Summer 2013
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is

The magazine of the Cystic Fibrosis Trust

Trunki founder Rob Law

CF Week 2013

Transformational treatments

Cystic Fibrosis why we're here

is magazine Summer 2013

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Regular

Features

Cystic Fibrosis is why we're here

Welcome

Welcome to the new-look is magazine designed and produced in our inspiring brand style.

Our new look is vital to help us communicate our cause to raise awareness of cystic fibrosis in the wider public and to raise more funds to invest in research and the work we do to support people with the condition.

All changes take a little time to get used to but I have been hugely encouraged by the range of positive responses we have received from many quarters.

The brand is designed to help us tell more impactful, personal stories about the commitment and dedication of those with cystic fibrosis and their families, and I am delighted that we are able to provide some of these in this edition.

Rob Law is our cover feature, and his is an extraordinary story of success and achievement in the face of numerous setbacks and personal tragedy. As the man who introduced the world to Trunkies, he has, for the first time, spoken publicly about his condition and I am sure he will be an inspiration to all those whose lives are touched by cystic fibrosis.

Two others who are achieving success in their respective careers are Nicholas Richards and Ben Mudge, and I am delighted they have agreed to share their experience in two new columns looking at nutrition and exercise – both crucial elements of ensuring a healthy life for people with cystic fibrosis.

New drugs and treatments are essential for longer, better lives, and the Trust's new research strategy, published at the end of April at an international scientific conference in London, sets our plans to help ensure they are developed and delivered to clinics as soon as possible.

You can read more about it on page 4, and about the latest developments in research across the world on page 18.

This is an extraordinary time for cystic fibrosis research with transformational treatments tackling the basic genetic defect now offering the prospect of a sea-change in the way the condition is treated. The first of these drugs, Kalydeco, is already being used by those with the G551D mutation, and you will be touched by reading the wonderful story of Tilda Black whose eight-year old daughter, Maisie, began taking Kalydeco earlier this year.

Thanks to the support of the wider community we have been able to lobby the NHS in England, Scotland, Northern Ireland, and Wales to provide the funding to enable all those eligible for Kalydeco to have access to it. It's a clear demonstration that when we act and speak together in support of the interests of people with cystic fibrosis we are a powerful voice.

And it shows that the Cystic Fibrosis Trust is only as strong and effective as the support we receive from people with cystic fibrosis, their families and the wider community. We work tirelessly to ensure everything we do is bringing maximum impact and benefit to the lives of people with the condition. But we continue to need your help, and I hope you can play a part in CF Week this month to raise funds. See our feature on page 10 to find out how you can help.

There is a great deal to do and to achieve for people with cystic fibrosis, and we are making progress on many fronts. So, on behalf of all at the Trust, I want to say a big thank you to all for your continued support.

Ed Owen
Chief Executive

Share your thoughts on is magazine on Facebook **facebook.com/cftrust**, Twitter **tweet@cftrust** or visit **cysticfibrosis.org.uk/forums**

News

Research strategy launches to widespread acclaim

New focus to extend and improve the lives of people with cystic fibrosis

On 29 April 2013, the Cystic Fibrosis Trust launched a new research strategy, focused on transforming the lives of people with cystic fibrosis. The launch took place at a scientific conference held jointly with the Wellcome Trust and attended by over 150 delegates, including influential figures from the world of science, research and biotechnology.

The strategy sets out our plans for research over the next five years, and details how we will achieve its bold and ambitious aims in order to ensure maximum impact from research we fund, including:

- enhancing the involvement of patients and families with research
- part-funding five research coordinators in CF clinics
- establishing venture and innovation awards totalling £1million, with at least half going to promote transformational research projects
- creating a cadre of at least 30 young scientists through the formation of six strategic research centres, which will solve key problems in cystic fibrosis
- trebling the number of clinical trials through investment
- developing a collaborative project with a biopharmaceutical company or major non-profit to develop new therapies.

The strategy and conference have been widely acclaimed by a number of figures.

Chief Medical Officer Dame Sally Davies spoke at the reception, and praised the work done by the Cystic Fibrosis Trust in developing a strategy with a patient focus at its heart, as well as acknowledging the Trust's 'proud history' of funding medical research.

Simon Denegri, Chair of INVOLVE – the UK's national advisory group on public involvement – and NIHR National Director for Public Participation and Engagement in Research, wrote on his blog: "I have to say I am highly impressed. In terms of content and dissemination they (the Cystic Fibrosis Trust) seem to have single-handedly shown the rest of the medical research charity sector how to embrace the future as a funder and a patient group."

And Andrew Fisher, Professor of Respiratory Transplant Medicine at Newcastle University and lead of the DEVELOP-UK study to increase organ availability in the UK said he was 'very impressed' with the strategy.

You can read the new strategy at **cysticfibrosis.org.uk/researchstrategy**
Share your comments at **cysticfibrosis.org.uk/forums**

Scientific conference proceedings now online

Footage from the scientific conference held on 29 April is now available to watch online courtesy of CF Unite.

The programme included talks from top scientists and clinicians on hot topics such as gene therapy and ex-vivo lung perfusion. Talks were streamed live so that people with cystic fibrosis and their families could find out about the latest research, and put their questions to the expert speakers.

If you'd like to watch the talks visit **cfunite.org**

Top right: The Research Strategy; right: microbiologist Jo Fothergill was one of the delegates at the research strategy launch; opposite page, top right: Kalydeko; bottom: Cystic Fibrosis Trust staff proudly display the new brand at our Bromley HQ.

Campaigning success as Kalydeco approved in Wales

A drug that will benefit people with cystic fibrosis who have the G551D mutation will be funded in Wales, following a campaign led by the Cystic Fibrosis Trust to overturn an initial negative recommendation. The campaign received widespread media coverage including BBC News, ITV news and in both regional and national press.

The Welsh Government had recommended against funding of Kalydeco, despite it having been made available across the rest of the UK. However following protests from the Cystic Fibrosis Trust, families directly affected by cystic fibrosis and clinicians in Wales, the decision was reversed and Health Minister Mark Drakeford announced in May that the drug would be funded.

We are grateful for the efforts of all those who supported our Campaign for Kalydeco, and have helped make this drug available across the UK.

See **page 19** to find out about the difference Kalydeco has made to one family

Treatment for most common cystic fibrosis mutation shows promise

Data announced in April suggests a combination of Kalydeco (ivacaftor) together with another compound called VX-661 shows promise as a treatment for the most common cystic fibrosis mutation, known as F508del.

Vertex Pharmaceuticals released results from a phase 2 clinical trial of the combination therapy, in which 'statistically significant' improvements in lung function were observed over 28 days. Vertex plans to conduct additional studies of VX-661, pending regulatory discussions.

Read our feature on **page 18** to find out about more new therapies in the pipeline for cystic fibrosis that target the faulty gene

New brand to help beat cystic fibrosis

On 26 March the Cystic Fibrosis Trust unveiled a new brand and website, ushering in a new era in the fight against cystic fibrosis. The brand provides a more effective way for us to talk about our work, and explain the impact of cystic fibrosis on people's lives. In turn, this will help us raise more awareness and more funds to support our vital work in research, clinical care, support and campaigning.

