



Haemoglobinopathy HCCs advice on risk assessment COVID-19 28.08.20

# Advice to clinicians on risk assessment for severe COVID-19 in patients with haemoglobinopathies and inherited rare anaemias

This information has been produced following virtual meetings with representatives from the Haemoglobinopathy Co-ordinating Centres (HCCs) for Haemoglobin Disorders, the Clinical Reference Group for Haemoglobin Disorders, the National Haemoglobinopathy Panel, NHS Screening Committee and NHSBT as well as national experts on Sickle Cell Disease, Thalassaemia and Rare Anaemias and patient groups.

This updates previous advice from this group and should be read in conjunction with government guidance advising that shielding should be paused from 1<sup>st</sup> August 2020. This document offers advice to Haemoglobinopathy practitioners if the situation arises that shielding is re-introduced. It also provides support for clinicians in having conversations about COVID-19 risk with patients.

This document covers

1. Background information
2. Guidance for discussions with children
3. Planned changes in risk assessment for adults
4. Guidance for discussion with adults
5. Advice to patients on employment/education
6. Additional patient information

It should be read in conjunction with 'Advice to clinicians on planning recovery from COVID-19' 26.06.2020. REF

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## *1 Background information*

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From March 2020 certain groups of 'clinically extremely vulnerable' people were advised to 'shield' to reduce their risk of COVID-19. The National Haemoglobinopathy Panel advised that all people with Sickle Cell Disease (SCD) and some of those with Thalassaemia and Rare Anaemias should shield. <https://b-s-h.org.uk/media/18244/hbp-hccs-response-to-covid-v9-200420.pdf>

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From the 1st August shielding was paused and those shielding have been encouraged to cautiously increase activity outside the home whilst maintaining social distancing. This governmental guidance is being regularly updated and the latest advice can be viewed here.

<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>.

Updated advice from August 13<sup>th</sup> states that:

- Patients may be advised to shield again if the situation changes and there is an increase in the transmission of COVID-19 in the community
- This may include local lockdowns
- The risks of serious illness from COVID-19 is low for most children and young people. Children and their families have been advised to speak to their paediatric specialist or general practitioner over the summer to discuss if they should be removed from the shielding list
- All members of the household of those who are identified as 'clinically extremely vulnerable' at the time of the flu vaccination programme are eligible for a free flu vaccination

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### *2 Guidance for discussions with children*

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The government advice says that evidence indicates that the risk of serious illness for most children and young people is low. In the future they expect fewer children and young people will be included on the shielded patient list. If a child or young person is removed from the shielded patient list, they will no longer be advised to shield in the future if coronavirus transmission increases.

The Royal College for Paediatrics and Child Health has produced some updated guidance on shielding (June 12<sup>th</sup> 2020) which should be used for discussions with children and their families and advises that clinicians should discuss this with their patients over the summer.

<https://www.rcpch.ac.uk/resources/covid-19-shielding-guidance-children-young-people>

This states that in principle:

- A small group of children who are 'clinically extremely vulnerable' due to their pre-existing condition will need to continue to shield.
- A further larger group of children exists who, due to their underlying condition, may need to shield. The decision to continue to shield would normally result from a discussion between the clinician, the child and their family.
- Splenectomy alone is not an indication for shielding

Conditions listed in the **red** categories confer the highest risk and individuals should be advised to continue to 'shield'.

Conditions listed in the **amber** categories below confer a less well defined risk and will require a case-by-case discussion to decide whether, on the balance of risks, an individual should be advised to

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continue shield. Not all of those in the categories listed below will need to shield. If following a discussion, the individual is advised not to shield, they should maintain stringent social distancing.

Individual risk assessment on which shielding advice is based should be undertaken by an experienced member of the local or specialist haemoglobinopathy clinical team (medical staff or clinical nurse specialists). This may be facilitated by a screening tool or checklist to ensure consistency. Input from other clinical specialties should be sought where relevant particularly if decisions on shielding are equivocal. It is recommended guidance on shielding is communicated verbally in the first instance and a record of the discussion documented in the patient's health record. A letter confirming the advice given should be sent to the patient/parent(s) and copied to the GP.

