Psychiatric complications of homozygous sickle cell disease among young adults in the Jamaican Cohort Study

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Background This study aimed to determine the prevalence of psychiatric disorder in young adults with homzygous sickle cell (SS) disease and in controls with normal haemoglobin, and to examine factors associated with psychiatric disorder.

Method The study design was cross-sectional. Subjects were aged 18–20 years: 63 with SS disease and 89 controls. The Psychiatric Assessment Schedule was used to determine psychiatric disorder at Index of Definition level 5.

Results Psychiatric disorder was identified in 18 (29%) SS disease patients and in 22 (25%) controls. In SS patients, psychiatric disorder was not related to illness severity but was associated with leaving school early, difficulties in social adjustment, impaired cognitive function and having previous psychiatric difficulties. Male SS patients with psychiatric disorder all had low body mass index (BMI <17.60). In controls, psychiatric disorder was associated with female gender, unemployment and difficulties in social adjustment.

Conclusions The prevalence of psychiatric disorder was similar in patients and controls, although associated factors tended to be different. The association with low BMI in SS men merits further study.

Homozygous sickle cell (SS) disease is a genetically determined condition affecting 0.3% of births among people of Caribbean origin and 1–2% of births among people from West Africa. In Britain, an estimated 5000 people have sickle cell disease (Streetly *et al*, 1993).

The abnormal sickle haemoglobin (HbS) tends to polymerise when deoxygenated, distorting the red cells, which are prematurely destroyed and may block flow in blood vessels. Rapid haemolysis results in anaemia, jaundice, increased gall stone formation and markedly expanded bone marrow activity. Abnormal red cells damage spleen function causing death from acute splenic sequestration or pneumococcal septicaemia. Death of bone marrow causes recurrent pain and swelling of the bones manifested by the typical painful crisis in adolescence and early adulthood. Damage to the skin circulation results in chronic leg ulceration around the ankles, arising most commonly in adolescence. Other serious complications include a pneumonia-like syndrome (acute chest syndrome), stroke, overwhelming infections, aplastic crisis and, eventually, chronic organ damage. There are complex derangements of growth in SS disease. Physical and sexual development is retarded, and although the height of many adult patients may exceed that of the normal population, body build tends to be slim with low body mass index (BMI). Although complications tend to recur with frequent hospital admissions, the disease is very variable and complications may be separated by long periods of good health. Average life expectancy is reduced.

PSYCHIATRIC MORBIDITY IN SICKLE CELL DISEASE

In childhood, girls have been observed to adapt to sickle cell disease better than boys, and younger better than older children (Hurtig & Park, 1989). Youngsters with sickle cell disease tended to withdraw socially, showed less body satisfaction and had more symptoms of depression than matched controls (Kumar *et al*, 1976; Morgan & Jackson, 1986). Asthenic body build has been associated with psychosocial difficulties, especially in adolescent males (Walco & Dampier, 1987; Knight *et al*, 1995). Both metabolic and psychological factors, such as parental attitudes and interactions with a sick child, may affect intellectual development (Swift *et al*, 1988; Wasserman *et al*, 1991).

Psychiatric disorders, such as anxiety and depression, occur in up to 40% of physically ill adult patients, four times more frequently than among the community at large (Bridges & Goldberg, 1984; Royal College of Physicians and Royal College of Psychiatrists, 1995). Recent advances in the treatment of sickle cell disease have been associated with increased survival (Lee *et al*, 1995), posing physical, psychological and social challenges to patients and clinicians. Among adults with sickle cell disease there are conflicting data on the frequency of psychiatric disorder and factors associated with it. One small study (n=22) suggested that 50% of painful crises followed life events and dysphoric feelings (Nadel & Portadin, 1977). Patients may become preoccupied with pain (Gil *et al*, 1990). Leg ulcers are associated with leaving school early, unemployment and lack of stable sexual relationships (Alleyne *et al*, 1977). Using the General Health Questionnaire and Social Functioning Schedule, Damlouji *et al* (1982) found no excess of psychiatric morbidity or psychosocial dysfunction among 30 sickle cell out-patients compared with a control group with diabetes mellitus. Level of psychosocial function was not associated with physical illness severity. Barrett *et al* (1988) found concerns about employment, finances, performance of normal daily activities, fear and anxiety regarding body deterioration, and lack of assertiveness in social relationships, although these were unrelated to physical illness severity. Depression affected 57% of 46 patients in one uncontrolled study using the Beck Depression Inventory, and was reported to predict emergency room attendance and hospital admissions (Belgrave & Mollock, 1991). In another study 45% of 109 patients scored above the 90th percentile on the Symptom Checklist (SCL-90) indicating psychiatric disorder (Thompson *et al*, 1992). Frequency of pain was associated with anxiety, and severity of psychiatric disorder with family difficulties and less

