Haemoglobinopathy HCCs response to Covid-19 V6 21st March 2020

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

Haemoglobinopathy Co-ordinating Centres

V6 21st March 2020

This information has been produced following virtual meetings with representatives from the Haemoglobinopathy Co-ordinating Centres (HCCs) and Clinical Reference Group (CRG) for Haemoglobin Disorders. This version supersedes previous drafts and will be reviewed weekly at the current time.

The most recent version will be displayed on the British Society of Haematology website (https://b-s-h.org.uk/about-us/news/covid-19-updates/) and on the NHSE website

https://www.england.nhs.uk/coronavirus/secondary-care/other-resources/specialty-guides/
Please ensure this is the most recent version before using. Please address queries to your Haemoglobinopathy Co-ordinating Centre lead. HCCs will also be able to access this via the National Haemoglobinopathy Panel SharePoint.

We recognise that the situation is changing very rapidly and as such guidance may becoming outdated within days.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.

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.

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). Offsite 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 (e.g. 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. Children with SCD are at increased risk and should not be attending school and should be socially distancing.

Work- explore options to work from home as appropriate

Social distancing: PHE advice applies to patients with Sickle cell disease – see link below. Patients with thalassaemia who are hyposplenic or if they are iron loaded (particularly cardiac iron) should be socially distancing.

Further information 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: The symptoms of acute chest syndrome and COVID-19 overlap 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. Clinicians should consider early top up transfusion if there are clinical concerns (e.g desaturation on exercise) and/or rapidly evolving chest involvement. There may be an increased need for emergency top up and exchange transfusion during this time. Services should develop plans for how they can provide emergency apheresis capacity.

Exchange transfusions: Manual exchange transfusion is not thought to be an aerosol generating procedure. If the unit has an SOP for manual exchange using a closed system, normal protection should be used. It may be advisable to use full PPE where there is no SOP and a closed system is not being used (dependent on local advice). There is no evidence that automated exchange transfusions are aerosol generating. Apheresis machines need deep cleaning after exposure to a Covid-19 patient (refer to manufacturers manual: Maintaining the Spectra Optia System, Cleaning and Disinfection).

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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 must urgently contact their clinical team for review. If they can't access their clinical teams and are acutely unwell they should attend the Emergency Department (A+E) in the usual way. If they present out of hours the patient or the admitting medical/haematology teams should be encouraged to contact the haemoglobinopathy teams. 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.

Red flag symptoms: Patients should be encouraged to attend the Emergency Department (A+E) or call 999 if any of the following occur:

- Uncontrolled pain >7/10 despite usual home analgesia
- Respiratory distress (new shortness of breath or increased breathlessness compared to baseline particularly at rest or on minimal exertion) +/- chest pain
- Persistent fever >38°
- Severe headache, confusion or neurological changes.

Clinicians should be aware that patients with SCD and Thalassaemia may present with these symptoms in the absence of Covid-19 and usual pathways for investigation and management should be followed.

Infants with SCD with fever or shortness of breath should follow standard procedure according to their providers policies; some are advised to call their local Emergency departments (A&Es) or the ward (if they have an open access policy)

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 the 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 with HbSS and SB°Thal needing their first TCD and patients with previous conditional or first abnormal TCD should be prioritised. TCDs in younger patients (especially those <10 years) should also be prioritised unless they are already on transfusion and stable, in which case their scans can be delayed. Additional advice to clinicians is currently being developed. Clinicians should discuss with their vascular

scientists about how this service can be provided. Clinicians should consider changing patients who are currently on transfusion for primary stroke prevention to hydroxycarbamide as per the TWITCH protocol.

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 (e.g. 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.

Non Transfusion Dependent Thalassaemia (NTDT): Patients who have had a splenectomy are at increased risk of Covid-19 and should follow recommendations for social distancing.

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. Patients who have had a splenectomy are at higher risk of Covid 19 and should socially isolate. 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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