Self-perceived stigma in young people with sickle cell disease: Associations with psychosocial distress

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Department of Medicine

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Declaration

I declare that the work in this thesis is my own and that I have appropriately acknowledged the work of others.
Dedication

This thesis is dedicated to my wife Obe and our sons, Victor and Kevin.
Acknowledgements

I would like to say a special thank you to my wife Obe and my sons Victor and Kevin without whose constant support, encouragement and allowance, I would not have been able to complete this Thesis. It has been a great family effort.

I would like to extend special thanks to my supervisors Dr Matthew Hodes and Professor Elena Garralda for their immense guidance, encouragement and fantastic mentoring.

Great thanks are due to the 93 young people who participated in the study and their families. I am also very grateful to the management of the Sickle Cell Society for facilitating recruitment through their membership database.

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Abstract

Background
Sickle cell disorders (SCD) are serious genetic blood conditions affecting mainly people of Black African origin. The disease is associated with serious physical complications and some affected persons have increased psychological difficulties. Self-perceived stigma is a putative risk factor for psychological distress in SCD but this had not been studied.

Aim
The primary aim is to estimate the prevalence of self-perceived stigma in young people with SCD and to explore its associations with psychosocial and illness variables. A secondary aim is to explore associations of other measures of psychological adjustment with illness and social indicators.

Methodology
Cross-sectional questionnaire survey of 93 young people with SCD aged 10-19 years (Mean 14 years). Questions on self-perceived distancing by others were used to assess stigma. Psychological difficulty was assessed with self-report Strengths and Difficulties Questionnaire (SDQ), Depressive symptoms were measured with the Short Mood and Feelings Questionnaire, self-esteem was assessed with the Rosenberg Self-Esteem Scale, and Family function was measured with the Family Assessment Device.

Results:
The respondents were evenly split in gender and almost all were of Black ethnicity (95%). However, they had better socioeconomic profile compared with average black families in the UK. Only 15% had self-perceived stigma.

Consistent with stigma theory, frequent ward admissions (i.e. measure of disruptiveness) and presence of leg ulcer (i.e. measure of visibility) predicted more self-perceived stigma. Self-perceived stigma in turn predicted more psychological difficulty (Total SDQ score).
Psychological symptoms were also associated with poor attitudes towards SCD, and by problematic family function.

**Conclusion**

To my knowledge, this is the first study to show that stigma theory applies to SCD and that self-perceived stigma is a significant predictor of psychological difficulty in this disorder. Thus, alleviating stigma could benefit psychological well-being in SCD.
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Introduction

Sickle cell disorders (SCDs) are blood disorders that predominantly affect people of black African ancestry as well as people of Mediterranean, Middle Eastern and Asian origins (Weatherall and Clegg, 2001). They are among the most common genetic conditions worldwide (Modell and Darlison 2008) and constitute the commonest monogenic diseases in the world (Weatherall 2008). SCDs are disorders of haemoglobin with the primary abnormality being distortion of the normal spherical shape of red blood cells into sickle shapes.

SCDs are associated with several, sometimes life-threatening physical complications. The primary pathogenesis of the complications is ischaemia caused by the trapping of sickle-shaped red blood cells in the microvasculature. Depending the affected organ, the consequent complications include ischaemic pain (which is the most typical and most distressing symptom) (Wethers 2000), bone necrosis, stroke, chronic leg ulceration, renal disease, and priapism. Other complications include acute chest syndrome, cholelithiasis, overwhelming infections, acute splenic sequestration, haemolytic and aplastic crises, and nocturnal enuresis (Dick 2008).

