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Research

Digestive pathologies associated with sickle cell disease in Lubumbashi: epidemiological and clinical aspects

Digestives diseases associated to sickle cell anemia in Lubumbashi: epidemiological and clinical aspects

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Résumé

Introduction: Sickle cell disease is a genetic disease of autosomal transmission linked to a structural abnormality of hemoglobin which results in the formation of hemoglobin S. The aim of our study is to collect cases of digestive pathologies encountered in sickle cell patients in Lubumbashi and to highlight their epidemiological and clinical characteristics. **Methods:** This is a retrospective, descriptive, cross-sectional study carried out at the Sickle Cell Disease Research Center in Lubumbashi. It involved the files of patients followed for sickle cell disease who presented with a digestive pathology during our 3-year period (from January 2015 to December 2017). Data collection was done using a survey form with different study parameters including: age, sex, reason for consultation, diagnosis, type of vaso-occlusive crisis, paraclinical examinations performed, hydroxyurea treatment. **Results:** We collected 206 files (N=206) of sickle cell patients who had digestive pathology out of a total of 403 files examined, which represents a frequency of 51.11% of digestive pathologies in sickle cell patients. Both sexes are represented with a slight female predominance (51.94%) and a sex ratio M/F: 0.92. The most represented age group is that between 1 and 6 years (32.52%), the average age: 11.8 years; standard deviation: 21.9; extreme ages: 13 months and 38 years. The reason for consultation is dominated by fever (60.67%), abdominal pain (44.66%) and digestive disorders (30.09%). Abdominal vaso-occlusive crises were found in 65 patients (31.55%) among whom 36 patients presented 1 crisis, 24 presented 2 and 5 patients presented 3. Intestinal pathologies were present in 121 patients (69.41%) dominated by intestinal parasitosis (found in 58 patients whose stool examination revealed 4 parasites: Yersinia enterocolitis, entamoeba histolytica, Giardia intestinalis and clostridium difficile); gastric pathologies were found in 105 patients (50.97%) divided into peptic ulcer (45 patients) and gastritis (60 patients); gallbladder pathology present in 40 patients (19.41%) including gallbladder lithiasis without cholecystitis 32 patients, lithiatic cholecystitis 5 patients and 3 cases of lithiasis of the main bile duct; only 1 case of acute pancreatitis diagnosed. The most common associated pathologies found in our study are respiratory with 169 cases (82.03%), oto-rhino-laryngological with 157 cases (76.21%), bone vaso-occlusive crises (146 cases or 70.87%), urogenital pathologies with 64 cases (31.06%) and malaria in 51 patients (24.75%). Splenic and hepatic lesions each constituted 47 cases (22.81%) and 18 cases (8.73%).

Ultrasound was requested in 79 patients but only 31 of them performed it, due to lack of financial means because it costs 20 US dollars locally. In cases of clinically evident splenomegaly, Jolly body was requested in 23 patients but only 2 patients performed it since it costs 10 US dollars. Routine blood counts include hemoglobin, hematocrit, inflammatory assessment and thick blood drop were performed in all our patients but liver function tests, stool and urine tests are recommended depending on the complaint. Of our 206 patients, only 60 of them were under hydroxyurea treatment (29.16%). **Conclusion:** Digestive pathologies are common in sickle cell patients and represent almost half of the sickle cell population. Unfortunately, the best care remains hampered by the obvious poverty of the population, limiting paraclinical examinations which are very useful in the digestive pathology encountered in sickle cell patients.

