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Opinion

Sickle Cell Disorder Co-Morbidity with Human Immunodeficiency Virus Infection

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Sickle Cell Disorder is an inherited hemoglobinopathy occurring mainly in West African populations.

The red blood cells (erythrocytes) in affected individuals assume a sickle shape thus compromising the cells' oxygen carrying capacity and life span. Hemoglobin S (HbS) is devastating in its homozygote form causing chronic anaemia and complications. The heterozygote form of the disorder often exists with normal Hemoglobin A i.e. HbAS and less frequently with Hemoglbin C as HbSC.

Levels of HbS in indigenous West Africans is attributed to the advantages of the heterozygous form; HbAS. HbAS confers some protection from faliciparum (malignant) malaria. Natural selection of HbAS due to survival from malaria compared to those with the homozygote HbAA has contributed to the existing 10 - 45% of young adults having HbAS in West African countries. Couples who each have HbAS have a 25% chance of giving birth to a child with HbSS. About 1.5% of children born in West Africa have HbSS.

Chronic anaemia is the major clinical feature of HbSS. Individuals with HbSS are susceptible to pneumococcal infection, salmonella osteomyelitis and high childhood mortality. As part of the management of their condition, majority of adults with HbSS have received blood transfusions in their lifetime. Unfortunately, blood transfusion services sometimes have sub-optimal HIV screening facilities and scientists may miss detecting HIV infection in the donor due to the "window period".

One would therefore infer that an individual with HbSS is exposed to the risk of acquiring HIV infection from blood transfusion. Individuals with HbSS and HIV infection would be expected to have an unusually high susceptibility to infections resulting from the combination of chronic anaemia and immunodeficiency. Progressively poor clinical prognosis could ensue.

THIS IS NOT SO! Individuals with HbSS who inadvertently acquire HIV infection have been informally observed to have same health challenges comparable to the period before the individual became infected with HIV!!! Literature review has not revealed any study confirming or contradicting this observed phenomenon. There is the suspicion of some level of resistance to potentiating effects of HIV infection in individuals that have HbSS.

Ongoing efforts on pre-marriage counseling and prenatal diagnosis will reduce the number of babies born with HbSS. Furthermore, improved HIV screening services for whole blood and increased use of blood components reduce transmission of HIV infection through blood transfusion. Co-morbidity of individuals with HbSS who have HIV infection would further reduce.

Research studies on persons with HbSS who have HIV infection may reveal a scientific breakthrough in physiologic barriers to devastating consequences of co-morbidity. Such studies are time-bound because of the aforementioned improvement in health services and subsequent reduction in existing study populations.

Scientists need to act now!!!

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