Treatment advances in developed countries have led to mitigation of some of these complications and hugely improved physical outcomes. This is exemplified by improved life expectancy from 14 years in the 1970s to more than 50 years currently (Claster & Vichinsky, 2003). Efforts are also being made in some developed countries (e.g. the UK) to use standards and guidelines to improve nation-wide access to evidence-based care for affected persons (Dick 2008). Unfortunately, as with most disorders, these advances have yet to filter to developing countries where most affected persons live. Consequently, life expectancy remains as low as 5 years in some areas (Serjeant 2005) such as sub-Saharan Africa where half of all affected children die before the age of 5 years (WHO 2006).

We know from many large scale epidemiological studies such as the Isle of Wight study (Rutter et al 1976) (consistently replicated since) that children with chronic physical disorders like SCDs are at increased risk of psychosocial difficulties. The risk is particular high for disorders involving the brain, of which SCDs is an example.
The ecological model of child development suggests that factors within the child and with the child’s environment are likely to contribute separately to the risk of mental disorder (Bronfenbrenner, 1979). Consistent with this model, several studies have shown that factors in the child such as severity or stage of disease are associated with psychosocial adjustment in children with SCD (Key et al 2001). Environmental factors, especially family function have also been linked with psychosocial adjustment in affected children (Barbarin et al 1999). Peer and broader social relationships are also important, including the way children view themselves in relation to others, and the stigma linked to illness may be expected to have an effect on children’s adjustment.

However, despite the plethora of studies on the psychosocial aspects of SCD in children, the potential primary role of stigma is yet to be studied. Stigma is an important aspect of the social environment, which could have significant influence on the adjustment of children affected by SCDs. Studies of other chronic physical disorders like epilepsy have shown the importance of stigma (Westbrook et al 1992).

Thus the primary objective of this study is to explore the level of perceived stigma among children with SCD. The second objective is to identify any associations between stigma and disease and psychosocial outcomes.

The role of psychological intervention in SCDs is increasingly being recognised and a guideline for this is being developed in the UK (Dick 2008). There is some evidence that Cognitive Behavioural Therapy may be helpful as an adjunct in pain management. However, despite encouraging results of psychological interventions in SCDs (Anie 2005), the evidence base is still limited (Anie and Green 2002). Thus identification of a wider range of putative risk factors for psychological distress in people with SCD could lead to new and more effective targeting of psychological interventions. Stigma is an example of a putative environmental risk factor that is yet to be studied in SCD; hence it is the focus of this study. As far as I am aware, this is the first study to specifically and directly examine self-perceived stigma in children with SCDs.
A cross-sectional questionnaire-based survey was the methodology chosen to achieve the objectives of the study. This methodology was considered appropriate and adequate given that the study is exploratory and the logistics of an alternative longitudinal design would be prohibitive.

To build on previous work and allow for comparison, I adopted the definition and measurement strategies for self-perceived stigma, which had been used reliably in two previous studies of children with epilepsy (Westbrook et al 1992) and stuttering (Blood et al 2003). 

The Thesis is structured into seven Chapters. The focus of each Chapter is outlined next.

In Chapter 1, I will discuss the biological aspects of SCDs. I will also present the epidemiology, course, treatment and physical complications of the disorder. The chapter will highlight the variability in the course and the huge differences in outcome in developed and developing countries. The Chapter will highlight improving international recognition of the huge burden of SCDs, which will hopefully translate into devotion of more resources to affected persons especially in developing countries.

In Chapter 2, I will discuss the concept of stigma and the development and elaboration of the term since the first seminal work on the subject by Goffman (1963). I will explore the concept of “stigma dimensions”, which are the attributes known to engender stigma when associated with a disorder or condition. I will show that two stigma dimensions (visibility and disruptiveness) could be applicable to SCDs. I will review the literature on stigma and SCDs, which showed very limited pre-existing data on the subject. I will also explore the methodological difficulties associated with studying stigma (such as socially desirable responding) and how this can be minimised.

In Chapter 3, I will explore the literature on psychopathology and psychosocial adjustment in SCDs. The chapter will apply both the bio-psycho-social and ecological models of adjustment to the literature. Given the particular clinical significance of
depression, I will devote a section to explore the risk factors, diagnostic difficulties and prevalence of depression in children with SCD. I will highlight differences in the data between developed and developing countries.