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Abstract

Introduction: sickle cell disease is a genetic disease with autosomal inheritance associated with haemoglobin structure abnormality which causes formation of of the hemoglobin S. The purpose our study was to collect data digestive diseases in patients with sickle cell disease in Lubumbashi and to highlight their epidemiological and clinical features. **Methods:** We conducted a retrospective, descriptive, cross-sectional study at the Research Center for Sickle Cell Lubumbashi Disease in patients All the records of on follow-up for sicklecell disease during our 3-year period (January 2015 to December 2017) were analyzed. Data were collected using including: age, the for consultation, diagnosis, the of vaso- a survey taking into account different study parameters occlusive crisis, the paraclinical examinations made, hydroxyurea type sex, reason treatment. **Results:** out of total of 403 medical records examined we found 206 cases (n=206) of sickle cell disease associated with digestive a disease, accounting for of of patients with sickle cell disease who suffered rom digestive diseases. Both sexes were represented with rate 51,11% a slight female predominance (51.94%) and sex ratio M/F of 0.92. The most represented 1-6 years (32.52%), the a a age ranges average age was the standard deviation was 21.9; the months and years. The reason for consultation was dominated by fever 11.8 years; extreme ages were 13 38 (60,67%), abdominal pain (44.66%) and digestive disorders (30,09%). Vaso-occlusive abdominal crises were found in 65 patients (31.55%) among whom 36 had only 1 crisis, 24 had 2 crises and had 3 crises. Intestinal diseases were found in 121 patients (69,41%) dominated by intestinal 5 parasites (found in 58 patients whose collection of stool samples showed 4 parasites: Yersinia enterocolitis, Entamoeba histolytica, Giardia intestinalis and Clostridium difficile). Gastric diseases were found 105 (50,97%) divided into peptic ulcer (45 patients) and (60 patients); in patients gastritis biliary vesicular disease was found in 40 patients (19.41%) including vesicular lithiasis without cholecystitis (32 patients), lithiasic cholecystitis (5 patients) and lithiasis the main biliary tract (3 cases); there was in 1 single case diagnosed with acute pancreatitis. The most common associated diseases in our study were respiratory diseases (169 cases;82,03%), oto-rhino-laryngological diseases (157 cases;76.21%), bony, vaso-occlusive crises (146 cases; 70,87%), urogenital diseases (64 cases; 31.06%) and malaria (51 patients; 24.75%). Hepatic diseases and diseases of the spleen were found in 18 cases (8.73%) and 47 cases (22,81%) respectively. Ultrasound was requested in 79 patients but only 31 of them underwent it because of the lack of financial means (it 20 U.S. dollars). costs In case the of clinically obvious splenomegaly, the search for Howell-Jolly bodies was requested in 23 patients but it only performed n 2 patients because it costs 10 U.S. dollars). Routine blood count, hemoglobin, hematocrit, was inflammatory assessment and thick drop examination were performed all but liver assessment, tests done in our patients on stool samples, urine test were recommended based on patient's complaint. Out of 206 patients, only 60 were under hydroxyurea treatment (29,16%). **Conclusion:** digestive diseases are common in patients with sickle cell disease and for almost half of patients with diagnosed account sickle cell disease. Unfortunately, best management is limited by poverty leading to less very useful paraclinical examinations in patients with digestive diseases resulting from sickle cell disease.

Key words: Sickle cell disease,digestive disease, frequency

Introduction

Sickle cell disease is a genetic disease that is transmitted autosomal linked to an abnormality in the structure of hemoglobin which results in the formation of hemoglobin S. It is one of the diseases most common recessive hereditary disorders worldwide and in sub-Saharan Africa [1, 2]. According to the World Health Organization Health, 7% of the population carries the abnormal globin gene and in some parts of the world up to 1% of newborns are affected by a hemoglobin pathology. This pathology is very common in the world but especially in the population of origin sub-Saharan Africa, America, the Antilles, India and the Middle East as well as the Mediterranean basin [3]. The annual incidence is estimated at 305,800 sickle cell patients born worldwide, 80% of whom are in Africa [4]. Newborn screening, systematic clinical monitoring and prevention of sepsis and multi-organ failure has contributed to increasing life expectancy among sickle cell patients in several countries; however, in limited resources where the majority of children with sickle cell disease are born, many continue to die in early childhood, often unaccounted for. diagnosed due to a lack of early detection and treatment programs [5]. Overall, sickle cell disease affects 300,000 children each year, the highest prevalence in areas where malaria is endemic, such as the East, Africa and South Asia. It is also estimated that in several African countries, 10 to 40% of the population is carrier of the sickle cell trait [6]. In 2010, the World Center for Disease Control with the program in charge of monitoring newborns which is in charge of sickle cell disease reported that the incidence of sickle cell disease was 73.1 cases per 1000 children

black, 3.0 cases per 1,000 white children, and 2.7 per 1,000 Asian, Hawaiian, or other island children recorded [7].

In the USA, it is very prevalent, affecting about 100,000 Americans [8]; It is the first disease detected in neonatology with 1 newborn affected in 1900 births, and there are 90,000 to 100,000 patients affected by major sickle cell syndrome in the United States of America [9]. In the Democratic Republic of Congo, recent epidemiological data have shown that 2% of newborns are homozygous for hemoglobin S and approximately 40,000 births of children with sickle cell disease are estimated each year year, while in the adult population the carriage of sickle cell trait amounts to approximately 25% [10, 11] but the disease remains little known with the consequence of high mortality in a country with limited resources [12, 13]. In a publication published in 2016 In the American Journal of Haematology, the authors celebrate the 40th anniversary of the sickle cell disease program, support which, in 40 years, has helped transform this pediatric disease, fatal in the 1970s, into a chronic adult disease nowadays [9].

This is a disease whose progression is punctuated by multiple complications which make it so serious [14]. We note in particular clinical manifestations such as chronic hemolytic anemia, vaso-occlusive phenomena, susceptibility extremes to infections, pain, pulmonary manifestations, cardiovascular [15], oto-rhino-laryngological complications [16]; renal [17], neurological manifestations [18, 19] and digestive manifestations which will be the subject of this present work. Sickle cell disease can affect several systems including the gastrointestinal tract. gastrointestinal and manifestations are usually secondary to small vessel infarction and occlusion of microcirculation. Ischemia manifesting as an abdominal crisis with pain severe which may reflect ulcerative disease, pancreatitis and rarely intestinal ischemia [20]. Patients with Sickle cell disease has a variety of gastrointestinal pathologies including gallstones, hepatitis, biliary sludge, hepatomegaly, painful crises, cirrhosis with a variety of etiologies. These and other entities related to gastrointestinal involvement in cases of Sickle cell disease is described emphatically based on its morphological and biochemical signs [21]. Gallstones are a very frequent chronic complication [22] and long recognized as a complication of sickle cell disease. A study Ultrasound in the UK showed that more than half of homozygous SS patients aged 10-65 years had a gallstone [23], in Jamaica, 13% had cholelithiasis [24]. The majority of sickle cell patients live in underdeveloped countries where endemic parasitic diseases are common and this may exacerbate their chronic anemic state [25], administration of hydroxyurea and transfusion are strongly recommended in several patients with sickle cell anemia [15]; transfusion should be given to correct anemia with a hemoglobin level below 5 g/dl because of hemoglobin S which has a low affinity to oxygen and ensures good oxygenation despite anemia [26].

