Summer 2017

Parents, families and friends meeting

Our next parents, families and friends meeting will be on Wednesday 13th September at 7.30pm, Seminar Room 2 in The Child and Women's Health building (Royal Devon and Exeter Hospital).

Please come along – cake provided!

Please could we ask for your help?

Did you know that we now look after over 180 patients with CF here in the Exeter CF Centre? All of those patients are seen anywhere between 6 monthly and weekly, always depending on how well or ill they are. If you are feeling unwell we can advise you and review you either by phone, or at the hospital if needed.

Due to the number of patients we have, and the number of clinic slots and rooms we have access to, we are struggling to fit everyone in. This is made more difficult by Bank holidays, on-call, non-CF commitments and people not attending or cancelling their appointments with no notice. So, if you know you cannot attend an appointment please let us know ASAP so we can offer it to someone else. However, it may be several weeks before we can fit you in. A lot of effort is put into our infection control measures to ensure that clinics are safe for you as a patient with CF. It is therefore important to try and come at the correct time to avoid the risk of cross infection. This applies to being both late or early.

The team is most grateful for your consideration of this and look forward to seeing you in clinic. Thank you.

Remember to drink!

In hot and humid weather those with CF will lose salt in their sweat and they are at risk of dehydration. This can cause tiredness, muscle aches and pains. It is important to ensure an adequate intake of salt and fluids:

Babies: Breast milk and formula milk are both good sources of fluid, salts and minerals. Offering additional frequent small feeds will help keep your baby's fluid levels topped up.

Children: Encourage frequent drinks, note avoid too much coca cola as this contains caffeine. Ice creams, ice lollies, frappes all add to fluid intake. Children may need reminding to drink with excitement of the holiday. Crisps, salty snacks and adding additional salt to food at the table will all boost intake.

If appropriate, salt supplements can be prescribed. Please ask your CF team if these are necessary.

Words from our physio

1. New patient leaflets released by the CF Trust on the Acapella Choice and the PARI PEP: https://www.cysticfibrosis.org.uk/the-work-we-do/clinical-care/supporting-clinicians/resources-for-clinicians/physiotherapy-leaflets

2. Protective covers for a TIVAD:
The CF Trust have been contacted by the dad of a boy with CF who is a really keen cricketer. They have had concerns about the port being hit. They have found a sports company who make a custom port guard. So far they have found this is working well, and the cost is under £100. The CF Trust are not endorsing this product in any way, as they don't know how effective it is, but they are aware of the risks of a port being hit or damaged when playing sports, so wanted to share this information. The company who make the guard: george@strettonfoxsports.co

3. Gym ball incidents reported by the Brompton:
Following some incidents relating to gym balls bursting: "To ensure that your gym ball remains in a safe condition please i) purchase a good quality gym ball which is anti-burst for your weight and your child's weight combined (remember they will grow) ii) store away from radiators iii) ensure that you clear a space for physio so the ball isn't bounced over toys etc and iv) ensure it is sufficiently inflated during use"

4. Consensus on physiotherapy management in CF 3rd edition 2017 recently released and available to download from the CF website.

A party with a difference...

http://www.devonlive.com/woman-who-was-never-meant-to-see-12th-birthday-celebrates-34th-year-with-big-party/story-30460183-detail/story.html

Healthy mind Healthy body

OneSmallStep is a new, free healthy lifestyle service which will deliver motivational, informative and guided small steps towards a happier and healthier lifestyle.

The four lifestyle behaviours that we have been commissioned to deliver are quitting smoking, reducing alcohol consumption, managing weight and getting more active.

The OneSmallStep service requires no paperwork and there is no formal referral process.

Simply visit **www.onesmallstep.org.uk** or call (freephone) 0800 298 2654 or 01392 908 139.

There is no eligibility criteria for accessing the service besides the customer being resident in Devon and being over the age of 18.

Let's talk poo!

Two new products are on the market to remove the embarrassing bathroom odours that can be an unwelcome symptom of cystic fibrosis. Air Wick's V.I.Poo and Poo~Pourri (poopourri.co.uk) are pre-poo eliminate sprays that help to trap nasty smells in the bowl for embarrassment-free flushing.

The protective barrier *traps* odour *under* the surface, *before* it ever begins. All you'll smell is a *refreshing* bouquet of natural essential oils. As the inventor of Poo~pourri says, 'you've got more important crap to think about'.

Upcoming research studies

Vege Alternative to Creon and Blue Poo! We are about to embark on a research study testing a new enzyme supplement called Lipromatase in people with CF over 7 yrs of age. Lipromatase has a plant origin and is more stable to stomach acid while not having high purine/uric acid levels. It is already used in the USA.

The study involves changing from Creon to Lipromatase or Pancrease guided by Penny and Marie. There are 70 CF centres taking part across the world, but only 3 in the UK.

Is there a downside? Well, at the beginning and then after a period of stabilisation (9 weeks), those taking part will need to stay in hospital for 4-5 days to follow a strict individualised diet while their poo is collected and frozen.

A blue dye (swallowed as capsules) is used to mark the beginning and end of the poo collection so that a detailed analysis of digestion can be made.

Those taking part will have all their expenses covered and will be paid for the inconvenience of being in hospital. Contact Penny or Marie if you are interested.

Alpine 2: Nebulised aztreonam is another antibiotic used to treat Pseudomonas lung infection. We plan to engage with a new study investigating whether it can be used to treat NEW isolates of pseudomonas in stable babies and children with CF.

It involves either a 2 or 4 week treatment period with close monitoring of sputum/cough swabs.It is exciting that we are continuing to look at new ways of treating Pseudomonas, which will give us new options. We will approach you if you or your child is eligible for this study.

Coming soon – the tidal wave of gene modifier therapies: Proteostasis PTI-428; Albatros GLPG2222; Vertex 101; HOPE-SPX101.

Seville ECFS Conference feedback 2017

The one theme that stood out for me was around promoting normalising life and the tasks of daily living for those with CF, as a coping mechanism. The theory is that treatments can be made to be an automatic action, rather than a chore. If they become a habit for the person who needs to do them, the burden is perceived to be less. For example, thinking of financial benefits that those with CF are eligible for can be thought of as pay for the hours that you need to spend on treatments. This theory is big at the moment, and later this year a research project (ACtiF- Adherence to treatment in adults with Cystic Fibrosis) begins that will explore this theory. An electronic CF Health Hub will be used to monitor and support people with CF. The Exeter CF Centre has been chosen as one of the research sites. 'Watch this space' for more information!