

## Mitigating the effect of the COVID-19 pandemic on sickle cell disease services in African countries



An estimated 5% of the world's population are carriers for trait genes for haemoglobin disorders, mainly sickle cell disease. Over 75% of the global burden of sickle cell anaemia occurs in sub-Saharan Africa, and sickle cell disease is the most prevalent inherited genetic disease with 10–45% of the population carrying the sickle cell gene. Advances in sickle cell disease management, including screening of newborn babies, pneumococcal prophylaxis, and hydroxyurea therapy have transformed sickle cell disease from a fatal childhood disease to a chronic condition of adulthood. Sickle cell disease remains a neglected tropical disease in Africa, characterised by a scarcity of specialist services and resultant high mortality for children younger than 5 years.<sup>1</sup> Furthermore, few African countries have epidemiological data on disease burden, which poses an additional barrier to effective health systems planning. This situation is compounded by the additional burden of the coronavirus disease 2019 (COVID-19) pandemic on existing resources.

As of April 15, 2020, 1 914 916 cases of COVID-19 have been confirmed and 123 010 people have died globally.<sup>2</sup> Increasingly, cases on the African continent are being observed; as of April 14, 2020, 10 759 COVID-19 cases have been confirmed in 45 countries with 520 deaths reported.<sup>3</sup> Many people are concerned about the burden that the COVID-19 pandemic will place on health-care systems generally and on specialist services particularly, including those for sickle cell disease.

Infection with severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) has shown different clinical courses across age groups.<sup>4</sup> Modelling of fatality in the early course of the outbreak from a preprint study showed case fatality ratios (CFR) increasing from 1.3% for people aged 50–59 years to 9.8% in those aged 70–79 years.<sup>5</sup> The increased CFR with older age has been attributed to increased comorbidities in these age groups, including pre-existing immunosuppression, diabetes, HIV, chronic obstructive pulmonary disease, and cardiovascular, liver, and renal conditions.

That a very different epidemic pattern for COVID-19 will be seen in Africa is possible. The demographics of many sub-Saharan countries are heavily skewed towards younger populations, suggesting potential for a milder

disease course than countries with a high proportion of older age groups; however, this might not always be the case, especially for countries with high burdens of pre-existing health conditions such as sickle cell disease. The ability of these often fragile health-care systems to manage COVID-19 has been discussed.<sup>6</sup> However, that many people younger than 50 years on the African continent live with potentially important comorbidities (eg, HIV, hypertension, and sickle cell disease), has not been discussed.<sup>7</sup>

Acute respiratory illnesses are a major cause of morbidity and mortality in patients with sickle cell disease who have an increased risk of asthma, pneumonia, and pulmonary vaso-occlusive disease, including acute chest syndrome. COVID-19 pathology is associated with reduced cellular oxygenation, pneumonia, and acute respiratory distress syndrome in severe cases. This pathological process is closely linked to an increased risk of vaso-occlusive complications in sickle cell disease, including acute chest syndrome. Specific diagnostic, treatment, and logistical challenges exist in meeting the health-care needs of people with sickle cell disease during the COVID-19 pandemic.

The influenza A H1N1 pandemic in 2009 highlighted increased susceptibility of children with sickle cell disease to severe complications of a respiratory virus, with an up to 50% increase in hospitalisation for this patient population.<sup>8</sup> Vaccination of people with sickle cell disease for influenza is a key intervention to prevent severe morbidity.<sup>9</sup> For SARS-CoV-2, people do not have previous immunity, and the development of a vaccine is many months away. An outbreak of COVID-19 in regions with a high prevalence of sickle cell disease will rapidly overwhelm existing resource-limited health-care services. These health-care systems should anticipate and prepare for a steep increase in young patients requiring high-intensity supportive care.

The risk of sickle cell disease complications and associated mortality can be mitigated through behavioural and pharmacological interventions. High-risk populations, including people with sickle cell disease, must be considered in the development of local preventative and health-care strategies. A crucial first step



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is to identify people who are at risk through screening. Uganda has shown that identifying people with sickle cell disease can be done with targeted screening;<sup>10</sup> however, these programmes need widespread adoption to identify the true numbers of people with sickle cell disease across the continent. Diagnostic and treatment resources should be rapidly scaled up in conjunction with investment in public health campaigns targeted to the at-risk sickle cell disease population. When an effective and affordable SARS-CoV-2 vaccine becomes available, people with sickle cell disease should be prioritised for immunisation because of their increased risk of mortality. This immunisation should be aligned with routine pneumococcal vaccination, which has proven to be effective.

The current focus on the COVID-19 pandemic in Africa and resource mobilisation provides unique opportunities for optimising health services for people with sickle cell disease in Africa and improving surveillance. The overlap in the epidemiology and clinical association of sickle cell disease with malaria, and bacterial and viral infections (including SARS-CoV-2), suggests that sickle cell disease should be included in the Integrated Management of Childhood Illness programme to improve outcomes. Provision for sickle cell disease diagnosis and treatment should be incorporated into national health systems programming, with an emphasis on delivering sickle cell disease services in the primary care setting. COVID-19 is expected to herald a global economic recession that might result in a contraction of international funding for health systems development in Africa.

The COVID-19 pandemic is currently overwhelming health systems in high-income countries and so could have an increased effect in low-resource settings in Africa where health services are already overstretched. The effect of COVID-19 on the global economy could cause recession both in Africa and around the world, posing a substantial threat to the delivery of health care in Africa. Every effort should be made to invest in primary health care, and integrate and align sickle cell disease diagnosis and treatment into existing health systems, rather than building new vertical programmes focusing only on sickle cell disease with interventions delivered separately from other health services.

African governments should leverage further funding resources to accomplish this aim. The preparation and mitigation stages of the pandemic also represent important opportunities to focus international efforts at rapidly scaling up sickle cell disease-related health-care infrastructure.

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