

Update from the Paediatric LSD clinical centres

Clinical support and homecare

Updated 26 March 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 news and Government and NHS guidance:

www.gov.uk/coronavirus

Shielding high-risk patients

Clinical centres are already reviewing all their patients and will be in contact with those patients deemed to be high risk.

This link gives government advice on shielding:

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 and Self-isolation

Social distancing for vulnerable groups and families

Social distancing measures are steps you can take to reduce the social interaction between people. This will help reduce the transmission of coronavirus (COVID-19).

The government website provides up to date information:

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 for symptomatic and unwell patients, or if someone you live with is symptomatic

In this instance you should self-isolate at home.

These links provide government and NHS self-isolation advice below for up to date information on what this means.

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

www.nhs.uk/conditions/coronavirus-covid-19/self-isolation-advice/

When to trigger self-isolation and 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 and ERT infusions

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.

Family chooses to strictly self-isolate due to 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.

Family chooses to undertake "more stringent social distancing"

If the family chooses this 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 Pompe Disease**
- Infantile LAL Deficiency
- 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 having a drug holiday

These patients 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

Such children 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 and 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 the accompanying 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.

www.mind.org.uk/information-support/coronavirus-and-your-wellbeing

Patient Support Group

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.

Pompe Support Network

www.pompe.uk

01730 231554

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