



## 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).

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### 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 minimise 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>

All patients with DBA are advised to adopt Social Distancing Measures, which includes being supported to work from home as per the current government guidance, even if the patient is a key worker.

- 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.

### **What is Shielding and who needs to adopt it?**

Shielding is a measure to protect people who are clinically extremely vulnerable by minimising all interaction between those who are extremely vulnerable and others. You are strongly advised to stay at home at all times and avoid any face-to-face contact for a period of at least 12 weeks but note that this period of time could change. This includes:

- Strictly avoid contact with someone who is displaying symptoms of coronavirus (COVID-19). These symptoms include high temperature and/or new and continuous cough.
- Do not leave your house.
- Do not attend any gatherings. This includes gatherings of friends and families in private spaces.
- Do not go out for shopping, leisure or travel and, when arranging food or medication deliveries if you live on your own, these should be left at the door to minimise contact.
- Keep in touch using remote technology such as phone, internet, and social media.

There will be exceptions to this, for example if you require a blood transfusion or an essential medical appointment. Your specialist team will help you plan this.

Patients are currently being contacted by letter or text from NHS England or their GPs. 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, the 29<sup>th</sup> March 2020 or been contacted by their GP, they should discuss their concerns with their GP or hospital clinician. The Haemoglobinopathy Coordinating Centre Leads and the Clinical Reference Group on Haemoglobin Disorders have been discussing this and provided greater clarity working with NHS-E.

In view of the complexity of identifying the correct DBA patients requiring Shielding and the anxiety it may generate wanting to know whether you or your child require Shielding we advise that this is discussed with your specialist team, if they do not contact you in the coming days.

It is recommended that Shielding is adopted by DBA patients in the following categories:

- on high doses of steroids defined as:
  - children: prednisolone (or equivalent)  $\geq 0.5$  mg/kg on alternate days or  $\geq 0.25$  mg/kg daily
  - adults: prednisolone (or equivalent)  $\geq 30$  mg on alternate days or  $\geq 15$  mg per day
- have an associated cellular or humoral immunodeficiency, or due to still being infants or too young to have their immune status assessed.
- have adrenal insufficiency on steroid replacement
- have iron overload defined as
  - T2\* <15 ms, previous or current impaired LV function or other cardiac complications due to the iron load or significant congenital heart disease due to DBA,
  - severe hepatic iron overload LIC >15 mg/g DW
  - the ferritin cannot be used as an assessment tool unlike other transfusion dependent anaemias as it often underrepresents the iron load.
- have had a BMT within 6 months or are still using immunosuppressive drugs

### **Is Shielding feasible?**

We recognised you may have many questions and concerns about how to institute Shielding. The feasibility of instituting many of the Shielding measures will depend on the age of the DBA patient. A substantial number of measures are not feasible for a child, but it appears that the risk of serious complications is higher for older people. In addition, a proportion of patients with DBA have learning or behavioural difficulties as part of the condition, which will also put limitations. It is important to take some time to think the implications of Shielding and discuss them both with those living with you and your specialist team.

The rest of your household are not required to adopt protective Shielding measures for themselves. They should support you in Shielding and follow guidance on Social Distancing for themselves.

You should consider the following matters:

- Minimise as much as possible the time other family members spend in shared spaces such as kitchens, bathrooms and sitting areas, and keep shared spaces well ventilated.
- Aim to keep 2 metres (3 steps) away from people you live with and encourage them to sleep in a different bed where possible.
- If you can, you should use a separate bathroom from the rest of the household. Make sure you use separate towels from the other people in your house, both for drying themselves after bathing or showering and for hand-hygiene purposes.
- If you do share a toilet and bathroom with others, it is important that they are cleaned after use every time (for example, wiping surfaces you have come into contact with). Another tip is to consider drawing up a rota for bathing, with you using the facilities first.

- If you share a kitchen with others, avoid using it while they are present. If you can, you should take your meals back to your room to eat. If you have one, use a dishwasher to clean and dry the family's used crockery and cutlery. If this is not possible, wash them using your usual washing-up liquid and warm water and dry them thoroughly. If you are using your own utensils, remember to use a separate tea towel for drying these.

We understand that it will be difficult for some people to separate themselves from others at home. Everyone in your household should regularly wash their hands, avoid touching their face, and clean frequently touched surfaces.

### **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. Blood transfusions are safe. NHSBT are working to maintain the blood supply and will liaise with hospitals in good time if problems are likely to develop to make the necessary arrangements. It may be that if the situation worsens significantly your transfusion threshold will need to be lowered from your usual Hb level, meaning your transfusions will be less frequent. 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.

**Red flag symptoms:**

You are encouraged to attend the Emergency Department (A+E) or call 999 if any of the following occur:

- Respiratory distress (new shortness of breath or increased breathlessness compared to baseline particularly at rest or on minimal exertion) +/- chest pain.
- Fever >38° C or 100.4° F lasting for at least one hour.
- severe headache, confusion or neurological changes.

***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.

**In case of diagnosis of COVID-19:**

Please remember, 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 is to identify those patients who require direct care because they have developed the infection in the lungs.

The use of paracetamol, and not ibuprofen, is at the current time the preferred option to treat the symptoms of coronavirus, unless your doctors have told you that it is not suitable for you.