British Journal of Clinical Psychology (1996), 35, 439–450 *Printed in Great Britain*

Psychological adjustment and family functioning in a group of British children with sickle cell disease: Preliminary empirical findings and a meta-analysis

Kenny Midence*
Department of Clinical Psychology, University College of North Wales, Bangor, Gwynedd LL57 2DG, Wales

Chris McManus
St Mary's Hospital Medical School, London

Peter Fuggle and **Sally Davies**
Brent Sickle Cell Centre, Central Middlesex Hospital, London

Sickle cell disease (SCD) is a family of inherited blood disorders of variable severity which have in common haemolytic anaemia, recurrent painful crises, end-organ failure and the risk of reduced life expectancy. In Britain, the condition predominantly occurs among families of African or Afro-Caribbean origin. This study examines the effects of the condition on the psychological adjustment and family functioning of 39 children with SCD and 24 control children. Participants were assessed on measures of behaviour, depression, self-esteem, IQ and reading skills, family relationships and maternal mental health. Assessment on the Family Environment Scale showed that SCD children came from families who reported more cohesion and less conflict than did the families of controls. Maternal mental health showed no significant differences between the SCD group and controls. Children with SCD showed an IQ deficit of five points, a difference that was not statistically significant. However, a meta-analysis of six studies in the literature, including this one, did show a highly significantly decreased intellectual ability. SCD children did not show significant differences from controls on measures of depression and self-esteem. However, differences in behavioural problems were found between the three groups, with the SC group showing more behavioural problems. Results of regression analyses suggest that maternal mental health is associated with children's behavioural problems.

Sickle cell disease (SCD) was recognized as the paradigm molecular disease in 1949 (Conley, 1980). The sickle gene is inherited in a mendelian recessive manner and the resulting sickle haemoglobin has a tendency to gel when deoxygenated, causing the pathognomonic sickle-shaped red blood cells. The patients suffer from chronic

* Request for reprints.

haemolytic anaemia, recurrent painful crises and end-organ failure including asplenia, stroke, avascular necrosis, chronic lung disease and chronic renal failure. The most common type of SCD, and generally the most severe, is the homozygous SS state but the interaction of sickle haemoglobin with other globin chain abnormalities also results in clinically significant, although often milder, disease, e.g. SC disease (SC). However, all types of SCD are extremely variable with respect to the frequency and severity of symptoms and pain crises (Platt *et al.*, 1991). There is a risk of sudden and unexpected death related both to infection as a result of hyposplenism and to acute vaso-occlusive events so that around 11 per cent of SCD patients do not survive to adulthood (Leikin *et al.*, 1989). There are around four million people with SCD globally (WHO, 1989) of whom over 6000 live in Britain and some 60000 in North America (Barnhart, Henry & Lusher, 1976).

Clinicians commonly express the opinion that children with SCD and their families experience more psychological and social problems than families where there is no chronic illness. Indeed, there is now a wealth of evidence indicating increased risk of psychological problems in children with chronic conditions (Lavigne & Faier-Routman, 1992; Weiland, Pless & Roghmann, 1992). However, the causal links between illness and disturbance remain unclear (Stein & Jessop, 1984), and a simple and direct relationship between chronic illness and poor psychological adjustment does not seem to exist (Midence, 1994).

Research on SCD has also provided some evidence for an increased risk of psychological maladjustment in children with SCD as compared with controls (Midence, Fuggle & Davies, 1993). However, this increased risk does not seem to be associated to illness severity (Hurtig, Koepke & Park, 1989; Hurtig & White, 1986). Furthermore, studies of intellectual ability in SCD children, which also considered aspects of severity, did not find any evidence of a significant relationship between intellectual ability and overall illness severity. For example, Fowler *et al.* (1986) found that SCD children scored 42 months below the chronological norms on a visual-motor integration test. They also scored lower than controls on several scales of the WISC-R. Ability scores were not correlated with measures of illness severity. Swift *et al.* (1989) also found that SS children scored 1 standard deviation below the controls on the subscales of the WISC–R and other measures of cognitive ability, but not on the tests of academic achievement and social competence. Scores were not significantly associated with severity measures.