In Chapter 4, I will discuss the detailed aims of the study and the four specific hypotheses I will test. I will describe the full study methodology including sample size calculations, recruitment strategies and the measurements used.

Results will be presented in Chapter 5 starting with descriptive results and ending with multivariate analyses including tests for the four study hypotheses.

In Chapter 6, I will discuss the main results in the light of the literature on stigma and previous data on the psychosocial adjustment of children affected by SCDs, and methodological limitations of the study.

I used the final Chapter (7) to draw conclusions from the entire work and make recommendations for both clinical work and future research directions.
Chapter 1
Sickle cell disorders:
Biology, epidemiology, course, complications and treatment

1.1. Introduction
Sickle cell disorders (SCDs) are among the most common genetic conditions worldwide (Modell and Darlison 2008) and the commonest monogenic diseases in the world (Weatherall 2008). SCDs are disorders of haemoglobin, which distort the normal spherical shape of red blood cells into sickle shapes; hence the name. The majority of affected persons are of black African ancestry but cases are seen among people of Mediterranean, Middle Eastern and Asian origins (Weatherall and Clegg, 2001). In this chapter, I will discuss the basic biology, epidemiology, course, complications and treatment of SCDs.

1.2. Biology
Haemoglobin comprises of four globin chains. Adult haemoglobin has two Alpha and two Beta globin chains while foetal haemoglobin has two alpha and two gamma chains. Mutations in the amino acid sequence of haemoglobin can result either in the production of abnormal globin chains (haemoglobinopathy) or reduced or inability to produce normal globin chains (thalassemia). SCDs are haemoglobinopathies, which are qualitative abnormalities of haemoglobin function. On the other hand, Thalaseamias are quantitative abnormalities of insufficient haemoglobin production. Haemoglobinopathies (e.g. SCDs) can co-exist with thalassemia (e.g. HbSThal). The more common and clinically significant haemoglobin variants include haemoglobin S, C, D, E, β thalasemia.

Haemoglobin gene variants are recessively inherited and heterozygous carriers are usually asymptomatic. This explains why affected children are usually born to asymptomatic parents. The serious haemoglobin disorder seen in SCD occurs in people who inherit harmful combinations of gene variants (Modell and Darlison 2008). The most common and severe SCD is HbSS. Other common combinations include HbSC and Hbβthalasaemia. Persons who inherit one abnormal haemoglobin
gene variant but whose other copy of the gene is normal are asymptomatic carriers (e.g. HbAS).

The fundamental pathology in SCD is that when deoxygenated, the abnormal haemoglobin variant deforms red blood cells into a crescent-shape. The abnormally shaped red blood cells become trapped in the microvasculature causing sludge and blockage. The impaired circulation results in ischaemic damage, which underlies the common complications in SCD. Affected individuals typically present with painful ischaemic crisis, which is the most distressing symptom (Wethers 2000). Severe forms of the disease are associated with serious and sometime life threatening complications including, severe anaemia, overwhelming infections, acute splenic sequestration, haemolytic and aplastic crises, acute chest syndrome, strokes, cholelithiasis, bone necrosis, chronic leg ulceration, and renal disease (Wethers 2000).

1.3. Epidemiology

Modell and Darlison (2008) have carried out the most recent and most comprehensive review of haemoglobin disorders I am aware of. Using a wide range of sources of epidemiological information, they showed that haemoglobin disorders constitute a significant health problem in 71% of countries including those that account for 89% of worldwide births. Their analysis also showed that over 300,000 infants are born per annum with these disorders of which the vast majority (83%) have sickle cell disorders. They concluded that haemoglobin disorders account for 3.4% of deaths in children less than 5 years of age (6.4% in Africa). Mordell and Darlison (2008) highlight that 5.2% of the world population and 7% of pregnant women carry a significant haemoglobin variant. Their analyses indicate that although HbS accounts for 40% of carriers, it causes 80% of disorders because of very high carrier prevalence in some areas.

