NHS
University Hospitals of Liverpool Group

Patient information

Information for Primary Care Professionals

Haematology Department

Information for Primary Care Professionals

Your patient is registered with the Specialist Haemoglobinopathy Team, based at the Royal Liverpool University Hospital. This document aims to summarise the available services locally, medications and their side effects, advice on drugs prescribed only by the Specialist Haemoglobinopathy Team, Immunisations and how to seek further advice.

Adult Haemoglobinopathy Services

Services for Adults with Haemoglobin Disorders (predominantly Sickle Cell Disease and Thalassaemia) are based at the Royal Liverpool University Hospital. Paediatric Services are delivered by the team at Alder Hey Children's Hospital, with whom we work collaboratively. All patients are offered Annual Clinical Reviews, in addition to regular follow-up appointments (the frequency depending on their individual clinical need). All patients have access to a haemoglobinopathy specialist nurse who they can contact during working hours, with access to Specialist Advice out of hours via the on-call Haematologist.

The Adult Haemoglobinopathy Team comprises:

Lead Consultant Haematologist
Haematology Specialist Registrar (Rotational Position)
Advanced Clinical Practitioner
Clinical Nurse Specialist x2
Specialist Psychologist
Advanced Clinical Pharmacist

Contact details: 0151 706 3397

All clinical correspondence is copied to the patients' GP, and other professionals that we are aware are involved in their care. Their GP is also provided with a copy of their Annual Review Summary and Acute Care Plan.

Prescriptions

It is advised that all patients with Sickle Cell disease and patients with Thalassaemia who have had a splenectomy should receive prophylactic antibiotics – currently Penicillin V 250mg twice daily (or Erythromycin 500mg bd if penicillin allergic). After discussion, some patients may elect not to take regular prophylactic antibiotics. **These patients must not be without an emergency supply of antibiotics – this will be communicated separately.** The majority of patients with Sickle Cell Disease will also require Folic Acid 5mg daily.

Patients with Sickle Cell Disease experience Acute Painful Crises, many of which are managed at home without the need for hospital attendance. As such, your patient will require a supply of analgesia (Paracetamol, weak opiate such as codeine or tramadol, NSAID if not contraindicated). A summary of their recommended first line analgesia will be included in their Annual Review Summary.

Please do not prescribe strong opiates for Sickle Cell related pain without discussion with the Specialist Team.

There will be some medications (such as Hydroxycarbamide and Iron Chelation agents) that we may prescribe for the patient. We will keep you informed about this and their dose changes etc. but will retain responsibility for prescribing and monitoring these agents.

Medications and their Side Effect Profiles

Please refer to the SmPC and BNF for further details or consult with the Specialist Haemoglobinopathy Team.

Patients taking these medications are monitored regularly by the Specialist Team, to ensure optimal dosing and monitor for side-effects. We will retain responsibility for prescribing these drugs.

Hydroxycarbamide

Hydroxycarbamide (Hydroxyurea) is currently the only licensed drug for the long-term control of Sickle Cell Disease. It is a cytostatic agent, with augmentation of Fetal Haemoglobin being probably its main mechanism of action. It is used to reduce the frequency of painful crises, reduce the risk of Acute Chest Syndrome, and has a role in the reduction of other chronic and acute complications.

Main side-effects are Myelosuppression (reversible), Nausea, Hyperpigmentation, Hair thinning, Teratogenicity (patients advised to stop Hydroxycarbamide after discussion with team three months before conceiving), Reduced spermatogenesis. There is no evidence to support an increased risk of malignancy.

Desferrioxamine (DFO)

Desferrioxamine is an iron chelation agent given by subcutaneous infusion. Although this can be demanding, results can be excellent if adequate adherence is maintained.
Main side effects are: Skeletal dysplasia, auditory and ophthalmic toxicity, nausea, vomiting, hepatic impairment, Yersinia and mucormycosis infections.

Deferasirox (DFX, EXJADE®)

Deferasirox is a once daily oral iron chelation agent, requiring dispersal in water, orange or apple juice. It is considered by many as the iron chelator of choice in Sickle Cell Disease.
Main side effects are: Increased creatinine, GI disturbance, rash, abnormal liver function tests.

Deferiprone (DFP)

Deferiprone is also an iron chelation product, currently only licensed in Thalassaemia Major, but has also been used in patients with Sickle Cell Disease.
Main side effects are: Agranulocytosis, Arthropathy, GI disturbance, increased ALT.

Immunisations

All patients with Haemoglobin Disorders should receive the Seasonal Influenza Vaccination unless contraindicated.

Patients with Sickle Cell Disease, and patients with Thalassaemia who have undergone splenectomy should also receive the following vaccinations:

- Normal childhood vaccination schedule.
- Hepatitis B vaccination (from one year of age).
- Pneumovax (from two years of age and then every five years).
- Haemophilus influenzae type B (Hib) vaccine.
- Meningococcal ACWY vaccine.
- Meningococcal B vaccine.
- BCG.
- Annual influenza.
- COVID.

All patients with Sickle Cell Disease, Thalassaemia Intermedia or Major or other Haemoglobin Disorders requiring transfusion should receive Hepatitis B Vaccination. The specialist team will advise on a patient-by-patient basis.

Travel Advice

Patients with Haemoglobin disorders should receive standard vaccination/prophylaxis prior travel according to the most recent guidance. Sickle Cell Disease and Thalassaemia do **not** provide adequate protection against malarial infection, and all patients should be advised to take malarial prophylaxis if traveling to a high-risk region.

Indications and arrangements for seeking specialist advice

If there are any queries regarding the management of this patient group, the Specialist Team (contact details above) are happy to advise. We are keen to remain updated about significant changes in medical condition, as there are often implications for the Haemoglobin Disorder both by the condition and its treatment. It is imperative we are consulted prior to any planned surgical intervention.

In working hours please contact us using the details above. Out of working hours please contact the Haematology SpR on-call via RLUH Switchboard (0151 706 2000).

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.

Further Information

National Haemoglobinopathy Panel - www.nationalhaempanel-nhs.net
Sickle cell Society – www.sicklecellsociety.org
United Kingdom Thalassaemia society - www.ukts.org

Contact Details:

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All Trust approved information is available on request in alternative formats, including other languages, easy read, large print, audio, Braille, moon and electronically.

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