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# Acute abdominal conditions in people with sickle cell disease: A 10-year experience in Port Harcourt, Nigeria

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## Abstract

**Background:** Abdominal crises (vaso-occlusive) are not infrequent in patients with sickle cell anemia. They usually present as acute abdomen. These patients none the less present with other causes of acute abdomen like others without hemoglobinopathy. Six cases of surgical acute abdomen in sickle cell disease patients treated in the University of Port Harcourt Teaching Hospital (UPTH) are presented.

**Materials and Methods:** Six sickle cell anaemia patients presenting with acute abdominal conditions from 1999 to 2008 (inclusive) in the University of Port Harcourt Teaching Hospital were studied retrospectively (two patients) and prospectively (four patients). After initial resuscitation, appropriate surgical procedures, namely, appendectomy (2), cholecystectomy (1), open drainage of splenic abscess (1), and release of adhesive band (1) were performed.

**Results:** Six cases were treated (female/male ratio 2:1). Four patients were in the 11–20 year age group. Two patients had appendectomy and two presented with acute cholecystitis/cholelithiasis, one of whom had open cholecystectomy. One young girl had drainage of splenic abscess and a 42-year-old male had division of obstructive adhesive band at laparotomy. There were two complications, namely, postoperative haemorrhage (in cholecystectomy patient) and acute chest syndrome (in one appendectomy patient). These were successfully managed with blood transfusion and antibiotic/oxygen therapy, respectively. There was no mortality.

**Conclusion:** Sickle cell anaemia patients are not exempt from acute abdominal conditions requiring surgery. A high index of suspicion coupled with repeated clinical examination is needed for early diagnosis and appropriate treatment.

**Keywords:** Acute abdominal conditions, Port Harcourt, sickle cell anaemia

## Résumé

**Background:** Crises abdominales (occlusion) ne sont pas rares chez les patients atteints d'anémie falciforme. Habituellement, ils présentent comme abdomen aigu. Ces controversées de patients moins présents avec les autres causes de l'abdomen aigu, comme d'autres sans hémoglobinopathie. Six cas de chirurgie abdomen aigu chez les patients atteints de drépanocytose traités dans l'Université de Port Harcourt hôpital d'enseignement (UPTH) sont présentés.

**Matériaux et procédés:** Six patients de l'anémie falciforme présentant des conditions abdominales aiguës de 1999 à 2008 (inclusivement), l'hôpital d'enseignement de l'Université de Port Harcourt ont été étudiés rétrospectivement (deux patients) et pour l'avenir (quatre patients). Après réanimation initiale, échéant interventions chirurgicales, à savoir, appendicectomie (2), cholécystectomie (1), open drainage d'abcès rate (1) et la libération de bande adhésive (1) ont été réalisées.

**Résultats:** Six cas ont été traités (femmes/hommes ratio 2: 1). Quatre patients étaient dans le groupe 11 à 20 ans. Deux les patients avaient appendicectomie et deux présentés avec aiguë cholécystite/cholélithiase, dont l'un avait ouvert cholécystectomie. Une jeune fille avait drainage d'abcès rate et un homme âgé de 42 ans a division of obstructive bande adhésive à laparotomie. Il y avait deux complications, à savoir, l'hémorragie postopératoire (en

cholécystectomie patient) et le syndrome pulmonaire aiguë (dans une appendicectomie patient). Elles ont été gérées avec succès avec du sang transfusion et antibiotique/oxygénothérapie, respectivement. Il n'y n'avait aucune mortalité. **Conclusion:** L'anémie falciforme patients ne sont pas exemptés de conditions abdominales aiguës nécessitant une intervention chirurgicale. Une haute un indice de suspicion couplée à l'examen clinique répétée est nécessaire pour un diagnostic précoce et le traitement approprié.

