Journal of Blood Disorders

Open Access

Austin Publishing Group

Special Article - Anaemia

# Feature of Older Patients with Sickle Cell Disease in the Congo

**Ngolet LO*, Atipo Tsiba Galiba FO, Malanda F, Ockouango Guelongo Ova JD and Elira Dokekias A**

Clinical Hematology Unit, Marien Ngouabi University, Republic of Congo

***Corresponding author:** Ngolet LO, Clinical Hematology Unit, Marien Ngouabi University, Brazzaville, Republic of Congo. BP: 32, Republic of Congo

**Received:** August 01, 2017; **Accepted:** September 26, 2017; **Published:** October 03, 2017

**Abstract**

**Background:** Despite the high mortality rate of patients with sickle cell disease during the childhood in Africa, their life expectancy is increasing. Consequently, the number of adults is growing. Hence, health providers need to be aware about their clinical and biological pattern.

**Patients and Methods:** We studied clinical and biological aspects of 47 patients with sickle cell disease and phenotype SS aged 40 years and over from 2011 to 2016.

**Results:** The mean age was 45 (range: 40- 59) years. The female population was older with a mean age of 47 (p=0.05). Among them 53.3% reported a miscarriage episode sickle cell related. Twenty three over 47 (48.9%) had a low monthly income. Older adults presented sever sickle cell related acute morbidity, with a number of 1.83 vaso occlusive crisis, 2 anemic crisis/ blood transfusions and 1.59 admission episodes per year blood transfusion procedure episodes. In our group 57.44% developed a chronic complication. The most frequent complications of older patients with sickle cell disease were a vascular necrosis of the femoral head (19.14%) and leg ulcer (8.51%). Elevated regurgitate jet velocity was noticed on 10 over 12 (83.33%) heart failure symptomatic patients. The mean hemoglobin rate was 7.3g/dL.

**Conclusion:** Sickle cell related acute morbidity is high in older patients. Chronic complications are more frequent in this group and elevated regurgitate jet velocity more common finding.

**Keywords:** Sickle cell; Congo; Older adults; Morbidity

## Introduction

Sickle cell disease is the most common inherited blood disorder in the Sub-Saharan where 75% of the 300,000 births worldwide live [1]. Few aspects of the sickle cell in the adulthood are known in the region because the mortality in the childhood is high. However despite our low resources, a slight decline in the mortality rate of SCD patients in Africa has been noticed due to improved supportive care such as malaria prophylaxis, SCD awareness, earlier diagnosis and better management care [2]. Consequently, childhood survival has improved and adult population with sickle cell disease is growing with the spectrum of clinical aspects that are for some unknown [3]. Reports of the adults' natural history are spares and clinical aspects are unknown in the continent [4,5]. One study of homozygous HbSS SCD patients over 30 years of age was performed In the Congo. Thus we undertook to study SCD patients over 40 years of age.

## Patients and Methods

### Data collection and assessment procedure

We recruited through outpatient clinic in the hematology department HbSS SCD adult patients 40 years over of age. This study was carried from 2011 to 2016. Data were obtained from adult individuals who were receiving their care from the hematology department or new patients that consulted for sickness. Patients with sickle cell β0 and SC genotypes were excluded in the study.

All patients provided informed consent in accordance with the Declaration of Helsinki. Research protocol was approved by the local institution ethic board (CERSSA). Each patient was interviewed and data were collected in the SCD medical history form. We obtained the following data:

**Sociodemographic:**

1. Age
2. Gender
3. Employment status
4. Income

The income of the family was interpreted based on the official lowest salary fixed by the Congolese government (XAF 90,000/ USD 153.53). The income was low when it was lower than XAF 90,000/ USD 153,53, middle when it was between than XAF 90,000/ USD 153,53 and XAF 500,000/ USD 852.96 and high when it was above XAF 500,000/ USD 852.96.

1. Number of children per family
2. Number of miscarriage SCD related for female patients

**Morbidity:** The number of crisis, hospitalization admissions and blood transfusion procedures that occurred during the study period.




**Citation:** Ngolet LO, Atipo Tsiba Galiba FO, Malanda F, Ockouango Guelongo Ova JD and Elira Dokekias A. Feature of Older Patients with Sickle Cell Disease in the Congo. J Blood Disord. 2017; 4(2): 1048.

**Table 1:** Clinical criteria.

| N= 47 | n(%) |
|---|---|
| **Number of admission episode per year** | |
| **Mean** | 1.56 |
| **Min-Max** | 0-9 |
| **Number of painful crisis per year** | |
| **Mean** | 1.83 |
| **Min-Max** | 0-12 |
| **Number of anemic crisis per year** | |
| **Mean** | 2 |
| **Min-Max** | 0-10 |
| **Number of blood transfusion episode per year** | |
| **Mean** | 2 |
| **Min-Max** | 0-12 |
| **Chronic complications frequency* n(%)** | 27/47 (57.44%) |
| **Leg Ulcer** | 4/47 (8.51%) |
| **Gall Bladder** | 2/47 (4.25) |
| **Avascular necrosis of the femoral head** | 9/47 (19.14%) |
| **ETRJV** | 10/12 (83.33%) |

*17/47 participants develop more one type of complication. One patient can develop more than one type of complication; ETRJV: Elevated Regurgitant Jet Velocity.

