# Sickle cell disease in surgery

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*Trop Doct* 2006 36: 200
DOI: 10.1258/004947506778604850

which is often fatal. An emergency cholecystectomy is then the only hope.

A variety of aetiologies may be found, which include cryptococcus, cytomegalovirus, microsporidia, as well as lymphoma and Kaposi Sarcoma.

Semi-elective cholecystectomy carries appreciable risks which are to some degree reduced by a laparoscopic technique if the expertise and technology are available, but still with considerable hazard. A percutaneous cholecystostomy may be a less hazardous option. Vague symptoms associated with ultrasonographic findings of a thickened gall bladder wall only are unlikely to be improved by operation.

Severe abdominal pain due to other HIV-related conditions may give rise to the suspicion of peritonitis and must be excluded in the differential diagnosis.

They include severe cystitis, a retroperitoneal abscess, necrotizing fasciitis of the abdominal wall, an abdominal wall abscess and intestinal wall haemorrhage from Kaposi Sarcoma mentioned above.

### Intestinal obstruction

Intestinal obstruction *per se* warrants a conservative approach of nasogastric drainage, restriction of oral intake and aggressive intravenous fluid replacement to correct fluid and electrolyte deficits. When colicky abdominal pain becomes constant and severe, or where there is no sign of resolution of obstruction in 48 h, or when signs of peritonitis supervene, then a laparotomy is indicated. In this respect, no difference exists between management of HIV-negative and -positive patients.

In HIV-positive patients the following specific causes of intestinal obstruction may be found:

Tuberculous adhesions/mass/intestinal stricture, Lymphoma of small bowel, Kaposi Sarcoma of small and large bowel, mesenteric lymphadenopathy and intussusception.

Tuberculous adhesions are often thick and unyielding (and may be detected as septa on ultrasonography in an ascites-filled abdomen). A tuberculous mass usually occurs in the right iliac fossa but any site may be affected, while an intestinal stricture occurs most commonly in the ileum and less so in the in the jejunum and can be found in both. These may be suspected on ultrasonography and confirmed by characteristic appearances on barium studies.

Great care should be taken when unravelling adherent loops of bowel because a postoperative enterocutaneous fistula is often fatal. This is particularly important where the aetiology is tuberculous.

Minimal surgical intervention is recommended, though sufficient to alleviate the obstruction. If abdominal tuberculosis can be confirmed, a conservative regimen of antituberculous therapy often results in complete resolution in subacute intestinal obstruction.

Complete bowel decompression prior to resection and anastomosis is recommended. Representative tissue should be sent for histological examination.

In a few cases, the signs of intestinal obstruction may be mimicked by a paralytic ileus thought to be related to an HIV neuropathy; in this circumstance, correction of fluid and electrolyte derangement should be followed by a conservative approach. As in pseudo-obstruction (Ogylvie's syndrome), the use of guanethidine and/or neostigmine can relieve the ileus completely.

### Discussion

The routine use of urinary catheters should be discouraged as the risk of sepsis far outweighs the advantages of an empty bladder. In men, a Paul's tubing (which can be fashioned from a condom) is perfectly adequate to monitor urine output.

Where the appendix is not seen to be the cause of peritonitis, its removal is not usually recommended: a faecal fistula may result.

Elective appendicectomy in asymptomatic individuals after successful conservative treatment of an appendix mass is also probably not indicated. Likewise, elective appendicectomy after drainage of an appendix abscess may not be warranted.

The use of drains is equivocal at best and they represent a foreign body; in HIV patients they are not recommended except where bowel or bladder fistulation is feared.

In cases of trauma the spleen should be preserved if at all possible; if splenectomy is inevitable, diced segments can be sutured *in situ* in an omental pouch, but the effects of further immune suppression may not thus be avoided.

# Sickle cell disease in surgery

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*TROPICAL DOCTOR* 2006; **36:** 200–202

### Introduction

Sickle cell disease (SCD) is a general term for a group of related genetic disorders caused by the presence of abnormal sickle haemoglobin (HbS) in which the red cells change shape upon deoxygenation because of polymerization of the abnormal sickle haemoglobin. The term sickle cell anaemia refers to the specific homozygous form of the disease, in which there are those with the disease and those who are carriers of the disease.

Most of the clinical manifestations of sickle cell anaemia can be explained by the propensity of the homozygous (HbSS) haemoglobin in affected red cells to sickle in an environment of hypoxia, hypothermia and dehydration. This causes vaso-occlusive episodes that result in drastically reduced blood flow to the tissues causing damage. Virtually any part of the body tissues and organs can be affected with the organs such as the spleen, liver, bone/bone marrow being particularly vulnerable.

This results in a high morbidity and mortality in persons with this condition. Life expectancy is to the fifth decade.