The importance of the role of the family relationships in the psychological adjustment of SCD children has been supported by Moise (1986), who found a significant correlation between family cohesion and children's adjustment. Hurtig & Park (1989) found that, although SCD children were at risk of increased behavioural problems, increased family cohesion contributed to coping competence in those children. Anderson, Weitzman & McMahon (1986) also showed that families of SCD children scored higher on family integration, cooperation, optimism and psychological stability. Recent studies have also suggested that maternal mental state and family characteristics in general are associated with behavioural problems in SCD children (Thompson, Gil, Burbach, Keith & Kinney, 1993) and chronically ill children (Walker, Ortiz-Valdez & Newbrough, 1989).

In addition, children with SCD seem to be at an increased risk of neurological

impairment due to strokes and subclinical cerebral infarction (Powars, 1975). Chronic hypoxaemia of the brain, at an early age, may also interfere with normal neurological development in SCD children which may result in later cognitive impairment (Brown, Armstrong & Eckman, 1993). Such deficits have been described in children with cyanotic heart disease (Newburger, Silbert, Buckley & Fyler, 1984) and with iron deficiency (Pollitt, Soemantri, Yunis & Scrimshaw, 1985) and a similar mechanism may contribute to cognitive impairment in SCD.

The present research arose out of the World Health Organization's collaborative group on 'Psychosocial aspects of patients and their families with haemoglobinopathies'. Despite pilot studies and ongoing work in the thalassaemias, particularly in Southern Europe, there is a dearth of information on the effects of SCD on the European population affected by this condition. This study is important over previous ones because it is the first of its kind to compare differences between British children with SCD and controls, and within the SCD population. The hypotheses examined in the present study examine differences between the SCD children and the control group and any differences within the SCD group (SS and SC). It was hypothesized that British SCD children may have increased behavioural problems, lower IQ and reading skills, reduced self-esteem and increased depression, and differences in maternal mental health and family dynamics as compared to a control group. Similar differences between the SS and SC groups were also investigated.

Finally, we have conducted a meta-analysis on all available published IQ data on SCD children. Previous research on the psychological and intellectual impact of SCD in childhood is both limited and inconsistent (Midence & Elander, 1994). For example, studies have provided different results concerning the presence and nature of intellectual deficits (e.g. Chodokoff & Whitten, 1963; Fowler *et al.*, 1986; Swift *et al.*, 1989; Wasserman, Wilimas, Fairclough, Mulhern & Wang, 1991). Therefore, a meta-analysis will provide a better understanding of the differences in intellectual ability between SCD children as compared to control groups.

Method

Design

The study used a cross-sectional case–control design using a clinic sample of children with SCD aged between six and 16 years and a group of non-affected controls of similar age, sex, ethnic background and educational provision. The children with SCD were obtained from the Haematology Department Sickle Cell Register of a North London hospital and included both regular clinic attendees and children who attended the department only once. The majority of children lived in the inner city area of North London. Control children were recruited from schools attended by the SCD children and were selected randomly by identifying children of similar age, sex and ethnic background within the same year group as the SCD child, and then selecting the one nearest the SCD child on the class register.

Participants

Thirty-nine children with SCD (24 SS and 15 SC) and 24 control children were recruited to the study. It was originally envisaged to obtain a control for each child in the study but only 24 controls were obtained due to time and funding constraints. Table 1 shows characteristics of the sample. There were no statistically significant differences between the three groups with respect to age, sex, ethnic background, marital status and social class background.

