Haemoglobinopathy HCCs response to Covid-19 V9 20th April 2020

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

Haemoglobinopathy Co-ordinating Centres

V9 20th April 2020

This information has been produced following virtual meetings with representatives from the Haemoglobinopathy Co-ordinating Centres (HCCs) 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We recognise that the situation is changing very rapidly and as such guidance may becoming outdated within days.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 and avoiding high risk social situations.

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. All patients with SCD and high risk patients with thalassaemia and other inherited disorders have been advised to 'shield'. Further advice on who should shield and how to shield is given here:

https://b-s-h.org.uk/media/18201/statement-from-the-haemoglobinopathy-co.pdf

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

Patients with thalassaemia and Diamond Blackfan anaemia who are not advised to shield should be stringent in socially isolating and should not go to work (even if they are key workers).

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.

Advice on Clinical Management

A) Management of patients with SCD/rare inherited anaemias and possible COVID-19 infection

It is essential that fever in patients with haemoglobinopathies is not presumed to be due to COVID-19 infection as these patients are hyposplenic and therefore also at high risk of infections from other causes.

Patients should let their specialist teams know if they have symptoms of COVID-19 (fever, cough) which they are managing at home or if they are admitted to hospital.

Patients with a fever: Patients with a fever of >37.8 require a clinical review, either virtually or in person. They should urgently contact their clinical team for review. If they can't access their clinical teams they should attend the Emergency Department (A+E) in the usual way. Standard of care for managing fever in haemoglobinopathy/SCD patients should be followed including examination, blood cultures and antibiotic therapy

The Emergency Department should test patients presenting with a fever for COVID-19

The Emergency Department/admitting medical teams should contact the haemoglobinopathy teams if any patients present to them with COVID-19 symptoms

If patients are discharged to self-isolate they should be given additional antibiotics, phone follow up should be arranged by the haemoglobinopathy team and they should be asked to present if they have worsening symptoms.

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)

B) Management of patients with SCD/rare inherited anaemia admitted to hospital with COVID-19 proven or suspect infection

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.

National Haemoglobinopathy Panel

Haemoglobinopathy HCCs response to Covid-19 V9 20th April 2020

C) Management of patients with SCD/rare inherited anaemia presenting to hospital with non-COVID-19 related 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.

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.

D) Management advice for outpatient care

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 4-6 monthly for hydroxycarbamide).

Offsite phlebotomy services should be utilised if possible.

Home delivery pharmacy services should be utilised if possible

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

Teams should set up a generic phone advice and 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).

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^{o}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.

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.

D) Management of Thalassaemia and Rare anaemias

Thalassaemia: Patients with thalassaemia who are at particularly high risk from COVID-19 will include those due to iron overload (T2* <15 ms, previous or current impaired LV function or other cardiac complication related to thalassaemia, severe iron overload LIC >15 mg/g DW or ferritin >3000 mg/L) or those with a splenectomy **in combination with** another risk factor for complications e.g. diabetes. Additional guidance is available on the management of patients with cardiac iron overload https://b-s-h.org.uk/media/18229/iron-chelation-therapy-covid-version-2-150420.pdf

Diamond Blackfan Anaemia: Additional advice has been produced on the management of patients with Diamond Blackfan Anaemia.
https://b-s-h.org.uk/media/18174/nhp-covid-19-dbaversion-224032020.pdf

Patients with Diamond Blackfan anaemia are at higher risk of complications from COVID-19 if they are on steroids as per NHP (National Haemoglobinopathy Panel) guidance (see link above), with an associated immunodeficiency (or due to age have not yet been assessed), have adrenal insufficiency on steroid replacement, have iron overload as per thalassemia criteria above or have had a BMT within two years or are still using immunosuppressive drugs.

Patients with other rare inherited anaemias e.g. pyruvate kinase deficiency, congenital dyserythopoietic anaemia are at increased risk of complications from COVID-19 if they have had a splenectomy **and** are at particularly high risk due to iron overload as per thalassaemia guidelines above.

National Haemoglobinopathy Panel

Haemoglobinopathy HCCs response to Covid-19 V9 20th April 2020

E) Advice on 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 and the clinical team immediately contacted for advice.

More detailed advice is available for patients with cardiac iron overload.

https://b-s-h.org.uk/media/18229/iron-chelation-therapy-covid-version-2-150420.pdf

F) Advice on Blood Transfusion

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

Exchange transfusions: There is no evidence that automated exchange transfusions are aerosol generating. The Spectra Optia should be cleaned following the regular procedures delineated in the user manual on the Terumo website: https://www.terumobct.com/, any further information please contact Terumo directly.

Manual exchange transfusions should be performed as a closed procedure, in which case it is not thought to be an aerosol generating procedure and the risk of blood spillage is low so standard protection should be used. If a closed system is not being used then there may be a risk of blood spillage and PPE may be necessary in line with trust guidance.

Blood supply: NHSBT are working to maintain the blood supply. Information from NHSBT regarding blood provision during the COVID-19 pandemic can be seen on the hospital and sciences website: https://hospital.blood.co.uk/business-continuity/coronavirus-covid-19/

An additional document has been produced in conjunction with NHSBT on supporting the transfusion needs of patients with inherited red cell disorders during the COVID-19 pandemic in England

https://b-s-h.org.uk/media/18230/supporting-the-transfusion-needs-of-patients-with-inherited-red-cell-disorders-during-the-covid-19-pandemic-in-england-v15420.pdf

This describes a phased approach to the pandemic. At the current moment routine transfusion treatments should continue. HCCs and SHTs should review their lists for regular transfusion on a weekly basis with respect to latest NHSBT advice and advice on shielding. The first phase includes advice about optimising blood use by relaxing advice on age of blood, using depletion exchange,

introducing hydroxycarbamide and optimising intervals between Group and Save samples and transfusion.

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. This is discussed in Phase 2 of the document. Each team should identify which transfusions could be delayed/deferred if this should become necessary due to blood shortages or staff illness.

G) Management of patients who have had a Haematopoetic 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. Pre-transplant workup investigations should be put on hold.

Patients with thalassaemia/rare red cell anaemias who have had a Haematopoietic Stem Cell Transplant or Gene Therapy/Editing within the last two years are at increased risk of complications from COVID-19 infection and should be advised to shield. Patients wish sickle cell disease who have had Haematopoietic Stem Cell Transplant or Gene Therapy/Editing or any patient who has had a Haematopoietic Stem Cell Transplant and is still on immunosuppression are advised to shield regardless of the time that has elapsed since the procedure.

Haemoglobinopathy Co-ordinating Centres

20th April 2020