VOLUME 1

ADOLESCENT COPING WITH SICKLE CELL DISEASE: THE ROLE OF PARENTAL UNDERSTANDING

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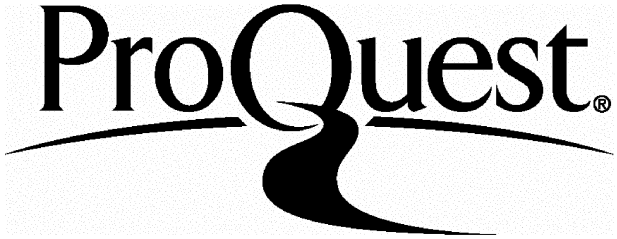

The study aimed to investigate 1) the adjustment status of adolescents with sickle cell disease 2) whether pain coping strategies were significant predictors of adjustment outcomes and 3) the contribution of parental understanding to adjustment outcomes for adolescents. Fifty-one parent-child dyads participated. The study design was cross-sectional and questionnaire-based within a structured interview format.

There was no evidence of increased psychological morbidity for adolescents with sickle cell disease when compared to population norms. Coping patterns of the sample were consistent with previous studies on adolescents with sickle cell disease. After controlling for age and frequency of pain, adolescents with high scores on Negative Thinking had more hospital admissions, more school absence, more adjustment difficulties and poorer quality of life.

The construct of parental understanding was operationalised in the study from parental ratings of adolescent coping strategies. After controlling for age and frequency of pain, parental understanding significantly predicted adolescent outcomes across the four domains of hospital admissions, school absence, adjustment and quality of life.

The findings highlight the importance of identifying parent-child patterns of communication in clinical work and strengthening the resources available within families that contribute toward successful adjustment of young people.

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Chapter One

INTRODUCTION

Overview of the Topic

Advances in paediatric medicine have meant that as well as the eradication of some common childhood illnesses, there are children surviving medical conditions that previously would have proved fatal. While there is considerable research interest in the psychological sequelae of chronic paediatric illnesses, comparatively scant attention has been paid to the impact of sickle cell disease upon children and their families. The literature suggests that it is during adolescence that children with sickle cell disease are most vulnerable to forming maladaptive coping strategies that may impair adjustment and lead to inappropriate use of health care in later life.

The present study aimed to develop a greater understanding of the coping patterns of adolescents with sickle cell disease and the relationship of these patterns to positive and maladaptive adjustment. More specifically, the study investigated the contribution of parental understanding of adolescent coping strategies to adolescent adjustment outcomes.

Conceptualising Paediatric Chronic Disease and Psychological Outcomes

As a consequence of medical advances, 'chronic disease' has become a phenomenon that presents new challenges to paediatricians. In particular, the psychological needs of children and their families demand an increasingly sophisticated approach by multidisciplinary and paediatric teams (Munson 1986). A definition of chronic disease proposed by Eiser (1990) is 'conditions that affect children for extended periods of time, often for life. These diseases can be managed to the extent that a degree of pain control or reduction in attacks...can generally be achieved. However they cannot be cured' (Eiser, 1990, p3). Generally, chronic conditions may be characterised by their protracted course and the range of diverse and adverse outcomes, from normal life expectancy to death (Bradford, 1997). Research into the prevalence of chronic childhood disorders indicates a rate of approximately 10-15 per cent in the general population (Pless and Nolan, 1991).

There is a debate within the literature about whether it is possible to develop a system of classification within chronic diseases in order to better understand the differential impact of particular diseases on outcomes, for example, child psychological adjustment (Bradford, 1997). Proposed models of classification can be broadly grouped into three areas: aetiology, disease characteristics or the severity of the disease. Fielding (1985) suggests five main causes of disease and argues that the psychological sequelae of chronic disease should be considered in relation to these causes: (i) chromosomal conditions, for example Down's Syndrome; (ii) abnormal heredity traits, for example, sickle cell or cystic fibrosis; (iii) interuterine factors or damage caused by infections or

other factors such as drug use; (iv) perinatal traumatic and infectious events, for example, damage to the central nervous system; (v) postnatal and childhood infections, mental illness and mental retardation. While this model usefully depicts the organisation of medical services, there is no conclusive evidence that the psychosocial impact of illness follows these diagnostic categories. Some studies have linked certain factors within groups of conditions to increased rates of psychopathology, for example, the increased risk of emotional and behavioural problems for children with neurological disorders (Rutter et al, 1970). But other researchers argue that a significant association between specific diseases and particular psychosocial difficulties has not been established (Pless and Perrin, 1985) and that there is not a 'psychological profile' that can be attached to a given disease.

An alternative approach to classification is the view that psychological outcomes are mediated not by the type of disease but by specific factors associated with the disease. Pless and Perrin (1985) suggest that considering outcomes in relation to the following categories may explain the variable impact of chronic disease: (i) the child's level of mobility/activity; (ii) whether the course of the disease is static or dynamic; (iii) the child's age at the time of the disease onset; (iv) whether the child's cognitive and sensory functioning is affected; (v) whether the disease is visible. Bradford (1997) notes that while this model has overall face validity it has not been subject to substantial empirical investigation, leading to inconclusive results about its application.

There has also been considerable interest in the relationship between disease severity and the impact upon child development and later adjustment in adolescence. It would seem intuitively correct to assume that the greater the extent of disease severity, the greater the corresponding degree of psychological distress and poor levels of adjustment. But the results from outcome studies across a range of conditions do not generate a consistent pattern of evidence. MacLean et al (1992) reported a linear relationship between clinical severity of childhood asthma and child adjustment but Perrin et al (1989) found children with moderate asthma to be better adjusted than children with either mild or severe forms of asthma. McArney et al (1974) found that poor adaptation in children with arthritis was more likely in children without disability compared to those children with disability. Wallander et al (1989) did not establish any link between severity of disease in cerebral palsy and spina bifida and child maladjustment.

Neither is there consistent evidence that child adjustment varies as a function of the particular disease of the child. In investigating the relationship of disease type to child adjustment, Wallander et al (1988) asked mothers of 270 chronically ill children aged 4-16 to complete the Child Behaviour Checklist (Achenbach and Edelbrock, 1983), a screening tool for emotional and behavioural difficulties. When compared against a normative group of healthy peers, the ill children did have higher behavioural and social competence problems but these adjustment problems did not vary in relation to their particular diagnosis. From a review of the literature, Pless and Perrin (1985) conclude that there is a commonality across the experience of childhood disease and that the

difficulties that children face vary only slightly from disorder to disorder, a perspective supported by other researchers (Varni and Wallander, 1988; Bradford, 1997). From the available evidence it does seem that families of children with chronic illness or physical disability do not show any marked indications of maladjustment compared to families with healthy children (Cadman et al, 1991). Family factors such as communication and quality of interaction, which have been associated with more positive outcomes among affected children, have also been shown to be related to the psychological well being of the family as a whole (Moos, 1984).

Summary of Paediatric Chronic Illness and Outcomes

Conceptualising psychological outcomes in relation to medical categories of diagnosis may have functional advantages in the organisation of clinical services to children and families. However there is no conclusive evidence that differential adjustment in children can be explained by disease type or disease severity. Overall, the balance of evidence suggests that these factors are not a reliable predictor of adolescent adjustment.

It may be more helpful to consider the specific challenges faced by children with chronic illness that are over and above the normal developmental tasks of childhood (Fielding, 1985), for example, the extent of disruption to school and family life from repeated hospital admissions or the experience of painful symptoms. These issues will be further considered in relation to what is known about the psychological sequelae of sickle cell disease, a chronic haematological disorder, for children and adolescents.

Medical Aspects of Sickle Cell Disease

Sickle cell disease (SCD) refers to a group of haemoglobin disorders and results from the inheritance of two abnormal genes related to haemoglobin formation. It affects an estimated 12,500 people in Britain, of whom over 9000 live in Greater London (Streetly et al, 1997). The majority of families affected are Afro-Caribbean and West African in origin but others are from the Mediterranean, Middle East and Asia. The disease is normally diagnosed within the first year of life and at least 150 children with SCD are born in Britain each year (Brozovic and Davies, 1987).

In this disease blood flow is obstructed in the small vessels of the venous system, causing oxygen deprivation and subsequent tissue injury. The nature and severity of this process, known as vaso-occlusion, is variable, but tissue damage is usually accompanied by pain. Patients can manage mild to moderate sickle cell pain at home but severe pain requires inpatient treatment and accounts for 91% of SCD-related hospital admissions, as opposed to other serious complications of the disease (Brozovic et al, 1987) which can include renal problems, acute chest syndrome, seizures and strokes. Phenotype of SCD is likely to be related to pain severity (Rucknagel, 1974). Although there is considerable variability, sickle cell anaemia is usually considered the most severe of clinical syndromes, followed by sickle beta thalassemia and haemoglobin SC disease respectively (Gil et al, 1989).

Until recently over 10% of children with SCD died in the first decade of life from severe infections (Brozovic and Davies, 1987) but significant advances in medical

management have reduced child mortality to approximately 1% of the sickle cell population. However, associated morbidity in children with SCD includes enuresis, delayed growth, chronic fatigue and delayed onset of puberty. Strokes can occur in children as well as adult patients. Furthermore, the severity of SCD is very variable. Some children have very few pain crises and remain symptom free whereas others may require frequent hospital admissions and experience a great deal of disruption to their daily life. Pain crises are sometimes preceded by factors such as infection, cold exposure, physical exertion or stress, but in many cases there is no apparent precipitating event.

There are at present no clinically effective methods to prevent the sickling process. Medical treatment for sickle cell crises is predominantly supportive and involves rapid analgesia, hydration and treatment of any identified triggers, such as infection and inflammatory conditions. Sickle cell disease requires medical monitoring for life and families are subject to illness-related stressors that impact upon their functioning and cohesiveness (Midence and Elander, 1994).

Psychological Aspects of Sickle Cell Disease in Children and Adolescents

Psychological aspects of SCD have received less research attention than some other chronic diseases affecting children. One probable reason for this is that SCD is less common than conditions like asthma or diabetes. Another may be linked to the patient population predominantly affected in the United Kingdom, that is, ethnic minorities, who may be marginalised in both healthcare provision and research interest. Overall,

what is known about the psychological adjustment of children and adolescents with SCD is based on a relatively small number of studies, of varying quality, few of which are based on British populations.

Review of Research Evidence

Conyard et al (1980) studied 21 adolescents in New York with SCD by obtaining ratings from social workers, parents and teachers about each child in relation to social behaviours. This study suggested a number of negative outcomes for young people, including isolation, dependence, emotional difficulties and withdrawal from peer and family relationships. Other descriptive studies include a survey of the experiences of families in Newham affected by sickle cell disease, where a range of positive and negative experiences was reported (Black and Laws, 1986).

Some evidence for psychosocial maladjustment of children with SCD has come from studies carried out in Nigeria. Akenzua (1990) used a revised version of the Paediatric Symptoms Checklist to look at emotional and behavioural disturbances in 204 children with sickle cell anaemia compared to 208 controls aged between 6 and 16 years. Thirty children with SCD (15%) scored at or above the cut-off point for detecting psychosocial problems compared with only two (1%) from the control group. Iloeje (1991) investigated psychiatric morbidity and intellectual functioning in 84 children with SCD and 84 healthy controls. No differences were found in intellectual functioning but the incidence of psychiatric morbidity in the SCD sample was significantly higher (ratings of 26% compared to 5% in the controls). In this study boys with SCD and older children

were rated higher for psychiatric morbidity. Taken together, these large-scale studies might suggest that children with SCD are at significantly higher risk than their peers for psychological maladjustment. But there is evidence that levels of emotional disturbance as measured by standardised Western questionnaires are high among the general Nigerian population (Midence and Elander, 1994). It is therefore difficult to interpret and apply these findings to a North American or European context.

Problems in adjustment for older children with SCD have been indicated in studies conducted in America. Morgan and Jackson (1986) measured body satisfaction, depression and social withdrawal in 24 young people aged between 12 and 17 with sickle cell anaemia compared against 24 healthy controls. Their results indicated that the SCD group were less satisfied with their bodies and showed more symptoms of depression and social withdrawal than the control group. The authors concluded that medical aspects of SCD such as retarded growth and delayed puberty were among factors that might predispose young people with SCD to psychological maladjustment by their interplay with the normal developmental tasks of adolescence.

However other similar types of studies have failed to find comparable differences in psychological adjustment between SCD children and healthy controls, or have found better adjustment in children with SCD. Kumar et al (1976) examined personal and social adjustment, anxiety and self-concept in 29 adolescents with sickle cell anaemia compared against a control group. No differences in personal and social adjustment were found but the SCD group showed higher scores in anxiety when compared to the

control group yet showed improved self-concept. Lemanek et al (1986) investigated behavioural characteristics of 30 SCD children and adolescents compared to healthy controls and found no significant differences. They used a battery of measures including the Behaviour Rating Profile and the Child Behaviour Checklist (completed by parents and doctors). While there was no evidence of problem behaviour among the SCD group, both groups manifested behavioural problems at the upper end of the normal range, which the authors attributed to socio-economic factors associated with poverty and minority status.

Some studies appear to suggest that SCD may be associated with subtle neurological deficits that might affect cognitive functioning. Fowler et al (1986) tested 28 children with SCD and 28 controls on the WISC-R ability scales and a visual-motor integration test. On the visual-motor integration test, the children with SCD scored 42 months below the chronological age norms for the test and showed low scores on several sub-scales of the WISC-R. While the control group performed better on both measures it was not clear from the study how to interpret the low scores of the SCD children, that is, whether these were a direct result of the illness or because of other factors such as social deprivation and poor school attendance. Differences in intelligence test performance between SCD children and controls have been identified in other studies. Swift et al (1989) looked at cognitive functioning in relation to illness severity in 21 children with sickle cell anaemia compared against 21 healthy siblings aged between 7 and 16 years. Cognitive abilities were assessed using specific subscales of the WISC-R along with other tests of memory, visual-motor skills, academic achievement and social

competence. The SCD children scored approximately one standard deviation below the controls on most of the WISC sub-scales and other measures of cognitive ability, but not on the tests of academic achievement and social competence. Test scores were not related to measures of illness severity. This suggests that SCD may be associated with some degree of cognitive impairment, even in the absence of neurological complications.

Summary of Research Evidence from Comparative Studies

There is no clear evidence for psychological morbidity in children with SCD when compared to healthy peers. There are methodological problems associated with the large-scale studies that demonstrate increased rates of morbidity in children with SCD and these findings have not been consistently replicated. Cognitive impairment has been identified, and this along with physical factors such as delayed development may indicate increased risk of adjustment problems for some children. However the evidence from studies comparing the adjustment of SCD children to healthy controls is inconclusive. It therefore seems pertinent to consider what is known about individual differences in adjustment within the child and adolescent population with SCD.

Factors Influencing Adjustment Outcomes in Children and Adolescents with SCD

Some researchers argue that the main limitation of comparative studies looking at adolescents with SCD and adjustment is that this design does not allow for the factors that might predispose children to psychological difficulties to be identified. Hurtig and White (1986) suggest that more 'within-group' studies are needed to identify those

children at higher risk and to specify factors associated with psychological maladjustment. Midence and Elander (1994) raise the question of why some young people with SCD cope well whereas others with the same condition experience social and psychological difficulties. In order to address this question, particular factors that may place the young person at increased risk must be identified, relating to SCD, characteristics of the child, their family and environment, along with the successful strategies that are used by young people for coping and adaptation.

