International Journal of Innovation and Scientific Research
ISSN 2351-8014 Vol. 69 No. 1 Oct. 2023, pp. 121-128

http://www.ijisr.issr-journals.org/

Sickle Cell Disease Thirty-Three Years of Research Experience in Gabon: A Comprehensive Review

Lucrèce M. Délicat-Loembet[1-2], Linda Sono D. E.[2-3], Arielle Mbila[2-3], and Dieudonné Nkoghe[3]

[1]Department of Biology, University of Sciences and Technology of Masuku, Franceville, Haut-Ogooue, Gabon

[2]NGO Sickle Cell Disease Organization of Gabon (SCDOGa), Franceville, Haut-Ogooue, Gabon

[3]Gabonese Ministry of Health, Libreville, Estuaire, Gabon

ABSTRACT: The extent of the scientific literature on sickle cell disease in Gabon over the last thirty years and the areas of study are not known. However, the exponential growth trend shows that there is an area of research that should be explored. The question here is to find out in which fields studies on sickle cell disease have already been carried out since 1990, 33 years of research. Because the environment can influence clinical signs in different patients, every African country is carrying out scientific research to improve the care of their patients. This review examines the history of research into sickle Cell Disease in Gabon. In Gabon, research into sickle cell disease is being carried out in all areas of epidemiology, screening, medical and psycho-clinical management, and pathologies associated with sickle cell disease and endangering the vital prognosis of Gabonese sickle cell patients. Traditional Gabonese pharmacopoeia plays a very important role in the management of this disease. Ultimately, local healthcare professionals should be familiar with the latest guidelines and resources specific to Gabon for the most accurate and up-to-date information on sickle cell disease management.

KEYWORDS: Sickle Cell Disease, Gabon, Research Developments, Management.

1 INTRODUCTION

Since the discovery of this autosomal recessive pathology by Herrick in 1910 [1], sickle cell disease has been the subject of several research studies. Discoveries about sickle cell disease have shown that it predominantly affects African populations [2], [3], yet research based on hemoglobinopathies and sickle cell disease (SCD) in particular is highly disparate. Because the environment can influence clinical signs in different patients, every African country is carrying out scientific research to improve the care of their patients. Each country moves at its own pace, and this is also the case in Gabon, where it is necessary to take stock of our knowledge of the disease. SCD is a real public health problem in Gabon. The aim of this systematic review is to take stock of the history of research into sickle cell disease in Gabon since 1990.

2 MATERIALS AND METHODS

After defining the title of our review on the history of sickle-cell anaemia in Gabon and the deadline for scientific publications, we proceeded with a targeted search on different research platforms databases. First, we conducted a preliminary literature search in order to evaluate the existence (in number and type) of publications concerning the chosen theme since 1990. In this way, we were able to define whether the chosen axes had already been developed previously. To conduct this work, we used documentary research in Medline PubMed, Google scholar, Google, and Mendeley electronic databases. Key words, such as sickle cell disease, sickle cell disease in Gabon, names of authors working on sickle cell disease in Gabon..., were used, and after reading the abstract or full text, we retained all the articles that best fit the objectives of our work. This selection work has been carried out for more than 4 months, i.e., throughout the writing of this review. The abstracts

or full text of articles that provide information on the subject matter were included and have given to the subdivision of the different sections or parts developed in the heading of the results.

3 RESULTS

3.1 EPIDEMIOLOGY

Sickle cell disease is a chronic blood disease that kills about two out of three children in Gabon before the age of five (5) years [4]. In Gabon, research into the epidemiology of this pathology is not very advanced, but some data do exist. Sickle cell disease in Gabon is similar to that observed in Central Africa, with an annual incidence of 1.34% according to a study carried out on newborns in the various provinces of Gabon [5], and a sickle cell trait carriage rate of 21.7% based on a study of over 4,000 individuals aged over 15 [6]. Sampling carried out during voluntary screening campaigns to assess the effectiveness of the rapid diagnostic test for sickle cell anemia with database comprised 299 patients, aged 6 months to 33 years, showed that homozygous HbAA phenotype was most prevalent (52.21%), followed by the homozygous S-allele (32.35%) and heterozygous AS (14.71%). There was also a small proportion of composite heterozygous SC (0.74%). the HbSS phenotype was more prevalent in children between 5 and 9 and between 10 and 14 years of age [7]. It should be pointed out that this study was carried out in Haut-Ogooué, and the majority of the participants came from this and surrounding localities. A study of Vierin Nzame et al., neonatal screening for sickle cell disease in Libreville, Estuaire province, showed that 1.80% of newborns were homozygous SS [8]. Despite this prevalence on the African continent, epidemiological data on the occurrence and evolution of these acute complications remains patchy, resulting in difficult management and follow-up of sickle cell patients [9]. Regarding the distribution of hemoglobin C and its prevalence in newborns in Africa, patches of median frequencies below 1% were predicted in Gabon [10].

