

Screening infants for sickle cell disease in sub-Saharan Africa: starting the journey to a sustainable model in primary care



In the WHO African region, an estimated 5% of deaths in children younger than 5 years result from sickle cell disease, and up to 16% of deaths in some west African countries.¹ The UN recognised sickle cell disease as a public health priority in 2009.² Haemoglobin disorders remain neglected in terms of global support and resourcing, in view of their increasing contribution to the global disease burden.³

Sickle cell disease in Africa is increasingly visible, with more affected infants surviving to later childhood after roll-out of *Haemophilus influenzae* and pneumococcal vaccinations. Increased survival of affected infants in African populations, where the birth prevalence of sickle cell disease can range from 0.5% to 2%, is leading to more affected children reaching older ages. Epidemiologists have shown that an infant mortality rate of 50 per 1000 livebirths, which is achieved in an increasing number of African countries, is the point at which inherited and congenital conditions become increasingly visible.⁴ These conditions mainly affect children, but sickle cell disease will increasingly become a long-term condition in adults in Africa too.

Population screening programmes for sickle cell disease are effective in the USA, Europe, and Brazil.⁵ In Africa, moving from small pilot studies, typically in large urban centres linked to specialised hospitals,⁶ to whole population national screening programmes has proved difficult. Identifying ways to introduce newborn screening alongside sustainable and scalable care pathways for affected infants is key to converting evidence into action. Ghana has led the way on developing a network of care for sickle cell disease across the country, and securing hydroxycarbamide for all children, but whole population screening has been slow to develop.

In *The Lancet Haematology*, Obiageli Nnodu and colleagues⁷ make a useful contribution on a possible scalable model for screening that could offer population coverage and access to care in rural and urban African settings. Nnodu and colleagues show that point-of-care testing is as sensitive and specific as the gold standard laboratory method, high performance liquid chromatography, in this setting. Validation of

two point-of-care test devices against existing high-performing specialist laboratory methods is excellent and an important step, recognising that minimising costs of tests is also key.

Perhaps most importantly for the sustainability of such initiatives, Nnodu and colleagues show the feasibility of including the testing process into existing immunisation programmes,⁷ which already have high uptake. This is a welcome start to developing the idea of a system-wide approach to embedding screening for sickle cell disease into existing care pathways.⁸ Integration within existing systems is important with limited resources, to enable scaling up of programmes and for sustainability, and it fits with government priorities focused on primary care for all. Other elements of a whole system are also important, including a population register and widespread education of both the public and professionals to address the understanding and stigma attached to this genetic condition, leading to parents' denial of the condition.^{6,7}

An effective and sustainable electronic population health record is also key to sustainability. Developments in immunisation and care information technology platforms, such as the BID Initiative in Tanzania and Zambia (funded by the Seattle-based PATH programme), could provide a national record system for all newborn babies and their mothers and be joined with other programmes, bypassing the multiple record systems in place in many highly resourced countries. A joined-up and easily accessible health record is important for completeness and timeliness of testing and follow-up of affected infants. Nnodu and colleagues report that, of 55 babies and infants younger than 9 months identified with sickle cell disease, 41 (75%) affected infants were enrolled into a care programme for free folic acid and penicillin, 36 (65%) were followed up for 9 months, and 29 (53%) agreed to continue to attend specialist care within the paediatric sickle cell programme of the teaching hospital. Primary care is likely to be the most realistic way to achieve large-scale access to care, although substantial education and training of frontline staff will be needed. Additionally, the stigma attached to this genetic condition highlights



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the importance of public messaging to support acceptance of the condition.

Moving forward requires clinicians and scientists working in this field to recognise that achieving sustainable, scalable, and impactful regional and national programmes in the African context will need us to draw on experience, expertise, and skills from a wide range of players, particularly of those who have delivered whole-population programmes, built wide consensus, and leveraged large-scale economies of scale for testing. Finally, since the arrival of an increased migrant population in Europe from Africa, development of consensus and wider collaboration on the approach to introduction of effective newborn screening for sickle cell disease in Europe is a model that can support the emerging African vision of collaboration for service development as distinct from western-led research collaborations.^{9,10}

AS declares no competing interests.

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