



COVID-19 Guidance for Patients with Diamond Blackfan anaemia

This document is a collaboration between the Haemoglobinopathy Co-ordinating Centres in England, offering guidance to patient with Diamond Blackfan anaemia (DBA) and their carers. It has been reviewed and agreed by representatives of the Haemoglobinopathy Co-ordinating Centres in England and the Clinical Reference Group for Haemoglobin disorders and DBA(UK).

Date: 04/06/2020 Version 4.0

What is Coronavirus and COVID-19?

Coronaviruses are a large family of viruses, which may cause illness in animals or humans. In humans, several coronaviruses are known to cause respiratory infections ranging from the common cold to more severe diseases such as Middle East Respiratory Syndrome (MERS) and Severe Acute Respiratory Syndrome (SARS). The most recently discovered coronavirus is called SARS-CoV-2 and causes coronavirus disease COVID-19.

The transmission mode is by spreading from person to person through:

- respiratory droplets spreading when coughing or sneezing
- close personal contact with an infected person (shaking hands or touching)
- touching something with the virus on it and then touching your eyes, nose or mouth with unwashed hands

What should I do?

It is essential to minimize its spread by taking the following measures:

- wash your hands with soap and water often – do this for at least 20 seconds
- always wash your hands when you get home or into work/school
- use hand sanitiser gel if soap and water are not available
- cover your mouth and nose with a tissue or your sleeve (not your hands) when you cough or sneeze and put used tissues in the bin immediately and wash your hands afterwards

General Guidance

Follow NHS England advice on <https://www.nhs.uk/conditions/coronavirus-covid-19/> and Public Health England on <https://www.gov.uk/government/publications/covid-19-guidance-on-social-distancing-and-for-vulnerable-people/guidance-on-social-distancing-for-everyone-in-the-uk-and-protecting-older-people-and-vulnerable-adults> and UK Government Guidance <https://www.gov.uk/government/publications/staying-alert-and-safe-social-distancing/staying-alert-and-safe-social-distancing>.

All patients with DBA are advised to adopt Social Distancing Measures. In addition:

- Discuss with lead medical provider possibility of remote consultations and postponement of routine monitoring tests and clinical consultations that are not essential.

- We encourage patients to let their specialist teams know if they have symptoms or have to self-isolate or, if they access 111, emergency services or are admitted to hospital.

The NHP group has carried out a real-time survey of confirmed and suspect COVID-19 cases and their outcomes in patients with red cell disorders. To the end of May, there have not been any reported patient with DBA in England affected by COVID19 and based on our data there is no good evidence that children with rare anaemias are at increased risk of severe complications of COVID-19. This is consistent with the finding of other international groups. As expertise has been accrued and the situation changes, the previous guidance has been updated.

Am I or is my child in the clinically extremely vulnerable group?

The Government guidance recommends shielding for the clinically extremely vulnerable group. Shielding is a practice used to protect extremely vulnerable people from coming into contact with coronavirus. This group includes patients in the following categories:

- Patients on high doses of steroids sufficient to significantly increase the risk of infection. This means taking prednisolone (or equivalent) ≥ 0.5 mg/kg/day or ≥ 20 mg per day. **Most patients with DBA responsive to steroids are on doses of prednisolone ≤ 5 mg/kg or ≤ 20 mg on alternate days and therefore do NOT fall in this group.**
- Patients with an associated cellular or humoral immunodeficiency, or if infants or too young to have their immune status assessed.
- Severe iron overload defined as cardiac T2* < 10 ms AND additional co-morbidity (diabetes, chronic liver disease).
- Patients who have had a BMT within 1 years or are still using immunosuppressive drugs

In view of the complexity of identifying the correct DBA patients falling into the extremely vulnerable group and the anxiety it may generate we advise that this is discussed with your specialist team.

Clinically Extremely Vulnerable Patients with DBA should follow the advice on <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> and register online for support using the link even if no additional support is needed right now.

What does Shielding mean?

Advice on Shielding has been updated:

1. Patients should work from home and children/young people should not return to school/educational establishments when these reopened.
2. Shielding means staying at home as much as possible and keep visits outside to a minimum (for instance once per day):
 - If you wish to spend time outdoors (though not in other buildings, households, or enclosed spaces) you should take extra care to minimise contact with others by keeping 2 metres apart.
 - If you choose to spend time outdoors, this can be with members of your own household. If you live alone, you can spend time outdoors with one person from another household (ideally the same person each time).

