2.HAEMOGLOBINOPATHY

Sickle Cell Crisis is a medical emergency.

Unwell patients with known or suspected sickle cell disease should be discussed with the Haematology Registrar or Consultant Haematologist on-call and admitted through the Emergency Department if necessary.

2.1 SCREENING

https://www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview

There is a national UK antenatal screening service offered for all pregnancies.

Cornwall and Devon are low prevalence areas and whether to test is based on a Family Origin Questionnaire.

Screening should occur by 10 weeks of pregnancy to allow prenatal diagnosis by 12 weeks + 6 days

The haematology department are responsible for processing the tests. Responsibility for follow up lies with the obstetric and genetic services.

Haemoglobinopathy screening is also included in the newborn blood spot tests.

The main purpose is to identify sickle cell disease early – identification and management improves outcomes – but the majority of haemoglobinopathies will be detected.

2.2 DETECTION:

The majority of adult patients will be aware if they have a significant haemoglobinopathy and will be under the care of a haematologist. However, those with heterozygous ('trait') or mild homozygous disease may be detected incidentally though screening or through an abnormal FBC performed for other reasons.

Variant haemoglobins are also commonly detected incidentally when HbA1C is requested. - Letters are generally sent out to GPs together with patient information leaflets where appropriate for potentially clinically significant findings.
Consider testing for those who have moved into the area, for children who did not undergo antenatal screening, and for family tracing.

Diagnosis is by capillary electrophoresis and confirmatory gel electrophoresis. Additional genetic testing may be required.

Haematology review is seldom necessary for these patients but knowledge is essential when affected people are considering having children, with the possibility of partner testing.

Patient friendly information leaflets available for various different carrier states (beta thalassaemia carriers, delta beta thalassaemia carriers, Hb OArab carriers, Hb C carriers, Hb D carriers, Hb E carriers, Hb Lepore carriers, Sickle cell carriers) at:

https://www.nhs.uk/conditions/thalassaemia/carriers/

2.3 PRE-OPERATIVE SCREENING

Patients who are of African or Afro-Caribbean heritage, or who have a family history of sickle cell trait or disease, should be offered diagnosis by haemoglobin electrophoresis before any general anaesthetic or elective procedure.

In an emergency a Sickledex test may be performed which detects both heterozygous (trait) and homozygous disease, with haemoglobin electrophoresis required to distinguish the two.

2.4 SICKLE CELL TRAIT

Explain to the person with sickle cell traits and/or their family/carers that:

- They should very rarely have symptoms. However, they are at risk of a vaso-occlusive episode if they become oxygen deprived. They should therefore:
- Avoid extreme exertion
- Avoid high altitudes, such as travelling in an unpressurized aircraft.
- Inform the anaesthetist that they are sickle cell carriers, if they are going to have an anaesthetic.
- They have 1 in 2 chance of passing the sickle haemoglobin gene to their child. If the other parent is also a carrier, there is a 1 in 4 chance that their child will have sickle cell disease.
- It is important to have malaria prophylaxis if they will be visiting an area where malaria is endemic. See the CKS topic on Malaria prophylaxis for more information**.**

2.5 REFERRAL

Please refer to the Bristol Guidance for the management of sickle cell crisis

https://uhbw.mystaffapp.org/diliboards/50/diliboard_contents/4353/document_view.pdf