**Mots clés:** Anémie falciforme conditions abdominale aiguë, Port Harcourt

## Introduction

The abnormal haemoglobin states have been known for many decades. The condition is genetically inherited with several varieties and worldwide distribution.<sup>[1]</sup> Genotypically homozygous (SS) individuals and double heterozygotes (SC, SB and SB+ thalassemias) have sickle cell disease. Such individuals are prone to painful vaso-occlusive crisis, chronic intravascular haemolysis and immune impairment.<sup>[2]</sup> The AS (heterozygous) state constitutes the trait and morphologically such individuals are normal. The incidence of the sickling trait varies widely from country to country. In West Africa, this varies from 10% in northern Ghana to 30% in northern Nigeria.<sup>[1]</sup> In Jamaica, a country populated predominantly by persons of West African descent, the prevalence of the sickle cell trait is 10%.<sup>[2]</sup>

The problems posed by a patient with sickle cell disease stem from vaso-occlusive crisis, intravascular haemolysis and immune impairment. The presentation is dependent on the organ(s) involved and may be a combination of symptoms and signs. Pain from vaso-occlusive crisis, anaemia, jaundice, and proneness to infection are typical features. Sepsis is in fact an ever-present problem in sickle cell disease and most of the time may be the precipitating factor in a crisis.<sup>[1]</sup> Pain emanating from abdominal organs will present with the acute abdomen. Such abdomen may be tender with guarding and rigidity and can be difficult to distinguish from the surgical acute abdomen.<sup>[1]</sup> Indeed, it is easy to misinterpret such a presentation as "ordinary sickle cell crisis" with consequent delay in diagnosis and treatment of a condition requiring surgical treatment.<sup>[3]</sup> When surgical intervention is necessary, proper attention is paid to the haemoglobin level, the problems of anaesthesia, hydration, acidosis, and risk of infection. Surgery is embarked upon only when the patient's clinical state is optimal.<sup>[4]</sup> Effective intraoperative and postoperative management is equally important for a satisfactory outcome. We present our experience with surgical management of six sickle cell patients in University of Port Harcourt Teaching Hospital (UPTH) over a 10-year period. The need for an increased index of suspicion and repeated clinical examination to determine whether surgical intervention is required is emphasized.

## Materials and Methods

Data were collected both retrospectively (1999–2001) for cases 1 and 2, and prospectively (2002–2008) for cases 3–6, who presented with acute abdominal pain. The 1999–2001 data were collected from theatre records and ward registers. A proforma was kept for the prospective cases. The data were analyzed with respect to age, gender, clinical presentation and diagnosis, investigations, surgical treatment, complications and outcome. As emergencies, all cases were initially resuscitated (adequate hydration, antibiotic therapy and pain relief once a definite diagnosis was made). Where a diagnosis could not be made immediately, patients were re-examined at regular intervals. All patients had general anaesthesia for surgery.

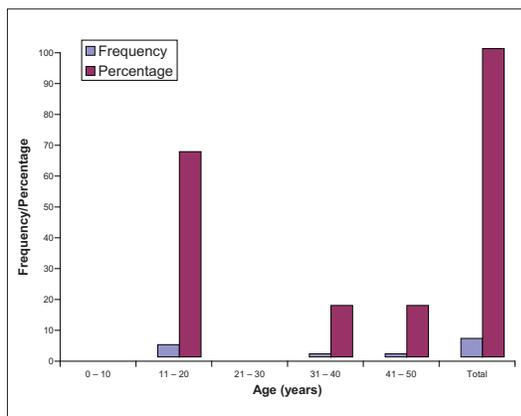
## Results

Six patients were treated over the study period. Females outnumbered males in a ratio of 2:1. Majority of the patients were in the 11–20 age group [Figures 1 and 2]. Two patients underwent appendicectomy for acute appendicitis [Table 1]. Both patients were initially assessed as having pain of vaso-occlusive crisis. The definitive diagnosis was made over a period of 24 hours with repeated clinical examination. At surgery, acute appendicitis was found in both cases and the diagnosis was confirmed by histopathology. Postoperatively, the 15-year-old developed chest symptoms with features suggestive of left lower lobe pneumonia. Chest radiograph showed patchy opacities. It was felt that the picture was consistent with the acute chest syndrome.

