



# Sickle Cell Disease Control Prospects the Emerging Necessities Due to Impactful COVID-19 Pandemic in Sub Saharan Africa

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## Review Article

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## Abstract

**Background:** People with sickle cell disease (SCD) seek health care services quite often as the condition is associated with high morbidity and mortality. The emergence of the COVID-19 pandemic has imposed diversion of health care attentions and resources from the usual demands. COVID-19 cases have occupied health care facilities, providers as well as care planners. This has affected the already dire state of SCD management in Sub Sahara Africa (SSA). It is necessary to look at options that have been examined and demonstrated to reduce the prevalence of SCD. The purpose of this review is to present prospects and methods that could be useful in reducing prevalence of SCD.

**Methodology:** The aims of the review were to document and discuss the methods that have been applied to reduce the prevalence of SCD so as to prospect for use in SSA. A review of the literature and targeting studies, presentations and information on control of SCD. The sources used were PubMed, PubMed Central, Google Scholar, and African Index Medicus. Bases of selection were any aspect of control of SCD by educational, counseling, awareness, prevention, testing and providing information. The prospects are explored, discussed and presented in this article.

**Keywords:** Education; Counseling; Sickle cell disease

## Introduction

Sickle Cell Disease is a genetic disease amenable to education and counseling as a means of control. In addition the SCD established prevalence in defined populations and geographical areas make it potential for considerations for control. Without treatment, which is inadequately available in low-income high-burden countries, it is observed that most children born with the disease die in their first years of life [1-3]. Currently coping with other diseases SCD included in the background of COVID 19 is far more demanding than could have been anticipated.

The SCD is the most prevalence hereditary illness in Sub Sahara Africa (SSA) [4-6]. Its clinical and pathological manifestations are due to homozygosity of the hemoglobin S (Sickle cell) gene [7-9]. Many SCD persons dependent on

constant attention provided by clinical health care teams. Clinical manifestations commonly occur at the age of 6–12 months [9-11]. The most common manifestations include painful occlusive events, haemolytic anaemia and end organ damage caused by vasculopathy and tissue ischemia [6,7,11-13]. Complications may be of sudden onset (sickle cell crisis), but a degree of sickling occurs most of the time, and it is this that leads to long-term organ damage. Sickle cell disease is a life-shortening condition although current interventions have contributed to patients living longer, often with attendant comorbidities [14].

## Burden of SCD

The United Nations General Assembly in 2009 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 [15,16]. Africa contributes between 75 to 85% of world's haemoglobinopathies cases where 6 to 9 million infants are born with SCD. The disease is associated with approximately 6.4% death rates of children in Africa in under 5 years old children. Of these about 70% of children are in Sub-Sahara Africa. While the HbSS is highly prevalent in areas of Sub-Saharan Africa, particularly West and East Africa [8-13] the prevalence of the carrier rates for HbS ranges from 5% to 40% among populations in these endemic areas [6,17-19].

Public health implications of sickle cell disease are significantly high morbidity and mortality impacting huge socio-economic burden on populations affected and the countries in the SSA [20-22].

### The Backdrop of COVID 19 Pandemic

The COVID 19 pandemic has diverted greater attention from other diseases; SCD included. Weak health care systems as observed in the SSA are worse off and persons with SCD have most certainly less attention than before the pandemic [14,23]. It is against this background that documented programmes that can reduce the prevalence of SCD is proposed for investment to reduce the overall burden of SCD in SSA [24,25].

### Control of SCD

**Counseling:** In SCD the purpose of counseling is to provide guidance for decision making process, regarding having children, helping to understand the disease SCD [26]. Genetic counseling helps to solve problems in the field of heredity, occurrence or risk of inheriting the disease [27,28]. Premarital counseling and testing is intended to reduce the prevalence of inheritable diseases such as SCD [29]. Importantly it has been shown to have a significant advantage over neonatal screening since it is aimed at primary prevention, the latter addresses secondary, or tertiary prevention [30]. It enables couples have prior knowledge of their SCD hemoglobin genotype and more chances of avoiding high-risk marriages [30]. Countries that have developed strategies and protocols for premarital counseling including Saudi Arabia, Greece, Cyprus, and Italy also have success stories with genetic counseling and testing [31-34]. However this required much support from the religious bodies to enforce premarital testing and counseling [35].

