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Sociodemographic Profile and Clinic of Polytransfused Sickle Cell Patients in Kinshasa: Case of the Center for Mixed Medicine and SS Anemia

Alain K. K. Ilunga[1,2,3*], Gisèle K. Kabengele[2], Tarcisse K. Kilara[2], Cagod B. Inkale[2,3], Isaac T. Woto[2], Simplice K. Makoka[2,3,4], Gloria Y. Kangite[1], Bernard Masikini[1], Rycklain B. Misengele[1], Christine M. Katuanda[1], Christian P. Vangu[1], Berry I. Bongenya[3,4], Dieudonné T. Nyembue[1,5], Erick N. Kamangu[2,3*]

[1]Institute for Research in Health Sciences, Kinshasa, Democratic Republic of the Congo
[2]Service of Molecular Biology, Department of Basic Sciences, Faculty of Medicine, University of Kinshasa, Kinshasa, Democratic Republic of the Congo
[3]"Focus HIV/AIDS" Research Group, Kinshasa, Democratic Republic of the Congo
[4]Faculty of Medicine, Bel Campus Technological University, Kinshasa, Democratic Republic of the Congo
[5]Oto-Rhino-Laryngology Service, Department of Specialties, University Clinics of Kinshasa, Faculty of Medicine, University of Kinshasa, Kinshasa, Democratic Republic of the Congo
Email: *alainilunga14@gmail.com, *erick.kamangu@unikin.ac.cd

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Abstract

Background: Sickle cell disease is an autosomal recessive hereditary hemoglobinopathy. Patients with sickle cell disease (HbSS, Sβ, SC phenotypes) with permanent hemolytic anemia very often require transfusions. **Objective:** To describe the socio-demographic and clinical characteristics of polytransfused sickle cell patients in Kinshasa. **Methods:** This is a descriptive cross-sectional study of polytransfused sickle cell patients followed at the SS Anemia Mixed Medicine Center (CMMASS). Socio-demographic and clinical data were recorded on survey sheets. The parameters of interest were the district, age, gender, linguistic area, level of education, marital status, religion and place of screening for sickle cell disease, according to age of screening, weight, number of transfusion and the clinical signs of major sickle cell disease. **Results:** the median age of polytransfused sickle cell patients is 29 years. The female gender is in the majority at 51.1%. The majority of polytransfused sickle cell patients (32.2%) come from the Mont Amba district and are from the Greater Kasai area (34.4%). All polytransfused sickle cell patients are single. The majority have high school level (56.6%), are from revivalist churches (75.6%),

and were first diagnosed with sickle cell disease at CMMASS (63.75%). The median age of screening was 13 years, the median number of transfusions is 24. Vaso-occlusive crisis is the main reason for consultation (77.7%). **Conclusion:** This study reveals that the median age of polytransfused sickle cell patients is 29 years. The majority of patients are female, single, from secondary school, praying in revival churches, from the district of Mont-Amba and from the Grand Kasaï area. Screening for sickle cell disease is done late, the median number of transfusions is 24. Vaso-occlusive crisis is the main reason for consultation for polytransfused sickle cell patients.

Subject Areas

Immunology

Keywords

Sickle Cell Patients, Polytransfused, Sociodemographic, Clinic, Kinshasa

1. Introduction

Sickle cell disease is an autosomal recessive hereditary hemoglobinopathy [1]. It is characterized by a structural anomaly of hemoglobin, in which the glutamic acid in position 6 on the beta chain is replaced by valine. The abnormal hemoglobin called HbS, results from the mutation of a base of the base triplet of the 6th codon of the beta gene. The normal 6th codon of the beta gene (GAG) is mutated into an abnormal codon (GTG) [1].

The frequency of this mutation is high worldwide, particularly in Africa. It is present in more than 120 million individuals [2] [3] [4] [5]. The Democratic Republic of Congo is the 3rd country in the world affected by sickle cell disease and the 2nd in Africa after Nigeria [6]. Each year, approximately 40,000 births of children with sickle cell disease are registered, and 2% of these newborns are homozygous [7].

This mutation induces instability in the structure of hemoglobin with the risk of polymerization and deformation of red blood cells resulting in hemolysis, vaso-occlusive crises (VOC) and endothelial inflammation [8].

Patients with sickle cell disease (HbSS, Sβ, SC phenotypes) have permanent hemolytic anemia and are at risk of developing chronic or acute organ damage (brain, lung, retina, kidney, etc.) that is sometimes severe [8] [9]. However, there is no date defining organ damages nor hemolytic anemia in patients in Kinshasa.