3.2 SCREENING

Sickle cell disease is an autosomal recessive hemoglobinopathy. It affects up to 2% of newborns in some sub-Saharan African countries. In most patients, the incidence of complications can be reduced if screening is carried out at birth [8]. Experience in this area began in 2007, when CIRMF, in collaboration with the International Atomic Energy Agency, introduced neonatal screening on 3,957 collected blood samples in all 9 provinces of Gabon [5]. Sickle cell screening in Gabon is carried out using several experimental techniques: Isoelectrophocalization [11], HPLC [6], hemoglobin electrophoresis on alkaline PH, the Emmel test, Capillary electrophoresis (rare) and, for nearly five years now, rapid sicklce cell desease screening tests [7]. This is a real advance that gives hope to practitioners, even if it is still difficult for people to have access to these tests because of their cost. It must also be said that lack of knowledge of the disease could be an obstacle to carrying out neonatal screening for sickle cell disease.Studies carried out among parturients have shown no association between knowledge of the mother's status and authorization to test their newborns [12]. If we tend to focus on neonatal screening at the moment, it's because we want to improve patients' life expectancy. But recent studies have shown that the discovery of sickle cell status can occur at any time, given the lack of a systematic screening system for sickle cell disease in Gabon. This situation contributes to uncertainty about the life expectancy of patients with undetected sickle-cell status.

3.3 CHARACTERISTICS OF THE GABONESE SICKLE CELL PATIENT

Clinical manifestations and course of sickle-cell anemia are variable. Knowledge about the factors, possibly geographical, which influence the prognosis are still few in number but also little known or not referenced in our context.

3.3.1 PAEDIATRIC AGE

Sickle cell disease with variable cost and manifestation is submitted presents a clinical picture which can be different depending on the country but also according to the type of environment in the same country. The clinical symptoms were observed and listed during a study carried out by Thuilliez, V. and al., in 1996 and identified the main causes of hospitalization: acute anemia (36 cases before the age of 5 years); painful crisis whose frequency increased with age (23% before 5 years, 35% between 5 and 10, 42% after 10 years); infections, essentially pulmonary occurring early, and bone infections at any age [13]. Although some patients die before their diagnosis, the fact remains that a certain type of symptom can be observed in these children received in consultation such as: pyrexia, anemia and/or more often 'hand-foot syndrome', hepatomegaly, splenomegaly after five years of age and jaundice [13]. Children and adolescents with hip pain and/or lameness when walking were identified. This could be explained by bone infarction which mainly affects the femoral head but can also affect the

humeral head but can also impact growth potential [14]. Leg ulcers are also manifestations observed in adolescent sickle cell patients and in young adults with sickle cell disease in Gabon [15].

Children living with sickle cell disease with SS-Sβ0 phenotype from 5 to 19 years of age from sub-Saharan Africa, frequently present growth retardation, often associated with delayed puberty. It is greater during puberty, due to frequent delayed puberty, and more marked in boys than in girls [16].

3.3.2 ADULT PATIENTS

Since 2006 the majority of studies carried out in adult sickle cell patients are carried out at the level of the hematology department of the university hospital center of Libreville. The sickle cell phenotype in adults is 95% SS type and approximately 2% SC type. It should be noted that the management of an adult sickle cell patient is not always obvious because the passage from the pediatric department to the hematology department is not subject to a particular procedure. Which should have been the case. The clinical characteristics of adults with sickle cell disease in Gabon are mainly: vaso- occlusive crises and cholelithiasis. Some other complications are also observed like: the basal hemoglobin level is between 7 and 9g/dl, pulmonary arterial hypertension and left ventricular hypertrophy [17].

3.4 TREATMENT AND MEDICAL FOLLOW-UP

Sickle-cell disease is third rank of admission motivations and it represents 13% of total admissions and first after 4 years of medical follow-up [18]. The management of sickle cell disease in our context is still very difficult depending on the areas and the availability of clinicians. However, certain management measures are put in place to improve the overall condition of the patient. In the case of drug management, folic acid is first on the list in 98% of cases [17], [19], for painful crises, paracetamol is used as first-line treatment but most often in self-medication and sometimes in combination with anti-inflammatories, in certain other cases, pain management is exclusively treated by the exclusive use of anti-inflammatory drugs. Taking hydroxycarbamine for background treatment is very rare in our context although the indication is proven for a large number of them. The explanation for the reluctance of patients to adhere to this disease-modifying treatment is mainly ignorance of the side effects linked to this product and its effect on fertility and the high price of this drug.

