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**Review article:**

# Study of surgical aspects of sickle cell disease - Pathophysiology with review of literature

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**Abstract:**

**Objectives :** To study the surgical presentations of sickle cell anaemia (SCA) in a sickle cell belt in India and to discuss the historical aspects and pathophysiology of sickling and its sequelae.

**Methodology :** One hundred patients attending the surgical outpatient were screened for Sickle cell disease and the signs and symptoms were recorded as to the type of crisis they presented with.

**Results:** Out of a hundred patients included in this study ninety patients presented with SCA and ten with Sickle cell trait (SCT). 53% patients presented with vasoocclusive crisis. Leg ulcers and abscesses formed the next most common modes of presentation. Wound infection and delayed wound healing were the most commonly occuring complications.

**Conclusion:** Patients of Sickle cell anaemia presented mainly with vasoocclusive crisis requiring a proper evaluation and careful management of surgical procedures. Distinguishing between vasoocclusive crisis or surgical cause of abdominal pain is paramount to avoid unnecessary surgical procedures. Surgical patients require proper hydration, blood transfusion and proper oxygenation to avoid post operative complications. Use of minimal invasive surgical techniques wherever possible obviates complication rate. Proper genetic counseling of susceptible population is essential.

**Keywords:** Sickle cell disease, vasoocclusive crisis, abdominal pain, drepanocytosis

## Introduction:

Sickle-cell disease (SCD), also called drepanocytosis, is a hereditary blood disorder, characterized by an abnormality in the oxygen-carrying haemoglobin molecule in red blood cells that leads to a tendency for the red cells to assume an abnormal, sickle-like shape under certain circumstances. Sickle-cell disease is associated with acute and chronic diseases, such as severe infections, attacks of severe pain ("sickle-cell crisis"), and an increased risk of death. Sickle-cell disease occurs when a person inherits two abnormal copies of the haemoglobin gene, one from each parent. Sickle cell anemia (SCA) results from substitution of a single amino acid, namely valine for glutamic acid in the beta globin gene at chromosome number 11.[1] The susceptibility of red cells to sickle correlates well with the concentration of sickle haemoglobin within the red cells. Several factors influence the course and severity of the disease. These include the presence or absence of alpha thalasaemia



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gene, oxygen content of the inspired air, cardiac and pulmonary status. Other factors such as dehydration, infection, acidosis and hypothermia may precipitate some of the disease complications. Several subtypes exist, depending on the exact mutation in each haemoglobin gene. A person with a single abnormal copy is usually asymptomatic and is said to have sickle cell trait. This prospective study in a sickel cell belt in India aims to study various surgical presentations in sickle cell disease patients coming to the department of surgery ; study various complications in sickle cell disease patients and to find the association between sickle cell disease and surgical presentation.

Historical aspects :

Sickle cell disease has been known to people from West Africa from several centuries. The tribal people gave unique names to it, which were onomatopoeic and imitated the cries and moans of the sufferer. All these names repeating syllables , possibly denoting the repetitive painful episodes. Some of the names included nwiiwii (Fante tribe), chwecheechwe (Ga Tribe), nuidudui (Ewe Tribe).One of the earliest papers describing SCD in the West was published in 1846 [2], in Southern Journal of Medical Pharmacology titled case of Absence of Spleen. It mentioned the autopsy findings in a slave who had been executed.

Symptoms of SCD could be tracked back to year 1670 in one Ghanian family.[3] The credit of discovery of sickle cell disease goes to Dr. James Herrick, who provided the first formal description of sickle cell anemia when he reported that the blood smear of a dental student at the Chicago College of Dental Surgery contained "pear-shaped and elongated forms" in 1910. Six years earlier (i.e. in 1904) he had examined a twenty year old Caribbean, Walter Clement Noel, a dental student who had been admitted to the hospital with cough and fever. Examination of the blood smear revealed the presence of unusual red cells which were described by Herrick in the following words, the shape of the red cells was very irregular, but what attracted attention was the large number of thin, elongated , sickle-shaped and crescent-shaped forms. Cook and Meyer (1915) were the first to suspect a genetic basis for the disease as three siblings of their patient had died due to severe anemia. Sickling was demonstrated by them both in the patient and her asymptomatic father. The term "sickle cell anemia" was first coined by Mason in 1922.

