Managing acute and chronic pain in sickle cell disease

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Acute painful episodes among patients with sickle cell disease may occur in any body part or several sites simultaneously. The reasons for inadequate pain relief for this patient group are complex. Prompt, accurate pain assessment is the cornerstone of effective pain management and a comprehensive, multidisciplinary team strategy is essential, particularly for managing patients with frequent acute pain or chronic pain.

Sickle cell disease (SCD) encompasses a group of haemoglobinopathies. There is currently no cure for adults with this hereditary disease, which mainly affects people of Afro-Caribbean origin and, to a lesser extent, Mediterranean, Middle Eastern and Asian groups. There are large numbers of people around the world who suffer from acute or chronic pain, or indeed both, as a result of SCD. Their quality of life is largely determined by the severity of their symptoms and the standard of their pain management. Unfortunately, for many people with SCD pain management is likely to be suboptimal at best and may well be extremely poor.

Reasons for inadequate pain relief among SCD patients are many and complex. One major reason is likely to be that, besides SCD being one of the most common hereditary disorders (Gribbons et al, 1995), it is poorly understood in areas with small populations of people of Afro-Caribbean origin. Lack of understanding can lead health care professionals to underestimate how much pain SCD patients suffer, and some SCD patients feel their reports of pain are not taken seriously (Alleyne and Thomas, 1994).

Another main reason for the poor quality of pain management is that SCD pain is often very poorly assessed and is consequently treated with inappropriate analgesics. This is particularly true for SCD patients with long-standing pain problems.

Pain in sickle cell disease

The hallmark of SCD is severe pain. This is caused by the tendency of sickle haemoglobin molecules to crystallise, distorting the red cells into crescent shapes on deoxygenation, occluding small blood vessels (Fig 1). Disruption of normal circulation leads to acute tissue ischaemia and secondary inflammation and, when prolonged, infarction of bone, joints and vital organs (Davies and Oni, 1997). Acute tissue ischaemia is associated with extreme pain, which may last from hours to weeks. The average duration is 5–7 days (Thomas et al, 1998) and the episodes are usually self-limiting.

Prolonged infarction can lead to a multitude of complications, such as organ damage, degeneration of the spine and joints, and ischaemic leg ulcers. These complications are associated with chronic pain and disability. Hydroxyurea has been used to reduce acute painful episodes (Charache et al, 1995).

Acute pain

Acute painful episodes may occur in any body part or at several sites simultaneously. They are often unpredictable and unavoidable, but may be precipitated by dehydration, cold, and physical and emotional stress such as surgery and pregnancy. They can also be brought on by hypoxia, chest infections and exposure to high altitude. Most patients manage pain satisfactorily at home with simple oral analgesics and present to hospital when pain becomes unbearable or they require professional help. Only a small proportion regularly use health services. Anie et al (2002) found that almost one-half of emergency visits and hospital admissions made by adult SCD patients were accounted for by 10 per cent of patients.

Vaso-occlusive pain is probably one of the most intense forms of pain. An acute episode of SCD pain could be likened to the pain of a myocardial infarction. For many patients, severe painful episodes indicate a threat of early and sudden death caused by vaso-occlusion and subsequent hypoxia and organ failure.

Chronic pain

Ongoing pain in SCD is usually due to orthopaedic problems and organ damage. Common causes include avascular necrosis (AVN) of the humeral or femoral head, and...
be locally recognised and used and therefore be familiar to patients. Whether patients present to their GP or A&E, assessment should be prompt to expedite pain relief and allow life-threatening syndromes, such as acute sickle chest syndrome, to be treated urgently. Patients with ongoing pain need a more in-depth assessment, including quality-of-life measures such as mood and function. An accurate diagnosis will be easier to attain using relevant investigations such as Doppler studies for leg ulcers and MRI scans for AVN. Patients with complex pain problems benefit from comprehensive assessment by specialists who understand the mechanisms and management of chronic pain.

Pain management
A comprehensive, multidisciplinary team strategy is essential, particularly for managing patients with frequent acute or chronic pain. Pain management guidelines and individual clinical management plans, which are shared between primary and secondary care teams, can promote continuity of care and patient confidence. The appropriate involvement of physiotherapists, occupational therapists and psychologists should also be enlisted. Good communication with other specialists can help reduce acute painful episodes and anxiety in acute-health care situations.

For example, pregnant patients benefit from careful pain management and planning for labour with the anaesthetist and obstetric team, and perioperative patients need analgesic management to be carefully coordinated by the theatre and recovery staff, ward nurses and pain team.

Most acute painful episodes can be managed at home, following a simple analgesic ladder. Mild pain may only warrant paracetamol and heat rubs. Non-steroidal anti-inflammatory drugs (NSAIDs), tramadol or a low-dose oral opioid, such as codeine or morphine, may be required for moderate pain. Most people with SCD will know that they should increase their fluid intake, keep warm and rest to aid recovery.

If the pain becomes more severe, medical help may be sought for stronger oral opioid doses, titrated against the level of pain. Where they are available, SCD day unit facilities allow prompt, effective assessment and management and help people avoid many A&E visits and hospital admissions (Cluster and Vichinsky, 2003).

