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Editorial

Sickle cell prevalence in the universe problem in 2024

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Sickle cell disorder (SCD) is an autosomal recessive inherited disorder characterized by sickle-shaped red blood cells. SCD is a monogenic disorder caused by a single nucleotide substitution at position 6 (glutamine is replaced by valine) of the β-globin gene, leading to polymerization of the resulting sickle hemoglobin variant. The SCD has both a heterozygous and a homozygous form and sometimes a double heterozygous form. Sickle cell trait, sickle cell anemia, and sickle beta thalassemia are the common forms of these disorders.

The most severe form is the symptomatic homozygous sickle cell disease and persons with mutation on one gene developed sickle cell traits/sickle cell carriers and are clinically asymptomatic. Sometimes, the double heterozygous behaves like the thalassemia intermedia phenotype. High Sickle Cell Disease burden was historically seen in malaria andemic regions of Africa, the Middle East, the Caribbean, and South Asia.

From 2000 to 2021, total births of babies with sickle cell disease globally increased by 13.7%. Globally, the number of people living with sickle cell disease increased by 41.4%. It is around 5.46 million in 2000 and increase to 7.74 million in 2021.[1]

Globally, the highest Sβ allele frequencies have been reported in India. India has been ranked the second affected country in terms of sickle β-globin thalassemia.[2]

As per the World Health Organization, 4.5 – 5% of the world's population are the traits of hemoglobinopathies. In India, there are 30 million carriers, and the mean prevalence is 3.3% across the nation.[3,4] The incidence of sickle cell anemia in India varies between 1% and 44%.[5]

Birth prevalence of SCD (child with age ≤ 1 year old) is usually measured through newborn screening tests. The birth prevalence was highest in sub-Saharan African countries (500–2000/Lakh population), followed by South America and the Caribbean Islands (20–1000/Lakh). In the European countries and the USA, the birth prevalence was ≤500/lakh population. The global hotspots of SCD are India, the Middle East, North-East Africa, and Sub-Saharan Africa. The prevalence of SCD was too scattered to provide consistent prevalence in each region through analysis. Mortality appeared high in low-resource settings, including India and Africa.[6]

In the previous issue of the Journal, Tina Katamea *et al.* added the scientific literature on the prevalence of sickle cell disease in Lubumbashi, Democratic Republic of the Congo.[7] They conducted newborn screening in the hospitals in Lubumbashi city in the Democratic Republic of the Congo. Newborn blood samples were examined by capillary electrophoresis. The prevalence of the sickle cell trait in newborns is 26.21%, and sickle cell disease is 5.01%. This is tertiary center data, which may not reflect the Democratic Republic of the Congo.

Several systemic studies were required to determine the exact prevalence of sickle disorder in developing countries.

REFERENCES

1. GBD 2021 Sickle Cell Disease Collaborators. Global, regional, and national prevalence and mortality burden of sickle cell disease, 2000-2021: A systematic analysis from the Global Burden of Disease Study 2021. Lancet Haematol 2023;10:E585-99.
2. Piel FB, Patil AP, Howes RE, Nyangiri OA, Gething PW, Dewi M, *et al*. Global epidemiology of sickle haemoglobin in neonates: A contemporary geostatistical model-based map and population estimates. Lancet 2013;381:142-51.
3. Balgir RS. The genetic burden of hemoglobinopathies with special reference to community health in India and the challenges ahead. Indian J Hematol Blood Transfus 2002;20:2-7.
4. Verma IC, Choudhry VP, Jain PK. Prevention of thalassemia: A necessity in India. Indian J Pediatr 1992;59:649-54.
5. Yaish HM. Thalassemia. Available from: https://www.emedicine.com/ped/topic2229.html [Last accessed on 2024 May 27].
6. Colombatti R, Birkegård C, Medici M. Global epidemiology of sickle cell disease: A systematic literature review. Hemasphere 2022;6(S3):3826.
7. Katamea T, Mukuku O, Mpoy CW, Mutombo AK, Luboya ON, Wembonyama SO. Newborn screening for sickle cell disease in Lubumbashi, Democratic Republic of the Congo: An update on the prevalence of the disease. J Hematol Allied Sci. 2023;3:120-4.

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