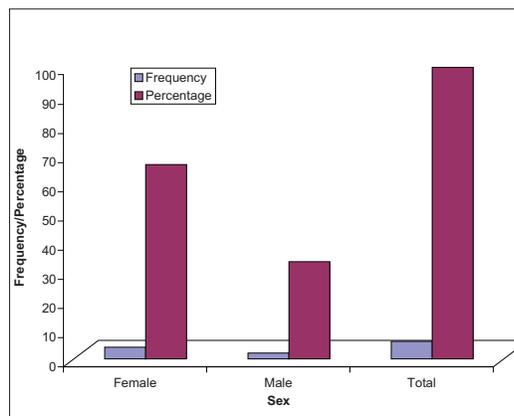
One female patient (34 years) and a male (17 years) also presented with acute right upper abdominal pain. Both had had recurrent pain in this part of the abdomen for 2 and 5 years, respectively. The attacks of pain had been associated with jaundice and the 17-year old male had telltale scarification marks over the right hypochondrium and epigastric region. A clinical diagnosis of cholecystitis was made. Abdominal ultrasound scan demonstrated multiple stones in both gall bladders. The haemoglobin levels were 11.3 and 7.2 g/dl, respectively. Triple antibiotic therapy (ampicillin/cloxacillin, gentamycin and metronidazole) was given. An urgent open

**Table 1: Particulars of six SS patients treated over a 10-year period**

Age	Sex	Diagnosis	Operation	Postoperative outcome
15	F	Splenic abscess	Laparotomy+drainage	Satisfactory
42	M	Intestinal obstruction	Laparotomy + release of adhesive band	Satisfactory
34	F	Cholecystitis/Cholelithiasis	Open cholecystectomy	Satisfactory
13	F	Acute appendicitis	Appendicectomy	Satisfactory
15	F	Acute appendicitis	Appendicectomy	Satisfactory
17	M	Cholecystitis/Cholelithiasis	Cholecystectomy awaited	



**Figure 1:** Age distribution of SS patients



**Figure 2:** Sex distribution of SS patients

cholecystectomy was done for the 34-year-old female by the 5<sup>th</sup> day of antibiotic treatment on account of continuing pain. Multiple pigment stones were found at surgery; she had 2 units of fresh whole blood transfused because of postoperative blood loss, which caused her haemoglobin to drop to 6.8 g/dl. The 17-year-old responded very well to antibiotic treatment; open cholecystectomy was scheduled. The surgery had to be cancelled on the appointed day because of industrial (strike) action by hospital workers. He is yet to report back for the operation.

A 15-year-old girl also came in with acute left hypochondrial pain, fever and jaundice. She had had recurrent pain at this site for over 3 years and this was evidenced by the presence of scarification marks over the left hypochondrium on examination. There was a tender mass over the left abdomen and this was shown to be cystic on ultrasound scan. Resuscitation was started with triple antibiotic therapy using ceftriaxone, gentamycin and metronidazole; 2 units of fresh whole blood were transfused as she was pale with haemoglobin of 4.6g/dl. At laparotomy 24 hours after resuscitation, a large abscess was found in the spleen; almost 90% of the spleen had been replaced by abscess cavity. The collection was drained and the remnant splenic tissue preserved. She made an uneventful recovery.

The sixth patient, a 42-year-old male without any previous surgery, was admitted with acute colicky

abdominal pain, vomiting and abdominal distension. His haemoglobin level was 9.1 g/dl and he showed moderate dehydration. A clinical diagnosis of acute intestinal obstruction was confirmed by plain supine/erect abdominal radiographs. He was resuscitated with intravenous fluids, nasogastric suction and antibiotics (ceftriaxone, gentamycin and metronidazole). At laparotomy, an adhesive band obstruction was found 16 cm from the ileocaecal junction. The band was divided.

The main complications were postoperative bleeding following urgent open cholecystectomy and acute chest syndrome following appendicectomy in the 15-year-old girl. The young girl developed chest symptoms and signs suggestive of left lower lobe pneumonia (later diagnosed as acute chest syndrome). Chest radiograph showed patchy opacities and she responded well to antibiotic treatment; no blood transfusion was given. There was no mortality.