The medical follow-up of the patient with sickle cell disease revolves around traditional long-term treatment (26%) and follow-up based on pharmaceutical products (73%) [19]. Nearly 1 in 3 subjects had no medical follow-up and no treatment. Since sickle cell disease is a chronic condition, its management in our context is essentially based on the prevention of acute or chronic complications. This prevention can only be effective if it is based on regular monitoring of the subject with sickle cell disease, allowing therapeutic and clinical monitoring [19]. Especially when 80% of deaths concerned children who were not followed up [13]. In our population there were 20.7% of patients who took iron every day. The prescription of iron supplementation is not systematic, but nevertheless concerns 1 out of 5 patients. Zinc supplementation concerns 3.7% of patients [19]. It has been reported in the literature that zinc supplementation had an effect on reducing the average number of infectious episodes and morbidity in sickle cell patients and improving growth and weight gain in children with sickle cell disease. Its use in Gabon is not sufficiently popularized and is not yet in prescription habits.

Managing pain in sickle cell patients is a must. The management of these patients must follow well-defined protocols that take into account the handicap caused by episodes of severe pain [20]. This is achieved by assessing pain using the DEGR and EVA scales, and by using analgesics from the WHO analgesic stepwise protocol. The pain may have multiple articular or osteoarticular locations and can be soothed by Level I analgesics in most cases, and by Level II in others. The WHO-recommended stepwise analgesic pain treatment used in this study has been shown to be effective in sickle cell pain crisis [20]. Access to Level III analgesics remains difficult in our context.

Blood transfusion is part of the means of combating sickle cell crisis in the event of anemia. Sickle Cell Desease is a major cause of maternal and neonatal mortality in tropical regions. Treatment consist of regular blood transfusion during pragnancy. In pregnant women, transfusion may be performed selectively (in function of the clinical status of the mother, fetus, and pregnancy) or prophylactically. However systematic prophylactic transfusion has been shunned due to the risk of transmitting infections and allo-immunization [21]. To avoid this kind of any problem, according to the author experiences the risks associated with more transfusions was offset by the greater safety margin and better efficacy of the prophylactic approach [21]. A study on the micronutrient status of Gabonese women at the time of pregnancy diagnosis revealed anemia in 71.2% of pregnant women. This anemia was accompanied by a drop in iron reserves in 11.1% of the women in this study. Folate and vitamin B12 deficiency were found in 2.6% and 6.6% of cases respectively [22]. The extent of the deficits found and their potential impact on pregnancy outcome suggest that they can be prevented by dietary diversification, educating young women about their eating habits and treating the infections responsible for blood spoliation [22].

A study to assess vaccination status and identify factors associated with vaccine compliance among adolescents attending school in Libreville. For adolescents with chronic illnesses such as sickle-cell anemia or epilepsy, this is an opportunity to correct care pathways [23]. Vaccination coverage of pupils at the start of secondary school is certainly better than that of the general population in Gabon. However, as the issues surrounding vaccination coverage of adolescents are not part of the Expanded Program on Immunization, they are not well understood by either parents or the education system. Awareness is needed, as these adolescents represent a growing proportion of future adults [23].

The management of priapism is also an important part of the care of male sickle cell patients in Gabon. this treatment is essentially surgical. Knowledge of the pathophysiological gic mechanisms of priapism is essential for understanding its indications and the various surgical methods of treatment [24]. In this way, the initial "symptom" priapism quickly becomes a "disease" priapism in its own right. independently of the triggering factor. In this respect, priapism is a genuine surgical emergency, because of its specific risk of acute ischemia of the corpora cavernosa, correlated with its duration, while at the same time searching for the triggering etiological factor in order to stop the pathological stimulation [25].

As regards the management of sickle cell disease by traditional medicine, several efforts must be made. Indeed, the WHO in 2004 reports that 80% of the populations of developing countries use traditional medicine, which is culturally well accepted, affordable and accessible; as a result, it encourages the development of a regulated traditional medicine policy [26]. This is an important step because nowadays several studies carried out in certain African countries (Congo et Madagascar) on the properties anti-sickling, anti-anaemic of pharmacological plants.

The main causes of death have been analyzed and the main ones are, severe anemia, infections and blood transfusion complications. Sickle cell disease is a real public health problem in Gabon. Mortality remains relatively high, but some deaths could be avoided with adequate care [27].

