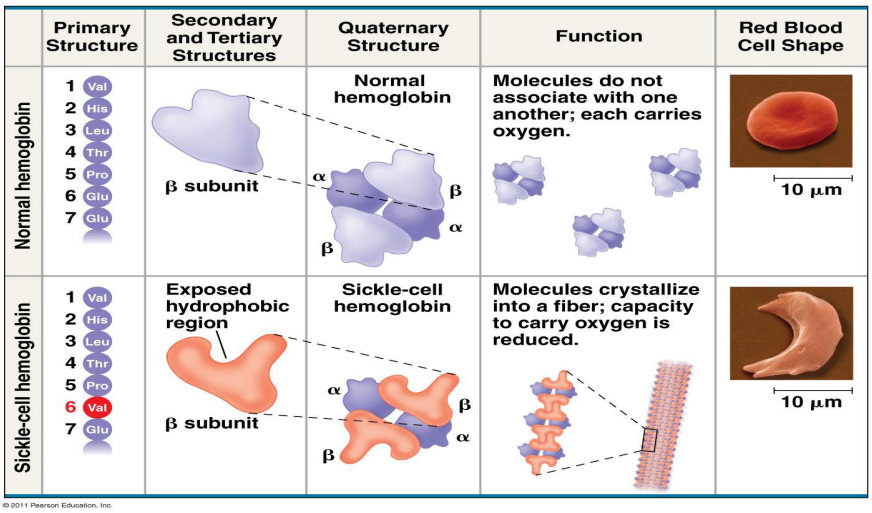
TAKING CARE OF PATIENTS WITH SICKLE CELL DISEASE
USEFUL INFORMATION FOR GENERAL PRACTITIONERS

PATHOPHYSIOLOGY

Sickle cell disease affects 1 in 2400 births annually in the UK and it has been characterised as the commonest and fastest growing genetic disorder.
In Croydon, approximately 800 people are affected by sickle cell disease, according to the Sickle Cell and Thalassemia Centre records.

Sickle cell disease arises from a mutation in the beta-globin gene, leading to a change in structure of haemoglobin. This leads to polymerization of the haemoglobin molecules into fibrins when they do not carry oxygen and a change in the red cell shape to sickle, leading to chronic haemolysis and vasoocclusion (picture 1).



Picture 1

As a consequence, multisystem complications arise, including painful crises, bone infarcts, strokes, retinopathy, chronic lung disease and pulmonary hypertension, coronary artery disease, chronic liver disease, hyposplenism and reduced immunity (picture 2).

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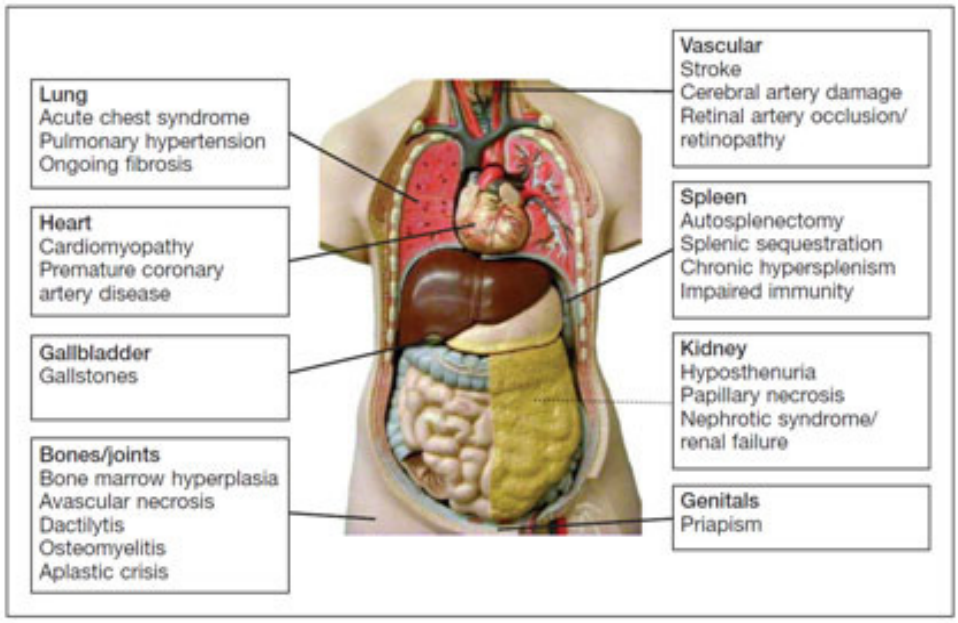

Picture 2

The most frequent genotypes associated with the disease are:

1. Homozygosity for the sickle beta globin gene (β^s) (HbSS)
2. Compound heterozygosity of the HbS and HbC genes (HbS/C disease)
3. HbS/β^0 or HbS/β^+ thalassemia

RECOMMENDATIONS

Sickle cell patients are in need of multidisciplinary and comprehensive care, with close communication and collaboration between community care and specialized hospital teams.