Table 1. Description of cases and controls

Measures	Sickle cell disease		Controls (%)
	SS (%)	SC (%)	
Males	7 (29)	6 (40)	9 (37)
Females	17 (71)	9 (60)	15 (63)
Mean age (SD)	10.01 (3.61)	9.49 (3.47)	10.09 (3.20)
Ethnic origin			
African	12 (50)	5 (33)	13 (54)
Afro-Caribbean	12 (50)	10 (67)	11 (46)
Parental marital status			
Single	10 (41)	9 (60)	11 (46)
Married	14 (59)	6 (40)	13 (54)
Social class			
Non-manual	9 (38)	9 (60)	15 (62)
Manual	3 (12)	2 (13)	4 (16)
Unwaged (housewife, student, unemployed)	12 (50)	4 (27)	5 (22)
Age at diagnosis			
< 12 months	15 (63)	12 (80)	
13–24 months	5 (21)	1 (7)	—
> 24 months	4 (16)	2 (13)	
Hospitalization due to crises (last 12 months, and more than 1 admission)	8 (33)	3 (20)	—
School absence in last term (one week or more)	6 (25)	4 (27)	10 (42)

Note. SS = Sickle cell anaemia; SC = Sickle cell disease.

Measures

All but one of the measures used in this study are standardized instruments with well-established and satisfactory reliability and validity characteristics. The only exception is the Great Ormond Street Self-Image Profile (GOSSIP), which is a children's measure of self-esteem, and for which no reliability and validity data are currently available (Dobbs, Monck & Rowley, 1986). The other measures used in the study were as follows: behavioural problems were assessed using the Rutter Behaviour Scales (teacher and parent versions) (Rutter, 1967; Rutter, Tizard & Whitmore, 1970); depression was assessed using the Depression Self-Rating Scale for Children (DSRSC) (Birleson, 1981); the Family Environment Scale (Moos & Moos, 1986), which has three subscales of family interaction, namely expressiveness, conflict and cohesion. This measure was completed by the mothers as prime carers. Intellectual ability was assessed using the British Ability Scales (BAS) (Elliott, Murray & Pearson, 1983) and reading skills (i.e. accuracy, comprehension and rate) using the Neale Analysis of Reading Ability–Revised (Neale, 1989). Finally, mothers' mental state was assessed using the General Health Questionnaire (GHQ) (Goldberg, 1978). This measure was also completed by the mothers.

Procedure

Ethical approval was obtained from the local ethics committee, and permission for schools to assist with the selection of controls was obtained from the local education authority. Families were contacted by letter and visited at home in order to explain the purpose and procedure of the study. A second visit

was arranged in order to carry out a parent and child interview and completion of the self-report measures. Data on each SCD child was obtained from the medical records about the severity of the condition, hospital admissions due to pain crises in the previous 12 months and neurological difficulties. The research psychologist also visited each child's school in order to obtain information from teachers and to carry out the psychometric assessment of each child.

Statistical analysis

Results presented in this paper are based on all available data as results remained consistent even when the analysis was repeated only on matched pairs. It had been anticipated *a priori* that SCD children would differ from controls, and this was assessed by planned contrast in a one-way ANOVA. Other differences between groups were assessed by Scheffé's *a posteriori* test. Regression analyses were conducted on the Rutter scales as dependent variables, and the independent variables in the regression equation were maternal education, marital status, maternal mental health and employment. Significant results are presented as standardized coefficients (β). The meta-analysis of intellectual functioning was carried out using the method of Rosenthal (1991) which combines effect sizes (averaged across all independent measures of intellectual ability) within each study included in the analysis.

Results

A summary of results showing mean (SD) scores for the SCD children and controls is shown in Table 2. A one-way ANOVA did not show any difference in IQ between the three groups ($F(2,55) = 1.009$, $p = .37$), and although the SCD children showed an IQ deficit compared with controls of approximately five IQ points, the difference did not achieve statistical significance ($t(55) = 1.42$, $p = .16$, two-tailed). Children with SCD generally scored lower than controls on the subscales of the reading test, although the difference only reached statistical significance for rate of reading where the SCD children were slower ($t(47) = 2.28$, $p < .05$).

However, results of the meta-analysis, comparing 234 SCD children with 206 controls from the studies included, showed a highly significant difference ($p < .001$), the weighted effect size (Hedge's g) being -0.302 standard deviations (Table 3). The effect remains significant if the data from the present study are omitted and also if the data from the most significant study are also omitted. Using the method of Rosenthal (1991), there was no evidence for heterogeneity between studies ($\chi^2(5) = 8.134$, n.s.). Power calculations suggest that an effect of this size requires, for an 80 per cent chance of detecting a significant effect at the .05 level, that studies should have 135 cases and 135 controls, thereby explaining why all these previous small individual studies had difficulty in demonstrating the phenomenon.