3.5 SICKLE CELL DISEASE AND SCHOOLING

Sickle cell disease is a pathology accompanied by several symptoms that can hinder the proper follow-up of schooling in the affected patient. Nearly 6 out of 10 children with sickle cell disease have normal schooling, 3 out of 10 children with sickle cell disease have poor schooling. These figures constitute a high proportion, but are unfortunately identical to those of the various surveys on the school career of children in general in Gabon [19].

In view of these data, it appears that sickle cell disease Sickle cell disease is therefore not in itself an obstacle to schooling, all children over the age of 5 were in school, but there is a large proportion of children with sickle cell disease who are behind in school.

Despite progress in the care of patients, many students with sickle cell disease face numerous crises every day which will unfortunately compromise their education and cause mental suffering [4]. Following a semi-structured interview with several students with sickle cell disease, it emerges that: sickle cell disease, because of its incurability, engages the student in long-term care which sometimes requires regular hospitalizations. This will have an impact on his school life and cause him a form of psychological weakening. This weakening is characterized by a number of affects and emotions such as sadness, shame, resignation, the feeling of being stigmatized and even death anxiety [4].

3.6 SICKLE CELL DISEASE AND OTHER PATHOLOGIES OR COMPLICATIONS

Sickle cell disease is associated with susceptibility to infections, as well as a predisposition to the occurrence of a number of complications that could alter the patient's general condition. Several research studies have highlighted these complications in Gabonese sickle-cell patients.

3.6.1 FEMORAL HEAD COMPLICATION

Like the humoral head, the femoral head can be affected by a bone infactus or Epiphyseal infarction that can be severely disabling for the patient. Such damage may not only affect the functional prognosis of the femoral head, but also its growth potential [14]. In fact, studies carried out between January 1989 and December 1999 (10 years) by Florent Mouba, J., and al., on a population of 22 adolescents sickle cell (SS) children presenting with difficulty walking and/or hip pain, showed that this complication has a definite impact on the lifestyle of the sickle cell child.

3.6.2 LEG ULCERS

A leg ulcer is one of the complications of sickle-cell anemia. It is a wound on the leg (including the ankle), which starts spontaneously or following a shock, and has still not healed 4 weeks after its appearance. A number of problems can arise from this complication, including aesthetic, psychological and socio-economic issues. Most of the time, patients affected by this pathology don't even know what they're suffering from. For some, it's a spell or an act of witchcraft, because it's considered an incurable wound. It is important to identify the risk factors for leg ulcers, present the clinical aspects and propose a management protocol for this complication [15].

3.6.3 GROWTH DELAY

It is known that children with sickle cell disease frequently show growth retardation, often associated with delayed puberty. A study of the growth of sickle cell children aged between 5 and 19 in five sub-Saharan African countries (Cameroon, Gabon, Ivory Coast, Mali and Senegal), compared with a control population [16]. In this study, the primary endpoint was stunting, defined as weight, height or BMI below the 5th percentile of WHO standards. The frequency of stunting was defined as a function of phenotype, age and sex, and an association was sought between stunting and the various clinical and biological parameters studied in the two sickle cell phenotype groups, using multivariate logistic regression. The main finding is that growth retardation remains frequent in children with sickle cell disease with the SS-Sβ0 phenotype aged 5 to 19 in sub-Saharan Africa [28]. It is greater during puberty, due to frequent delayed puberty, and more marked in boys than in girls, the latter presenting height and weight measurements equivalent to those of the controls from the age of 18 years. Growth retardation is associated with anemia and hyperhemolysis. However, this growth retardation observed in the sickle cell patient would not be without consequences in the long term for the sickle cell patient.

3.6.4 SICKLE CELL DISEASE AND OCULAR PATHOLOGIES

Macular ischemia refers to the extent of the macular capillaries from the perifoveolar capillary anastomotic arch. It is responsible for the occurrence of neovessels and associated complications (hemorrhages, retinal detachment). Macular ischemia is a rare but classic complication of sickle cell disease. the study of the fundus of the eye of a patient with sickle cell disease of Gabonese nationality aged 24 with a sudden drop in visual acuity of the left eye to 6/10th P2, showed a macular infactus in the left eye fundus [29]. According to the authors, Sickle cell disease would be primarily responsible for peripheral retinal capillary occlusion leading to the formation of preretinal neovascularization. Involvement of the posterior pole is rarer and can manifest as a macular infarction responsible for a sudden drop in visual acuity. This is reversible with appropriate treatment.