### Education

The role of education in SCD is to provide knowledge, information and resources to enable minimizing disease burden and health consequences [36]. It puts into context the physical, psychological, mental and social wellbeing [36-

38]. Aspects of education provide clinical information and explains modifiers of genetics, economic and environmental of those affected [39].

### Education and Counseling

Influence of health education and genetic counseling has been examined and are essential components of any SCD program [40]. The education contextualizes culture, religion, health beliefs and attitudes that have significant impact on outcomes of the burden of SCD [35]. Education both increases the returns to social connection and reduces the extent of religious belief. The positive effect of education on social connection is the result of both treatment and selection: schooling creates social skills and may increase people's utility from engaging in other social activities such as church attendance. The negative effect of education on religious belief occurs because secular education emphasizes secular beliefs that are at odds with many traditional religious views. As such education may define the roles of community leaders who substantially influence reproductive decisions made by individuals and families [9,41].

However genetic counseling has shown mixed results as demonstrated by a study in Bhoga, et al. that only one third of the parents practiced birth control hence limiting their family size [41]. Half of them carried on as before exposure to counseling a scenario replicated in other studies as well [28,41,42]. However in this cohorts the study brought out social issues as a result of sickle cell disease. One fifth of the 45 marriages ended in separation, divorce or the husband taking another wife [41,43,44]. It was also noted that parents that had one affected child with one or two normal healthy siblings, long crisis-free periods in the affected child, and a stable marriage took responsible decisions after the counseling [41,44]. On the other hand, the opposite was true for parents with more than one affected child, no surviving normal child, polygamy and a non-caring husband. These highlight the social issues surrounding sickle cell disease and the impact of social support [41]. The SCD congregates in ethnic groups and while this is a good marker of the populations at risk it opens room for forms of discrimination [45]. In addition intermarriage has created the abnormal Hb where it initial did not occur [46].

A study in Ghana advised the development of SCD materials as essential to the training program [47]. The training materials provided in different languages and factor in gender differences be considered. In addition recommend peer to peer education for prospective couples was advised and could be achieved within existing youth groups. This study further recommended that an educational policy for SCD information should be incorporated into the school curriculum and into training for teachers, health workers,

and genetic counselors [47].

### Education in Religious and Traditional Settings

Premarital SCD counseling to be widely acceptable to most communities should include, youth engagement, churches, mosques, and traditional issues [48,49]. In addition SCD programs to engage the participation of traditional leaders, such as chiefs and SCD [50] messaging at religious and special traditional events, such as confirmation, baptism, and puberty rites [50,51].

### Media and Entertainment

The role of the media in broadcasting SCD information has been discussed. Suggestions made of inclusion in favourite television programs and interactive theatre with role play; documentary movies; and animated cartoons. Popular presenters on radio stations also could be enlisted to improve awareness [47,50,52].

### Healthcare Provider Forums

This is to include clinical, health care facilities ensuring to introduce SCD during provision of health care such as in preconception care and prenatal diagnosis [53]. In adults sessions only cultural, customary and community dedicated rituals be part of the counseling process [54,55]. A study in Nigeria presented the importance of genetic counselor personal values in peoples interactions [56,40]. Traditionally, genetic counseling has been considered a “value neutral” profession, in which the provider’s beliefs or values should not infringe on patient autonomy. But values, which we may rely on even subconsciously, can impact “presentation of facts and options to patients , if and how they engage patients in consideration of the moral consequences of their decisions, and how they respond to ethically challenging situations [57]. Because of this reality, genetic counseling cannot truly be considered a value-neutral or culture-neutral profession [58,59].

### Discussion

This review has been compelled by the current situation particularly in the SSA that is faced with high prevalence of SCD and the effect of the COVID-19 pandemic [57,60,61]. The central thesis is that primary control of SCD would reduce the numbers of SCD and free health care providers to other emerging health care needs. There are published show case results of studies on the control SCD that could be considered to control SCD prevalence [62].

