birthday approached. He says: "I really loved the personalised chalkboards I had seen online, but I wanted something more; I wanted big, I wanted hand-made and I wanted something that would last. I set myself the challenge of making a totally personalised A1 chalkboard."

'String-a-Ling-a-Ding-Dong' was born, and to date Colin has made over 200 pieces (the largest over a metre tall!) with future orders in his books "right up to 2022".

Colin in his workshop

Colin came across the Helen Barrett Bright Ideas Awards on Facebook, but considered not applying, until his health took a turn for the worse and he had a change of heart. "I had a tough time over the Christmas period and suffered pneumonia, pleurisy and a collapsed lung, which resulted in getting IVs for the

Colin's chalkboard art

first time in 17 years," he remembers. Along with issues with his eyes caused by his CF-related diabetes, Colin's health changes "brought things into perspective a bit and made me start taking my health a lot more seriously."

He says: "On the last day I decided to put a pitch together. What did I have to lose? I really didn't think I stood a chance of getting any sum of money."

Instead, Colin received £2,500 to insulate his garage, where he does the majority of his work: "In the wintertime especially, the garage is freezing cold, and I really can't spend more than an hour or so out there. If I were to insulate and heat the garage, then I'd be able to use the space year-round." While Colin still has a full-time job and hopes to continue working on 'String-a-Ling-a-Ding-Don' alongside it as long as he can, he is aware that this might not always be possible:

"If the time comes and I have to give up my work then I hope to be able to work away at my own business."

Find out about the winners of the latest round of Helen Barrett Bright Ideas Awards and see how you can apply at cysticfibrosis.org.uk/brightidea

25 years in CF care

This year sees the CF service at the Norfolk and Norwich University Hospital turn 25. We spoke to Alison Betteridge, a paediatric CF nurse who has been at the service since the very beginning, about the changes she has seen in CF care and the lives of her patients, from the introduction of blood screening to the emergence of cross-infection rules.

Alison (left) has been with the hospital's CF service since the beginning.

"Cystic fibrosis is so much more than a respiratory condition – I have been involved in helping people select pets, consider career options, and even choose prom dresses that don't show their portacaths off too much."

Like most CF professionals, in her 25 years in CF care, Alison has enjoyed a close relationship with her patients, a journey she began thanks to some early support from the Cystic Fibrosis Trust.

Before moving into CF care, Alison worked in various areas of paediatric care, and met many children with cystic fibrosis. When she started in the CF role at the Norfolk and Norwich in 1994, she had 35 patients, who were seen across the county. When this number grew to 50, the hospital's service was classed as a CF centre, and later underwent the inspection and validation process instigated by the Trust.

From small beginnings…

Back then, Alison recalls, the service had no designated funding stream. Today, it has grown to include a part-time pharmacist, a CF psychologist, a CF social worker, two consultant dieticians, a physiotherapist, a CF coordinator and a clinical fellow. This, Alison believes, is partly down to the growth of the UK CF Registry, then called the Dundee Database until the Trust began managing it in 2007.

Alison remembers consultants entering data onto the database "using floppy disks on a designated PC," but over the years she has found she spends a lot more of her day inputting data on her patients into the database herself. The value of this extra work, she says, "becomes evident when we see how this has led to transparent funding of the service and an incredible tool for research to help improve the care of both current and future patients."

The Norfolk and Norwich has been part of the regional screening programme for CF since the 1980s and Alison had the privilege to care for the first person in the world to be diagnosed with CF through blood screening.

Prior to Alison's post being created, parents with children who were suspected of having CF would have to wait a long time for a diagnosis to be confirmed. Alison says: "We have never swayed from our original standard of informing parents of a new diagnosis in a timely manner," and today she and her team do their best to keep the time from parents knowing something has been detected in their baby's newborn screening test to getting a firm diagnosis to 24 hours or less.

Alison says: "The service manager would perform the tests, and a letter would be typed and sent out to parents, who had to reply to the date to attend a clinic appointment. This could all take some time. Once I was in post I was able to telephone families to arrange sweat tests within the space of a day and often undertake these in the family's home. They could then come to the hospital the same afternoon to discuss the diagnosis with the consultant and specialist nurse."

