

COVID-19 and Red Cell Disorders (Sickle Cell Disorders, Thalassaemia and other inherited anaemias)

We are writing to you regarding COVID-19 and what that may mean for you. Clearly this is an evolving situation and this information may change, so it is important you keep up to date with recommendations. UK government advice is available at: <https://www.nhs.uk/conditions/coronavirus-covid-19/>
<https://www.gov.uk/government/collections/coronavirus-covid-19-list-of-guidance>

Information is also available on the Sickle Cell Society (www.sicklecellsociety.org) and UK Thalassaemia Society (www.ukts.org) webpages. Please always check you are reading the most recent version of any document. Note there is separate information regarding Diamond Blackfan Anaemia.

This is an unprecedented time for the NHS. Medical and nursing teams will face intense pressures over this period. Your regular clinical staff are already being moved to look after sick adults outside of Haematology; your Consultants are working extremely hard with national colleagues to ensure the best possible care is offered to our patient cohort during this period. Most teams have very few junior doctors at present so are extremely stretched and, in some cases, unwell themselves. Our aim is to provide you with as much information as possible so you can take the right precautions regarding your own health and understand when and when not, to come to hospital. It is important that you read this advice carefully. The specialist team is available for advice but please understand they may not be able to respond quickly to anything other than urgent medical queries.

Basic information about the virus:

A novel coronavirus called SARS-CoV-2, of animal origin, has emerged and the infection called Coronavirus Diseases 2019 (COVID-19) started spreading worldwide. Incubation period from the time from exposure to symptom development is between 2-14 days. Avoiding exposure by adhering to recommended hygiene procedures, isolation of SARS-CoV-2 infected persons and social distancing are the only prevention strategies. There are no approved treatment options and there is no available vaccine, although work is being done on both.

How does the virus affect those with sickle cell disease or thalassaemia?

The knowledge we have about how COVID-19 will affect those living with Sickle Cell Disease and Thalassaemia is evolving constantly. We are in constant communication with health authorities across the globe as well as within and between our networks. In light of this, advice may change in the upcoming days, weeks and months.

Data from China and Italy suggests that children have a milder form of the disease than adults, although we do not understand why this is the case. That does not mean that the young do not get sick, only that the chance is higher the older you get. Only 2 in every 100 diagnosed cases of coronavirus in China have been in children aged <18 years. The Italian experience in Milan and Turin can be seen as model for transfusion dependent thalassaemia patients in a high-risk area. In Italy there has been limited impact in patients with thalassaemia. Similarly, the Monza haematology, oncology and BMT paediatric service has seen limited impact in children with serious haematological disorders.

We do not have enough information yet on how Covid-19 affects sickle cell patients. More information will likely be available in the next few weeks as cases increase. We are planning to collect information in collaboration with other European countries and the USA.

In England, the care of patients with haemoglobinopathies is coordinated through Haemoglobinopathy Coordinating Centres (HCCs) with guidance from the Clinical Reference Group (CRG) for Haemoglobin Disorders, part of NHS England. Your Specialist Team is represented on these groups and, as new information is available, we will update guidance for patients the North West and North Eastern/Yorkshire networks.

What You Need to Know About the Coronavirus (COVID-19)

The pandemic is real.

We are all at risk of getting infected. Individuals with sickle cell disease who get Covid-19 are at higher risk of developing sickle complications such as painful crisis and acute chest syndrome and people who have had their spleen removed have a weaker immune system which might lead to secondary infection. People with excess iron in their body (iron overload) may not be able to manage infections as well as others. Those with co-existing health problems such as diabetes, heart disease and asthma seem to be worse affected.

How can I avoid getting infected?

SHIELDING

The latest government guidance strongly advises people with serious underlying health conditions to “Shield” at home. Shielding is a measure to protect people by minimising all interaction between those who are extremely vulnerable and others. They are strongly advising people with serious underlying health conditions which put them at very high risk of severe illness from coronavirus (COVID-19) to rigorously follow shielding measures to keep themselves safe. This guidance includes people with Sickle Cell and others at high risk (see below). You may have already received a letter already from the government about this.