The COVID-19 pandemic has brought into focus the needs to have common action paths ,knowledge and

established concerns of what could be done or options on health in general [62,63]. For those who already have disease conditions and possibilities of getting another, the reasons for preparedness is clearly necessary. The usual clinic, admission attention may just not be available in situations such as that imposed by the COVID 19 pandemic [62,64]. The review demonstrates evidence from studies, surveys, and observations on the control of SCD. The impact in the control have shown mixed results. Since we propose these methods for Sub-Saharan Africa many of the cited studies are from Africa. Few were from India, USA and Europe [25,65,66].

Advanced countries with significant population of patients with SCD have evolved functional genetic and counseling protocols, with remarkable reduction on disease prevalence [67]. This feat however does not appear to be replicable in a number of SAA countries. In the SSA genetic understanding and screening is not a common practice and the diagnosis is usually made SCD cases present with a severe complication [68]. Even when tragedies such as two or more miscarriages, still births, or children die in infancy, many at times health care providers do not consider genetic counseling [14,35].

In some of the countries in SSA with huge burden of SCD such as Nigeria utilization of genetic counseling and testing SCD is not common [9,69,70]. Lack of this counseling not only adversely affect health policy planning but also continue to fuel the already high carriage rate of the S gene [14,41,44,69]. Clearly a formal educational counseling delivery of control would be preferred to urgently address SCD control [28,42].

Education of the populace to increase the understanding of SCD and its impact since the most important challenge is to raise the awareness on its causes and prevention through health education [21,71]. In many countries of SSA, no genetic counseling clinic is known as at the time of this review. In many developed countries, genetic testing and genetic counseling have been commonly done to identify carriers [72-74].

A strong premise of this body of work is that education is a cultural process [75,76]. Schools are the primary vehicle for transmitting knowledge and skills as well as the values, practices, and culture of a society [76]. In which case a form of curriculum developed to encompass all the requirements and avoid eminent challenges [77]. Mounting evidence demonstrates the benefits of creating an educational environment that is relevant to and reflective of realities, background, and culture [77,78] discussed examples of successful programs. This research shows that cultural and ethnic identity mitigate negative experiences, increasing self-confidence, self-esteem, and resiliency among both children and adults [77]. At the collective level, culture is related to

the survival of distinct practices and also the functioning of social and family networks and support systems that may contribute to internal sustainability and vitality of social groups [77,79,80]. Many areas of human service have capitalized on these inherent benefits by integrating culturally specific practices or approaches into the delivery of health, social work, counseling, and other services through formal education [76,80-82]. What may be less obvious is that all educational systems and institutions are rooted in a particular cultural worldview [81,82].

Counseling about a genetically transmitted condition consists of providing accurate information that responds not only to the preceding questions but to all questions, both verbalized and implied, that relate to the effect of that condition on the individual's physical and emotional well-being and his present and potential family [42,83].

Counseling be part of the education program as an essential components of any SCD control model to ensure that results are effectively communicated by trained healthcare workers to those affected and their families [79]. These are also important within the context of culture and religion because health beliefs and attitudes of the general public have a significant impact on outcomes, and community leaders substantially influence reproductive decisions made by individuals and families [41,49,84].

An educational program would have requirements for mitigating counseling limitations as illustrated in a study in Ghana [37,85]. Counseling for sickle cell be approached as a process of basic education or giving information. Training needs of counselors and counselees would have to be in such an educational curriculum [42].

This was demonstrated by a workshop for a sickle cell genetic counselor training in Ghana [37]. Knowledge and education of the general public about SCD with special emphasis on the youth appeared to be essential [40]. Public health education improved knowledge and SCD screening and counseling uptake among them. Areas of emphasis would be stressed and those considered adverse mitigated [40]. As some studies have shown that regardless of their SCD or SCT status, women generally placed a high value on motherhood and their ability to have children, and this could be influenced by culture and religious beliefs [34,86].

