Open Access Library Journal
2022, Volume 9, e9132
ISSN Online: 2333-9721
ISSN Print: 2333-9705

Reasons for Consultation of Sickle Cell Patients at the Center for Mixed Medicine and SS Anemia

Paulin Mulowayi[1,2*], Berry Bongenya[3,4], Ben Bulanda[3,4], Elvis Kateba[4], Steve Kodondi[1,2], Christian Vangu[1,2], Blezz Nkolomoni[1,2], Erick N. Kamangu[4,5*]

[1]Institute for Research in Health Sciences, Kinshasa, Democratic Republic of Congo
[2]Centre for Mixed Medicine and SS Anemia, Kinshasa, Democratic Republic of Congo
[3]Faculty of Medicine, Bel Campus Technological University, Kinshasa, Democratic Republic of Congo
[4]"Focus HIV/AIDS" Research Group, Kinshasa, Democratic Republic of Congo
[5]Department of Basic Sciences, Faculty of Medicine, University of Kinshasa, Kinshasa, Democratic Republic of Congo
Email: *drmikapaul@gmail.com, *erick.kamangu@unikin.ac.cd

How to cite this paper: Mulowayi, P., Bongenya, B., Bulanda, B., Kateba, E., Kodondi, S., Vangu, C., Nkolomoni, B. and Kamangu, E.N. (2022) Reasons for Consultation of Sickle Cell Patients at the Center for Mixed Medicine and SS Anemia. *Open Access Library Journal*, **9**: e9132.
https://doi.org/10.4236/oalib.1109132

Received: July 22, 2022
Accepted: September 27, 2022
Published: September 30, 2022

Abstract

Background: Sickle cell disease is an autosomal recessive genetic disease characterized by a point mutation. In the Democratic Republic of Congo, approximately 40,000 newborns are born with sickle cell disease. **Objective:** The objective of the study is to determine the predominant reasons for consultation of sickle cell patients at the Center of Mixed Medicine and SS Anemia (CMMASS) in Kinshasa. **Methods:** This study was conducted at CMMASS. It is a retrospective study carried out on the files of elderly sickle cell patients who came for consultation in the period from January 1, 2018 to December 31, 2020. Any well-kept file of sickle cell patients over the age of 18 with a documented electrophoresis result having consulted during the study period was considered for work. The parameters of interest in this work were the different reasons for consultation. **Results:** 2463 SS homozygous sickle cell patients had been seen in consultation at the CMMASS. 1350 patients (54.8%) were women; thus giving a sex ratio of 1.21 in favor of women. The most represented age group is that of 18 to 25 years with 1148 patients (46.6%). The most common reasons for consultation in the series were osteo-articular pain (61.75%), fever (24.48%), pallor (12.59%), headache (7.8%), and cough (2.56%). **Conclusion:** Bone and joint pain was the most dominant reason for consultation among homozygous sickle cell patients at the CMMASS during the period from January 1, 2018 to December 31, 2020.

Subject Areas

Hematology

Keywords

Sickle Cell Patients, Consultation, CMASS, Kinshasa

1. Introduction

Sickle cell disease is an autosomal recessive genetic disease characterized by a point mutation of the globin b chain in position 6 of the GAG-GTG codon, the replacement of valine by glutamic acid which induces the synthesis of hemoglobin s responsible for vaso-occlusive crises and chronic hemolysis [1].

According to the WHO, nearly 5% of the population carries a gene responsible for a hemoglobin abnormality. Africa is an original home. Recent estimates predict more than 400,000 births with Major Sickle Cell Syndrome (MDS) in 2050 [2], 75% of which will be recorded in sub-Saharan Africa and India [3].

The Democratic Republic of Congo (DRC) is the third most affected country in the world by sickle cell disease after India and Nigeria, respectively the first and second most affected countries in the world, and the second country in Africa after Nigeria. It is estimated that 20 million Congolese (*i.e.* 25% to 30% of the population) are carriers of the sickle cell gene. The prevalence of the HBss is 3% and represents a heavy burden for those affected and their families. In the meantime, around 40,000 newborns are born with sickle cell disease in the DRC [4] [5] [6].