## Discussion

It is not surprising that the majority of patients were in the age group 11–20 years. It is only in recent times that the average life expectancy of the sickle (SS) cell patient has been improved by advances in medical treatment. By 1973, the life expectancy of a patient with sickle cell disease was about 14 years; but 30 years later, this has improved to about

50 years because of improved management of the condition.<sup>[4,5]</sup>

Typically, patients with SS disease present with intermittent haemolytic and vaso-occlusive attacks of which pain is a prominent feature. When abdominal, it is very easy (or tempting) to ascribe such pain to what could be described as “normal pain of crisis”. Adopting such an attitude could prove disastrous when the condition turns out to be the one that actually requires surgical treatment. The way to prevent such an occurrence is to continue to monitor and observe these patients with repeated clinical examination. Such clinical re-assessment should be complemented by any ancillary investigations that may be considered necessary. Investigations like abdominal ultrasound scans and radiographs could be found useful as was our experience in these series.

Acute appendicitis is the commonest cause of the acute abdomen in UPTH.<sup>[6]</sup> This is the experience of authors from other centres, both locally and foreign.<sup>[7]</sup> Even when a known SS patient presents with pain of vaso-occlusive crisis, the possibility of appendicitis should therefore still be considered when the clinical features are suggestive. It has been noted that appendicitis can mimic pain of haemolytic/vaso-occlusive crisis with the pain localized to the right lower quadrant.<sup>[3]</sup> The two patients who had appendectomy were initially thought to have “abdominal pain of sickling crisis”. Of the nine cases of acute appendicitis in sickle cell disease patients managed by Al-Salem *et al*, six were perforated, the high incidence of perforation being attributed to the blocking of appendiceal vessels by sickled red blood cells.<sup>[8]</sup> The two cases in this study were, however, not perforated.

Owing to repeated haemolytic episodes, patients with SS disease are prone to develop gall stones over time. Such stones tend to be of the bilirubinate (pigmented) variety. The prevalence of gall stones in this condition is said to increase with age, ranging from 9% in the 5–7 age group to 83% in adults.<sup>[2,9]</sup> When such a patient presents with right upper abdominal pain and features of cholecystitis, no time should be wasted in performing an abdominal ultrasound scan. In our environment where malaria is endemic, red blood cell destruction from chronic haemolytic attacks is common. Pigment stone formation, therefore, occurs over the years.<sup>[3]</sup> The SS patient is consequently probably “doubly” exposed to forming pigment stones.

There is no controversy about the need to remove symptomatic gall bladder stones as was the situation in two cases in this study (one is yet to

have cholecystectomy). Many children with gall bladder stones are asymptomatic;<sup>[3]</sup> a great number of surgeons now remove asymptomatic stones in SS disease patients.<sup>[10,11]</sup> This approach has been regarded by some other workers as controversial.<sup>[12]</sup> In a study of 226 children with SS disease, Duncan *et al*. found that only 1% per year of these patients with gall stones developed symptoms.<sup>[9,13]</sup> Prophylactic cholecystectomy in asymptomatic children has been criticized by some other authors too.<sup>[14]</sup> The basis of such criticism is that common bile stones are not prevented, the majority of adults with gall stones do not develop complications and that preservation of the gall bladder may have a protective effect against right-sided colonic carcinoma.<sup>[3]</sup> With introduction of minimal access surgery, laparoscopic cholecystectomy has been advocated as the procedure of choice since it reduces perioperative morbidity. However, although laparoscopic cholecystectomy gives improved surgical outcome in SS disease, the cost of equipment and disposables places this mode of treatment out of reach of the general public in most developing countries.<sup>[2]</sup>

These patients are at an obvious surgical risk, especially when subjected to general anaesthesia. Such risk stems principally from hypoxia; other factors are hypotension, obstructed airway, hypothermia and unaccepted haemoglobin level. There is also the problem of insufficient time for preoperative preparation and optimization. The choice of general anaesthesia, with rapid sequence induction, intubation, muscle relaxation and controlled ventilation with 50% inspired oxygen as was done in all these cases ensured adequate oxygenation and normocarbia. Close monitoring and adequate hydration with warm intravenous fluids was maintained throughout the perioperative period. Despite the challenges, with careful perioperative anaesthetic management, patients with sickle cell disease may undergo multiple surgical operations with successful outcomes as shown by one of the patients who had caesarean section in May 2001, open cholecystectomy in 2002, and repair of a symptomatic supraumbilical hernia 1 year later.