Although SCD children showed higher mean scores on the Rutter scale for parents, a one-way ANOVA did not show significant differences between the three groups ($F(2,60) = .878$, $p = .42$), and no significant differences were found between the SCD and control groups ($t(60) = 1.74$, $p = .08$). However, with regard to the Rutter scale for teachers, a one-way ANOVA showed a significant difference between the three groups ($F(2,57) = 3.69$, $p < .05$), and between the SCD and controls ($t(57) = 1.90$, $p < .05$). A Scheffé's *a posteriori* comparison of all three groups with alpha = .05 showed a difference between the SC and controls with the SC group showing more behavioural problems than the other two groups. No significant differences were found between the three groups on the depression scale ($F(2,50) = 1.96$, $p = .15$), and no evidence of differences in self-esteem was found

Table 2. Means and standard deviations of SCD sample and controls on all measures

Measures	Sickle cell disease		Controls	Sig.
	SS	SC		
British Ability Scale	101.7 (16.4)	100.8 (12.4)	106 (10.3)	n.s.
Reading test (RT) accuracy	9.06 (2.79)	7.45 (2.77)	9.22 (2.41)	n.s.
RT comprehension	9.11 (2.76)	7.09 (3.39)	8.27 (1.88)	n.s.
RT rate	9.06 (2.79)	7.91 (2.17)	10 (2.56)	SCD vs. C $p < .05$
DSRSC	3.85 (2.54)	3.57 (2.38)	5.78 (5.09)	n.s.
Rutter scale (parents)	3.68 (2.79)	4.53 (3.86)	3.08 (3.29)	n.s.
Rutter scale (teachers)	1.79 (2.26)	3.66 (2.93)	1.50 (2.06)	SS vs. SC vs. C $p < .05$ SCD vs. C $p < .05$
GOSSIP	28.5 (1.8)	27.2 (3.1)	27.1 (2.8)	n.s.
FES: Cohesion	7.71 (0.81)	8.07 (1.22)	6.08 (2.61)	SS vs. SC vs. C $p < .001$ SCD vs. C $p < .001$
Expressiveness	5.87 (1.48)	5.20 (1.74)	5.13 (1.42)	n.s.
Conflict	1.45 (1.10)	1.80 (0.67)	3.83 (2.31)	SS vs. SC vs. C $p < .001$ SCD vs. C $p < .001$
GHQ (mothers)	0.71 (1.63)	1.60 (3.20)	0.83 (2.95)	n.s.

Note. SS = Sickle cell anaemia; SC = Sickle cell disease; C = Controls.

between the three groups ($F(2,51) = 1.92$, $p = .155$) or between the SCD and controls ($t(51) = 1.08$, $p = .283$).

Although the mothers of the SC group showed higher mean scores on the GHQ, there was no difference between the three groups on maternal mental health ($F(2,59) = .60$, $p = .55$), and there was no suggestion that the mothers of children with SCD showed significantly higher scores on the GHQ ($t(59) = .47$, $p = .63$). However, as the scores of the SC group were higher regarding behavioural problems and maternal mental health, a multiple regression analysis was conducted to explore any causal relationships between variables that may account for these high scores in the SC group. Results of the regression analyses suggested that behavioural problems, as reported by mothers, were predicted by maternal mental health ($\beta = .230$, $p < .05$) and mother's marital status ($\beta = .357$, $p < .05$). Results also suggested that behavioural problems, as reported by teachers, were predicted by poor maternal mental health ($\beta = .325$, $p < .05$).