The substantial global burden of SCD suggested by Modell and Darlison (2008) is supported by other country specific estimates of prevalence. For example, the American National Heart, Lung and Blood Institute estimates that SCD affects 70,000 – 100,000 persons in United States. They estimate a prevalence of 1 in 500 among African American births and 1 in 36,000 Hispanic American births. About 2 million

Nigeria has the highest prevalence of SCDs worldwide because it is the most populous Black Country. About 25% of Nigerian adults carry the sickle cell gene and up to 150,000 babies are born per year with SCDs (WHO 2006, Akinyanju 1989).

In the UK, estimates of the number of affected persons range from 6500 to 12,500 (Bennett 2005). The new-born screening programme in England has produced more accurate birth prevalence data. Between 2004-2005, the screening programme identified 250 babies with SCDs and 6500 carriers of the sickle cell trait in England. The birth prevalence was 1:1500. The authors estimated that when UK-wide data becomes available, the national birth prevalence of SCD will be about 1:2000-1:2,500. They noted that SCDs are as common as cystic fibrosis in England although SCD cases are concentrated in London and other urban areas (Streetly et al 2008).

The natural geographical distribution of sickle cell trait mirrors that of endemic malaria. Heterozygous carrier status for the sickle cell gene provides some protection against malaria (Crompton et al 2008). Thus, given the significant mortality associated with malaria, the protective advantage for carriers leads to their selective survival, which results in continuing propagation of the gene.

Developed countries have much fewer numbers of people with SCD. They also have access to more advanced treatments and prenatal screening programmes (Weatherall 2008). On the converse, low income countries, particularly those of Sub-Saharan Africa and Asia have some of the highest prevalence of SCDs and very limited expertise and resources for managing these disorders. Without greater recognition of the serious and large scale burden of SCDs by governments and international health agencies, an ever increasing numbers of people in many low income countries will continue to lack access to effective treatment and screening programme, resulting in avoidable suffering and death. However, SCDs are now attracting the level of public attention they deserve. For example, the United Nations General Assembly adopted a resolution on 22nd December 2008 to recognise SCDs as public health problems and
declared that 19\textsuperscript{th} June of each year would be Sickle Cell World Day

1.4. Course and outcome of SCD

The course of SCDs is variable with some individuals severely disabled and others running a mild course. It is difficult to predict the course for particular individuals. The largest longitudinal study of SCDs (The Cooperative Study of Sickle Cell Disease) reported that dactylitis, severe anaemia, and leucocytosis in very young children with sickle cell disease (SCD) predicted a more severe course and increased the risk of later adverse outcomes. However, this finding has not been validated in other studies. For example, a recent cohort study in USA which followed 168 children with SCDs for 7 years (Quinn et al 2008) found no relationship between early clinical predictors and later adverse outcomes. Most subjects who experienced adverse events during this study were actually predicted to be at low risk for those events. The study found that no subject who was predicted to be at high risk actually experienced an adverse outcome.

Although more affected individuals are surviving for longer, in low income countries, SCDs is still responsible for a considerably high proportion of under 5 mortality. This amounts to 5\% of under 5 deaths in Africa and up to 16\% in some West African countries. In some areas of sub-Saharan Africa, half of all affected children die before the age of 5 years usually from anaemia and infections (WHO 2006).