In Kinshasa, sickle cell patients are treated in specialized centers such as the Center for Mixed Medicine and SS Anemia (CMMASS), which is a public utility center managed by the Ministry of Health and Hygiene. However, there is not data on the frequent reasons of consultation of the patients in the center. Hence the present study was carried out on the files of homozygous sickle cell patients regularly followed at CMMASS-Kinshasa, with the objective of determining the reasons for consultation of these patients.

2. Methods

Setting and type of study

This study was conducted at CMMASS, which is an institution specializing in the management of sickle cell disease in Kinshasa, DRC. It is a retrospective study carried out on the files of patients who came for consultation in the period from January 1, 2018 to December 31, 2020.

Study population

It was made up of the records of sickle cell patients aged 18 and over who came for consultation at the CMMASS during the period in question.

Inclusion criteria

Any well-kept records of sickle cell patients over the age of 18 with a documented electrophoresis result who consulted during the study period were considered for work.

Parameter of interest

The parameters of interest in this study were the different reasons for consultation of sickle cell patients (osteo-articular pain, abdominal pain, chest pain, pallor, dyspnea, jaundice, fever, cough and others) at the CMMASS during the period.

3. Results

In the period from January 1, 2018 to December 31, 2020, according to patients' consultation files at the center, 2463 SS homozygous sickle cell patients had been seen in consultation at the CMMASS. One thousand three hundred and fifty (1350) patients (54.8%) were female while 1113 (45.2%) were male; thus giving a sex ratio of 1.21 in favor of women. The most represented age group is that of 18 to 25 years with 1148 patients (46.6%); followed by that of 26 to 33 years old with 719 patients (29.2%), from 34 to 41 years old with 586 patients (23.8%) and over 42 years old with 10 patients (0.4%). Table 1 presents the data mentioned above.

The most represented reasons for consultation, in the patients' fines, in the series were grouped according to the syndromes: pain reasons with osteo-articular pain for 1521 patients (61.75%), infectious reasons with fever for 603 patients (24.48%), hemolytic patterns with pallor for 310 patients (12.59%), menongo-encephalic patterns with headaches for 192 patients (7.8%), and other patterns such as cough for 63 patients (2.56%). Table 2 presents the data mentioned above in more detail.

4. Discussion

The objective of this work was to determine the reasons for consultation of sickle cell patients at the CMMASS in the period from January 1, 2018 to December 31, 2020. According to the inclusion criteria, 2463 files of sickle cell patients had been selected for the work at the CMMASS.

One thousand three hundred and fifty patients (54.8%) were female while

Table 1. Gender and age of the population.

Parameters	n	%
Gender		
Female	1350	54.8
Male	1113	45.2
Interval of age (years)		
18 - 25	1148	46.6
26 - 33	719	29.2
34 - 41	586	23.8
>42	10	0.4

Table 2. Motifs of consultation.

Parameters	n	%
Reasons for pain		
Osteo-articular pain	1521	61.75
Abdominal pain	387	15.71
Generalized pain	303	12.30
Chest pain	131	5.31
Swelling	30	1.22
Reasons for the infectious syndrome		
Fever	603	24.48
Hepatomegaly	67	2.72
Malleolar wounds	33	1.34
Splenomegaly	29	1.18
Reasons for hemolytic syndrome		
Pallor	310	12.59
Fatigue	291	11.82
Jaundice	222	9.01
Dyspnea	186	7.55
Dizziness	41	1.66
Reasons for meningoencephalic syndrome		
Headaches	192	7.80
Seizures	67	2.72
Hemiplegia	26	1.06
Reasons for Digestive Syndrome		
Vomiting	74	3.00
Diarrhea	20	0.80
Other reasons for consultation		
Cough	63	2.56
priapism	58	2.35
Vaginal pruritus	20	0.81

1113 (45.2%) were male; thus giving a sex ratio of 1.21 in favor of women. This slight female predominance in the present study is also found in various studies in the DRC [7] [8] [9]. On the other hand, other authors find no predominance between the two sexes [10] [11]. These differences would be related to the demographic data of different countries because the transmission of sickle cell disease is not linked to sex [12] [13]. The most represented age group is that of 18 to 25 years with 1148 patients (46.6%); followed by that of 26 to 33 years old

with 719 patients (29.2%), from 34 to 41 years old with 586 patients (23.8%) and over 42 years old with 10 patients (0.4%).

