

Guidance regarding iron chelation therapy in patients haemoglobin disorders or rare anaemias and cardiac iron overload if unwell with presumed or confirmed COVID-19 infection

Consensus document from the Haemoglobinopathies Co-ordinating Centres.

There **is a small number of patients with haemoglobin disorders or rare anaemias who have myocardial iron overload** and are at high risk of cardiac decompensation in the context of high fever.

This is a consensus view on how to manage these high risk patients. Most centres will have 2 or 3 patients in this situation only:

Each patient should be managed on a case by case basis and specialist advice should be sought through the HCC thalassaemia lead for your region.

General advice for patient with thalassaemia:

Patient is isolating at home with presumed COVID infection

The general recommendation for all transfusion dependant thalassaemia major patients has been to interrupt iron chelation therapy if the patient is febrile until the fever has resolved and/or the cause of fever has been medically assessed. However there are circumstances under which stopping chelation in the context of fever can be harmful, particularly when cardiac iron is increased, so each case must be reviewed by a clinician familiar with thalassaemia and chelation management.

All patients who are self isolating due to symptoms or an infected household member will need to contact their transfusion team and let them know they are in isolation so that transfusion therapy can be planned appropriately. Patients self isolating will not know if they have COVID-19 or not at the moment.

Patients with Cardiac T2* < 10 ms and proven or suspected Covid-19 infection:

Patient is on Intravenous iron chelation therapy through a central venous access device or on oral iron chelation (monotherapy or combination therapy)

Daily contact with patients by specialist team either via email or telephone.

- If symptoms mild and patient not febrile advise to continue chelation and liaise with team on a daily basis.
- If patient has fever > 37.8 °C then they should be admitted:
 - Cardiac status must be assessed
 - Focused echo for LV function & TRjet Vmax (if any TR)
 - BNP if symptoms of breathlessness (a negative BNP rules out cardiac failure)
 - Baseline ECG
 - Other causes of fever should be ruled out as per normal recommendations in TM such as infected lines, Gram negative bacteraemia or Yersinia infection.

- If fever is due to above then manage as per local protocols and guidelines for acutely unwell thalassaemia patients.
- **Patients where other causes are excluded and are COVID-19 POSITIVE** (*this information may not be available immediately and may take several days*) should continue on intravenous iron chelation with desferrioxamine and stop the oral iron chelation medication until the infection resolves.
- Careful monitoring is needed of patients clinical state and the risk of cardiac decompensation is high if T2* is particularly low or if pre-existing low ejection fraction.

Cardiac T2* > 10-20 ms and proven or suspect Covid-19 infection

- If symptoms are mild and no fever then advise to continue iron chelation and daily contact with the team.
- **If febrile they should present for face to face assessment at the hospital using the processes defined by the centre and possibly a COVID swab.**
- If no cardiac symptoms and cardiovascularly stable, then patients should aim to continue chelation therapy as soon as they have been assessed by their specialist centre and acute bacteraemic infection has been reasonably excluded
- If patient develops palpitations, ankle swelling and worsening shortness of breath then to contact their team and attend hospital as a matter of urgency, be admitted and managed as per those patients with T2* <10 ms.
 - Echo, ECG & BNP at baseline

Pathway for advice for sick patients with Transfusion dependent thalassaemia and at risk of heart failure:

SHT lead to contact the HCC for Thalassaemia and speak to haemoglobinopathy consultant on call out of hours or directly with the lead for thalassaemia dependant on local protocols.

This document was produced by a sub-group of specialists from the Haemoglobinopathy Co-ordinating Centres. Additional comments from Professor Malcolm Walker (UCLH)