The management of sickle cell disease often requires the use of blood transfusion to correct anemia or prevent certain complications. No data were found on the profile and the characteristics of those polytransfused patients.

Thus, the present study aims to describe the socio-demographic and clinical characteristics of polytransfused sickle cell patients in Kinshasa aged over 5 years in the Center of Mixed Medicine and SS Anemia (CMMASS).

2. Methods

2.1. Type, Scope and Period of Study

This is a cross-sectional descriptive study of polytransfused sickle cell patients at the SS Anemia Mixed Medicine Center (CMMASS). This center is a specialized care center located in the municipality of Kalamu, a public reference hospital for the care of sickle cell disease recognized in Kinshasa, Democratic Republic of Congo.

This study took place from January 14, 2022 to March 14, 2022.

2.2. Study Population, Sampling and Selection Criteria

The study population is made up of polytransfused sickle cell patients who were consulted at the CMMASS, who have received at least 2 transfusions in the last 6 months, aged over 5 years and who had agreed to participate in the study.

The size of the sample is convenient based on the duration of the inclusion period which was of 2 months (from January 14, 2022 until March 14, 2022).

2.3. Parameters of Interest

The parameters of interest included the socio-demographic data of the patients such as district, age, gender and linguistic space, level of education, marital status, religion and place of screening for sickle cell disease, and clinical elements such as screening age, weight, number of transfusions and clinical signs of major sickle cell disease.

2.4. Statistical Analysis

The information on the subjects participating in the study as well as the results were recorded on the survey sheets. The data collected were entered on Windows Excel version 2016. The analysis of the results was done on SPSS version 2020 for Windows. The statistical tests used in this study are: the student test for quantitative variables and the chi-square (x^2) test for qualitative variables.

The results are expressed as mean and standard deviation. The tables were reformatted in Excel.

2.5. Ethical Consideration

This study obtained the approval of the Ethics Committee of the School of Public Health of the Faculty of Medicine, University of Kinshasa (ESP/CE/1108/2021) and the agreement of CMMASS officials before its start.

The consent of the guardian was sought for the patients under 18 years. Polytransfused sickle cell patients were informed of the study and gave their signed agreement on an informed consent form before collecting the data.

3. Results

The sample consists of patients' files of 90 polytransfused sickle cell patients who

consulted CMMASS during the study period and who freely agreed to participate. Data were extracted from the individual files of patients.

3.1. Sociodemographic Data

Most polytransfused sickle cell patients are in the interval of 16 to 25 years at 46.7% followed by that of 6 to 15 years with 37.8% then that of 26 to 35 years with 13.3%, and finally that of 36 to 45 years and that of 46 to 55 years with 1.1% each. The median age was 29 years with extremes of 6 to 50 years.

The female gender is dominant at 51.1% against 48.9% of the male gender, thus giving a sex ratio of 1.04.

The majority of polytransfused sickle cell patients come from the district of Mont-Amba at 32.2% followed by the district of Lukunga at 26.7% then from the district of Funa at 24.4% and finally from the district Tshangu at 15%.

Most polytransfused sickle cell patients are from the Grand Kasai area at 34.4%, followed by the Grand Bandundu area at 23.3% and Central Kongo at 16.6%. Grand Equateur represents 13.3%, Grand Kivu 5.5% and the provinces of Angola 2.2%.

The data presented above are included in Table 1.

All polytransfused sickle cell patients are single for those of marriageable age.

The majority of polytransfused sickle cell patients are at secondary level at 56.6% followed by primary level at 28.8%, university students at 13.3% and illiterates at 1.1%.

The majority of polytransfused sickle cell patients are revival churches at 75.6% followed by the Catholic Church at 13.3% followed by the Protestant Church at 7.8% finally the Kimbanguist religion and the Muslim religion each have 1.1%.

The majority of polytransfused sickle cell patients were diagnosed with sickle cell disease at the CMMASS at 63.75% followed by the National Institute for Biomedical Research (INRB) at 8.75%. The Biomedical Center of Matonge (CBM) and the General Reference Hospital of Kinshasa (HGRK) at 5% each.

The results mentioned here above are presented in Table 2.

3.2. Clinical Data

Screening for sickle cell disease first took place between 1 to 5 years at 61.53% followed by the age group from 6 to 10 years at 26.92% then the age group from 11 to 15 years at 10.2% finally of the age group from 21 to 25 years at 1.28%. The median age of patients screening for sickle cell disease was 13 years.