Illness Severity, Age and Gender

Hurtig (1986) reported on a preliminary study of 12 SCD children aged between 8 and 16 as part of a five year project to assess personal and social adjustment as a function of illness variables. Measures of personality, intelligence, family and social functioning and performance at school were obtained for each child. The data suggested that for older children (over 12 years), those with more frequent pain crises, hospital admissions and visits to casualty appeared to have lower self-esteem, lower IQ, poorer social competence, more behavioural problems and a more external locus of control. The most important factor associated with poor adjustment outcomes in this study was the age at which a diagnosis was made, that is, the earlier the diagnosis was made in the adolescent's life, the poorer the overall adjustment.

However a follow-up study from the same research group using a larger sample failed to reproduce these findings. Hurtig et al (1989) looked at the relationship of illness variables to the same range of social and behavioural characteristics in 70 children with

sickle cell anaemia, aged between 8 and 16 years. These results showed that age and gender were better predictors of psychosocial adjustment than measures of illness severity. The highest level of behavioural problems and psychological maladjustment were among older boys, irrespective of the duration of illness, patterns of pain crises and levels of medical care required. Similar findings were reported by Hurtig and White (1986) in their study of 50 young people with SCD aged between 8 and 16. Measures of adjustment included the WISC-R, the revised Child Behaviour Profile and structured interviews with children and parents. They found that boys were doing generally less well than girls and that their problems were in the domains of social and behavioural adjustment, rather than intellectual or personality development.

Overall, severity of illness appears not to be reliably associated with adjustment outcomes and is not correlated with scores on WISC sub-scales (Fowler et al, 1986; Swift et al, 1989). In terms of the gender differences identified, it is possible that the traditional activities of adolescent boys may conflict more readily with the limitations imposed by SCD. Factors such as over-exertion, exposure to cold and change in temperatures have all been linked to precipitating painful sickle crises. Sport and strenuous physical activity may be more important for boys to maintain than girls and therefore boys may be more likely to 'risk' the onset of crises for the sake of participating in activities as well as suffer more adjustment difficulties from the particular restrictions SCD may bring. Moreover for boys, adolescence is a period where peer approval may be linked with prowess at sport and physical development and these are areas that may be significantly impaired for young people with SCD.

Family Environment

Factors relating to the family environment of children with SCD might be expected to play an important role in the development of successful coping. Generally it has been shown that increased cohesion in families facilitates problem-solving skills and more open expression of emotion (Minuchin et al, 1975). Studies of families affected by SCD show results that are broadly similar to findings from families and other chronic illnesses. Moise (1986) examined individual and family factors in 33 children with SCD aged 8-16 in order to identify those factors that were associated with successful adjustment. Information was obtained about the children, their family and progress at school using a battery of measures, including the Norwicki-Strickland Internal-External Scale, the Stressful Life Events Checklist and the Child Behaviour Profile. This study found that better adjusted school-aged children with SCD came from more cohesive families while those with poorer performance at school came from highly stressed families. The results also showed that better personal and social adjustment were associated with a more positive self-concept and an internal locus of control.

Hurtig and Park (1989) looked at coping in adolescents with SCD as a function of family styles and found that fewer behavioural problems among boys and greater coping competency among girls were associated with cohesive and expressive families. This supports earlier studies that suggested a child's difficulty in expressing anxiety, for example, may be due to cues picked up from parents that some topics are taboo (Whitten and Fischoff, 1974), and emphasised the importance of open discussion of feelings and concerns between parents and children with SCD.

It should also be noted that the quality of a child's family environment may be related to external factors such as housing conditions or parental employment status. Lemanek (1986) suggested that the behavioural problems found among the young people in their study could be attributed to social and economic factors associated with poverty and minority status, rather than family or SCD characteristics. Overall, very little is known about the implications of SCD for families despite the large literature on chronic illness. Midence and Elander (1994) argue that differences in family structure and dynamics, religion, formal and informal networks of support and experiences of racism mean that findings from the literature on family adaptation to chronic illness cannot necessarily be generalised across ethnic groups.

Parenting

Parents have reported feeling anxiety, guilt, overprotective impulses, and hopelessness during their children's sickle pain crises (Maxwell and Streetly, 1999). It is reasonable to assume that the presence of a chronic illness places strain on the parents' relationship although rates of divorce are not higher than among comparable families without chronic illness (Sabbeth and Leventhal 1984). In general mothers bear the main burden of care for a chronically ill child reflecting traditional patterns of childcare (Eiser, 1990). In a study of 209 children with different chronic illnesses, Jessop et al (1988) found that poor mental health among mothers was related to their perception of the severity of their child's condition, absence of emotional support and their own physical health. Problems cited by mothers of SCD children include constant worry about the

child's next pain crisis and the need to take time off work to manage their child's pain crises at home (Black and Laws, 1986).

The contribution of fathers to overall patterns of family functioning in children with SCD (and in chronic illness more generally) has not been widely investigated yet their influence may be of critical importance, as children learn strategies for coping and achieving independence. Midence and Elander (1994) point out that fathers and male partners of mothers, as well as cultural patterns of parenting, should be incorporated in any model that attempts to explain the mechanisms involved in the coping and management of SCD in families.

School

Many researchers have identified regular school attendance and participation in related activities as essential elements in positive child adjustment to chronic illness. For children with SCD however, the evidence suggests that school attendance along with participation in sports, are the activities most affected by pain crises (Walco and Dampier, 1990). Time is also missed from school because of infections, disease-related problems or the need to attend routine follow-up appointments at hospital (Shapiro et al, 1990). Fowler et al (1985) looked at school achievement and absenteeism in 270 children with chronic illnesses, some with SCD, and found that an average of 20 days had been missed from school during the previous academic year and that achievement was generally low. Shapiro et al (1990) reported that nearly half of the total days of school missed by children with SCD were due to pain crises, although most of the days were spent at home without attending hospital or clinic. Furthermore, Hurtig et al

(1989) found that the child's self-report of pain frequency was a significant predictor of school performance even after controlling for illness variables.

Apart from the risks of academic underachievement, children who are frequently absent from school may come to feel isolated from their friends and peer group and in some cases may be reluctant to return. Conyard et al (1980) found that adjustment to school and participation in school activities was poor in SCD children, especially boys. Many of these children were prone to isolation, dependence and withdrawal from relationships with peers. Hurtig et al (1989) suggest that those children who experience frequent and unpredictable pain crises may have the most difficulty in achieving optimal academic success. Older boys seem to be particularly at risk of poor school performance, more so than older girls and younger boys (Hurtig, 1986). As discussed, some studies have shown that children with SCD score lower on tests of IQ than controls (Swift et al, 1989), but links with lower attainment may also be due to the secondary effects of absence from school and lowered expectations of achievement among affected children.

Summary of Evidence: Adjustment Factors in SCD Children and Adolescents

As in studies within the general paediatric literature, severity of illness is not consistently linked to adverse adjustment outcomes for children with SCD. Older children, especially adolescent boys appear to be the group most at risk of social and behavioural adjustment problems. Cognitive factors and absence from school are also risk factors for difficulties in adjustment. Positive functioning has been associated with cohesive and expressive families but the role of parenting in adjustment is not clear.

The relationship of coping strategies to positive and maladaptive adjustment will now be considered in order to further examine the factors that are associated with differential outcomes in young people with SCD.

The Relationship of Coping Strategies to Adjustment

The coping patterns established in childhood may greatly influence that individual's coping ability and adjustment as an adult (Ross and Ross, 1984), including adult utilisation of healthcare and in the case of sickle cell management, the use of crisis hospital admissions for analgesia. Many adult patients manage pain on their own at home by increasing fluid intake, resting and taking oral analgesics (Rosse, 1983). Others cope less well and lead more limited lives, sometimes unable to work. Many of these patients are depressed, anxious and preoccupied with physical symptoms (Whitten and Fischoff, 1974), and become overly reliant on healthcare services for their pain management. There is evidence that a small percentage of the affected population consume a disproportionate amount of healthcare resources, both in Britain (Maxwell et al, 1999) and in the United States (Westerman et al, 1997).

Research with other pain populations has found that coping strategies are associated with response to pain even after controlling for disease severity, for example in patients with osteoarthritis (Keefe et al, 1987), and with chronic low back pain (Turner and Clancy, 1986). Kennedy et al (2000) examined coping in a longitudinal study of 87 adults with traumatic spinal cord injury. This study found that after controlling for functional independence, age gender and socio-economic variables, the coping

strategies utilised six weeks post-injury predicted a significant proportion of adjustment at one year post-discharge, thereby demonstrating a significant predictive relationship between coping strategies and psychological distress.

Models of Coping and Psychological Adjustment

Research into psychological well-being and coping strategies draws on a number of models which provide a theoretical framework for considering this relationship. General theories of health psychology, such as the Health Belief Model (Marteau, 1989) emphasise the importance of beliefs about the extent to which an individual is responsible for his or her health and able to take a more active role in its management. Lazarus and Folkman (1984) outlined a transactional model of stress and illness, that describes emotions as outcomes or reactions to cognitively mediated transactions within the environment. The model proposes that the individual mobilises a series of coping strategies which aim to either change a situation for the better (problem-focused coping strategies) or to regulate the level of emotional distress (emotion-focused coping strategies). Self-regulation theory (Leventhal et al, 1980) proposes that individuals reduce health threats in ways that are consistent with their illness.

Coping with illness has also been conceptualised in relation to the processing of information (Leventhal et al, 1984). This model suggests that the processing system is divided into two independent systems of coping, one concerned with the objective problems of the threat or illness, and the other concerned with emotional reactions. These systems begin to interact as the individual adapts to each situation that the

environment presents. In more recent theoretical developments, Ferguson and Cox (1997) concluded that a distinction should be made within three aspects of coping: behaviour, style and function. They propose that what individuals choose not to do as a coping strategy, along with what they choose to do, is important in defining the functional nature of coping and must be considered when examining the relationship of coping to adjustment outcomes.

Research Evidence on Coping Patterns and Sickle Cell Disease

In relation to SCD, several early studies suggested that coping strategies may be important in explaining some of the variability in adjustment to SCD pain. Denial has been associated with fewer painful episodes (Nadel and Portadin, 1977) while low levels of social and occupational activity has been associated with more frequent hospitalisation and drug treatments (Vichinsky et al, 1982). However many of the early studies that describe the adjustment of adults with SCD were limited in their methodology, as no formal measures were used to define coping abilities, disease severity was not controlled for and very small sample sizes were used.

Gil et al (1989) adopted a more rigorous methodology than previous pain coping studies with SCD adults, to test the hypothesis that coping strategies predict certain outcome measures; pain, levels of activity, healthcare use and psychological distress. The Coping Strategies Questionnaire (CSQ), a standardised measure of pain coping strategies developed for patients with low back pain (Rosenstiel and Keefe, 1983) was adapted by the Gil research group for use with sickle cell populations. The CSQ was administered

to 79 adult patients attending a sickle cell outpatient clinic along with measures of psychological distress, disease severity and the inclusion of demographic variables (Gil et al, 1989). Patients also completed a structured pain interview to assess frequency of sickle pain episodes in the previous 9 months, associated reduction of activity, use of outpatient emergency services and number of hospital admissions.

The two factors in the CSQ, Coping Attempts and Negative Thinking/ Passive Adherence, accounted for 74% of the variance in responses. Patients with high scores on Coping Attempts appeared to cope with pain in an active way, using a variety of cognitive and behavioural coping strategies. Patients with high scores on Negative Thinking and Passive Adherence, appeared to engage in negative thinking patterns, including catastrophising and self-statements of fear and anger, as well as to adhere passively to physiological pain coping methods typically recommended by physicians such as increasing fluid intake and resting.

Even after controlling for demographics and disease severity, patients high on Negative Thinking and Passive Adherence reported significantly more painful episodes. In comparison with older patients, younger patients tended to rate their pain as more severe. Patients with sickle cell anaemia had more painful episodes compared to patients with haemoglobin disease or sickle thalassemia syndromes. Patients high on Negative Thinking and Passive Adherence had significantly higher percentages of activity reduction and higher levels of psychological distress in comparison with patients low on that factor. Patients low on Coping Attempts had significantly higher percentages of activity reduction when compared to patients with high scores on this

factor. Patients high on Negative Thinking and Passive Adherence had more visits to casualty departments and more frequent hospital admissions when compared to patients with low scores on this factor, even after controlling for age, sex, number of acute events and frequency of painful episodes.

The significance of age in this study is of particular relevance. Young adults tended to rate their pain as more severe and they had significantly more casualty visits and hospitalisations. This finding is consistent with an earlier study showing a higher incidence of painful crises among young adults under 35 with SCD (Baum et al, 1987). Gil et al (1989) suggest it may be that young adulthood is a stage of life where pain episodes tend to be more frequent or in which coping with pain is particularly difficult. It is also possible that with increasing age, adults learn more effective coping strategies for dealing with pain.

Summary of Evidence: Coping Strategies and Sickle Cell Disease in Adult Patients

Positive psychological well-being appears to be associated with a range of coping strategies characterised by a tendency to positively reinterpret events and actively engage in dealing with the stressor, rather than to disengage. In relation to coping with SCD, the findings from research using the Coping Strategies Questionnaire are consistent with studies from other pain populations, that is, that pain coping strategies are significant predictors of psychosocial and physical function. After controlling for pain frequency and severity, Coping Attempts was associated with greater activity levels and reduced levels of psychological distress. Increased use of Negative Thinking

and Passive Adherence was associated with more pain, more hospital admissions, greater reduction in activity and increased psychological distress.

In adult studies, younger patients report increased pain and more healthcare use than older patients. It is a minority of adult SCD patients who rely on hospital care for pain management, a strategy that is associated with poor overall adjustment. It is therefore important to understand the process of early adjustment to the disease and how specific pain coping strategies and early adjustment may be related to subsequent adult adjustment.

Child Pain Coping Strategies and Child Adjustment

Childhood Pain in Sickle Cell Disease

Sickle cell pain, coped with at home, is a common experience for a large proportion of children and is likely to be under-reported in routine outpatient clinics (Fuggle et al, 1996). Pain in children with SCD can be measured through self-report of the child and parent (Varni et al, 1987). However there is some evidence that both child and parent tend to under-report pain and that diaries maintained by the child over a period of time give more reliable estimates (Gill et al, 1997). Estimates of the average number of painful episodes in school age children, from 7 years to adolescence, range from once every two weeks to once per month (Hurtig and White, 1986). How a child copes with pain will greatly contribute to his or her overall psychosocial adjustment (Gil et al, 1989).

While there is some literature on describing the strategies children use to cope with pain (McGrath, 1987; Ross and Ross, 1988; Siegel and Smith, 1989) there are only a small number of studies examining pain in children with SCD. Hurtig and White (1986) administered structured interviews to parents of children with SCD and found that although pain intensity tended to be similar across different age groups, healthcare use was greater in younger children.