Medical advances have improved life expectancy of people affected by SCDs in developed countries from 14 years in the 1970s to more than 50 years currently (Claster & Vichinsky, 2003). In the United State, average life expectancy in 1994 was 42 years for men and 48 years for females (Platt et al 1994). The outcome is not universally poor in all low income countries. For example, in Jamaica, life expectancy for people with SCD in 2001 was 53 years for men and 58 years for women (Wierenga et al 2001). On the contrary, life expectancy for people affected by SCDs in some low income countries can be as low as 5 years (Serjeant 2005).
1.5. Complications and treatment
Complications in SCDs arise principally from ischaemic damage from blockage of small blood vessels by sickle shaped red cells. Affected individuals typically present with painful ischaemic crisis, which is the most distressing symptom (Wethers 2000). The disorder is associated with serious and sometimes life threatening complications including overwhelming infections, acute splenic sequestration, haemolytic and aplastic crises, acute chest syndrome, stroke, cholelithiasis, bone necrosis, chronic leg ulceration, and renal disease, priapism, and nocturnal enuresis (Dick 2008). SCD complications involving the brain are significant. Up to 11% of affected persons suffer overt strokes and up to 20% have evidence of ischaemic brain damage on MRI by the age of 20 years (Pegelow et al 2002).

Due to the high contribution of pneumococcal infections and malaria to mortality in SCDs, routine treatment includes pneumococcal immunisation and prophylactic penicillin and antimalarials (in malarious areas). Because of the high turnover of red blood cells and increased haemopoiesis, folic acid supplementation is also routinely advised. Adequate nutrition and fluid intake is helpful. Specific treatments for complications such as analgesia for pain, blood transfusions for anaemia are sometimes required. In the UK, standards and guidelines for clinical care have been introduced to improve access to evidence-based care for people affected persons by SCDs throughout the country (Dick 2008).

For suitable patients, allogeneic haematopoietic cell transplantation (HCT) is currently the only treatment with curative potential for SCDs (Michlitsch and Walters 2008). Recent studies of HCT show an event-free survival of 85% after human leucocyte antigen (HLA)-identical sibling transplantation for SCDs (Michlitsch and Walters 2008). However, HCT is limited by the risk of serious complications including graft failure, recurrent disease, graft-versus-host-disease (GVHD), and infections. Also the availability of expertise and supportive treatment limit this intervention to few centres in developed countries.

Hydroxyurea has been shown to reduce the clinical severity of SCDs in adults (Odievre et al 2008). The mechanism of action is thought to be related to increased
production of foetal haemoglobin. A recent review of trials of hydroxyurea in children found that the medication increased foetal haemoglobin in most children and reduced the number of vaso-occlusive crises, hospitalisations, frequency of acute chest syndrome and rate of blood transfusion. The medication was well tolerated in the long-term. It was noted that response to hydroxyurea in children did not always correlate with foetal haemoglobin levels suggesting that other mechanisms may be involved in its therapeutic effect. The main side effect of hydroxyurea is myelosuppression. The risk of malignancy with long term use is a concern although there is no evidence of this in people taking the medication for SCDs.

As already noted, patients in developed countries with better access to these advanced treatments have substantially improved life expectancy and quality of life.

The role of psychological intervention in SCDs is recognised and a guideline for this is being developed in the UK (Dick 2008). There is some evidence that Cognitive Behavioural Therapy may be helpful as an adjunct in pain management. However, despite encouraging results of psychological interventions in SCDs (Anie 2005), the evidence base is still limited (Anie and Green 2002). Thus identification of a wider range of putative risk factors for psychological distress in people with SCD (such as perceived stigma) could lead to new and more effective targeting of psychological interventions. Stigma is a good example of putative risk factors that are yet to be explored in SCD.

1.6. Summary
SCDs affect a large number of people worldwide. Although these disorders are much more common in developing countries, particularly in sub-Saharan Africa, migration means more cases are increasingly being seen in developed countries and in all parts of the world. Thus awareness of the physical treatment and psychosocial complications is essential in all countries. While SCDs carry risk of serious and sometimes life threatening complications, the severity is variable; hence some affected persons can lead a relatively healthy life. Recent treatment advances have improved life expectancy although most of this benefit is still limited to developed countries. The most distressing symptom is painful ischaemic crisis, and there is some
evidence that psychological interventions can be helpful in managing these. A better understanding of psychological factors associated with SCD could lead to new and effective targeting of psychological interventions.
Chapter 2.