Table 3. Meta-analysis of studies on intellectual functioning in SCD

Study	N_{SCD}	$N_{Controls}$	No. of measures of intellectual ability	Mean effect size (Hedge's g)	t	p
Chodorkoff & Whitten (1963)	19	19	2	−0.381	−1.174	n.s.
Fowler *et al.* (1986)	28	28	6	−0.517	−1.934	n.s.
Swift *et al.* (1989)	21	21	21	−1.018	−3.299	< .005
Iloeje (1991)	84	84	1	−0.0481	−0.312	n.s.
Wasserman *et al.* (1991)	43	30	5	−0.275	−1.156	n.s.
This study	39	24	4	−0.295	−1.137	n.s.
Total	234	206		Weighted mean = −0.302	Combined $t = -3.679$	< .0001

There were also differences between the groups with respect to family functioning with a significant difference between the three groups on the cohesion scale ($F(2,59) = 7.30$, $p < .001$) and between the SCD group and controls ($t(59) = 3.82$, $p < .001$). A Scheffé's test showed a difference between the SS and SC groups and controls respectively. Both SS and SC groups showed higher scores on the cohesion scale than controls. There was also a significant difference between the three groups on the conflict scale ($F(2,59) = 14.28$, $p < .001$) and between the SCD and control groups ($t(59) = 5.15$, $p < .001$). A Scheffé's test showed that the control group had higher conflict scores than the SS and SC groups. There was no suggestion that the families of SCD children showed different degrees of expressiveness.

Discussion

Some important issues emerge from this study. First, with respect to intellectual ability, our results suggested a non-significant trend of subtle yet clinically significant deficits on an IQ measure. Results of the meta-analysis of other published data confirmed this finding which indicated that children with SCD, in general, perform less well on measures of intellectual ability as compared with their peers. The size of this deficit has been estimated at approximately five IQ points, which would indicate a twofold increase of risk for significant learning difficulties and the need for remedial education compared with their peers.

However, it should be emphasized that the majority of children with SCD do not show intellectual deficits and function well within mainstream education. This is supported by our results with respect to reading skills where children with SCD showed only minor deficits with respect to reading skills compared with their peers. The observed intellectual deficits suggest that either anaemia or sickle damage to the brain including cerebrovascular accidents and subclinical infarction (Hindmarsh, Brozovic, Brook & Davies, 1987) may have neurological consequences in children with SCD although the available evidence does not support a clear association between markers for more severe presentation of the disorder and increased risk of intellectual deficits. Swift *et al.* (1989) reported no evidence of an age effect on IQ and suggested that the observed cognitive impairment might be a consequence of early neurological events before the age of seven. A high priority for further research is to gain a better understanding of the possible neurological mechanisms involved in order to improve outcome in future (Midence & Elander, 1994).

With respect to the other measures used, there was no evidence of increased frequency of depressive symptoms or lower self-esteem in the SCD group compared with controls. The presence of behavioural problems on the Rutter scales is of particular importance, the effect principally being behavioural problems rather than emotional problems, with the SC children showing a difference from controls, whereas the SS children were similar to controls. The mean scores of the SC group on both Rutter scales were higher than the SS and control groups. Furthermore, close inspection of individual items revealed that the SC group significantly differed from the other two groups in a number of items related to the conduct subscale (fights/quarrelsome, often disobedient, tell lies, very restless, and cannot settle to

anything). Iloeje (1991) has suggested that the presence of subtle neurological problems may also be responsible for the behavioural problems in SCD children.

Findings on the effects of chronic illness on children with mild conditions have been provided by other researchers (Bruhn, Hampton & Chandler, 1971; McAnarney, Pless, Satterwhite & Friedman, 1974; Markova, Lockyer & Forbes, 1980; Perrin, MacLean & Perrin, 1989), and different explanations have also been suggested. For example, Wright (1983) has attributed this kind of finding to the concept of overlapping, which suggests that children who are on the borderline of being healthy and having an illness are less uncertain about the group they belong to (health or illness). Garson, Benson, Ivler & Patton (1978) have also suggested that parents of children less severely affected by their condition are perceived to need less support, and this may lead to greater psychological risk in children.