One study examined the frequency and severity of sickle cell pain, its impact on quality of life and methods of coping for 25 children with SCD aged 6-16 years who were matched with non-affected peers (Fuggle et al, 1996). The study further aimed to determine whether children with SCD could discriminate between sickle pain and other childhood ailments. Each child was asked to complete a diary every day for four weeks, recording information about their wellbeing, common childhood symptoms such as headaches, pain events and their impact on different types of activity and the coping methods adopted. The children with SCD were asked whether their pain was related to the disease or not.

In the study, children with SCD showed similar amounts of minor ill health (excluding sickle pain) and coped in a similar way, for example talking to a parent. There was no evidence that SCD children tended to put themselves in a sick role by interpreting minor health events in a more anxious or sensitised way than control children did. They were also able to discriminate accurately between sickle related pain and other types of pain. Pains perceived to be due to SCD were significantly different from non-sickle pain in terms of location, intensity and overall severity. Sickle pain in the study sample

occurred on average once every 14 days, nearly all episodes were managed at home and increased the risk of the child not attending school by seven times. Fuggle et al (1996) conclude that systematic assessment of child pain and coping methods needs to be focussed on supporting successful pain management and child resilience in the home environment.

Child pain coping strategies were assessed by Ross and Ross (1984), who studied a sample of children with pain, including children with SCD. In this study only a small proportion of the sample reported using self-initiated strategies to cope with pain. Of the strategies reported, distraction, thought stopping and relaxation were among the most common. However Gil et al (1991) point out that combining groups of pain populations makes it extremely difficult to identify the specific strategies used by children with SCD. Studying pain coping strategies in children with SCD pain is particularly important, because of the relationship between childhood patterns of coping and later adult adjustment (Ross and Ross, 1988).

Coping Strategies in Children with SCD

The specific coping strategies that children with SCD use to cope with pain were specifically examined along with their role in relation to child adjustment, in the domain of activity reduction, psychological distress and use of healthcare services (Gil et al, 1991). The study also set out to investigate the relationship of parental pain coping to child adjustment and the relationship between parent and child pain coping strategies.

72 parent-child dyads participated in the study, making it one of the largest study samples of SCD children.

In the study, the version of the CSQ validated with an adult SCD population (Gil et al, 1989) was administered separately to each child and parent. Children were instructed to complete the CSQ for how they coped with SCD pain while parents were instructed to complete the CSQ for how they cope with other types of pain, such as headaches. Pain status, activity level, healthcare utilisation, psychological distress and child adjustment were measured along with phenotype of SCD and illness severity.

Parents reported frequent painful episodes for their child that resulted in reductions in household, school and social activities and required regular contact with healthcare services. There were no significant relationships between disease severity, pain status, activity reduction or healthcare utilisation.

Children with high scores on Coping Attempts appeared to cope with pain in an active way using a variety of cognitive and behavioural coping strategies, including diverting attention, calming self-statements and re-interpreting pain sensation. Children with high scores on Negative Thinking appeared to engage in negative thinking patterns including catastrophising and self-statements of fear and anger, as well as isolation in response to pain. Children with high scores on Passive Adherence seemed to rely on concrete coping strategies typically recommended by healthcare professionals for SCD pain management, such as increasing fluid intake and resting. There was a significant positive correlation between the child's age and use of Passive Adherence.

No significant gender differences were found or in the coping strategy factor scores between children from higher socio-economic groups compared to children from more deprived families.

After the effects of age and frequency of painful episodes were controlled for, children high on Coping Attempts had fewer casualty visits while children high on Passive Adherence had more frequent casualty visits. Age accounted for significant proportions of the variance in duration of painful episodes, where older children had longer painful episodes compared to younger children. Children high on Passive Adherence and Negative Thinking had significantly higher percentages of household activity reduction. Frequency of painful episodes and age did not account for any significant proportions of the variance in activity reduction measures.

Children and adolescents high on Negative Thinking were higher on overall levels of distress. Parents of children with more frequent painful episodes reported a higher level of internalising behaviour problems in their children. Adolescents high on Negative Thinking were more psychologically distressed in comparison to adolescents low on Negative Thinking. The strategies parents themselves used to cope with their own pain experiences were associated with their child's SCD adjustment. Parents high on Passive Adherence had children who were higher on Passive Adherence. Parents high on Coping Attempts had children who were lower on Negative Thinking.

Changes Over Time in Child Coping Strategies

The extent to which pain coping strategies measured at one point in time predicted subsequent adjustment in children and adolescents was further examined by the Gil research group (Gil et al, 1993). Eight-seven children and adolescents completed a baseline assessment of pain coping strategies using the CSQ. Seventy parents completed a structured interview assessing their child's healthcare use and activity reduction during painful episodes over the follow-up period of 9 months. Disease severity and socio-economic variables were also recorded.

There was significant positive correlations for children between baseline and follow-up scores on all three coping strategy factors, Coping Attempts, Negative Thinking and Passive Adherence. This suggests that pain coping strategies were relatively stable over time in this group. For adolescents, the correlation between baseline and follow-up scores was significant for Coping Attempts, but the correlations for Negative Thinking and Passive Adherence were not significant. There appears therefore to be less stability and more change in the adolescent group. Coping strategies used by children and adolescents predicted significant proportions of variance in their adjustment beyond that accounted for by pain frequency, disease severity and age.

Summary of Evidence: Child Coping Strategies, SCD and Adjustment

Taken together, these findings are similar to those discussed from adult SCD studies, in which adults high on Negative Thinking and Passive Adherence had lower levels of activity, more healthcare use and higher levels of psychological distress (Gil et al,

1989). Additionally, these findings are consistent with the general paediatric literature, that children using more negative self-statements were unsuccessful in dealing with painful medical procedures (Siegel and Smith, 1989). Coping strategy factors were not related to pain frequency or duration of painful episodes.

In adults with SCD patterns of pain coping appeared relatively stable over time (Gil et al, 1992), particularly true for Negative Thinking and Passive Adherence. While there was some degree of stability in coping strategies in younger children, the relationships were not as strong as those found in adults with SCD (Gil et al, 1993). Older children were higher on Passive Adherence and tended to be higher on Negative Thinking in comparison to younger children. Given that Passive Adherence is associated with more maladaptive adjustment, that is more frequent healthcare use, greater reduction in activity and higher levels of psychological distress, these findings suggest that as children pass through adolescence, they rely more on passive pain coping strategies and engage more in negative thinking patterns. Adolescence may be a period when ineffective coping strategies become entrenched (Gil et al, 1991).

Pain coping strategies in children and parents can be reliably assessed using questionnaire methods yet most of the studies on coping strategies in children have used interview procedures and behavioural observation methods (Siegel and Smith, 1989). The use of questionnaires such as the CSQ provide a systematic and simple means to assess and compare parent and child coping strategies. Agreement between parent and

child, as a contributing factor to child coping and adjustment outcomes, is now considered.

The Relationship of Parent-Child Agreement to Child Adjustment

Parental Contribution to Child Patterns of Coping

There are few studies that examine the parental contribution to patterns of child coping and adjustment in sickle cell disease. In other pain populations, coping behaviours in children have been shown to be influenced by factors such as encouragement from their mothers (Dunn-Geier et al, 1986).

Using a series of measures and structured interviews Kliewer and Lewis (1995) examined the contribution of parenting variables to the general coping processes of 39 children and adolescents with SCD. Parents reported on the coping suggestions they made to their children, their own coping strategies and family cohesion while children rated their general coping strategies and level of hope. After controlling for age, gender and disease severity, children's hope was positively associated with active coping suggestions by parents. Children's active coping was associated with a cohesive family environment. Parents who used low levels of cognitive restructuring as a coping strategy had children who used high levels of avoidance coping. These findings are consistent with the concordance reported between parent and child pain management strategies (Gil et al, 1991).

Parent-Child Agreement in Assessment of Child Psychopathology

Reports on concordance between parent and child about child psychological disorders have examined the degree of agreement on specific symptoms as well as diagnoses (Klein, 1991). Investigators have used the Kappa statistic, which controls for chance agreement, to estimate concordance between child and parent information for categorical data, and correlations for continuous measures.

One of the early studies used a structured interview schedule, to report on 307 children and adolescents aged between 6 and 16 years, most of whom were drawn from a psychiatric outpatient population (Herjanic and Reich, 1982). Children were interviewed independently of their mothers. The great majority (70%) showed very poor agreement between parent and child. There was no domain of functioning where the child and parent provided consistent information and for the most part, different symptoms were elicited from mothers and children. On the whole, mothers were more likely than their children to report conduct problems, while children were more likely to report subjective states, including physical symptoms such as headaches or dizziness.

In an inpatient Israeli adolescent population, mother-child agreement was examined in 40 interviews (Apter et al, 1989). Correlations between parent and adolescent reports on scales of depressed mood, conduct problems and thought disorder were adequate but for anxiety, concordance was low. Although agreement was high for some depressive symptoms, it was on the whole poor. Edelbrock et al (1986) evaluated 299 psychiatric outpatients and inpatients aged between 6 and 18 and their parents to assess

concordance about behavioural problems and affective/anxiety disorders. Parent-child agreement for behavioural problems was modest and for the affective/anxiety domain was very poor. In line with other studies, parents reported higher rates of conduct problems and lower frequencies of affective/anxious symptoms than child reports.

Reich et al (1982) reported better agreement for older children (12-16 years) than for a younger age-group but Klein (1991) notes that no statistical contrasts were conducted in this study. Another study using children aged between 6 and 18 years found that significantly greater agreement occurred in the 14-18 year age-range compared to 6-9 years (Edelbrock et al, 1985). No significant differences were found between other age-groups. It may be that older children are more reliable informants and therefore provide a higher rate of concordance with their parents but the findings from these studies were not replicated in other investigations (Weissman et al, 1987). No consistent significant effects of gender have been identified in studies on parent-child agreement.

Parental Awareness of Child Cognitions

The psychological adjustment of healthy siblings was investigated in relation to their cognitions about their brother's or sister's illness, to parental awareness of these cognitions and to three other parental factors; parental distress, parental social support and amount of parental care demanded by the illness (Smith et al, in press). Sixty-two well siblings and mothers of children with a range of chronic diseases completed a series of standardised questionnaires and separate structured interviews.

In this study, the level of agreement between well siblings and mothers about the siblings' cognitions varied by type. There was a high level of agreement on items concerning well siblings' attitudes to the illness or the ill child. Less agreement was apparent for items about the well siblings' reactions to family functioning. Mothers rated well siblings as having more negative cognitions about the illness than reported by the siblings themselves. Poorer sibling adjustment was associated with lower maternal awareness of their cognitions.

These findings indicate that children most at risk of poor adjustment were likely to have a parent who was less aware of their thoughts about living with the illness. It also supports models of stress and coping that suggest successful adaptation to illness is, in part, a function of the way in which the family manages the differing cognitions about the illness of each of its members (McCubbin and Patterson, 1983).

Summary of Parent-Child Agreement and Child Adjustment

There is some evidence that parent's coping suggestions to their children are associated with adjustment outcomes for children. In children with SCD children's hope was positively associated with active coping suggestions by parents. Parents with low levels of cognitive restructuring had children with high levels of avoidance coping. There are no consistent findings about the rates of parent-child concordance in reporting child psychopathology. Agreement is generally poor, even for observable behaviours, and there is no one type of information for which parent-child agreement is consistently high. Poorer adustment of children has been linked to lower maternal awareness of child cognitions. The precise way that cognitive and family factors link together to influence

child adjustment remains unclear and it seems relevant to further investigate the role of parental awareness in child adjustment outcomes.

General Aims of the Study

The present study aimed to investigate the coping patterns of British young people with sickle cell disease, the relationship of coping patterns to adjustment and to identify the previously unresearched role of parental understanding in adolescent adjustment outcomes. Parental understanding was operationalised using measures of parental agreement and of the parental ideal of child coping strategies for sickle cell pain.

The research questions were:

1. What is the adjustment status of adolescents with sickle cell disease in relation to adjustment norms?
2. Are pain coping strategies significant predictors of adjustment outcomes for adolescents with sickle cell disease living in the United Kingdom?
3. Does parental understanding contribute to differential adjustment outcomes for adolescents with sickle cell disease?

Chapter Two

METHOD

Overview

Fifty-one parent-child dyads were recruited from two paediatric outpatient clinics providing care for children with sickle cell disease. Parents and children were seen at home for the study and each asked to complete a series of questionnaires and a short interview. The parent interviews asked about parental perception of the impact of sickle cell disease upon their child's development. A version of the Coping Strategies Questionnaire (CSQ) was administered twice to the participating parent to elicit parental awareness of their child's coping strategies with sickle cell pain (Parental Agreement) and to elicit the parent's ideal view of optimal coping strategies (Parental Ideal).

Children were asked to complete the CSQ-Sickle Cell Disease Version for Children, the Strengths and Difficulties Questionnaire (SDQ) and the Generic Child Quality of Life (GCQ) measure. Child interviews asked about the child's experience of sickle cell disease, both negative and positive.

Sample

The sample consisted of 51 families with a child aged between 10 and 17 with sickle cell disease who were registered outpatients of one of two London hospital paediatric clinics. Fifty-four families were identified in total by the Consultant Paediatricians, 24 from Hospital 1 and 30 from Hospital 2. The age-range of children was selected so that children could complete questionnaires independently and to include the adolescent sickle cell population, where the literature reports the majority of adjustment problems. There were no exclusion criteria except for the requirement that the parent and child spoke English. All families were of Afro-Caribbean or Black African origin, reflecting the predominant groups in Britain affected by sickle cell disease. The families' socio-economic group was classified using an index of social deprivation, a combined rating of the educational background of the participating parent, the standard of housing and household income.

The researcher approached the two Consultants responsible for care of children with sickle cell disease and obtained consent for the study to be carried out on the patient groups. Each Consultant sent an introductory letter to all families, explaining the aims of the study and indicating that the researcher would be in contact with them shortly. This letter was followed up by telephone contact from the researcher, when more information was given about the study and questions answered. If the parent agreed to participate with their child an appointment was made and a confirmatory letter was sent with an information sheet.

Of the 54 families identified, 51 families agreed to participate in the study. Three families declined to participate, one because the parent was in hospital, one because the family were about to go abroad and the other because of a current disagreement with medical staff about the adolescent's care. Of the adolescents, 32 were girls (62.7%) and 19 were boys (37.3%). The age range of adolescents was 10-17 and the mean age was 12.96 (SD = 2.02).

Design

The design was cross-sectional and questionnaire-based using a one-group sample. A control group was not used because the aim of the study was to identify the specific factors associated with differential adjustment outcomes within a child sickle cell population.

Procedure

The interview was completed with one parent and their child in families' homes. In most cases the parent interviewed was the mother. Separating the parent and child during the interviews was initially considered, because of a concern that the child's responses may be influenced by the presence of the parent. This was explored in the pilot interviews and the conclusion was that the cost of imposing this as a condition in the interviews was too high. In some cases families were living in crowded circumstances, and so there were practical limitations on such a condition, but neither did it seem acceptable to families to ask to see the child separately. There were real concerns that this may have affected the progress of the study, in the sense of

engendering a mistrust of the interview process. In many cases the parent did in fact leave the room having completed the questionnaires before the child had finished, and the interview with the child was conducted separately. There were no differences observed in the participation of the child in the research process whether or not the parent was present.

