The first standards for adults with sickle cell disease (SCD) were launched this month to provide guidance on the minimum levels of care expected (Sickle Cell Society, 2008).

Developed by an expert working party and published by the Sickle Cell Society, they recommend care follows the network model with hospital-based SCD specialist centres acting as ‘expert resources’ for local hospitals. Sickle cell and thalassaemia community (SCaT) centres/services, working with GP practices, will underpin integrated health and social care in the community to provide most care close to home.

This home care will be provided either by specialist nurses attached to SCaT centres or appropriately trained district nurses. Patient self-care will play a key role, so it is vital they understand how to manage pain and minor complications. Both patients and nurses need a thorough understanding of complications that require urgent attention.

THE DISEASE
SCD is the most common and fastest-growing genetic disorder in England (NHSAntenatal and Newborn Screening Programme, 2006). It comprises a range of conditions related to a haemoglobin mutation. Adult haemoglobin consists of two alpha globin and two beta globin chains wrapped around a haem (iron containing) molecule. In SCD a mutation in the beta chain means the haemoglobin has a lower affinity for oxygen.

People who inherit the mutation from both parents or the mutation from one and thalassaemia (lack of a beta chain) from the other will suffer sickling. Those who inherit the mutation from only one parent will be SCD carriers and rarely display symptoms.

SCD is most common in people of African descent but occurs in all malarial areas of the world with a prevalence of 10–30% as people with the mutation are resistant to malaria. As a result of migration, SCD now affects one in 2,400 babies in England and its prevalence is growing (NHS, 2006).

Sickle blood cells have a shorter lifespan than normal blood cells. Haemolysis and adhesion to blood vessel walls causes damage in the vasculature, and ultimately cerebrovascular incidents and end organ failure. Frequent painful attacks and other complications will reduce life expectancy to an average of 42 for men and 48 for women (Platt et al, 1994). It is vital to minimise complications and identify them promptly.

Although newborns are screened for SCD and standards for caring for children with the disease were published in 2006 (NHSANSP, 2006) these are the first standards for adult care. As a result there have been dramatic differences in the type and quality of care provided for adults.

COMMUNITY MANAGEMENT
In line with the National Service Framework for Long Term Conditions recommendations (Department of Health, 2005), the adult standards recommend care is delivered through networks built on existing services. They promote a multidisciplinary approach to care for patients close to or at home.

In high prevalence areas large hospitals may work together to provide specialised services across the network. In low prevalence areas an SCD specialist centre will supervise and share care with local hospitals, and there will be a greater role for primary and community care teams supported by SCaT community centres/services.

All patients should be seen at a specialised centre soon after diagnosis and
at least annually for review. But routine checks and acute and ongoing clinical care for some less complex complications should be available in local hospitals.

Access to dedicated community services will vary. High prevalence areas will often have SCaT community centres/services run by specialist nurse counsellors working with other health and social care providers. These will offer advice to patients and guidance to staff in more local community services with whom the patient may be in more regular contact. All patients should have telephone details of the nearest SCaT centre.

Each SCD specialist acute care clinical centre should have at least one such community care centre to act as a one-stop shop. These should offer screening services, information, a disease management counselling service, genetic counselling services and a facility for referral for prenatal diagnosis, benefits and psychosocial service advice, and facilities for support groups.

They can also offer courses for health professionals who work with patients with SCD, nurse-led clinics, day care pain management facilities and/or nurse home visiting to support self-management.

Some areas will have specialist nurse counsellors or outreach nursing teams to support patients managing uncomplicated pain crises at home. In others areas district nursing services can be trained to advise patients at home.

EDUCATING PATIENTS

Key to effective management is the patient having a clear understanding of their condition. They should be given information on how to minimise crises and manage symptoms, including uncomplicated pain, at home. They should also be provided with a range of pain killers.

Both patients and carers need to be aware of which symptoms require urgent medical attention. Access to the Expert Patients Programme (EPP) will aid this. Information on local patient groups and relevant national voluntary organisations, such as the Sickle Cell Society, is also useful.

All patients must have a named key contact to telephone for advice when necessary. This may be a specialist nurse counsellor, a ward or day-unit based nurse, or a member of the medical team.

MANAGING COMPLICATIONS

Most acute presentations to hospital by patients with SCD are due to painful crises. These account for 90% of resulting admissions. Other acute complications are less common but the morbidity and mortality associated with them is high (see box).

With an acute painful crisis, the first dose of a potent analgesic must be administered within 30 minutes of the patient’s presentation to the hospital clinical area (including time spent in triage) and the on-call haematologist notified immediately.

Pain management protocols agreed by the local SCD specialist centre should be easily accessible in clinical areas such as the emergency room, day hospitals and ward areas. In some cases patients may have individualised pain management protocols determined by the haematologists and nurse clinicians in charge of their care.

Uncomplicated painful crises are frequently accompanied by low-grade fever. Life-threatening infections are more common, so temperatures of >38°C should be investigated by blood/urine culture and chest X-ray and antibiotic treatment started immediately.

Acute chest syndrome (ACS) is a common form of acute lung injury that may lead to acute respiratory distress syndrome and death, so local protocols for painful crisis should include features suggestive of ACS with action to be taken.

ACS symptoms include tachypnoea, chest pain, cough or shortness of breath, generally with fever, in the presence of a new infiltrate on chest X-ray, although X-ray changes often lag behind clinical signs. ACS may not present immediately but can progress rapidly. Awareness, anticipation of its development and intervention with treatments such as oxygen and blood transfusion is key to management.

Half of all SCD patients will experience at least one ACS episode during their lives, so all patients and their carers should be aware that potential ACS symptoms, such as chest pain and shortness of breath, require urgent medical assessment. Nurses in the haematology, accident and emergency and acute medical teams should also be educated to recognise and manage ACS.

COMPILATIONS OF ACUTE SICKLE CELL DISEASE

- Painful crisis – responsible for most acute presentations to hospital
- Fever – requires investigation when >38°C because of the risk of life-threatening infections.
- Acute chest syndrome (ACS) – may lead to acute respiratory distress syndrome and death. May not present immediately. Nurses need to be aware of the symptoms and how to manage them.
- Acute abdomen pain – various causes, including gallstones, which occur in 70% of adults.
- Acute anaemia – full blood count and reticulocyte count required for any acutely unwell patient.
- Acute neurological symptoms – stroke is common (>5% prevalence). There should be specific guidelines for its management.
- Osteomyelitis – easily confused with a painful crisis.
- Acute renal disease – manifests as inability to urinate. Can cause dehydration.
- Acute priapism – can lead to erectile dysfunction. Early presentation <4 hours is vital for a successful outcome.

REFERENCES


For additional clinical information log on to nursingtimes.net and click on to NT Clinical and Archive.