EDITORIAL

Caring for Children with Sickle Cell Disease

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Sickle Cell Disease encompasses a range of conditions that are characterized by the inheritance of two abnormal genes that are responsible for haemoglobin with one of them coding for haemoglobin S. With this in mind, it is obvious that this genetic condition is ubiquitous in the world as a result of interracial and interethnic marriages, but finds its outstanding expression in people of African parentage where the haemoglobin S is most prevalent.[1]

The scope of adverse events related to this condition is as a result of genetically defective haemoglobin that crystallizes out in oxygen tension lowering circumstances as is the case with infections and other conditions that are responsible for hypoxia. The one prominent event and most distressing is the vaso- occlusive (painful) crisis which accounts for most hospitalizations.

This occurrence so often heralds the presence of the ailment. I vividly remember a number of infants aged around six months, in excruciating pain and inconsolable while parents wondered desperately what the problem was all about. And once the diagnosis disclosed, they would ask questions that carelessly concealed their fears of the child's compromised growth and impending death, among others, in view of the multiple complications inherent to the condition.

A sense of guilt and visible unpreparedness to deal with this calamity would take a merciless grip on their lives. Many, in their denial, would ask for a second confirmatory test with a narrow hope of a comforting negative outcome.

It goes without saying that low income settings in Africa and Zambia in particular can only be amplifiers of health and socio economic challenges faced by people affected by sickle cell disease. The socio-cultural aspect of the disease is of paramount importance as many still need to be enlightened about the condition and its mode of inheritance so as to help them cope with it.

It is obvious that parents with a better understanding of the condition will observe measures that would enhance the quality of life of the affected child and that of the entire household.

With the improvement of quality care fostered by follow up programmes such as the one that has been in existence at the University Teaching Hospital (Lusaka, Zambia) for a number of years, morbidity and mortality related to Sickle Cell Disease (SCD) have been curtailed resulting in many patients affected with this ailment reaching adulthood. They are now faced with challenges related to this age group such as employment and marriage. Issues of reproductive health and employment, among many others, turn into a source of anxiety as disclosure of their status might constitute an added stressful tight spot.[2]

In as much as it is the sufferer's predicament, it also affects the immediate family members as they would want to be assured of the well being of their kindred.

This calls for efforts to reduce the deleterious effects of Sickle Cell Disease on both the affected child and the family by putting in place strategies such as improved awareness, disease prevention, early detection through genetic counseling (i.e. premarital screening) and neonatal screening as advocated at the sixtieth session of WHO-Afro held in Malabo, Equatorial Guinea in September 2010.

REFERENCES

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