The HCC Leads and the CRG have issued further guidance as to which patients should practice shielding. We are recommending that these patients should be following advice on shielding:

- All patients with sickle cell disease (e.g HbSS, HbS Bthal, HbSC and so on). It does not apply to patients with sickle cell trait (sickle cell carriers)
- Patients with thalassaemia who are at particularly high risk due to severe iron overload (T2* <15 ms, Liver iron >15 mg/g DW or ferritin >3000 mg/L), those with previous or current heart complications or those who have had their spleen removed **and** have another risk factor for such as diabetes or high blood pressure.
- Patients with Diamond Blackfan anaemia who are on steroids, for treatment or replacement of low adrenal function or have immune deficiency), those who have iron overload as for thalassaemia above or who have had a BMT within 6 months or are still using immunosuppressive drugs.
- Patients with other rare inherited anaemias e.g. pyruvate kinase deficiency, congenital dyserythropoietic anaemia who have had a splenectomy **and** are at particularly high risk due to iron overload as per thalassaemia guidelines above

Patients have been contacted by letter or text from NHS England or their GPs. If you think you fall into one of the categories of extremely vulnerable people and have not received a letter or been contacted by your GP, you should contact the hospital team (see below).

Government information on how to shield is available at the link below:

<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

This is what everyone else is doing. The best way to slow the spread of the infection is to not get infected in the first place. One of the terms you may have heard is “social distancing.”

The government has announced Social Distancing measures. Please see information at the link below:

<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>

Guidelines for washing and disinfection

- Wash your hands more often than usual, for 20 seconds using soap and hot water, particularly after coughing, sneezing and blowing your nose, or after being in public areas where other people are doing so. Use hand sanitiser if that's all you have access to.
- To reduce the spread of germs when you cough or sneeze, cover your mouth and nose with a tissue, or your sleeve (not your hands) if you don't have a tissue, and throw the tissue away immediately. Then wash your hands or use hand sanitising gel.
- Clean and disinfect regularly touched objects and surfaces using your regular cleaning products to reduce the risk of passing the infection on to other people.

Common Questions

What should I do if I am unwell? How do I know if I have COVID-19?

The only way to know for sure that you have coronavirus is to get tested. However, there are currently not enough tests for everyone at the moment and testing is only available for people who are sick and in hospital. As the weeks progress, we hope that the capacity to test will increase and we will be able to get a better picture of those who are infected, who are not infected and who have been infected and are now immune.

Symptoms:

The most common symptoms of coronavirus (COVID-19) are: recent onset of a new continuous cough and/or high temperature. Patients with a fever of $>37.8^{\circ}\text{C}$ or 100°F require a clinical review, either over the phone or in person. You should call 111, inform them and then urgently contact your clinical team at the same time for review. This is because we need to know what is happening with you and also because you may have a different reason for these symptoms such as other types of infection and we don't want that to be missed by those less familiar with your disorder. We will decide what you need to do based on your symptoms and knowledge of your particular case. **Always tell NHS 111 or any hospital doctors about your blood condition.**

We will try to treat you in the community where possible but appreciate this may not always be possible or desirable. If you have these symptoms, however mild, and we tell you to stay at home, do not leave your house for 7 days from when your symptoms started unless directed otherwise by ourselves or NHS 111.

If your symptoms worsen during home isolation or are no better after 7 days, contact NHS 111 online **and** contact your clinical team. For a medical emergency dial 999. Always tell the teams about your blood disorder.

Management of acute painful crisis

You should treat your pain as usual with paracetamol and codeine. If you are experiencing severe sickle cell pain (crisis) not resolving with paracetamol and/or codeine please attend A&E as usual. You may have read that concerns have been raised whether ibuprofen may make things Covid-19 infection worse but there is enough not scientific evidence to show this at present. If you are having a crisis and have a fever or suspect Covid-19 infection, other agents e.g. paracetamol should be considered in preference to ibuprofen, if possible, until we know more. Remember to contact the haematology team if you have a fever, respiratory symptoms or anything else that is unusual. **Please do let us know if you are unwell at home or if you are admitted to hospital elsewhere.**

What should I do if someone in my household is unwell or I am well but have symptoms?

Please refer to the government guidance:

<https://www.gov.uk/government/publications/covid-19-stay-at-home-guidance>

If you live alone and you have symptoms of coronavirus illness (COVID-19), however mild, stay at home for 7 days from when your symptoms started. If you live with others and you or one of them have symptoms of coronavirus, then all household members must stay at home and not leave the house for 14 days. The 14-day period starts from the day when the first person in the house became ill.