Methods

This was a descriptive and retrospective study spread over a period of 3 years, from January 2015 to December 2017. It was carried out at the sickle cell research center in Lubumbashi. This center located in Lubumbashi is a public scientific and technical establishment created in November 1982 in Kinshasa; the opening in Lubumbashi was made in 2011 by decree ministerial. It has a dual mission of taking charge of the sickle cell disease and to conduct research to improve specific treatments for sickle cell disease. Data collection was carried out using an anonymous survey form that examined the files of sickle cell patients of all ages and sexes having consulted the center for a digestive pathology diagnosed during our study period from January 2015 to December 2017 a period of 3 years. Thus the study parameters were: age; sex, reason for consultation, objective signs, diagnosis clinical, paraclinical examinations carried out, associated pathologies, treatment with hydroxyurea. Thus were excluded from our study any patient who did not develop a digestive pathology during our study period and anyone who is not part of our study period. Thus 403 patient files were examined, among which 206 had experienced digestive pathology and were selected for the present study; this therefore constituted our sample size. The collected data were entered and analyzed with Word et Excel 2010; Epi-info 3.2.

Results

We collected 206 cases of digestive pathologies from a total of 403 files examined during the 3 years of our study period.
or a frequency of 51.77% of cases. The most affected age group is 1 to 6 years with 32.5% of cases followed by
7-11 years (21.4%) and 12-16 years (20.9% of cases) (Table 1). The mean age is 11.8±21.9 years. The female sex predominates with
51.95% of cases versus 48.05% for men (Table 1); sex
Male/female ratio: 0.92 The reasons for consultation are multiple
(Table 2); fever was found in 125 patients or 60.67%, abdominal pain was found in 92 patients (44.66%),
digestive disorders found in 62 patients (30.06%), the
abdominal bloating in 10 patients (4.85%). Only 116
patients out of 206 (56.31%) had digestive signs while 90 patients (43.68%) had no digestive complaints despite the
diagnostic confirmation of digestive pathology by routine paraclinical examinations in sickle cell patients. Depending on the
diagnoses, abdominal vaso-occlusive crises represented 65 cases (31.55%); intestinal digestive pathology represented 121 cases (58.73%) divided into intestinal parasitosis (58 cases or 28.15%), digestive disorders (55 cases or 26.69%), functional colopathy (5 cases or 2.42%), inguinal hernia (22 cases or 10.67%) and appendicitis (1 case or 0.48%); gastric pathology 105 cases (50.97%) distributed in gastritis (60 cases or 29.12%) and gastroduodenal ulcer (45 cases or 21.84%); gallbladder pathology found in 40 patients (19.41%) divided into gallbladder lithiasis (32 cases or 15.53%), cholecystitis (5 cases or 2.42%) and cholecystolithiasis (3 cases or 1.45%); splenic lesions were found in 47 patients (22.81%) divided into 27 cases of splenomegaly (13.10%), 14 cases of asplenia functional (6.79%), 4 cases of splenic sequestration (1.94%) and 2 cases of hypersplenism); and liver damage 18 cases (8.73%) divided into 16 cases of hepatomegaly (7.76%), hepatic sequestration and viral hepatitis B each 1 case (0.48%) each.

Acute pancreatitis was discovered in only 1 patient (0.48%).
(Table 3). The associated pathologies found are:
respiratory pathologies (82.03%), vaso-occlusive crises
bone (146 cases or 70.87%), oto-rhino-laryngological (76.21%), malaria (51 cases or 24.75%), urogenital pathologies (64
cases or 31.06%), aseptic necrosis (21 cases or 10.19%), acute hemolytic crises (13 cases or 6.31%), pathologies
kidney disease (6 cases or 2.91%) and heart disease (3 cases or 1.45%) (Table 4). Biological tests systematically requested are made up of an inflammatory assessment (white blood cells, sedimentation rate, C-reactive protein, leukocyte formula), the rate hemoglobin, a thick drop. These were requested in all our 206 patients. Other biological tests are requested depending on the clinic, these are fresh stool tests
performed in 79 patients (38.34%), stool culture in 15 cases (7.28%), urinary infectious assessment using cytobacteriological examination in 6 patients (2.91%), transaminases requested in 43 patients
(20.87%), the body of joly requested in 23 patients (11.16%), serology to hepatitis A in 3 patients (1.45%), hepatitis B in 2
patients (2.90%), lumbar puncture in 80 patients (38.83%).
Ultrasound was requested in 79 patients (38.34%) for the diagnosis of pathologies such as gallstones, cholecystitis, splenomegaly, hepatomegaly but only 31 patients performed it. Of our 206 patients, only 60 of them were under treatment with hydroxyurea, i.e. 29.12% of cases.