In 1923 Taliafero and Huck hypothesized that, the sickle cell disease is an inherited disease and the mode of inheritance is based on Mendelian recessive character. In 1924, it was Sydenstiker , who gave the first details of clinical description of sickle cell disease and introduced the term crisis seen during the course of the disease.

Hahn and Gillespiell (1927) documented the role of reduced oxygen tension and reduced pH in in-vivo production of sickle cells. Waugh and Servier et al (1930) showed that the sickling process is directly dependent on hypoxia and increased carbon dioxide which induce the process of sickling. The study of Diggs et al (1939) revealed the irreversible sickle cells which do not revert to its original shape after deoxygenation. Sherman (1940) reported that the sickling of red blood cells in the absence of oxygen is caused by a change in the hemoglobin molecule structure. Janet Watson (1948) suggested that the



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presence of fetal hemoglobin in the red blood cells of sickle cell newborns is the reason they do not show disease symptoms. Physical chemist Linus Pauling and associates (1949) explained how protein electrophoresis showed that the sickle cell hemoglobin differed in structure from the normal hemoglobin moeity. This was the first time that the cause of a disease was linked to a change in protein structure. Vernon Ingram and J.A. Hunt (1956) sequenced sickle hemoglobin and showed that glutamic acid at position 6 was replaced by a valine in sickle hemoglobin. Using the known information about amino acids and codons that coded for them, he was able to predict the mutation in sickle cell anemia. This made sickle cell anemia the first genetic disorder whose molecular basis was known. Konotey Ahulu 5 (1974), presented the summary of clinical manifestations of the disease and classified the crisis.

Walter Gilbert and Frederick Sanger ( 1977), working separately in the United States and England, developed a new technique for rapid DNA sequencing. This led to the identification of the mutation in the beta globin gene. Dr. Charache reported that the anticancer drug hydroxyurea is the first to reduce the frequent, painful complications that characterize sickle cell disease.

Geographical distribution of sickle cell disease [4]:

SCD presents a major medical problem in tropical Africa, the Caribbean, the Middle East and the Indian subcontinent. It exists in all countries of Africa and in areas where Africans have migrated.[5] The transatlantic slave trade was largely responsible for introducing the sickle cell gene into the Americas and the Caribbean. However, SCD had already spread from Africa to Southern Europe as a result of the slave trade, so it also manifested in Portuguese, Spaniards, French Corsicans, Sardinians, Sicilians, mainland Italians, Greeks, Turks and Cypriots. Although the disease is endemic only in certain parts of the world and in ethnic minorities of the inner city areas of the western world, greater population mobility makes recognition of this disease of paramount importance in surgical practice. This study was done in an endemic area in India.

SCD appears in most of the Near and Middle East countries including Lebanon, Israel, Saudi Arabia, Kuwait and Yemen.[6] The condition has also been reported in India and Sri Lanka. SCD is an international health problem and truly a global challenge.

The prevalence is quite variable, but it is estimated that 8% of black population in America and 40% of the population in certain countries of tropical Africa have the sickle cell trait. In Jamaica, 8% of black people carry the sickle cell gene.

The prevalence of the disease in the United States is approximately 1 in 5,000, mostly affecting Americans of Sub-Saharan African descent, according to the National Institutes of Health.[4] In the United States, about 1 in 500 African-American children born will have sickle-cell anemia.

