Haemoglobinopathy HCCs update COVID-19 05.11.20

Updated advice to clinicians regarding COVID-19 in patients with haemoglobinopathies and inherited rare anaemias

Summary

- Children with sickle cell disease (SCD), thalassaemia and other inherited anaemias and most adults with thalassaemia and other inherited anaemias are not 'clinically extremely vulnerable' and do not need to shield unless they have significant complications and have been specifically identified by their clinical team. They should attend school normally and follow the recommendations for other children in their school.
- All adults with sickle cell disease (SCD), small numbers of children with SCD and small numbers of those with thalassaemia and other inherited anaemias are 'clinically extremely vulnerable'. Patients on the 'clinically extremely vulnerable' list should receive an e-mail or letter from the Government confirming their status and containing further advice.

This updates previous advice from this group from 19th October 2020.

This document covers

1. Updated advice on shielding
2. Advice on children and young people
3. Guidance for discussions with adults
4. Patient information

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 and the National Haemoglobinopathy Panel as well as national experts on Sickle Cell Disease, Thalassaemia and Rare Anaemias.

This document refers to updated governmental guidance from 4th November 2020 on shielding and protecting people who are clinically extremely vulnerable from COVID-19. 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

1 Updated advice on shielding

Following the announcement for new national restrictions from 5th November 2020, all individuals in England have been advised to stay at home except for specific purposes. Additional advice has been

issued for those who are 'clinically extremely vulnerable' and at highest risk of becoming very unwell if they catch COVID-19.

This is not a return to the very restrictive shielding advice issued earlier in the year but patients are urged to follow extra precautionary shielding measures to help keep themselves safe. This remains advice, not the law. This advice applies from 5th November to 2nd December 2020 or until the lockdown is stopped.

The main advice is:
Work: You are strongly advised to work from home. If you cannot work from home, you should not attend work for this period of restriction. The formal shielding letter from the government may be used as evidence for your employer.

Socialising: You should stay at home as much as possible, except to go outdoors for exercise or to attend essential health appointments. You may meet with one other person from outside your household or support bubble but try to keep contact to a minimum and avoid busy places.

Education: There is a very low risk of children becoming very unwell from COVID-19 and most children originally identified as clinically extremely vulnerable no longer need to follow this advice. Families should consult their specialist to discuss if their child is still classified as clinically extremely vulnerable. Very few children with haemoglobin disorders and inherited anaemias will be classified as clinically extremely vulnerable.
Children who live with someone who is clinically extremely vulnerable should still attend school

Travel: You should avoid all non-essential travel by private or public transport.

Shopping: You are advised not to go to shops. Shop online or ask others to collect shopping for you.

Further information on how to access support can be found in the letter or email that will be sent to patients or at:
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

2 Advice for children and young people

The government advice says that evidence indicates that the risk of serious illness for most children and young people is low. The evidence in children and young adults with haemoglobinopathies has also shown this. Children and their families were advised to speak to their paediatric specialist or general practitioner over the summer.

The Royal College for Paediatrics and Child Health has regularly updated guidance on shielding for children < 18 years, most recently on 22.09.20. The guidance can be seen here.

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

Paediatricians have been asked to review the lists of patients classified as clinically extremely vulnerable and remove patients who are inappropriately listed.

Haemoglobinopathy HCCs update COVID-19 05.11.20

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). The discussion with the children and their families should be clearly documented. Further advice is available in the risk assessment document from 28.08.20 https://static1.squarespace.com/static/5e8ca9bcda00561f349fa870/t/5f4e0a27ad09bd44e5b86ef7/1598949931180/HBP+HCCs+advice+on+risk+assessement+28+08+20.pdf

The majority of children and young people with haemoglobinopathies and inherited anaemias have been removed from the shielding lists and are no longer classified as 'clinically extremely vulnerable'. They will NOT need to shield. It is essential that the shielding lists are kept up to date to ensure we don't cause harm by keeping children away from school unnecessarily.