3.6.5 BACTERIAL INFECTIONS

Bacterial infection in children with sickle cell disease should be a major concern for clinicians, especially in terms of its management. A retrospective study over a 2-year period (January 1994-December 1995) was carried out in the pediatric department of HPO, showing bacterial infections in the subjects included in the study. the blood culture was positive 11 times and the germs isolated were: *Salmonella spp*., *Streptococcus pneumoniae, Haemophilus influenzae, Klebsiella pneumoniae and Staphylococcus aureus* [30], the population was aged 3 months to 16 years. On cytobacteriological examination of urines, the germs isolated concerned *Echerichia coli, Klebsiella pneumoniae, Enterobacter cloacae, Salmonella Tephy and Cytrobacter freundii.*

3.6.6 HYPERSPLENISM

Sickle cell anemia is a hereditary genetic disorder responsible for a sickle-shaped deformation of the red blood cells. It can give rise to numerous complications, including splenic sequestration and hypersplenism (a functional disorder of the spleen characterized by a pathological increase in the activity of certain functions. It usually involves increased sequestration or destruction of blood-forming elements, which can lead to cytopenia). Therapeutic treatment of this disorder includes splenectomy, which induces a state of hyposplenia or asplenia with sometimes fatal consequences, especially in sickle-cell patients [31]. In order to gain a better understanding of this situation in Gabon, a retrospective study was carried out between 2016 and 2019. The indication for surgery was hypersplenism (8 cases), splenic sequestration (3 cases), and splenic infarction (2 cases). The authors concluded that the indication for splenectomy in sickle cell disease is dominated by hypersplenism. In our context, total splenectomy by laparotomy seems to be the rule [31].

3.6.7 MALARIA

A study carried out at the Hôpital d'instruction des armées Omar Bongo Ondimba has shown infection (82%) dominates the acute complications found, with malaria in the lead (72%), followed by pneumopathy (22%) [9]. Besides its importance as a public health threat, SCD holds a special place in human population biology as a paradigmatic example of selective advantage of the heterozygotes, leading to balancing selection. Malaria is the first pathology observed in sickle cell patients in our malaria context, characterized by fever in 35% of cases [30]. An increase of 10% in *P. falciparum* malaria prevalence is associated with an increase by 4.3% of SCT (Sickle Cell Trait) carriers. An increase of 10 y of age is associated with an increase by 5.5% of SCT carriers. These strong associations show that malaria remains a selective factor in current human populations, despite the progress of medicine and the actions undertaken to fight this disease [32]. This recent epidemiological study in Gabon, Africa, demonstrated a positive correlation between the prevalence of Plasmodium falciparum and the prevalence of individuals with SCT, implying that malaria continues to serve as a selection factor for SCT in the current world.

The geographical superposition of distribution of sickle cell disease and malaria has malaria has become a classic in the field of cumulative pathologies infectious disease genetic disease still raises fundamental questions and motivates numerous and motivates numerous scientific research. Malaria, like sickle cell anemia affecting the red blood cell, lead to hemolytic anemia [33]. People with sickle-cell anemia (SS) are therefore not protected from severe malaria, just like people without the sickle-cell trait [34].

3.7 SICKLE CELL DISEASE AND PHARMACOPOEIA

In recent years, we've seen a surge in the number of new dietary supplements under the name of Alicament. Alicament is a word formed from two words: food and medicine. These products are presented as solutions to multiple health problems, such as sickle cell disease. Sickle cell anemia is a genetic disease caused by a defective gene that deforms the red blood cells. It manifests itself in painful seizures, anemia and increased susceptibility to infections. So these new products are touted as having the merit of relieving sickle cell crises. An interdisciplinary approach in the search for a solution to this disease has become necessary. Agri-food companies, phyto-drug laboratories and grandmother's recipes are faced not only with increased competition, but also with the demands of consumers who have become vigilant about the sanitary quality of these products.

In Gabon, Wavé-fortex is a food powder formulated from 24 wild Gabonese plants and patented by the African Intellectual Property Organization (OAPI) [35]. It has also seen the innovation of an alicament under the name KINGUELINE designed to prevent and relieve crises in sickle cell patients. Kengueline's antifalcemic effect could be due to the anthoxyanes it contains, but also to the presence of elements from the Lamiacae and Zingiberaceae families.

But grandma's recipes from Gabonese pharmacopoeia are still the most widely used. This is how certain plants such as: *Alchornea cordifolia* (Djeka) is a species of plant in the Euphorbiaceae family, *Hibiscus sabdariffa* (Bissap), *Manihot esculenta* (cassava leaves), *Moringa oleifera* (moringa). The risks to the patient of using these plants are overdosing, since plants such as manioc contain cyanide, and can cause cyanide poisoning. As for Bissap, if the dose is not respected, it can lead to liver problems. But it should be noted that indigenous populations have more or less mastered the ancestral use of these products, which are the only ones to which people living in rural areas have access.