The Red Blood Cell :

Red blood cells (RBC) are the most common type of blood cells and the vertebrate organism's principal means of delivering oxygen ($O_2$) to the body tissues via the blood flow through the circulatory system. They take up oxygen in the lungs and release it while squeezing through the body's capillaries. Human, mature red blood cells are flexible biconcave disks that lack a cell



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nucleus and most organelles. 2.4 million new RBC' s are produced per second. The cells develop in the bone marrow and circulate for about 100–120 days in the body before their components are recycled by macrophages. Each circulation takes about 20 seconds. Approximately a quarter of the cells in the human body are RBC" s. RBC"s consist mainly of hemoglobin ( Hb), a complex metalloprotein containing heme groups whose iron atoms temporarily bind to oxygen molecules ($O_2$) in the lungs and release them throughout the body.[7]

The RBC"s have other functions that are equally important for human beings and are usually less emphasized :

a) When RBC undergo shear stress in constricted vessels, they release adenosine tri phosphate (ATP) which causes the vessel walls to relax and dilate so as to promote normal blood flow.

b) When their Hb molecules are deoxygenated, RBC"s release S-nitrosothiols which acts to dilate vessels, thus directing more blood to areas of the body depleted of oxygen.

c) It has been recently demonstrated that RBC"s can also synthesize nitric oxide enzymatically, using L-arginine as substrate, just like endothelial cells. Exposure of RBC"s to physiological levels of shear stress activates nitric oxide synthetase and export of nitric oxide, which may contribute to the regulation of vascular tone.

d) RBC"s can also produce hydrogen sulfide, a signaling gas that acts to relax vessel walls. It is believed that the cardio protective effects of garlic are due to erythrocytes converting its sulfur compounds into hydrogen sulfide.

e) RBC"s also play a part in the body's immune response: when lysed by pathogens such as bacteria, their hemoglobin releases free radicals which break down the pathogen's cell wall and membrane, killing it.

Type of Hb present in the red cells differs between fetal, neonatal and adult life. Analysis of globin chains from any purified Hb demonstrated that the tetramer was composed of two pairs of non-identical globin chains, belonging to the alpha and non alpha gene (beta-globin) gene family.

How is sickle cell anemia inherited?

Sickle cell anemia is inherited as an autosomal (meaning that the gene is not linked to a sex chromosome) recessive condition whereas sickle cell trait is inherited as an autosomal dominant trait. This means that the gene can be passed on from a parent carrying it to male and female children. In order for sickle cell anemia to occur, a sickle cell gene must be inherited from both the mother and the father, so that the child has two sickle cell genes ( Figure 1). The inheritance of just one sickle gene is called sickle cell trait or the "carrier" state. Sickle cell trait does not cause sickle cell anemia. Persons with sickle cell trait usually do not have many symptoms of disease and have normal hospitalization rates and life expectancies. When two carriers of sickle cell trait mate, their offspring have a one in four chance of having sickle cell anemia.[8]

Pathophysiology of sickle cell disease and crisis [9]:

In SCD the normal adult Hb(A) is replaced by the defective Hb (S). SCD occurs in individuals who are homozygous for a single nucleotide substitution in the beta globin gene, which renders the hemoglobin (Hb S) much less soluble than normal hemoglobin (HbA).The sickle mutation substitutes thiamine for adenine in the sixth



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codon of the beta gene (GAG ,GTG), thereby encoding valine instead of glutamine in the sixth position of the beta chain. This minor change results in profound changes in molecular stability and solubility of the Hb molecule.

Deoxygenated HbS undergoes a pronounced decrease in solubility and increase in viscosity. Deoxy HbS polymers in the cell exist as highly ordered fiber aggregates which fill the cell and distort it in to classic sickle shape or other elongated forms. It is the presence of polymer that causes the reversible, oxygen linked changes in the rheological properties of the disease. Impedance to blood flow is the key rheological determinant of pathophysiology of sickle cell disease.

Kinetics of sickling:-

A red cell spends approximately 1 to 2 seconds in the arterial circulation and 1 second to return to lungs.[11] This is called the delay time. Crises occurs when the delay time is shortened or capillary transit time lengthened enough to increase significantly this probability of crisis. Thus, if the delay is longer than 15 seconds; the cell can return to the lung and be reoxygenated before any significant polymer-ization.If the delay time is between 1 to 15 seconds gelation occurs while the cell is in the venous circulation.