Tackling stigmatization that is quite common in African communities where patrimonial societies tend to blame mothers for their child's ill health. A study in rural Kenya revealed that fathers attributed "bad spirits (pepo mchafu) [85] that comes to attack the child" due to mothers, who then became economically disadvantaged as a result of their child's SCD [87-89]. A societal education could be a way of

ameliorating such notions as it dedicate sessions to improve understanding of the impacts of societal context [41,90-92]. Education about SCD and interventions to address underlying economic and social disparities could potentially alleviate stigmatization and negative misconceptions [92-94].

The timing of education is important for uptake. Such as the screening of couples at risk of producing children with serious SCD. The Virginia Sickle Cell Anemia Awareness Program began identifying such couples as early as 1970 [44]. Ideally, a screening program to inform people about childbearing risks should be performed well in advance of childbearing age, or at least prior to choosing mates [28,41]. This has bearing in timing and may also vary from one community to the other, factors that programs should incorporate during the designing [35,38,95].

The SSA communities have varied cultural customs and ways of perception of inherited conditions such as SCD. This makes it challenging to counseling alone [43,94]. To these, formal education with curriculum for regional and national objectives may work compared to community based education [43,96]. However it is imperative to understand the community unique cultural, social, customary and economic drivers that be factored in the delivery of such a curriculum [96].

Genetic counseling for SCD enjoys huge acceptance in some situations and through education such communities could be identified and provisions made. Consistent application of genetic counseling coupled with the establishment of permanent centers for this is advocated to be vital in reducing the prevalence of SCD as evidenced in the Indian population [64]. While in Nigeria's study, a three and a half year follow up of 45 families with at least one affected child from Ibadan community to investigate the effectiveness of counseling both general and genetic ,most parents sought advice for problems concerning their affected child, his management, education and future prospect in marriage [79]. One great fear is of recurrence and marital problems caused by the child's condition need to be sorted out by general curriculum based education [30,44].

In SSA with remarkable paucity of competencies counseling about a genetically transmitted condition would not meet the desired requirement [97,98]. Furthermore education allows for appreciating and understanding a culture unique to a particular national group. As in this case the SSA inhabitants of each country have their own language, beliefs, health system, style of government, values, mode of dress, communication network, and manifest a variety of behaviors that set them apart from people in other nations [38]. These wide diversities can be captured in a joint curriculum for educating the inhabitants of the SSA [58].

## Challenges

The two general major ways of controlling SCD in this review are counseling and education [53]. Notables in both methods are or insignificant content and context with regards to culture, customs, social, and environment of the communities in the reviewed materials. We hypothesize that the minimal impact documented therefore was related to little attention given to these aspects of life that influence attitude, practice and knowledge of a people. Many of these also delivered as consisting of giving pieces of advice. While at individual level their influence may be evident but at community and regional level the influence may not be substantial. On the other hand if education and counseling are implemented in a structured format and delivered within an educational system the impact would be enhanced.

Inadequate attention to the facts that the SSA is largely patriarchal and there would be need to institutionalize SCD control within the health education system rather than counseling usually to individuals, families or small groups representing communities. We believe that these are challenges due to targeting specific populations. A national curriculum based education rather than individual counseling would be more productive than selecting a focus group. The SSA has religious and spiritual heritage as such people naturally are sensitive to intrusions by different religious and spiritual traditions. Education may go to some length communicating interest, understanding, and respect to those who have spiritual beliefs that are different from their own. They seek to learn more about the spiritual beliefs and cultures of clients with whom they work [53]. It is not realistic to assume that a counselor can possibly assimilate both the quantity and quality of theological information about religious systems to hold court with their clients all the time. Rather education about the basics of faith communities allows a bridge from which to illicit and draw out the experience with dignity and genuine respect for diversity [99].

## Conclusion

This review portrays education and counseling as the main prospects for the control of sickle cell disease. It is considered that method of delivery be a formal education curriculum that captures most of the life governance of the SSA such as social, cultural, economic and spiritual contexts and diversity of the populations. It is incumbent on us to be prepared as a region to urgently address these prospects to reduce the prevalence of SCD.

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