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Survey of Haemoglobin Genotypes in Children at Ilorin

J O ADEWUYI† AND E A AKINTUNDE††

SUMMARY

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Haemoglobin electrophoretic patterns were determined in 1,296 apparently healthy children aged between one month and fourteen years. Homozygous SS (Hb SS) was found to have a prevalence of 1.5% and heterozygous SC (Hb SC) one of 1.3%, giving a combined prevalence of 2.8% in the population studied. The prevalence of sickle cell disease (HbSS and HbSC) was found to vary with age. There was peak prevalence of HbSS at age 4 years, with a sharp drop at age 8 years; this suggests an increase in mortality and/or morbidity from the age of about 8 years. Whereas, prevalence of Hb SS remained low after this critical age, there was a stepwise rise in that of Hb SC to a smaller peak at the age of about 13 years.

Introduction

ONE of the factors known to contribute to the persistence of a high frequency of the sickle haemoglobin gene in tropical Africa is the partial protection from falciparum malaria infection enjoyed by carriers of the abnormal gene.[1] Variations in prevalence of sickle cell trait and disease in children in different parts of the tropics may therefore, be partly determined by the intensity and mode of falciparum malaria transmission in the region. Thus, Fleming et al[2] found that the prevalence of sickle cell trait in the Sudan Savanna region of Nigeria where falciparum malaria is hyperendemic was slightly higher (28.9%), than in the rainforest belt of Nigeria (24.7%) where the parasite is holoendemic. This was attributed to the more rapid acquisition, in a holoendemic situation, of partial immunity and the consequent quicker disappearance of the survival handicap among children with normal haemoglobin.[2]

University of Ilorin Teaching Hospital, Ilorin

Department of Paediatrics

† Senior Lecturer,

Department of Haematology,
†† Principal Medical Laboratory Technogist.

Correspondence: Dr J O Adewuyi

The prevalence of sickle cell trait and disease has hitherto not been widely reported in the Guinea Savanna, a transitional zone between the Southern rainforest and northern Sudan and Sahel regions of Nigeria. This study was conducted in Ilorin the capital of Kwara State, to document the prevalence of sickle haemoglobins in the Guinea Savanna area.

Materials and Methods

The subjects were apparently healthy children between the ages of one month and fourteen years. Children under two years were recruited at the Infant Welfare Clinics where babies are brought for routine immunisation, while those aged three to five years were chosen from Nursery schools. Primary schools provided subjects aged 5-11 years and secondary schools, those aged 11 – 14 years. For a good cross section of children within the region institutions sampled were widely spread and included elite and non-elite schools. To ensure inclusion of children living in rural environment, institutions were sampled from Ganmo and Shao two rural communities on the outskirts of Ilorin township. A small adult sample consisting of school teachers and parents above the age of twenty-five years was also examined for comparison.

Blood was obtained by finger pricks and drawn into a pair of heparinised capillary tubes. From one, blood films were made and packed cell volume determined while the other was used to determine haemoglobin genotypes. Haemoglobin electrophoresis was carried out on cellulose acetate strips using Tris-borate buffer at pH 8.4 according to the system developed by Schneider[3] and used by Serjeant *et al* for screening samples of cord blood for detection of sickle cell disease in Jamaica.[4]

Results

One thousand two hundred and ninety six children and one hundred and ten adults were examined. The distribution of Haemoglobin (Hb) genotypes in the 1296 children and 110 adults is shown in Table I. Eight hundred and ninety three (68.9%) of the children had Hb genotype AA; two hundred and ninety five (22.8%) had As and sixty eight (5.2%) had Hb AC. Hb genotypes SS and SC were almost equal at 1.5% and 1.3% respectively making a total of 2.8% in the one month to fourteen years aged group for the two forms of sickle cell disease (Hb SS and Hb SC). Table I also shows that the peak prevalence of Hb SS occurred at age 4 years (3.75%), falling slightly thereafter, until age 8 years, when it dropped sharply to nearly zero. A similar peak in the prevalence of Hb SC occurred at age 5 years (3.45%) with a sharp fall to zero at age 7 years. However, although the prevalence remained low for Hb SS after age 8 years (Fig 1), for Hb SC there was a gradual rise from age 10 years to another, though smaller peak (2.5%) at age 13 years.

Discussion

Mortality due to sickle cell disease is minimal below the age of six months,[5] and the frequencies of the Hb genotypes below the age of six months reflect the relative frequencies at birth. Thus in the present study, it is apparent 4.4% of babies born in the population have Hb SS or SC (Table 1) This value is higher than the 2.06% of newborns with Hb SS to SC found by Fleming in Northern Nigeria.[2] However the gene frequency