If the delay time is less than approximately 1 second, gelation can occur while the cell is in one of the narrow vessels of microcirculation. Because the cell is much less deformable, it may not get permanently stuck. Sickling is not an instantaneous phenomenon and the process of molecular polymerization can be divided into various stages for its understanding. The delay period between deoxygenation and polymerization has been termed as nucleation process, in which HbS tetramers form small aggregates without any change in the internal viscosity. When these aggregates reach a critical mass, a rapid addition of free Hb unit occurs to form fibers, which then align to form a tactoid.[12,13]

.Under physiological conditions , the delay between complete deoxygenation and erythrocyte sickling is nearly 2 seconds, but it is influenced by other factors such as change in Hb concentration, the presence of hemoglobin other than HbS, temperature, pH ; 2,3,__Diphosphoglycerate (DPG) and Mean Corpuscular Haemoglobin Concentration (MCHC) levels.[14] Among these factors MCHC plays an important role. Small increments in deoxy -Hb concentration (e.g., those that occur with loss of cell, water) profoundly shorten the delay time, thereby, promoting sickling.

Sickle cell crisis:-

The term sickle cell crisis used to describe several independent acute conditions occurring in patients with SCD . SCD could result in anemia and crisis that could be of many types including vasooclusive crisis, aplastic crisis, sequestration crisis, hyper hemolytic crisis. Most episodes of sickle cell crisis last between five and seven days.

A) *Vasoocclusive Crisis:-*

It consists of a sudden attack of bone pain usually in the limbs, joints, hand, chest and abdomen. Painful crises are more frequent in children and it tends to decrease with advancing age. The vicious cycle is in the in-vivo sickling, causing increased blood viscosity and hypoxia



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which further increases the viscosity causing further stasis. Organs with a high oxygen uptake (i.e. brain and kidney) and organs which have sinusoidal vessels and slow flow rate (Spleen, liver, bone marrow) are more prone for this crisis. The crisis may persist for few hours to several days.

Venous thrombosis has been observed in superficial and deep veins of the extremities, splenic and meningeal veins and dural tissues. The thrombi in these veins are extension of intravascular clots. Arterial and pulmonary emboli are common in sickle cell disease who present with cough, haemoptysis, expectoration and pleural rub. Necrosis of bone marrow may lead to mobilization of fat leading to fat emboli which lodges in pulmonary arteries leading to infarction.

B) *Haemolytic Crisis:*

It is characterized by a precipitous drop in the erythrocytic values with increase in serum bilirubin, urine urobilinogen , faecal stereobilinogen and hypercellularity in the bone marrow. There is an increased number of immature cells in the peripheral blood . Cases with splenomegaly are more prone to hemolytic crisis.

C) *Aplastic or Hypoplastic Crisis:*

It is characterized by a sudden decrease of cellularity and maturity of cells in bone marrow. Hence it is associated with a striking decline in values of erythrocytes, a low reticulocyte count, leucopenia and thrombocytopenia . After a few days the marrow is replaced by intense erythropoietic hyperplasia. Normoblasts and reticulocytes appear in the peripheral blood and their number increases rapidly in circulation. Hb crisis to its original level over 3 weeks. Folic acid deficiency and infection with parvo virus are thought to be the precipitating factors.

D) *Sequestration Crisis:*

This is characterized by a sudden fall in Hb level with acutely enlarged spleen due to rapid pooling and sudden sequestration of large amount of blood in the spleen. This is the characteristic of sickle cell beta thalassaemia disease. It also occurs in sickle cell anaemia. There is a sudden drop in Hb level with hypovolemic shock and death in some children.

Observation :

This study of 100 patients (n=100) of sickle cell disease revealed a male preponderance (n=64) and female (n=36) (Table 1). The predominant age of presentation in this study was >40 years. However there were four patients below the age of 10 years. Abdominal symptoms, vomiting, fever and weakness formed the main presenting complaints. Pain in abdomen was present in 67 (67%) patients in this study. 69 patients complained of generalized weakness, fever and cardiorespiratory symptoms which accounted for 58 and 26 patients respectively (Table 2). Haematuria, bony pain, discoloration of digits and ulcer over the body constituted a smaller group.

