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Letter to the Editor

Sir,

Preventable Morbidity and Mortality in Children with Sickle Cell Disease and Fever: the need for a National Protocol

Children with sickle cell anaemia and its variants are at much higher risk than normal for morbidity and morbidity. This is due to the complications associated with sickle cell disease of which overwhelming sepsis is the most common. This problem has been recognized for at least, 30 years. Furthermore, as developing countries struggle to meet the millennium goals and reduce the mortality in children younger than five years, the contribution of sickle cell anaemia to childhood deaths is likely to increase unless specific interventions are developed to target the disease.

Each year, an estimated 200,000 children in Africa are born with sickle cell anaemia, and more than 70 percent die before the disease is diagnosed.[1] The incidence of bacterial sepsis in our affected population in Nigeria has been estimated to be as high as 60 percent with a case fatality rate of 25-35 percent.[2] This number is not encouraging in the face of millennium goals and the vision of reducing the national child mortality rate, and it is therefore unacceptable.

Paediatric haematologists in highly industrialised countries such as the USA have developed elaborate protocols for preventing, evaluating and treating febrile children with sickle haemoglobinopathies with the aim of avoiding and reducing morbidity. There has been a push to adopt these protocols in developing countries but express transfer of knowledge and practice from these developed countries is not practicable or useful in the face of poor infrastructure or resources. Furthermore, there has been a decade long argument that the organisms that cause sepsis in our population are different from those in the developed world. While studies in the USA have shown that children with SCD are highly vulnerable to *Streptococcus pneumoniae, Haemophilus influenzae* and Salmonella,[3] the few studies carried out in sub-Sahara Africa have indicated otherwise. In studies carried out in Nigeria and Uganda, S. aureus, E. coli, Klebsilla species, and non typhi salmonella species have dominated.[4-7] In a more recent study carried out in Nigeria in 2007, the three commonest organisms isolated were S. aureus, *Haemophilus influenzae* and S. epidermidis.[8] But studies carried out in the Congo and most recently in Kenya, have found organisms similar to those described in studies carried out in the USA.[9,10] The reasons given for this discrepancy include the suggestion that *Staphylococcus aureus* and *Escherichia coli* are easier to culture than fastidious organisms such as *Streptococcus pneumoniae* and *Haemophilus influenzae* and in laboratories with little resources, this differential culture sensitivity might bias the distribution of the pathogens detected. Secondly, most of the studies have examined patients with existing diagnosis of SCD in a region in which infections with H.influenzae and pneumococcus occur predominantly in young children most of whom die before a diagnosis of sickle cell disease is made.[9]

With the above in mind, various studies have been carried out that could help in formulating a protocol plan. Most recent research carried out in Kenya has established that children on daily prophylactic penicillin are at considerably decreased risk of death from sepsis.[9] Even more encouraging, a Cochrane study carried out by Oniyangi *et al* showed that malaria prophylaxis plus antibiotics even further decreased morbidity and mortality in children with SCD.[11] Also established by Akinyanju *et al* is that children with SCD, if cared for under a special sickle cell programme where there are laid down guidelines in the management of a febrile sickle cell patient and other sickle cell crisis from the time of diagnosis, fare much better with less financial burden and stress on the family.[12,13] National neonatal screening for sickle cell disease, which is presently carried out in parts of Kenya, Ghana and Benin Republic, allows implementation of treatment that includes early initiation of malaria and bacterial prophylaxis and introduction of vaccines which together reduce morbidity and mortality in the first five years of life.[1,9]

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In Africa, where more than 80 percent of all children with this disease are born, a scarcity of data has impeded development of evidence-based guidelines. The general national literature has almost been completely silent concerning this problem despite its importance and a general high awareness of the significance of fever in children with SCD among paediatricians. Hence the need for well documented epidemiological studies with careful analysis of results which can help direct health policies.[14]

It is time for paediatricians to be persuaded of an urgent necessity to give priority to research related to the health of children with sickle cell disease in the country. They should consider establishing protocols for preventing, evaluating and treating febrile children with sickle haemoglobinopathies because "prevention is indeed better than cure".

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