It can occur in any individual of any colour or ethnicity, but is most common in races who live in areas affected by malaria: in East and Central Africa the prevalence is reported as between 25 and 30% around Lake Victoria.

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It can occur in any individual of any colour or ethnicity, but is most common in races who live in areas affected by malaria: in East and Central Africa the prevalence is reported as between 25 and 30% around Lake Victoria.

While SCD is considered a medical disorder and not a surgical condition the effects of the disease impact on the surgical care of the affected patient, both with regard to the treatment of the specific vaso-occlusive events and also on elective surgical procedures carried out on those with SCD (sicklers). Surgery can be a risky undertaking in sicklers, but with good perioperative planning it can safely be undertaken.

## Clinical presentation

Clinical manifestations of the disease are variable and include repeated painful vaso-occlusive, haemolytic, aplastic, metaplastic episodes and sequestration crises. Complications affect any part of the body.

## Diagnosis

The disease is diagnosed clinically, based on a high level of suspicion, which may be based on the family history and where the individual came from.

There is often a persistent anaemia (6–9 g/dL) and signs of haemolysis including jaundice may be present.

Abnormal blood forms are detected by haemoglobin electrophoresis.

## Surgical aspects of SCD

The surgical aspects of SCD relate to both the effects of Sickling Crises on different tissues or organs of the body and to the precautions which must be taken in order to minimize the possibility of Sickling Crises when undertaking elective surgery.

### *Effect on organ systems*

As previously mentioned, any tissue or organ can be affected. The most significantly affected are:

*Eye* Retinopathy is a significant problem in 10–20% of the patients with SCD. The peak age of onset is in the twenties. This complication can produce sudden loss of vision. Laser photocoagulation has been used in an effort to prevent retinal haemorrhage. Another common problem is retinal detachment, particularly as a sequel to retinal haemorrhage.

*Spleen* Splenic sequestration crisis results from the acute entrapment of large amounts of blood in the spleen. Given the lower morbidity and mortality in adults, splenectomy is rarely a consideration. Normally surgery is indicated in hypersplenism associated with pancytopenia (anaemia, neutropaenia and thrombocytopaenia), splenic abscess and, less commonly, massive splenomegally.

Patients for splenectomy should receive polyvalent pneumococcal vaccine two weeks before elective surgery and as soon as possible after emergency splenectomy. This is followed by prophylactic use of penicillin until adulthood or for at least two years to prevent overwhelming postsplenectomy infection (OPSI).

Splenic abscess develops in 12–20% of patients with SCD. Pus taken for culture and sensitivity taken from splenic abscesses may be negative particularly if patients are given antibiotics.

The definitive treatment of splenic abscess is splenectomy; however, percutaneous drainage with low morbidity and mortality is possible.

*Gastro-intestinal system* Ischaemic colitis is common among the under eighteens and should always be considered in an SCD patient in crisis and who develops severe acute abdominal pain, rectal bleeding and signs of peritonism. Initial management includes nasogastric tube decompression, broad-spectrum antibiotics and haemodynamic support including exchange transfusion and resection of ischaemic bowel. Because of the risks associated with surgery the following differential diagnoses should always be considered: vertebral bone infarction with pressure on nerve roots, mesenteric and retroperitoneal lymph nodes, splenic infarction and abscess, hepatitis and hepatic abscess, acute pancreatitis and thrombosis of the portal system.

*Cholelithiasis* Cholelithiasis prevalence among SCD patients varies from 34 to 70% in the USA and around 30% in Jamaica and Africa and this increases to over 95% at 35 years of age. Cholecystectomy is indicated in these patients, with a reported morbidity of 37% for Open cholecystectomy and 7% for Laparoscopic cholecystectomy (LC). Choledocholithiasis occurs in 14–18% of patients and it is recommended that the common bile duct be explored during cholecystectomy.

LC is increasingly being offered for SCD patients with asymptomatic cholelithiasis and is the technique of choice with some series recording very low morbidity and no mortality.

Individuals with SCD may also develop hepatic crisis, which can lead to acute hepatic failure.

*Bone* Acute bone marrow necrosis is now commonly recognized as a complication of SCD, in part due to improved methods of detection. In the past the diagnosis could only be made by bone marrow biopsy or inferred from the complications that resulted and, if the necrosis occurred in regions of the marrow that were not easily biopsied, the diagnosis was almost impossible to confirm.

This has changed with the introduction of magnetic resonance imaging (MRI) techniques.

Pulmonary fat emboli can complicate bone marrow necrosis. Fat emboli can trigger respiratory insufficiency or even acute chest syndrome.

Avascular necrosis (AVN) of bone, which is distinctly different from acute bone marrow necros, is a common problem in patients with SCD. The areas most frequently affected are cortical bone of the acetabulum, the head of the femur and the head of the humerus. Humeral and femoral head core decompression is performed by some orthopaedic surgeons with success. This invasive procedure should be reserved for patients with more advanced cases of AVN. The efficacy of this is yet to be proven.