The majority of patients weighed between 46 to 55 Kg at 33.3% followed by the group weighed from 36 to 45 Kg at 22.2%, that of 16 to 25 Kg at 17.8%, that of 26 to 35 kg at 14.4%, and finally that of 56 to 65 kg at 11.1%.

The majority of polytransfused sickle cell patients have a history of 2 to 10 transfusions at 54.4% followed by those with a history of 11 to 19 transfusions at 31.11% then those with a history of 20 to 28 transfusions at 6.6% finally those

Table 1. Distribution of polytransfused sickle cell patients according to district, age, gender and linguistic area.

Age in years	**N = 90**	**%**
6 - 15	34	37.80
16 - 25	42	46.70
26 - 35	12	13.30
36 - 45	1	1.10
46 - 55	1	1.10
Gender	**N = 90**	**%**
Male	44	48.90
Female	46	51.10
Districts	**N = 90**	**%**
Funa	22	24.40
Lukunga	24	26.70
Mont-Amba	29	32.20
Tshangu	15	16.70
Linguistic Space	**N = 90**	**%**
Grand Equateur	12	13.30
Grand Oriental	4	4.40
Grand Kasaï	31	34.40
Grand Kivu	5	5.50
Grand Bandundu	21	23.30
Kongo Central	15	16.60
Grand Katanga	0	0.00
Other (Angola)	2	2.20

Table 2. Distribution of polytransfused sickle cell patients according to Civil Status, Level of education, Religion and place of screening for sickle cell disease.

Marital status	**N = 90**	**%**
Single	90	100
Married	0	0.00
Free Union	0	0.00
Education level	**N = 90**	**%**
Illiterate	1	1.10
Primary	26	28.88
Secondary	51	56.60
University	12	13.33

Continued

Religion	N = 90	%
Catholic	12	13.33
Protestant	7	7.70
Revival Church	68	75.50
Muslim	1	1.10
Kimbanguist	1	1.10
Screening centers	**N = 80**	**%**
CMMASS	51	63.75
INRB	7	8.75
CUK	2	2.50
HGRK	4	5.00
CBM	4	5.00
Saint Joseph	1	1.25
Others	**11**	**13.75**

with a history of 29 to 37 transfusions at 5.5% and a history of 38 to 46 transfusions at 2.2%. The median number of transfusions was 24.

Polytransfused sickle cell patients consulted more for vaso-occlusive crisis at 77.77% followed by hemolytic crisis at 71.11% then for control at 15% finally for infection at 11%.

The data presented here above are included in Table 3.

4. Discussion

The objective of this study was to describe the sociodemographic and clinical characteristics of polytransfused sickle cell patients. Ninety polytransfused sickle cell patients were included in the study.

4.1. From a Socio-Demographic Point of View

The age of polytransfused sickle cell patients varies between 6 and 50 years. The age group from 16 to 25 is more dominant with 46.7%. The median age was 29 years old. These data corroborate with those carried out by Mwanaut I in Kinshasa over a period of 11 years, in 180 polytransfused sickle cell patients from the CMMASS whose median age was 28 years [10] and those of the Ngo SF study in Dakar, carried out on a period of 13 years, in 129 polytransfused sickle cell patients from the clinical hematology department of the National Blood Transfusion Center in Dakar; the median age was 27 years [11].

It is important to note that hemoglobin S almost completely replaces hemoglobin F around 12 to 48 months. This thus promotes the occurrence of sickle cell crises indicating a blood transfusion that are often more serious and more frequent during early childhood and adulthood. The number of blood transfusions

Table 3. Distribution of polytransfused sickle cell patients according to screening age, weight, number of transfusions and clinical signs (diagnoses).

Age of screening	**N = 78**	**%**
1 - 5	48	61.53
6 - 10	21	26.92
11 - 15	8	10.25
16 - 20	0	0.00
21 - 25	1	1.28
Weight	**N = 90**	**%**
16 - 25	16	17.80
26 - 35	13	14.40
36 - 45	20	22.20
46 - 55	30	33.30
56 - 65	10	11.10
Number of transfusion	**N = 90**	**%**
2 - 10	49	54.40
11 - 19	28	31.11
20 - 28	6	6.66
29 - 37	5	5.55
38 - 46	2	2.20
Diagnostic	**N = 90**	**%**
CVO	70	77.77
CH	64	71.11
Infection	11	12.22
Normal	14	15.55
CVO et CH	52	57.77
CVO et infection	9	10.00
CH et infection	8	8.88
CVO, CH et infection	7	7.77

also increases with age due to the absence of fetal hemoglobin and due to the vulnerability of these patients [12].

