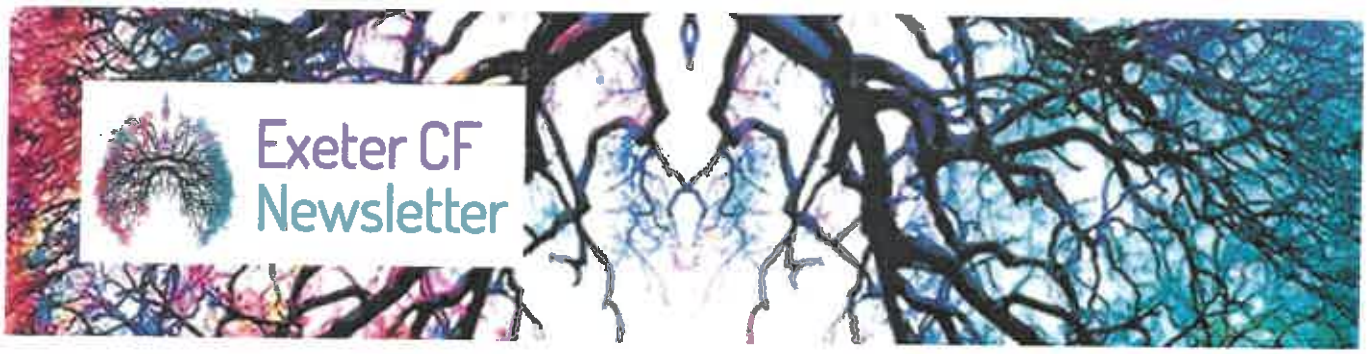

Summer 2019

New resources for children of parents with CF

We know from speaking to parents who have cystic fibrosis that explaining the condition to their children can be tricky. The CF Trust has worked with families affected by CF and CF clinicians, to publish two stories about Seb and Rosie, who both have a parent with cystic fibrosis. Both stories are roughly aimed at children aged 5-9 years old and can be found at: https://www.cysticfibrosis.org.uk/the-work-we-do/publications/rosie-and-seb

Seb's and Rosie's stories aim to encourage conversations about cystic fibrosis and how it can affect the lives of people with the condition and their families. Both books include things to chat about with your child after you have read the story. You can order the stories as printed, hardback books, download them as PDFs to read on your computer, phone or tablet, or watch as motion storybooks. The books were funded by Jeans for Genes, a charity that raises money for the care of children and families who are affected by genetic disorders.

The Digital CF Generation

Would you like to be involved in the development of a new digital tool for enjoyment, health, and well being? Researchers at the University of Exeter are setting up a patient and public involvement group to help guide their work. This group can provide their perspectives and feedback on some of the ideas the researchers have and creative content that might be used in the tool, to make sure that the research is relevant to people who have cystic fibrosis. The group will either work one-to-one with the researcher or as a group via the telephone, Whatsapp, or Skype.

No programming skills are required! – Just your enthusiasm for digital technology and an interest in voicing your opinion. We are specifically inviting people who have CF who are between 12 and 18 years of age, but we would also like to invite parents and carers. If you would like to be involved or you would like to find out more, please get in touch with Dr Sam van Beurden at the University of Exeter – 01392 726 440 or s.b.vanbeurden@exeter.ac.uk

I-neb reminder

Green chamber – Pulmozyme

Grey chamber – Promixin (mixed with 1ml of 0.9% saline), Colistin, Salbutamol

Lilac chamber – Hypertonic saline (x2 fills), Tobramycin (x2 fills)

For those who use an I-neb but are no longer taking Promixin:

New discs and chambers will now be supplied by the hospital rather than by Phillips. You will be supplied with new chambers every 12 months and discs as required. Please be patient with us during this transition, we may need some prompting!

Creon shortages

There have been some issues with the supply of Creon 25,000. These should now be resolved but it may be helpful to have the helpline number for Creon issues:

Customer Services (Mylan)
01707 853 100

Combined multivitamin preparations now on the formulary: DEKAs Plus & Paravit-CF

We are pleased to announce that DEKAs Plus & Paravit-CF all-in-one multivitamins designed for those with Cystic Fibrosis are now available and can be prescribed by your GP. This will replace Multivitamins BPC, vitamin A&D, vitamin E and vitamin K which can all be stopped. Note: some patients will need to continue taking additional Vitamin D preparations such as Cholecalciferol, Calcichew D3 and ADCal D3. In your next clinic, CF dietitians will review and change your vitamin regimen to the most appropriate all-in-one preparation. For Infants 1-4 years Paravit CF liquid, this is a clear, odourless preparation. The other great thing is it does not stain clothes! For Older Children and adults DEKAS Plus, either as a Chewable tablet or a soft gel capsule. Samples of DEKAS will be available in clinic. You will be advised at your clinic appointment about the appropriate change and your GP informed of the necessary prescribing instructions.