You can find out more about why we rebranded on **page 8**

CF Week 2013 is almost here

Help us raise funds and awareness from 24 – 30 June

The Cystic Fibrosis Trust's national fundraising and awareness week is just around the corner, and plans are fast taking shape.

This year we are raising awareness of the work we do in the field of transplants. We're shouting about the need to increase rates of lung transplantation in people with cystic fibrosis, and the importance of better post-transplant care.

As usual, the week will be full of fun events throughout the country, including National Get Britain Bouncing Day on 27 June (part of our Big Bounce campaign) and the Big Cake Bake.

Find out how you can get involved at **cysticfibrosis.org.uk/cfweek** or read our special CF Week feature on **page 10**

First phase of Personal Independence Payment (PIP) comes into force

April 2013 saw the first phase of the introduction of the Government's new system of financial benefits to assist people with disabilities with living and mobility costs.

Currently the change from the previous Disability Living Allowance (DLA) to the new PIP system only affects people aged 16 to 64, living in certain areas. Anyone concerned can check whether they are affected by visiting **www.gov.uk/pip-checker**.

The Cystic Fibrosis Trust's website contains up-to-date information about the roll-out of PIP, including how to claim and what supporting information is needed. A guide to the assessment criteria was published on our website in May.

We are supporting the Disability Benefits Consortium (DBC)'s campaign to make sure that PIP assessments are fair and fit-for-purpose. The DBC is asking supporters to write to their MP and tell them which company will be handling assessment in their area, and to highlight a number of concerns. Visit **tinyurl.com/PIPaction** to add your voice.

More information is available at **cysticfibrosis.org.uk/pip**

Clinical conference to bring CF teams together

Event to promote best practice in new NHS environment

On 25–26 September, the Cystic Fibrosis Trust will be holding its annual conference for cystic fibrosis clinical teams. The two-day event will be held at Manchester Conference Centre.

Under the heading 'Moving forward together in a new era' the conference will bring together cystic fibrosis consultants, allied health professionals and others involved in the provision of cystic fibrosis care at specialist CF centres and clinics across the UK. The event provides a unique opportunity for networking, knowledge sharing and promoting best practice, in the context of a changing NHS, and a new era of transformational medicines targeting the basic defect in cystic fibrosis (see our feature on **page 18** for more on this).

Day 1 is a conference for clinical teams, and includes:

- plenary sessions on transformational treatments and transplantation, hot topic sessions addressing emerging issues in cystic fibrosis care;
- specialised workshops on a range of issues including adherence, fungal disease and nutrition;
- a case presentation competition.

The conference will be followed by an evening reception and dinner with inspirational speaker.

Day 2 is an invite-only forum for specialist CF centre directors and clinical leads, at which key issues around running specialist CF services, as well as wider issues that could impact CF care and treatment, are discussed and debated.

Although the conference is only open to cystic fibrosis clinicians, some of the key talks and presentations will be available for viewing online. Details will be advertised nearer the time.

If you are a cystic fibrosis clinician working in the UK, visit **cysticfibrosis.org.uk/conferences** for more information, to view the programme and to register

Famous faces kick off about cystic fibrosis

MPs claim first win against Cystic Fibrosis Trust celebrity team in the tenth Nicky's Whisper football match

MPs including Clive Betts, Jim Murphy and Ian Murray enjoyed their first win against a team featuring celebs such as Omid Djalili, Alastair Campbell, Dalton Grant, MUSE bassist Chris Wolstenholme and Daybreak sports reporter Gavin Ramjaun, in the tenth Nicky's Whisper football match in aid of the Cystic Fibrosis Trust, held at Queens Park Rangers' Loftus Road ground on 21 May.

The annual football match is held in memory of Nicky West, an inspirational young woman with cystic fibrosis and former ambassador for the Trust who did much to raise awareness of the condition. The game gained widespread media coverage, featuring in the *Daily Telegraph*, *Independent on Sunday* and *Sunday Express* newspapers, among others, and Omid Djalili was also interviewed on Talk Sport. We are delighted to reveal that the anniversary match has raised £37,000 towards our work.

Chris Wolstenholme said: "It's amazing to be involved in such a great charity. Thanks so much for asking me and well done on all the good work."

Successful telephone appeals to continue

Since the launch of the Cystic Fibrosis Trust's telephone fundraising appeals in July 2012, over 1,000 of our supporters have pledged on average more than £100 each per year to support our vital work.

Due to the success of this form of fundraising, we plan to continue our telephone fundraising appeals in 2013–2014.

We are grateful for the widespread support of this new initiative, which is an important way for us to communicate about the impact and importance of our work, and the real difference that our supporters make.

Footballer Danny Fox to help tackle cystic fibrosis

Southampton defender Danny Fox is joining forces with the Cystic Fibrosis Trust to raise awareness and funds, after the condition claimed the life of a childhood friend last year.

Danny attended the tenth Nicky's Whisper football match at Loftus Road in May, and has agreed to become a patron of the Cystic Fibrosis Trust, pledging to raise £25,000 during his first year of support. During CF Week, Danny will be visiting a specialist CF centre to meet cystic fibrosis patients and the teams looking after them.

Danny said: "I want to help raise awareness and funds to stop cystic fibrosis from cutting people off in the prime of their lives." We're delighted to have him on board.

Marathon effort

Thank you to the 220 brave runners who took part in the London Marathon for the Cystic Fibrosis Trust on 21 April and completed the 26.2 mile route through the Capital, including Simon, pictured. The Marathon is a huge boost to the vital work of the Cystic Fibrosis Trust – over £500k is expected to be raised (including Gift Aid) from this year alone. We are hugely grateful to all our runners, and the fantastic cheering crews supporting them every step of the way.

Inspired to take part in the 2014 London Marathon? Find out how at **cysticfibrosis.org.uk/vlm**

Opposite page: Big Cake Bake; this page, top: Nicky's Whisper; right: Southampton FC defender Danny Fox is supporting the Cystic Fibrosis Trust; far right: Simon running in the London Marathon.

Who we are

In March we launched our new brand and website, revealing a bold new look as we approach our 50th anniversary year

The rebrand is vital to help us stand out in an increasingly crowded charity marketplace, to raise more awareness and more funds, writes Director of Marketing Tamsyn Clark.

We created our new brand to help us beat cystic fibrosis. It's more than simply a new look, it changes the way we talk about cystic fibrosis, what we do and how we do it. We want to be a more dynamic, action-oriented organisation, and we hope you agree these steps will help achieve that.

Like many charities, we've struggled in recent years to maintain our income. We have a very loyal and committed supporter base, but between 2009 and 2011 our income fell by more than £500,000. We need to better explain the seriousness and impact of cystic fibrosis to the wider public. This will help us attract new audiences and better engage with our current supporters.

"We want to be a more dynamic, action-oriented organisation"

Research involving our community and key stakeholders showed that supporters felt awareness and understanding of cystic fibrosis among the general public was very low. For us to progress as an organisation, our brand must enhance the public's understanding, so we can engage their support and raise more money for the fight against cystic fibrosis.