**Table 2:** Biological criteria.

| Parameters | Mean values (range) |
|---|---|
| **Hb (g/dL)** | 7.3 (4.7-9.1) |
| **MCV (fl)** | 87.1(68-111) |
| **MCH (pg)** | 29.1(23.6-35.8) |
| **WBC (G/L)** | 12.4(5.4-31.1) |
| **Platelets (G/L)** | 342 (159.7-550.2) |
| **LDH (UI/L)** | 663.1 (221-2083) |

We also recorded chronic complications SCD related noticed at the time of the inclusion in the study as leg ulcer, a vascular femoral necrosis and Gall Bladder. Chronic complications were either obtained from patients' medical records or diagnosed at the time of the visit in the hematology department for symptomatic patients.

Heart ultrasound was performed on symptomatic patients that were presenting a congestive heart failure or dyspnea. Tricuspid regurgitate jet velocity was considered elevated (ETRJV) when the velocity was above 2.5m/sec as per current guidelines of the American Society of Echography [6].

5 milliliters of blood for complete blood count and 5 milliliters for analysis of LDH were sampled for each patient. Samples were obtained on patients in stationary period (no painful or anemic crisis) for at least 3 months.

Painful crisis was defined as an unpredictable intense pain that generally lasts hours to a few days.

We defined anemic crisis was defined by a decrease of at least 2g/dL from the steady state hemoglobin The most common process leading to this complication in adult patients is the Hyperhemolysis crisis. Transient red cell aplasia and acute splenic sequestration are other causes of anemic crisis and are more frequent in children.

Blood transfusion was indicated when the hemoglobin rate was below 6g/dL of the patient or if there is sufficient physiological derangement such as: heart failure, dyspnea, hypotension or marked fatigue related to hemoglobin concentration fall.

A total of sixty two patients over 40 years of age consulted during the study period. Among them 47 (75.81%) agreed to participate to the study

### Statistical analysis

For the description of each quantitative variable, the mean, range, frequencies and percentages were calculated. Student's test was used for the comparison of variable. A significance level of $p \leq 0.05$ was considered.

## Results

### Sociodemographic

47 patients over 40 years of age and were identified, of which 17 (36.2%) were males while 30 (63.8%) were females. The median age of the population was 45 years (extremes: 40 and 59 years, $X^2$, p=0.04). Female patients had higher median age 47 years (extreme: 40 and 59 years, $X^2$= 17.05, p=0.05) compared to male ones: 44 years (extreme 40 and 57 years, $X^2$ = 14.65, p=0.10). Twenty five were unemployed (53.2%) in which 19 females (76%) and 6 males (24%). The total monthly income was low for 23 patients (48.9%), middle for 15 (31.9%) and high for 9 participants (19.1%), p=0.04. The low income was more frequent in the female population (58.3%) than the male one (p= 0.02). The average number of children per family was 0.5 for SCD women versus 1.6 for <male (p=0.01). Sixteen women over 30 (53.3%) reported one episode of miscarriage related to sickle cell.

### Clinical history

The clinical characteristics of SCD patients over age 40 are described in the Table 1.

### Biological characteristics

Laboratory determinants of older adult patients with SCD are reported in the Table 2.

## Discussion

The present paper describes the clinical and biological features of SCD patients over 40 years of age. We acknowledge on a series of limitation in our study. First, SCD renal, eye abnormalities, brain imaging, pulmonary evaluation and infectious complications have not been documented due to lake of diagnosis facilities and lake of funding. Second, we did not study the co-morbidities. They may in this range of age, additionally to SCD, influence the length of life expectancy. Third, the cross sectional did not give us tools to elucidate why this population survives longer.

The median age of the population studied was 45 years. The female population was older with a mean age of 47 years. This epidemiological distribution is a picture of the demographics in the Congo and is not relative to any inherent biological characteristics. Manpreed et al. in a similar study in the USA found, an older population with a median age of 49 years [3]. The longer life expectancy, in Western countries, is partially the result of a systematic universal newborn screening (NBS), pneumococcal infection prevention, early detection of complications as stroke and more recently, use of Hydroxyurea in adult [7-9]. Despite the lake of such interventions in Africa, a slight decrease of the childhood mortality has been noticed due to a better management of the disease, annual SCD awareness and malaria prophylaxis. Consequently, life expectancy of patients with SCD is increasing.

19.1 and 31.9%, have respectively high and middle income which is higher that the demographic distribution of the population in the Congo [10]. Tolo-Diebkile in Ivory Coast noted the same distribution [5]. In low resources countries where there is not any medical coverage system, the level of income is proportionally linked to quality and accessibility of care, thus outcome and length or survival [4,5].

