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Clinical Severity of Sickle Cell Anaemia in Children in the Gambia: A Cross-Sectional Study

Background: Sickle cell anaemia (SCA) in children demonstrates a broad range of clinical manifestations and serious complications. Assessment of disease severity in specific populations is necessary to plan services and optimise care.
Aim: To describe the clinical severity of SCA and associated sociodemographic and clinical factors in children in Gambia.
Methods: The presence of lifetime complications was confirmed by history and review of medical charts. We determined clinical severity using a validated scoring system and related the severity to sociodemographic and clinical factors.
Results: In 130 study participants, ages ranged from 5 to 15 years with a mean (SD) age of 9.74 (2.81) years. Eleven (8.5%) children had had acute chest syndrome, 7 (5.4%) avascular necrosis of the femoral head, 6 (4.6%) gallstones, 5 (3.8%) stroke and 1 (0.8%) priapism over their lifetime. Disease severity was classified as mild in 108 (83.1%) children, moderate in 17 (13.1%) and severe in 5 (3.8%). Age, age at diagnosis, sex, ethnicity, social class, and treatment with hydroxyurea was not significantly correlated with SCA clinical severity (P values 0.10-0.84).
Conclusion: The high proportion of children with mild disease may be due to the high prevalence of Senegalese ?-haemoglobin haplotype in the Senegambia subregion. However, the presence of moderate or severe disease in almost 1 in 5 children calls for concerted efforts in SCD care in this region.