Please remember to give/take the vitamin at the same time as a drink or meal to help absorption, especially if you use pancreatic enzyme replacement therapy (Creon). Blood vitamin levels will be monitored at annual review or earlier if deemed necessary and dose adjustments made if required.

The best questions attract the best answers – a word from the CF Trust

Seeing CF portrayed on TV and in the media can bring up questions, worries or concerns that people affected by CF might not have thought about before. If you are a parent, you might find your child asking questions that you find hard to answer or becoming aware of aspects of CF that you haven't discussed with them before. Every individual and family are different in the way they talk about difficult or sensitive issues, but being able to share worries or ask questions with people you love and trust can be really helpful. We have prepared this guide to support you in these conversations. Your CF team will also be able to support you and your family with any concerns.

It can help to ask open questions to start a conversation...

- "How does what you've seen or read compare to your experience of life with cystic fibrosis?"
- "What questions do you have about what you've seen? What parts of the film stuck with you? What questions did it bring up for you?"
- "Did you find out anything new about CF? How do you feel about that?"
- "Have you spoken to your friends about the film? What did they think about it? What questions do you think your friends might have about cystic fibrosis if they saw this?"
- "Is there anything you want to talk to your CF team about at your next appointment? How can I help you raise that with them?"
- "How CF specialists deal with cross-infection is very different in the United States – how do you think life for people with CF in the United States might be different from your own?"

Sometimes it can help to talk about ways you might feel, to encourage someone else to share their thoughts.

- "Some people might find that a bit worrying... How do you feel about it?"
- "That made me feel a bit uncomfortable – how are you feeling about it?"
- "I felt that [...] was portrayed really well – which bits did you enjoy?"
- "I found how they dealt with [...] really surprising – did anything catch you off-guard?"

Different topics and themes affect everyone in different ways, and it can be surprising what strikes a particular chord with different people, so you may find that what upsets you might not affect someone else, and that's OK – the important thing for everyone is to have the space and support to talk about it.

Everyone with CF is different, in terms of their health itself but also how they handle the challenges it can bring and how they feel about the condition. TV, books and in some cases, what we see online, is usually dramatised and 'creative licence' is used to make facts fit the story. Sometimes we need to talk it through to work out what is reality and what is drama, and how that makes us feel. We may see different views and perspectives of CF within just one film, book, video etc. It can be useful to talk about which of these someone does/doesn't identify with, and the fact that each one is a specific character created for the purpose of a story – in real life we are all a mixture of different views, and these can change over time as we grow, age and have different life experiences.

Research update

Here is a short summary of current and upcoming trials running at Exeter:
Adult trials that are open to new patients:
Living with CF: A Quality of Life measurement for people aged > 16 years with CF, in the UK CF Registry. Taking part involves completing an online questionnaire at annual review. Is your annual review coming up? If so, please get in contact if you would like to hear more about this study.
Hope-1 Study: This study is looking at the effect of a new nebulised treatment vs placebo (a dummy treatment) on lung function in people 18-50 years old with an FEV1 between 50-80%. Trial involvement lasts around 2 months.

Paediatric trials that are open to new patients:
Gilead Alpine 2: This study is looking at whether 14 days or 28 days Aztreonam Lysine (Cayston) is effective and safe for treating new onset Pseudomonas infection in children who haven't grown Pseudomonas in the last two years (or ever).
CF Start: This study is looking to compare two strategies for giving antibiotics to new babies with CF, "Prevent and Treat" flucloxacillin prophylaxis (standard care in the UK) vs "Detect and Treat" antibiotics, prescribed depending on microbiology results.

Upcoming trials:
Vertex NextGen: This study is looking at the effect of a new triple therapy treatment in those aged 12 years+. This trial is planned to start in the Autumn.
Proteostasis: This study is looking at the effect of a new triple therapy treatment in adults 18+. This trial is planned to start in the Autumn.
OligoG: This study is looking at a new inhaled treatment for treating chronic pseudomonas in those aged 12 years+. Date of recruitment starting in Exeter not yet confirmed, but you can find further information about the study here: https://oligogpivotalcf.eu