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## Patient information

# Haemoglobinopathy Disorders - Living With Sickle Cell Disease Information and Advice for Patients and Carers

## Haematology Department

It is important for people with sickle cell disease to learn as much as possible about their condition so they know how to keep themselves well and avoid things that can cause a sickle cell crisis.

### **The triggers for a crisis include:**

- Dehydration.
- Not having your recommended medications and vaccinations.
- Being too cold or too hot.
- Excessive physical exertion/stress.
- Emotional stress and anxiety.

### **How you can avoid the triggers for a sickle cell crisis.**

#### **Keep hydrated**

You can become dehydrated if you are not drinking enough fluids, especially on a hot day, or have diarrhoea and/or vomiting.

- Drink three to four litres of fluid per day in the form of juice, squash or water.
- Only have tea and coffee in moderation as they increase the amount of urine you pass.
- Only drink alcohol in moderation as it can cause dehydration.

If you are unable to drink sufficient fluids because you are vomiting please contact the Sickle Cell Team (SCT) for advice.

#### **Have your recommended medications and vaccinations**

If you have been prescribed medications it is important that you take these as directed. You are also advised to take the following:

#### **Folic acid**

Folic acid is needed to help your body make red blood cells. A normal balanced diet should contain adequate folic acid but it is recommended that you take a 5mg supplement of folic acid each day.

## **Penicillin**

People with sickle cell disease have a spleen that doesn't work properly. This is called functional Hyposplenism. The spleen is a gland in the abdomen (tummy) and is part of the immune system which helps the body to fight infections.

If your spleen isn't working properly you will be more prone to developing infections, particularly from certain types of bacteria, including pneumococcus bacteria.

**There are measures mentioned below that may reduce your risk of infection.**

Your doctor will recommend that you take phenoxymethylpenicillin (penicillin V) 250mg twice a day. This is an antibiotic that is very good at destroying the types of bacteria your spleen would normally destroy. This is aimed to reduce the risk of infection that these bacteria can cause.

If you are allergic to phenoxymethylpenicillin (penicillin V) then another type of antibiotic will be offered.

If you have any concerns or questions regarding taking these tablets then please discuss it with your doctor or specialist nurse.

Find further information about these medications, including the possible side effects, in the manufacturer's leaflet that comes with them.

It is also important that you know the warning signs of an infection so that you can seek help quickly. Your specialist nurse or doctor will give you more information about these.

## **Vaccinations**

You should have had all the immunisations in the childhood immunisation programme and should continue to have the following booster doses:

**Pneumovax** - every five years to protect against the pneumococcus bacteria.

**Influenza (flu)** - once a year to protect against the flu virus.

**Hepatitis B** - to protect against hepatitis B which can be transmitted through infected blood.

**Meningivac** - This gives protection against meningococcus types A and C which cause meningitis. A single dose should be given if not already received as part of child immunisation schedule.

**Haemophilus Influenzae Type B** - A single dose should be given if not already received as part of child immunisation schedule.

Your sickle cell team will liaise with your family doctor (GP) regarding your up to date vaccinations.

## **Avoid getting too hot or cold**

Extremes of temperature can trigger a sickle cell crisis. To avoid this make sure you wrap up warm in cold weather and try to keep cool in hot weather and drink plenty to make sure you keep hydrated.

## **Avoid excessive physical stress**

It is important that you exercise regularly, but make sure you only exercise within your limits. Excessive exercise can trigger a sickle cell crisis.

## **Avoid emotional stress and anxiety**

If you feel stressed or are finding it difficult to cope with the various challenges of work, relationships, finances etc. please talk to your specialist nurse or doctor about this. It is important that you get the support you need to reduce emotional stress and anxiety.

## **Eat a healthy diet**

Like anyone else you should eat a healthy, balanced diet with plenty of fresh fruit and vegetables (these contain folic acid among other nutrients).

**Please do not take any iron supplements without talking to your specialist nurse or doctor first.**

## **Travel advice**

Travelling with sickle cell disorder is not usually a problem but it is important to plan your trip carefully.

Ensure you take a sufficient amount of medication on holiday with you as some pain medications may be difficult to get abroad.

You also need to check whether you need to have any extra vaccines before you travel or take any extra medications such as anti-malaria tablets.

It is also vital that you get adequate travel insurance that will cover you if you have any medical problems related to your sickle cell disease whilst abroad.

If you are travelling by plane, make sure you drink plenty of non-alcoholic fluids, keep warm and go for short frequent walks up and down the aisle if your journey is long.

Whilst abroad it is important that you keep hydrated; if you are not sure about the cleanliness of the water then drink bottled water, particularly if you develop diarrhoea and/ or vomiting.

## **Feedback**

Your feedback is important to us and helps us influence care in the future.

Following your discharge from hospital or attendance at your outpatient appointment you will receive a text asking if you would recommend our service to others. Please take the time to text back, you will not be charged for the text and can opt out at any point. Your co-operation is greatly appreciated.

## **Contact details**

**If you have any questions or concerns please contact the Sickle Cell Team.**

### **The Team**

**Dr D Simcox                      Consultant Haematologist**

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