PERSPECTIVE

Functional meals offer prospective therapies for sickle cell patients in Nigerian children

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ABSTRACT

One of the structural hemoglobinopathies, Sickle Cell Disease (SCD), also known as Sickle Cell Anaemia (SCA), is caused by a single nucleotide shift from GAG to GTG, which converts glutamate into valine in the amino acid of a -globin chain of Haemoglobin (Hb). Red Blood Cells (RBCs) with this unique mutation become disordered due to changes in their shape and other clinical circumstances. The only treatment options for SCA in the 1980s were occasional red blood cell transfusions, painkillers, and penicillin prophylaxis, which were frequently saved for serious, life-threatening consequences. Four different medications have now been approved by the US Food and Drug Administration (FDA) for use in the prevention and treatment of SCD clinical problems. The present study discusses the therapeutic effects of readily available functional food as one of the therapies or an adjunct therapy to address the sickle cell crisis in Nigerian children due to limitations (adherence, safety, adverse effects) of existing therapies in the prophylaxis and treatment of SCD complications in Nigerian children as well as their inaccessibility to approved drugs.

Key Words: *Hemoglobinopathies; Sickle cell disease; Single nucleotide shift; Glutamate; Red blood cells*

INTRODUCTION

A collection of hereditary disorders affecting the red blood cell is known as Sickle Cell Disease (SCD). The combination of sickle thalassemia and sickle cell anaemia is frequently referred to as drepanocytosis (HbSS). Sickle cell carriers with heterozygous genotypes (HbAS) have offspring with a 25% probability of inheriting a homozygous sickle genotype (HbSS) or a 75% homozygous normal genotype (HbAA). Herrick classified sickle cell disorder as a haematological illness in 1910, and Linus Pauling established its biochemical pathology in 1949. Epidemiological data on SCD has primarily been documented among the populations of Jamaica, India, and the US, with less figures in Africa, where the trait of SCD is thought to be most prevalent. It is especially prevalent among the black people of Africa and several other races that live in the Middle East, India, and the Mediterranean. Additional research on SCD in children has shown that the majority of cases occur in developing nations, with an estimated 200,000 sickle genotype births every year in Sub-Saharan Africa. One of the leading causes of infant death in Africa is sickle cell disease. 80% of the world's SCD patients reside in Sub-Saharan Africa By 2050, 88% of this percentage is predicted to exist. With over 150,000 births per year and a mortality rate of 50%–90% before the age of five, Nigeria has the highest burden of sickle cell disease in both Africa and the globe. Excess mortality is a result of inadequate medical care, inadequate education, ignorance, poverty, and poor nutritional decisions. For instance, historical and scientific documents from West Africa indicate many names that the three primary Nigerian tribes use to identify SCD children. The Yorubas, who predominated in the west, referred to them as "abiku," which is Yoruba for "sufferers" or "children that bring grief." They were known as "Ogbanje" by the Ibos and "Sankara-miji" by the Hausas. It is the most common intrinsic condition in Nigeria, affecting roughly 4 million people at a prevalence of 2% at birth, although more than 40 million people exhibit features associated with sickle cell disease. About 75% of African children with SCD are found in Nigeria. Although they were normal weight at birth, babies with SCD experience weight loss over the first year, which gradually persists until maturity. This is coincidentally followed by a delay in both males' and girls' skeletal maturation, as well as a delayed menstrual cycle in girls. When the gene encoding the human -globin component experiences a single nucleotide base change, the hereditary sickle cell derangement is made visible at the molecular and genetic levels. This substitution results in the Aberrant Haemoglobin (HbA) replacing the hydrophilic amino acid (glutamic acid) at position 6 with a hydrophobic amino acid (valine), which leads to the illness state of sickle red blood cells. When deoxygenated, the hydrophobic substitution that has taken place makes Sickle Cell Haemoglobin (HbS) insoluble. The continuously forming HbS molecules polymerize to long, crystalline intracellular masses of fibres, which