What about my regular treatments?

Hydroxycarbamide: There is no evidence that being on hydroxycarbamide would increase risk of severe illness with COVID-19. It is important that you remain on your usual hydroxycarbamide dose to reduce the risk of crises and avoid hospital admissions. For patients in stable health, we can extend the interval between blood monitoring to avoid coming to hospital. We will advise those who need blood tests during this time how to get them. We will try and arrange for local delivery of your medicine.

If you are on hydroxycarbamide and running out of supply, please email us at least 1 week in advance on haemoglobinopathy@mft.nhs.uk

Blood transfusion: We are planning to continue giving transfusion to those who are on regular programmes. There is a lot of work going on at NHS Blood and Transplant to make sure we have enough blood as the blood supply as the number of donors may drop as people become ill. If this becomes an issue we will call you to rearrange transfusions.

Most units will call you prior to transfusions to check you are still well. If you have symptoms but are well, transfusion may be deferred. If you are unwell, they will discuss with the medical staff as to the best course of action. Please do not miss appointments.

Clinic appointments: These will be done virtually (by telephone) or delayed. Routine ophthalmology, audiology, echocardiography imaging for iron overload assessments will not be ordered unless absolutely necessary. Any urgent investigations will still be requested as required.

Trans-cranial Doppler (TCD) screening: These may also need to be postponed but we may bring in some patients for these scans. Guidance on who to needs scans has been given to paediatric teams.

Iron chelation: Routine monitoring of iron overload and chelation drugs should be continued. For patients on regular transfusions, outpatient review should be co-ordinated to take place at the same time as transfusion. We will consider if routine MRI monitoring for iron overload can be postponed (e.g. in stable patients, on long term chelation).

If a fever develops, **all chelation agents should be stopped** and you should contact your clinical team.

If you are on iron chelation and running out of supply, please email us at least 1 week in advance on haemoglobinopathy@mft.nhs.uk

What other things can I do to make social distancing more manageable and be prepared?

- Please check to see if you have sufficient supplies of medication and are not about to run out.
- We would discourage you from stockpiling large amounts of painkillers. Even if you cannot get to the hospital there are ways in which your team would be able to get a prescription to you.
- Contact your GP to ask about getting extra medications to have on hand in case. Manchester Foundation Trust is exploring home delivery of medications that we usually dispense for you.
- Be sure you have over-the-counter medicines (e.g. paracetamol and codeine)and medical supplies (e.g. tissues) to treat fever
- Make sure you have a thermometer to take your temperature.
- Take your prescribed medications unless you have been told otherwise
- Have enough household items and groceries on hand so that you will be prepared to stay at home for a period of time that could be many weeks.
- Stay in touch with others by phone or email. You may need to ask for help from friends, family, neighbours, etc. if you become ill.
- Think ahead about who will watch your children, other loved ones, or pets if you become too sick.
- Make sure your vaccinations are up to date

What about local arrangements?

Local arrangements may change. This will depend on the make-up and resources of your local team, the degree to which they are redeployed into acute frontline care. Please contact them in the usual manner, saving clinical questions for the clinical team and administrative questions for the administrative team. Given the pressures on the clinical team, we will be prioritising those with clinical rather than more general queries.

What about clinical trials?

Research is being rapidly refocused on COVID-19. Large numbers of existing clinical trials are being paused until further notice to support this effort and to allow clinically trained researchers and support staff to support front line care. You will be informed separately and individually by your research teams about this.

Contacting us

Main hospital switchboard: **0161 276 1234**

MRI Clinical Nurse Specialists: **0161 701 6683, 0161 271 4574** (9am – 5pm)

- Please leave a voice message.
- If you do not get an answer, they may be dealing with an emergency. Please leave your name phone number so they can return your call.
- Out of hours: please leave a message and they will return the call during the next working day.

Alternatively email queries to haemoglobinopathy@mft.nhs.uk and we will aim to respond by the next working day.

For urgent enquiries eg if you think you have infection and are unwell please call switch board and ask to be put through to the Haematology Registrar on 4445 bleep. Please these doctors are very busy so can only deal with urgent enquiries.

Haematology Day Unit: **0161 276 4808** or ext **64808** via switchboard