Discussion

Our study on the epidemiological and clinical aspects of digestive pathologies in sickle cell patients collected 206 cases out of
403 patient files examined. The frequency of digestive pathology among sickle cell patients in Lushois is 51.11%. Without
give numerical values for all digestive pathologies, the authors are unanimous on the high frequency of these in the
sickle cell disease.

In relation to the age group: the most affected age group in our series is that between 1-6 years with 32.5% of cases
with an average age of 11.8 years for extreme ages of 13
months and 38 years old. In Ouagadougou, Solange Odilet Diarra Ye finds an average age lower than ours of 7 years and 7.9±3.87 years respectively, this being explained by the fact that their study only concerned the children up to 15 years old [27, 28] while ours included patients of all ages; The most affected age group is 6-
10 years with 41% [28].

By sex: Our study reveals a slight female predominance with a sex ratio of 1.08 in favor of women. But in relation to sex, studies are contradictory. Some show a male predominance [28, 29]; others a

female predominance [29-32]; still others an equal distribution

between the two sexes [33, 34]. We believe that the predominance of one or the other kind is a matter of chance.

Related to the reason for consultation: reasons for consultation in sickle cell patients with pathology digestive are multiple. The general signs are in the foreground dominated by fever in our series with 60.67% of cases while Sonia [32] reports a fever in 95.5% of cases, a very high frequency superior to ours; this is due to the fact that his study focused on infections in sickle cell patients. Only 116 patients out of 206 (56.31%) had digestive clinical signs dominated by abdominal pain (44.66%) then abdominal bloating (4.85%) while 90 patients (43.69%) had no complaints during of the consultation. Sonia [32] reports 23.3% of abdominal pain, low compared to ours probably for the reason that her study was not focused on the digestive tract like ours.

In relation to the etiological diagnosis: depending on the diagnoses, abdominal vaso-occlusive crises represented 65 cases (31.55%) in our series, on the other hand Roger Dodo in Benin reported 12.7% for abdominal vaso-occlusives [35]; this difference could be explained by the fact that his study was focused solely on sickle cell emergencies while ours focused on urgent or non-urgent digestive pathologies.
In patients with vaso-occlusive crisis, acute sickle cell hepatic crisis is reported in 10% of patients [36];
Clinically it can present as an attack of cholecystitis with fever, hypochondralgia, jaundice [37].

Intestinal digestive pathology represented 121 cases (58.73%). in our study. We found intestinal parasitosis (58 cases or 28.15%) dominated by Yersinia enterocolitis (28 cases) and Entamoeba histolitica (19 cases) and Giardia intestinalis (11 cases), functional colopathy (60 cases or 29.12%), febrile enteritis (2 cases or 0.97%), inguinal hernia (22 cases or 10.67%) and appendicitis (1 case or 0.48%); in sub-Saharan Africa, the authors report frequencies of 38.8% for Giardiasis as well as 27.8% and 25.9% for amoebiasis [25-33]. Digestive tract parasites are found in all age groups in Sonia [32] and are dominated by Gardia intestinalis (8 cases out of 17) and Entamoeba histolitica (4 cases out of 17) also protozoa as in our series; this confirms the data reported by authors that the majority sickle cell patients live in underdeveloped countries where endemic parasitic diseases are common, which can exacerbate their condition of chronic anemia [25]. This could prove to be real since we have found malaria as one of the most frequent associated pathologies with 24.75% of patients. This rate is slightly low compared to that reported by Roger Dodo [35] of 27.5% of cases but higher than that found by Sonia [32] by 16.5% and Mbika [38] by 18%. The interactions between sickle cell disease and malaria remain observed and the belief that sickle cell patients are protected remains widespread [39]; this which proves the controversy of this belief. Heterozygous patients whose red blood cells contain hemoglobin A and S would be strongly protected from malaria [40-41] but the global distribution and frequency of the beta mutation S currently reflects the historical incidence of deaths due to malaria [6]. In relation to biological examinations, they are requested according to the clinic, these are the fresh stool examinations carried out in 79 patients (38.34%) while in the study of Sonia [32] examination Parasitological examination of stools was carried out in 16.5% of patients, stool culture 15 cases (7.28%) against 13.5% in Sonia [32], the results urinary tract infection made by cytobacteriological examination in 6 patients (2.91%) against 19.3% in Sonia [32], transaminases requested in 43 patients (20.87%), the body of Joly requested in 23 patients (11.16%), hepatitis A serology in 3 patients (1.45%), hepatitis B in 2 patients (2.90%), lumbar puncture in 80 patients (38.83%). Gastric pathology 105 cases (50.97%) distributed in gastritis (60 cases or 29.12%) and gastroduodenal ulcer (45 cases or 21.84%); this rate seems frequent compared to the non-infected population sickle cell disease. There is no clear finding suggesting that the incidence of peptic ulcer in sickle cell disease is higher than in the general population despite the depressive nature of this disease [42], however dyspeptic symptoms are frequent in sickle cell patients [42]. Lee MG finds on examination digestive endoscopy an anomaly in 20 out of 51 patients who presented abdominal pain divided into 14 cases of duodenal ulcer, 4 cases of gastric ulcer and 2 cases of gastric ulcer [43]. The etiology of these ulcers is associated with reduced mucosal resistance secondary to repeated ischemia during sickle cell disease [44]. The gallbladder pathology found in 40 patients (19.41%) divided into gallbladder lithiasis (32 cases or 15.53%), cholecystitis (5 cases or 2.42%) and cholecystolithiasis (3 cases or 1.45%); the hepatobiliary system is one of the organs abdominal muscles most affected by sickle cell disease and liver damage is observed in 10 to 40% of cases in series sickle cell disease [4]. The frequency of gallstone disease in sickle cell disease increases with age and severity of the disease.

