SUMMARY

Background  Sickle cell anaemia is a severe, chronic genetic disorder with a very high prevalence in Nigeria and other parts of the world. This study related Hb F levels to indices of severity in sickle cell anaemia so as to investigate the potential of therapeutic modalities directed towards ameliorating the disease by raising the levels of Hb F.

Methods  The study was carried out between November 2006 and March 2007. One-hundred and sixty patients who met the inclusion criteria were recruited into the study as they attended the haematology clinic of the Aminu Kano Teaching Hospital. The Aminu Kano Teaching Hospital (AKTH), a three hundred bed tertiary health facility located in Kano state, North-Western Nigeria. Established in August 1988 it serves a large catchment area including Kano, Jigawa, Katsina, Zamfara, Yobe and Bauchi states with a total population of over twenty-five million people. The total number of admissions to the hospital average about ten thousand per year. During the clinic visit a questionnaire was administered to each patient to obtain and document socio-demographic data and clinical history and to ascertain previous hospital admissions for a painful crises or blood transfusion during the preceding twelve months. Each patient was thoroughly examined to check for the presence and degree of characteristic ‘sickle cell facies’. These were evaluated using the simple grading system adopted by Konotey-Ahulu. Long-term skeletal complications such as digital shortening, collapse of the femoral head causing a limp, and chronic leg ulcers were checked for. A blood sample was taken for the estimation of haemoglobin F and a full blood count. Blood sampling was deferred to a subsequent date for patients who were not in the steady-state.

Results  Patients with higher Hb F levels were found to be older and to have higher haemoglobin levels, fewer incidences of leucocytosis, acute painful crises, blood transfusion and admission to hospital.
Conclusions A higher Hb F level ameliorates the clinical severity of sickle cell anaemia.