HHS Public Access

Author manuscript

N Engl J Med. Author manuscript; available in PMC 2020 May 29.

Published in final edited form as:

N Engl J Med. 2017 July 20; 377(3): 302–303. doi:10.1056/NEJMc1706325.

Sickle Cell Disease

Baba Inusa, F.R.C.P.C.H., D.C.P.(Hematology),
Guy's and St. Thomas' NHS Foundation Trust, London, United Kingdom

Joyce Popoola, F.R.C.P., Ph.D.,
St. George's University Hospitals NHS Foundation Trust, London, United Kingdom

Ambroise Wonkam, Ph.D., D.Med.
University of Cape Town, Cape Town, South Africa

TO THE EDITOR: The review of sickle cell disease by Piel et al. (April 20 issue)[1] is timely and highlights the need to address the lack of research about this disease in sub-Saharan Africa. The authors rightly state that in the past two decades, childhood mortality has been reduced in sub-Saharan Africa, but the survival data cited by Piel et al. were derived from a single-site study performed almost four decades ago.[2]

Two-year follow-up data from a pilot cohort study in Nigeria (Table 1) show that survival among children with sickle cell disease remains poor in sub-Saharan Africa.[3] There are no conclusive data to support the use of chemoprevention in addition to insecticide-treated bed nets for prophylaxis against malaria in patients with sickle cell disease.[4]

With regard to Figure 3 in the review by Piel et al., multiple data suggest that the Cameroon haplotype of the β-globin gene (*HBB*) is associated with a more severe phenotype than the Benin haplotype; thus, in the figure, the Cameroon haplotype should have been to the right of the Benin haplotype. In addition to fetal hemoglobin (HbF)-promoting loci and the coinheritance of α-thalassemia that are established genetic modifiers of sickle cell disease, data also provide support for genetic risk markers of renal dysfunction in *APOL1* and *HMOX1*[5] and of cholestasis in *UGT1A1*.

Acknowledgments

Dr. Inusa reports receiving a grant (N253) from the European Community, United Nations Development Program, Joint Migration and Development Initiative. No other potential conflict of interest relevant to this letter was reported.

References

1. Piel FB, Steinberg MH, Rees DC. Sickle cell disease. N Engl J Med 2017;376:1561–73. [PubMed: 28423290]
2. Molineaux L, Fleming AF, Cornille-Brøgger R, Kagan I, Storey J. Abnormal haemoglobins in the Sudan savanna of Nigeria. III. Malaria, immunoglobulins and antimalarial antibodies in sickle cell disease. Ann Trop Med Parasitol 1979;73:301–10. [PubMed: 315213]

baba.inusa@gstt.nhs.uk.

3. Inusa BP, Daniel Y, Lawson JO, et al. Sickle cell disease screening in northern Nigeria: the coexistence of B-thalassemia inheritance. Pediat Therapeut 2015;5:262.

4. Sundell K, Jagannathan P, Huang L, et al. Variable piperaquine exposure significantly impacts protective efficacy of monthly dihydroartemisinin-piperaquine for the prevention of malaria in Ugandan children. Malar J 2015;14:368. [PubMed: 26403465]

5. Geard A, Pule GD, Chetcha Chemegni B, et al. Clinical and genetic predictors of renal dysfunctions in sickle cell anaemia in Cameroon. Br J Haematol 2017 5 3 (Epub ahead of print).

Table 1.

Two-Year Follow-up of Infants Who Received a Diagnosis of Sickle Cell Disease at 0 to 6 Months of Age.*

Finding	Infants with Sickle Cell Disease (N = 48)	Controls (N = 96)	Total (N = 144)
	number (percent)		
Alive	26 (54)	72 (75)	98 (68)
Died	1 (2)	1 (1)	2 (1)
Family relocated	12 (25)†	7 (7)	19 (13)
No telephone in home	6 (12)	13 (14)	9 (13)
Family's telephone switched off	3 (6)	3 (3)	6 (4)

*Data are from the Sickle Cell Cohort Study: A Sustainable Pilot Scheme (http://www.migration4development.org/en/projects/sickle-cell-cohort-study-sustainable-pilot-scheme) conducted in Abuja, Nigeria.

†P=0.003 for the comparison with controls.