The study took approximately one hour to complete. Parents completed a structured interview while the child began to complete the CSQ, and then completed the adapted version of the CSQ twice. Upon completion of the CSQ the child completed the Strengths and Difficulties Questionnaire (SDQ), the Generic Quality of Life measure (GCQ) and a brief structured interview. When Parent Version 1 (Parental Agreement) of the CSQ was administered, parents were asked to complete it based on what they thought their child did or thought to cope with sickle cell pain. When Parent Version 2 (Parental Ideal) of the CSQ was administered, parents were asked to complete it based on what they thought were the best things that the child should do or think to cope with sickle cell pain, as if they were advising them on the best strategy.

A pilot study with five families was conducted. The content and procedure of the questionnaires and interviews appeared to be acceptable and relevant and there were no major amendments.

Ethical Considerations

Ethical approval from the study was obtained from both hospitals (Appendix 1). At recruitment and when families were interviewed, the researcher discussed with parents and children the aims and objectives of the study. Families were advised that a summary of the findings of the study would be sent out to them and that a seminar would be hosted to present and discuss these results, to which all participating families would be invited.

Written consent was obtained from the participating parent and child on separate consent forms that also asked for verification that the study had been adequately explained to them. The information sheets and consent forms (Appendix 2) stated that families' decision to participate or not, or to later withdraw from the study, would not affect their care or treatment from the hospital in any way.

Measures

Child Coping Strategies

The coping strategies of children with sickle cell disease were assessed using the Coping Strategies Questionnaire-Sickle Cell Disease Version for Children (Gil et al, 1991). This is a scale adapted from the Coping Strategies Questionnaire (CSQ) designed for patients with low back pain (Rosenstiel and Keefe, 1983). The adapted CSQ comprises 78 statements and consists of the13 sub-scales, each with 6 items. The child rates each item on a 7 point Likert-type scale, to indicate how often they use each

strategy, where 0 = never and 6 = always. A sample item is the statement 'When I feel pain I tell myself it doesn't hurt.'

The sickle cell version has several additional sub-scales to assess cognitive, behavioural and physiological strategies particularly relevant to sickle cell disease (Appendix 3). The strategies incorporated under each coping factor, Coping Attempts, Negative Thinking and Passive Adherence are shown below in Table 1.

Table 1
Coping Strategies for Coping Domains from the Coping Strategies Questionnaire

Coping Attempts	**Negative Thinking**	**Passive Adherence**
Diverting attention	Catastrophising	Resting
Reinterpreting pain	Fear self-statements	Taking fluids
Ignoring pain sensations	Anger self-statements	Praying and hoping
Calming self-statements	Isolation	Heat/cold/massage
Increased behaviour activity		

Operationalising Parental Understanding: Parental Agreement of Coping

The CSQ-Sickle Cell Disease Version for Children was modified for the purposes of this study. 26 items were selected from the revised Coping Strategies Questionnaire, two from each of the 13 sub-scales (Appendix 4). The CSQ- Sickle Cell Disease Version for Parents (1) was rated in the same way as the child version, where parents rate each item on a 7 point Likert-type scale, to indicate how often they think their child uses each strategy, where 0 = never and 6 = always. A sample item is the statement 'when my son feels pain, he does something active, like playing outside.' The rationale for amending the version given to the parent was because this was to be administered

twice, and it was considered unlikely that completing a 78-item scale twice would be acceptable to participants.

Operationalising Parental Understanding: Parental Ideal View of Child Coping

The CSQ-Sickle Cell Disease Version for Parents (2) included the same 26 items as the first version given to parents (Appendix 4). It was administered with instructions given to parents that while the items were the same, it should be completed as if the parent was giving the child advice about the best thing to do or think when coping with sickle cell pain. Parents are asked to rate each item on a 7 point Likert-type scale, where 0 = never and 6 = always. A sample item is 'when my son feels pain he should rub the parts of his body that hurt.'

Demographic Variables

In the structured interview with the parent (Appendix 4) the following information was collected: gender of the child and participating parent; age of the child; family composition living at home; in single mother households the level of contact the child had with the father; the presence of siblings and other relatives with sickle cell disease; pre or post natal diagnosis of the participating child; a deprivation index rated from educational background of the participating parent, the standard of housing and household income; the frequency of painful episodes for the child in the previous week and month; the number of lifetime hospital admissions and the hospital attended by the child.

Strengths and Difficulties Questionnaire

Child adjustment was assessed using the Strengths and Difficulties Questionnaire (SDQ) (Goodman, 1997). The self-report version was used (Appendix 3) in order that adolescents could independently complete this measure. It is a brief behavioural screening questionnaire that asks about 25 attributes, some positive, some negative. The SDQ additionally has items that indicate positive attributes, for example 'I am helpful if someone is hurt, upset or feeling ill.' It includes items such as feeling bullied within peer relationships and asks about symptoms of hyperactivity, such as inattention and impulsivity. The child is asked to rate each item on a 3 point Likert-type scale, marked 'not true,' somewhat true' and 'certainly true.'

There are clear differences in this measure when compared against the widely used Rutter questionnaires (Rutter et al, 1970) and the Child Behaviour Checklist (CBCL) (Achenbach, 1991). For example, one difference is in length that potentially could affect the psychometric properties of the SDQ; the CBCL has 118 items on psychopathology alone. The SDQ has been evaluated against the CBCL to assess whether the brevity of the SDQ was achieved at the cost of reduced reliability and validity (Goodman, 1999). In this study mothers completed the SDQ and CBCL on 132 children drawn from psychiatric and dental clinics. Scores from the SDQ and CBCL were highly correlated and equally able to discriminate between psychiatric and dental cases.

Generic Child Quality of Life Measure

Quality of life for each child was assessed using the Generic Child Quality of Life measure (GCQ) (Collier and Mackinlay, 1995). The child responds to a set of 25 statements by circling one of five figures of a boy/girl, each shown under five ratings: always, often, sometimes, hardly ever and never (Appendix 3). The child is asked to indicate which child is 'most like you' in response to a statement. A sample item is 'A group of girls find out they are all different in how much of the time they worry about things. Tick the girl most like you.' The researcher then folds the pages of the questionnaire to reveal a new set of boy/girl figures and to conceal the completed answers. The child is asked to work through the statements again, this time indicating the child they would 'most like to be' in response to the 25 statements. The child's perception of their quality of life is measured by a calculation of the discrepancies between ratings of actual and ideal life.

The Generic Child Quality of Life measure was developed to allow comparison between chronically ill children and the general child population (Collier and Mackinlay, 1997). Many other quality of life measures suitable for use with children are disease-specific which do allow for the assessment of symptom interference and control. This advantage is less relevant where a chronic medical condition is primarily managed at home and children attend hospital for routine outpatient review, as in the case of sickle cell disease. In these circumstances an assessment of quality of life which starts with the medical condition's symptoms and treatments appears to be less suitable

than one which starts with quality of life for the general child population. The GCQ has showed an acceptable reliability (Collier and Mackinlay, 1999).

Number of Days of School Absence

In the structured interview with parents (Appendix 4) parents were asked to estimate the number of days off school the child had had in the previous year because of problems with sickle cell. The child was asked for his/her own estimate in the Appraisal measure (Appendix 3).

Paediatric Rating of Hospital Admissions

The two Consultant Paediatricians were asked to complete a form for each participating child under their care (Appendix 5). The information collected was SCD phenotype, a rating of the appropriateness of each hospital admission for pain relief in the last two years (1 = not at all appropriate and 5 = very appropriate) and the total number of hospital admissions for each child in the previous two years.

Child Appraisal Measure

At the end of the written measures the child participated in a structured interview (Appendix 3). The child was asked to specify the number of painful sickle cell episodes in the past week and month, what type of activities the pain had prevented the child from doing and the number of lifetime hospital admissions. The child was asked to think about what would be different if he/she did not have sickle cell disease and whether there was anything positive about having sickle cell disease. Children were

asked to rate how much of a difference (big/small/none) that sickle cell had made to the following areas: how the child thinks or feels about themselves, how people see them, what they could or could not do, how they got on with other children and how they got on with schoolwork.

Plan of Data Analysis

The Results chapter begins with descriptive and exploratory data analysis of the demographic and illness variables and the ratings by parent and child of the effect of SCD upon the child's life. The scores obtained from the CSQ completed by parent and child are presented with computations of parental understanding of coping strategies used by the child. The research questions are examined using correlational and multiple regression techniques.

The descriptive data section provides a summary of the mean scores, standard deviations and range of scores for measures of adolescent adjustment, comparing these to standardised norms. Statistical associations were explored between the demographic and illness variables and the adolescent adjustment outcomes using simple bivariate correlations.

Adolescent coping strategies were assessed by a summary of CSQ scores in each of the three coping factors identified in the Gil et al (1991) study; Coping Attempts, Negative Thinking and Passive Adherence. Parental understanding was operationalised by combining the measures of Parental Agreement and Parental Ideal. Parental Agreement

of child coping strategies was assessed by calculating the discrepancy between the CSQ scores of parental understanding of adolescent coping strategies and the adolescent scores in each of the three coping domains. Parental Ideal view of coping strategies that would be optimally used by their child was assessed by calculating the discrepancy between the parental ideal rating and the adolescent scores in each of the three coping domains.

Multiple regression analyses were carried out to identify those variables that predicted variance in the adolescent adjustment scores and to specifically determine the contribution of parent-child agreement in relation to adolescent coping strategies and adolescent adjustment.

Chapter Three

RESULTS

Demographic Characteristics of Participants

Table 2 presents the demographic characteristics of the study sample. Of the participating adults, 45 (88.2%) were the mother of the adolescent. Four were the father (7.8 %), three of whom were single parents, one through bereavement and two through separation. Of the remaining two carers, one was the older brother of the adolescent and one was a foster mother. High deprivation index scores (range of 3-9) indicated a high level of socio-economic deprivation, calculated from ratings of educational background of the parent, household income and quality of housing. The mean deprivation index score was 5.31 (SD = 1.29).

Table 2
Demographic Characteristics of the Study Sample

Ethnicity of Adolescents	
Black African	33 (64.7%)
Afro-Caribbean	15 (29.4%)
Mixed Race	3 (5.9%)
Age Adolescent Diagnosed with SCD	
pre-natal screening	6 (11.8%)
post-natal < one year	35 (68.6%)
one year and older	10 (19.6%)
Number of Siblings living at home	
0	11 (21.6%)
1	17 (33.3%)
2	11 (21.6%)
3	6 (11.8%)
4+	6 (11.8%)
Presence of Siblings with SCD	
siblings with SCD	18 (35.3%)
no siblings with SCD	22 (43.1%)
no siblings at home	11 (21.6%)
Family Composition	
dual parent household	17 (33.3%)
single mother/ child has contact with father	21 (41.2%)
single mother/ child no contact with father	5 (9.8%)
father single parent	3 (5.9%)
other (partner died/ foster parent/other carer)	5 (9.8%)
Deprivation Index (Maximum Range = 3-9)	
3	4 (7.8%)
4	12 (23.5%)
5	10 (19.6%)
6	15 (29.4%)
7	9 (17.6%)
8	1 (2.0%)
Clinic Attended	
Hospital 1	23 (45.1%)
Hospital 2	28 (54.9%)

n = 51

Adolescent Adjustment

The adjustment of adolescents with SCD was assessed using two measures: the Strengths and Difficulties (SDQ) measure and the Generic Children's Quality of Life (GCQ) measure.

The SDQ consists of four sub-scales measuring different problem areas and one sub-scale assessing the level of pro-social behaviours. The pro-social sub-scale was scored in the opposite direction to the other sub-scales. Table 3 summarises the scores obtained from the SDQ. The mean score obtained from the study sample was 12.16 (SD = 5.74) consistent with the range found in a normal population (Goodman, 1997) and there was a consistent pattern across the profile of sub-scale scores.

Table 3
Summary of Strengths and Difficulties Scores

	M	**SD**	**Range of Scores**
Total Difficulties Score	12.16	5.74	1-25
Sub-Scale Scores:			
Peer Problems	1.76	1.67	0-7
Conduct Problems	2.53	1.92	0-9
Hyperactivity	4.14	2.11	0-9
Emotional Symptoms	3.73	2.36	0-9
Pro-Social Behaviours	7.88	1.77	4-10

The clinical range indicating borderline or abnormal scores in a low-risk population is 17-40 for the Total Difficulties score. Table 4 compares the study sample with population norms (Goodman 1997), showing that the clinical range in SCD adolescents in the study was consistent with a normal population.

Table 4
Clinical Range of the Total Difficulties Score in the SDQ

Total Difficulties Score	**Study Sample** (n = 51)	**Population Norms**
Normal (0-16)	39 (76.5%)	80%
Borderline/Abnormal (17-40)	12 (23.5%)	20%

Adolescent adjustment was also measured using the total score obtained from the GCQ measure. Quality of life was assessed from two ratings by the adolescent; a rating of the current perception of their lives based on a series of issues presented in the questionnaire and a rating of how they would like their life to be. The discrepancy between the rating of each item's actual and preferred score (actual self and preferred self) was calculated to give a total quality of life score for each adolescent. Discrepancies were not given a negative or positive value as it was the absolute size of the discrepancy that was of interest, not a directional value. Scores were transformed so that a high score indicated a high quality of life. The mean total quality of life score obtained was 75.27 (SD = 10.79). The range of scores obtained was 44-98 from a maximum score of 100.

Because the GCQ measure has less supporting data regarding its reliability and validity than the SDQ, the scores obtained in the present study were compared against those found in 720 children drawn from a UK school population, aged 6-14 (Collier and MacKinlay, 1999). The mean Quality of Life score that this population obtained was 74.18 (SD = 9.94) which is similar to the present sample.

Adolescent Pain Status and Use of Healthcare Services

Adolescent pain frequency and use of hospital services was assessed by information collected from the adolescent, the parent and the consultant paediatrician.

Adolescents and parents were asked about the number of lifetime hospital admissions for SCD pain and the frequency of SCD pain that adolescents had experienced in the previous week and month. Table 5 summarises the data from adolescent and parental report. There was a broad consensus between parent and adolescent report of adolescent pain status.

The majority of adolescents reported either few or very frequent hospital admissions for SCD pain. A range of pain frequency was reported, from no pain at all to pain episodes every day. However 38 adolescents (74.5%) reported either no pain or one episode of pain over the previous week. Because of the high parent-child concordance the adolescent estimate for painful episodes in the previous month was the variable used in subsequent analyses as the measure of pain frequency.

Adolescents were further asked about whether their recent experience of SCD pain had affected their normal level of activities. Twenty-eight adolescents (54.9%) reported a reduction in normal activities caused by recent experience of SCD pain. Twelve adolescents (23.5%) reported no reduction in their normal levels of activity while 11(21.6%) had not experienced any painful episodes within the previous week or month.