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effective pain-coping strategies. These formal studies of psychiatric disorder in sickle cell disease have been limited by many factors. Imprecise genotype diagnosis has led to pooling of data from almost asymptomatic genotypes with more severe forms of the disease. Patients studied have been selected from out-patient clinics, which may have excluded relatively asymptomatic patients. Control data have either been absent or have used standardised norms derived from dissimilar populations. Response rates have often been unreported, leading to uncertainty about the representativeness of the findings. Psychiatric disorder has been assessed by unstructured interviews or self-rated questionnaires, which are both unreliable, the latter also frequently including questions about somatic function, thus tending to overestimate psychiatric disorder in patients with physical illness (Mayou & Hawton, 1986). Because of these methodological shortcomings, it is not possible to draw well-founded conclusions about either the frequency of psychiatric disorder in young adults with sickle cell disease, or factors associated with it.

The present study attempts to clarify this picture by the use of a standardised interview assessment of psychiatric disorder in a representative group of patients with SS disease and a matched normal control group.

METHOD

Subjects

All subjects have participated in the Jamaican Cohort Study based on the neonatal diagnosis of all cases of sickle cell disease among 100 000 consecutive non-operative deliveries at the main Government Maternity Hospital between June 1973 and December 1981 (Serjeant *et al*, 1986). This screening programme detected 315 infants with sickle cell phenotype. The first 125 were each matched with two infants of the same sex born closest in time to the index cases at the same hospital, but with a normal haemoglobin (AA) genotype, providing a group of 250 controls. Patients and controls were of similar racial groups, and predominantly of West African origin. All children were followed prospectively, monthly in infancy, and three-monthly thereafter, but also attending at any time if unwell, and all receiving free medical care. Of the first 125 with a sickle cell phenotype, subsequent follow-up revealed some had other genotypes, leaving 107 with SS disease. By the study date (January–June 1994), 31 (29%) had died, and 13 (12%) had emigrated, leaving a study group of 63 SS disease patients aged 18–20 years.

Of the 250 controls with an AA phenotype, four were subsequently found to have the β-thalassaemia trait, leaving a group of 246 with AA genotype. By the study date, six (2%) had died, 50 (20%, not statistically different from SS group) had emigrated and 32 (13%) had defaulted (all in early childhood), leaving a potential control group of 158. Precisely age-matched controls were available for 42 of the SS patients. Another 47 were selected at random from the remaining controls, within the age range of the index group.

The term genotype is used specifically to refer to the haemoglobin genotype and not to differences in gender. All subjects were physically well at the time of interview. All assessments were made by one interviewer (C. H.).

Assessments

Socio-demographic data were collected as follows: stable visiting (courting) or cohabiting relationships; number of children; urban or rural residence; employment status and educational experience. Employment of the main breadwinner in the household, domestic amenities (flush toilet, running water to house and electricity supply), and the number of people per room were used as an indication of socio-economic class according to Jamaican criteria (Smith, 1984; STATIN, 1990).

Physical illness data included medical history from birth obtained from clinic records, including a history of leg ulceration for a minimum duration of three months. Disease severity during the previous six months was inferred from number of painful crises, sick visits to the clinic, admissions to hospital and number of 'clinical events', that is episodes of physical illness not necessarily presenting to the sickle cell clinic.

