

Clinical haematology

Hydroxycarbamide treatment in adults with sickle cell disease (SCD)

Information for patients, relatives and carers

Introduction

This leaflet has been provided to answer some of the questions you may have about treatment with hydroxycarbamide (also known as hydroxyurea) in sickle cell disease.

Together with advice from your specialist team this will help you to make an informed decision about taking hydroxycarbamide.

What is hydroxycarbamide?

Hydroxycarbamide is a drug that has been used for many years to treat blood disorders and certain types of cancer. It has also been found to be beneficial in sickle cell disease (SCD).

Findings from a research study conducted in the USA in the 1990s first showed that many sickle cell patients taking hydroxycarbamide have fewer painful crises, chest crises and blood transfusions. More recent evidence shows that hydroxycarbamide can also improve life expectancy in SCD.

Hydroxycarbamide is usually recommended if:

- you have had three or more hospital admissions in the past year for a sickle cell crisis
- have regular crises at home affecting your work or normal daily life
- have had two or more chest crises

How hydroxycarbamide works in SCD

Hydroxycarbamide works in several ways, by:

- Increasing the number of red blood cells containing fetal haemoglobin (HbF). Having more HbF is a good thing in SCD. During the first few months of life red blood cells contain mostly HbF. This protects against sickling. As the level of HbF falls, the benefit wears off. Some people with SCD naturally produce more HbF throughout their lives. They have fewer problems and live longer. Hydroxycarbamide stimulates production of HbF which reduces sickling – this effect may take a few months.
- Helping to keep red cells well-hydrated and reducing their stickiness. This may prevent a crisis developing. These effects occur quite quickly, which may explain why some people feel better and experience less pain within a few weeks of starting treatment.
- Reducing the number of white blood cells which is often high in people severely affected by SCD. These cells produce chemicals that can cause inflammation and trigger a crisis. Having fewer white cells makes sickling less likely.

How to take hydroxycarbamide

Before you start treatment, please read the medication leaflet provided by the pharmacy.

- Hydroxycarbamide comes in 500 mg capsules and is taken by mouth once a day.
- After starting treatment, the dose of hydroxycarbamide is usually increased gradually. This will be monitored and adjusted when you attend the outpatient clinic. The maximum dose depends on your weight, blood counts and how you respond to the drug. For adults, the maximum dose is usually three to four capsules per day.
- Hydroxycarbamide should be taken either before or after food at about the same time each day. The capsules should be swallowed whole, with plenty of water.
- Take care not to break open the capsules – if this happens accidentally, wash your hands straight away.
- If you have difficulty swallowing the capsules, ask your doctor or pharmacist if hydroxycarbamide can be provided in a different form.
- It is important to take hydroxycarbamide as prescribed to ensure you get the most benefit from treatment and avoid unwanted effects.
- If you miss a dose, take the next dose as usual and do not double it.
- If you have been unable to take hydroxycarbamide regularly, it is important that you tell your doctor or clinical nurse specialist (CNS) at your next clinic appointment.
- If you think you may run out of capsules before your next clinic appointment, contact your CNS. They will be able to arrange a prescription.
- If you take too much hydroxycarbamide by mistake, you should contact the haematology team urgently for advice.

How to store and dispose of hydroxycarbamide

- Store in a dry place at room temperature.
- Make sure it is in a secure location, out of the reach of children and pets.
- Return any unused medication to your hospital or local pharmacy.
- Do not throw the capsules in the bin or flush them down the toilet.

Side effects of hydroxycarbamide

Most people tolerate hydroxycarbamide treatment well but like all medicines there are some potential side effects. The risk of side effects is reduced by seeing you regularly in the clinic and checking your blood counts. Most side effects are reversible on stopping or reducing the medication.

Low blood counts are a side effect you should be aware of. If this happens you may notice you:

- are more tired or short of breath due to a fall in haemoglobin
- bruise or bleed more easily due to low platelets (a type of blood cell important in blood clotting)
- develop mouth ulcers or pick up infections more easily due to low white blood cells

If you experience any of the symptoms listed above or develop a fever greater than 38°C (100.4°F), you should:

- contact the haematology 24/7 triage renal and haematology triage unit (RHTU) on 020 3311 7755
- or go to the nearest A&E immediately.

