Evaluation of haematological findings in 50 Bahraini patients with sickle cell disease and in some of their parents

M A BUHAZZA*, A B BIKHAZI†, AND F P KHOURI‡

From the *College of Health Sciences, State of Bahrain; and †the Department of Physiology and ‡the Department of Laboratory Medicine, American University of Beirut, Lebanon.

SUMMARY The haematological findings in 50 Bahrainis with sickle cell disease are reported. This establishes the existence of the Hb S gene in Bahrain. The mean Hb F level in the Bahraini patients was 13.8%, a value lower than that encountered in sickle cell homozygotes from Kuwait and Saudi Arabia.

The clinical manifestations of sickle cell anaemia result from the presence of Hb S in the erythrocytes of sickle cell patients. In addition to Hb S the erythrocytes of sickle cell patients may also contain variable amounts of fetal haemoglobin.¹ Increased amounts of fetal haemoglobin apparently protect the erythrocytes from sickling, thereby reducing the secondary clinical manifestations.² Saudi Arabs and other populations in the Middle East exhibit a benign type of sickle cell disease compared with Blacks in Africa and the Americas.³ This appears to result from the presence of relatively large amounts of fetal haemoglobin in their red blood cells.⁴ To our knowledge, no similar studies have been carried out on the Bahraini Arab population. The present investigation was undertaken to determine the existence of the sickle gene among Bahraini Arabs.

Materials and methods

CONTROL SUBJECTS

Cord blood from 21 Bahraini newborn infants and venous blood from 20 Bahraini clinically normal adults were collected and studied for the presence of abnormal haemoglobins. The procedure used for haemoglobin analysis was the microzone method for haemoglobin electrophoresis.^{5,6} Fetal haemoglobin values were determined by the alkali denaturation method of Singer *et al.*⁷ Quantitative estimation of haemoglobin A₂ was obtained by column chromatography on diethyl amino ethyl cellulose (DEAE).⁸

The latter test was carried out only on blood from the adult group.

Fifty Bahraini subjects with sickle cell disease were initially identified with the haemoglobin solubility test followed by haemoglobin electrophoresis⁹ on cellulose acetate. Fetal haemoglobin was measured as for the control group. Other standard haematological tests were performed according to the procedures of Dacie and Lewis.⁹ The patients' age at the time of the study ranged from 11 months to 30 years. There were 22 females and 28 males.

FAMILY STUDY

Family study was possible in only 21 mothers and 16 fathers of 27 patients, as the rest refused the examination, and in one instance the father was dead.

Results

CONTROL SUBJECTS

The electrophoretic findings in the control group are summarised in table 1. In all infants, the haemoglo-

TABLE 1 Data on control subjects.

	Electrophoretic pattern	Hb F AD (%)	Hb A ₂ (%)	Hb S (%)
Infants (21)*	A + F	50-70 (59.4 ± 1.2)		
Adults (17)*	A	0.8-3.0 (1.48 ± 0.14)	1.7-3.1 (2.04 ± 0.08)	
Adults (3)*	A + S	1.7-2.0 (1.8 ± 0.08)	1.3-2.2 (2.04 ± 0.08)	32-36 (34.3 ± 1.2)

*Number in parentheses indicates the number of control subjects tested.

Received for publication 11 May 1984.

Accepted for publication 28 September 1984.

bin pattern was AF. Seventeen out of 20 adult blood samples showed normal electrophoretic patterns. The remaining three subjects showed the AS pattern; their Hb F and Hb A₂ values were within the normal range. The findings in these three adults are compatible with the sickle cell trait, so they were excluded from the study.

Table 2 shows the haematological data on the 50 patients with sickle cell disease. All showed two bands of haemoglobin on electrophoresis identified as Hb S and Hb F; Hb A was absent. Seven patients had Hb F levels within the normal range, 17 had Hb F <10%, and in the remaining 26 patients Hb F was >10%. No statistically significant difference was found in the haematological findings between males and females within the different age groups of these patients

FAMILY STUDY

The results are summarised in table 3. SA electrophoretic patterns were revealed in 16 mothers and 15 fathers. Two mothers and one father showed SF patterns and their Hb F values were 18.3%, 5.0%, and 4.4%, respectively. The last three mothers showed negative sickling tests and their electrophoretic pattern was normal. Most of the parents were consanguineous.