How our brand works

Our name is staying. Rather than simply creating a new logo, we have developed a more effective way of communicating the impact of cystic fibrosis, and what we are doing to beat it.

Our brand can be used in various ways depending on our audience. Using the 'is' at the end of 'cystic fibrosis' we've developed a set of key phrases to communicate different aspects of cystic fibrosis. Our phrases include:

- **Cystic fibros*is* beatable**
- **Cystic fibros*is* a lifelong challenge**
- **Cystic fibros*is* crying out for a cure**

These messages can work across our different channels, such as on our website, and in fundraising and campaigning materials. They can also be used by people affected by cystic fibrosis to convey its impact on their lives.

We believe our new brand will help deliver more income for the charity alongside our improved fundraising strategy. Other charities have proven that a well-articulated brand can produce greater fundraising results and in a competitive charity market, we believe investment is vital to continue making a difference for people living with cystic fibrosis.

The feedback so far

We've received lots of feedback on our new brand and website. We're delighted that so many of you love our new look – and those that don't, we equally value your opinions. Your comments will help us improve the brand as we roll it out over the coming weeks and months, so keep them coming on our web forums and social media.

With our new brand, we can shout even louder about what cystic fibrosis 'is'. This will help us raise more funds to achieve even greater impact on the lives of everyone living with cystic fibrosis. We look forward to working with all our supporters and stakeholders to maximise the opportunities our new brand presents. ■

What is cystic fibrosis to you?
During our brand launch we ran a Facebook competition for you to submit your own IS statements. We loved reading your entries; congratulations to winner Ellis who got the most likes for his statement, and has had his IS statement made into his very own t-shirt!

Find out more about the rebrand at **cysticfibrosis.org.uk/brandfaqs**

Cystic Fibrosis why we're connecting

245,000

posts and counting on our online forum

To reduce the risk of cross-infection, people with cystic fibrosis must avoid meeting each other face to face.

The Cystic Fibrosis Trust online forum gives them the chance to meet others online, discuss the latest treatments and research or simply share their experiences.

Join the conversation at cysticfibrosis.org.uk/forum

Are you ready for

Our national fundraising and awareness week takes place from Monday 24 – Sunday 30 June. Can you help us shout louder about cystic fibrosis and make it a huge success?

The week will be packed with great events across the UK. Whether you take part in an existing event or set-up your own, get involved and help us highlight this year's theme, transplantation.

Here are some ideas to get you started:

Get bouncing!
Thursday 27 June is National Get Britain Bouncing Day! Order your Big Bouncing Kit now, including:

- a guide to Big Bounce
- a poster
- stickers
- sponsorship forms.

For every £15 you raise we'll give you a beach ball, or for every £30 you'll get a free space hopper!

If you have a child with cystic fibrosis, you can get a discount on a trampoline through our corporate supporter Supertramp.

Find out more at **cysticfibrosis.org.uk/bigbounce** or contact our events team on 0300 373 1100 or **events@cysticfibrosis.org.uk**

"This year's theme is transplantation"

Cupcake anyone?
The Big Cake Bake is back, supported by celebrity chef James Martin and sponsored by Western Power Distribution. Download or order your guide to holding a Bake at **cysticfibrosis.org.uk/bigcakebake**. You can also download our 'How to…' factsheet, poster, ingredients cards and invitations – everything you need to get started!

Every penny you raise will help us make a daily difference to the lives of those with cystic fibrosis and the people who care for them.

- £15 could over the cost of running our helpline for one hour, offering support for everyone affected by cystic fibrosis.
- £80 could fund a day of research into detecting early signs of lung disease.
- £250 could help someone with cystic fibrosis manage their travel costs when being assessed for lung transplant.

Take part in our CF Week raffle!
Buy a ticket for our CF Week raffle and you could win a top prize of £2,000 – or one of many other fantastic prizes. You can also sell tickets to your friends, family and co-workers and help us raise funds. To order tickets call our Supporter Care team on **0300 373 1040**.

Create your own event
CF Week is a chance to get creative and have some fun by holding your own fundraising event. Visit **cysticfibrosis.org.uk/howto** for some great ideas and important information on event safety and cross-infection.

Your Regional Fundraising Manager can provide advice and support, as well as fundraising materials showcasing our fantastic new brand, from banners and posters to collection tins and balloons.

Visit **cysticfibrosis.org.uk/inyourarea** to find your local contact

CF Week 2013?

Raising awareness of transplantation

This year's theme is transplantation, an emotive but important subject. During CF Week we will be highlighting our work in this area, the need for more lung transplants for people with cystic fibrosis, and the importance of better post-transplant care.

Thanks to improvements in specialist care and treatment, people with cystic fibrosis are living longer, healthier lives, but many will still reach a point where they need a lung transplant to prolong their life.

The success rate of lung transplants for people with cystic fibrosis is encouraging, but the procedure carries a number of risks, including rejection or infection. A transplant will not always be the most appropriate treatment for someone who is seriously ill.

There are currently around 80 people with cystic fibrosis awaiting a heart or lung transplant, or both. There is a shortage of suitable donor organs, and around a third of all those on the waiting list for a lung transplant will not live to receive a donor lung.

"Since going on the transplant list, I can't think of anything else, every time the phone rings I wonder if it's 'that call'." *Emily Icke, who has been on the transplant list since November 2012.*

Improving lung transplant rates

We are co-funding pioneering research to make previously unsuitable donor lungs fit for transplant. The DEVELOP-UK study, being led by Newcastle's Freeman Hospital, uses an Ex-Vivo Lung Perfusion (EVLP) circuit which pumps nutrients and oxygen through the donor lung. This research could change how lung transplantation is performed, increasing the supply of donor organs and reducing the number of patients dying while awaiting a transplant. By the end of the study more than 100 patients who may not have received a donor lung otherwise will have had access to a transplant and a chance at a longer life.

"My life is so much better now I've had my transplant; thanks to research funded by the Cystic Fibrosis Trust I got my call. I hope everyone who needs new lungs gets their chance!" *Philippa Bradbury, one of the first people to receive a transplant in the EVLP pilot.*

We were able to fund the pilot study for this research because of funding from the Robert Luff Foundation. In 2009 the Foundation granted us a further £500,000 to expand the study.

The British Transplantation Society said the technique could "dramatically" increase the number of lungs used. Results are expected in early 2016.

As part of CF Week we will be launching a debate to gather the views of stakeholders and highlight specific issues affecting transplantation for people with cystic fibrosis. Ultimately, we want to ensure that all those that are suitable for a lung transplant receive one and that those who do receive a transplant have the best possible outcomes. To join in the consultation email **enquiries@cysticfibrosis.org.uk**

Keep up to date with all the latest CF Week news at **cysticfibrosis.org.uk/cfweek**

Support us by remembering someone special

Many donations received at the Cystic Fibrosis Trust are made in memory of a loved one, perhaps as collections at a funeral instead of flowers, or as an anniversary gift. There are many ways family and friends can be involved in giving in memory, and we have produced some information and materials that may be helpful at this time.

- Our new leaflet gives details of different ways of supporting the Cystic Fibrosis Trust in memory of a loved one.
- Our donation envelopes provide a very simple way to help organise a collection, but will also allow us to benefit from Gift Aid.

