

Statement from the Haemoglobinopathy Co-ordinating Centres for Haemoglobin Disorders

The latest government guidance strongly advises people with serious underlying health conditions to rigorously follow shielding measures.

This guidance includes 'People with rare diseases and inborn errors of metabolism that significantly increase the risk of infections (such as SCID and homozygous sickle cell).

<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>

We have reviewed this guidance and offer clarification on groups of patients who should be included.

In line with government guidance 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 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.
- Patients with Diamond Blackfan anaemia who are on steroids as per NHP guidance, with an associated immunodeficiency (or due to age have not yet been assessed), have adrenal insufficiency on steroid replacement, have iron overload as per thalassaemia criteria above or 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 are currently being contacted by letter or text. The guidance states that if patients think they fall into one of the categories of extremely vulnerable people and have not received a letter by Sunday 29 March 2020 or been contacted by their GP they should discuss their concerns with their GP or hospital clinician. It is likely therefore that haematologists will be contacted by patients for clarification of their status (as many of you have been already). If you are asked to decide if a patient should be shielded or not, then please use the advice above. If you have further queries, please contact your haemoglobinopathy co-ordinating centre.

If individual trusts would like to send out the letters to their patients this can be done using the appropriate template, alternatively they may wish to ask their patients to contact them after Sunday 29 March 2020 if they have not had a letter and think they should have had one.

In view of the nuanced advice for patients with thalassaemia and Diamond Blackfan Anaemia we suggest clinicians proactively contact patients within these groups who they feel are at highest risk and require shielding.

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) or to school.

Haemoglobinopathy Co-ordinating Centres for Haemoglobin Disorders