Table 5
Summary of Adolescent and Parental Report of Adolescent Pain and Healthcare Use

	Adolescent Report	Parental Report
Number of painful episodes previous week		
0-1	38 (74.5%)	41 (80.4%)
2-3	9 (17.7%)	5 (9.8%)
4+	4 (7.8%)	5 (9.8%)
Number of painful episodes previous month		
0-3	35 (68.7%)	38 (74.5%)
4-6	10 (19.5%)	8 (15.7%)
7+	6 (11.8%)	5 (9.8%)
Number of lifetime hospital admissions for pain relief		
0-2	16 (31.4%)	16 (31.4%)
3-4	13 (25.5.%)	7 (13.7%)
5-10	6 (11.8)	9 (17.6%)
11+	16 (31.4%)	19 (37.3%)

n = 51

The consultant paediatrician responsible for the adolescent's care was asked to specify SCD phenotype, the number of hospital admissions in the previous two years, rate the appropriateness of these admissions and rate the overall appropriateness of the adolescent's use of outpatient services. Table 6 summarises the data from paediatric report of adolescent healthcare use. The majority of adolescents had sickle cell anaemia (SS), the homozygous form of SCD that has a highly variable clinical picture. SC disease (SC) is far less common than sickle cell anaemia in the UK.

The number of hospital admissions ranged from 0 to 21 but 31 adolescents (60.8%) had one or no admissions in the previous two years. Paediatric ratings of appropriateness of adolescent healthcare use were on a scale of 1 (not at all appropriate) to 5 (very appropriate). Twenty-eight adolescents (54.9%) had not been admitted to hospital for

SCD pain in the previous two years so ratings were given for the remaining 23 adolescents (45.1%). Of these, 19 adolescents (82.6%) were given ratings of 4 or 5, indicating high paediatric agreement with the appropriateness of hospital admissions for pain relief. In relation to use of outpatient clinic services, paediatric ratings indicated less satisfaction with the appropriate use of medical care, where 17 adolescents (33.4%) were given a rating of 1 or 2. Comments provided about low ratings most commonly indicated concerns about high rates of defaulting outpatient clinic appointments.

Table 6
Paediatrician Report of Adolescent Healthcare Use

	Adolescents
SCD Phenotype	
Sickle Cell Anaemia	45 (88.2%)
SC Disease	5 (9.8%)
Sickle Thalassaemia	1 (2.0%)
Hospital admissions for pain relief in previous two years	
0	28 (54.9%)
1-2	8 (15.7%)
3-5	7 (13.7%)
6-10	5 (9.8%)
11+	3 (5.9%)
Paediatric rating of appropriateness of admissions (1 = not at all appropriate 5 = very appropriate)	(n = 23)
1	0 (0%)
2	2 (8.7%)
3	2 (8.7%)
4	11(47.8%)
5	8 (34.8%)
Paediatric rating of appropriate use of outpatient services (1 = not at all appropriate 5 = very appropriate)	
1	3 (5.9%)
2	14 (27.5%)
3	6 (11.8%)
4	16 (31.4%)
5	12 (23.5%)

n = 51

Adolescent Appraisal of the Impact of SCD

In a brief structured interview, adolescents were asked to rate their perception of the effect of SCD upon the following areas; their self-image, their perception of how others saw them, the types of activities affected, friendships and their progress at school. Adolescents were asked to say whether SCD had made a big, small or no difference to these areas (Table 7). The area of least concern for adolescents was the effect of SCD on friendships where the majority felt SCD had no effect on this area. However some adolescents said that peers at school thought SCD was infectious and that this had caused them distress. The most problematic area was the effect on activities, with sport and outdoor recreation most frequently cited as limited by SCD. Half the sample indicated that SCD affected the way they felt about themselves.

Table 7
Adolescent Appraisal of SCD Effects

	Big	**Small**	**None**
Self-Image	8 (15.7%)	17 (33.3%)	26 (51.0%)
Perception of Others' View	5 (9.8%)	22 (43.1%)	24 (47.1%)
Types of Activities	17 (33.3%)	27 (52.9%)	7 (13.7%)
Friendships	1 (2.0%)	4 (7.8%)	46 (90.2%)
School Progress	14 (27.5%)	13 (25.5%)	24 (47.1%)

Adolescents were also asked to specify whether there were any positive aspects of living with SCD. Thirty-seven adolescents (72.5%) said there was nothing positive about living with SCD while 14 (27.5%) said there were some positive differences. Of

the positive differences cited, these included the experience of dealing with doctors and hospitals increasing self-confidence; that sickle cell community groups had arranged holidays and social activities for families affected by SCD and some adolescents felt that SCD had given them an increased understanding/sensitivity about other people who are sick or disabled.

Parental Appraisal of the Impact of SCD

Parents were asked to rate the effect of SCD (big, small or none) upon their child's self-image, personality, activities and friendships (Table 8). As reported by the adolescents, parents rated the area of friendships as the least problematic and reported that the greatest impact of SCD was upon their child's activities.

Parents were also asked to rate their level of concern regarding the effect of SCD upon their child's progress at school. Twenty-one parents (41.2%) said they were very concerned, citing reasons such as school absence and worry that SCD was an added pressure on their child, affecting their ability to concentrate on schoolwork. Fifteen parents (29.4%) said they were fairly concerned while 15 parents said that they were not at all concerned about their child's progress.

Table 8
Parental Appraisal of the Impact of Sickle Cell Disease

	Big	**Small**	**None**
Self-Image	10 (19.6%)	14 (27.5%)	27 (52.9%)
Personality	16 (31.4%)	15 (29.4%)	20 (39.2%)
Types of Activities	23 (45.1%)	18 (35.3%)	10 (19.6%)
Friendships	4 (7.8%)	8 (15.7%)	39 (76.5%)

Parents were also asked to give an overall rating on the effect of SCD on their child's life, from presenting few problems (1) to being a very significant problem (10) for their child. The mean parental rating was 5.78 (SD = 2.79). Twenty-six parents (51%) gave a rating of between 7 and 10.

School Absence

The impact of SCD upon school attendance was assessed by parental report of the number of days absent as a result of SCD related problems in the previous twelve months. There was a wide variation in school attendance with the number of absent days ranging from 0-139. The mean number of days absent was 19.2 (SD = 27.87) but over half the sample (56.9%) had been absent ten days or less from school in the previous year. Thirty-three (64.7%) parents cited managing their child's SCD pain at home as the main reason for school absence compared to 9 families (17.6%) where the adolescent was admitted to hospital. Nine adolescents had no school absence caused by SCD in the previous 12 months. Table 9 summarises the data on school attendance.

Table 9
Parental Report on SCD Related School Absence

Number of Days Absent from School in 12 Months	**Adolescents (n = 51)**
0-5	18 (35.3%)
6-10	11 (21.6%)
11-20	10 (19.6%)
21+	12 (23.5%)

Adolescent Coping Strategies

Adolescent CSQ Scores

The mean ratings for the Coping Strategies scores completed by the adolescents are presented in Table 10. These results are also compared to CSQ scores obtained by a sample of 72 SCD African-American children in North Carolina (Gil et al, 1991). The American sample was aged between 7 and 17 years with a mean age of 11.1 years (SD = 3.3), 25 of whom were in the age-range of 13-17. Researchers based at the recruiting hospital developed the use of the CSQ for children and adults with SCD and have reported a number of studies from these populations.

The researchers reported mean scores for each component of the coping domain, for example, diverting attention, calming self-statements (Coping Attempts), catastrophising and fear self-statements (Negative Thinking). The overall mean scores given in Table 10, taken from the Gil (1991) data were calculated by multiplying the mean scores reported by the number of component items in the CSQ and then summed for each coping domain. Overall, the mean scores show a broadly similar pattern, although the Coping Attempts scores are higher in the present sample.

Table 10
Comparison of CSQ Scores between Study Samples

	Study Sample M	**Gil et al (1991) M**
Coping Attempts	92.0	72.84
Negative Thinking	57.02	45.84
Passive Adherence	88.71	81.70

The 26 items that correspond to the CSQ Parent Version were then examined. Scores were compared to assess whether scores obtained from the adapted shorter version corresponded to those obtained from the 78 item CSQ (Table 11). Adolescents rated on a scale of 0 (never) to 6 (always) how often they used each strategy to cope with SCD pain. High scores indicate that the adolescent utilised that domain of coping frequently.

Table 11
Comparison of Adolescent CSQ Scores on the 78 item Version and 26 item Version

	CSQ 78 Item Version			CSQ 26 Item Version		
	M	**SD**	**Range of Scores**	**M**	**SD**	**Range of Scores**
Coping Attempts	92.0	29.96	19-150	28.47	10.36	4-48
Negative Thinking	57.02	23.70	12-114	18.98	8.34	2-39
Passive Adherence	88.71	14.73	54-113	33.78	5.59	21-45

To examine the concordance between the child version of the CSQ (78 items) and the items selected for the parent version (26 items) simple bivariate correlations were performed (Table 12). Because there were strong correlations found in the two sets of scores for all the coping domains of Coping Attempts, Negative Thinking and Passive Adherence, the scores obtained from the 26 items in the child CSQ were used for the subsequent analyses. There was also a positive correlation found between the domains of Coping Attempts and Passive Adherence.

Table 12
Correlation of Adolescent CSQ scores on the 78 item Version and 26 item Version

	Coping domain (26 item)		
Coping domain (78 item)	Coping Attempts	Negative Thinking	Passive Adherence
Coping Attempts	.928**	.093	.470**
Negative Thinking	.017	.907**	.058
Passive Adherence	.455**	.175	.837**

**p <0.01

Parental CSQ Scores: Adolescent Coping Strategies

Table 13 compares adolescent scores obtained from the CSQ with Parental Agreement scores obtained from the CSQ. Parental Agreement was measured from the rating given by the parent for each item, where they indicated to what extent they thought their child used each strategy to cope with sickle pain. Comparison of mean scores indicates that parental agreement ratings showed a similar pattern across coping domains to the ratings given by adolescents.

Table 13
Parental Agreement CSQ Scores on Adolescent Coping Strategies

	Parental Agreement of Current Coping		**Range of Scores**	**Adolescent Scores**		**Range of Scores**
	M	**SD**		**M**	**SD**	
Coping Attempts	24.02	10.51	6-50	28.47	10.36	4-48
Negative Thinking	20.92	10.68	0-44	18.98	8.34	2-39
Passive Adherence	32.61	8.11	12-47	33.78	5.59	21-45

Table 14 compares adolescent scores obtained from the CSQ with Parental Ideal scores obtained from the CSQ. Parental Ideal was measured from the rating given by the parent on each item that reflected their opinion on the best type of strategy to deal with SCD pain. Comparison of mean scores indicates that parental ideal ratings showed a similar pattern across coping domains to the ratings given by adolescents.

Table 14
Parental Ideal CSQ Scores on Adolescent Coping Strategies

	Parental Ideal View of Coping		**Range of Scores**	**Adolescent Scores**		**Range of Scores**
	M	**SD**		**M**	**SD**	
Coping Attempts	33.12	10.29	8-51	28.47	10.36	4-48
Negative Thinking	10.31	7.93	0-42	18.98	8.34	2-39
Passive Adherence	39.18	6.05	25-48	33.78	5.59	21-45

Operationalising Parental Understanding

Parental Agreement of child coping strategies was computed by subtracting the total number of adolescent scores for each of the coping domains from the parental scores for each of the coping domains. Scores were not given a positive or negative value as it was the absolute size of the discrepancy that was of interest, not a directional value. Low scores indicated a high consensus between parent and child while high scores indicated a low level of parental agreement. Table 15 presents the scores obtained for the difference between parental and adolescent view of adolescent coping strategies (Parental Agreement).

The discrepancy between the parent's ideal view of coping strategies and their child's actual coping strategies was computed by subtracting the total number of adolescent scores for each of the coping domains from the scores obtained for the parental ideal view of coping strategies. Low scores indicated a high consensus between adolescent coping strategies and the parental ideal view, while high scores indicate a large discrepancy between adolescent coping strategies and the parental ideal view. Table 15 shows the scores obtained for the discrepancy between adolescent coping strategies and the parental ideal (Parental Ideal).

Table 15
Mean Differences between CSQ Parental and Adolescent Scores

	Parental Agreement		Range of Scores	Parental Ideal		Range of Scores
	M	SD		M	SD	
Coping Attempts	10.41	7.10	1-33	11.98	8.66	0-37
Negative Thinking	10.14	6.64	0-26	11.18	7.95	0-34
Passive Adherence	7.14	5.89	0-28	8.65	5.52	0-23

Subset of Adolescents in Borderline/Abnormal Range of Total Difficulties Score

The data obtained from the SDQ measure suggest that the majority of adolescents with SCD in the study were no different from a normal population, indicating that SCD of itself does not contribute significantly to adjustment. Further analyses were conducted on the group of 12 adolescents (23.5%) who scored in the borderline/abnormal range of

clinical functioning to compare pain frequency, school absence, hospital admissions and quality of life (Table 16).

Table 16
Comparison between SDQ Borderline/Abnormal Subset and Normal Adjustment Group

	SDQ Borderline/Abnormal Group (n = 12)	SDQ Normal Adjustment Group (n = 39)
Mean Quality of Life Score (SD)	67.67 (11.45)	77.62 (9.55)
Number of days absent from school in previous 12 months		
0-5	3 (25.0%)	15(38.5%)
6-10	4 (33.3%)	7 (17.9%)
11-20	2 (16.7%)	8 (20.4%)
21+	3 (25.0%)	9 (23.2%)
Number of painful episodes previous month		
0-3	8 (66.7%)	27 (69.2%)
4-6	3 (25.0)	7 (17.9%)
7+	1 (8.3%)	5 (12.9%)
Hospital admissions for pain relief in previous two years		
0	3 (25.0%)	25 (64.1%)
1-2	5 (41.7%)	3 (7.7%)
3-5	2 (16.7%)	5 (12.8%)
6-10	1 (8.3%)	4 (10.2%)
11+	1 (8.3%)	2 (5.2%)
Paediatric rating of appropriateness of admissions (n = 9) (1 = not at all appropriate 5 = very appropriate)		(n = 14)
1	0 (0.0%)	0 (0.0%)
2	2 (16.7%)	0 (0.0%)
3	1 (8.3%)	1 (7.1%)
4	4 (33.3%)	7 (50.0%)
5	2 (16.7%)	6 (42.9%)
Paediatric Rating of appropriate use of outpatient services (1= not at all appropriate 5 = very appropriate)		
1	1 (8.3%)	2 (5.1%)
2	5 (41.7%)	9 (23.1%)
3	1 (8.3%)	5 (12.8%)
4	3 (25.0%)	13 (33.3%)
5	2 (16.7%)	10 (25.7%)

Overall there was a similar pattern of findings between the SDQ subset and the whole sample in school absence, pain frequency, hospital admissions and paediatric rating of admissions. However, the Quality of Life score was lower for the subset. More adolescents in the subset received very low ratings by paediatricians for their use of outpatient services, indicating inappropriate healthcare use. Half the subset were given a rating of 1 or 2 compared to just over one quarter of the normal adjustment group.

Adolescent coping scores, parental agreement and parental ideal scores from the CSQ were then compared. Table 17 compares the mean scores (and standard deviations) obtained in the three coping domains between the SDQ subset and the normal adjustment group.