Physical examination included leg ulceration and height and weight to calculate BMI. Normal BMI is 20–25 and a BMI below 17.5 is an ICD–10 requirement for a diagnosis of anorexia nervosa (World Health Organization, 1992).

Psychiatric disorder was assessed in a two-stage procedure. The General Health Questionnaire (GHQ-28; Goldberg & Hillier, 1979), a self-rated measure, was used as a screening instrument after adaptation of some idioms and phrases to Jamaican English. The questions were read to subjects with the questionnaire in front of them. To avoid missing chronic disorders, the GHQ was scored using the CGHQ method (Goodchild & Duncan-Jones, 1985). The GHQ was piloted on the first 30 subjects, using scores of four or above as a threshold for proceeding to second-stage psychiatric assessment interview. Twenty of these subjects were interviewed, identifying psychiatric disorder in seven subjects, all of whom had scored eight or above on the GHQ. The threshold was therefore raised to seven or above for the remainder of the project. For all subjects exceeding threshold scores on the GHQ, the Psychiatric Assessment Schedule (PAS; Dean *et al*, 1983) was used to determine psychiatric caseness. The PAS is derived from the Present State Examination (PSE), a standardised, structured psychiatric interview for eliciting and rating psychiatric symptoms present during the month before interview (Wing *et al*, 1974). The PAS contains neurotic symptom items from the PSE and screens for psychosis. Criteria for rating symptoms, and analysis of data using the CATEGO computer program, are the same as for the PSE (Wing & Sturt, 1978; Dean *et al*, 1983). Psychiatric morbidity is identified using the Index of Definition (ID), with ID level 5 as the criterion for psychiatric caseness among community populations, and ID level 6 indicating more severe disorder (Bebbington *et al*, 1991). Cultural and geographical factors were carefully considered in rating symptoms.

The only reliable, available index of past psychiatric difficulties was the Child Behaviour Checklist (CBCL; Achenbach & Edelbrock, 1983), administered to almost all SS subjects but to only 47 (53%) controls in 1990–1991. Psychosocial assessments included life events, social adjustment and psychoactive substance use. Life events within the previous six months were assessed using the List of Threatening Experiences (Brugha *et al*, 1985), a 12-item checklist designed to identify life events likely to have a marked long-term threat to mental health, including bereavement, loss of relationships, unemployment and criminal activity (arrest, police caution, court appearance). Operational criteria were defined for each item. A semi-structured interview was used to elicit recall of the event and its date. Social adjustment was assessed with the Modified Social Adjustment Scale self-report questionnaire (SAS-M; Cooper *et al*, 1982). It is divided into seven social role areas, each of which may be scored independently and an

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overall adjustment score calculated. Higher scores indicate worse adjustment. Language was modified and questions were read to subjects. Psychoactive substance use was assessed using a semi-structured interview based on DSM–III–R (American Psychiatric Association, 1987) criteria for abuse and dependence. Cognitive function was assessed with standard psychometric tests: digit span (Wechsler, 1945); immediate and delayed verbal memory of a 21-item story (derived from Wilson *et al*, 1985); increasing lengths of fist/flat-hand tapping sequences (details available from the author); immediate and delayed visual memory of two abstract line drawings (Wechsler, 1945); verbal fluency, naming fruit and animals, for one minute each (Lishman, 1987) and Corsi blocks, a test of visuo-spatial abilities (Lezak, 1976). Results of intelligence quotient (IQ) testing using the Wechsler Intelligence Scale for Children and the Wechsler Adult Intelligence Scale, performed in 1990–1991, were available for most subjects (Knight *et al*, 1995).