Results from the regression analyses suggest that children of single mothers with poor maternal mental health are more likely to show more behavioural problems. Maternal mental health has been associated with significantly higher levels of reported child behavioural problems (Fendrich, Warner & Weissman, 1990; Hammen, Burge & Stansbury, 1990; Walker *et al.*, 1989). Thompson *et al.* (1993) also found that although SCD children's psychological adjustment was not related to illness severity, children with behavioural problems had mothers with significantly higher anxiety and depression. The researchers concluded from these findings that the overall adjustment of SCD children could be improved by reducing maternal anxiety. 'Social-ecological factors' including the social environment and family resources (Wallander, Varni, Babani, Banis & Wilcox, 1989), maternal education and marital status (Walker *et al.*, 1989), all seem to be related to maternal mental health and children's behavioural adjustment. In line with previous studies, the present findings seem to suggest that maternal characteristics may make a significant contribution to children's adjustment. The importance of the quality of the home environment and maternal mental state in the direct or indirect prediction of children's social, emotional and cognitive development has been pointed out by Wallander *et al.* (1989). Fergusson & Lynskey (1993) have also suggested that 'the weight of the evidence tends to suggest that the association between maternal depression and disruptive childhood behaviours arises largely or wholly because the social factors (social disadvantage, stress, marital problems) that give rise to increased risks of depressive symptoms in women are also independently associated with increased risks of childhood problem behaviours'.

Family relationships in the present study were marked by increased cohesiveness and reduced conflict, although it must be emphasized that the differences between the groups were self-reported rather than observed differences in family interaction. This finding is in line with a previous observation by Anderson *et al.* (1986) who also showed that families of SCD children scored higher on family integration, cooperation, optimism and psychological stability within the family. The presence of a chronic illness in the family may lead to enhanced family relations or, alternatively, it may reflect family patterns of enmeshment and overprotectiveness, which may lead to an increased resistance to acknowledging and reporting family differences and conflict. Further research will be needed to understand the mechanisms that may underlie this observation.

The present study is not without methodological difficulties. First, it would have been desirable to obtain a larger sample and a matched control for each SCD child. Second, additional comparison groups (including one selected for comparable illness-related effects), and a sibling control group would have made results more satisfactory. However, it is difficult for one clinical centre to carry out such a design with a wide variety of physical conditions (Lavigne & Faier-Routman, 1992). Variability in adjusting to chronic illness is evident in studies examining adjustment within specific conditions (Midence, 1994), and, as suggested by Thompson *et al.* (1993), future research on SCD, and chronic illness in general, needs to identify the processes that are associated with good and poor psychological adjustment, an approach already taken by these researchers in the USA. Hurtig & White (1986) maintain that investigations of differences between chronically ill children and controls do not provide any information regarding those factors associated with positive psychological functioning and adjustment. They have also pointed out the need for a within-group approach to identify specific groups of children at risk and specific variables associated with psychological maladjustment. Undoubtedly, better designed studies are needed to understand these processes, and to act on the findings to improve the lives of SCD sufferers, especially those at risk. It is hoped that this first study of SCD in the UK will encourage European researchers to move the field forward.

Acknowledgement

The authors would like to thank the two anonymous reviewers for their helpful comments.

References

Anderson, J., Weitzman, M. & McMahon, L. (1986). Psychosocial adjustment, family functioning, and sickle cell disease. *American Journal of Diseases of Children*, **140**, 297.

Barnhart, M. I., Henry, R. L. & Lusher, J. M. (1976). *Sickle Cell.* Upjohn: A Scope Publication.

Birleson, P. (1981). The validity of depressive disorder in childhood and the development of a self-rating scale: A research report. *Journal of Child Psychology and Psychiatry*, **22**, 73–88.

Brown, R. T., Armstrong, F. D. & Eckman, J. R. (1993). Neurocognitive aspects of pediatric sickle cell disease. *Journal of Learning Disabilities*, **26**(1), 33–45.

Bruhn, J. G., Hampton, J. W. & Chandler, B. C. (1971). Clinical marginality and psychological adjustment in haemophilia. *Journal of Psychosomatic Research*, **15**, 207–213.