Table 17
CSQ Scores of SDQ Borderline/Abnormal Subset and Normal Adjustment Group

	SDQ Borderline/Abnormal Group (n = 12)		**SDQ Normal Adjustment Group (n = 39)**	
Adolescent Coping	**M**	**SD**	**M**	**SD**
Coping Attempts	26.92	(11.84)	28.95	(9.98)
Negative Thinking	25.17	(5.72)	17.08	(8.14)
Passive Adherence	33.67	(3.75)	33.82	(6.08)
Parental Agreement				
Coping Attempts	13.33	(11.40)	9.51	(5.02)
Negative Thinking	10.67	(7.76)	9.97	(6.36)
Passive Adherence	5.17	(3.66)	7.74	(6.33)
Parental Ideal				
Coping Attempts	11.58	(8.76)	12.10	(8.74)
Negative Thinking	17.25	(8.42)	9.31	(6.89)
Passive Adherence	6.75	(5.79)	9.23	(5.38)

The pattern of CSQ scores obtained between the SDQ subset and the normal adjustment group was similar. In adolescent coping scores, the SDQ sub-group showed higher scores in the domain of Negative Thinking. The pattern of parental understanding scores was similar between the two groups. In the parental agreement scores, lower parent-child agreement was found in the SDQ subset in the domain of coping attempts. In the parental ideal scores the SDQ subset obtained higher scores in the domain of negative thinking, indicating a greater discrepancy between adolescent coping and the parental ideal.

Coping Strategies in Adolescents

The relationship between adolescent coping strategies and demographic variables and each of the outcome measures was examined using simple bivariate correlations.

Demographic Variables and Coping Strategies

No significant correlations between CSQ scores, age, ethnicity, age of diagnosis, deprivation, family composition or number of siblings were found. There was a positive correlation between gender and use of Negative Thinking (r = .298, p < .05), where girls were more likely to use this factor more than boys. There was a negative correlation between Quality of Life and the presence of siblings with SCD (r = -.292, p < .05).

Pain Frequency, Hospital Admissions and Coping Strategies

Because the parent and child report of painful episodes had high concordance, the child estimate of painful episodes in the previous month was used as the dependent variable in the calculation. There was a negative correlation between pain frequency and the use of Coping Attempts (r = -.373, p < .01), where adolescents with high scores on this factor reported less painful episodes.

The relationship of the number of hospital admissions for pain relief to coping strategies was examined using the paediatrician report of hospital admissions in the previous two years as the dependent variable. There was a positive correlation between the number of hospital admissions and the use of Negative Thinking (r = .351, p < .05) where adolescents with high scores on this factor had more hospital admissions according to hospital records.

School Absence and Coping Strategies

A positive correlation was found between the extent of school absence and Negative Thinking (r = .323, p <. 05) where a higher number of school days missed was associated with high scores on Negative Thinking. No other correlations were found between school absence and coping strategies.

Strengths and Difficulties and Coping Strategies

A positive correlation was found between the Total Difficulties score and Negative Thinking (r = .496, p < .01). No other correlations were found between the SDQ and coping scores.

Quality of Life and Coping Strategies

A negative correlation was found between Quality of Life score and Negative Thinking (r = -.492, p < 0.01) where high ratings obtained on Quality of Life correlated with low scores on Negative Thinking. A positive correlation was found between Quality of Life and Coping Attempts (r = .292, p = 0.05). No other correlations were found between Quality of Life and coping strategies.

Relationship between Outcome Variables and Measures of Parental Understanding

The relationship between hospital admissions, school absence, SDQ, Quality of Life and the measures of parental understanding was examined using simple bivariate correlations.

Hospital Admissions and Parental Understanding

No significant correlations were found between the number of hospital admissions in the previous two years and the measures of parental understanding.

School Absence and Parental Understanding

A significant positive correlation was found between the number of days missed at school because of problems with SCD and Parental Agreement in the domain of Coping Attempts ($r = .320$, $p < .05$). Low parental scores, indicating high level of agreement were associated with a lower number of days absent from school. No other significant correlations were found.

Total Difficulties Score and Parental Understanding

A positive correlation was found between the Total Difficulties score from the SDQ and Parental Ideal in the domain of Negative Thinking ($r = .481$, $p < .01$). High discrepancy scores were associated with higher Total Difficulties scores. No other significant correlations were found.

Quality of Life and Parental Understanding

A negative correlation was found between the Quality of Life score and Parental Ideal in the domain of Negative Thinking ($r = -.515$, $p < 0.01$). No other significant correlations were found.

Predicting Hospital Admissions, School Absence, Adjustment and Quality of Life from Adolescent Pain Coping Strategies and Measures of Parental Understanding.

A series of hierarchical regression analyses was conducted for each of the outcome variables in order to assess the relation between adolescent adjustment, adolescent coping strategies and the contribution of parental understanding about coping strategies to adjustment.

Predictor variables were entered in three steps; a) child age and child report of pain frequency in the previous month, b) adolescent CSQ scores and c) parental agreement of their child's coping strategies (Parental Agreement). The analysis was repeated for steps a) and b) and then the discrepancy between adolescent coping and the parental ideal view of coping was entered (Parental Ideal). Table 18 summarises the results for the regression analyses.

Table 18
Summary of Regression Analyses for Hospital Admissions, School Absence, SDQ and Quality of Life with Parental Understanding Variables Entered Separately

	R2	R2 change	F for R2 change	Overall F value
Hospital Admissions				
Age and Pain Frequency	.128	.128	3.524	3.524*
Coping Strategies	.294	.166	3.524	3.746**
Parental Agreement	.327	.033	.689	2.552*
Parental Ideal	.302	.008	.160	2.271*
School Absence				
Age and Pain Frequency	.339	.339	12.297	12.297***
Coping Strategies	.427	.089	2.323	6.719***
Parental Agreement	.522	.094	2.765	5.730***
Parental Ideal	.503	.075	2.112	5.303***
Total Difficulties Score				
Age and Pain Frequency	.000	.000	.006	.006
Coping Strategies	.294	.294	6.235	3.744**
Parental Agreement	.402	.108	2.538	3.532**
Parental Ideal	.380	.086	1.951	3.220**
Quality of Life				
Age and Pain Frequency	.020	.020	.496	.496
Coping Strategies	.375	.354	8.498	5.390**
Parental Agreement	.483	.108	2.925	4.898***
Parental Ideal	.433	.059	1.445	4.011**

*p<.05 **p<.01 ***p<.001

NB Parental Agreement was entered after (i) age and pain frequency and (ii) coping strategies.
Parental Ideal was entered after (i) age and pain frequency and (ii) coping strategies.
The degrees of freedom for the F value were 8,42.

Parental Ideal scores were then added to the regression analyses, after age, pain frequency, CSQ scores and Parental Agreement were entered. It can be seen from Table 19 that Parental Ideal was positively correlated with the outcome variables but did not contribute significantly to the overall regression equation in the absence of Parental Agreement.

Table 19
Summary of Regression Analyses for Hospital Admissions, School Absence, SDQ and Quality of Life with Parental Understanding Variables Entered Sequentially.

	R2	**R2 change**	**F for R2 change**	**Overall F value**
Hospital Admissions				
Age, Pain, CSQ & Agreement	.327	.327	2.552	2.552*
Parental Ideal	.330	.002	.048	1.742
School Absence				
Age, Pain, CSQ & Agreement	.522	.522	5.730	5.730***
Parental Ideal	.573	.051	1.541	4.749***
Total Difficulties Score				
Age, Pain, CSQ & Agreement	.402	.402	3.532	3.532**
Parental Ideal	.421	.019	.428	2.580*
Quality of Life				
Age, Pain, CSQ & Agreement	.483	.483	4.898	4.898***
Parental Ideal	.493	.010	.269	3.450**

* p<.05 ** p< .01 ***p< .001

NB Parental Ideal was entered after (i) age and pain frequency (ii) coping strategies and (iii) parental agreement.
The degrees of freedom for the F value were 11,39.

Chapter Four

DISCUSSION

Overview of Main Findings

The study set out to identify the coping patterns of adolescents with sickle cell disease and to establish the relationship of these patterns to positive and maladaptive adjustment. The contribution of parental understanding to adolescent adjustment was examined using the measures of Parental Agreement and Parental Ideal obtained by parental ratings of child coping strategies.

The study showed five main findings.

1. The majority of adolescents living with sickle cell disease were no different from a healthy population when compared to adjustment and quality of life norms.

2. The pattern of coping scores obtained from the Coping Strategies Questionnaire were broadly similar to findings from an American study sample of SCD adolescents, although the present sample showed higher scores on Coping Attempts.

3. The contribution of adolescent coping styles to outcome measures varied across the domain of coping. Adolescents who showed high scores on Negative Thinking had more hospital admissions, increased school absence, more adjustment difficulties and a

poorer quality of life. Adolescents with high scores on Coping Attempts reported less painful episodes and an improved quality of life.

4. The construct of parental understanding was operationalised by measuring the discrepancy between adolescent coping and parental agreement of adolescent coping, and the discrepancy between adolescent coping and the parental ideal of coping. The contribution of parental understanding also varied across coping domain. In the domain of Coping Attempts, high Parental Agreement scores were associated with decreased school absence. In the domain of Negative Thinking, high Parental Ideal scores were associated with more adjustment difficulties and a poorer quality of life.

5. Taken as a whole, parental understanding significantly predicted adolescent outcomes across the four domains of hospital admissions, school absence, adjustment and quality of life, after age and pain frequency were controlled for.

Research Question 1: What is the adjustment status of adolescents with sickle cell disease in relation to adjustment norms?

From the study, it is clear that maladjustment is not an inevitable outcome for adolescents living with SCD. The study population taken as a whole mirrored both adjustment norms and ratings of quality of life when compared to a healthy adolescent population. Just under one quarter of the study sample scored within the borderline or abnormal adjustment range, consistent with population norms.

This is an important finding of the study and draws attention to the inconclusive evidence discussed earlier about the relationship of disease, disease severity and

psychological adjustment (Wallander et al, 1989). The presence of SCD as a chronic illness is undoubtedly a stressor to adolescents and their families but the study provides no evidence that SCD accounts for higher rates of psychiatric morbidity. This is consistent with previous research where there is no clear evidence of increased rates of morbidity or adjustment disorders in young people with sickle cell disease (Midence et al, 1993).

It is also consistent with findings from the general paediatric literature that families with children with chronic illness or disability do not show any marked indications of maladjustment compared to families with healthy children (Cadman et al, 1991). The study is consistent with previous research that has failed to find adjustment differences when comparing SCD children to healthy controls (Lemanek et al 1986). The results imply that it is not particularly useful to attempt to conceptualise an SCD 'psychological profile' (Pless and Perrin, 1985) but to consider the findings in relation to the specific types of challenges that SCD presents to adolescents, over and above the normal developmental tasks (Fielding, 1985).

In this context, the experience of sickle cell disease, as reported by adolescents and parents in the study, indicates the considerable resilience of families in managing an unpredictable and complex condition. These findings are now reviewed.

Families

Only a small number of studies have looked at the experience of black families and chronic disease (Midence and Elander, 1994). In the study, two thirds of the sample were of Black African origin and one third were of Afro-Caribbean origin. Over a third of families had more than one child with SCD living at home. While half the sample were families with a single mother as sole parent living at home, the majority of these families (41.2%) reported frequent contact between the father and children. Asking about contact from fathers was included in the research interview because of the expected high rate of 'single parent' households. This raises important issues about the contribution of fathers to families that may otherwise remain invisible in research outcomes, and is particularly pertinent to assumptions behind evaluating the contribution of parenting to child adjustment and family cohesiveness.

In terms of social deprivation, half of the sample scored 6 or more (in a possible range of 3-8) indicating that families with SCD face multiple stressors to do with low income and poor quality housing, but that there is not a linear relationship between these factors and adolescent adjustment.

Illness Variables

Consistent with the illness patterns of SCD reported in the literature, the study sample reported a wide range of pain frequency and lifetime hospital admissions. Some parents commented that there had been an intense period early in the child's life where hospital admissions were very frequent for their child, but that by early adolescence their child

was experiencing significantly less painful crises. Of the adolescents who had had hospital admissions in the previous two years, paediatricians considered the majority of these admissions to be appropriate and for the whole sample, outpatient use was generally considered by paediatricians to be appropriate. This is an important issue because of the way in which sickle cell patients as a group may be perceived to be overly reliant on hospital care. The study is consistent with other findings that indicate the majority of families manage sickle pain at home (Fuggle et al, 1996).

Where paediatricians expressed concern about outpatient use this was not, as might have been expected, for inappropriate presentations for pain relief, except in one instance where parental anxiety was cited. Adolescents received low ratings for healthcare use either in relation to under-utilisation, where there was a feeling that pain was being managed at home with insufficient medical support, or that there was frequent defaulting of appointments and a concern that the adolescent would be lost to follow-up by the clinic. Of the families who are monitored by SCD clinics, it appears that it is a small minority of adolescent patients who present management difficulties to healthcare services.

School

School absence and concern about their child's progress at school was cited by the majority of parents as a significant worry. Nearly half the sample had more than 10 days absence due to SCD in the preceding 12 months and 9 families (17.6%) reported absences of 30 days or more. As in other studies (Shapiro et al, 1990), the main reason

for time away from school was pain crises that were generally managed at home without attending hospital or the clinic. Over a quarter of adolescents felt that SCD was having a significant impact upon their progress at school, citing missed days and worry about the onset of painful crises. The secondary effects of school absenteeism (Swift et al, 1989), for example, disruption to routine and peer-group relationships were also significant in accounting for some of the parental and adolescent concern about lowered educational attainment.

The study also suggests that school absence may be a sensitive measure of adjustment and family environment. Two of the three adolescents that estimated over 100 days absent from school in the previous 12 months scored in the abnormal adjustment range.

Activities

While progress at school was rated as of some concern to over half the adolescents, the impact of SCD upon sports and outdoor activity was considered by the majority as having the greatest effect upon their daily lives. This is consistent with other findings (Walco and Dampier, 1990; Conyard et al, 1990), and is unsurprising given the life stage of the sample. The majority of the sample was girls, whereas other studies (Conyard et al, 1990) have linked the effects upon sports activity to posing particular difficulties for boys. Many adolescents cited the onset of fatigue and anticipatory worry about the onset of painful crises as the main inhibitory factors in limiting their participation in sports. Parents talked about the difficulties in trying to support their child's participation in

activities as much as possible while being aware of the need for limitations. One carer said of their child 'She thinks she can do everything, *I* worry more.'

Self-Image

Just under half of both parents and adolescents said that SCD had some effect on the way adolescents felt about themselves. Some parents said that they felt SCD had affected their child's confidence. Adolescents cited the effect of SCD upon their growth and physical development as affecting the way they felt about themselves, for example looking younger and smaller than their peers, and parents cited worries that their child was more vulnerable to being bullied. Some adolescents mentioned continued problems with enuresis as the main cause of distress.

Others' Perception

Over half of the adolescents felt that SCD did affect to some extent the way that other people saw them, referring to their physical development. A number of adolescents talked about feeling 'different' from their peers and not wanting people to know that they had SCD. One girl said 'I act like a normal person without telling' and there was a feeling from many parents and adolescents that SCD was poorly understood in schools and in their communities, and that there remained a stigma to the condition. One boy, who had been hospitalised many times, said that SCD meant that 'you don't grow up to be a big person, but you have to be a strong person, strong-willed, and that makes you a 'big' person.'

Research Question 2: Are pain coping strategies significant predictors of adjustment outcomes for adolescents with sickle cell disease living in the United Kingdom?

