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Unusual Features of Haemolytic Crisis in Sickle-Cell Anaemia

Sir,

The case report by Olarenwaju, Ogunfowora and Njokanma [1] merits some comments. In the absence of jaundice, will it be correct to describe the crisis as haemolytic? Is acute splenic sequestration (ASS)not a more accurate description? ASS in children with homozygous sickle-cell disease (HbSS) typically presents with severe pallor, reticulocytosis and spienomegaly of varying degrees. It is one of the common causes of morbidity and mortality in young children with HbSS disease.[2] It can follow a mild febrile illness, or may be associated with a severe septicaemic illness such as pneumococcal pneumonia.In a prospective study by Topley *et al,*[3] symptoms were found to be non-specific and may be mild or absent, even in fatal cases. It is most common between the ages of six months and two years, but can occur within the first five years of life. It is thus, my opinion that ASS is the diagnosis that best fits the patient in question.

Acute splenic sequestration tends to recur and mortality increases with recurrence.[3] Management of this condition is either by chronic blood transfusion or splenectomy. Splenectomy is recommended by Seeler and Shwaiki;[4] it is preferred because of the problems associated with chronic blood transfusion. Moreover, in most tropical countries, blood for transfusion may not be easily available when most urgently needed. Also, as shown by the work of Rogers, Serjeant and Serjeant,[5] the immune functions of the spleen are compromised with episodes of ASS. Two years ago, I managed a 13-month old male child with HbSS who, within a seven-month period, received three emergency blood transfusions for acute falls in haemoglobin. After the third transfusion, an elective splenectomy was performed and till date, this patient has not been transfused again.

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1 Olarenwaju DM, Ogunfowora CB and Njokanma FO. Unusual features of haemolytic crisis in sickle-cell anaemia. *Nig J Paediatr* 1992; **19**: 44-6.

2 Rogers DW, Clarke JM, Cupidore L, Ramlal AM, Sparke BR and Serjeant GR. Early deaths in Jamaican children with sickle-cell disease. *Br Med J.* 1978; **1**: 1515-6.

3 Topley JM, Rogers DW, Stevens MCG and Serjeant GR. Acute splenic sequestration and hypersplenism in the first five years in homozygous sickle-cell disease. *Arch Dis Child* 1981: **56**: 765-9.

4 Seeler RA and Shwiaki MZ. Acute splenic sequestration crisis (ASS) in young children with sickle-cell anaemia. *Clin Pediatr* 1972; **11** 701-4.

5 Rogers DW, Serjeant BE and Serjeant GR. Early rise in 'pitted' red cell count as a guide to susceptibility to infection in childhood sickle-cell anaemia. *Arch Dis Child* 1982; **57**: 338-42.