Normally, patients for surgery under general anaesthesia are, among other requirements, expected to have minimum haemoglobin of 10g/dl. Patients with SS disease, however, normally have a haemoglobin level below this value, i.e., usually between 7 and 8g/dl. At this steady-state value of haemoglobin, blood transfusions are not routinely required. Preoperative blood transfusions are given only if admission haemoglobin levels are more than 1g/dl below steady state.<sup>[2]</sup> If such blood is to be given, it should be fresh (to retain platelet activity), as stored blood is acidotic and hyperkalemic.

These are conditions that can precipitate sickling.<sup>[1]</sup> Moreover, there should be clear indications for such blood transfusions.<sup>[4]</sup> The two transfusions given in this study were for haemoglobin levels of 4.6 and 6.8g/dl, respectively. A straight transfusion, where additional blood is given without removing sickled blood, is advised when haemoglobin level is less than 8 or 9g/dl. On the other hand, an exchange transfusion is advocated for situations with higher haemoglobin levels.<sup>[4]</sup> Exchange blood transfusion reduces the number of sickle cells and increases the number of normal red cells, thereby reducing the frequency of sickling episodes.

The spleen in SS disease is prone to chronic organ damage as a result of repeated infarctions. The resulting fibrosis may produce splenic atrophy and even autosplenectomy.<sup>[4]</sup> On the other hand, an area of splenic infarction can suffer secondary infection and subsequently form an abscess<sup>[15]</sup> as it happened in the 15-year-old girl in this study. The attempt to preserve even the little splenic tissue was meant to help reduce the risk of development of the dreaded overwhelming post splenectomy infection.

Open cholecystectomy is yet to be performed for the 17-year-old boy whose surgery was cancelled because of an industrial (strike) action by the hospital workers. Cancellation of surgical cases in hospitals in this environment is not an uncommon occurrence. The reasons for such cancellations may be patient, hospital or surgeon related.<sup>[16]</sup> The inconvenience, pain and extra cost to patients by such cancelled operations cannot be quantified.

As regards the complications, it was not very clear why the 34-year-old female had such significant postoperative bleeding requiring 2 units of fresh blood. It is possible that because surgery was done on an urgent basis (5<sup>th</sup> day after starting antibiotic therapy), the acute inflammation had not settled satisfactorily. It is known that elective cholecystectomy has a lower complication rate than emergency intervention.<sup>[17]</sup> The postoperative chest complication in the 15-year-old girl was later diagnosed as an acute chest syndrome on account of patchy opacities that were demonstrated on chest radiograph. The acute chest syndrome is the second most common cause of admission and a leading cause of mortality in all age groups with sickle cell disease.<sup>[4]</sup> In their report on biliary surgery in sickle cell disease in Jamaica, Duncan *et al.* found the acute chest syndrome as the principal source of postoperative morbidity and mortality.<sup>[2]</sup>

Mortality figures for surgery in sickle cell disease patients are generally low with improvement in

medical care. In a study of 1079 surgical patients published in 1995, perioperative mortality was low at 1.1%.<sup>[18]</sup> A contemporary publication of a series of 604 cases also noted a rate of acute sickle cell exacerbations of approximately 15% with a mortality rate of 0.3%.<sup>[19]</sup> In 33 patients undergoing 50 surgical procedures, Oduntan and Isaacs recorded only 2 deaths, one unrelated to the abnormal haemoglobin state, while in the other, it could not be entirely excluded.<sup>[20]</sup> We did not record any mortality in our series although the number is rather low.

In conclusion, because SS disease is associated with pain of vaso-occlusive crisis, there is a tendency to ascribe acute abdominal pain of surgical importance to such attacks. A high index of suspicion coupled with repeated clinical examination is required if catastrophic consequences are to be avoided in the management of these patients. We also recommend multidisciplinary management involving haematologists and anaesthesiologists for these patients. This will decrease the overall complication rates and improve clinical outcome.

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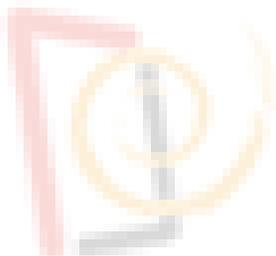
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