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PB2215 GLOBAL EPIDEMIOLOGY OF SICKLE CELL DISEASE: A SYSTEMATIC LITERATURE REVIEW

Topic: 26. Sickle cell disease

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Background: Sickle cell disease (SCD) has a high prevalence and social impact worldwide, with a high mortality within the first three years of life. A high prevalence has been reported in sub-Saharan Africa, the Mediterranean, the Middle East, and India. With global migration, prevalence is also increasing in other geographic regions, but there are no definitive data on global prevalence and mortality of SCD.

Aims: To perform a systematic literature review (SLR) of available evidence on the epidemiological burden (prevalence, birth prevalence and mortality/life expectancy) of SCD.

Methods: A comprehensive literature search of bibliographic databases and proceedings from key conferences was performed to identify peer-reviewed studies reporting on SCD epidemiology, from 1 January 2010, to 30 December 2020 (congress abstracts 2018 to 2020 only). The SLR protocol followed PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. Two independent reviewers screened titles and abstracts based on predefined inclusion and exclusion criteria, including reporting of relevant population data and at least one outcome of interest. Multiple data elements were extracted, including number of people screened and diagnosed, prevalence, birth prevalence, mortality/survival/life expectancy, geographic region, years of data collection, genotype and age category. Study quality was assessed with a version of the GRADE (Grading of Recommendations, Assessment, Development and Evaluations) framework, adapted to observational studies. Meta-analyses using a binomial normal random-effects model were carried out to estimate the prevalence and birth prevalence globally and regionally (when sufficient data were available).

Results: Of 1581 journal articles and 482 abstracts screened, 115 publications met the inclusion criteria; 43 included data on birth prevalence, 24 on prevalence and 33 on mortality or life expectancy.

Birth prevalence of SCD (children ≤1 year old), mostly measured through new-born screening, was highest in several sub-Saharan Africa countries (500–2000/100,000), South America and Caribbean Islands (20-1000/100,000); in USA and European countries found in the SLR, birth prevalence was ≤500/100,000.

Overall findings on regional prevalence indicated that sub-Saharan and North-East Africa, India and the Middle East are global SCD “hotspots”. However, prevalence data were too sparse to provide consistent prevalence numbers per region through meta-analysis.

Mortality appeared particularly high in low-resource settings, including Africa and India; although, different methods of data presentation and source populations make comparisons across studies difficult.

There are limitations to this SLR due to gaps in the published literature, including inconsistent reporting of SCD genotypes, diagnosis criteria and study settings. In addition, there was paucity of peer-reviewed publications on life expectancy and prevalence from European countries with assumed high prevalence. A search of global and local registries is ongoing to supplement the published literature.

Summary/Conclusion: This SLR has provided some insights into the global epidemiology of SCD, confirming earlier studies of areas with a high prevalence. However, this SLR confirms that there is a great need for resources for additional studies with uniform data collection on prevalence and mortality to ensure an increased awareness of SCD among public health policy makers worldwide.