SICKLE CELL DISEASE - GETTING THE ACT RIGHT

Guest Editorial
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Since the first description of sickle cell disease (SCD) in 1910 in the United States of America (USA) in a young man of African descent [1], a lot of effort was dedicated to understanding the disorder such that within 40 years of its first description, its epidemiology and pathophysiology had been well described [2,3]. Despite this knowledge, it did not immediately translate into improved identification of and care for persons with SCD. This led to Ranney to remark in her 1972 editorial; “The fact that the biochemical abnormality and its pathological consequences are so much better delineated in sickling than in most other serious diseases has led to the notion that the most notable deficit in our care of sicklers is in the delayed application of our knowledge to the treatment of patients with sickle cell disease and to the detection of individuals with sickle cell trait.” [4].

As of 2019, Zambia is still grappling with the issues of diagnosis of the condition. We still have individuals being diagnosed with sickle cell anaemia well into their adulthood and we still do not know the prevalence of the sickle cell gene in our population. Twenty years ago, it was understandable to have a delayed diagnosis of sickle cell disease because the diagnosis required laboratory facilities. One needed at least a microscope or a centrifuge and a proficient laboratory person to diagnose the disease. Today, the development of easy to use point of care test kits for sickle cell diagnosis makes it inexcusable for one to be diagnosed with the condition in adulthood let alone in late childhood.

The World Health Organization (WHO) estimates that 5% of the global population carries the trait genes for sickle cell disease or thalassaemia, the major haemoglobin disorders and can be as high as 25% in some regions. It is further estimated that over 300,000 babies are born each year with a severe disorder of haemoglobin of which 200,000 are sickle cell disease [5]. Sickle cell disease is the major haemoglobin disorder in Africa while thalassaemia predominates in the Mediterranean and Asia. Many children with severe forms of sickle cell disease die before the age of 5 years either from severe anaemia, malaria and invasive bacterial infection [6].

Having realised that SCD is a major health burden on the African region, the World Health Organization adopted a strategy that aims to reduce the incidence, morbidity and mortality due to SCD [7]. Core to this strategy is i) Understanding the prevalence of the sickle cell gene in the population, ii) Early identification of persons with SCD and iii) Early interventions for persons with SCD to reduce morbidity and mortality.

We have a functioning system for transportation of dried blood spot (DBS) specimens for early infant diagnosis of HIV infection. We can leverage on this system to transport samples for infants and children requiring laboratory confirmation of the sickle cell status. It has been shown that DBS specimens can be used for haemoglobin (Hb) electrophoresis [9]

We have several health facilities that have the equipment for Hb electrophoresis such as UTH Adult, Ndola Teaching Hospital and Arthur Davison Children’s Hospital. These could be used as initial hubs for the SCD programme. Skilled laboratory personnel capable of doing Hb electrophoresis and quantification are there.

We have knowledgeable healthcare providers capable of providing comprehensive healthcare to persons living with SCD. This is evidenced by the establishment of specialized sickle cell clinics in facilities like ADH and Livingstone Teaching Hospital.

In her speech to the Minister of Health on World Sickle Cell Day, Lwimba Kasongo, a person living with SCD, made a passionate appeal for the institution of comprehensive healthcare services for persons living with SCD at all health care levels.

In his response, the Minister of Health represented by the Permanent Secretary – Technical Services, Dr Kennedy Malama, assured the SCD community that the condition was being given priority. He tasked technical experts led by Dr Catherine Chunda to come up with a Zambian Strategy for SCD.

We have easy to use point of care tests such as the SickleSCAN® test kit which has been evaluated for use in Zambia. This point of care device can be used by any healthcare provider to identify persons with the sickle cell gene or SCD [8]. With this kit we can then screen all antenatal mothers for the SCD. We can use this kit to screen all children presenting with anaemia at any health facility. In so doing we would be i) Generating information about the prevalence of the sickle cell gene in the Zambian population. We have health informatics systems that can be used to manage this data and feed into the overall Zambian strategy on SCD. ii) Identifying newborns that would require further testing for SCD, iii) Identifying children with SCD and intervening early.

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Whilst we may not have all the resources to implement a fully functional “well oiled” SCD programme, we do however have enough resources to implement ac-

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tivities that will have a positive impact on our knowledge of the condition in our population, on the reduction of childhood mortality from SCD and the quality of life persons living with SCD.

IT'S JUST A MATTER OF GETTING THE ACT RIGHT