Literature review (1): Stigma and sickle cell disease.

2.1. Introduction

In this chapter, I discuss the concept of stigma and the dimensions that determine the stigmatising potential of disorders. I apply the stigma dimensions to SCDs to explore to what extent they are applicable to this condition. I review the literature for empirical evidence of stigma in SCDs and interactions with other types of social disadvantages. The chapter includes a discussion of the social model of disability as a critique of discourses of stigma, which could unwittingly present stigmatised persons as victims. The chapter is not intended as an exhaustive sociological or anthropological exploration of the concept of stigma. Instead, I have prioritised aspects of the concept that are relevant and applicable to SCDs.

2.2. Concept of stigma

Worldwide, the contribution of stigma to the burden of physical disorders is increasingly being recognised (ILAE/IBE/WHO 2003). For disorders such as SCDs that are already associated with potentially serious physical complications, any additional social and psychological distress engendered by stigma is likely to substantially increase associated disability and hardship.

It is customary to commence discussions of stigma with reference to Erving Goffman’s seminal work on the subject. Goffman (1963) described stigma as an attribute that is deeply discrediting. He described how possessing the stigmatising attribute fundamentally intrudes on how others perceive the individual. In the ensuing transaction, the person with the attribute subsequently internalises the associated discredit thereby changing his or her own perception of the attribute. The stigmatised person feels he has been transformed from a normal to a tainted person. For example, the person with the attribute may start to anticipate discriminatory behaviour from others and may experience a reshaping of their emotions and beliefs about themselves and society. Goffman also described how the stigma process could extend to other
people without the attribute but who are connected to the stigmatised person (e.g. relatives). He referred to this as courtesy stigma.

Since Goffman’s work, other sociologists have extended the characterisation of stigma. Of the several types of stigma that have been described, the concepts of Enacted and Perceived stigma described by Jacoby (1994) appear most relevant to the work presented in this thesis. These two types of stigma are particularly important in understanding the impact of stigma on affected individuals and in thinking of appropriate interventions.

According to Jacoby (1994), enacted stigma describes the actual experience of negative and discriminatory behaviour by others against the person with the stigmatising attribute. The resulting distress in the affected individual is clearly linked to an actual experience of ill-treatment. Thus interventions to reduce enacted stigma would be more effective if directed at changing the negative and stereotypical attitudes of the perpetrators.

On the contrary, Jacoby (1994) described perceived stigma as a subjective belief or anticipation that having the stigmatising condition will lead to discrimination by others. The belief may be related to previous experiences of enacted stigma or may not be founded on actual experience (Scambler 2004). According to Heatherton and colleagues, even when the stigmatising attribute is not obvious, those who perceive themselves to be stigmatised often experience psychological distress and have a negative view of themselves (Heatherton, et al., 2003). It is as if the affected persons develop a different view of the world and different way of interpreting events and experiences influenced by possession of the stigmatising attribute (Scambler 2004).

Perceived stigma can have serious disabling consequences due to the tendency by affected individuals to take, sometimes, extra-ordinary measures to conceal their attribute (Scambler 2004; Scambler and Hopkins 1986). Typical consequences of these efforts to avoid disclosure include isolation and loss of social and economic opportunities (Leary et al 1998). The importance of recognising perceived stigma lies in the potential for psychological treatment. For example, because the underlying mechanism in perceived stigma may involve distorted cognitive appraisal, the
associated psychological distress and avoidance could be amenable to Cognitive Behavioural Therapy (Kent 2000).

The impact of perceived stigma can be serious on affected persons. Even for life threatening diseases, perceived stigma could lead affected individuals to make deliberate and seemingly irrational decision not to seek help (Sadavoy et al 2004). Consistent with Goffman’s work, both enacted and perceived stigma can also apply to third parties with links to the stigmatised individual (courtesy stigma).

