

Paediatric advice on clinical support and homecare (26.03.2020)

The clinical centres are working tirelessly to manage the huge volumes of calls and enquiries from our patient communities. For many they have had to either cancel all clinics and non-emergency appointments or move them to telephone appointments. These are unprecedented times and many of our doctors and nurses have already been, or are on standby, to be called to the front line.

Each centre has an answerphone to ensure that they do not miss any calls. Please bear with them as they filter through the many calls that are coming through on a daily basis.

The current advice is that critically ill LSD patients should not be transferred to the specialist centres and should continue to receive treatment at their local hospital as appropriate. Advice and support should be sought by your specialist centre as required. Keep to hand your specialist centre telephone numbers, any medical alert cards, care plans or leaflets for medical staff that explain your condition. Speak with your closest family members and advocates to ensure they know where information is kept'.

Please be aware that advice is changing daily and it is important that you keep yourselves up to date with the latest government advice:

<https://www.gov.uk/coronavirus>

Categories of patients

Extremely vulnerable high-risk patients

Clinical centres are already reviewing all their patients and will be in contact with those patients deemed to be high risk to discuss individual circumstances. Please find below the link to the government advice on shielding.

<https://www.gov.uk/government/publications/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19>

Social distancing -vulnerable groups, families and everyone

Social distancing measures are steps you can take to reduce the social interaction between people. The government is currently asking all people to social distance themselves. This is to help reduce the transmission of coronavirus (COVID-19). Please use the government website link below for up to date information.

<https://www.gov.uk/government/publications/covid-19-guidance-on-social-distancing-and-for-vulnerable-people/guidance-on-social-distancing-for-everyone-in-the-uk-and-protecting-older-people-and-vulnerable-adults>

Self-isolation -symptomatic and unwell or someone you live with is symptomatic

In this instance you should self-isolate at home. Please see the link to the NHS self-isolation advice below for up to date information on what this means.

<https://www.gov.uk/government/publications/covid-19-stay-at-home-guidance/stay-at-home-guidance-for-households-with-possible-coronavirus-covid-19-infection>

<https://www.nhs.uk/conditions/coronavirus-covid-19/self-isolation-advice/>

When to trigger self-isolation / shielding:

The paediatric centres will follow government advice and support patients who are shielding or choose to self-isolate based on government advice. Specialist centres may contact some families to clarify whether their underlying condition constitutes a high risk group that would benefit from shielding but this will be highly individual, patient-specific advice.

Homecare / ERT infusions

If a patient is self-isolating because of COVID-19 symptoms - nurse visits for ERT infusions are paused for the duration of self-isolation (currently 14days). If patients can become independent, that would be advantageous, but homecare resources are not available for large-scale, short-term training.

If a family chooses to strictly self-isolate because of underlying vulnerability - your specialist centre will have an individual discussion with you about ERT and the risk/benefit balance of taking a prolonged "drug holiday". They will offer reassurance to patients and families undergoing a "drug holiday" that this is a decision in their best interest, based on the greater risk to them of being infected with COVID-19, than any modest or moderate effect of missing treatment for a period of time.

If a family chooses to undertake "more stringent social distancing" because of underlying vulnerability, but not full family self-isolation, it may be appropriate for some patients to continue receiving ERT as long as homecare companies can deliver this.

This is felt to be essential for;

- Infantile LAL Deficiency
- Infantile Pompe Disease
- Type III Gaucher
- some MPS patients being prepared for stem cell transplantation.

CLN2 patients receiving intrathecal cerliponase in hospital will also continue to be, prioritised for this treatment].

If Homecare Company staffing becomes critically low, then patients with other diseases may be required to reduce their dosing frequency.

Patients with indwelling venous access devices who are having a drug holiday should have these flushed at the longest interval compatible with keeping the specific device in operation. This may appear to contradict the statement in relation to infusions, but it is a balance of risk decision based on how critical to the patient's health the individual procedural visit is.

Critically ill lysosomal disease patients should not be transferred to their LSD specialist centre unless there are clear clinical reasons for this. They should as a rule continue to receive treatment at their local hospital as appropriate.

Discussions regarding missed infusions

We will defer any discussion of catch-up infusions until the crisis is over.

Keeping in touch with self-isolating patients

We will keep in contact with self-isolated patients by regular telephone calls, the frequency to be determined by need and staff availability.

Patients enrolled on an MAA

We expect guidance and further information to be available from NICE and NHSE within the next few days.

Patients waiting to start ERT

Patients will be reviewed by their treating clinician. There may be delay in establishing patients on ERT due to overstretched resources and the risk of bringing patients into the hospital.

Patient / carer wellbeing

We acknowledge that this is a very anxious time for many of our patients, carers and their families and the wellbeing of you all is important to us. Please find attached a resource called first aid to worry which gives some helpful advice to help with those feelings on anxiety. In addition to this there is a link to mind who also have resources to support you.

<https://www.mind.org.uk/information-support/coronavirus-and-your-wellbeing/>

Please do not forget your patient support group is there to support, advise and to be a listening ear. We are all here for you.

To be reviewed on 27.03.2020

Attachment

First Aid for Worry

It is normal to feel anxious right now because worrying things are happening. This anxiety is a normal, human response that people across the country and the world are sharing right now.

You can try these things to help you to manage your worries.

- Only check the news once or twice a day. Although it can feel like you are doing something useful when you check the news, this may actually maintain your anxiety.
- Interrupt worrying thoughts by noticing: 5 things you can see, 4 things you can hear, 3 things you can feel, 2 things you can smell, 1 thing you can taste.
- Stay active. Plan small, useful tasks to do each day.
- Look out of the window. How many different shades of green can you see? Or blue? Or another colour?
- Listen to all the noises you can hear outside the room. Now listen to all the noises you can hear inside the room. Listen to the noises your body is making - can you hear your own breathing? Listen to that for a minute or two.
- When you wash your hands, do it mindfully. Notice the feeling of the warm water, the slipperiness of the soap, the sensation of one hand moving on the other and the water running over your hands. Dry them gently and notice this sensation. You can do this for other activities, such as showering or washing up.
- Talk to a friend or family member about something you both enjoy. Start off by asking to have a conversation about this, and asking not to mention the coronavirus.
- Try a breathing exercise: Breathe in for the count of four, and then out for the count of four. Do this gently for a few minutes. Notice the feeling of the air moving in and out of your nose and your chest rising and falling.
- Offer encouragement and support to someone else.

- At the end of each day, write down three things you are grateful for today.