

CREATION OF A LARGE RESEARCH COHORT, INCLUDING NEW-BORN INFANTS AND ADULTS

Rationale:

Over the past 30 years well-studied cohorts of patients with SCD have been developed in the US, Jamaica and Brazil yielding crucial insights in the clinical course of the illness and the value of prophylactic and therapeutic interventions. On the contrary, very limited data exist on both survival and clinical course of SCD in Africa. Estimates of survival to the age of 5, for example, range from 10 to 70%, and the incidence of many severe complications, such as stroke in children, is hardly documented at all. While it is known that the spectrum of complications from SCD among African patients is overall similar to what is observed in industrialized societies, specific exposures in tropical Africa, such as endemic malaria, may influence the clinical course in ways that are not yet well understood. Well-characterized cohorts are equally necessary for studies of the influence of modifying genes – particularly those associated with HbF, cytoprotective enzymes – and the pathophysiologic consequences of high rates of hemolysis.