"The technology used for treating people with CF is also unrecognisable from that of a quarter of a century ago." **- Alison**

Learning from the best

When considering the advice she would give to someone interested in a career in CF care today, Alison says: "Undertake as much training as possible but accept that parents will always be more expert than you in the day-to-day management of the condition. They are also a huge support to each other."

While, Alison notes, roles in CF care tend to be grouped together with roles in other respiratory conditions, she says that newcomers to CF care soon realise that life with CF is more than a set of symptoms. In Alison's experience, it's the little things that count: "Having the right brand of ketchup when you are in for IVs can be as important as any other aspect of the CF experience for these youngsters."

Alison at the UK CF Registry meeting 2019

"Some changes, while positive, have created unexpected challenges, the biggest being the introduction of cross-infection rules." **- Alison**

Sometimes that level of care even came at a price - Alison fondly recalls an event that took place on the CF ward during the winter.

"When a certain patient was sad that he was in hospital when it snowed, we gathered him a washing-up bowl of fresh snow as he so wanted to build a snowman. We felt pretty gullible when instead of building the said snowman in his room he made snow balls to throw at whoever came in to check his IV infusion!"

A changing landscape

A lot has changed over the years in CF care, and for Alison this includes everything from how information is recorded – "we no longer expect to get microbiology results two weeks after clinic on reams of paper to stick in a patient's notes" – to the ability of people with CF to access information.

Alison says: "I am eternally grateful to the Cystic Fibrosis Trust for their well-presented and researched literature, from a couple of sheets of A4, photocopied on low-quality paper, to professionally-produced booklets that you can happily share with parents knowing that they contain current and relevant information."

The technology used for treating people with CF is also unrecognisable from that

of a quarter of a century ago. Alison said: "Lung-function testing no longer involves thermal paper and a four-inch screen but an interactive assortment of incentives on PCs and laptops."

Some changes, while positive, have created unexpected challenges, the biggest being the introduction of cross-infection rules.

"It is often hard to wave children goodbye... but it is a joy when they see you out with their own children and recall the time they spent in our care." **- Alison**

When Alison began her position, she says, "children arrived at clinics and gathered in a play area where they could get drinks and crisps and play together with the brio train set. They had physio side by side in one area and parents had a chance to catch up with other parents that they knew well."

These vital cross-infection control rules, Alison says, have had a huge impact on face-to-face support for children and families, but "the Cystic Fibrosis Trust have looked at innovative ways to address this."

Burden of care for people with CF and their families has also increased. Alison remembers "tentatively starting a few children on DNase," a treatment that helps lung clearance, and even remembers her first patient to start taking the treatment. Today, however, Alison and her team have a selection of "10 to 12 nebulised formulations with a choice of four nebulisers, some with four different fill chambers."

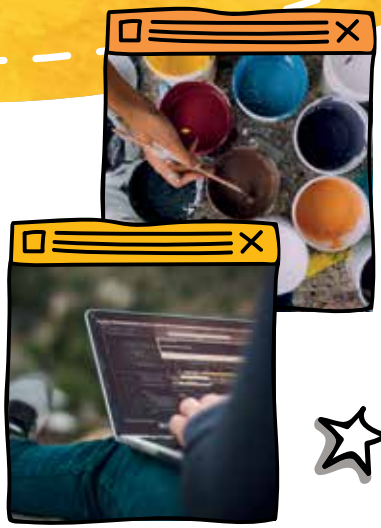
As people with CF live longer, Alison is seeing the children she cared for grow up and have their own families: "It is often hard to wave children goodbye when they graduate to care of the adult team, but it is a joy when they see you out with their own children and recall the time they spent in our care.

"I feel particularly privileged to have been a small part in the lives of many families who have children with this condition and I have learned so much from them."

cysticfibrosis.org.uk

Cystic Fibrosis Trust

Building Brighter Futures



A new programme of skill-building and creative online workshops and talks for anyone aged 10-18 living with CF is available.

Topics ranging from mental health and wellbeing to campaigning, coding and the creative arts will be covered in these interactive workshops. Everything is digital so participants can connect with other young people who have CF too!