3.8 ISOLATED INITIATIVES AND IMPLICATIONS FOR SOCIAL WORK INTERVENTION

Despite the Gabonese government's growing awareness of the importance of this pathology, materialized by the creation of the National Sickle Cell Disease Program in the mid-2000s, care is still based on individual initiatives, led by a few passionate and dedicated people united in associations or NGOs. Although very important in the process of reducing sickle cell births, genetic counseling is non-existent, prenatal diagnosis is still prohibitively expensive and inaccessible to the majority of couples, neonatal diagnosis is still at the experimental stage, there is no specifically dedicated service, transfusion treatment is still problematic.

Improving care requires a proactive policy that focuses on the organization of neonatal screening, the creation of specialized centers run by multidisciplinary teams, and the provision of free medication, vaccines and complementary follow-up examinations [36].

Over the past twenty years, a growing number of associations and NGOs have come together to fight sickle cell disease. There are still many unexplored areas in the multidisciplinary management of sickle cell disease. As part of its support for research, information and education on sickle-cell anemia, the Sickle Cell Disease Organization of Gabon (SCDOGa) NGO, which has already been in existence for five years, has set itself the task of carrying out studies on the type of mass screening that

can be adapted to our context and, above all, to our country's level of development, requiring fewer resources, less time for experimentation and shorter training periods [7].

This commitment also involves understanding the dynamics and level of medical care as well as the socio-economic situation of people living with sickle cell disease [37]. Which showed that people with sickle cell disease in Gabon Screening for sickle cell disease is not included in the habits of the population, only 26% of the study population has already been screened only 7.8% of households screened their children. The population of Franceville was found to be poor overall, with a monthly income of less than 150,000. However, there was no correlation between monthly income and sickle cell disease screening and between monthly income and having a sickle cell child. Low income could negatively influence the medical management of a person living with sickle cell disease [37].

4 CONCLUSION

It is therefore observed that with setup of facilities that promote regular medical check-ups, people living with sickle cell anemia will improve their quality of life by reducing the number of premature deaths and complications. These facilities can then effectively manage pain, Infections and also offer patient and caregiver education. Usage of Hydroxyurea can be introduced as this medication has been shown to reduce the frequency and severity of painful episodes and complications. Other interventions include availability of blood in case of blood trafusion, Oxygen therapy for management of acute events, Diet and hydration, Educationa and Councelling and also formation of support groups that help manage the social aspect of dimistifying, myths and misconceptions about sickle cell disease. Finaly, local healthcare professionals should be familiar with the latest guidelines and resources specific to Gabon for the most accurate and up-to-date information on sickle cell disease management.

ACKNOWLEDGMENTS

The authors gratefully acknowledge the significant contribution to this project of Selina Olwanda Ogweno