However, it is important to recognise that despite the possibility of more negative self-appraisal by some stigmatised persons, this is by no means universal. On the contrary, other stigmatised persons show resilience and are able to ward off negative threats to their self esteem (Heatherton, et al., 2003).

Stigma is a ubiquitous and diffuse concept (Weiss et al. 2001), which lends it use to a wide range of diverse processes that have in common a sense of social rejection (Coker 2005). Related concepts, which are sometimes used loosely to infer stigma include, social rejection, negative attitude, prejudice, discrimination, and social embarrassment. Also stigma has cultural and situational dynamism; hence what is considered stigmatising in a particular historical or cultural context may not be at a different time or place and could even become a positive attribute.

This thesis is focused on “Felt or Perceived” stigma. This is in part because examining this type of stigma could potentially lead to development of individually targeted psychological interventions for people with SCDs. While proponents of the social model of disability would argue that the distress associated with stigma is due to societal attitudes and that interventions should therefore focus on changing societal perceptions, there is good evidence from conditions like epilepsy (Westbrook et al 1992) to suggest that self-perceived stigma is also associated with direct psychological distress to affected persons. This association justifies studying and intervening directly against self-perceived stigma.
2.3. Impact of stigma

Stigma associated with mental and physical illnesses have adverse impact on a range of outcomes. The serious impact of stigma is probably best illustrated with reference to HIV-AIDS. Several studies have shown a close association between HIV-AIDS related stigma (measured by desire for social distance) and reduced utilisation of voluntary counselling and testing (VCT) services (Hutchinson and Mahlalela 2006, Iliyasu et al. 2006, Babalola 2006), and disclosure of HIV status (Kilewo et al. 1999). This combination of reduced VCT and disclosure have serious implications for controlling HIV transmission. Uys (2003) found that in South Africa, relatives of patients with terminal AIDS could not be given appropriate emotional support because the terminally ill patients had refused permission to discuss their HIV-AIDS.

Stigma also interferes with grieving. Frohlich (2005) noted that in South Africa, some relatives of patients who have died from HIV-AIDS avoid grieving openly for fear of courtesy stigma. Cluver and Gardner (2007) found that in addition to other stressors like bereavement, physical abuse, poverty and loss of contact with remaining family members, enacted stigma was a significant contributor to the distress of children orphaned by HIV-AIDS in South Africa.

With respect to conditions more comparable to SCDs, several studies have shown associations between self-perceived stigma and higher levels of psychological and emotional distress in psoriasis (Richards et al 2001, Leary et al 1998), vitiligo (Kent 2000), and epilepsy (Westbrook et al 1992; Austin et al 2004; Adewuya et al 2006).

2.4. Epilepsy as a model for stigma in chronic physical disorders like SCD

Epilepsy presents a good model for exploring stigma in chronic conditions like SCD. Several studies of epilepsy in both adults and children have demonstrated that the condition is highly stigmatising. These findings apply to studies that explored both enacted and self-perceived stigma (Jacoby 1994) and those that measured social distance (Austin et al 2002), and prejudice (Fernandes et al 2007) against people with epilepsy. Also the findings have been shown in both developed (Westbrook et al 1992) and developing (Adewuya et al 2006) countries. The world-wide recognition of the prevalence and adverse impact of stigma on people with epilepsy prompted the
international campaign “Out of the shadows” by the World Health Organisation, International League Against Epilepsy, and International Bureau for Epilepsy in 1997 (ILAE/IBE/WHO 2003). This thesis is not intended for detailed discussion on epilepsy; hence only illustrative literature on children is highlighted.

In a test of theoretical model of stigma among children with epilepsy in United States, Westbrook and colleagues found a high prevalence of self-perceived stigma and adverse impact on self esteem (Westbrook et al 1992). This study demonstrated a reliable methodology for assessing self-perceived stigma, which I adapted for this study. Also in United States, a large scale survey of school children found significant social distance towards peers with epilepsy. For example, 69% of the sample would not date an affected person (Austin et al 2002).