Conditions listed in the categories below will require a case-by-case discussion to decide whether, on the balance of risks, a child should be advised to continue shield. Not all children and young people in the categories listed below will need to shield. If following a discussion, a child is advised not to shield, they should maintain stringent social distancing.

Any decision on shielding should balance the clinical and social impact of shielding - weighing the benefit of keeping children and young people with underlying co-morbidities safe whilst protecting the most socially vulnerable, due to family and social circumstances, who may risk additional harm from continued shielding. There may be other patients who do not fit these categories below, but for whom secondary care clinicians feel, after discussions with families, that shielding is advisable. We advise contacting their tertiary specialists for advice.

	<b>Clinically extremely vulnerable: shielding strongly recommended</b>	<b>Case-by-case discussion on possibility of continuing to shield</b>	<b>Shielding no longer recommended</b>
<b>Sickle cell disease</b>	Children who have had a BMT within one year or are on continuing immunosuppression	Additional co-morbidities of concern e.g progressive critical neurovasculopathy, severe or symptomatic heart failure  A history, within the preceding 12 months, of either one or more chest crisis requiring ITU treatment or two of more chest crises requiring treatment	All other children with sickle cell disease

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<b>Thalassaemia</b>	Children who have had a BMT within one year or are on continuing immunosuppression	Severe cardiac iron overload (T2 * $<10$ ms) plus additional co-morbidity causing concern (eg splenectomy)	All other children with thalassaemia
<b>DBA</b>	Children who have had a BMT within one year or are on continuing immunosuppression	Associated immunodeficiency, severe iron overload (as per thalassaemia definition) or are on prednisolone (or equivalent) $\geq 0.5$ mg/kg/day	All other children with DBA
<b>Other rare inherited anaemias, e.g. pyruvate kinase deficiency, congenital dyserythropoietic anaemia</b>	Children who have had a BMT within one year or are on continuing immunosuppression	Severe cardiac iron overload (T2 * $<10$ ms) plus additional co-morbidity causing concern (eg splenectomy)	All other children with inherited anaemia

Clinicians should discuss with children and their families/carers their removal from the shielding list and update the shielding lists accordingly. Patients can only be removed from the shielding patient list by their GP or specialist, following consultation (via letter, phone call or face to face discussion) with the child and their family, and other clinicians where appropriate.

It is likely that the majority of children and young people with haemoglobinopathies and inherited anaemias will be removed from the shielding lists and will NOT need to shield if shielding is re-introduced. It is essential that the shielding lists are kept up to date to ensure we don't cause harm by keeping children away from home unnecessarily.

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### *3 Planned changes to risk assessment in adults*

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Current advice for adults (as of August 13<sup>th</sup> 2020) is that patients who are on the list of 'clinically extremely vulnerable' could be advised to shield again if there is an increase of COVID-19 in the community. This could be national advice or may be local advice if there are local lock-downs.

The government are developing a national risk assessment tool for adults. This is based on large data sets that identify risk factors for severe COVID-19 infection. This will include factors such as age, renal disease, diabetes and cardiovascular disease. Sickle cell disease will also be included as a risk factor. Patients will be given a risk score based on all of these risk factors. It is expected that this tool will be available in September and once this is available clinicians who look after adults with Haemoglobinopathies and Rare Anaemias will need to work with their patients to produce a risk score and provide advice on risk.

The protocol of this risk assessment tool is given here.

<https://www.medrxiv.org/content/10.1101/2020.06.28.20141986v1.full.pdf>



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This tool is based on large data sets and as we look after rare conditions and rare complications of these conditions, these may occur too infrequently to influence this national tool. We have developed some guidance on rare complications which should be taken into consideration when discussing risk scores. This is based on data from national data collection (Telfer et al 2020 Haematologica in press), from observational data and from expert opinion.