Analysis

All data were compared between genotypes, and psychiatric cases (ID level 5) and non-cases were compared within genotypes. For categorical variables, differences were assessed using the χ^2 test with Pearson's continuity correction or Fisher's exact test for small numbers. For scored variables that were usually not normally distributed, the non-parametric Mann–Whitney test was used. Forward stepwise logistic regression was used to ascertain the extent to which variables independently contributed to psychiatric caseness. In male SS patients, the association of psychiatric disorder and low BMI was explored. Rothman's (1990) argument against making corrections for multiple comparisons was followed. He states that in a logically designed project, data are not collected at random, but because they are relevant to the investigation. Thus, making such corrections may exclude significant data. Thresholds and validity coefficients were determined for the GHQ (Goldberg & Williams, 1988). The Statistical Package for the Social Sciences (SPSS/PC+4.0; SPSS Inc., 1990) was used for data analysis.

RESULTS

All 152 (100%) subjects approached agreed to participate in the study: 63 SS (31 male) and 89 AA (44 male). Social circumstances

Table I Some characteristics of SS patients and AA controls

Characteristic	Patients *n*=63 or [*n*] no. (%) or median (interquartile range)	Controls *n*=89 or [*n*] no. (%) or median (interquartile range)
Demographic/social		
Male	31 (49)	44 (19)
Visiting/cohabiting relationship	33 (52)**	69 (78)
Have at least 1 child	4 (6)**	24 (27)
Unemployed	24 (38)	20 (23)
Left school before 17 years	14 (22)	20 (23)
Physical illness		
Leg ulcers ever	24 (38)	
Current leg ulcers	15 (24)	
⩾1 painful crisis in previous six months	15 (24)	
⩾1 sick visit in previous six months	44 (70)***	21 (24)
⩾1 clinical event in previous six months	51 (81)***	30 (34)
⩾1 admission in previous six months	6 (10)**	0
Body mass index		
Male	[30] 17.7 (16.5–19.2)***	[44] 20.3 (19.1–22.0)
Female	[32] 18.5 (17.2–20.0)***	[45] 20.5 (19.0–23.0)
Past psychiatric difficulties		
Child Behaviour Checklist abnormal	[62] 22 (36)	[47] 17 (36)
Life events in previous six months		
⩾1 death	10 (16)	15 (17)
Break-up of relationship	7 (11)	10 (11)
Unemployed ⩾ one month	26 (41)	30 (34)
Loss of job	2 (3)	9 (10)
Modified Social Adjustment Scale		
Work/studies	[38] 1.3 (1.2–1.9)	[65] 1.5 (1.2–1.8)
Housework	[59] 1.5 (1.2–1.8)	1.4 (1.2–1.8)
Social and leisure	2.1 (1.8–2.5)	2.0 (1.7–2.4)
Extended family relationships	2.2 (1.9–2.7)	2.3 (1.9–2.6)
Overall adjustment score	1.9 (1.6–2.3)	1.9 (1.6–2.1)
Current use of alcohol and cannabis		
Alcohol	45 (73)	42 (67)
Cannabis	20 (33)	7 (11)
Current cognitive function		
Digit span forwards	[62] 7 (6–8)	6 (6–8)
Digit span reverse	[62] 4 (3–6)	4 (3–5)
Immediate verbal memory	[61] 6 (4–8)	[85] 6 (5–8)
Delayed verbal memory	[61] 8 (6–10)	[85] 8 (7–11)
Fist/flat sequence	[62] 5 (4–5)	5(4–6)
Immediate visual memory	[62] 4 (2–5)*	5 (3–5)
Delayed visual memory	[62] 3 (4–5)	4 (3–5)
Verbal fluency	[62] 24 (20–27)	24 (21–28)
Corsi blocks	[62] 5 (5–6)	5 (5–6)
Previously measured IQ[1]	[56] 76 (70–84)**	[35] 85 (78–94)

*$P<0.05$, **$P<0.01$, ***$P<0.001$, *v.* control.
1. The American (rather than the Jamaican) scoring system was used to score IQ tests, producing inappropriately low scores for the study population. Results cannot, therefore, be compared directly with other studies.