REFERENCES

[1] A. Chiabi and M. Beyeme-Owono, «Epidémiologie de la drépanocytose,» *Clin. Mother Child Heal.,* vol. I, no. 1, pp. 6–8, 2004.
[2] I. Rusanova, Epidemiology of sickle cell disease (SCD). 2013.
[3] P. McGann and C. Hoppe, «The pressing need for point-of-care diagnostics for sickle cell disease: A review of current and future technologies,» *j,* vol. 67, 2017, doi: 10.1016/j.bcmd.2017.08.010.
[4] U. D. E. L. A. Narcissique, «VIVRE AVEC LA DRÉPANOCYTOSE EN MILIEU AU REPLI SUR SOI Firmin Marius Olivaint NYAMA Université Omar Bongo de Libreville Ces hospitalisations fréquentes et parfois longues vont donc avoir un impact considérable sur leur scolarité et occasionner par conséqu,» pp. 74–86, 2019.
[5] L. M. Delicat-loembet *et al.,* «Neonatal Screening of Sickle Cell Disease in Gabon : A Nationwide Study,» vol. 11, pp. 1–7, 2022, doi: 10.35248/2167-0897.22.11.376.Citation.
[6] L. M. Délicat-Loembet *et al.,* «Prevalence of the sickle cell trait in Gabon: a nationwide study.,» *Infect. Genet. Evol. J. Mol. Epidemiol. Evol. Genet. Infect. Dis.,* vol. 25, pp. 52–56, Jul. 2014, doi: 10.1016/j.meegid.2014.04.003.
[7] L. M. Delicat-loembet, A. D. N. Mabiala, and U. Bisvigou, «Diagnosis of Sickle Cell Disease in Gabon Using Sickle SCAN ® : A Point-of-Care Blood Test,» vol. 6, no. 2, pp. 1–9, 2022, doi: 10.21106/ijtmrph.315.
[8] Y. VIERIN NZAME, I. BOUSSOUGOU BU BADINGA, J. KOKO, P. BLOT, and A. MOUSSAVOU, «Dépistage néonatal de la drépanocytose au Gabon,» *Med. Afr. Noire,* vol. 59, no. 2, pp. 95–99, 2012.
[9] R. Okoue Ondo *et al.,* «Complications aiguës des syndromes drépanocytaires majeurs: expérience de l'Hôpital d'instruction des armées Omar Bongo Ondimba,» *J. Eur. des Urgences Réanimation,* vol. 34, no. 2, pp. 82–91, 2022, doi: https://doi.org/10.1016/j.jeurea.2022.08.003.
[10] F. B. Piel *et al.,* «The distribution of haemoglobin C and its prevalence in newborns in Africa,» *Sci. Rep.,* vol. 3, pp. 1–8, 2013, doi: 10.1038/srep01671.
[11] «Neonatal Screening of Sickle Cell Disease at the Centre Hospitalier Universitaire de Libreville: Epidemiological and Clinical Aspects EC PAEDIATRICS Research Article,» vol. 11, no. 2019, pp. 24–29.
[12] L. E. Mombo, L. K. Makosso, C. Bisseye, K. Mbacky, J. M. Setchell, and A. Edou, «Acceptability of neonatal sickle cell disease screening among parturient women at the Paul Moukambi Regional Hospital in rural Eastern Gabon, Central Africa.,» *Afr. J. Reprod. Health,* vol. 25, no. 3, 2021.
[13] V. Thuilliez, V. Ditsambou, J. R. Mba, S. Mba Meyo, and J. Kitengue, «Aspects actuels de la drepanocytose chez l'enfant au Gabon,» *Arch. Pediatr.,* 1996, doi: 10.1016/0929-693X (96) 87087-4.

[14] J. Florent Mouba, M. Mimbila, L. E. Lentombo, J. F. Thardin, and A. Ondo, «Nécrose aseptique de la tête fémorale chez l'enfant drépanocytaire: Expérience de Libreville (Gabon),» *Cah. Sante,* vol. 21, no. 2, 2011, doi: 10.1684/san.2011.0244.

[15] J. F. Mouba, C. Mengue, L. Eko, and A. Ondo, «Ulcères de jambe chez le drépanocytaire à Libreville (Gabon): Aspects cliniques et prise en charge,» *Med. Sante Trop.,* vol. 24, no. 1, 2014, doi: 10.1684/mst.2014.0300.

[16] L. Alexandre *et al.,* «Étude de la croissance chez les enfants drépanocytaires de 5 pays d'Afrique subsaharienne au sein de l'étude CADRE,» *La Rev. Médecine Interne,* vol. 37, pp. A66–A67, 2016. doi: https://doi.org/10.1016/j.revmed.2016.10.016.

[17] M. Igala *et al.,* «Clinicobiological features and management of adult sickle cell patients at Libreville University Teaching Hospital,» *Pan Afr Med J,* vol. 23, no. 8, pp. 61–66, 2022.

[18] V. Thuilliez and Y. Vierin, « [The importance of sickle cell anemia in a pediatric environment in Gabon],» *Le poids la drepanocytose en milieu Pediatr. au Gabon.,* vol. 9, no. 1, pp. 45–60, 1997, [Online]. Available: http://ovidsp.ovid.com/ovidweb.cgi?T=JS&PAGE=reference&D=med4&NEWS=N&AN=9432415.

[19] S. Minto'o Rogombé *et al.,* «Article Original Suivi Médical et Scolarité de l'Enfant Drépanocytaire au Gabon Relation between medical monitoring and schooling of sickle cell children in Gabon,» *J. Med. Heal. Sci.,* vol. 19, no. January, pp. 74–78, 2018.

[20] A. Moussavou, Y. Vierin, C. Eloundou-Orima, M. Mboussou, and M. Keita, «Prise en charge de la douleur drépanocytaire selon les critères de l'Organisation mondiale de la santé,» *Arch. Pediatr.,* vol. 11, no. 9, 2004. doi: 10.1016/j.arcped.2004.03.100.