In Jamaica, the prevalence is estimated at 12% in the 5-7 year old group and 23% in the 11-13 year old group [24]. This corroborates with our observations and this is due to chronic hemolysis releasing bile pigments which alters the balance of the Admirant-Small triangle thus causing stones.

biliary. Liver damage 18 cases (8.73%) divided into hepatomegaly (16 cases or 7.76%), hepatic sequestration (1 case or 0.48%) and viral hepatitis B (1 case or 0.48%) while
Johnson [36] found 32% chronic liver abnormalities in biology including hepatitis, chronic passive liver congestion, liver disease
alcoholic liver disease, collagen vascular disease, and sarcoidosis; he found 19% hepatitis while Koskina [45] found 39% involvement
hepatic, 9 cases of painful hepatomegaly (22%), 8 cases of cholestasis. Acute hepatic crisis in sickle cell disease is reported in 10% of patients with vaso-occlusive crisis [46]; clinically it may present as a cholecystitis crisis

with fever, hypochondralgia, jaundice [37]. Our low results on liver damage are probably underestimated because of
financial limitations of paraclinical ultrasound and biology. Splenic lesions found in 47 patients
(22.81%) divided into splenomegaly (27 cases or 13.10%), functional asplenia (14 cases or 6.79%), splenic sequestration (4 cases or
1.94%) and hypersplenism (2 cases or 0.97%). Sonia [32] reports a frequency of splenomegaly and hepatomegaly higher than ours
or 42.1% and 30.1% respectively. Our low rate could
can be explained by the fact that out of 79 patients only 31 patients have performed the ultrasound which was therefore not systematic, some asymptomatic patients who did not have the ultrasound could have a subclinical splenomegaly. The spleen is almost always affected in sickle cell disease according to Ellen C. with micro-infarcts in
the first 36 months of life leading to splenic atrophy, chronic liver diseases can be due to hemosiderosis and
hepatitis [46]. We believe that the hepatomegaly found in our patients was caused by hemosiderosis unfortunately
the non-subsidy of the research center limits the examinations
complementary to the responsibility of the family itself. We do not have found only 1 case out of 206 cases while Johnson found that acute hepatitis affects 10% of patients admitted for abdominal pain crisis [36]. Only 60 patients out of 206 were under treatment with hydroxyurea, i.e. 29.12%. Hydroxyurea would reduce
complications of sickle cell disease, such as painful crises [47]. Treatment with hydroxyurea has been observed
with statistically significant correlation to the reduction of hemoglobin S level, with an elevation of fetal hemoglobin [48].

Conclusion

Sickle cell patients are prone to digestive pathologies; our study revealed the high frequency of digestive pathologies in sickle cell patients of 51.1%. The clinical presentation is dominated by fever, abdominal pain and digestive disorders but sometimes the patient is asymptomatic, the discovery of the pathology
digestive system being done through routine or monitoring examinations. Unfortunately, the high cost of certain paraclinical examinations
limits the best care in a population with a
manifest poverty.

Current state of knowledge on the subject

- Sickle cell disease is the most common genetic disorder in the world;
- It is mainly located in sub-Saharan Africa;
- Digestive pathologies are common in sickle cell patients.

Contribution of our study to knowledge

- The objective frequency of digestive pathologies in the sickle cell patients from Lubumbashi;
- The epidemiological and clinical characteristics of digestive pathologies in sickle cell patients;
- The limits of the best support for these digestive pathologies.

Conflicts of interest

The authors declare no conflict of interest.

Authors' contributions

Manix Ilunga Banza wrote the article. Jules Panda Mulefu is
the initiator of the article and the designer of the research protocol. Read Ipani Lire contributed to the writing of the article. Yannick Tietie Ben N'dwala and Israël Badypwila Tshiamala contributed to the collection of data.
data. Vincent de Paul Kaoma Cabala participated in reading the article. All read and approved the final version of the manuscript.