Acute painful episodes in hospital
Thorough pain assessment will indicate the level and type of analgesia required for each individual on each visit. Treatment should be fast, aggressive and closely monitored. Patients will benefit from reassurance that they are believed when they report pain and that medication will be delivered swiftly. Gas and air must not be used frequently or for prolonged periods because it can precipitate megaloblastic anaemia in patients with haemolytic anaemias, such as SCD.

Patients with coexisting chronic pain should continue

Quality of life
Recurrent episodes of severe acute pain and unrelenting chronic pain can be profoundly disabling and depressing. Pain in SCD patients is associated with reduced scores on quality-of-life measures such as physical function, role limitations due to physical problems, social functioning and general health perceptions (Anie et al, 2002).

People with SCD may have been aware of their shortened lifespan since they were children and have negative coping strategies (Thomas and Taylor, 2002). Hospital pain management can disempower patients due to staff misconceptions about pain levels and fears of opioid addiction. Strategies that empower patients to manage stressors better and encourage patient involvement may go some way to counter this (Maxwell et al, 1999).

Pain assessment
Prompt, accurate pain assessment is the cornerstone of effective pain management. It should include pain site, duration, score, character, exacerbating and relieving factors, associated symptoms, previous analgesia and physical examination. Ideally, the assessment tool would be locally recognised and used and therefore be familiar to patients. Whether patients present to their GP or A&E, assessment should be prompt to expedite pain relief and allow life-threatening syndromes, such as acute sickle chest syndrome, to be treated urgently. Patients with ongoing pain need a more in-depth assessment, including quality-of-life measures such as mood and function. An accurate diagnosis will be easier to attain using relevant investigations such as Doppler studies for leg ulcers and MRI scans for AVN. Patients with complex pain problems benefit from comprehensive assessment by specialists who understand the mechanisms and management of chronic pain.

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Guided reflection

Use the following points to write a reflection for your PREP portfolio:

- Outline your area of expertise and why you read this article;
- Summarise the main points the article makes about pain in sickle cell disease;
- Identify a new piece of knowledge you have learnt about pain in sickle cell disease;
- Consider how you might use this knowledge in your future practice;
- State how you intend to follow up what you have learnt from this article.

Opioids

Opioid-agonist drugs are the mainstay of treatment for acute SCD pain. Patients whose pain is severe enough to warrant hospitalisation usually require opioids.

The consensus view suggests that morphine is usually the most suitable opioid, certainly for new patients and children. Pethidine has become outmoded due to the short duration of action and its unattractive side-effects (Ballas, 2001).

In most cases, opioids can be given orally. The dose must be titrated appropriately to reflect the drug’s therapeutic duration of action and the intensity of the pain. Severe painful episodes often necessitate very high and frequent doses of opioids.

Parenteral opioids may be required for a short period of time to aid rapid titration of analgesia and bring the pain under control. People with SCD often have poor venous access and so it is preferable to administer parenteral opioids via subcutaneous cannula rather than intravenously.

Some SCD patients prefer to receive patient-controlled analgesia (PCA) to promote fast and predictable pain relief and give themselves a degree of control over their pain (Johnson, 2003).

The subcutaneous route should also be used for PCA. Diamorphine is useful due to its solubility, allowing a small volume, such as 5–10mg per ml, to be administered subcutaneously. Typically, bolus doses of diamorphine need to be higher than in postoperative PCA regimens, for example 5–30mg, and lockout times longer, which can be 20–60 minutes.

Other opioids, such as hydromorphone, may be used for patients who experience intolerable side-effects to morphine or diamorphine. Alternatively, slow-release oxycodone offers a fast onset of analgesia with prolonged beneficial effect. Fentanyl lozenges could be used for a short, initial period, offering a speedy analgesic onset that is comparable to opioid injection.

Managing chronic pain

Thorough pain assessment will indicate the type of pain management approaches that are most likely to be effective. Patients should always be encouraged to engage in activities that will help them manage their own pain and boost their confidence rather than make them dependent on health care professionals. Most local leisure centres run affordable classes, such as yoga and hydrotherapy, which can be enjoyable and effective.

Many people with SCD learn to avoid swimming because cold water can precipitate acute painful episodes. They may therefore need a lot of reassurance that the warm water in a hydrotherapy pool will enable them to safely and comfortably exercise aching joints.

Occupational therapists and physiotherapists play an important role in showing patients how to achieve optimum physical function and independence.

Psychologists may be able to use cognitive behavioural therapy to help SCD patients with anxiety and depression stemming from sources such as disability, job absences and financial difficulties (Thomas et al, 1998).

Pain specialists may need to be enlisted early on to ensure that patients receive comprehensive assessment and appropriate chronic pain management. Topical treatments and non-pharmacological approaches should be considered early on.

Selected analgesics are likely to include those with anti-inflammatory, antineuralgic and local anaesthetic properties. Degenerative conditions such as AVN may be amenable to more invasive approaches such as neural blockade and surgery. Patients with progressive muscular-skeletal conditions should be referred to pain management, orthopaedics and rheumatology specialists.