To receive the leaflet or order donation envelopes, please contact Sam Loughnane on **020 8290 8030** or email **inmemory@cysticfibrosis.org.uk**. You can also order donation envelopes on our website at **cysticfibrosis.org.uk/inmemory**

Trunki Daddy

Rob Law on beating the Dragons – and cystic fibrosis

It's a chilly March morning in Bristol, and we've arrived at the offices of Trunki for a photoshoot and filming session with the company's CEO and founder Rob Law.

Not sure what Trunki is? You may not be familiar with the name, but if you've passed through an airport, chances are you've seen one. The brightly coloured suitcases for children defied an infamous thrashing on BBC's *Dragon's Den* to become a commercial hit; a Trunki suitcase is now sold every minute, and in just seven years of trading the company's reach has expanded to 97 countries across the world.

The unassuming stone exterior of Trunki HQ belies the colourful and creative hub inside. Pink, yellow, blue and fetching cow-print suitcases line the walls, new swimming bags, booster seats and pillows dot the office and spacemen peer out of lifts… there's even a giant orange slide between floors. These are no ordinary offices…

Rob's lightbulb moment came when travelling, and observing the lack of child-friendly luggage available. In the middle of a design degree at the time, it didn't take long to get working on a solution, and the first Trunki prototype was soon born (it still sits proudly amongst the gleaming new models at Trunki HQ).

"I knew I was on to a good idea," Rob says. "We did extensive user testing, and both kids and parents loved the product."

But it took some time to convince retailers that Trunki was not 'toy luggage', and Rob was repeatedly rejected by the UK's largest luggage and toy manufacturers. Undeterred, he continued to knock on doors, all the while tinkering with the prototype to ensure it was the best it could possibly be.

Six years after graduating with first-class honours from Newcastle University, and with a grant from the Princes Trust, Rob was able to manufacture his first batch of Trunki suitcases and begin selling the product in the UK.

The business suffered a number of major setbacks in the early days, from faulty production, to a crucial supplier going bust, to the terror alerts in 2006 that saw a ban on airplane hand luggage. But the biggest turning point for the company came in 2006, when Rob appeared on the BBC's Dragon's Den.

"Trunki needed publicity – and money. I really thought they couldn't turn it down," he recalls.

Alas, things did not turn out quite as planned. Despite an excellent start, Dragon Theo Paphitis managed to break the Trunki's strap within seconds of picking it up. The Dragons were all 'out', and to add insult to injury, Theo's scathing comments were used to advertise the episode before it was aired.

> "A Trunki is sold every minute"

Commiserating in the pub after filming, Rob was convinced the escapade had cost him the business he'd worked so hard to get off the ground. But thanks to some quick thinking PR, things took an unexpected turn. Rob added a survey to the Trunki website, asking for the public's feedback on his product. The night the episode aired, 2,000 people filled it out. Words of support – and Trunki orders – came pouring in.

Within weeks, Rob got through the doors of what was his business's holy grail – the John Lewis luggage department. ▸

Opposite: Rob Law at Trunki HQ in Bristol, March 2013; right: Rob and various Trunki models.

"I never looked back," says Rob. And things certainly run a lot more smoothly nowadays. With 15 different Trunki models and five sub-brands, and a turnover last year of over £6 million – for the Dragons, Trunki really was the one that got away.

In 2010, Rob was awarded an MBE. But his career highlight was when Trunki won the best small-to-medium enterprise at last year's National Business Awards.

"There is not much time to reflect on success"

"It's the most prestigious award we have won to date," he says. "I'm so proud that we have been able to achieve the results responsibly with our innovative products and business model. The most satisfaction I get from business is working alongside my great team, seeing us achieve phenomenal things and punching well above our weight."

Success is certainly not something Rob takes for granted. Bitter experience has taught him that building a global brand is not easy.

Why I wanted to film Rob's story

The film about Rob's life was produced on a pro-bono basis by Tim Miller, a former executive producer at ITV. Tim explains why he was inspired to film Rob's story.

"I was lucky enough to have seen Rob present the Trunki story and was completely blown away. It was an almost unbelievable tale of determination and the indomitable human spirit – and that was just the business story. When you consider that he's achieved all this while contending with cystic fibrosis, it almost defies belief.

"I thought it was truly inspiring – as did another CF parent who also heard Rob speak, and felt that more people should know about it. You can't control the extent to which you're affected by cystic fibrosis, but Rob's story does remind us that – with or without the condition – perseverance can pay off."

"With challenging economic conditions and managing our continual growth, I'm always kept on my toes. People often say I must be incredibly proud of what we've achieved, but there is always the next challenge round the corner and the to-do list never gets shorter! There is not much time to reflect on success."

Rob's story is perhaps all the more remarkable given he has cystic fibrosis. Whilst he doesn't make a point of disclosing this, he is happy to share his story, despite the devastating impact it has had on his life. In 1994 Rob's twin sister Kate lost her life to the condition, aged just 15. Her death was a defining moment for Rob, both personally and professionally.

"I learnt very early how precious life is and I was determined to make the most of my time," he says. "I promised myself I would not let cystic fibrosis beat me."

The fact that Rob has had to overcome so many hurdles could explain why, despite his success, he remains resolutely down to earth. When it comes to business, he clearly has a steely drive and determination – but it comes with a healthy dose of humour too. He may be a CEO, but prefers his informal job title – Trunki Daddy. The office where Rob and his team of 30 work is fondly referred to as the 'mothership'. And Rob doesn't mind admitting that Trunki appeals to his less serious side.

"When you spend over half your waking life working, it seems a shame not to make it fun and enjoyable – it makes a big difference to the team's morale and helps us recruit top talent. At the end of the day business is all about people."

Rob admits that juggling the demands of cystic fibrosis with those of his business can be a challenge.

"Cystic fibrosis can grab you when you least expect it," he says. "Staying fit and healthy enough to run a business isn't always easy. I spend two hours a day staying on top of my condition – it creates a huge demand on my time."

'Staying fit and healthy' is something of an understatement; Rob regularly takes part in extreme sporting events, from marathons and triathlons to his

Left:Tim Miller; above: Rob is interviewed at Trunki HQ.

The Sixty-Five Roses Club: one year on

Rob Law is patron of the Sixty-Five Roses Club. Launched in May 2012, the club is designed for donors who support the Cystic Fibrosis Trust with gifts of between £1,000 and £5,000 annually.

Members receive personalised updates each year to show the work they are supporting, and are part of an exclusive group invited to an annual reception and lecture during CF Week. This provides an opportunity to meet other like-minded supporters, many of whom are affected by cystic fibrosis, and to hear from experts about our work.

Since its launch just over a year ago, the club has attracted 26 committed members – including Rob – who have donated over £30,000 towards our vital work. We're very grateful for this support and delighted that our first year has been such a success.

What we hope to achieve

Cystic fibrosis is a lifelong challenge. In the more than 70 years since the first diagnosis, improvements in the understanding, management and treatment of cystic fibrosis have taken it from a disease of infancy to a long-term condition. The strides made have increased median predicted survival in the UK to 41 years – but it's not good enough. We're here to beat cystic fibrosis and make a daily difference to the lives of people with the condition, and those who care for them. With your help, we can win this fight.