- You should stay alert when leaving home: washing your hands regularly, maintaining social distance and avoiding gatherings of any size.
- You should not attend any gatherings, including gatherings of friends and families in private spaces, for example, parties, weddings and religious services.
- You should strictly avoid contact with anyone who is displaying symptoms of COVID-19 (a new continuous cough, a high temperature, or a loss of, or change in, your sense of taste or smell).

Is Shielding feasible if you are living in a family group?

Siblings and other members of the household are not required to adopt protective shielding measures for themselves but should avoid going to school and work where possible. We understand that it will be impossible for families with children to separate themselves at home. We advise that families try their best to follow the guidance as best possible and encourage everyone in the household to regularly wash their hands, avoid touching their face and clean frequently touched surfaces.

Am I or is my child in the clinically vulnerable group and what does it mean?

Clinically vulnerable patients are those considered to potentially have higher risks than the general population. Our review of the available data suggests that patients in this group no longer need to strictly shield.

This group should follow the Government guidance Staying alert and safe (social distancing) for the whole population about staying at home and staying away from others: <https://www.gov.uk/government/publications/staying-alert-and-safe-social-distancing/staying-alert-and-safe-social-distancing>.

Children and young people in the vulnerable group can attend school when schools re-open for their year group. Adults, however, should continue to work from home if possible or alternatively placed in low risk environments. Give their potential for increased vulnerability; it is very important that patients still follow strict social distancing measures and good hand hygiene. Siblings and other household members should also all stay at home and stay away from others as much as possible.

Patients with DBA are classified as Clinically Vulnerable if they fall in any of the following categories:

- Patients 50 years and above
- Severe cardiac iron overload $T2^* < 10$ ms, with no additional co-morbidities and adherent with therapy
- Severe cardiac iron overload ($T2^* > 10$ ms but < 12 ms).
- Severe - moderate iron overload ($LIC > 30$ mg/g DW and $T2^* > 12$ ms) AND additional comorbidity.
- Patients receiving steroid treatment.
- Patients with congenital heart disease as per British Congenital Cardia Association guidelines in https://www.bcca-uk.org/pages/news_box.asp?NewsID=19495710 (infants < 1 year with unrepaired congenital heart disease requiring surgery or catheter intervention, chronic cyanosis (oxygen saturations $< 85\%$ persistently), patients with

severe cardiomyopathies requiring medication, and patients with congenital heart disease on medication to improve heart function).

Do I need to make changes to my treatment?

No changes to your usual treatment are required. However, you should avoid if possible changing to a new form of treatment unless essential.

- **Patients on regular transfusions** should remain on the same schedule. Chelation treatment and its monitoring should continue as usual, though you can expect MRI monitoring to be postponed if your iron control is good. If a fever develops, you should stop all chelation agents at once and contact your specialist team as you normally would.
- **Patients on steroid** treatment should remain on the same steroid regimen.
- **Patients in haematological remission** do not require additional monitoring.

The exceptions are the following:

- We do not recommend patients starting a steroid trial because of the suppression of the immune system with high doses of steroids. The steroid trial best postponed until the risk of Coronavirus has ceased.
- We do not recommend undertaking a bone marrow transplant for transfusion dependent DBA and the likelihood is that your specialist team will postpone this. There may be circumstances in which a bone marrow transplant still needs to go ahead (for examples patients who have ceased to produce blood altogether or who have abnormal cells) but these circumstances are not common and will be discussed by the specialist team.

In case of diagnosis of COVID-19:

All patients with DBA need urgent assessment if unwell or have fever both to be screened for COVID-19 and to rule out non-COVID-19 complications, which can be as or more severe (e.g. bacterial infections causing sepsis). Following the assessment, you may not necessarily need to be treated in hospital if you contract COVID-19. Some patient will recover at home after a mild illness.

It is important that you contact and inform your specialist team about symptoms or self-isolation or, following contact with 111, emergency services or admission to hospital. However, do not delay accessing emergency services in order to inform the specialist team. This can always be done afterwards. The aim of the initial assessment is to identify those patients who require direct care because they have developed the infection in the lungs and to ensure you do not have an alternative complication that requires treatment.

If receiving steroid treatment: the dose of steroids usually needs to be increased during the period of time you are unwell and this needs to be discussed with the team managing your case.

If receiving chelation treatment: you should stop chelation treatment at once and discuss this with your specialist team, as with any other febrile episode, unless you have been previously asked not to do this because of severe iron loading in your heart.

You can take paracetamol or ibuprofen when self-medicating for symptoms of COVID-19, such as fever and headache, and should follow [NHS advice](#) and contact your specialist team if you have any questions.