Most of the patients afflicted by AVN are in their twenties or thirties with the condition bad enough to warrant joint replacement. The decision to proceed with joint replacement is difficult as artificial joints are not well tolerated by patients with sickle syndromes. As many as one-third SCD patients require revision arthroplasty within four years of joint replacement.

Osteomyelitis often occurs at the site of necrotic segments of bone and requires bone biopsy for definitive

diagnosis. It should be treated with appropriate antibiotics for four to six weeks, given intravenously for best effect.

*Priapism* Priapism is a result of impaired blood flow from the corpus spongiosum of the penis, leading to prolonged erections. Priapism lasting more than 3 or 4 h is a medical emergency since it can produce impotence. Decongestive measures should be undertaken, but the results of treatment are variable and it is reported that treatment by irrigation of the ventral vein of the penis risks inducing impotence in up to a third of affected individuals.

*Skin ulcers* About 30% of patients with SCD disease develop skin ulcers with the most common site being over the lateral malleoli, often extending into the underlying subcutaneous tissue. They are prone to infection and treatment may take months. Conservative treatment with rest, elevation, dry dressings and antimicrobial ointment application is recommended as skin grafting frequently fails owing to poor blood supply.

### Considerations for elective surgery

Life expectancy among individuals affected by SCD is to the fifth decade, mainly due to improved medical care. As these individuals live longer they get affected by surgical conditions seen in the non-SCD population and thus we can expect more surgical intervention in these individuals.

*Perioperative considerations* In general, one needs to avoid factors that may precipitate a sickling crisis such as dehydration, sepsis, altitude, acidosis and hypoxia. The anaesthetist must be made aware of the condition and appropriate preparation of the patient undertaken. Higher levels of oxygen delivery are required.

The level of preparation will vary with the severity of affection by the particular individual and the magnitude of the surgery to be undertaken.

*Infection prophylaxis* Infection is a leading cause of death in patients with SCD. Hyposplenism due to splenic auto infarction is a major contributor. Hyposplenism is not the sole cause of the defective host defence as evidenced by the fact that overwhelming sepsis is the leading cause of death of children under three years of age.

It is recommended that all children up to age 16 years be placed on prophylactic penicillin at a dose of 250 mg twice a day. Patients with allergies to penicillin should be treated with erythromycin. The role of prophylactic penicillin in adults with SCD is unclear. Adults develop overwhelming sepsis, but at a much lower frequency than children.

*Immunization* Immunization with the pneumococcal vaccine is standard practice both in adults and children with SCD. Several studies suggest that immunization provides protection where splenic function has been lost. The more recently available 23-valent vaccine provides broader coverage than earlier versions, although the duration of protection is unknown. More recently a vaccine against *Hemophilus influenzae* has entered the clinical arena. The efficacy of this vaccine in SCD is unknown. Given the serious nature of *H. influenzae* infections in these patients, many specialists, particularly pediatricians, now routinely immunize their patients against this organism. Immunization against viral influenza is a common practice. Recently an effective vaccine against hepatitis B was developed. Since patients with SCD are likely to require one or more transfusions in their lifetime, immunization against hepatitis B is a reasonable precaution.

*Blood transfusion* SCD patients awaiting surgery will usually require a blood transfusion and may be divided into four groups depending on the magnitude of the intended surgery and the severity of the condition.

*Group one*
Patient well, no risk factors, procedure short, minimal perioperative complications.
*Top up Hb to $>7.0\,g/dL$, regardless of HbS levels.*

*Group two*
No special risk factors, intermediate risk surgery, e.g. herniorrhaphy.
*Top up Hb to 9–11 $g/dL$, irrespective of HbS levels.*
Hb not to exceed $12\,g/dL$.

*Group three*
H/O chest crisis, recent painful crisis, scheduled for major surgery, e.g. laparotomy.
*Exchange transfusion if HbS levels $<30\%$.*
Top up Hb to $12\,g/dL$.

*Group four*
CNS crisis, e.g. stroke, on regular transfusion programme, scheduled for major surgery, e.g. craniotomy/thoracic surgery.
*Exchange transfusion if HbS level at $<30\%$.*
Top up Hb to $12\,g/dL$.

## Conclusions

The adoption of a multidisciplinary approach to the care of individuals with SCD has led to a decrease in morbidity and mortality from the disease so that life expectancy is now up to 50 years. Surgery has a significant role in the treatment of the disease, both in dealing with sickling crises and electively. It has been shown that with the proper precautions, including adequate blood transfusion, elective surgery may be safely carried out. The use of minimally invasive surgery is up to five times safer than conventional surgery.