MCT Insights - Sickle Cell Disease in West Africa

Sickle cell disease also known as Sickle Cell Anemia is a common genetic disorder caused by an abnormal hemoglobin. The mutant genes of the disease are inherited from both the father and the mother.
Sickle cell disease (SCD) has been recognized as a public health priority since 2005. It is a hemoglobinopathy that is observed today worldwide. It is even recognized as the most widespread genetic disease in the world. The high frequency of healthy carriers (having inherited a mutant gene from only one of the two parents) in some regions leads to a high rate of newborns affected by this condition. According to WHO, the genes causing haemoglobinopathies are found in about 5% of the world's population[1]. Each year, more than 500 000 sickle cell children are born, including 300 000 in Africa, half of children die in Africa before the age of five.[2]

Table: case of some countries of West Africa

	Mali[3]	Sénégal[4]	Burkina Faso[5]	Ivory Coast[6]	Ghana[7]	Nigeria[8]
Global prevalence of the S gene	12%	8 - 10%	**28,9%**	14%	30 %	24%
Prevalence Patients	**3%**	0,5%	1,3%	2%	2%	2%

We have examined the case of some West African countries (see table above). In Mali, the prevalence of the S gene is estimated at 12% with 3% of known sickle cell patients[9]. It is in Burkina Faso that the prevalence of the S gene is the highest, estimated at 28.9%. Ivory Coast and Senegal are affected with a prevalence of 14% and 8-10% respectively for all forms, of which 2% in Ivory Coast and 1.3% in Senegal manifest the disease form. These figures may be below the reality despite the progress made in epidemiological surveillance in recent years in Africa. Many of these patients escape the formal health care system.

To get an idea of the number of potential patients with the prevalence cited in the table above, we made a simple estimate with the current population of some countries. The results are quite surprising, the estimated number of patients with SCD that Nigeria is supposed to manage is more than 3,900,000 sickle cell patients, for the Ivory Coast nearly a million, Ghana and Mali about 580,000. This gives us an idea of the challenge.

The manifestations of SCD are well known to doctors, major sickle cell syndrome includes about 3 types of manifestations: chronic hemolytic anemia; vaso-occlusive phenomena; susceptibility to infections (especially encapsulated germs).
Standard treatment includes supportive care, intravenous infusion, antibiotic therapy and simple blood transfusion or exchange transfusion. In Bamako, Research center named “Centre de Recherche et de Lutte contre la Drépanocytose (CRLD)” has a trans-cranial

Doppler ultrasound for screening abnormalities of the cerebral circulation. For other complications, admission to the hospital is required.

Unfortunately, vast majority of patients are in reality children whose parents cannot afford proper medical treatment. This leads to improper medical follow-ups of the pathology and therefore is impacting patient outcome as SCD complications are numerous, unpredictable and serious. According to Professor Dapa Diallo, General Director of CRLD in Bamako, these children, especially those with the severe form of the disease do not reach adulthood. Most of them die before the age of five in terrible suffering.[10]

Those lucky enough to cross this age will face serious growth problems and other complications due to lack of adequate care. According to the Professor, anemia alone ruins many families because the expenses involved are unsustainable.

This raises this notion of equity in access to quality care. The governments of West African countries have rather limited resources. Most of the affected countries have programs to fight sickle cell disease. Unfortunately in these countries, these good initiatives are limited in scope. For Mali for example, there are only four services that are entirely dedicated to sickle cell patients, including only one outside Bamako.

Because of its high frequency, its significant lethality and unacceptable in these countries, associations against SCD have been created. They play an important role by conducting sessions of animation, information and sensitization of the populations for a better knowledge of the disease. One of the concerns of the associations is the customary practices, such as consanguineous marriage which is not preceded by premarital checkup, which suggests that the prevalence rate of the disease is increasing more and more.

Screening campaigns are not often initiated because the usual diagnostic methods such as electrophoresis of hemoglobin are very expensive and therefore difficult to implement in mass screening campaigns. New SCD Screening with a rapid diagnostic test will improve significantly disease diagnosis. Prenatal screening is not yet in effect.

