ABSTRACT

Background/objective: Sickle cell disease (SCD) is a monogenic disease with multiple phenotypic expressions. Previous studies describing SCD clinical phenotypes in Nigeria were localized, with limited data, hence the need to understand how SCD varies across Nigeria. Method: The Sickle Pan African Research Consortium (SPARCO) with a hub in Tanzania and collaborative sites in Tanzania, Ghana and Nigeria, is establishing a single patient-consented electronic database with a target of 13,000 SCD patients. In collaboration with the Sickle Cell Support Society of Nigeria, 20 hospitals, with paediatric and adult SCD clinics, are participating in patient recruitment. Demographic and clinical information, collected with uniform case report forms, were entered into Excel spreadsheets and uploaded into Research Electronic Data Capture software by trained data clerks and frequency tables generated. Result: Data were available on 3622 patients enrolled in the database, comprising 1889 (52.9%) females and 1434 (39.6%) children ≤15 years. The frequencies of Hb SS, Hb SC and Hb Sβ thalassemia in this data set were 97.5%, 2.5% and 0% respectively. Sixty percent, 23.8%, 5.9%, 4.8% and 2.5% have had bone pain crisis, dactylitis, acute chest syndrome, priapism and stroke respectively. The most frequent chronic complications were: leg ulcers (6.5%), avascular necrosis of bone (6.0%), renal (6.3%) and pulmonary hypertension (1.1%). Only 13.2% had been hospitalized while 67.5% had received blood transfusion. Conclusion: These data on the spectrum of clinical phenotypes of SCD are useful for planning, improving the management of SCD across Nigeria and provide a foundation for genomic research on SCD.

Keywords: Clinical phenotypes; Haemoglobinopathy registry; Nigeria; Sickle cell disease.