Visit cysticfibrosis.org.uk/brighterfutures

Supported by BBC Children in Need

To comply or not comply?

19-year-old Nubi explores adherence to treatment from the perspective of a young person with CF at a crossroads in her life.

Some of my earliest memories of clinic are of consultants, physiotherapists and dieticians telling me that "I must take my medications" or I could get "very poorly". So, I took them. My parents were instructed to make sure I took them, so they made sure. I knew what I was taking my medications for, but I never really questioned 'why' or 'how', I just did as I was told.

People have said to me, "stop complaining it's only a few tablets" but it's SO much more than that. I struggled with adherence in my teenage years, particularly with taking my vitamins and doing my physiotherapy. I had the most amazing paediatric team and the most amazing parents, but I was so afraid to speak up about how I was feeling and that I was struggling with all the medication. I was lying through my teeth to not only my team and parents, but myself. I didn't want to believe I was finding this so difficult. After all, it's a few pills, right?

Transitioning to adult care was when I realised I needed to speak out about what I was struggling with. My vitamin levels were at an all-time low, my mental health was quite low and my physical health was not good. I didn't want to struggle any more or feel resentful towards my medication or my cystic fibrosis.

After speaking to my CF team, my family and some friends with CF, I realised that I was NOT alone; I was quite normal.

"I didn't want to believe I was finding this so difficult. After all it's a few pills, right?"

I think a lot of people are nervous to admit that they are having a few struggles with the demand of a typical 'CF routine', whether they're children, teenagers or adults.

But remember this: CF is a full-time job you didn't ask for. Jobs have requirements and demands, and can be stressful and time-consuming. If CF was your day job and you found something difficult, would you speak out or suffer in silence? You'd probably speak out! Do the same with cystic fibrosis.

> **"...I realised that I was NOT alone; I was quite normal."**

If you are struggling to manage your adherence and are worried about asking for advice, here are a few tips from Nubi:

1. Whether you are a parent of a child with CF, a teenager or adult, speak to your CF team about how you're feeling.
2. This sounds a bit childish maybe, but make medication charts! For children, you could make them more inviting with gold stars, stickers and rewards. For adults, it could be as basic or creative as you want, but it gets the job done!
3. Don't let it consume you. Make sure you have other means of enjoyment such as exercise, walking, cooking or listening to music!

We are all trying our best, but sometimes this can feel like it's not enough. If you ever feel like that about your CF and compliance, speaking up is the first step to getting the help you need.

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

Audrey Eade, age 49, recently went on holiday abroad and took some time out to share how she keeps organised, the importance of salt and water in hot places and owning your beach bod!

1

Double check your meds and get someone else to check them! Take a doctor's letter for airport security.

2

Familiarise yourself with the local doctor/pharmacy. Just in case!

3

Stay hydrated; drink plenty of water, take your salt tablets and have lots of fresh fruit and salad. Stick to the factor 50, especially if you are on immunosuppressants.

4

Own your beach bod - scars and all! There are all shapes and sizes on the beach.

5

Squid: choose foods that cry out to be salted!

6

Hot? Get up early and enjoy a sunrise walk for some exercise.

7

Relax, rest and enjoy. Feel smug that you never forgot anything and didn't end up at a doctor's.

Find out more about travelling and cystic fibrosis by visiting cysticfibrosis.org.uk/travel

Cystic Fibrosis worth a minute of your time

Will you plant a seed this September?

Helen Standley was the beloved Nanny to grandson George, who has cystic fibrosis, and supported the Trust during her lifetime in his honour. She is described by her family as "one of the most wonderful people on the earth. Other people came first, herself second."

Helen ensured she made provision for the family she loved, and she also included a gift to the Trust in her will, helping to plant a seed of a gift that will help us continue to grow our support for people with cystic fibrosis for years to come. We are truly grateful for her support.

Nearly a quarter of the work undertook to help people living with CF last year was made possible thanks to supporters who included gifts in their wills.

Visit cysticfibrosis.org.uk/freewills to find out more.

Fighting for a Life Unlimited