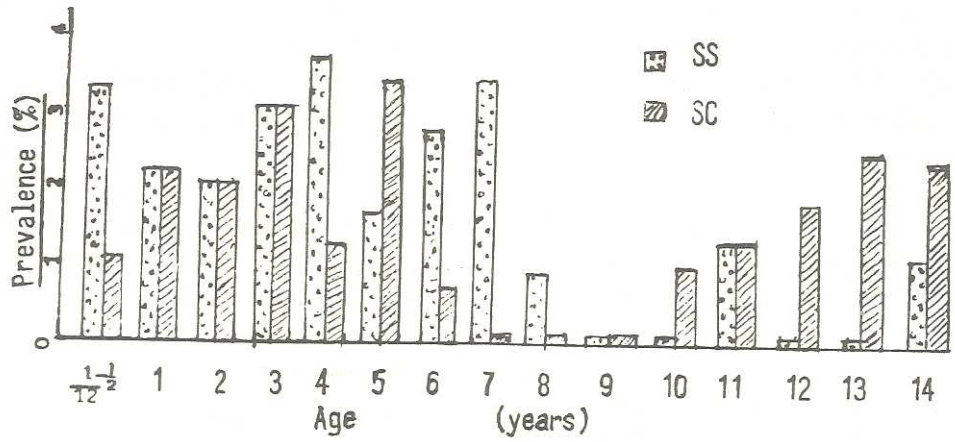

Fig. 1 Prevalence of Hb SS and HB SC by age in Ilorin

TABLE I
Distribution of haemoglobin genotype in Ilorin

Age in years	Total No	Hb AA No of cases	% of total	Hb AS No of cases	% of total	Hb AC No of cases	% of total	Hb SS No of cases	% of total	Hb SC No of cases	% of total	Hb CC No of cases	% of total	SS + SC % of total
1/12 – 1/2	90	65	72.2	15	16.6	5	5.5	3	3.3	1	1.1	1	1.1	4.4
1/2 – 1	45	27	60.0	13	29.0	3	6.6	1	2.2	1	2.2	0	0.0	4.4
2	46	30	65.2	12	26.1	2	4.3	1	2.1	1	2.1	0	0.0	4.2
3	32	22	68.8	7	21.8	1	3.1	1	3.1	1	3.1	0	0.0	6.2
4	80	59	73.7	16	20.0	1	1.25	3	3.75	1	1.25	0	0.0	5.0
5	58	44	75.8	8	13.8	3	5.2	1	1.72	2	3.45	0	0.0	5.2
6	141	95	67.4	34	24.1	7	4.9	4	2.8	1	0.7	0	0.0	3.5
7	84	57	68.0	21	25.0	3	3.5	3	3.5	0	0.0	0	0.0	3.5
8	122	88	72.0	24	19.8	8	6.5	1	0.8	0	0.0	1	0.8	0.8
9	90	61	68.0	24	26.6	4	4.4	0	0.0	0	0.0	1	1.1	0.0
10	100	68	68.0	24	24.0	7	7.0	0	0.0	1	1.0	0	0.0	1.0
11	73	39	53.4	23	31.5	9	12.3	1	1.3	1	1.3	0	0.0	2.6
12	165	116	70.3	41	24.8	5	3.0	0	0.0	3	1.8	0	0.0	1.8
13	81	62	76.5	14	17.3	3	3.7	0	0.0	2	2.5	0	0.0	2.5
14	89	66	67.5	19	23.5	7	8.6	1	1.1	2	2.3	0	0.0	3.4
0–14	1296	893	68.9	295	22.8	68	5.2	20	1.5	17	1.3	3	0.2	2.8
Above 25	110	76	70.0	28	25.0	6	5.0	0	0.0	0	0.0	0	0.0	0.0

for the S – gene calculated from the observed genotype frequencies below the age of six months in Ilorin is 0.105 which is significantly lower than the value of 0.128 calculated from Fleming's figures for Garki in Northern Nigeria ($p > 0.01$).

The prevalence of sickle cell trait, HbAS, rose from 16.6% at 6 months to 29.0% at one year, while HbAA dropped by a similar margin over the same period. This marked change during the second half of the first year of life suggests that the Hb AS children

survive better over this period, which coincides with the time of high mortality among children with falciparum malaria infection. It can therefore be inferred, in agreement with earlier observations,[2] that the S-gene gives partial protection against falciparum malaria parasitaemia and infection. The point is further proved by the drop in the prevalence of Hb AS back to 13.8% at the age 5 years, indicating a reduction in the survival advantage of Hb AS children as both Hb AS and Hb AA children acquire partial immunity to malaria after about 5 years exposure to the parasite.

The change in frequency of Hb SS and SC with age is interesting. Morbidity studies have tended to stress the vulnerability of the sickle cell disease patient in early childhood. Kaine[6] observed that the incidence of infections and sickle cell crisis among sickle cell anaemia patients fell progressively with age with a sharp decline after the age of 10 years. Mortality pattern was however, not discussed. Other workers in Africa and elsewhere have also found that morbidity in terms of frequency of admissions to hospital,[7] incidence of painful crisis,[8] and frequency of clinic visits,[9] is greater in children below five years of age. Although there is scanty data on mortality patterns in children with sickle cell disease, it is believed that mortality is also higher in early childhood. Up to two thirds of children with sickle cell anaemia are said to die by the age of five years.[10] The fall in frequency of Hb SS and SC genotypes among apparently health children from age 8 years in the study, would suggest that mortality and/or morbidity is significantly increased at this age. This may represent a new trend probably resulting from better medical care in early childhood which has not been extended to middle childhood occupied by the stressful years of primary and junior high school. Further mortality studies are required to shed more light on the situation.

The significant reappearance of Hb SC in the healthy children population from the age of 13 years would tend to support the observation that sickle cell HbC disease is a clinically milder form of the diseases than sickle cell anaemia especially beyond childhood.[11]

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