Paintings

References

1. Burnham-Marusich AR, Ezeanolue CO, Obiefune MC, Yang W, Osuji A, Ogidi AG et al. . Prevalence of Sickle Cell Trait and Reliability of Self-Reported Status among Expectant Parents in Nigeria: Implications for Targeted Newborn Screening. Public Health Genomics. 2016; 19(5): 298-306. **PubMed** | **Google Scholar**

2. Piel FB, Howes RE, Patil AP, Nyangiri OA, Gething PW, Bhatt S tool . The distribution of haemoglobin C and its prevalence in newborns in Africa. Sci Rep. 2013; 3: 1671. **PubMed** | **Google Scholar**

3. Mario N, Sala N. Biological diagnosis of hemoglobinopathies. French-speaking Review of Laboratories. 2016; 481: 36-39. **Google Scholar**

4. Piel FB, Patil AP, Howes RE, Nyangiri OA, Gething PW, Dewi M. et al. . Global epidemiology of sickle haemoglobin in neonates: a contemporary geostatistical model-based map and population estimates. Lancet. 2013 Jan 12;381(9861):142-51. **PubMed** | **Google Scholar**

5. Chakravorty S, Williams TN. Sickle cell disease: a neglected chronic disease of increasing global health importance. Arch Dis Child. Janv 2015; 100(1): 48-53. **PubMed** | **Google Scholar**

6. Piel FB, Patil AP, Howes RE, Nyangiri OA, Gething PW, Williams TN et al. . Global distribution of the sickle cell gene and geographical confirmation of the malaria hypothesis. Nat Commun. 2 Nov 2010; 1: 104. **PubMed** | **Google Scholar**

[PubMed] 7. Ojodu J, Hulihan MM, Pope SN, Grant AM, Centers for Disease Control Control and Prevention (CDC). Incidence of sickle cell trait United States, 2010. MMWR Morb Mortal Wkly Rep. 12 Déc 2014; 63(49): 1155-8. **PubMed** | **Google Scholar**

8. Hassell KL. Population estimates of sickle cell disease in the U.S. Am J Prev Med. Avr 2010; 38(4 Suppl): S512-521. **PubMed** | **Google Scholar**

9. Chaturvedi S, DeBaun MR. Evolution of sickle cell disease from a life-threatening disease of children to a chronic disease of adults: The last 40 years. Am J Hematol. Janv 2016; 91(1): 5-14. **PubMed** | **Google Scholar**

10. Tshilolo L, Aissi LM, Lukusa D, Kinsiama C, Wembonyama S, Swan B et al. . Neonatal screening for sickle cell anaemia in the Democratic Republic of the Congo: experience from a pioneer project on 31 204 newborns. J Clin Pathol. Janv 2009; 62(1): 35-8. **PubMed** | **Google Scholar**

11. Agasa B, Help K, Opara A, Tshilumba K, Dupont E, Vertonghen F And al . Prevalence of sickle cell disease in a northeastern region of the Democratic Republic of Congo: what impact on transfusion policy? Transfus Med. Févr 2010; 20(1): 62-5. **Google Scholar**

12. De Montalembert M, Tshilolo L. [Is therapeutic progress in the management of sickle cell disease applicable in sub-Saharan Africa?]. Med Trop (Mar). Dec 2007; 67(6): 612-6. **PubMed** | **Google Scholar**

13. Driscoll MC. Sickle cell disease. Pediatr Rev. Juill 2007; 28(7): 259-68. **PubMed** | **Google Scholar**

14. M'pemba L, N'Zingoula S. The vaccination status of homozygous Congolese children with sickle cell disease. Med Afr Noire. 2004; 51: 37-41.

15. Yawn BP, Buchanan GR, Afenyi-Annan AN, Ballas SK, Hassell KL, James AH et al. . Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. JAMA. 10 Sept 2014; 312(10): 1033-48. **PubMed** | **Google Scholar**

16. Tshilolo L, Bahwe M, Vertongen F. ENT and oral anomalies observed in sickle cell patients
Zairois. Nouv Rev Fr hematol. 1995; 37(1): 4.

17. Bartolucci P, Habibi A, Stehlé T, Di Liberto G, Rakotoson MG, Gellen-Dautremer J. et al. . Six months of hydroxyurea reduces albuminuria in patients with sickle cell disease. J Am Soc Nephrol. 2016; 27(6): 1847-53. **PubMed** | **Google Scholar**

18. Neonato MG, Guilloud-Bataille M, Beauvais P, Bégué P, Belloy M, Benkerrou M tool . Acute clinical events in 299 homozygous sickle cell patients living in France. French Study Group on Sickle Cell Disease. Eur J Haematol. Sept 2000; 65(3): 155-64. **PubMed** | **Google Scholar**

19. Tshilolo L, Mukendi R, Girot R. Drepanocytosis in the south of the Zaire: study of two series of 251 and 340 patients followed between 1988 and 1992. Archives of Pediatrics. 1994; 3(2): 104-11. **PubMed** | **Google Scholar**