The longer survival is also determined by the Hb rate level that influences the severity of the disease [11-13]. Low hemoglobin rate is associated with high morbidity and mortality of SCD [11-13]. In our series, the mean of hemoglobin (7.3g/dl) was higher than the average of the younger Congolese SCD population [4,5,14].

It seems that our patients are more symptomatic compared to other studies. Losada et al in Trinidad reported that 72% of their cohort did not develop any acute crisis in the past two years while in our series, 75% had developed one [15]. Surprisingly, Manpreet et al in the US, documented higher frequency of vaso occlusive crisis and hospitalization admission episodes [3]. The diversity of the genotypes included in the studies does not allow us to make consistent comparison.

Chronic complications are reported to be more common in older adults and ETRJV seems to constitute the most frequent chronic complication [3,16]. Because the Heart ultrasound was underwent only on 12 patients over 47, we are unable to give an accurate estimation of the vasculopathie. However, the morbidity was found on 10/12 (83.33%) which may presume a high incidence of the ETRV in our cohort.

## Conclusion

SCD survivor rate in Africa is growing up and clinical and biological characteristics need to be known and better understand. Genetics characteristics and other factors may explain longer survival that request further studies.

### References

1. World Health Organization. Management of birth defects and haemoglobin disorders: report of a joint WHO-March of Dimes Meeting. Geneva: World Health Organization. 2006.
2. Grosse SD, Obame I, Atrash H, Amendah DD, Piel FB, Williams TN. Sickle cell disease in Africa. 2011; 41: S398-S405.
3. Manpreet KS, Cohen A. Aging in sickle cell disease: co-morbidities and new issues in management. Hemoglobin. 2015; 39: 221-224.
4. Elira Dokekias A, Nzingoula S. Profil du drépanocytaire homozygote après l'âge de 30 ans. Med d'Af Noire. 2001; 48: 411-418.
5. Tolo-Diebkilé A, Koffi Kouassy G, Sawadogo D, Kouakou B, Siransy-Bogui L, Mamadou YS, et al. Drepanocytose homozygote chez l'adulte ivoirien de plus de 21 ans. Cahiers Santé. 2010; 20: 63-67.
6. Reeves ST, Glas KE, Eltzchig H, Pathew JP, Rubenson DS, Hartman GS, et al. Guidelines for performing a comprehensive epicardial echocardiography examination: recommendations of the American Society of Echocardiography and the society of Cardiovascular Anesthesiologists. Anest Analg. 2007; 105: 22-28.
7. Morris J, Dunn D, Beckford M, Grandison Y, Mason K, Higgs DR, et al. The hematology of homozygous SCD after the age of 40 years. Br J Haematol. 1991; 77: 382-385.
8. Telfer P, Coen P, Chakravorty S, Wilkey O, Evans J, Newell H, et al. Clinical outcomes in children with sickle cell disease in living in England: a neonatal cohort in East London. Haematologica. 2007; 92: 905-912.
9. Steinberg MH, Barton F, Castro O, Pegelow CH, Dallas SK, Kutlar A, et al. Effect of hydroyurea on mortality and morbidity in adult sickle cell anemia. J Am Med Assoc. 2003; 289: 1645-1665.
10. A propos du PNUD en Republique du Congo.
11. Redelsperger MM, Bardakdjlan-Michau J, Neonato MG, Girot R. Diagnostic biologique des syndromes drépanocytaires. In: Girot R, Begué P, Galacteros F, editors. La drépanocytose. John Libbey eutrotext. 2003; 13-29.
12. Nagel RL, Rao SK, Dundq Belkhodja Q, Connolly MM, Fabry ME, Georges A, et al. The hematologic characteristics of sickle cell anemia bearing the Bantu haplotypes: the relationship between (G) γ and HbF level. Blood. 1987; 69: 1026-1030.
13. Tshilolo L, Summa V, Gregory C, Kinsiama C, Bazeboso JA, Avvisati G, et al. Fotal haemoglobin, erythrocytes containing foetal haemoglobin, and hematological features in Congolese patients with sickle cell anaemia. Anemia. 2012; 6: 1-6.
14. Kocko I, Ngolet LO, Galiba Atipo FO, Okco LT, Malanda F, Elira Dokekias A. Valeurs de l'hémogramme du drépanocytaire adulte Congolais en période intercritique. HSD. 2016; 17: 63-66.
15. LosadaR, Bravo I, Capildeo K, Charles K. Is the group of older sickle cell patients from Trinidad and Tobago different? Am J Hematol. 2006; 81: 219-220.
16. Mc Kerrel TDH, Cohen HW, Billet HH. The older sickle cell patient. Am J Hematol. 2004; 76: 101-106.

J Blood Disord - Volume 4 Issue 2 - 2017
**ISSN 2379-8009** | www.austinpublishinggroup.com


**Citation:** Ngolet LO, Atipo Tsiba Galiba FO, Malanda F, Ockouango Guelongo Ova JD and Elira Dokekias A. Feature of Older Patients with Sickle Cell Disease in the Congo. J Blood Disord. 2017; 4(2): 1048.