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### *4 Guidance for discussions with adults*

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Whilst we wait updated advice on the updated risk assessment tool, it is important to remember that **advice on 'clinically extremely vulnerable' patients is advisory rather than mandatory**. The reason for the advice is to keep patients safe and allow them to best protect themselves from the risk of COVID-19.

We recognise that shielding and social distancing are going to become more and more challenging over time for social, employment and psychological well-being. They will also impact on the need of patients to access long term health care. The challenge is how we balance the ongoing risk of COVID-19 with the risks of shielding/social distancing. People will need to make complex assessments about the ongoing risks and benefits of shielding and social distancing and make a decision that feels right for them. This advice will take into consideration the growing experience of dealing with COVID-19 in this patient population and aims to help clinicians and patients to assess risk.

Haemoglobinopathy clinicians will need to advise their patients in two ways. Firstly, we will need to help interpret the new guidance (when available) with respect to each patient's individual clinical scenario and advise them whether they fall into the higher risk category or not. Secondly, we will need to have conversations with them interpreting the impact of this clinical risk with respect to their social, educational, employment and mental health needs. Individuals should then be able to consider their own risk profile depending on age, ethnicity and their underlying condition, take into consideration their own personal circumstances and their own risk appetite and balance this against the current risk in the community (which may change).

We have listed below factors which (on current evidence) confer increased risk of serious effects of COVID-19 and may be helpful in supporting discussions about clinical risk. It is likely that these will be updated as more evidence becomes available.

Current evidence shows that the largest risk factor for serious outcomes from COVID-19 is age. Among people already diagnosed with COVID-19, people who were 80 or older were seventy times more likely to die than those under 40. Risk of dying among those diagnosed with COVID-19 was also higher in males than females; higher in those living in the more deprived areas than those living in the least

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deprived; and higher in those in Black, Asian and Minority Ethnic (BAME) groups than in White ethnic groups. This latter fact is of particular importance as almost all patients with sickle cell and thalassaemia are within the BAME population and hence are at increased risk from COVID.

Public Health England have produced a document 'Disparities in the risks and outcomes of COVID-19.' 2<sup>nd</sup> June 2020. <https://www.gov.uk/government/publications/covid-19-review-of-disparities-in-risks-and-outcomes>

Conditions listed in the **red** categories confer the highest risk of serious effects of COVID-19.

Conditions listed in the **amber** categories below are likely to cause an increased but less well defined risk and will require a case-by-case discussion.

Conditions listed in the **green** categories are unlikely to cause an increased risk of serious effects of COVID-19

	<b>Highest risk of serious effects of COVID-19</b>	<b>Likely increased risk of serious effects of COVID-19</b>	<b>Not likely to cause increased risk of serious effects of COVID-19</b>
<b>General risk factors</b>	<p>Organ transplant recipient; on chemo or radiotherapy; being treated for a haematological cancer; HSCT in past 12 months/on immunosuppression;</p> <p>Severe lung condition (cystic fibrosis, severe asthma or severe COPD); on</p> <p>On immunosuppressants (such as high doses of steroids); have a serious heart condition and pregnant;</p> <p>ESRF on renal replacement therapy</p>	<p>Diabetes; IHD;</p> <p>Obesity (BMI 40+);</p> <p>Increasing age**;</p> <p>Liver disease (hepatitis, cirrhosis);</p> <p>HTN</p> <p>Pre-existing cardiac disease</p> <p>Auto-immune disease, on immunosuppression</p>	Splenectomy
<b>Sickle cell disease</b>	Severe cardiac iron overload (T2 * < 10 ms) AND additional co-morbidity (e.g. diabetes, chronic liver disease)	<p>Patients 50 years and above</p> <p>CKD3 and over;</p> <p>Pulmonary hypertension by echo criteria &gt; TRV max &gt;2.5ms; (NB &gt;2.9 ms places patients at most risk)</p>	