Chodokoff, J. & Whitten, C. F. (1963). Intellectual status of children with sickle cell anemia. *Journal of Pediatrics*, **63**(1), 29–35.

Conley, C. L. (1980). Sickle cell anemia: The first molecular disease. In M. Wintrobe (Ed.), *Blood, Pure and Eloquent.* New York: McGraw-Hill.

Dobbs, R., Monck, E. & Rowley, S. (1986). Self-esteem questionnaire for children and adolescents. Dept. of Psychological Medicine, Hospital for Sick Children, Great Ormond Street, London.

Elliott, C. D., Murray, D. J. & Pearson, L. S. (1983). *British Ability Scales.* NFER-NELSON.

Fendrich, M., Warner, V. & Weissman, M. M. (1990). Family risk factors, parental depression, and psychopathology in offspring. *Developmental Psychology*, **26**(1), 40–50.

Fergusson, D. M. & Lynskey, M. T. (1993). The effects of maternal depression on child conduct disorder and attention deficit behaviours. *Social Psychiatry and Psychiatric Epidemiology*, **28**, 116–123.

Fowler, M. G., Whitt, J. K., Redding Lallinger, R., Nash, K. B., Atkinson, S. S., Wells, R. J. & McMillan, C. (1986). Neuropsychologic and academic functioning of children with sickle cell anaemia. *Developmental and Behavioural Pediatrics*, 9(4), 213–220.

on, A., Benson, R. S., Ivler, L. & Patton, C. (1978). Parental reactions to children with congenital art disease. *Child Psychiatry and Human Development*, 9(2), 86–94.
lberg, D. (1978). *Manual of the General Health Questionnaire*. NFER-NELSON.
men, C., Burge, D. & Stansbury, K. (1990). Relationship of mother and child variables to child tcomes in a high-risk sample: A causal modelling analysis. *Developmental Psychology*, 26(1), 24–30.
lmarsh, P. C., Brozovic, M., Brook, C. G. D. & Davies, S. (1987). Incidence of overt and covert urological damage in children with sickle cell disease. *Postgraduate Medical Journal*, 63, 751–753.
ig, A. L., Koepke, D. & Park, K. B. (1989). Relation between severity of chronic illness and ljustment in children and adolescents with SCD. *Journal of Pediatric Psychology*, 14, 117–132.
ig, A. L. & Park, K. B. (1989). Adjustment and coping and adolescents with sickle cell disease. *nnals of the New York Academy of Sciences*, 565, 172–182.
tig, A. L. & White, L. S. (1986). Psychosocial adjustment in children and adolescents with sickle ll disease. *Journal of Pediatric Psychology*, 11(3), 411–427.
e, S. O. (1991). Psychiatric morbidity among children with SCD. *Developmental Medicine and Child Neurology*, 33, 1087–1094.
igne, J. V. & Faier-Routman, J. (1992). Psychological adjustment to pediatric physical disorders: . meta-analysis review. *Journal of Pediatric Psychology*, 17(2), 133–157.
tin, S. L., Gallagher, K., Kinney, T. R., Sloane, D., Klug, P., Riada, W. & the Cooperative Study f Sickle Cell Disease. (1989). Mortality in children and adolescents with sickle cell disease. *Pediatrics*, 4(3), 500–508.
Anarney, E., Pless, I. B., Satterwhite, B. & Friedman, S. E. (1974). Psychological problems of hildren with chronic juvenile arthritis. *Pediatrics*, 53, 523–528.
rkova, I., Lockyer, R. & Forbes, C. (1980). Self-perception of employed and unemployed iaemophiliacs. *Psychological Medicine*, 10, 559–565.
dence, K. (1994). The effects of chronic illness in children and their families: An overview. *Genetic, Social, and General Psychology Monographs*, 120(3), 309–326.
dence, K. & Elander, J. (1994). *Sickle Cell Disease: A Psychosocial Approach*. Oxford: Radcliffe Medical Press.
idence, K., Fuggle, P. & Davies, S. (1993). Psychosocial aspects of sickle cell disease (SCD) in childhood and adolescence: A review. *British Journal of Clinical Psychology*, 32, 271–280.
oise, J. (1986). Towards a model of competence and coping. In A. L. Hurtig & C. T. Viera (Eds), *Sickle Cell Disease: Psychological and Psychosocial Issues*. Urbana, IL: University of Illinois Press.
oos, R. H. & Moos, B. S. (1986). *Family Environment Scale. Manual*. Palo Alto, CA: Consulting Psychologists Press, Inc.
eale, M. D. (1989). *Neale Analysis of Reading Ability*. NFER-NELSON.
ewburger, J. W., Silbert, A. R., Buckley, L. P. & Fyler, D. C. (1984). Cognitive function and age at repair of transposition of the great arteries in children. *New England Journal of Medicine*, 310(23), 1495–1499.
errin, J. M., MacLean, W. E. & Perrin, E. C. (1989). Parental perceptions of health status and psychologic adjustment of children with asthma. *Pediatrics*, 83(1), 26–30.
att, O. S., Thorington, B. D., Brambilla, D. J., Milner, P. F., Rose, W. F., Vichinsky, E. & Kinney, T. R. (1991). Pain in sickle cell disease: Rates and risk factors. *New England Journal of Medicine*, 325(1), 11–16.
ollitt, E., Soemantri, A. G., Yunis, F. & Scrimshaw, N. S. (1985). Cognitive effects of iron deficiency anaemia. *Lancet*, i, 158.
owars, D. (1975). Natural history of sickle cell disease—The first ten years. *Seminars in Haematology*, 12, 3.
Rosenthal, R. (1991). *Meta-Analytic Procedures for Social Research*. Newbury Park, CA: Sage.
Rutter, M. (1967). A children's behavioural questionnaire for completion by teachers: Preliminary findings. *Journal of Child Psychology and Psychiatry*, 8, 1–11.
Rutter, M., Tizard, J. & Whitmore, K. (1970). *Education, Health and Behaviour*. London: Longmans.
Stein, R. E. & Jessop, D. J. (1984). Relationship between health status and psychological adjustment among children with chronic conditions. *Pediatrics*, 73(2), 169–174.