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Table 2 Variables associated with psychiatric caseness compared with non-cases

Characteristic	SS patients no. (%) or median (interquartile range)		AA controls no. (%) or median (interquartile range)	
	Case (*n*=18 or [*n*])	Non-case (*n*=45 or [*n*])	Case (*n*=22 or [*n*])	Non-case (*n*=67 or [*n*])
Demographic/social				
Male	8 (44)	23 (51)	6 (27)	38 (56)
Female	10 (56)	22 (49)	16 (72)*	29 (43)
Visiting/cohabiting relationship	9 (50)	24 (53)	16 (72)	53 (79)
Unemployed	10 (56)	14 (31)	11 (50)***	9 (13)
Leaving school before 17 years	8 (44)*	6 (13)	15 (68)	54 (81)
Physical illness[1]				
Leg ulcers ever	6 (33)	18 (40)		
Current leg ulcers	4 (22)	11 (24)		
⩾1 painful crisis in previous six months	4 (22)	11 (24)		
⩾1 sick visit in previous six months	14 (78)	30 (67)	6 (27)	15 (22)
⩾1 clinical event in previous six months	14 (78)	37 (82)	11 (50)	19 (28)
Body mass index				
Male	[8] 16.4 (15.7–17.5)**	[22] 18.3 (17.4–19.8)	[6] 21.4 (19.9–23.4)	[38] 20.2 (18.8–21.7)
Female	[10] 19.2 (18.0–20.2)	[22] 18.2 (17.0–19.4)	[16] 20.2 (18.7–25.0)	[29] 20.9 (19.1–23.4)
Past psychiatric difficulties				
Child Behaviour Checklist abnormal	12 (27)*	10 (59)	[15] 7 (47)	[32] 10 (31)
Life events in previous six months				
⩾1 death	3 (17)	7 (16)	4 (18)	11 (16)
Unemployed ⩾ one month	11 (61)	15 (33)	10 (46)	20 (30)
Break-up of relationship	1 (6)	6 (13)	5 (27)	5 (8)
Modified Social Adjustment Scale				
Housework	[15] 1.8 (1.7–2.2)***	[44] 1.3 (1.0–1.7)	1.8 (1.3–2.2)*	[64] 1.4 (1.2–1.8)
Social and leisure	[17] 2.4 (2.2–3.0)***	2.0 (1.7–2.3)	2.4 (2.1–3.2)***	1.9 (1.7–2.2)
Extended family relationships	[17] 2.6 (2.0–3.0)	2.0 (1.8–2.5)	2.7 (2.4–3.2)***	2.1 (1.7–2.4)
Overall adjustment score	2.3 (1.9–2.6)***	1.7 (1.5–2.1)	2.1 (1.9 – 2.7)***	1.8 (1.6–2.0)
Current use of alcohol and cannabis				
Alcohol	13 (72)	29 (64)	17 (77)	48 (72)
Cannabis	1 (6)	6 (13)	5 (23)	15 (22)
Current cognitive function				
Digit span forward	[17] 6 (5–9)	7 (6–8)	6 (6–8)	6 (6–8)
Digit span reverse	[17] 4 (3–7)	4 (3–6)	4 (3–5)	4 (3–5)
Immediate verbal memory	[17] 4 (3–7)*	[44] 6 (5–9)	[21] 6 (5–8)	[64] 6 (5–8)
Delayed verbal memory	[17] 6 (4–9)	[44] 9 (6–10)	[21] 9 (7–11)	[64] 8 (7–10)
Fist/flat sequence	[17] 4 (3–5)**	5 (4–6)	5 (4–6)	5 (4–6)
Immediate visual memory	[17] 4 (2–5)	4 (2–5)	5 (2–5)	5 (3–5)
Delayed visual memory	[17] 3 (2–4)	4 (3–5)	4 (3–5)	4 (3–5)
Verbal fluency	[17] 21 (19–26)	26 (21–27)	24 (21–26)	24 (21–28)
Corsi blocks	[17] 5 (4–6)	5 (5–6)	6 (4–6)	5 (5–6)
Previously measured IQ	[17] 73 (67–76)*	[39] 77 (71–85)	[11] 91 (84–99)	[24] 83 (77–92)