Clinical signs of presentation included pallor as a distinct forerunner followed by abdominal tenderness and hepatosplenomegaly ( Table 2). There were thirteen cases of various grades of splenomegaly of which nine cases presented in crisis ( Sequestration and Hemolytic). One case presented as Aplastic crisis diagnosed due to pancytopenia and erythropoietic hyperplasia on bone marrow exam.

This study revealed that the vasoocclusive crisis was the predominant mode



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of presentation (53%) whereas 37% of cases presented with surgical conditions without manifesting crisis ( Table 3).

Haemoglobin electrophoresis revealed 10% of patients demonstrating sickle cell trait Haemoglobin AS ( Hb AS) and 90% demonstrating Haemoglobin SS ( Hb SS) ( Table 4). There was no history of malaria in patients of the former. Leg ulcers and abdominal emergencies formed half of the case presentation. Although priapism is a common form of presentation this series had only one such case. The modes of surgical presentation in this series was leg ulcers and abdominal pathologies like appendicitis, cholecystitis, intestinal perforations and intestinal obstruction. Visceral malignancies were seen in nine patients. Abscesses made up for ten patients ( Table 5).Ten cases of Sickle cell trait were detected on routine screening of patients attending the outpatient department for surgical problems of hernia, carcinoma penis, urolithiasis.

Discussion :

Sickle cell disease (SCD) is a single gene disorder causing a conglomeration of severe systemic symptomatology characterized by chronic anaemia, acute painful crisis episodes, organ infarction and chronic organ damage and by a significant reduction in life expectancy. SCD manifestation lies in the malarial belts of the tropics where the carriers are protected against death from malaria. Hemoglobinopathies are a highly prevalent group of hereditary disorders of hemoglobin (Hb) characterized by the presence of an abnormal β-globin chain (sickle cell disease) or a decrease or absence of α - or β –globin chains (thalassemias). The sickle hemoglobin (HbS) is the most common Hb variant.[16] It would be prudent to mention that SCD can be categorized into two arms; one representing Sickle cell trait (SCT) and the other Sickle cell anaemia (SCA). The SCT patients are usually asymptomatic and represent the carrier state. This series had 10 patients with SCT and 90 with SCA.

In India there are various pockets of sickle cell belt scattered all over the nation with a predominance in central India. Sickle cell gene was first identified among the tribals of Nilgiri Hills by Lehman and Cutbush (1952). Thereafter , more than 300 tribal groups have been screened to detect the sickle cell gene. The prevalence varies considerably among different tribal groups ranging from 0-35%. In certain states like Madhya Pradesh, Orissa, Chattisgarh, Jharkhand, Gujarat and Maharashtra it forms a major public health problem. 1981 census revealed the prevalence of Hb S in various populations studied, Rao (1988) estimated the expected number of sickle homozygotes as 1,31,375 in India and the expected number of sickle cell heterozygotes was 24,34,170. Due to a recent surge in population migration the spread of SCD has seen a diverse spread making it difficult to identify and counsel the affected families and making surveillance difficult.

Vaso-occlusive phenomena and hemolysis are the clinical hallmarks of SCD. This study deals with the various surgical manifestation of SCD. As surgical management of these patients is associated with high morbidity and mortality, a close interaction between medical and surgical branches is paramount.

Abdominal pain is a common mode of presentation during sickle cell crises. Its



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pathogenesis is not fully understood but it presents a diagnostic challenge as it mimics a wide spectrum of surgical emergencies that are encountered in day to day surgical practice.[17]. It is of vasocclusive origin, often termed "girdle syndrome" because of the circumferential distribution of the pain. Sixty seven patients in this series presented with this symptom of which twenty seven patients had to be wheeled up for surgery on emergency basis for gastroduodenal perforation, small bowel perforation, bowel obstruction, cholecystitis and appendicitis.