1: Care:

People with sickle cell disease need to be followed up by haematology specialists. Please ensure they are referred to and regularly reviewed by haematologists.

2: Regular medication:

All patients with sickle cell disease need to take Folic acid regularly, since they have increased and constant haemolysis and have increased needs of folic acid.

3: Infection prevention:

Due to hyposplenism, patients with sickle cell disease are at an increased risk of infections, especially from encapsulated organisms (such as Pneumococcus, Meningococcus and Haemophilus Influenzae).
They need to be up to date with the following vaccination schedule:

a) Annual seasonal flu vaccination
b) Pneumococcal Vaccine every 5 years

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c) Once-off tetravalent Meningococcal Vaccine ACWY
d) Once off Haemophlus Influenza b vaccine (if not done at childhood)
e) Hepatitis B vaccination (provided patient is not immune and not a HBV carrier)

Please ensure that a copy of the patients' vaccination status is sent annually to the Haematologist following up the patients, to keep in their records.

In addition to the above, it is recommended that patients take antibiotic prophylaxis daily with penicillin V 250 mg BD or erythromycin 250 mg BD if penicillin allergic. For patients that refuse to take antibiotic prophylaxis regularly, it is advised that they hold a weeks' course of antibiotics in reserve in case they develop a febrile illness and to get medical advise as soon as possible to prevent sepsis.

4: Pain management:

For the treatment of sickle pain, paracetamol and NSAIDS and opioid analgesics are used (NSAIDS are used provided there is no history of renal damage or proteinuria more than trace in urine dipstix). Strong opioid analgesics are sometimes required for severe pain. Please note that pethidine is NOT recommended by NICE for the treatment of sickle pain due to the risk of neurological complications (seizures).
Sickle patients are sometimes prescribed by the Haematology specialists with a short course (5-7 days) of opioid analgesics to use at home in case of sickle pain but are advised to attend the hospital A&E if the pain is severe and/or not responding to oral analgesics. Long term use of strong opioids should be avoided if possible. Please contact the Haematology consultant if a patient is requiring long term opioids to control sickle pain.

For prescription of opioid analgesics, it is advised that only a single healthcare professional provides prescriptions, provided there is constant communication between primary care and haematology specialists, in order to avoid duplication and over-prescription.
Please be aware of adverse effects of opioid analgesics, including respiratory suppression, cardiovascular compromise, nausea, constipation, pruritus and dependency. Please monitor these patients closely and liaise with the Haematologist in charge of their case if further information is needed.

5: Specialized Treatment:

Sickle cell patients with more severe disease phenotype will be offered the following specialized treatments:

1. Red cell transfusions or exchange transfusions: These will be undertaken at the hospital at regular intervals. Patients on regular transfusions will be closely monitored for evidence of iron overload (with regular blood tests and special imaging).
   To prevent iron overload from developing, patients may be started on iron chelation treatment (Deferoxamine s/c, deferiprone or deferasirox po). These agents will be started by the Haematology specialist in charge

of the patient's case and monitoring will be performed at the Haematology Clinic. However, please be aware of possible adverse effects that these medications may cause, such as GI disturbance, abnormal liver and renal function tests, visual and hearing decline and leukopenia (specific to deferiprone). The haematology doctor will inform you of any special measures or monitoring needed for these patients.

2. Hydroxycarbamide: This is an oral cytotoxic agent that is effective in sickle cell disease, helps increase Hb levels; prevents recurrence of painful crisis and more serious complications of the disease. The decision to initiate this treatment lies with the Haematology Specialist managing the patient, after consultation and consent by the patient. Initiation of this treatment will be done at the Hospital and regular monitoring of blood parameters is required to achieve the maximum tolerated dose that will offer the greatest clinical benefit.

If a sickle patient is on Hydroxycarbamide, please encourage them to be compliant and ensure that they attend regular haematology clinic appointments for monitoring of blood parameters, since Hydroxycarbamide may cause low blood counts (such as neutropenia) and liver and renal test abnormalities.

After a period of time when the patient is stable on a Hydroxycarbamide dose, you may be asked to take on prescriptions as part of a shared care agreement. This will be a separate process, involving patient consent and agreement between GP and Haematology Specialist. In this case, detailed information will be sent to you prior to agreeing on shared care.

Patients with Sickle Cell Disease require multidisciplinary care to maintain a good quality of life. Please ensure their easy and timely access to primary care and remain in close communication with the Haematologist or Nurse Specialist.

We hope that you find the above information helpful.
If you need further information, please……..

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