20. Gage TP, Gagnier JM. Ischemic colitis complicating sickle cell crisis. Gastroenterology. Janv 1983; 84(1): 171-4. **PubMed** | **Google Scholar**

21. Krauss JS, Freant LJ, Lee JR. Gastrointestinal pathology in sickle cell disease. Ann Clin Lab Sci. Févr 1998; 28(1): 19-23. **PubMed** | **Google Scholar**

22. Embury SH. Sickle cell disease: basic principles and clinical practice. New York: Raven Press. 1994; 902. **Google Scholar**

23. Bond LR, Hatty SR, Horn ME, Dick M, Meire HB, Bellingham AJ. Gall stones in sickle cell disease in the United Kingdom. Br Med J (Clin Res Ed). 1987 Jul 25;295(6592):234-6. **PubMed** | **Google Scholar**

24. Webb DK, Darby JS, Dunn DT, Terry SI, Serjeant GR. Gall stones in Jamaican children with homozygous sickle cell disease. Arch Dis Child. Mai 1989; 64(5): 693-6. **PubMed** | **Google Scholar**

25. Ahmed SG, Uraka J. Impact of intestinal parasites on haematological parameters of sickle-cell anaemia patients in
Nigeria. East Mediterr Health J. Sept 2011; 17(9): 710-3. **PubMed** | **Google Scholar**

26. Porter JB, Huehns ER. Transfusion and exchange transfusion in sickle cell anaemias, with particular reference to iron metabolism. Acta Haematol. 1987; 78(2-3): 198-205. **PubMed** | **Google Scholar**

27. Ouédraogo-Yugbaré SO, Tiendrebeogo J, Koueta F, Sawadogo H, Dao L, Ouédraogo P. Major sickle cell syndromes in children aged 0 to 15 years in Ouagadougou: tool
genetic markers and clinical characteristics. Pan African Medical Journal. 2014;19:215. **Google Scholar**

28. Yé D, Kouéta F, Dao L, Kaboret S, Sawadogo A. Management of sickle cell disease in a pediatric setting: experience of the Charles-de-Gaulle pediatric university hospital in Ouagadougou (Burkina Faso). Cahiers de Santé. Apr 2008; 2:
071-075. **Google Scholar**

29. Nacoulma E, Kam L, Gue EE, Kafando E, Ayereroue J, Blot I. Evaluation of the vaccination status of children with sickle cell disease in the city of Ouagadougou (Burkina Faso). Study and development notebooks French-speaking research / Health. 2006; 16(3): 155-60. **Google Scholar**

30. Wierenga KJ, Hambleton IR, Wilson RM, Alexander H, Serjeant BE, Serjeant GR. Significance of fever in Jamaican patients with homozygous sickle cell disease. Arch Dis Child. Févr 2001; 84(2): 156-9. **PubMed** | **Google Scholar**

31. Latoundji S, Anani L, Ablet E, Zohoun I. Morbidity and mortality sickle cell disease in Benin. Med Afr Noire. 1991;38(8/9): 571-6.

32. Douamba S, Nagalo K, Tamini L, Traoré I, Kam M, Couéta F. et al. [Major sickle cell syndromes and infections associated with this condition in children in Burkina Faso]. Pan Afr Med J. 2017; 26:7. **PubMed** | **Google Scholar**

33. Gbadoé AD, Atsou K, Agbodjan-Djossou OA, Tsolényanu E, Nyadanu M, Dogba AD et al. . [Ambulatory management of sickle cell disease: evaluation of the first year follow up of patients in the pediatric department of Lomé (Togo)]. Bull Soc Pathol Exot. Mai 2001; 94(2): 101-5. **PubMed** | **Google Scholar**

34. Mukinayi BM, Kalenda DK, Mbelu S, Gulbis B. [Awareness and attitudes of 50 congolese families affected by sickle cell disease: a local survey]. Pan Afr Med J. 2018; 29: 24. **PubMed** | **Google Scholar**

35. Dodo R, Zohoun A, Baglo T, Mehou J, Anani L. [Emergency treatment of sickle cell diseases in the Blood Diseases Department at the Koutoukou Maga National Teaching Hospital, Cotonou, Benin]. Pan Afr Med J. 2018; 30 : 192. **PubMed** | **Google Scholar**

36. Johnson CS, Omata M, Tong MJ, Simmons JF, Weiner J, Tatter D. Liver involvement in sickle cell disease. Medicine (Baltimore). Sept 1985; 64(5): 349-56. **PubMed** | **Google Scholar**

37. Rushikesh S, Taborda C, Chawla S. Acute and chronic hepatobiliary manifestations of sickle cell disease: A review. MWorld J Gastrointest Pathophysiol. 2017 Aug 15; 8(3): 108-116. **PubMed** | **Google Scholar**

38. Mbika Cardorelle A, Okoko A, Mouko A. [Vaso-occlusive crisis of sickle cell child in Brazzaville drugs news]. Arch Pediatr. Mars 2010; 17(3): 295-6. **Google Scholar**