Discussion

The findings reported in this manuscript are the first on sickle cell disease from the State of Bahrain. A total of 50 cases was investigated. Family studies of both parents were possible in only 15 instances, and in another seven only one parent was available for

TABLE 2 Haematological data on 50 sickle cell disease patients.

	Mean	Range	Remarks
Hb G (%)	8.9±0.22	5.4–11.5	Less than mean in 20 patients
PCV (%)	26.9±0.7	15–35	
Platelet count (μl)	209 000±13 700	92 000–620 000	Increased in 2 patients
WBC (μl)	12 200±1120	4000–34 400	More than 10 000 μl in 24 patients
RBC (10 ⁶ /mm ³)	(3.02±0.07)10 ⁶	(2.0–3.9)10 ⁶	
MCV (fl)	88.4±0.9	71–107	
MCH (pg)	29.1±0.3	26–33	
MCH (%)	33.0±0.3	29–43	
Reticulocyte (%)	5.7±0.79	0.3–20.5	Raised in 12 patients
HB F (%)	13.8±1.58		

Electrophoretic pattern S+F in all patients.

TABLE 3 Results of family studies.

Patient No	Sickle cell test		Electrophoretic pattern		Remarks
	Mother	Father	Mother	Father	
1	+ve	+ve	S+A	S+A	
2	+ve	+ve	S+A	S+A	Parents are related
3	+ve	+ve	S+A	S+A	Parents are related
4	+ve	—	S+F	—	Hb F of mother 18.3%
5	+ve	+ve	S+A	S+F	Hb F of father 4.4%
6	+ve	Dead	S+A	—	
7	+ve	—	S+A	—	Parents are related
8	—ve	+ve	A	S+A	
9	—ve	—	A	—	
10	+ve	+ve	S+A	S+A	
11	+ve	+ve	S+A	S+A	Parents are related
12	+ve	+ve	S+F	S+A	Hb F of mother 5.0%
13	—	+ve	—	S+A	Parents are related
14	+ve	+ve	S+A	S+A	
15	—ve	+ve	A	S+A	Parents are related
16	+ve	+ve	S+A	S+A	
17	+ve	+ve	S+A	S+A	
18	+ve	—	S+A	—	4 patients are sibs
19	+ve	+ve	S+A	S+A	2 patients are sibs
20	+ve	+ve	S+A	S+A	2 patients are sibs
21	+ve	—	S+A	—	Parents are related
22	+ve	+ve	S+A	S+A	

examination. Among the total number of parents examined, three out of 37 were themselves also homozygous for the disease and three mothers had no Hb S and were presumably β thalassaemia heterozygotes. The remaining 31 parents were Hb S heterozygotes.

The results of this study also showed that Hb F levels ranged from normal to 54.2%, with a mean of about 14%. This is lower than those encountered in Kuwait¹⁰ and Saudi Arabia^{4 11 12} where the disease is relatively benign.

In conclusion, the above findings have established the existence of sickle cell disease in Bahrain. The presence of three heterozygotes for the S gene in a control group of 20 adults suggests a high heterozygote frequency among Bahrainis. Further work with more adequate sampling is needed in order to determine the exact incidence of the sickle cell gene in this country.

The authors wish to acknowledge the Ministry of Health in the State of Bahrain for financial support. Thanks and appreciation are also extended to the management, staff, and students of the College of Health Sciences in Bahrain, and to Dr V V Deshmakh and the staff and technicians of the Department of Pathology, Salmaniya Medical Centre, Bahrain for technical help.

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Correspondence and requests for reprints to Dr Anwar B Bikhazi, Department of Physiology, American University of Beirut, Beirut, Lebanon.