[21] D. R. Moussaoui, L. Chouhou, K. Guelzim, J. Kouach, M. Dehayni, and H. S. Fehri, « [Severe sickle cell disease and pregnancy. Systematic prophylactic transfusions in 16 cases].,» *Drepanocytose majeure grossesse. Transfusions Prophyl. Syst. a Propos 16 cas.,* vol. 62, no. 6, pp. 603–606, 2002, [Online]. Available: http://0-ovidsp.ovid.com.wam.city.ac.uk/ovidweb.cgi?T=JS&PAGE=reference&D=med4&NEWS=N&AN=12731306.

[22] E. N.-M. a Félix Ovono Abessolo a, Jean-Pierre Ngou-Mve Ngou b, Jacques Bang Ntamack c, Alexandrine Sylvie Nsi a, Jean-François Meye c, «No Volume 2011, Issue 436, November 2011, Pages 67-71 Title Statut en micronutriments de la femme gabonaise lors du diagnostic de sa grossesseMicronutrient status in Gabonese women at the time of the diagnosis of the pregnancy,» pp. 3–6.

[23] U. Bisvigou *et al.,* «Assessment of vaccination status and booster vaccinations in adolescents attending school in Libreville, Gabon,» *Pan Afr. Med. J.,* vol. 35, pp. 1–10, 2020, doi: 10.11604/pamj.2020.35.74.20024.

[24] L. Falandry, «Traitement chirurgical du priapisme experience de 56 cas en milieu Africain,» *Med. Trop.,* vol. 60, no. 1, 2000.

[25] L. Falandry *et al.,* « [Surgical treatment of priapism: experience of 56 cases in an African setting].,» *Med. Trop. (Mars).,* vol. 60, no. 1, 2000.

[26] World Health Organization (WHO), «WHO Traditional Medicine Strategy 2014-2023,» *World Heal. Organ.,* pp. 1–76, 2013, doi: 2013.

[27] J. Koko, D. Dufillot, J. M. Ba-Meyo, D. Gahouma, and F. Kani, «Mortalite des enfants drepanocytaires dans un service de pediatrie en Afrique Centrale,» *Arch. Pediatr.,* 1998, doi: 10.1016/S0929-693X (98) 80003-1.

[28] L. Alexandre-Heymann *et al.,* «Prevalence and correlates of growth failure in young African patients with sickle cell disease,» *Br. J. Haematol.,* vol. 184, no. 2, 2019, doi: 10.1111/bjh.15638.

[29] B. Sautiere, V. Thoorens, J. Copin, C. Rose, J. F. Rouland, and P. Labalette, «462 Infarctus maculaire chez une patiente porteuse d'une drépanocytose homozygote SS,» *J. Fr. Ophtalmol.,* vol. 31, p. 150, 2008. doi: https://doi.org/10.1016/S0181-5512 (08) 71060-X.

[30] J. Koko, F. Kani, A. Reymond-Yeni, G. Onewin-Andjanga, A. Moussavou, and D. Gahouma, «Infections bacteriennes chez l'enfant drepanocytaire a Libreville,» *Archives de Pediatrie*. 1999. doi: 10.1016/S0929-693X (00) 86994-8.

[31] D. M. K *et al.,* «Articles originaux,» vol. 20, 2022.

[32] E. Elguero *et al.,* «Malaria continues to select for sickle cell trait in Central Africa,» *Proc. Natl. Acad. Sci.,* p. 201505665, 2015, doi: 10.1073/pnas.1505665112.

[33] A. Centrale, C. International, D. R. Biomédicales, D. F. Cirmf, L. Docteur, and J. Paul, «En direct 2,» pp. 27–28, 2011.

[34] A. Kawahara *et al.,* «Successful Treatment of Acute Chest Syndrome with Manual Exchange Transfusion in a Patient with Sickle Beta+-thalassemia,» *Intern. Med.,* vol. 58, no. 11, pp. 1629–1634, 2019, doi: 10.2169/internalmedicine.1753-18.

[35] A. N. Lepengue, A. Souza, J. F. Yala, J. Lebamba, J. Mavoungou, and B. M'batchi, «Etude De Quelques Caracteristiques Physicochimiques Et Biochimiques De Wavé-fortex Un Complement Alimentaire Naturel Du Gabon,» *Eur. Sci. Journal, ESJ,* vol. 12, no. 33, p. 508, 2016, doi: 10.19044/esj.2016.v12n33p508.

[36] J. Koko, «Prof. Jean KOKO Département de Pédiatrie Faculté de Médecine Université des Sciences de la Santé (USS) Libreville (GABON),» no. April 2013, 2015, doi: 10.13140/2.1.1350.4806.

[37] Delicat-Loembet. LM *et al.,* «Sickle Cell Disease in Southeast Zone of Gabon a Central African Country : A Socio-Economic Study,» vol. 3, no. 2, pp. 942–949, 2023.