Editorial

Disparities in sickle cell disease management: quest for global protective immunity

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The worldwide burden of sickle cell disease (SCD) is enormous and a major global health concern. The SCD is a congenital hematologic disorder characterized by deformed red blood cells, acute episodic attacks of pain and pulmonary compromise and widespread organ damage due to ischemia. Repeated episodes of ischemia lead to serious dysfunction of the spleen, which results in susceptibility to severe infection and sepsis. The pathogenesis of this chronic life-long inflammation is poorly understood and the SCD progression was a death sentence, most patients die at young age. Now, more than 90% live until adulthood, posing new issues and challenges.

In the current global political scenario, healthcares, and the quality of healthcare, are unequally distributed. Race, insurance status, socioeconomic status, gender, etc. are associated with unequal distributions of healthcare and disparities in research and medical treatment. In the United States, African-Americans are affected by a number of healthcare disparities. Unfortunately, those of us who work in the area of SCD are well-aware of the problem of disparities as SCD is not the disease of white people. There are disparities in: – Federal and private funding for research and medical care; the availability of comprehensive centers for healthcare and the quality of the interpersonal aspects of hospital care. Both disease-based and race-based discrimination negatively impact the overall outcome of SCD. Disease-based, but not race-based, discrimination appears to have further negative consequences on patient's self-efficacy in managing their disease and their adherence with doctor's advice.

SCD patients are frequently seen in emergency rooms or require admission for treatment of pain, infection, or other complications. Readmissions to hospitals are receiving enormous attention from Medicare and other payers; with hospitals being penalized for readmissions for specific diagnoses such as SCD. Treating SCD, with its frequent need for hospitalization; is quite expensive. For example, the cost of hospitalizations for treating acute pain alone is now estimated at -$1billion, according to Johns Hopkins hematologist Dr. John Strouse. Despite some advances in research and drug development for sickle cell, significant gaps remain in the translation of this research from bench to bedside, and a far more concerted effort by the public and government agencies are needed to address this major health disparity.

In past few years, a state of significant immune dysregulation has been observed with the progression of SCD. This progressive immune dysregulation impair the normal immune response in SCD patients which might pose an underlying threat to non-sickle healthy human populations. Several studies in past decade have indicated the more health complications and deaths in SCD patients from cancer and infectious diseases. Our lab also identify such a mechanism of immune-dysregulation where progressive loss of major Histocompatibility complex II correlate with the high susceptibility for infections and early death in SCD mouse model. These important findings raise critical questions in terms of global immunity. It has been known that when an infectious agent infects an immunocompromised or person with dysfunctional immunity such as SCD, the new form of pathogen most likely will be much more virulent. The gene-mutation rate will be much higher in such situations. This theory warrant further research to understand how few SCD patients in a community can affect herd immunity of large population?

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Conflict of interest

The author declares no conflict of interest.