An important finding from the study is that the pattern of adolescent coping scores is broadly consistent with other samples of young people with SCD (Gil et al, 1991). The majority of empirical studies that have looked at adolescent coping with SCD, using the CSQ and other standardised measures, have been conducted in America. The present study is a comparable sample of British ethnic minority young people and it is encouraging that the use of the CSQ provided a systematic and simple means to compare parent and child coping strategies, that was both reliable and acceptable to participants.

The study further tested the use of an adapted brief version of the CSQ because it was unlikely that parents would have been willing to complete a 78-item questionnaire twice (part of the methodology for measuring agreement). As reported, strong correlations between the adapted version and the full version were found, supporting the use of a more acceptable length of questionnaire to participants for future studies.

A central aim of the study was to assess whether the coping strategies used by adolescents with SCD would predict certain outcome measures: school absence, healthcare use, psychological distress and quality of life. Increased use of Coping Attempts, multiple cognitive and behavioural coping strategies including distraction and calming self-statements, was associated with lower episodes of pain frequency and increased quality of life. This finding is consistent with research with SCD adults (Gil et

al, 1989) and findings from studies of children with other painful conditions (Siegel and Smith, 1989).

The relationship identified between Negative Thinking and more hospital admissions, greater school absence, poorer adjustment and poorer quality of life also replicates the pattern of findings from the Gil group. Adolescents with high scores in this domain typically engaged in negative thinking patterns such as catastrophising and self-statements of fear and anger.

Interestingly, there was no relationship identified between Negative Thinking and Passive Adherence, but a positive correlation was shown between Passive Adherence and Coping Attempts. The items in the Passive Adherence domain included reliance on concrete strategies, such as fluids, taking hot baths, and following standard medical advice for pain management but also contained statements of relying on faith in God and praying as a coping strategy. It is possible that these statements are considered as positive and active coping attempts by communities where religious faith has a central role, and may reflect a greater cohesiveness in families. Passive Adherence was not linked to any negative outcomes, such as increased healthcare use and greater reduction in activity, as it was in the American research population (Gil et al, 1991).

Overall, the study showed that coping strategies used by adolescents to cope with sickle cell pain have predictive power in accounting for adjustment outcomes beyond that accounted for by age and frequency of painful episodes. As in the general paediatric

literature and in previous sickle cell research, disease severity was shown not to be a reliable predictor of adjustment.

Research Question 3: What is the contribution of parental understanding to adjustment outcomes for adolescents with sickle cell disease?

Overall, the comparisons of mean scores obtained by adolescents and parents across the three coping domains showed high consensus about coping patterns, where parents rated their view of how their child coped with SCD pain. This is also consistent with the concordance between parent and adolescent report about pain frequency reported in the study.

High Parental Agreement scores in Coping Attempts were significantly associated with less days off school because of problems with SCD. This finding supports the indication that school absence may be a more sensitive measure of adjustment than hospital admissions, since the majority of families reported managing painful episodes at home.

Comparisons of mean scores between Parental Ideal and adolescent ratings indicated moderate understanding in the domains of Coping Attempts and Passive Adherence. In the domain of Negative Thinking there was a significant discrepancy in scores, indicating a large discrepancy between adolescent coping strategies and Parental Ideal. In terms of outcomes, poor adjustment and poor quality of life was associated with high discrepancies in parental ideal and adolescent scores in the domain of Negative Thinking.

Taken as a whole, the scores obtained from the parental understanding measures are low, indicating high parent-child consensus about coping with pain. Parental understanding appears to make a significant contribution to adolescent outcomes after age and pain frequency have been controlled for.

As reported earlier, estimates of parent-child concordance in the literature regarding psychiatric symptomatology have been inconsistent, although there is some evidence that in groups of older children, rates of agreement increase (Reich et al, 1982). Agreement for discrete symptoms between parent and child is poor as reported in the literature, even when these are for observable behaviours, such as bed-wetting (Klein, 1991). Parent and child reports about child sickle cell pain tend to underestimate pain frequency when compared to diaries of pain kept by the child (Gill et al 1997). The high concordance reported in this study therefore needs cautious interpretation.

The overall contribution of parental understanding to adjustment is consistent with findings from children with a chronically ill sibling, where poor adjustment was associated with lower maternal understanding of their cognitions (Smith et al, in press).

Limitations of the Study

In the analysis a number of tests were performed on the data and the possibility that some of the significant results obtained were due to Type 1 error cannot be excluded. The statistical analysis was also limited by the small sample size, the age-range of children that spanned eight years and the fact that there was not an even gender distribution. The literature indicates increased adjustment risk for older children and boys, and exploration of these issues from the data was limited by the small numbers.

The sampling procedure relied on the paediatrician providing a list of adolescent patients who had attended for outpatient care within the previous two years. This procedure inevitably biased the research toward those adolescents in contact with healthcare services, and this contact may in itself be an index of adjustment. There was anecdotal evidence from the clinics that a number of families had been lost to follow-up and the study did not provide for any analyses of the group of adolescents that are not being monitored by paediatricians.

The sample was therefore a sample of clinic attenders, not a community sample of adolescents with sickle cell disease. There is some criticism within the SCD literature that research systematically excludes individuals with infrequent hospital contact on the assumption that they experience little or no severe pain (Serjeant, 1995). Given that the majority of families affected by sickle cell manage pain at home, there is likely to be a cohort of adolescents who were not current outpatients and who may have a different experience from the study sample of coping and adjustment.

It is also likely that parental understanding and adolescent adjustment will mutually affect each other. There are many other variables that are not measured in the present study that may be contributing to adjustment outcomes, for example the security of attachment. It is important to stress that parental understanding is a far more complex construct than the perameters of the study suggests. What is presented is one way of operationalising this construct but it is clearly not the only way to measure parental understanding. Furthermore, while the results are statistically significant, the clinical significance of the findings is modest.

Clinical and Professional Implications

Research with other pain populations suggests that enhancing pain coping effectiveness may be beneficial (Peterson, 1989). Children who take a more active approach to pain by using a variety of coping and behavioural strategies may be better equipped to manage the unpredictable painful episodes that may happen over the course of their lives. Gil et al (1991) discuss the need to evaluate the efficacy of cognitive-behavioural treatment programmes that offer early intervention, to specifically train SCD children and their parents to use strategies that are associated with Coping Attempts.

However there is a contradiction inherent between a prescriptive approach and the implications of the present study. An intervention based on training children and parents to have 'adaptive' cognitions to stressors may be unhelpful, or even harmful, if it disrupts parental understanding or family patterns of communication (Smith et al, in press).

The findings also support what is widely regarded as good clinical practice in child and family psychology services. Parent-child interaction work is based on facilitating communication between parent and child, and supporting the parent toward a greater understanding of the child's needs. The construct of parental understanding implies that in successful adaptation, there is a positive quality of communication between parent and child. The contribution of parental understanding that has been identified in the study offers further insight into the complex processes involved in the adjustment of adolescents and child mental health.

Future Research

The results from the study invite further exploration into the components of parental understanding. The addition of measures for example, of family cohesiveness, may help to further refine the measurement of parental understanding. The development of parental influence on children's coping processes over time in a longitudinal study may also yield some important data.

For the adolescents in the study who showed borderline and abnormal scores in adjustment it would be of interest to further explore the relationship of sickle cell disease and vulnerability factors to maladjustment with a larger sample.

Finally, there appears to be little empirical research with families with sickle cell who are not in contact with hospital services. A comparative study between clinic attenders and non-attenders may further develop our understanding about factors influencing differential adjustment in young people with sickle cell disease.

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TABLES

APPENDIX 1

Ethics Committee Correspondence

CREED
Clinical Research, Education,
Effectiveness, & Development

Telephone: 0171-288 5675 (24 hr service)
0171-288 5676
0171-288 5677
Fax Number: 0171-288 5674

The Whittington Hospital
Highgate Hill
London N19 5NF

Ref: 99-14

Ms L A French
Clinical Health Psychology

22nd April 1999

Dear Ms French

99-14 Adolescent coping with Sickle Cell Disease: The role of Parental Understanding

I am pleased to inform you that the above named project has been **approved**.

Approval is for two years from the date of this letter. Extension of this period will be dependent on the submission of a brief synopsis of the project together with an estimation of the time required for its ultimate completion.

The Ethics Committee will require an annual report on the progress of the study, and a copy of the completed study together with any consequent publication. In addition, the Committee must be informed, by the completion of the relevant form, of any untoward or adverse events which occur during the course of the study. The person who provided independent review of the original protocol should also be sent information regarding adverse events.

The Ethics Committee must be informed of, and approve, any proposed amendment to your initial application which has a bearing on the treatment or investigation of patients or volunteers.

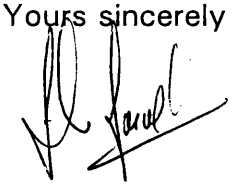
A copy of the patient consent form and information sheet must be lodged in the clinical notes.

Furthermore, whilst I am sure that every effort is already made to preserve the confidentiality of any patient information used in this study, could you please ensure that the team of investigators are aware that everyone who has access to patient information appreciates the importance of maintaining confidentiality particularly in respect of the use of computers and the statutory regulations laid down in the Data Protection Act 1998.

In terms of the managerial and financial implications associated with the study, where these relate to additional costs for the Trust, Mr Rob Hurd (Management Accountant, Finance Department, Whittington Hospital), will be in contact with you to discuss the Trust's schedule of charges for research projects. **Approval of these issues must be obtained from your general manager before the study commences.**

In any correspondence regarding the study please quote the above Ethics Committee reference number.

Yours sincerely



Mr John Farrell
Chairman - Local Research and Ethics Committee

Enfield Haringey
HEALTH AUTHORITY
Where health matters

Chairman: Peter Dixon
Chief Executive: Christine Outram

24 December 1999

Ms L French
260 Ferme Park Road
Crouch End
London N8 9BL

Dear Ms French,

725 – Adolescent coping with sickle cell disease: The role of parental understanding

Acting under delegated authority, I write to inform you that Local Research Ethics Committee considered the above study on 20 December 1999 and **it was approved** subject to the following proviso:

1. Amend consent form to read, "I have read and understood …"

Confirmation that the amendments have been included in the study is required before commencement of the study.

Yours sincerely

C Hamilton

Christine Hamilton
LREC Administrator
(on behalf of the LREC Chairman)

Holbrook House, Cockfosters Road
Barnet, Hertfordshire EN4 0DR
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4 January 2000

Christine Hamilton
LREC Administrator
Enfield & Haringey Health Authority Local Research Ethics Committee
Holbrook House
Cockfosters Road
Barnet
EN4 0DR

Dear Ms Hamilton

725-Adolescent Coping with Sickle Cell Disease: The Role of Parental Understanding

Thank-you for confirmation that the above study was approved at the meeting of the LREC on 20 December 1999 subject to amendment of the consent form for parents.

I enclose the amended consent form and the patient information sheet that indicates approval has been obtained from the Ethics Committees from Enfield and Haringey Health Authority and the Whittington Hospital NHS Trust.

Yours sincerely

Lesley French

APPENDIX 2

Information Sheet and Consent Form

INFORMATION SHEET FOR PARENTS & CHILDREN

Adolescents and Coping with Sickle Cell Disease

This study is designed to look at how adolescents understand and cope with sickle cell disease. We would like parents and children to help us with this study by completing some questionnaires and a short interview at home or at the clinic, at a time convenient for you. All the information we collect remains completely confidential and your name is not recorded on the questionnaires. We hope that the study will help the clinic to provide an improved service for families affected by sickle cell disease.

You do not have to take part in this study if you do not want to. If you decide to take part you can change your mind at any time without having to give a reason. Your decision to help in this study or not will not affect your care from the hospital in any way.

All studies that involve asking patients to help are reviewed by an ethics committee before they begin. This study was reviewed by the Whittington Hospital NHS Trust Ethics Committee and by the Enfield and Haringey Health Authority Local Research Ethics Committee. The study was approved by both committees.

If you would like further information about the study please contact:

Lesley French
telephone 0171 530 2450
Department of Clinical Health Psychology
University College London.

CONSENT FORM FOR CHILDREN

Adolescents and Coping with Sickle Cell Disease

Investigator: Lesley French
University College London

Please complete by circling either YES or NO

1. The study has been explained to me and I understand it.

YES NO

2. I agree to take part in this study.

YES NO

3. I know that I can come out of the study at any time.

YES NO

Signed: .....

Your Name in Block Letters: .....

Investigator's Signature: .....

Date: .....

CONSENT FORM FOR PARENTS

Adolescents and Coping with Sickle Cell Disease

Investigator: Lesley French
University College London

Please complete by circling either YES or NO

1. I have read and understood the information sheet about the study.	YES	NO
2. I have had an opportunity to ask questions about this study.	YES	NO
3. I have received enough information about this study.	YES	NO
4. I understand I can withdraw from this study at any time, without having to give a reason, and this will not affect my care in any way.	YES	NO
5. I agree to take part in this study with my child.	YES	NO

Signed: .....

Your Name in Block Letters: .....

Investigator's Signature: .....

Date: .....

APPENDIX 3

Adolescent Questionnaires

Coping Strategies Questionnaire -Sickle Cell Disease Version for Children

Below is a list of things that children have reported doing or thinking when they feel pain. I want you to indicate, using the scale below, *how much you do or think each item on the list when you feel sickle cell pain.*

0 = you never do that when you feel pain, **3** = you sometimes do that when you feel pain and **6** = you always do that when you feel pain.