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change the erythrocyte's initial biconcave shape into a sickled shape cell. Hemolytic anaemia and tissue disruption caused by the sickled cells' obstruction of blood arteries highlight the magnitudes of this remodelling. Vascular necrosis, hyposthenuria, proliferative retinopathy, priapism, aplastic crises, pulmonary illness, and nephropathy are clinical manifestations of this syndrome. In the majority of patients, the effects are permanent and include recurrent pain attacks and progressive organ malfunction that shortens biologic life expectancy. Reactive oxygen species are also thought to be very important in the pathophysiology of SCD. The development of SCD-related organ problems may be significantly influenced by the chronically elevated oxidative stress in SCD. In terms of pharmacology, numerous medications have surfaced with various targeting routes. However, the bulk of them have not shown any effect in clinical studies, and the ones that have generated hopeful outcomes are less common, making them unavailable to low-income nations like Nigeria. Up to 80% of people in Africa rely on conventional plant-based medicine, which has few side effects, to meet their basic medical needs, according to WHO (2002). Almost every known culture in Nigeria has long used phytomedicine as efficient treatments for the prevention and treatment of a variety of health issues. According to studies, polyphenols, alkaloids, flavonoids, and other phenolic substances have antisickling and antioxidant actions that can help to normalise the abnormal redox equilibrium that causes sickle cell crisis in infants. Functional foods, or foods having bioactive substances that can boost a person's health beyond rudimentary diets or serve as a measure for prevention or management of chronic diseases, are typically used to describe such foods including polyphenols. However, the use of well-known medications and possibly curative procedures will largely be restricted to high-income nations that can afford to purchase them, leaving low-income nations who are suffering greatly due to the lack of these medicines out in the cold. In this review, we highlight the widely accessible special functional meals that can be utilised to help Nigerian children who are suffering from the sickle cell crises. Deoxygenated sickle cell red blood cells (RBC) have a sticky patch on their surface that attracts additional haemoglobin molecules, forming a polymer of mutant Sickle Cell Haemoglobin (HbS) molecules. The RBC membrane is then altered and deformed, resulting in sickling, by the polymers aggregating to form an insoluble, lengthy chain of hard fibre (bundle). The hydrophobicity of valine, according to a paper by Piccin and colleagues, results in this sickling because, during polymerization, it attracts hydrophobic areas of neighbouring -chains. This sickle-like characteristic is the distinguishing factor that triggers downstream processes such as hemolysis, vaso-occlusion, sterile inflammation, and sickle RBC symptoms .Studies from the past and the present have shown a connection between HbS poly-merization, poor blood rheology, hemolysis, and sickle RBCs' enhanced adhesiveness with inflammatory cells and vascular endothelium, as well as vaso-occlusion activation. Vaso-cellular occlusion's and molecular mode of action, however, is still unknown. Additionally, research has demonstrated that a variety of inflammatory and/or environmental factors, including TNF, hypoxia, heme, dehydration, Hb, infection, hypoxia, acidosis, lipopolysaccharide, and others, can cause blood vessels to occlude. According to reports, the autonomic nervous system and vaso-occlusion are linked or connected, and these inputs or triggers may result in painful events. It has also been explained that not all vascular beds or organs exhibit the samecellular and molecular paradigm of vaso-occlusion. Another crucial nutritional supplement for SCD crises may be walnuts, flaxseeds, and chia seeds. According to Daak and colleagues, omega-3 fatty acids found in flaxseeds, chia seeds, and walnuts significantly improve the general health of children with SCD by lowering vaso-occlusive crises. Its anti-adhesive, anti-aggregatory, vasodilatory, and anti-inflammatory characteristics are probably to blame for this. Polyunsaturated Fatty Acids, which include omega-3 and omega-6 fatty acids, are needed for the Red Blood Cell Membrane's (RBCM) synthesis and repair (PUFAs). Lack of PUFA-rich diets may result in problems with the RBCM's repair system, which would delay the maturity and development of the nervous system and various other sensory systems. Recent investigations have clarified how vascular thrombosis and inflammation caused by sickle vasculopathy contribute to the development of sickle cell problems. Omega - 3 fatty acid diet supplementation has recently been shown to regulate RBC membrane, boost NO generation, reduce inflammation, and inhibit adhesion molecules in sickle cell mouse investigations. Given the aforementioned characteristics, one can speculate that omega-3 fatty acids express or possess the special properties of crizanlizumab and L-glutamine. Thus, the probability of complications or severe symptoms of SCD and the degree of anaemia decrease with increasing blood levels of Omega-3 fatty acids. Since the beginning of SCD, numerous medicinal roots, leaves, and seeds that possess advantageous phytochemical components have been used to lessen unpleasant and sickling events. Antioxidant plant extracts that are also essential as antisickling agents include the leaf extracts of *Carica papaya*, *Parquetina nigrescens*, *Fagara zanthoxyloides*, and *Cajanus cajan*. Extracts from *Carica papaya* leaves have shown promise in *in vitro* studies for decreasing hemolysis and maintaining the integrity of the erythrocyte membrane.