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Sickle cell disease among children in Africa: An integrative literature review and global recommendations

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ABSTRACT

Sickle cell disease (SCD) is a genetic blood disorder affecting red blood cells, with high morbidity and mortality rates. The United Nations has recognized SCD as a global public health concern, and the World Health Organization (WHO) recommends that 50% of member states will have established SCD control programs by 2020 (World Health Organization, 2006).

This paper presents an integrative review of 63 references related to SCD among children less than 18 years of age in Africa, published between 2000 and 2015. The review focuses on the incidence, prevalence, morbidity, and mortality; current practices and challenges related to screening, diagnosis, and treatment; and recommendations for practice, policy, and research to improve health outcomes of children with SCD in Africa.

There have been significant improvements in the morbidity and mortality rates for children with SCD in high resource countries such as the United States due to factors such as early diagnosis through newborn screening programs, prophylactic therapy, comprehensive care programs including hydroxyurea therapy, and bone marrow transplant. Many of these interventions can confer the same benefits to SCD patients in Africa. Newborn screening for SCD, developing partnerships between high resource countries and countries in Africa to support training of healthcare workers, research, and sharing of knowledge can help to reduce the SCD burden in Africa.

Contents

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1. Introduction

Sickle cell disease (SCD) is a genetic blood disorder affecting red blood cells, with high morbidity and mortality rates. Sickle haemoglobin (HbS) is a structural variant of normal adult haemoglobin (HbA) (Chakravorty & Williams, 2015). SCD includes a series of pathological genotypes resulting from the inheritance of HbS. SCD affects 20–25 million people globally, and 50–80% of infants born with SCD in Africa die before the age of 5 years (Aygun & Odame, 2012). It is estimated that 240,000 children are born with SCD annually in sub-Saharan Africa (Makani et al., 2011).

The United Nations General Assembly has recognized SCD as a global public health concern due to the morbidity and mortality caused by the disease and the significant social and economic impact that results (United Nations General Assembly, 2009). The purpose of this paper is to review the literature related to SCD among children less than 18 years of age in Africa. The review focuses on the incidence, prevalence, morbidity, and mortality of SCD among children in Africa; current practices and challenges related to screening, diagnosis, and treatment; and recommendations for practice, policy, and research to improve health outcomes of children with SCD in Africa based on the literature and on global guidelines.

The integrative review method proposed by Whittemore and Knafl (2005) guided the process used for this review. This method consists of five steps: (a) identification of the problem; (b) identification of search strategies; (c) evaluation of data quality; (d) data analysis; and (e) synthesis and presentation of the data. The inclusion criteria included full-text reports, web pages, or articles (research-based or descriptions of existing programs or policies), published between 2000 and 2015, and focused on SCD prevalence, morbidity, mortality, screening, treatment, and/or policy guidelines for children less than 18 years of age in Africa. Data-bases included in the initial search included Medline, CINAHL, PubMed, and Google Scholar, using the key words: "Sickle Cell Disease (SCD)", "Sickle Cell Anemia (SCA)", "SCA/SCD Screening, Treatment, and Policies," cross-listed with keywords "Children" and "Africa." Additional sources were identified by examining reference lists of each paper that was reviewed. A total of 63 references were identified that met inclusion criteria.

After the articles were retrieved, their strengths and limitations were assessed by both co-authors, and key concepts were identified. This paper presents a synthesis of the articles reviewed, and a discussion of implications of the findings for practice, policy, and future research. The articles were analyzed and grouped according to the following categories to facilitate the final synthesis: epidemiology, prevalence, and global health significance; mortality rates; morbidity; genetic counseling and newborn screening; management; and recommendations for policy, research, and global partnerships.

2. Epidemiology

Of the 330,000 babies born with a major hemoglobinopathy worldwide, 275,000 have SCD, making it the major global hemoglobinopathy (Aygun & Odame, 2012; Modell & Darlison, 2008; Weatherall, 2011). SCD patients in the developed world account for only 10% of the world's SCD patient population (Aygun & Odame, 2012). In 2008, Aliyu et al. (2008) reported United Nations estimates that there are between 20 and 25 million people worldwide living with SCD, of which 12–15 million live in Africa. It is estimated that 75–85% of children born with SCD are born in Africa, where mortality rates for those under age 5 range from 50% to 80% (Aygun & Odame, 2012; Makani et al., 2011).

The highest prevalence of sickle-cell trait (SCT) in Africa occurs between the latitudes of 15° North and 20° south, where the prevalence ranges between 10% and 40% of the population (Agasa et al., 2010). In 2010, Rawezula reported results of a study of records of over 2000 newborns at a hospital in Tanzania (Rwezaula, 2010). Findings indicated that 18.2% of the neonates had abnormal hemoglobin levels and that the incidence of abnormal hemoglobin levels differed based on the geographical regions of the newborns' parents. The incidence of SCT among infants whose parents were from the coastal areas was 35.6%, compared with 6.7% for infants whose parents were from the northern region.

The incidence of the SCT in Cameroon, the Democratic Republic of Congo, Gabon, Ghana, and Nigeria ranges from 20% to 30%, and in some parts of Uganda, the prevalence is 45% (Afolayan & Jolayemi, 2011; Agasa et al., 2010; Anie, Egunjobi, & Akinyanju, 2010; Serjeant & Ndugwa, 2004; World Health Organization, 2006). Chakravorty and Williams (2015) suggested that there are few places where the carrier rate for SCD is greater than 25% because of the disadvantages conferred by homozygosity.

Because many births occur outside of hospitals and many children die before diagnosis with SCD, there are limited statistical data on the incidence of SCD in Africa (Serjeant, 2010; Serjeant & Ndugwa, 2004). There is a need for additional research to gather accurate data about SCD prevalence in order to direct appropriate health care policies (Munyanganizi, Cotton, Vertongen, & Gulbis, 2006; Odunvbun, Okolo, & Rahimy, 2008). One approach to generating more accurate data about the incidence and prevalence of SCT and SCD was reported in 2010 by Piel et al. (2010), who developed a geo-statistical mapping model based on the frequency of the hemoglobin S (HBS) allele and population data.

It has been suggested that one factor associated with the high incidence of SCD in tropical Africa is the protection against Plasmodium malaria associated with having the SCT (Aygun & Odame, 2012). However, that protection seems not to extend to people with SCD (Komba et al., 2009; Rahimy et al., 2003). The theory that the SCT offers some immunity against the malaria parasite was found to be strong in tropical Africa through a geostatistical mapping study, but could not be explained in other parts of the world (Aidoo et al., 2002; Piel et al., 2010). Migration from Africa and other regions with high rates of SCT and SCD has contributed to the global spread of SCD to areas such as North America and the Caribbean (Weatherall, 2011; Wonkam et al., 2011).

3. Mortality rates

Mortality rates associated with SCD vary widely across the globe. Children born in high-resource countries with major