Swift, A. V., Cohen, M. J., Hynd, G. W., Wisenbaker, J. M., McKie, K. M., Makari, G. & McKie, V. C. (1989). Neuropsychologic impairment in children with sickle cell anaemia. *Pediatrics*, **84**(6), 1077–1085.

Thompson, R. J., Gil, K. R., Burbach, D. J., Keith, B. R. & Kinney, T. R. (1993). Role of child and maternal processes in the psychological adjustment of children with sickle cell disease. *Journal of Consultant and Clinical Psychology*, **61**(3), 468–474.

Walker, L. S., Ortiz-Valdez, J. A. & Newbrough, J. R. (1989). The role of maternal employment and depression in the psychological adjustment of chronically ill, mental retarded, and well children. *Journal of Pediatric Psychology*, **14**(3), 357–370.

Wallander, J. L., Varni, J. W., Babani, L., Banis, H. T. & Wilcox, K. T. (1989). Family resources as resistance factors for psychological adjustment in chronically ill and handicapped children. *Journal of Pediatric Psychology*, **14**(2), 157–173.

Wasserman, A. L., Wilimas, J. A., Fairclough, D. L., Mulhern, R. K. & Wang, W. (1991). Subtle neuropsychological deficits in children with sickle cell disease. *American Journal of Pediatric Hematology/Oncology*, **13**(1), 14–20.

Weiland, S. K., Pless, I. B. & Roghmann, K. J. (1992). Chronic illness and mental health problems in pediatric practice: Results from a survey of primary care providers. *Pediatrics*, **89**(3), 445–449.

World Health Organization (1989). Report of the VIth Annual Meeting of the WHO Working Group on the Feasibility Study of Hereditary Disease Community Control Programmes (Hereditary Anaemia). WHO/HDP/HA/89.2.

Wright, B. (1983). *Physical Disability: A Psychosocial Approach*. New York: Harper and Row.

Received 18 November 1994; revised version received 27 June 1995