How you can help

The Sixty-Five Roses Club is about you, and provides an opportunity to support one of our broad areas of work – clinical care, medical research or support services. If you are interested in becoming a member of the Sixty-Five Roses Club, our next annual lecture is being held in London on Tuesday 26 June, so you are welcome to come along and find out more. We ask for a three-year commitment to the Sixty-Five Roses Club as this will help us plan for the future, so that we can keep working for change, developing better treatments and, ultimately, a cure.

If you would like to become a member of the Sixty-Five Roses Club or have any questions please contact Lindsey Burke, Major Donor Manager, on **020 8290 8035** or email **lindsey.burke@cysticfibrosis.org.uk**

The phrase '65 Roses' comes from a small child's attempt to pronounce cystic fibrosis (CF) and is now a term used by the many in the CF community when raising funds for the Cystic Fibrosis Trust.

biggest challenge yet – a sub-five-hour half Iron Man – applying the same dedication and discipline to his training as he does to his business.

"I get a real buzz from giving it my all; crossing the finishing line knowing I could not have gone any faster gives me a huge sense of achievement and also gives the single finger to cystic fibrosis."

"Cystic fibrosis can grab you when you least expect it"

So where next for Trunki? Rob is keen to grow it into '*the* global brand for families on the go.' Accordingly, the company is currently developing a new children's travel gear category.

But it's not all work work work. "I'm enjoying having my weekends and holidays back," Rob admits, "and spending more time with my partner Kathryn and our two cats." (Though you won't be surprised to learn he's squeezing in training for five more triathlons in his spare time).

The filming wraps, and Rob is straight in to another meeting. We leave the 'mothership', passing by cabinets of trophies and awards on the way out. With Rob at the helm, and a new range of Trunki products about to launch, no doubt there will be many more to come. ■

Words: Jacqueline Ali

You can watch Rob in our new film 'Cystic fibrosis… not going to stop me' on the Cystic Fibrosis Trust website. Share your thoughts on our Facebook page, Twitter or our website forums.

If you've been inspired by Rob's fitness challenges, check out **cysticfibrosis.org.uk/events** to see the range of challenge events you can take part in to raise funds for us.

Targeting the root cause of cystic fibrosis

Therapies that target the genetic defect in cystic fibrosis are creating a buzz. Dr David Sheppard (DS), a CF Trust-funded scientist at the University of Bristol, talks to is Editor Jacqueline Ali (JA) about his painstaking work developing new drugs to tackle the root cause of cystic fibrosis

JA Tell me about your research in Bristol.

DS I'm fascinated by CFTR, the protein that goes wrong in cystic fibrosis. CFTR forms a gated channel for chloride ions (one part of salt) to pass in and out of cells. I seek to understand how this protein works, how it goes wrong in cystic fibrosis and how drugs alter its activity. This knowledge is essential to develop new therapies that target the root cause of cystic fibrosis.

I'm also interested in alternative approaches to therapy that bypass the loss of CFTR activity in cystic fibrosis, by designing and building molecules to shuttle chloride across cell surfaces.

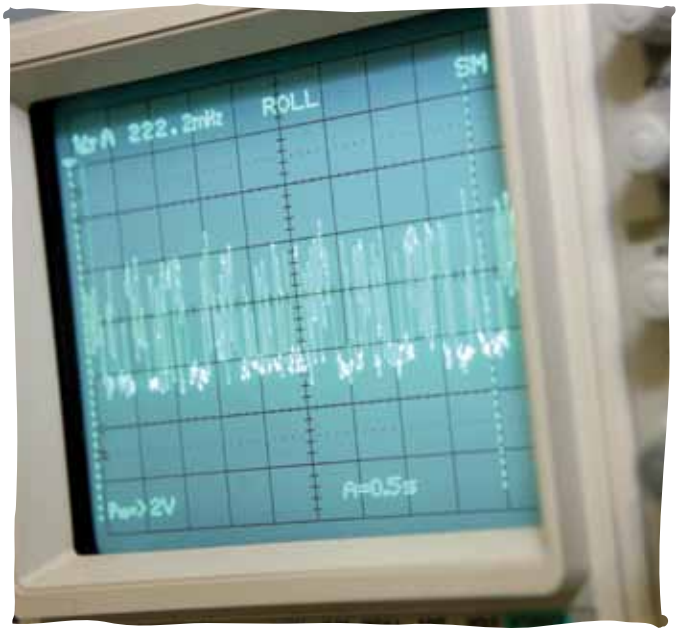
JA Why is CFTR so important in cystic fibrosis?

DS Because CFTR doesn't work properly in cystic fibrosis, normal flow of salt and water in and out of cells is disrupted. This leads to the build up of thick sticky mucus in organs, causing damage and attracting infection, which leads to serious problems for people living with cystic fibrosis. If we can correct the action of CFTR we could prevent the consequences of gene defects.

JA How do you study CFTR?

DS As chloride ions stream through open CFTR channels, they generate tiny electrical currents. We measure these currents to monitor CFTR activity. By studying single CFTR proteins fluttering open and closed in real time, we learn how gene defects produce faulty channels and how drugs make these faulty channels work better. It's very painstaking work, but extremely rewarding because of what we learn.

Opposite: Dr David Sheppard is studying the function and activity of CFTR, the protein that goes wrong in cystic fibrosis; right: electrical currents generated by chloride ions are used to monitor CFTR activity.



JA What impact do the different CF gene mutations have on the CFTR channels?

DS We can identify precisely how different gene defects lead to faulty CFTR channels. Some defects prevent CFTR delivery to the cell surface. Others obstruct chloride ion flow through the CFTR channel. And some hinder the opening of CFTR's gate – as if the gates are rusty and don't want to open. We use similar approaches with drugs to learn how they restore activity to faulty CFTR channels.

"I'm looking forward to the day when the structure of CFTR is identified"

JA What are potentiators and why are they important?

DS Potentiators are one class of drugs that target faulty CFTR. Just as oil makes a rusty gate open more easily, so CFTR potentiators allow CFTR's gate to work properly. This type of drug only works when faulty CFTR is present at its correct location the cell surface. Ivacaftor is the first CFTR potentiator to become available for individuals living with cystic fibrosis. By itself, it is not a treatment for the most common cystic fibrosis mutation F508del. However, ivacaftor is highly effective at restoring function to G551D, a gene defect present at the cell surface, but with almost no activity. When ivacaftor was tested in the clinic, it produced a robust and sustained improvement in lung function. In combination with other treatments to deliver faulty CFTR to the cell surface, CFTR potentiators will have a central role in future therapies by boosting greatly the activity of faulty CFTR.