The most represented age group is that of 18 to 25 years with 1148 patients (46.6%); followed by that of 26 to 33 years old with 719 patients (29.2%), from 34 to 41 years old with 586 patients (23.8%) and over 42 years old with 10 patients (0.4%). In this series, the slight predominance of 46.6% compared to other age groups could be explained by the better understanding of the pathology by older subjects who better apply preventive measures and are therefore less affected by certain acute complications.. This argument corroborates the article by Chetcha Chemegni B *et al.* which reports a predominance in the age group between 15 - 24 years or 60% in their series [14] [15]. Even more, it is explained by the life expectancy of sickle cell patients. Indeed, in the United States, their life expectancy would be reduced by 25 to 30 years compared to the black American population in general [16]. In 1991, Latoudji *et al.* found in Benin a 30-year survival rate of 18.66% in homozygous sickle cell subjects [17].

The most represented reasons for consultation in the series were grouped according to the syndromes: pain reasons with osteo-articular pain for 1521 patients (61.75%), infectious reasons with fever for 603 patients (24.48%), hemolytic patterns with pallor for 310 patients (12.59%), menongo-encephalic patterns with headaches for 192 patients (7.8%), and other patterns such as cough for 63 patients (2.56%). Osteoarticular pain was the most common reason for consultation with 61.75%. Our result is superior to that of Ibrahim Keita [18], who found osteoarticular pain, pallor and jaundice with respectively 41.7%, 37.5% and 17.5% of cases.

5. Conclusion

This study reveals that osteoarticular pain was the most dominant reason for consultation in homozygous sickle cell patients, followed by fever and pallor at the CMMASS during the period from January 1, 2018 to December 31, 2020.

Conflicts of Interest

The authors declare no conflicts of interest.

References

[1] Mwanaut, I.M., Bongenya, B.I., Chuga, D., Kabasele, J.Y.D., Kateba, E.T., Okonda, M.O., Mukoka, S.K., Tshibumbu, C.K., Bulanda, B.I. and Kamangu, E.N. (2019) Seroprevalence of HIV, HBV Infections and HIV/HBV Coinfection in Polytransfused Adult Sickle-Cell Disease: Case of Center of Mixed Medicine and SS Anemia of Kinshasa. *ARC Journal of AIDS*, **4**, 18-21.

[2] Piel, F.B., Steinerg, M.H. and Rees, D.C. (2017) Sickle Cell Disease. *The New England Journal of Medicine*, **376**, 561-573. https://doi.org/10.1056/NEJMra1510865

[3] Piel, F.B., Hay, S.L., Gupta, S.D., Weatherall, D.J. and Williams, T.N. (2013) Global Burden of Sickle Cell Anemia in Children under Five, 2010-2050: Modeling Based on

Demographics, Excess Mortality, and Interventions. *PLOS Medicine*, **10**, e1001484. https://doi.org/10.1371/journal.pmed.1001484

[4] Agasa, B., Bosunga, K., Opara, A., Tshilumbu, K., Dupont, E., Vertogen, F., Vertongen, F., Cotton, F. and Gulbis, B. (2010) Prevalence of Sickle Cell Disease in Northeastern Region of the Democratic Republic of Congo: What Impact on Transfusion Policy? *Transfusion Medicine*, **20**, 62-65. https://doi.org/10.1111/j.1365-3148.2009.00943.x

[5] Tshlolo, L., Aissi, L.M., Lukusa, D., Kinshiama, C., Wembonyama, S., Gulbis, B. and Vertongen, F. (2009) Néonatal Screening for Sickle Cell Anemia in the Democratic Republic of Congo; Experience from a Pioneer Project on 31204 Newborns. *Journal of Clinical Pathology*, **62**, 35-38. https://doi.org/10.1136/jcp.2008.058958