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		<p>Chronic sickle lung – on ambulatory oxygen;</p> <p>Liver – sickle hepatopathy</p> <p>Acute sickle presentation requiring ICU support in past 12 months</p> <p>Two or more chest crises requiring treatment in the past 12 months</p> <p>Iron overload as per thal guidelines</p> <p>Severe neurological disease eg progressive critical neurovasculopathy,</p>	
<p><b>Thalassaemia</b></p>	<p>Severe cardiac iron overload (T2* &lt; 10 ms) AND additional co-morbidity (eg diabetes, chronic liver disease)</p>	<p>Patients 50 years and above;</p> <p>Severe cardiac iron overload T2* &lt; 10ms, with no additional co-morbidities and adherent with therapy;</p> <p>Cardiac iron overload (T2* 10-20ms);</p> <p>Severe - moderate iron overload (LIC &gt; 30mg/g dw and cardiac T2* &gt; 10 ms)</p> <p>PLUS additional comorbidity</p>	<p>Splenectomy</p>

DBA	Patients who have had a BMT within one year or are on continuing immunosuppression	Associated immunodeficiency, severe iron overload (as per thalassaemia definition) or are on prednisolone (or equivalent) $\geq 20$ mg/day*	
Other rare inherited anaemias, e.g. pyruvate kinase deficiency, congenital dyserythropoietic anaemia	If they are at particularly high risk due to iron overload as per thalassaemia guidelines above.		Splenectomy

\*Most patients with DBA responsive to steroids are on doses of prednisolone  $\leq 5$  mg/kg or  $\leq 20$  mg on alternate days and therefore do NOT fall in this group.

\*\*Age >70 years has been highlighted in the general population, but in view of the high rates of comorbidities in this patient group, age >50 years is likely to be associated with increased risk.

Pregnant patients should be managed in accordance with the RCOG and government guidance, with updated risk assessments undertaken with employers. It is acknowledged that evidence is only just being gathered in this population hence this criteria will remain under regular review and be updated based on it.

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### *5 Advice to patients on employment/education*

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Patients may also have risk factors relating to their work/education environment. **For all patients clinicians should review continue social distancing, discuss their commute and work/school environment and the potential risks:**

- a. Patient in a public/client facing/interacting: such as healthcare, education, travel, childcare
  - i. OH assessment advised
  - ii. Recommend as per PHE website – to work from home/redeployment to a less client facing area.
  - iii. If patient willing to accept risk and return to work suggest we advise use of appropriate PPE.
- b. Travel
  - i. Avoid public transport if possible
  - ii. If must use public transport then consider travel outside peak hours
  - iii. Avoid transiting through busy thoroughfares (e.g. shopping malls at peak times)
- c. Parents with children returning to school – await government advice for Sept:
  - i. Per government advice continued social distancing,
  - ii. Discuss staggered/distanced drop off for very young with the schools



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- iii. Continued good hygiene measures hand washing etc
- iv. Keep outer wear separate

As lockdown is relaxed it is likely that individuals will need to return to work or education. National recommendations will need to be followed but general principles are discussed below.

### *Employment*

Workers should work from home if they are able to do so and employers are encouraged to support this. Once shielding of highly vulnerable individuals is stopped and for non-shielded patients, if patients decide to return to work employers should make special accommodations for vulnerable individuals at work if working from home is not possible. This will include robust social distancing within the work environment, the use of teleconferencing and the use of protective equipment. As restrictions are lifted vulnerable individuals will be able to resume working normally but will need to maintain physical distancing while in public places.

### *Education*

Children of school age should be following national recommendations on school attendance

### *Carers*

In some cases carers of highly vulnerable children or adults, particularly those in high risk occupations, have asked to or been advised to work from home whilst family members are shielding. As restrictions are lifted they should continue to follow national recommendations and should discuss concerns with health professionals. Once shielding of highly vulnerable individuals is stopped and for carers of non-shielded patients, as they return to work employers should make special accommodations including robust social distancing within the work environment, the use of teleconferencing and the use of protective equipment. As restrictions are lifted these carers will be able to resume working normally but will need to maintain physical distancing while in public places. Non-shielded individuals should avoid public transport whilst safety concerns remain.

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## ***6 Additional patient information***

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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.