JA Tell me about your latest grant from the Cystic Fibrosis Trust

DS With Steve Husbands from the University of Bath, we are investigating an innovative approach to drug therapy for those with the F508del mutation. To restore function to F508del, at least two drugs are required. One drug is needed to deliver F508del to its correct location, the cell surface. This type of drug is called a corrector. Because F508del forms a channel with a faulty gate, a CFTR potentiator is also needed to correct chloride transport by F508del. There is evidence that some chemicals possess the properties of both correctors and potentiators. We would like to know exactly how these chemicals work and learn whether we can develop better chemicals of this type. This is very much basic research, but it might lead to work to develop innovative therapies that target the root cause of cystic fibrosis. I'm very excited about this project.

JA What does the future hold for CFTR research?

DS There are lots of exciting possibilities. I'm greatly looking forward to the day when the structure of CFTR is identified. With this information, we will be able to design drugs that dock specifically with CFTR. I'm also working with Professor Tony Davis, a chemist at the University of Bristol to develop artificial transporters. This work is still at an early stage, but we have been able to show that artificial transporters shuttle chloride across cell surfaces and another group has tested them in cystic fibrosis cells. So perhaps one day artificial transporters might be used to treat cystic fibrosis and other diseases caused by faulty ion transport. ■

Transformational treatments

Dr Sheppard and colleague Yiting Wang of the University of Bristol summarise the new and emerging therapies that could revolutionise future treatment of cystic fibrosis

Today, most cystic fibrosis treatments tackle disease symptoms. However, future therapies will treat its root cause – faulty genes and proteins. Last year, ivacaftor (Kalydeco), the first of these new transformational therapies, became available to those with the G551D gene defect. It is hoped that ivacaftor will be the first of many transformational treatments to benefit people with cystic fibrosis. Here, we discuss some of the different approaches being explored to develop such therapies.

Gene therapy

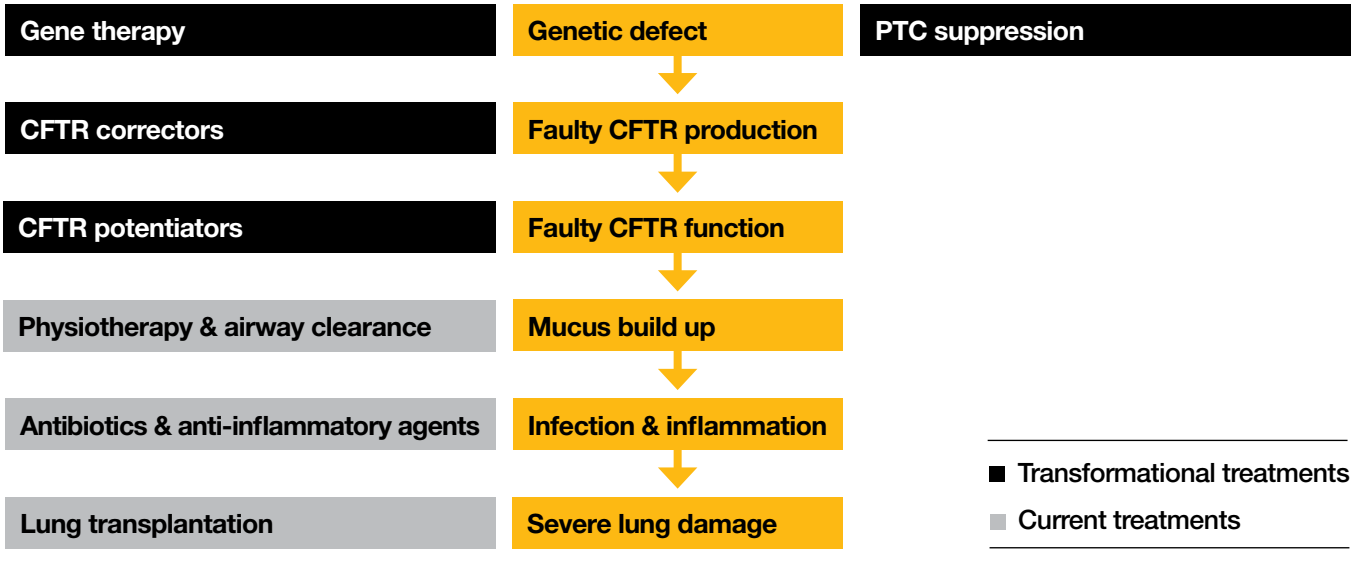
The UK CF Gene Therapy Consortium is currently conducting a year-long clinical trial, where a new copy of the CFTR gene is introduced into the lungs of people with cystic fibrosis using fat droplets. The results of this trial – expected 2014 – will be a key milestone in the development of transformational treatments. Separately, the Consortium is working to develop alternative, more efficient methods to deliver the CFTR gene into lung tissues using modified viruses.

PTC suppression

In the UK, a small number of people with cystic fibrosis have gene defects that introduce unwanted 'stop signals', (called premature termination codons – or PTCs) into the CFTR gene. These signals prevent CFTR protein from being made inside the cell. Because certain antibiotics block unwanted stop signals, drugs have been developed that silence these signals. One such drug, PTC-124 (ataluren), developed by PTC Therapeutics, has been tested in the clinic with some success. In the future, drugs like ataluren will likely be used alone or with other transformational therapies to restore CFTR function.

CFTR modulators

Correctors: The gene defect F508del prevents CFTR reaching the cell surface. Previous work showed that growing cells in the lab at room temperature delivered F508del to the cell surface, where it had some function. Cooling is not a suitable treatment for cystic fibrosis, but drugs that mimic the effects of cooling have considerable potential. CFTR corrector is the name given to this class of drugs.



With support from the US CF Foundation, Vertex Pharmaceuticals developed VX-809 (lumacaftor), the first CFTR corrector to be tested in the clinic. Although VX-809 was safe to use, it was without effect on lung function. Other CFTR correctors are now under development.

Potentiators: There is great interest in ivacaftor, the first approved transformational drug therapy and the first of a class of drugs called CFTR potentiators. These drugs boost CFTR activity at the cell surface by promoting the opening of the gate that controls chloride flow through CFTR. Although this class of drugs is primarily designed to restore function to gene defects that obstruct gate opening (e.g. G551D), it is likely that they will benefit multiple gene defects when used in combination with other transformational treatments.

Corrector-potentiator combination therapy: Some gene defects cause multiple problems for CFTR. For example, F508del not only prevents the delivery of CFTR to the cell surface, it hinders opening of CFTR's gate. To tackle these different F508del defects, CFTR correctors and potentiators are being tested in combination. In the lab, ivacaftor and VX-809 strongly boosted chloride transport by F508del. Based on encouraging clinical data, the US drug regulator, the FDA, awarded ivacaftor and VX-809 "Breakthrough Therapy Designation" to accelerate therapy development. Vertex Pharmaceuticals is now planning a large international clinical trial of ivacaftor and VX-809. This is the CF trial to watch in 2013.

Future therapies

Research in many different areas is likely to speed up the development of transformational therapies. The successful transplant of a trachea grown from an individual's own stem cells in 2008 argues that one day it might be possible to regenerate organs damaged by cystic fibrosis. Alternatively, genetic engineering might be used to repair gene defects within cells, or synthetic biology used to design artificial transporters to shuttle chloride across cell surfaces. These are just some of the exciting areas under investigation that might yield future transformational therapies. ■

In January it was announced that ivacaftor (Kalydeco) would be funded in Scotland through a special fund for orphan medicines. For eight-year-old Maisie Black, just two months on the drug have been life-transforming, as mum Tilda explains.