39. Perignon A, Botterel F, Farrugia C, Foulet F, Bachir D, Galacteros . F et al.C-02 Malaria and homozygous sickle cell disease. Medicine and Infectious Diseases. June 2008;38:S140. **Google Scholar**

40. Allison AC. Protection afforded by sickle-cell trait against subtertian malareal infection. Br Med J. 6 Févr 1954; 1(4857): 290-4. **PubMed** | **Google Scholar**

41. Taylor SM, Cerami C, Fairhurst RM. Hemoglobinopathies: slicing the Gordian knot of Plasmodium falciparum malaria pathogenesis. PLoS Pathog. 2013; 9(5): e1003327. **PubMed** | **Google Scholar**

42. Meshikhes AW. Gastroenterological manifestations of sickle cell disease. Saudi J Gastroenterol. Janv 1997; 3(1): 29-33. **PubMed** | **Google Scholar**

43. Lee MG, Thirumalai CH, Terry SI, Serjeant GR. Endoscopic and gastric acid studies in homozygous sickle cell disease and upper abdominal pain. Gut. Mai 1989; 30(5): 569-72. **PubMed** | **Google Scholar**

44. Rao S, Royal JE, Conrad HA, Harris V, Ahuja J. Duodenal ulcer in sickle cell anemia. J Pediatr Gastroenterol Nutr. Janv 1990; 10(1): 117-20. **PubMed** | **Google Scholar**

45. Koskinas J, Manesis EK, Zacharakis GH, Galiatsatos N, Sevastos N, Archimandritis AJ. Liver involvement in acute vaso-occlusive crisis of sickle cell disease: prevalence and predisposing factors. Scand J Gastroenterol. Avr 2007; 42(4): 499-507. **PubMed** | **Google Scholar**

46. Ebert EC, Nagar M, Hagspiel KD. Gastrointestinal and hepatic complications of sickle cell disease. Clin Gastroenterol Hepatol. June 2010;8(6):483-9; quiz e70. **PubMed** | **Google Scholar**

47. Meremikwu MM, Okomo U. Sickle cell disease. BMJ Clin Evid. BMJ Clin Evid. 2016 Jan 22; 2016. pii: **2402.PubMed** | **Google Scholar**

48. Shome DK, Al Ajmi A, Radhi AA, Mansoor EJ, Majed KS. The effect of hydroxyurea therapy in Bahraini sickle cell disease patients. Indian Journal of Hematology and Blood Transfusion. Mars 2016; 32(1): 104-9. **PubMed** | **Google Scholar**

Table 1: Distribution of cases by age and sex

Variables	Effective	Percentage (%)
Age group (years)		
1-6	67	32,5
7-11	44	21,4
12-16	43	20,9
17-21	21	10,2
22-26	20	9,7
27-31	7	3,4
32-36	3	1,5
37-41	1	0,5
Sex		
Male 99		48,1
Female 107		51,9

The most affected age group is 1-6 years with 32.5% of cases; mean age: 11.8 ± 21.9 years; extreme ages: 13 months and 40 years
The female gender slightly predominates over the male with 51.9% of cases; the male/female sex ratio: 0.92

Table 2: Distribution of cases according to reasons for consultation

Reason for consultation	Workforce	Percentage (%)
No complaints 43.68	90	
Abdominal pain 44.66	92	
Fever 60.67	125	
Digestive disorders 30.09	62	
Abdominal bloating 4.85	10	
Hematemesis **4.36**	**9**	

In 90 patients, no complaints were mentioned during the consultation
while fever was the most common reason for consultation with 60.67% and abdominal pain was present in 44.67% of patients.

Table 3: Distribution of cases according to diagnoses

Etiologies	**Staff** 65	**Percentage (%)**
Vaso-occlusive abdominal crisis		31,55
Mild vaso-occlusive crisis	52	25,24
Moderate vaso-occlusive crisis	11	5,33
Severe vaso-occlusive crisis	2	0,97
Intestinal parasitosis	58	28,15
Functional irritable bowel syndrome	10	4,85
Appendicitis	1	0,48
Gastritis	60	29,12
Peptic ulcer	45	21,84
Gallstone disease	32	15,53
Cholecystitis		2,42
Cholecystolithiasis	5	1,45
Splenomegaly	3 27	13,10
Functional asplenia	14	6,79
Splenic sequestration	4	1,94
Hypersplenism	2	0,97
Hepatomegaly	16	7,76
Hepatitis	1	0,48
Liver sequestration	1	0,48
Inguinal hernia	22	10,67

Abdominal vaso-occlusive crises represented 31.55% (25.24% of mild crises, 5.33% of moderate crises and 0.97% of severe crises), intestinal parasitoses (28.15%)

Table 4: Distribution of cases according to associated pathologies

Pathologies (Systems)	**Staff**	**Percentage %**
Respiratory 82.03		
Otorhinolaryngology 76.21		
Urogenital 31.06	169,157,64	
Locomotor system 70.87	146	
Cardiovascular 1.45		
Malaria 24.75	3 51	