*$P<0.05$, **$P<0.01$, ***$P<0.001$, *v.* non-case.
1. Number of hospital admissions too small for further analysis.

assessed by the proportion with urban residences (89% SS *v.* 81% AA), employment of main breadwinner (unskilled: 53% SS *v.* 48% AA), domestic amenities (all amenities: 68% SS *v.* 63% AA) and density of persons per room (median, interquartile range 2, 1–3 SS *v.* 2, 2–3 AA) were similar in both genotypes.

Comparing some characteristics between the two genotype groups (Table 1), SS patients were less likely to have stable relationships or to have children, were more likely to have illnesses and hospital admissions, had lower BMI and a lower previously measured IQ. No differences

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Table 3 Logistic regression models for psychiatric caseness

Step	Variable	χ^2 (for improvement) d.f.=1	P	Subjects correctly classified (%)
Patients (n=61)				
0	No variables			73.8
1	Cognitive function test: fist/flat sequence	8.75	0.0031	80.3
2	Past psychiatric difficulties: Child Behaviour Checklist abnormal	4.09	0.044	81.6
Controls (n=89)				
0	No variables			75.3
1	SAS-M: extended family relationships	25.48	<0.001	79.8
2	⩾1 clinical events in previous six months	6.19	0.013	83.2

Patients – analysis also included the following variables: unemployment, leaving school early, unemployment (⩾one month in the previous six months), SAS-M social and leisure role area, immediate verbal memory, IQ and SAS-M housework role area were excluded because of small numbers.
Controls – analysis also included the following variables: gender, having children, unemployment, break-up of relationship, SAS-M social and leisure activity role area.

between patients and controls occurred in past psychiatric difficulties as measured by the CBCL, life events, SAS-M scores, use of alcohol or cannabis, or current cognitive function, apart from a modest impairment of immediate visual memory in SS patients.

Some data showed significant gender differences (all $P<0.05$). More women than men were continuing in education (29 (37%) *v.* 12 (16%)), only men admitted to criminal activity (1 (3%) SS men *v.* 8 (18%) AA men) and fewer women used alcohol (women 46 (60%) *v.* men 61 (81%)) and cannabis (women 3 (4%) *v.* men 24 (32%)). Fewer SS women than AA women had children (4 (13%) SS *v.* 21 (47%) AA).

Prevalence of psychiatric disorder

The prevalence and severity of psychiatric disorder using four criteria of morbidity did not vary between genotypes: ID level 5 (18 (29%) SS *v.* 22 (25%) AA); ID level 6 (9 (14%) SS *v.* 12 (14%) AA); ID level 5 plus DSM–III–R diagnosis of psychoactive substance abuse or dependence (19 (30%) SS *v.* 25 (28%) AA); and total GHQ score (median (interquartile range) 7 (3–12) SS *v.* 7 (5–12) AA).

Variables associated with psychiatric caseness (Table 2)

In SS disease psychiatric caseness was associated with leaving school early, past psychiatric difficulties (CBCL), low BMI in males, two tests of cognitive function (immediate verbal memory and fist/flat sequence) and previously measured IQ, but not with physical illness. In AA controls, psychiatric caseness was associated with female gender, unemployment and higher SAS-M scores for extended family relationships. In both genotypes, caseness was associated with higher SAS-M scores for housework, social and leisure activity and overall adjustment, but not with life events or psychoactive substance use.

Logistic regression models for psychiatric caseness (Table 3) indicated significant contributions from the fist/flat sequence cognitive function test and past psychiatric difficulties (CBCL) in SS disease, and SAS-M extended family relationships role area score and one or more clinical events in the previous six months in AA controls.