My daughter Maisie has now been on ivacaftor for just over eight weeks and we feel that there has been a huge improvement in her symptoms. When she was discharged from Yorkhill hospital, a day after starting ivacaftor, she still had a very productive sounding cough and a crackle in her chest.

"Maisie has been given a lifeline with ivacaftor."

She had been in hospital for three weeks for IV antibiotics and was undergoing very aggressive physiotherapy, but without much improvement.

Within the first couple of days of taking ivacaftor Maisie developed a runny nose then with physio would cough up what our physiotherapist advised us was old mucus. Within a week and a half the crackle – which we were told would not go – could not be found on examination. At her first clinic since starting ivacaftor (one month in) her FEV1 had increased from 98% to 104%, her height is up 3.5cm and her weight was up almost 2kg.

The biggest improvement that we see is in her digestive system. Maisie has always suffered hugely with digestive issues and bowel obstructions but is now having normal bowel movements. Following discussion with her doctors we have now taken her off Senna and Metronidazole and are gradually reducing her Movicol. She used to be in pain after eating almost every day; this has not happened once since starting ivacaftor. It has made a huge difference in the way she feels. She is full of energy.

We feel a weight has been lifted and although treatment-wise not much will change we now feel so much more positive for the future. At eight years old Maisie has already been through so much; she has been given a lifeline with ivacaftor. As a family it will change all of our lives."

Join the discussion on Kalydeco and other transformational treatments on our web forums – visit **forum.cysticfibrosis.org.uk**

TORPEDO-CF

Can you help eradicate *Pseudomonas*?

TORPEDO-CF is a clinical trial that opened in 2010. It aims to recruit 280 adults and children with cystic fibrosis to help work out which is the best treatment for early infection with *Pseudomonas*.

In some CF centres, treatment with intravenous antibiotics is considered the best way to eradicate *Pseudomonas*. Other centres prescribe oral antibiotic treatment with ciprofloxacin. Both intravenous and oral antibiotics are effective in treatment of *Pseudomonas*. What we don't know is whether there are important differences, especially in success of eradication, between these two strategies. If there are differences, and one of these treatments is clearly superior to the other, then this information would be very helpful to cystic fibrosis patients infected with *Pseudomonas* and the CF teams that look for and treat early infection with this organism.

"Having recruited two young people with CF attending our paediatric centre, I must say I have been very impressed how straightforward and easy the enrolment and randomisation process is for TOREPDO-CF. Our patients are very interested in the study and motivated to contribute to establishing the evidence we need to work out which regimen is most effective"

Dr Tim Lee, Leeds Teaching Hospitals NHS Trust

The importance of clinical trials

In December 2012 a medical research conference was held, which people with cystic fibrosis, their families and parents were invited to attend by internet link. Viewers were able to send in questions live to the panel of expert cystic fibrosis researchers. The talks and discussions were broadcast live over the internet and can be viewed online at **http://cfunite.org/events-archive/uk-cf-clinical-trials-meeting**.

The meeting was held mainly to emphasise to CF teams and to people affected by cystic fibrosis that participation in research is one of the most important ways to improve health and outcomes in cystic fibrosis. There is a need to compare one treatment with another so that evidence can be built up that demonstrates the best way to treat each aspect of the condition. One of the important ways to do that is to make sure that people with cystic fibrosis and their families are aware of the research that is going on and are aware of the studies that they are able to take part in.

"Participation in trials is one of the most important ways to improve health and outcomes in cystic fibrosis"

TORPEDO-CF is an excellent example of a national trial that should be available to all patients of any age that become infected with *Pseudomonas*. The trial has now recruited 115 children and adults from the 69 UK CF centres.

If you would like further details about the study, please contact your own CF team, visit **www.torpedo-cf.org.uk or email torpedo@liv.ac.uk**

"This research is long overdue"

Zoë and Steve Elliott, from Nottingham, have two-year-old twins, Alexander and Isobel, who were both diagnosed with cystic fibrosis shortly after they were born. They hope TORPEDO-CF will give parents of children with cystic fibrosis firm evidence of whether IV antibiotics really do offer the best treatment option.

Zoë said: "When we were invited on to the trial I was initially really surprised that this research hadn't already been done because in our experience IV antibiotics are generally perceived among parents of children with CF to be the best way of eradicating the infection. As a mum, if it's a toss-up between being at home with a nebuliser for three months or having your child in hospital having invasive treatment for two weeks I know what my gut instinct tells me — it's home every time. But it would be reassuring to have some firm evidence to allow us to make a more informed choice and that's why this research is long overdue." ■

Above: the Elliot family from Nottingham is part of TORPEDO-CF.

Improving care and treatment

The UK Cystic Fibrosis Registry Review 2012

Noreen Caine, Registry Consultant at the Cystic Fibrosis Trust, explains the purpose of the UK CF Registry Review, and highlights how the Registry is helping improve care and treatment of people with cystic fibrosis.

The UK CF Registry is a powerful tool that helps us monitor and audit CF care in the UK. It is used to determine best care and treatments, and helps us plan clinical trials of new therapies and provision of future care. The success of the CF Registry is due largely to the commitment of the 99% of UK CF patients who have consented to their routine clinical information being entered onto the Registry, and the CF centres that enter the data.

In recognition of the need to provide feedback to people with cystic fibrosis, their carers and clinical staff, the Registry Review 2012 has been developed. It aims to show how data collected is supporting continuing improvements in the safety and quality of care. In particular, the Review highlights how the Cystic Fibrosis Trust is working with the NHS to inform the planning and commissioning of services for patients and working with industry to improve the safety of medicines.

> "The success of the Registry is due largely to the commitment of CF patients who consent to their data being entered"

Working with the NHS to improve the quality of care

The NHS in England and Scotland have contracts with the Cystic Fibrosis Trust for the provision of information from the Registry to inform the planning and commissioning of services. The purpose is to improve the quality and efficiency of care provided.

In April 2013, a new national funding system for cystic fibrosis in NHS England was implemented, which should lead to greater consistency and fairness in funding CF care. Payment by Results (PbR) will mean that people with cystic fibrosis receive care funded according to individual needs, including the drugs they are prescribed. Information from the Registry helps the NHS ensure that funding of services is based on good information about disease severity.

Top right: the Registry will help ensure the safety of new drug therapies for cystic fibrosis.

Working with industry to make medicines safer

Once new drugs have completed clinical trials and are licensed for use in the NHS, drug companies have to monitor the long-term safety of the drug in everyday practice, usually over a five-year period.

The Cystic Fibrosis Trust has been approached by several companies wishing to use the Registry to perform long term safety studies after the introduction of new therapies; this is on the recommendation of the European Medicines Agency.

Our main objectives in agreeing to this programme of work are to:

- improve the quality of these long-term safety studies.
- ensure that patients gain early access to new drugs which have been through a rigorous safety assessment.
- secure funding to help in the running costs of the Registry.

Which drugs?