As duodenal ulcer (DU) in sickle cell populations does not appear to be related to high acid output, it is believed that the aetiology is related to the decreased mucosal resistance caused by repeated ischaemic infarcts secondary to sickling crises.[18,19]Organs having a sinusoidal vascular pattern are commonly responsible for abdominal pain due to sequestration of red cells. Acute splenic sequestration crisis, splenic infarction and abscesses, hepatobiliary causes such as biliary colic, acute cholecystitis, hepatic crisis and liver abscesses are common, which has been corroborated in this study with twenty two cases.[20]Anaemia and its various sequelae are commonly seen particularly in the paediatric age group and alongwith total leukocytic count could be used as a prognostic indicator of a more severe disease in adulthood . A low level of haemoglobin and an elevated total leukocytic count are ominous signs of an impending higher mortality and morbidity in childhood or adult life.[21] These can be used as a simple tool for screening children in high prevalence areas. In this study we had only four children below the age of 10 years who had Hb <10 Gm% but when the entire sample is considered it had sixty one patients having Hb < 10 Gm% out of eighty five patients having clinical pallor. Thus a prospective study using these parameters would be helpful. We encountered ten children with non surgical presentation of SCD. They presented with dactylitis, fever and chest symptoms and were treated by the department of pediatrics.

Vasoocclusive crisis form a major symptom of SCD characterized by bone and joint pains, chest pain and abdominal pain caused by vascular thrombosis and emboli. This produces capillary stasis, formation of plugs of red cells, thrombi, and vascular occlusion causing infarction and ischemic necrosis. This is known as "logjam" phenomena. More than half of the patients in this series (53%) presented with vasoocclusive crisis. This symptom of pain has been also used by various authors to determine mortality and morbidity by using "pain-rate" as an indicator. The "pain rate" is calculated by dividing the number of episodes of pain by the number of patient-years. Episodes that manifested within a two-week period were counted as one episode . Patients with an average of three or more episodes per year had a higher mortality rate than those with fewer than three episodes per year. A high pain rate is a marker for early death in these young adults particularly above 20 years of age.[22]

In this series ten patients presented with aplastic, sequestration and hemolytic crisis. Leg ulcers also form a frequent mode of presentation. They develop in 25 to 100 % of patients with SCA during their lifetime. The incidence of leg ulcers in the United States is about 10 per 100 patient years. These ulcers are resistant to



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treatment, are recurrent and cause considerable physical and psychosocial disability.[23] This occurs due to sickling of red cells leading to blocking of capillaries and ischaemia. Injury to red cells leads to release of integrins leading to platelet aggregation and adherence of sickled cells to vascular endothelium leading to vascular occlusion. They confront treatment challenges, but a proper surgical management including regular dressings and proper antibiotics can obviate majority of complications. The patients with pain in this series were treated by analgesics, maintaining hydration and oxygen inhalation with which the patients responded well.Tissue anoxia, thrombosis and necrosis is usually the causative factor. Superficial abscesses underwent incision and drainage. Splenic abscesses were treated with spleenectomy as there is no point in preserving a non-functioning spleen ( functional asplenia) that exists in the majority of patients.[24] Hepatic abscesses are due to secondary enteric infection of hepatic infarcts. A single case of hepatic abscess was detected in this series which was treated by CT guided aspiration and antibiotics. Hepatic abscesses are rare according to various reports.[25,26]

Many of the patients presenting to the surgical department require surgical intervention. Despite the best of facilities and expertise the perioperative complications are quite high and hence prophylactic blood transfusions, which decrease the frequency of most complications in patients with SCA, are also frequently given as a part of perioperative management.[27]The objective of blood transfusion is not only to dilute sickling but to alter the HbS level to about 50%. Studies of transfusion therapy have demonstrated a reversal of organ dysfunction inspite of a relatively high HbS levels. In this series 18 patients required blood transfusion and 82 patients did not. However no elective preoperative transfusion was given.

It was observed that wound healing was delayed in majority of surgical cases probably as a result of tissue hypoxia. There was no mortality in this series. A measure of fetal hemoglobin (Hb) is a fairly good indicator of mortality which was not done in this series. The risk of early death was inversely associated with level of fetal Hb. This association was revealed by studies that indicated an increased survival of patients with sickle cell anemia who had fetal Hb levels above the 75th percentile.[28]

Although medical line of management is equally important it was not discussed here.

