

Advice on COVID-19 in patients with Sickle Cell Disease and Thalassaemia

Haemoglobinopathy Co-ordinating Centres

17th March 2020

We recognise that the situation is changing very rapidly and as such guidance may become outdated within days. Please use NHSE (<https://www.england.nhs.uk/coronavirus/>) and PHE websites (<https://www.gov.uk/government/topical-events/coronavirus-covid-19-uk-government-response>) in addition to your local trust guidelines to provide updated guidance. All staff and patients should be advised to follow national policy guidance on hand washing, testing, self-isolation, avoiding high risk social situations and so on.

This information has been produced following virtual meetings with representatives from the HCCs and NHSBT. We will aim to update it regularly.

It will be added to a Sharepoint shortly.

Overview

Patients with both SCD and Thalassaemia are likely to be at increased risk of complications from COVID-19. Patients at highest risk include the elderly (>50 in our population), those with a history of respiratory or cardiac disease and those with other co-morbidities.

Patients should follow generic NHS advice for self-isolation if they are exposed and remain well. They should let their clinical team know.

General guidelines for management

Move to virtual consultations where possible.

Postpone non essential investigations

Patients on hydroxycarbamide or iron chelation will need regular monitoring to continue but this could be done virtually and on an extended schedule (maximum interval 12 weekly for hydroxycarbamide). Off site phlebotomy and pharmacy services should be utilised if possible.

Patients should not attend outpatients or day unit if they have temperature/respiratory/coryzal symptoms

Encourage patients to let their specialist teams know if they have symptoms or have to self-isolate ; or if they are admitted to hospital

Teams should consider if they have resources to maintain regular contact via telephone with patients who are self-isolating and setting up a generic email for patient queries, which will be manned by clinical staff.

Teams should consider setting up mechanisms of communication between nearby trusts/networks so that they can provide clinical advice in the event of staff sickness. (eg WhatsApp or email groups)

What to say to patients

Refer patients to up to date advice on the NHSE and PHE websites. There are also resources on the UKTS and Sickle Cell Society web pages (<https://ukts.org/heads-up/coronavirus-information/> and <https://www.sicklecellsociety.org/coronavirus-and-scd/>) and further resources will be added to the National Haemoglobinopathy Panel web pages as they are made available.

The following should also be discussed:

Delay non-essential travel

School: follow issued advice by the authorities as per other children

Work: explore options to work from home as appropriate

Further evidence is given here: <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> (16th March 2020)

Frequently asked questions/Specific Haemoglobinopathy issues

Sickle cell disease and Acute Chest syndrome: There is an overlap of symptoms of acute chest syndrome and COVID-19 and infection with COVID-19 may increase the risk of acute chest syndrome. Clinicians should be extra vigilant for this complication and should treat patients fulfilling the criteria for acute chest syndrome (respiratory signs and symptoms, abnormal Chest X-Ray) as per national guidance. This will include treatment with top up or exchange transfusion. There may be an increased need for emergency transfusion during this time. Services should develop plans for how they can provide emergency apheresis capacity.

Manual exchange transfusions: This may be an aerosol generating procedure. Therefore full PPE ensemble (including FFP3 respirator), long sleeved disposable gown should be used in both proven AND SUSPECTED cases who need manual exchange, until further evidence is available. There is no evidence that automated exchange transfusions are aerosol generating.

Patients with a fever: Patients with a fever of >37.8 require a clinical review, either virtually or in person. They should be advised to call 111, inform them and then urgently contact their clinical team for review. If self-isolating they should be given additional antibiotics, phone follow up should be arranged and they should be asked to present if they have worsening symptoms.

Management of acute pain: Patients should be encouraged to treat pain as usual but to contact their clinical team if they have a fever or respiratory symptoms.

Hydroxycarbamide: There is no evidence that being on hydroxycarbamide would increase risk of COVID-19 as long as there is no related myelosuppression. Patients should be urged to remain on their usual hydroxycarbamide dosages to maintain good health and avoid hospital admissions. It may be advisable to avoid routinely starting or dose escalating hydroxycarbamide to reduce need for repeated phlebotomy and hospital visits until the situation has stabilised. For stable patients it is reasonable to extend interval between blood monitoring.

Annual reviews: These should be done virtually or delayed. Routine ophthalmology, echocardiography, imaging should not be ordered at present. Urgent investigations should be requested as appropriate.

Trans-cranial Doppler (TCD) screening: These may also need to be postponed but services should consider how they can continue to provide this for essential groups. Patients needing their first TCD, patients with previous conditional TCDs, younger patients should be prioritised.

Iron chelation: Routine monitoring for iron overload and for the effects of iron chelation should be continued. For patients on regular transfusions, outpatient review should be co-ordinated to take place at the same time as transfusion. Clinicians should consider if routine MRI monitoring for iron overload can be postponed (eg in stable patients, on long term chelation). If a fever develops, all chelation agents should be stopped.

Ibuprofen: Concerns have been raised about the role of ibuprofen in this condition. In febrile patients with suspected Covid-19 infection other agents should be considered in preference to ibuprofen if possible until further evidence is available. Please refer to PHE/NHSE sites for most up to date advice.

Blood supply

NHSBT are working to maintain the blood supply and will update clinical teams if problems develop or are anticipated. At the current moment routine transfusion treatments should continue. It may be necessary to prioritise patients for transfusion and we may need to limit units of blood given, to offer top up rather than exchange transfusion or postpone non-essential transfusions. NHSBT will inform trusts when or if this becomes necessary.

HCCs and SHTs should review their lists for regular transfusion on a weekly basis with respect to latest NHSBT advice. Each team should identify which patients could be delayed/deferred if this should become necessary due to blood shortages or staff illness.

The risk of transfusion transmitted infection is currently thought to be low.

Current BSH advice recommends using blood that is <10 days old for top up transfusion and <7 days old for exchange transfusion in sickle cell disease and <14 days old in thalassaemia. As there is little evidence to support these AND as there may be blood shortages, clinicians and transfusion staff should consider relaxing these criteria for transfusion and using older blood. Using recently donated blood may be more important in emergency exchanges than in elective exchanges. Extended phenotype matching should take priority over age of blood.

Haemopoetic Stem Cell Transplantation /Gene therapy

Transplantation and gene therapy for non-urgent non-malignant conditions (SCD and thalassaemia) will be halted until the situation stabilizes.

Rare anaemias

There is currently no evidence that conditions such as pyruvate kinase deficiency will be exacerbated by Covid-19 and patients should follow national guidelines. There are specific guidelines on the management of Diamond Blackfan anaemia.

Haemoglobinopathy Co-ordinating Centres/Clinical Reference Group for Haemoglobin Disorders

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