CRLD plays a key role on the regional fight against sickle cell disease, by welcoming patients from the region but also from neighboring countries of Guinea and Mauritania. This center now manages a cohort of more than 8 000 sickle cell patients[11]. This also means that the center is in constant need of partnership and funding. This is practically the case in other countries of the sub-region. The CRLD works through programs to improve patient care, early detection, training of health professionals, awareness and education, and the development of clinical research.

Despite the presence of brilliant researchers in the field, there is still low participation of the West African community in medical research on sickle cell disease. Indeed very few clinical studies on SCD are currently conducted in West Africa. As per international clinical trial databases, only two observational studies sponsored by an NGO called "Cardiologie et Développement" (active in Ivory Coast, Mali and Senegal) was noted. We count 6

interventional studies active in 2018 in West Africa (2 sponsored by AstraZeneca in Ghana, 1 sponsored by Global Blood Therapeutics in Ghana and in Nigeria and 3 studies from Vanderbilt University Medical Center in Nigeria)[12].

However, West Africa would be a very interesting option for the pharmaceutical industry to address the recruitment challenge facing most clinical studies on genetic diseases. This would also allow patient alternatives in many cases.

Written by Mahamadou Koné MD,

Clinical Research Associate at MCT (MCT is a regional CRO specialized on Africa and Middle East) based in Bamako/ Mali.

[1] **WHO.int ; Drépanocytose,** Rapport du Secrétariat; PDF, page 1. Url : http://apps.who.int/gb/archive/pdf_files/WHA59/A59_9-fr.pdf accessed 10-21-18

[2] **Hémoglobinoses** - Médecine tropicale, pDF ; Page 1 ; Url: http://medecinetropicale.free.fr/cours/hemoglobinoses.pdf accessed 10-21-18

[3] **Sangho H et al.** [Management of sickle cells disease by households in Bamako]. PubMed: Mali Ms. d. 2009; 24 (3): 53-6. Url: https://www.ncbi.nlm.nih.gov/pubmed/20093217 accessed 10-21-18

[4] **Seck M et al.** [Profile of stroke of SC sickle-cell patients in Dakar: a case-control study with SS sickle cell form]. Pubmed, Mali Me d . 2017; 32 (4): 7-12. Url: https://www.ncbi.nlm.nih.gov/pubmed/30079643 accessed 10-21-18

[5] **Simpore J et a**l. [Glucose-6-phosphate dehydrogenase deficiency and sickle cell disease in Burkina Faso]. Pubmed, Pak J Biol Sci. 2007 Feb 1; 10 (3): 409-14. Url: https://www.ncbi.nlm.nih.gov/pubmed/19069510 accessed 10-21-18

[6] **Tossea SK et al.** [Cross sectional study on prevalence of sickle cell alleles S and C among patients with mild malaria in Ivory Coast]. BMC Res Notes. 2018 Apr 2;11(1):215. Url : https://www.ncbi.nlm.nih.gov/pubmed/29609623 accessed 10-21-18

[7] **Kyerewaa Edwin A, Edwin F, Etwire V**. Controlling Sickle Cell Disease in Ghana--ethics and options. Pan Afr Med J. 2011;10:14. Url: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3282939/#__ffn_sectitle accessed 10-21-18

[8] **WHO.int ; Drépanocytose,** Rapport du Secrétariat; PDF, page 1. Url : http://apps.who.int/gb/archive/pdf_files/WHA59/A59_9-fr.pdf accessed 10-21-18

[9] **Sangho H et al.** [Management of sickle cells disease by households in Bamako]. PubMed: Mali Ms. d. 2009; 24 (3): 53-6. Url: https://www.ncbi.nlm.nih.gov/pubmed/20093217 accessed 10-21-18

[10] **Sangho H et al.** [Management of sickle cells disease by households in Bamako]. PubMed: Mali Ms. d. 2009; 24 (3): 53-6. Url: https://www.ncbi.nlm.nih.gov/pubmed/20093217 accessed 10-21-18

[11] **Pierre Fabre Foundation.** Web page, Url: https://www.fondationpierrefabre.org/fr/suivre-notre-action/10-ans-dengagement-dans-la-lutte-contre-la-drepanocytose accessed 10-21-18

[12] **Sources :** Web site https://clinicaltrials.gov/ct2/home ; Web site https://www.clinicaltrialsregister.eu/ctr-search/search accessed 12-03-18