The choice of which companies to work with is driven by the potential benefits to patients of the drugs coming to market. These choices, and the development of agreements with the companies that safeguard the integrity of the Registry, is work that is being conducted under the aegis of the CF Registry Steering Committee with study protocols approved by the European Medicines Agency.

Each company will pay the full costs of their study and the income received provides an opportunity, for the first time, to pay a Registry Support Grant to each clinical centre. ■

The Registry Review 2012 and a Working with Industry leaflet are available on our website at **cysticfibrosis.org.uk/registry**

Food for thought

To maintain a healthy weight and better fight off infections, people with cystic fibrosis must pay special attention to their diet. In the first of two new columns, chef Nicholas Richards explains why he was inspired to create a new website to provide nutritional advice to those with the condition.

I am 25 years old and was diagnosed with cystic fibrosis at birth. My career began in IT, but I got bored of sitting at a desk all day. I began working as a chef, and then kitchen manager, in a carvery restaurant. Unfortunately, the long hours and the demands of the job took their toll on my health. I lost my job, and, temporarily, my interest in cooking. But I picked myself up and moved to London to work as sous-chef. It was the best kitchen I've worked in – fresh ingredients, and my ideas on the menu. I learnt so much about cooking, and became a true chef. Soon after I moved back up north to an independent restaurant where I now work.

"My fiancée stops me forgetting my Creon!"

I developed my website **cffoodtoday.com** to provide high calorie, nutritious meal ideas to people with cystic fibrosis. Between 80–90% of people with cystic fibrosis can't absorb the nutrients from food, so they need a tailored diet with a higher number of calories. Without this, low body weight and malnutrition may occur.

There is lots of information out there about healthy eating and weight loss, but it's harder to find information about high calorie meals. Through my website I hope to help people with cystic fibrosis maintain a healthier, balanced diet, rather than think fast food is their only option.

Although my weight has always been quite stable, I find weight gain difficult. My fiancée keeps me on track with a food plan, and stops me forgetting my Creon when going places! I try and average around 5,000kcal each day. Maintaining body weight is often hard with cystic fibrosis, especially as some medication we take to look after other parts of the body can affect our appetite. I find watching cookery programmes helps. You could also browse cook books for ideas and inspiration. If you're unable to cook, could you help out in the kitchen or watch while someone is cooking? The smells in the kitchen can help your appetite and start the ball rolling.

I'm about to add two new tracking features to my website to help people keep an eye on their weight and also their food and Creon intake, which I hope will help at clinic appointments to help people with cystic fibrosis and their dietitians better understand and improve their health. Everyone with cystic fibrosis should have access to a specialist CF dietitian, to help them maintain a healthy weight and stay as well as possible.

Expert comment

Sarah Collins: Specialist CF Dietitian, The Royal Brompton Hospital, London

For the past 25 years dietitians have recommended a high fat, high calorie diet as a standard part of nutritional care of people with cystic fibrosis; this has resulted in improved nutritional status and survival. The majority of people with cystic fibrosis should be able to achieve good growth and nutritional status by following these recommendations. It is important that all children and adults with CF receive regular dietary reviews with a CF specialist dietitian to ensure optimal nutritional status is achieved and maintained.

The CF Trust's nutritional consensus document is currently being reviewed taking into account current evidence for best dietetic practice; this will provide an up-to-date resource for dietitians and other health care professionals working in cystic fibrosis. It will be published in 2013. ■

The Cystic Fibrosis Trust publishes factsheets on nutrition for babies, children and adults with cystic fibrosis. Visit **cysticfibrosis.org.uk/publications** to download

Above: chef Nicholas Richards; right: a high calorie diet is recommended for people with cystic fibrosis.

Fighting fit

Exercise is a great way to help keep the lungs clear in cystic fibrosis. In the second of two new columns, personal trainer and model Ben Mudge shares his philosophy on health and exercise.

I am a 23-year-old personal trainer and fitness model, and I am grateful for this opportunity to share with you what I believe to be one of the most important aspects of life – exercise.

I have always been interested in sports, though I pursued a career in the film industry after leaving school. But I quickly realised that the long hours did not allow much time for exercise or sports in my life, and my fitness levels and weight dropped rapidly. If I wanted to stay healthy something had to change, so I joined a gym and made regular training a part of my weekly routine.

I noticed positive changes in my weight, my health and my confidence almost straight away. A friend then asked me to help him out with a training programme as well, which I found I really enjoyed. This led me to where I am now – a qualified Personal Trainer – in my opinion one of the greatest jobs in the world. I get to help people become healthy and improve their lives through exercise and nutrition. I am also a sponsored athlete by both Reflex Nutrition and Better Bodies Clothing.

Above: personal trainer Ben Mudge.

> **"I noticed positive changes in my weight, health and confidence almost straight away"**

Last year I tested myself a bit more and applied for *Men's Health* Cover Model Competition. This required me to really look at my diet and to take my training to the next level. I got to the final five from thousands of applicants – an experience I won't forget.

I am often asked how I motivate myself to train nearly every day. The simple answer is I like to be healthy (and to look good). My biggest push to train is knowing that nothing worth having comes easily – sometimes you have to put in the work when no-one is watching.

My experiences have taught me that it is vital to look after your body through exercise plus a healthy nutrition plan. Without these two things I would not be where I am today. Did I mention I have cystic fibrosis? I may have it in my life, but I'm determined not to let it have my life. I look upon cystic fibrosis as a reason to exercise, not an excuse not to.

Stay tuned for some benefits, tips and ideas on how to make exercise part of your life.

Facebook: **Ben Mudge**, Twitter: **@mudge90**, YouTube: **mudge90**

Expert comment

Dr Judy Bradley, Professor of Physiotherapy and Lisa Kent, Research Associate – University of Ulster

There is increasing evidence that exercise and physical activity can play an important role in the management of cystic fibrosis. Where possible, adults with cystic fibrosis should aim for at least 150 minutes a week of moderate intensity physical activity, such as brisk walking. Children with cystic fibrosis should aim to be physically active for at least 60 minutes per day (NICE 2009). These targets can be achieved in a number of short, 10-minute bouts throughout the day.

You don't need to go to the gym to get physically active. For those less interested in training programs, or maybe even a little intimidated by the gym scene, activities that are part of your daily routine, such as walking upstairs instead of getting the lift, can be of benefit. Reducing the time spent sitting or lying every day is also important for our general health.

Your CF team can provide individualised advice on how to get more active, and what type of activity would most meet your needs. Many CF teams are involved in studies to further explore the benefits of exercise and physical activity, as well as strategies to help people become and stay more active. Ben is part of our research team in Belfast developing a physical activity program for children with cystic fibrosis and their families. These studies are important to ensure that patients receive optimal advice and have access to evidence-based intervention. ■

Cystic Fibrosis why we're listening

3,000

calls are answered by the helpline every year

The Cystic Fibrosis Trust helpline provides high-quality, impartial information and advice for people with cystic fibrosis and their carers.

If you have questions about benefits, welfare grants, genetic screening, or the latest research and treatments – or simply want someone to talk to – we're here, on the end of the line.

Call our helpline: 0300 373 1000
9am–5pm, Monday to Friday

cysticfibrosis.org.uk/helpline