

BROADSHEET

Management of sickle cell crisis

BRITISH ASSOCIATION FOR ACCIDENT AND EMERGENCY MEDICINE GUIDELINES FOR THE MANAGEMENT OF SICKLE CELL CRISES

REMEMBER AT ALL TIMES—The patient or relative generally knows if he/she has sickle cell disease and is usually knowledgeable about the disease. The pain of sickle cell crisis is genuine, can be excruciating, and can be greater than that of childbirth. Sickle cell disease patients often have a higher pain threshold than others.

DISEASE DEFINITION:

Those at risk include patients with an ethnic background from Africa, West Indies, India, the Mediterranean, or the Middle East. Sickle cell crisis may present with a variety of clinical syndromes including severe pain in limbs, chest, abdomen, and/or systemic conditions. The pain is often severe but limb, hip or back pains may be vague in the early stages of an attack. The pain is caused by blockage of small vessels by sickled red blood cells. The immediate result of sickling is a reduction in oxygen delivery to tissues. The local fall in oxygen saturation may be precipitated by a number of factors, such as minor infections.

MANAGEMENT AIMS:

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| 1. Parenteral analgesia within 15 minutes | 6. Obtain expert haematological advice early and/or contact the patient's base hospital if any of the following is present: | |
| 2. IV fluids within 30 minutes | a. shock | d. silent abdomen |
| 3. Admit to ward within two hours | b. pregnancy >12 weeks | e. organ involvement |
| 4. Warmth. Reassurance | c. dyspnoea | f. vomiting/dehydration >10% |
| 5. Identify the cause of pain | | g. abdominal pain/distension |
| | | h. you are inexperienced in the management of this condition |

TREATMENT OF THE PAINFUL CRISIS:

TRIAGE NURSE

Place on a trolley
Check oxygen saturation
Give oxygen
Wrap in warm blankets if patient feels cold

Triage to resuscitation room if:

- SaO₂ <85%
- Neurological deficit
- Hgb <5
- Shock
- Organ involvement
- Priapism

Obtain an interpreter if needed
Ask for urgent medical assistance
Check Sickle Cell Disease Register if available
Contact patient's base hospital for information

A&E DOCTOR

Give high flow oxygen. Ensure adequate ventilation. Obtain IV access.

Take bloods for FBC, reticulocytes, group/save, urea/electrolytes, viral titres, blood cultures.

Commence IV infusion. (1 litre 0.9% NaCl over 3 hours in an adult)

Do not x ray sites of pain.

Give amoxycillin 500 mg and penicillin G 1200 mg IV

In case of allergy give clarithromycin/erythromycin and seek advice.

Give IV morphine except in case of allergy/pregnancy/liver disease (seeking advice regarding liver disease)

Body weight

30-50 kg

>50 kg

If unable to obtain IV access, give morphine 0.1 mg/kg IM/SC. Repeat after one hour if required. If morphine contraindicated, seek advice urgently. DO NOT USE PETHIDINE ROUTINELY.

Dose of morphine

morphine 5 mg

morphine 10 mg

SECONDARY SURVEY:

There are certain recognised syndromes and complications which may occur during sickle cell crises.

CHEST SYNDROME

Suspect from chest pain, tachypnoea, chest wall tenderness and SaO₂ <95%. Often misdiagnosed as clinical signs and chest x ray changes may present late. Look for signs of coincidental ileus. Monitor arterial gases. If pA0₂ <9 kPa contact anaesthetist, consider CPAP. If pA0₂ <8 kPa ask haematologist about possible exchange transfusion.

INFECTIONS

Prone to pneumococcus and parvovirus B19 especially if hyposplenic. Pyrexia may be only sign. Do blood cultures, sputum cultures, throat swab, MSU/urinalysis. Discuss with consultant microbiologist and haematologist.

NEUROLOGICAL

Strokes, fits and TIAs can present with no associated pain. Discuss these with a haematologist.

ABDOMINAL SYNDROME

Usually pain with no peritoneal signs and normal bowel sounds. Sometimes like surgical abdomen. Do abdominal x rays and serum amylase. Keep nil by mouth and monitor carefully. If the patient is on desferrioxamine and develops diarrhoea, stop the drug. Check yersinia titre, and treat if positive.