0	**1**	**2**	**3**	**4**	**5**	**6**
Never			Sometimes			Always

When I feel pain....	**What Ido or think**
1. I try to get some sleep.	_____
2. I imagine that the pain is outside of my body.	_____
3. I take a hot or cold bath.	_____
4. I think of things that I enjoy doing.	_____
5. I try to think years ahead, what everything will be like after I've got rid of the pain.	_____
6. I read.	_____
7. I avoid people.	_____
8. I realise that most people don't really care.	_____
9. I don't like to be with people.	_____
10. I try to think of something pleasant.	_____
11. I drink twice as much as I usually do.	_____
12. I rub the parts of my body that hurt.	_____
13. I increase my fluid intake.	_____
14. I tell myself it doesn't hurt.	_____
15. It is terrible and I feel that it is too much to take.	_____

0	1	2	3	4	5	6
Never			Sometimes			Always

When I feel pain....	**What Ido or think**
16. I try to drink some water or juice every hour.	_____
17. I think it is not fair that I have to live this way.	_____
18. I do something I enjoy, such as watching TV.	_____
19. I try to drink a lot of water.	_____
20. I worry that I am having a heart attack or some other physical problem.	_____
21. It is terrible and I feel it is never going to get any better.	_____
22. I take a hot or cold shower.	_____
23. I think no one wants to hear about my problems.	_____
24. I go off by myself.	_____
25. I go to bed.	_____
26. I try to be alone.	_____
27. I rely on my faith in God.	_____
28. I count numbers in my head or run a song through my mind.	_____
29. I worry that my disease is getting worse.	_____
30. I know I need to get away from everyone.	_____
31. I pretend it is not a part of me.	_____
32. I rub painful areas.	_____
33. I use ice packs to help relieve the pain.	_____
34. I think of things in my head to keep my mind off the pain.	_____
35. I go to a quiet place where I won't be bothered.	_____

0	1	2	3	4	5	6
Never			Sometimes			Always

When I feel pain....	**What I do or think**
36. I think of people I enjoy doing things with.	_____
37. Although it hurts, I just keep on going.	_____
38. I think that if I can't be healthy then no one else should be.	_____
39. I tell myself that I can get over the pain.	_____
40. I try to be around other people.	_____
41. I ignore it.	_____
42. I have faith in doctors that someday there will be a cure for my pain.	_____
43. I think that I don't deserve this.	_____
44. I just go on as if nothing happened.	_____
45. I tell myself to be brave and carry on despite the pain.	_____
46. I worry all the time whether it will end.	_____
47. I just think of it as some other sensation, such as numbness.	_____
48. I know others don't understand.	_____
49. I don't pay any attention to it.	_____
50. I drink five or more glasses of water or juice a day.	_____
51. I worry that I am really going to get sick.	_____
52. I relax my muscles.	_____
53. I do anything to get my mind off the pain.	_____
54. I am afraid I am going to die.	_____
55. I lay down on the bed or couch in order to relax.	_____

0	1	2	3	4	5	6
Never			Sometimes			Always

When I feel pain.... **What I do or think**

56. I pretend it is not in my body. _____

57. I feel I can't go on. _____

58. I pretend it is not there. _____

59. I pray to God it won't last long. _____

60. I feel I can't stand it anymore. _____

61. I think about happy times in the past. _____

62. I do something active, like playing outside. _____

63. I try to feel distant from the pain, almost as if the pain was in someone else's body. _____

64. I drink as much juice or water as I can. _____

65. I know I'll have to go to the hospital or see my doctor. _____

66. I spend time resting. _____

67. I know someday someone will be here to help me and it will go away for a while. _____

68. I don't think about the pain. _____

69. I leave the house and do something, such as playing with friends. _____

70. I try to rest. _____

71. I am sure there is something wrong. _____

72. No matter how bad it gets, I know I can handle it. _____

73. I tell myself I can't let the pain stand in the way of what I have to do. _____

74.I use a heating pad. _____

0	1	2	3	4	5	6
Never			Sometimes			Always

When I feel pain....	**What I do or think**
75. I don't let it bother me.	_____
76. I feel my life isn't worth living.	_____
77. I pray for the pain to stop.	_____
78. I try to make the pain feel like something else.	_____

Based on all the things you do to cope with your pain, on an average day, *how much control do you feel you have over it?* You can circle any number along the scale below.

0	1	2	3	4	5	6
No control			Some control			Complete control

Based on all the things you do to cope with your pain, on an average day, *how much are you able to <u>decrease</u> it?* You can circle any number along the scale below.

0	1	2	3	4	5	6
Can't decrease it at all			Can decrease it somewhat			Can decrease it completely

Thank-you.

Strengths and Difficulties Questionnaire

For each item, please mark, Not True, Somewhat True or Certainly True. Please give your answers on the basis of how things have been for you over the last six months.

	Not True	Somewhat True	Certainly True
I am considerate of other people's feelings.			
I am restless, I cannot stay still for long.			
I get a lot of headaches, stomach-aches or sickness.			
I usually share with others (food, games, pens etc.)			
I get very angry and often lose my temper.			
I am rather solitary. I usually play alone or keep to myself.			
I usually do as I am told.			
I worry a lot.			
I am helpful if someone is hurt, upset or feeling ill.			
I am constantly fidgeting or squirming.			
I have at least one good friend.			
I fight a lot. I can make other people do what I want.			
I am often unhappy, down-hearted or tearful.			
Other people my age generally like me.			
I am easily distracted. I find it difficult to concentrate.			
I am nervous in new situations. I easily lose confidence.			
I am kind to younger children.			
I am often accused of lying or cheating.			
Other children or young people pick on me or bully me.			
I often volunteer to help others (parents, teachers, children).			
I think things out before acting.			
I take things that are not mine, from home, school or elsewhere.			
I get on better with adults than with people my own age.			
I have many fears. I am easily scared.			
I see tasks through to the end. My attention is good.			

Overall, do you think that you have difficulties in one or more of the following areas: emotions, concentration, behaviour or being able to get on with other people?

No	Yes Minor Difficulties	Yes Definite Difficulties	Yes Severe Difficulties

If Yes, do these difficulties upset or distress you?

Not at all	Only a little	Quite a lot	A great deal

Do these difficulties interfere with your life in the following areas?

	Not at all	Only a little	Quite a lot	A great deal
Home Life				
Friendships				
Classroom Learning				
Leisure Activities				

Do these difficulties make it harder for those around you (family, friends, teachers, etc.)?

Not at all	Only a little	Quite a lot	A great deal

Thank-you.

Quality of Life Measure-Version for Girls

This is a story about 5 girls who are all friends. They talk about lots of things and find out they all have different feelings. I would like you to think about each thing that the friends talk about and tick the girl most like you (example sheet overleaf).
They talk about....

1. having fun-they find out that one girl always has fun, one often has fun, one sometimes has fun, another hardly ever has fun and one girl never has fun. Tick the girl most like you.
2. being happy and smiling
3. how often they worry about things
4. how often they spend time with friends
5. how often they have enough friends
6. how much of the time other people understood how they felt
7. how much of the time they are picked on
8. how often they help others
9. how often they hurt other people
10. how often they get upset
11. how often they feel bored
12. how often they can go to someone if they have a problem
13. how much of the time they like their parents
14. how much of the time they think their parents love them
15. how often they are told off
16. how often they are allowed to choose for themselves
17. how much of the time they feel happy with their life
18. how often they are really ill
19. how often this stops them from doing things that they want to do
20. how happy they are about the way they look
21. how often they feel different from other children
22. how often they try hard with their work
23. how often they are told off by the teacher
24. how often they feel more clever than other children
25. how often they are good at sport

Now fold the page at the dotted line. You will see a new row of girls.
Answer the questions again, but this time **tick the girl that you would most like to be.**
Start again at number 1 and work through to number 25.

Thank-you.

	always	often	some-times	hardly ever	never
1					
2					
3					
4					
5					
6					
7					
8					
9					
10					
11					
12					

	always	often	sometimes	hardly ever	never
13					
14					

	always	often	sometimes	hardly ever	never
15					
16					
17					

	always	often	sometimes	hardly ever	never
18					
19					

	always	often	sometimes	hardly ever	never
20					
21					

	always	often	sometimes	hardly ever	never
22					
23					

	always	often	sometimes	hardly ever	never
24					
25					

ADOLESCENT COPING WITH SICKLE CELL DISEASE

MEASURE OF APPRAISAL FOR COMPLETION WITH CHILD

1. I want you to tell me how much pain you have from sickle cell.
How many times in the past week have you had sickle cell pain?.....
How many times in the last month?.....
Has the pain stopped you from doing any of the things you had planned to? YES NO
(If yes) What sorts of things did the pain stop you from doing?

2. How many times have you been in hospital for sickle cell pain in your life?
1-2? 3-4? 5-10? More than 10?.....

3. I wonder if it is possible for you to think of yourself without sickle cell disease. What would be different for you if you didn't have it?

4. I want you to tell me how much of a difference having sickle cell has made to you.
You can say whether it's made a big, small or no difference at all to these questions.

	BIG DIFFERENCE	*SMALL DIFFERENCE*	*NONE*
a) how you think or feel about yourself?	.....	.....	.....
b) how other people see you?	.....	.....	.....
c) what you can or you can't do?	.....	.....	.....
d) how you get on with other children?	.....	.....	.....
e) how you get on with schoolwork?	.....	.....	.....

Prompt for comments.

5. I wonder whether there is anything at all about having sickle cell that has made a difference to you in a positive or a good way?

Thank and finish.

APPENDIX 4

Parent Questionnaires

ADOLESCENT COPING WITH SICKLE CELL DISEASE

INTERVIEW WITH PARENT

Participating Parent MOTHER FATHER OTHER RELATIVE (specify)

Participating Child MALE FEMALE Date of birth_____

To begin with, I would like to ask you for just a few details about your family.

1. Who lives at home with your child?
Child's mother.....father.... child's brothers (number).....child's sisters.....
Other family members.....

2. If your child's father does not live with you is there contact between him and the child?
If so, how frequent is this?

3. How old was your child when you discovered he/she had sickle cell disease? .....

4. Do you have any other children with sickle cell disease? YES NO
If so, what age are they?.....(sons).....(daughters)

5. Do you have any other relatives that you know with sickle cell disease? YES NO

If so, what relation are they to you?.....

6. Which of the following ethnic groups best describes you and your child?

	PARENT	CHILD
Afro-Caribbean		
Black African		
Mixed Race		
Other		

Thank-you. I would now like to ask you about some things to do with school

7. Where does your child go to school?.....

8. How many days off school do you think your child had in the last 12 months because of problems with sickle cell?.....
What problems did your child have with sickle cell that made him/her miss school?

9. How concerned are you that sickle cell disease is going to affect your child's progress at school?

VERY CONCERNED FAIRLY CONCERNED NOT AT ALL CONCERNED
Why do you say that?

10. What age were you when you left school?.....
Did you go on to do any further training or education?
Do you work outside the home? YES NO
(If father living at home) does..... father work? YES NO N/A

Housing Status: OWNER OCCUPIED COUNCIL-POOR COUNCIL -ADEQUATE

Now there are a few questions about the effect of sickle cell on your child.

11. How much sickle cell pain has your child had in the past week?.....
And in the past month?.....

12. How many times has your child had to stay in a hospital ward for sickle cell pain in his/her life?

1-2?.....3-4?.....5-10?.....More than 10?.....

13. On a scale from 1-10, tell me <u>how big a problem</u> you think sickle cell disease is for your child?
1= not a problem at all and 10 = a very big problem.

1 2 3 4 5 6 7 8 9 10

14. Finally, how <u>much</u> of a difference do you think that having sickle cell has made to your child's life. Please tell me whether it has made a big, small or no difference at all to the following areas. Your child's.....

	BIG DIFFERENCE	*SMALL DIFFERENCE*	*NONE*
a) personality?	.....	.....	.....
b) sense of self?	.....	.....	.....
c) activities?	.....	.....	.....
d) friendships?	.....	.....	.....

Prompt for comments.

Thank and continue with written questionnaires.

Coping Strategies Questionnaire - Revised

Sickle Cell Disease Version for Parents (1)

Below is a list of things that children have reported doing or thinking when they feel pain. I want you to indicate, using the scale below, *how much you think your son does that activity or says those things to himself when he feels pain*, just imagining how he might answer.
0 = he never does that when he feels pain, **3** = he sometimes does that when he feels pain and **6** = he always does that when he feels pain.

0	**1**	**2**	**3**	**4**	**5**	**6**
Never			Sometimes			Always

When my son feels pain.....	**What my son does or thinks**
1. He imagines that the pain is outside of his body.	_____
2. He takes a hot or a cold bath.	_____
3. He thinks of things that he enjoys doing.	_____
4. He avoids people.	_____
5. He drinks twice as much as he usually does.	_____
6. He rubs the parts of his body that hurt.	_____
7. It is terrible and he feels it is never going to get any better.	_____
8. He goes to bed.	_____
9. He tries to be alone.	_____
10. He counts numbers in his head or runs a song through his mind.	_____
11. Although it hurts, he just keeps on going.	_____
12. He thinks that if he can't be healthy then no one else should be.	_____
13. He tells himself that he can get over the pain.	_____
14. He ignores it.	_____

0	1	2	3	4	5	6
Never			Sometimes			Always

When my son feels pain.... **What my son does or thinks**

15. He has faith in doctors that someday there will be a cure for his pain. _____

16. He thinks that he doesn't deserve this. _____

17. He tells himself to be brave and carry on despite the pain. _____

18. He just thinks of it as some other sensation, such as numbness. _____

19. He worries that he is going to get really sick. _____

20. He is afraid he is going to die. _____

21. He prays to God it won't last long. _____

22. He does something active, like playing outside. _____

23. He drinks as much water or juice as he can. _____

24. He doesn't think about the pain. _____

25. He tries to rest. _____

26. He is sure that there is something wrong. _____

Based on all the things your son does to cope with his pain, on an average day, *how much control do you think he has over it?* Please circle the appropriate number.

0	1	2	3	4	5	6
No control			Some control			Complete control

Thank-you.

Coping Strategies Questionnaire - Revised

Sickle Cell Disease Version for Parents (2)

Now I want you to go through the list again, using the same scale, and indicate *how you think your son should think or do things when he feels pain.* Imagine that you were giving him advice on the best thing to do or think when he is experiencing sickle cell pain.

0 = he should never do that when he feels pain, **3** = he should sometimes do that when he feels pain and **6** = he should always do that when he feels pain.

0	**1**	**2**	**3**	**4**	**5**	**6**
Never			Sometimes			Always

When my son feels pain.....	**The best thing he should think or do**
1. He should imagine that the pain is outside of his body.	_____
2. He should take a hot or cold bath.	_____
3. He should think of things that he enjoys doing.	_____
4. He should avoid people.	_____
5. He should drink twice as much as he usually does.	_____
6. He should rub the parts of his body that hurt.	_____
7. It is terrible and he should be feeling that it is never going to get any better.	_____
8. He should go to bed.	_____
9. He should try to be alone.	_____
10. He should count numbers in his head or run a song through his mind.	_____
11. Although it hurts, he should just keep on going.	_____
12. He should think that if he can't be healthy then no one else should be.	_____
13. He should tell himself that he can get over the pain.	_____
14. He should ignore it.	_____

0	1	2	3	4	5	6
Never			Sometimes			Always

When my son feels pain.... **The best thing he should think or do**

15. He should have faith in doctors someday there will be a cure for his pain. _____

16. He should think that he doesn't deserve this. _____

17. He should tell himself to be brave and carry on despite the pain. _____

18. He should think of it as some other sensation, such as numbness. _____

19. He should be worrying that he is going to get really sick. _____

20. He should be afraid that he is going to die. _____

21. He should pray to God that it won't last long. _____

22. He should do something active, like playing outside. _____

23. He should drink as much water or juice as he can. _____

24. He shouldn't think about the pain. _____

25. He should try to rest. _____

26. He should feel sure that there is something wrong. _____

Based on all the things your son does to cope with his pain, on an average day, *how much control do you think he has to decrease it?* Please circle the appropriate number.

0	1	2	3	4	5	6
Can't decrease it at all			Can decrease it somewhat			Can decrease it completely

Thank-you.

APPENDIX 5

Paediatric Rating Form

Adolescent Coping and Sickle Cell Disease: The Role of Parental Understanding

PAEDIATRIC RATING FORM

1. Name of child _____ (Study number_____)

2. Type of sickle cell disease _____

3. Number of child hospital admissions for pain relief 1998 _____

1999 _____

4. Overall, how appropriate do you think the hospital admissions for pain relief were? Please circle a number on the scale below.

1	2	3	4	5
NOT AT ALL APPROPRIATE				VERY APPROPRIATE

5. Overall, how appropriate do you think this child's use of medical outpatient services for sickle cell disease is? Please circle a number on the scale below.

1	2	3	4	5
NOT AT ALL APPROPRIATE				VERY APPROPRIATE

Thank-you.