You may not be aware if your blood counts are low to start with, so it is important to attend your clinic appointments for these to be checked. You can discuss any other symptoms that you are concerned about with your doctor or CNS at these appointments.

Other unwanted effects that can occur include:

- an upset stomach (diarrhoea, constipation or sickness)
- poor appetite
- headaches or dizziness
- allergic reaction such as skin rash
- skin and nail pigmentation
- hair thinning
- skin ulceration

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- abnormal liver or kidney function tests.

Most of these are uncommon.

There have been concerns that hydroxycarbamide may increase the risk of developing leukaemia and other cancers. However, after more than 20 years' experience of its use in SCD internationally, there is no evidence to support this.

Taking other medicines with hydroxycarbamide

It is safe to take hydroxycarbamide with most other medications. You should let your doctor, CNS or pharmacists know about all other medications you are taking or are prescribed.

Live vaccines (for example, yellow fever vaccine) should be avoided.

Eat and drink as usual

Eat a balanced, healthy diet and drink plenty of water.

You can drink alcohol while taking hydroxycarbamide

You should not exceed the maximum recommended weekly limits of 14 units of alcohol for men and women.

If you or your partner would like to become pregnant

Hydroxycarbamide treatment does not stop you or your partner from having children. If you are planning to have children, you should discuss this first with your haematologist or CNS. Hydroxycarbamide must be stopped for at least three months before conception to avoid any risk to the baby. While you are on treatment it is important to use reliable contraception. If you or your partner becomes pregnant while taking hydroxycarbamide contact your doctor or CNS immediately for further advice.

There is no evidence that taking hydroxycarbamide reduces fertility, although in men the sperm count may be reduced while taking the drug. This usually reverses on stopping treatment. As a precaution we advise you to consider sperm analysis and storage before starting treatment.

Breastfeeding while taking hydroxycarbamide

You should avoid hydroxycarbamide while breastfeeding because it is secreted in breast milk and could affect your baby.

Other special precautions

Sunscreen – remember to use sunscreen to protect your skin. It may become more sensitive to the sun while you are taking hydroxycarbamide.

Condoms – use condoms if you are taking hydroxycarbamide, even if another form of

contraception is used. This protects you or your partner from exposure to the small amounts of the drug present in semen or vaginal secretions and any possible harm that might result.

Consent

Before you start treatment with hydroxycarbamide your doctor or CNS will seek your consent and may ask you to sign a consent form to confirm that you have understood the benefits and risks.

Contact details for advice and further information

If you need urgent advice, contact:

- your CNS on 020 8383 8553
- the RHTU, open 24 hours a day, seven days a week: 020 3311 7755
- a hospital doctor – ask the hospital switchboard for the ‘red cell registrar’ or ‘red cell consultant’: 020 3313 1000

How do I make a comment about my visit?

We aim to provide the best possible service and staff will be happy to answer any of the questions you may have. If you have any **suggestions** or **comments** about your visit, please either speak to a member of staff or contact the patient advice and liaison service (**PALS**) on **020 3313 0088** (Charing Cross, Hammersmith and Queen Charlotte’s & Chelsea hospitals), or **020 3312 7777** (St Mary’s and Western Eye hospitals). You can also email PALS at imperial.pals@nhs.net The PALS team will listen to your concerns, suggestions or queries and is often able to help solve problems on your behalf.

Alternatively, you may wish to complain by contacting our complaints department:

Complaints department, fourth floor, Salton House, St Mary’s Hospital, Praed Street
London W2 1NY

Email: ICHC-tr.Complaints@nhs.net

Telephone: **020 3312 1337 / 1349**

Alternative formats

This leaflet can be provided on request in large print or easy read, as a sound recording, in Braille or in alternative languages. Please email the communications team:
imperial.communications@nhs.net

Wi-fi

Wi-fi is available at our Trust. For more information visit our website: www.imperial.nhs.uk

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