SICKLE CELL CRISIS

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Very few patients with sickle cell diseases (HbSS, HbSC, HbSBthal) live in the Kingston Hospital catchment area. Many live in adjacent catchment areas and are registered at St. George's or the Hammersmith Hospitals, or are students at Kingston University. They may become unwell locally and attend our A and E department. They may carry with them a 'Haemoglobinopathy Card'. This will often give helpful advice on aspects of treatment.

PAIN CRISIS

The most common type of crisis presents as agonising and relentless pain. The pain may be localised to a single long bone (typically in the juxta-articular area), present symmetrically in several limbs, or involve the axial skeleton (lumbar spine, ribs or pelvis).

In the Accident and Emergency Department

Assessment

1. Patients with sickle cell disease should be triaged as urgent. Pre-analgesia assessments should be kept to a minimum. Our target time for presentation-to-medical assessment is within 30 minutes. **Pain control should be given within that time.** Pain, respiratory rate and sedation level should be measured every 20 minutes until satisfactory pain relief has been achieved and then at least every 4 hours.
2. For pain management, titrate intravenous morphine in increments of 2mg doses every 5 minutes (up to a maximum of 10mg), while waiting for PCA to be set up. Give intravenous paracetamol 1g if >50kg (< 50kg: 15 mg/kg every 4-6 hours). Contact Pain Team or the On call Anaesthetist on bleep 040 to set up Patient Controlled Analgesia (PCA) morphine, or fentanyl if patient has allergy or sensitivity. **Follow the Patient Controlled Analgesia policy on PIMS.**

Notes:

1. Pethidine should not be given unless specifically indicated on the patient's haemoglobinopathy card: it is associated with grand mal seizures in susceptible patients.
2. If a new patient or a patient without a haemoglobinopathy card requests pethidine and refuses any alternative, then he/she should be referred directly to the Haematology team.
3. Nitrous oxide (Entonox) should not be given: in patients with sickle cell disease it can cause an acute, irreversible neuropathy

After analgesia, perform a full medical assessment. This should include:

- clinical examination focusing on the chest, abdomen and CNS
- measurement of body temperature, BP, pulse and respiratory rate,
- pulse oximetry measuring O_2 saturation,
- taking blood for full blood count, U&Es, blood culture and group and save
- requesting and reviewing a chest x-ray if the pain is in the chest. Do not x-ray painful bones as it is rarely useful
- checking for clinical signs of any of the life threatening crises (see below).

Action

1. Give oxygen. Ensure airway and ventilation and then start 24% O_2 at 4L/min via a facemask. If pulse oximetry shows saturation of < 92%, increase concentration of inhaled O_2.
2. Fluids. Give at a rate of 1L every 6 hours. Because of problems with venous access give orally if at all possible. For children get paediatric advice.
3. Antibiotics. Fever is usual in crisis and infection is often present. Start an antibiotic as per the following guidelines:

- For patients who are admitted with uncomplicated painful crisis without specific evidence of infection but who develop pyrexia, commence oral Co-Amoxiclav 625mg TDS, after cultures (blood, urine and any other source that is indicated) have been taken.
- If penicillin **allergic** or there are chest signs, or an abnormal CXR, give cefuroxime 1.5 grams IV TDS (unless there is renal impairment) and clarithromycin 500mg PO BD.
- If symptoms/signs of focal infection are present (e.g. tonsillitis, UTI) consult the hospital antimicrobial policy for drug of choice
- Stop prophylactic penicillin if any additional antibiotics cover for pneumococcus.
- Patients on desferrioxamine (DFO) who have diarrhoea should be started on ciprofloxacin immediately (after checking records that they are not G6PD deficient) and the DFO stopped. Ciprofloxacin can be stopped if Yersinia infection has been excluded.

Admission

If the patient is to be admitted immediately, contact the Bed Manager and advise the haematology team. No patient admitted with sickle cell crisis should be placed on a ward outside the Medical Unit. After admission to the ward, Pain team will review patient on the ward. Do not give other potent analgesia while patient is on PCA unless advised by the pain Team

The patient should wait no more than 4 hours in A&E. If, for unavoidable reasons, this delay is extended then the patient should:

1. be given a 2 hourly programme of analgesia
2. have fluid input maintained
3. have antibiotic regime maintained
4. be observed regularly to ensure all vital signs are maintained.

If a patient is discharged from, or leaves A&E, then:

- contact the GP and let him/her know of the attendance and assessment. This may be done by telephoning Balham Health Centre on 0208 700 0615.
- give the patient sufficient analgesia to ensure effective pain management until the patient may see their GP or a specialist nurse counsellor.

LIFE-THREATENING CRISIS

Patients can present with a variety of other acute manifestations which may be rapidly fatal if not recognised and treated quickly.

INFECTION

Patients prone to sickling have reduced splenic function and are at risk of overwhelming septicaemia (pneumococcus, meningococcus, rarely haemophilus) even if taking penicillin prophylaxis. Peak risk is in childhood. The patient may present with fever, shock, seizures, coma, meningitis (often with delayed CSF pleocytosis) or even profuse diarrhoea. Early IV antibiotics (broad-spectrum beta-lactams such as amoxicillin or cefotaxime) and volume support are vital.

SPLENIC SEQUESTRATION

During infection children may suffer a rapid fall in haemoglobin and growth of the spleen – changes often noted by the mother. Death can result from hypovolaemia and anaemia. Early transfusion is vital.

CHEST CRISIS

Assess each patient with acute painful sickle cell episodes for acute chest syndrome if he/she exhibits one or more of the following signs: abnormal respiratory signs or symptoms, chest pain, fever, or signs or symptoms of hypoxia. It sometimes begins as a pain crisis affecting ribs or shoulders. Treat with fluids and oxygen; observe arterial oxygen tensions – a falling PaO_2 will require exchange transfusion which needs expert advice.

GIRDLE SYNDROME

If sickling occurs in the splanchnic bed, the patient may develop abdominal pain with rigidity, loss of bowel sounds and increasing icterus. IV fluids are vital. A surgeon should be consulted to exclude other abdominal events, but surgery should be with-held unless unavoidable, and then only after exchange transfusion.

CEREBRAL SICKLING

Patients can present with strokes, fits, coma, bizarre behaviour or psychosis. Sickling should be excluded in any susceptible patient presenting with such signs. IV fluids are vital and early exchange transfusion a possibility.

BLOOD TRANSFUSION

In a patient with Sickle Cell Disease, blood transfusion can be dangerous. Never give a simple transfusion for anaemia (except in those sequestrating) without reducing the HbS level by exchange. If this precaution is not taken, the blood viscosity will be increased and the patient made worse. Get haematological advice.

SURGERY

Do not plan or carry out surgery without first assessing the patient with the Haematology Team. Special pre- and post-operative care, often including blood exchange, is essential to optimise outcome.