[6] Ware, R.E., Montalembert, D.M., Tshilolo, L. and Abboud, M.R. (2017) Sickle Cell Disease. *The Lancet*, **390**, 311-323. https://doi.org/10.1016/S0140-6736(17)30193-9

[7] Thiam, L., Dramé, A., Zokébé, I.C., Niokhor, F.D., Seck, N., Boiro, D., Abdoulaye, A.N., Basse, I., Niang, B., Deme, I.L., Sylla, A., Diagne, I. and Ndiaye, O. (2017) Profils épidémiologiques, cliniques et hématologiques de la drépanocytose homozygote SS en phase inter critique chez l'enfant à Ziguinchor, Sénégal. *Pan African Medical Journal*, **28**, Article 208. https://www.doi.org/10.11604/pamj.2017.28.208.14006 https://doi.org/10.11604/pamj.2017.28.208.14006

[8] Samira, O. (2015) Drépanocytose homozygote chez l'enfant à l'hôpital provincial de Tanger: A propos de 10 cas. Thèse Méd Rabat, Université Mohammed V de Rabat, Rabat.

[9] Nacoulma, E.W.C., Sakande, J., Kafando, E., Kpowbié, E.D. and Guissou, I.P. (2006) Profil hématologique et biochimique des drépanocytaires SS et SC en phase stationnaire au Centre Hospitalier National Yalgado Ouedraogo de Ouagadougou. *Mali Medical*, **21**, 8-11.

[10] Thuilliez, V. and Vierin, Y. (1997) Le poids de la drépanocytose en milieu pédiatrique au Gabon. *Santé Publique*, **3**, 45-60. https://doi.org/10.1016/0929-693X(96)87087-4

[11] Dreux, O. (2012) Education thérapeutique pour les enfants drépanocytaires: Justifications à la mise en place et initiation de ce projet au CHU de Grenoble. Thèse Méd, Université Joseph Fournier, Grenoble.

[12] Nacoulma, E.W.C., Bonkoungou, P., Dembelele, Ye, D. and Kam, L. (2006) Les drépanocytoses majeures dans le service de pédiatrie du centre hospitalier universitaire Sourou Sanon de Bobo Dioulasso. *Medecine d'Afrique Noire*, **53**, 694-698.

[13] Kouakou, B., N'dhatz, E., Nanho, D.C., Sangare, A., Sanogo, I. and Tolo, A. (2006) Profil évolutif de la drépanocytose homozygote suivie: Expérience du service d'hématologie clinique du CHU de Yopougon. *Medecine d'Afrique Noire*, **53**, 5-10.

[14] Chetcha Chemegni, B., Ngouefo, G., Ngo Sack, F., Ngouadjeu, E., Kouemeni, L. and Kaptué Noche, L. (2017) Complications aigues de la drépanocytose aigues. *Journal Marocain des Sciences Médicales*, **21**, 18-23.

[15] Diop, S., Mokono, S.O., Ndiaye, M., Touré Fall, A.O., Thiam, D. and Diakhaté, L. (2003) La Drépanocytose homozygote après l'âge de 20 ans: Suivi d'une cohorte de 108 patients au CHU de Dakar. *La Revue de Médecine Interne*, **24**, 711-715. https://doi.org/10.1016/S0248-8663(03)00220-0

[16] Latoudji, S., Anani, L., Able, E. and Zahou, I. (1991) Morbidité et mortalité drépanocytaire au Benin. *Medecine d'Afrique Noire*, **38**, 8-9.

[17] Platt, O.S., Brambilla, D.J., Rosse, W.F., Milner, P.F., Castro, O., Steinberg, M.H. and Klug, P.P. (1994) Mortality in Sickle Cell Disease: Life Expectancy and Risk Factors for Early Death. *The New England Journal of Medicine*, **330**, 1639-1644.

https://doi.org/10.1056/NEJM199406093302303

[18] Keita, I. (2020) Aspects épidémio-cliniques de la drépanocytose dans le service de pédiatrie de l'hôpital de Sikasso, Bamako.