Children and young people or their families who are unclear about their shielding status should contact their GP or their specialist team for discussion.

Children and young people with haemoglobinopathies and rare anaemia should continue to follow the advice issued to the public as a whole.

We encourage young people who are in further education to work with their local university/education provider to maintain their education in a Covid secure way and clinicians should support them in assessing their individual risk.

---

3 Guidance for discussions with adults

Adapted from advice issued on 28.08.20
https://static1.squarespace.com/static/5e8ca9bcda00561f349fa870/t/5f4e0a27ad09bd44e5b86ef7/1598949931180/HBP+HCCs+advice+on+risk+assessement+28+08+20.pdf

The 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 our patient population is heterogeneous and that levels of risk will vary between patients. Haemoglobinopathy clinicians will need to assist patients in interpreting their personal level of 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, taking into consideration their own personal circumstances and their own risk appetite. This guidance aims to help clinicians to support patients with decision making.

The following table demonstrates which groups are at highest risk of severe COVID-19 infection from current evidence. Other risk factors include increased age, male sex, living in more deprived areas and Black, Asian and Minority Ethnic (BAME) groups.

Haemoglobinopathy HCCs update COVID-19 05.11.20

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

National Haemoglobinopathy Panel

Haemoglobinopathy HCCs update COVID-19 05.11.20

		Iron overload as per Thal guidelines Severe neurological disease eg progressive severe cerebrovascular disease,	
Thalassaemia	Severe cardiac iron overload (T2 *< 10 ms) AND additional co-morbidity (eg diabetes, chronic liver disease)	Patients 50 years and above*; Severe cardiac iron overload T2*< 10ms, with no additional co-morbidities and adherent with therapy; Cardiac iron overload (T2* 10-20ms); Severe - moderate iron overload (LIC > 30mg/g dw and cardiac T2* > 10 ms) PLUS additional comorbidity	Splenectomy
DBA	Patients who have had a BMT within one year or are on continuing immunosuppression	Patients 50 years and above*; associated immunodeficiency**, severe iron overload (as per thalassaemia definition) or are on prednisolone (or equivalent) ≥20 mg/day***	
Other rare inherited anaemias, e.g. pyruvate kinase deficiency, congenital dyserythopoietic anaemia	If they are at particularly high risk due to iron overload as per thalassaemia guidelines above.		Splenectomy

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

**** as per PID UK recommendations: https://www.ukpin.org.uk/docs/default-source/default-document-library/ukpin_risk_stratification_covid19_finalac6baa9cd4eb6fe9b40eff00005026c1.pdf

***Most patients with DBA responsive to steroids are on doses of prednisolone ≤0.5 mg/kg or ≤20 mg on alternate days and therefore do NOT fall in this group.

Pregnant patients should be managed in accordance with the RCOG and government guidance, with updated risk assessments undertaken with employers on it.
https://www.rcog.org.uk/globalassets/documents/guidelines/2020-10-14-coronavirus-covid-19-infection-in-pregnancy-v12.pdf

4 Patient information

Patient resources:

Gov.uk guidance: 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

UKTS: https://ukts.org/heads-up/coronavirus-information/

Sickle cell society: https://www.sicklecellsociety.org/coronavirus-and-scd/

DBA: http://diamondblackfan.org.uk/2020/03/covid-19-guidance-for-patients-with-diamond-blackfan-anaemia/

Oxford University Hospitals: Covid-19: Day to day Life: making decisions about your risk of exposure
https://www.ouh.nhs.uk/patient-guide/leaflets/files/68440Plife.pdf

Additional staff resources:

National Haemoglobinopathy Panel website https://www.nationalhaempanel-nhs.net/publications/ijlpzyz21d9qy0jdj0byy055d1fcha

The British Society for Haematology https://b-s-h.org.uk/about-us/news/covid-19-updates/.

Public Health England 'Disparities in the risks and outcomes of COVID-19'
https://www.gov.uk/government/publications/covid-19-review-of-disparities-in-risks-and-outcomes