Low BMI in SS males

A striking observation was the association of psychiatric caseness with low BMI. All eight male SS psychiatric cases had a BMI below 17.60 compared with seven of 22 non-cases. Comparison of the features in male SS patients with BMI <17.60 and ⩾17.60 showed the lower BMI group had similar age distribution and physical illness severity to the higher BMI group, but fewer visiting or cohabiting relationships (5 (33%) low BMI *v.* 12 (80%) higher BMI; $P<0.05$) and worse scores for the SAS-M social and leisure activity role area (median (interquartile range) 2.4 (1.8–3.0) low BMI *v.* 1.8 (1.6–2.1) higher BMI; $P<0.05$) for the delayed verbal memory test (7 (5–9) low BMI *v.* 9 (8–11) higher BMI; $P<0.05$), and for previously measured IQ (73 (64–76) low BMI *v.* 83 (76–85) higher BMI; $P<0.01$). Compared with non-cases (Table 4), psychiatric cases within the low BMI group showed significantly worse scores for SAS-M social and leisure activity role area, and for cognitive function tests immediate verbal memory, delayed verbal memory and verbal fluency.

Validity coefficients for the GHQ (CGHQ scoring)

Validity coefficients were within the expected range (Goldberg & Williams, 1988; SS patients: threshold 8/9, sensitivity 88%, specificity 80%; AA controls: threshold 9/10, sensitivity 82%, specificity 90%).

DISCUSSION

The Jamaican Cohort Study provides a unique opportunity for the study of psychiatric problems in SS disease. It is a representative sample of patients, including not only the severely symptomatic, who form most of the clinic populations on whom studies are usually performed, but also mildly affected cases who might not otherwise come to medical attention. Assessing every single SS patient in the controlled part of the study has further avoided symptomatic selection. All patients and controls have been followed from birth in an identical manner, including the provision of social support.

Cultural and social factors

In the absence of locally developed Caribbean instruments for assessing psychiatric disorder, the GHQ was considered an appropriate screening instrument, having been used both in many cultural groups and in the physically ill with high sensitivity and specificity (Goldberg & Williams, 1988). The PAS was derived from the PSE for which cross-cultural validity and reliability have been established (World Health Organization, 1975).

The Modified Social Adjustment Scale has not previously been used cross-culturally, although its content appeared to be appropriate for the Jamaican Cohort Study. In addition, previous research suggested that

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Table 4 Low BMI (<17.60) male SS patients: psychiatric cases (n=7) compared with non-cases (n=7) for social adjustment and cognitive assessment

	Case	Non-case
	Median (interquartile range)	
Modified Social Adjustment scale		
Housework	1.8 (1.5–2.1)[1]	1.5 (2.2–1.2)[1]
Social and leisure	2.9 (2.4–3.8)*	1.8 (1.6–2.4)
Extended family relationships	2.6 (2.0–2.8)	2.0 (1.9–2.0)
Overall adjustment score	2.4 (2.1–2.9)*	2.0 (1.6–2.1)
Current cognitive function		
Immediate verbal memory	3 (3–5)*	7 (5–10)
Delayed verbal memory	5 (4–8)*	8 (7–12)
Verbal fluency	19 (16–25)*	24 (21–31)
Previously measured IQ	73 (52–76)	70 (66–76)[2]

1. n=6.
2. n=5.
*P<0.05, v. non-case.

scores on this instrument were not correlated with social class, age or gender and that self-assessments compared well with assessments made by informants, even among psychiatrically disturbed subjects (Cooper *et al*, 1982).

The List of Threatening Experiences has been used in different cultural groups in England (Bebbington *et al*, 1981, Brugha *et al*, 1985). In the present study, deaths of close relatives and friends were unrelated to psychiatric disorder in both genotype groups. This lack of association was unexpected as other studies have indicated strong associations between major life events and psychiatric disorder (Brown & Harris, 1978). Other factors such as religious beliefs and practice may be important in coping with difficulties. In addition, frequent positive responses to the PAS question, "Have you had the feeling that something terrible might happen?", suggested that realistic fear, of violence, road accidents, hurricanes and earthquakes, which are all relatively frequent in Jamaica, outweigh the effects of individual life events.