The female gender is in the majority at 51.1% with a sex ratio of 1.04. This predominance is also found in the study by Mwanaut I in Kinshasa [10], Ya Pongombo S.M in Lubumbashi [13] and Nacoulma in Ouagadougou in Mali [14]. On the other hand, other authors in the DRC including Abdala K.A [15] and Mashako MR [16] and in Africa Diagne and Mabiala report a slight predominance of the male sex [17] [18]. Finally, others find no predominance between

the two sexes; this is the case of Thuilliez [19] and Dreux [20]. These differences would be related to the demographic data of each country because the transmission of sickle cell disease is not linked to sex [21] [22].

These sickle cell sufferers are 75.5% revival churches [23]. This is probably related to the proliferation of revival churches [23] [24] as the transmission of sickle cell disease is not linked to religion.

Screening is done at 63.75% at CMMASS because it is a public hospital of reference for the management of sickle cell disease recognized in the city of Kinshasa and all Kinshasa attends there as this study proves.

Most polytransfused sickle cell patients are from the greater Kasai area at 34.4%. This finding seems to corroborate what Tshilolo [25] and Ya Pongombo S.M [13] have described.

Indeed, Tshilolo is the first to observe that the former colonial "Luba Empire" represented 2/3 of the case. In the work of Ya Pongombo S.M [13], the Luba Empire represents 68.3% of cases. Tshilolo locates this group of majority tribes (Luba Kasaï, Luba Katanga, Songye, Hemba and Lunda), beyond the Lualaba River and in the center south of 2 Kasaï provinces [25].

On the other hand Mashako MR [16] finds in North Kivu that it is the Kongo ethnic group which predominates with 21.7% of cases followed by the Kasaï area with 18.8%. However, the geographical distribution of the beta S gene does not show any difference between the different territories of the country [25].

All polytransfused sickle cell patients are single. This is almost similar to the results obtained by Mwanaut IM [10] and can be explained firstly by new recommendations imposing a premarital check-up, in this case hemoglobin electrophoresis, on anyone who aspires to marriage and secondly by several campaigns of sensitization initiated by the National Program for the Fight Against Sickle Cell Disease (PNLCD), aimed at reducing or even bringing to zero the number of new cases of SS Anemia in the city of Kinshasa in particular and in the DRC in general [10].

It should be noted that pregnancy remains a high-risk situation for both mother and fetus and its management requires multidisciplinary care [1]. Hence a certain fear of being able to commit to marriage to a sickle cell sufferer.

4.2. From a Clinical Point of View

Screening for sickle cell disease of cases under study took place at 61.53% between 1 to 5 years old followed by 26.92% in the age group of 6 to 10 years old. The median age was 13 years old.

These results show that the diagnosis of sickle cell disease is made with a delay in our environment. This delay would certainly be linked to the lack of parental awareness of sickle cell disease and its inaugural manifestations, but also to the use of traditional medicine during the first manifestations [10].

The median number of transfusions was 24. Tshilolo, Mukinayi BM [26] and Ya Pongombo SM [13] show that in Africa as in the DRC the first transfusion takes place around 2 years. The number of blood transfusions increases with age

due to the absence of fetal hemoglobin and due to the vulnerability of these patients [12].

Polytransfused sickle cell patients consulted for vaso-occlusive crisis in isolation (36.4%) or associated with other sickle cell crises at 77.77%.

These results are in line with other studies that have found vaso-occlusive crises to be the leading cause of hospitalization in sickle cell patients [14] [27] [28].

5. Conclusions

This study reveals that the median age of polytransfused sickle cell patients is 29 years. The female gender is in the majority. Polytransfused sickle cell patients mostly come from the district of Mont Amba and are from the greater Kasai area.

All of the polytransfused sickle cell patients are single, the majority have high school level, pray in revival churches and were diagnosed with sickle cell disease for the first time at CMMASS. Screening for sickle cell disease is done late, the median number of transfusions is 24. Vaso-occlusive crisis is the main reason for consultation for polytransfused sickle cell patients.

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Conflicts of Interest

The authors declare no conflicts of interest.

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