APLASTIC CRISIS

Characterised by: high output CCF, decreased haematocrit, and reticulocytes associated with parvovirus B19 infection. Siblings may be source of parvovirus. Senior haematology opinion essential.

SEQUESTRATION

Presents with severe anaemia. Age <5 years generally splenomegaly. Age >5 years generally hepatomegaly. May need urgent transfusion. Seek immediate advice from a consultant haematologist.

MANAGEMENT OF CHILDREN:

Seek expert assistance early. IV access may be difficult; start oral rehydration immediately. Give paracetamol early orally/rectally. Morphine sulphate SC as follows: up to 1 month, 0.150 mg/kg; 1-12 months, 0.200 mg/kg; 1-12 years, 0.1-0.2 mg/kg. Guide to weight in kg for children aged less than 10 years = (age + 4) × 2. Provide good emotional support for the child and parents. Some children may appear angry and rebellious but try and imagine what they have been through.

PITFALLS:

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| • Do not underestimate the requirement for pain relief. | • Do not transfuse the patient, except under direct advice of a haematologist. | • Treat early to avoid late complications. |
| • Avoid damage to veins if IV access is very difficult and take patient's advice on the best vein. | • Watch for signs of organ involvement. | |
| • If IV access impossible, try NG or rectal fluids. | • Look for signs of advancing crisis. | |

DEFINITIVE CARE:

Admit to medical/paediatric ward for ongoing treatment and further haematological opinion.

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Commentary

WHY THESE GUIDELINES

Greater population movement in recent times has meant that a patient can present with a sickle cell crisis anywhere in the UK. These guidelines will be particularly useful for those hospitals which deal with this condition infrequently. These patients are often disadvantaged in accident and emergency departments, where they present less frequently, because the underlying pathological severity of the condition is easy to underestimate.

WHAT IS A SICKLE CELL CRISIS?

In the inherited condition of the gene causing an amino acid substitution in haemoglobin from the normal adult HbA to HbS, the homozygote (SS) has sickle cell anaemia and the heterozygote (AS) has sickle cell trait. In the deoxygenated state HbS molecules can polymerise causing red blood cells to sickle. Sick cells block small blood vessels to cause infarction but are also fragile and therefore haemolyse. Sickle cell crises may be from thrombosis, marrow aplasia, sequestration, or haemolysis.

PROBLEMS IN MANAGEMENT

(1) *Pain without clinical signs* means that it is difficult for the uninitiated to take the pain seriously. Assessment is made even more difficult because the patient may be irritable due to pain and hypoxia. Generous, early, and frequent analgesia is required. In severe pain, morphine is the drug of choice. Patients like

pethidine but its use should be avoided because of the greater possibility of addiction, the possibility of metabolites causing fits, and the long half life which makes patient controlled analgesia difficult. Patients may say that they have allergic reactions to morphine in an attempt to persuade the doctor to use pethidine so this history should be properly pursued. Ketorolac trometamol has been shown to be useful in the USA but centres in the UK have found it less useful because patient acceptance has been poor.

(2) *Hydration* can be difficult by the intravenous route because of vein closure due to frequent cannulation. Profuse oral drinks may tide over the problem temporarily in patients with normal bowel sounds, as may rectal rehydration. The external jugular or any other vein can be used in an emergency.

(3) *The need for speed* because of the risks of rapid deterioration, infarction, and infection is paramount.

CONCLUSION

We hope these guidelines find national acceptance and implementation. Rapid effective management of sickle cell crises will result in a dramatic reduction of avoidable distress and morbidity.

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Faculty of Accident and Emergency Medicine

Consultant appointments, January to April 1998

Mr Richard Bailey	Chesterfield and North Derbyshire Hospital
Mr Martin Edgell	Sharoe Green Hospital, Preston
Mr R Pullinger	John Radcliffe Hospital, Oxford
Dr G L Campbell-Hewson	Addenbrooke's Hospital, Cambridge
Mr Peter Thomas	Milton Keynes General Hospital
Dr Fionna Poyner	Northampton General Hospital
Mr David Small	Bedford Hospital
Dr George Olverman	Macclesfield District General Hospital
Mr I H Jones	Royal Bolton Hospital
Dr Ruth Spedding	Warrington Hospital
Mrs Carol Gray	North Staffordshire Hospital
Dr P A Kenny	University Hospital, Lewisham