REFERENCES


NSAIDs

NSAIDs, administered orally and rectally, make excellent adjunct therapy due to their effectiveness in reducing bone pain, their anti-inflammatory action and possible opioid-sparing effect. It is important to rule out contraindications to NSAIDs such as impaired renal function, which is not uncommon in SCD patients. Topical NSAID preparations and Cox-2 inhibitors should be considered in patients prone to dyspepsia and those requiring frequent or long-term therapy.

NSAIDs such as ketorolac and parecoxib can be given for short periods in patients who are unable to tolerate other routes. However, intramuscular diclofenac should be avoided because the injection can be painful to administer and repeated use can lead to sterile abscesses.

Drugs may need to be given to relieve and prevent analgesic side-effects. These include antiemetics, laxatives and antipruritics, such as hydroxyzine 25–50mg. Intravenous fluids help to decrease blood viscosity, improve blood flow and reduce risk of renal compromise. Warming with heat pads and supplemental oxygen (where the oxygen saturation has dropped below about 90 per cent) play an important role in improving oxygenation and perfusion, and reducing tissue necrosis.
Antineuralgics
Tricyclic antidepressants such as amitriptyline have been used to treat neuropathic pain for many years. They produce analgesia by inhibiting the uptake by nerves of catecholamines, such as noradrenaline and serotonin, thus increasing levels of these neurotransmitters in synapses. Tricyclic antidepressants are not used to offer pain relief by reducing depression, as their analgesic effect is produced at lower doses and more quickly than their antidepressant effect.

Patients should be made aware that these drugs are also used for people experiencing depression and that it will take a few weeks of treatment before they will feel the benefits. Doses usually start at 25mg at night and are increased gradually. One of the side-effects of these drugs is sedation, but this can benefit patients who have difficulty sleeping due to chronic pain. Some people experience intolerable drowsiness, dry mouth or constipation and these drugs are contraindicated for people with cardiac tachyarrhythmias.

The newer antiepileptic drugs are another important group of antineuralgic drugs. Gabapentin is licensed for the treatment of neuropathic pain. Its exact mode of action is not yet fully understood, although it appears to target calcium channels. It has been shown to reduce pain in a wide range of neuropathic pain conditions and induce better sleep, mood and quality of life (Serpell and the Neuropathic Pain Study Group, 2002). Side-effects tend to be uncommon, transient and mild, and include dizziness, lethargy, headache and confusion.

Capsaicin cream is derived from chilli peppers and causes a depletion of the neurotransmitter substance P from sensory nerves. It should be applied sparingly four times a day. Compliance can be limited by a burning sensation when applied, but anecdotal reports show SCD patients tend to like the feeling of heat it generates. Patients need to be educated about how to apply it correctly.

Local anaesthetics
Bupivacaine 0.25–0.5 per cent can be safely applied topically to leg ulcers during dressing changes to reduce pain during the procedure and for a few hours after.

Opioids
Tramadol can be a useful analgesic for chronic pain, since it has both weak opioid agonist and monoaminergic effects and is considered to have a low addiction potential (Sindrup and Jensen, 1999).

Since it is not a controlled drug, it is suitably accessible for patients. There is mounting evidence of the efficacy of opioids in neuropathic pain (Rowbotham et al, 2003), particularly when used in conjunction with tricyclics and/or anticonvulsants.

Consequently, pure opioids, such as morphine and oxycodone, are being used more for neuropathic pain, which is refractory to other analogesics, not amenable to surgery and significantly impairs a patient’s quality of life. For example, severe AVN of the hip causes immense pain on movement.

Buprenorphine transdermal patches offer a promising new option that would appear to have a substantially lower risk of sedation and respiratory depression than other strong opioids in clinical use (Budd, 2002). However, long-term opioid regimens for chronic SCD pain must be based on comprehensive assessment and rationalisation of benefit, be regularly monitored and should not be given parenterally, in line with recommendations from the Pain Society, Royal College of Anaesthetists, Royal College of General Practitioners and Royal College of Psychiatrists (2004).

Non-pharmacological approaches
Transcutaneous electrical nerve stimulation (TENS) and acupuncture, when delivered by experienced practitioners, represent safe therapies with broad anecdotal evidence of efficacy. Few complementary therapies are supported by scientific evidence of efficacy and are therefore rarely provided by the NHS.

However, SCD patients with pain that responds poorly to other treatments, or who experience unpleasant side-effects, may well wish to explore all the available options. Nurses can encourage patients to be realistic about the likely outcomes and assist them in accessing those therapies they feel will help aspects of their quality of life. Since stress and anxiety exacerbate pain, it makes sense for nurses to offer patients reassurance and support with relaxation.

Neural blockade
There is little evidence supporting neural blockade for chronic SCD pain, but it can represent a useful part of a comprehensive multidisciplinary approach where the benefits and risks have been carefully assessed. In particular, suprascapular blocks and lumbar plexus blocks can relieve AVN pain in the shoulder and hip respectively. Chemical lumbar sympathectomy may be performed to aid cutaneous perfusion and healing of some leg ulcers caused by SCD.

These procedures should be done by experienced pain specialists, following careful assessment of their appropriateness for individual patients.