Conclusion :

Although the mainstay of management of SCA is medical, the affected patients present with a myriad of surgical problems to the general surgeon, posing a formidable diagnostic challenge. Introduction of minimally invasive techniques and awareness of the various surgical manifestations of SCA alongwith adequate precautions to guard against factors predisposing to vasoocclusive crisis will greatly contribute to a reduction in mortality and morbidity associated with surgery in this high risk group of patients. Maintaining hydration, oxygen saturation in blood, analgesics help in adults. In children complete immunization and penicillin prophylaxis contribute to a complication free life. Finally, genetic counseling can be helpful for parents and families to prevent SCD.



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Table 1:

| Age (In Years) | Male | | Female | |
|---|---|---|---|---|
| | No | % | No | % |
| 0-10 | 3 | 3 | 1 | 1 |
| 11-20 | 9 | 9 | 4 | 4 |
| 21-30 | 13 | 13 | 9 | 9 |
| 31-40 | 7 | 7 | 6 | 6 |
| >40 | 32 | 32 | 16 | 16 |
| Total | 64 | 64 | 36 | 36 |

Table 2 :

| Symptoms | No. Of Patients* | Signs | No. Of Patients* |
|---|---|---|---|
| Abdominal pain | 67 | Pallor | 85 |
| Abdominal distension | 30 | Icterus | 3 |
| Vomiting | 44 | Edema | 12 |
| No F/M | 20 | Ascitis | 10 |
| Hematuria | 7 | Signs of dehydration | 30 |
| Fever | 58 | LN | 9 |
| Chest pain | 7 | Tenderness | 48 |
| Cough | 12 | Guarding | 29 |
| Breathlessness | 3 | Rigidity | 2 |
| Palpitation | 4 | Hepatomegaly | 8 |
| Discoloration of digits | 4 | Splenomegaly | 13 |
| Generalized weakness | 69 | Lung signs | 18 |
| Bony pain | 35 | Lung signs | 18 |
| Lump in abdomen | 17 | | |
| Ulcer over body | 15 | | |

*Categorized by predominant signs and symptoms of presentation as they may overlap.

Table 3:

| Crisis | No. of patients |
|---|---|
| Vasoocclusive | 53 |
| Hemolytic | 4 |
| Sequestration | 5 |
| Aplastic | 1 |
| No crisis but present with different surgical condition | 37 |



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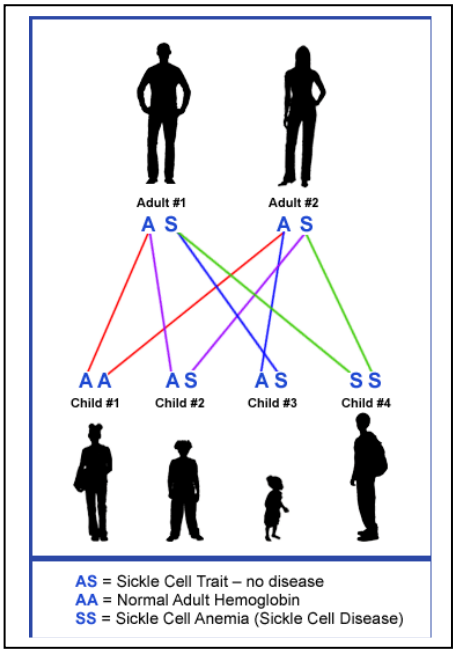
Table 4:

| Haemoglobin electrophoresis | Patient | Percentage (%) |
|---|---|---|
| Haemoglobin AS | 10 | 10 |
| Haemoglobin SS | 90 | 90 |

Table 5 :

| Surgical presentation | Patients |
|---|---|
| 1) Leg ulcers | 20 |
| 2) Abdominal emergencies | 27 |
| 3) Abscesses<br>a) Breast.<br>b) Subcutaneous.<br>c) Splenic.<br>d) Hepatic | 10<br>03<br>04<br>03<br>01 |
| 4) Priapism | 01 |
| 5) Visceral malignancies | 09 |
| 6) Splenomegaly | 13 |
| 7) Others ( abdominal lump, Raynauds phenomenon, Hematuria, Inguinal hernia, Carcinoma of penis) | 20 |
| Total | 100 |



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