Formal comprehensive assessment of cognitive function was not possible with the time restraints of the present study. The small number of brief tests used aimed to be culturally appropriate and unrelated to educational attainment.

The control group

Contrary to expectations, there were high but similar rates of psychiatric disorder in both genotype groups. Control group rates considerably exceeded other estimates of psychiatric disorder in physically healthy populations (Royal College of Physicians and Royal College of Psychiatrists, 1995). This is unlikely to be a spurious result since assessment was made by a single interviewer using a standardised, structured psychiatric interview. However, these rates may not be representative of psychiatric disorder in the general population: control subjects differ from their peers by having taken part in the life-long medical follow-up study. This may have inculcated an abnormal focus on physical symptoms compared with other young people, making the association in the control group between psychiatric morbidity and clinical events difficult to interpret. Poor social circumstances and a generally urban residence may also be associated with the high rates, persistent social difficulties being associated with psychiatric disorder (Brown & Harris, 1978). Increasing rates of depression have been observed in both richer and poorer countries worldwide and have been associated with the break-up of traditional family structures, economic depression, unemployment and urbanisation (Abas *et al*, 1994), all of which may be important in the present study. The higher rate of psychiatric disorder identified among women compared with men is well recognised (Weissman & Klerman, 1977).

The sickle cell group

Rates of psychiatric disorder were lower than those identified in other studies of sickle cell disease which used self-rated instruments (Damlouji *et al*, 1982; Belgrave & Mollock, 1991; Thompson *et al*, 1992); these methods are known to give higher estimates than interview-based measures (Mayou & Hawton, 1986).

There was no association between psychiatric disorder and conventional measures of physical illness severity. This is consistent with previous observations that patients with chronic physical illness such as sickle cell disease, cystic fibrosis and carcinoma may cope better by denying their disease (Nadel & Portadin, 1977; Cowen *et al*, 1984; Greer, 1991). Use of denial may also partially account for the similar rates of psychiatric disorder in SS patients compared with controls.

Although unemployment has been reported to be associated with psychosocial distress in sickle cell disease (Barrett *et al*, 1988), it was not associated with psychiatric caseness in the present SS group. Compared with controls, both cases and non-cases among the SS group were less likely to have visiting or cohabiting relationships. This was consistent with previous observations that sickle cell patients may withdraw socially and have difficulty making social relationships (Kumar *et al*, 1976; Barrett *et al*, 1988).

Although previously measured IQ was lower in SS patients compared with controls, current cognitive function scores were similar for both groups. This is compatible with the suggestion that cognitive impairment results from cerebral damage in early childhood, and improvement may occur with time (Wasserman *et al*, 1991). However, among psychiatric cases, cognitive function was impaired in SS patients, but not in controls. Severity of psychiatric disorder is unlikely to be responsible for this as both SS and AA groups had the same distribution of cases between ID levels 5 and 6.

Low-BMI SS males

One of the most striking observations in the present study is the association of psychiatric disorder with low BMI in SS males. Although numbers are small, our results suggest that cognitively impaired males with low BMI are more likely to be psychiatrically ill than their non-cognitively impaired, low-BMI counterparts. In addition, among low-BMI males, the lack of visiting or cohabiting

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relationships and difficulty with social and leisure activities, suggest that these young men may have difficulties in interpersonal relationships with peers. These findings suggest that young men with low BMI and cognitive impairment may have psychological and social difficulties and point the way to an important area of future research.

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CLINICAL IMPLICATIONS

- High rates of psychiatric disorder were found in both SS patients and controls.
- Psychiatric disorder in young SS adults was not associated with severity of physical illness.
- Among SS males, psychiatric disorder was associated with low BMI and cognitive impairment.

LIMITATIONS

- All subjects have been part of a life-long study; this may influence psychiatric morbidity among both patients and controls.
- Social measures have not been validated in Caribbean populations and this may lead to problems in interpreting some of the data.
- There were only small numbers of SS males with low BMI.

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