A recent study in Nigeria found a negative association between self-perceived stigma and school achievement of adolescents affected by epilepsy (Adewuya et al 2006). Also, a review from the same region in Africa (Baskind and Birbeck 2005) found epidemiological, anthropologic and sociologic evidence that epilepsy attracts a very negative public perception.

Other reviews of the literature on epilepsy and stigma in adolescence have found that stigma is common and adversely affects the quality of life of affected young people (e.g. MacLeod and Austin 2003).

2.5. Stigma dimensions and application to SCDs

Stigma dimensions predict how others are likely to respond to the possession of a potentially stigmatising attribute. Thus these dimensions help to understand why certain attributes and not others become stigmatising. Katz (1981) and Jones et al (1984) described several interrelated stigma dimensions including: Visibility, Threat or Peril, Chronicity, Responsibility, and Disruptiveness.

The dimension “Visibility” refers to the extent the attribute is obvious, concealable, or aesthetically challenging to others. In general, stigma theory predicts that the more visible and disfiguring an attribute the more stigmatising it is likely to be. Some
people with SCD have easily recognisable physical manifestations such as jaundice, leg ulcers, and delayed physical development (Dick 2008). Severe cases, especially where effective treatments are not widely available, may be associated with gross physical signs such as gnathopathy (Wessberg et al. 1980), and bossing of the forehead (Acquaye et al. 1985). While widespread access to disease-modifying treatments (Atweh and Schechter, 2001) has made these gross signs uncommon in Western countries, they are still common signs in many developing countries where the vast majority of people with SCD live. Thus the visibility of the physical signs of SCDs increases the potency for stigma.

The stigma dimension of “Threat or Peril” is to do with the perceived danger posed to others by virtue of a person possessing the attribute. Consistent with this dimension, it is well recognised that having a potentially fatal infectious disease such as Tuberculosis is stigmatising. In some non-Western societies, inaccurate beliefs that associate SCD with peril are still common. For example, a recent survey of secondary school students in Nigeria found that 9% of the students believed that SCDs are infectious (unpublished data). Also in Nigeria, 8% of relatives attributed SCDs to malevolent spirits of reincarnation ((Ohaeri and Shokunbi 2001; Nzewi 2001). SCDs are therefore likely to be stigmatised in these settings where a significant proportion of the society hold threat-promoting views about the condition. Hinshaw (2005) suggests that the kind of demonological attributions described above are associated with increased stigma.

The dimension of “Chronicity” predicts that long lasting conditions would be more stigmatising than acute short-lived disorders that leave no permanent marks. SCD disease is essentially a chronic life-long disorder with only a small chance of cure for a minority of affected persons (through bone marrow transplantation). Although some affected persons are able to enjoy prolonged periods of good health, the underlying genetic disorder does not change (except for a minority who undergo successful bone marrow transplantation). Atkin and Ahmad (2001) found that among young people with SCDs, even when they are stable and free from acute events, they still worry about their future health. This partly explains why the treatment paradigm for the condition is sometimes described in principles of palliative care (Bevan 1998; Ballas 2005). The chronic nature of SCDs therefore suggests increased likelihood of stigma.
The stigma dimension of “Disruptiveness” describes the extent to which possessing the attribute interferes with interpersonal relationships. Disruptiveness is also related to other dimensions like Chronicity, as more severe and long-standing disorders tend to be also more disruptive. Although the course of SCDs are variable and many affected persons live relatively healthy undisrupted lives, a proportion require frequent hospitalisation as a result of different acute illness episodes particularly pain (Wethers 2000). In addition, some experience even more frequent but less severe episodes not requiring hospital admission but nonetheless necessitating rest at home. The limitations imposed by these illness episodes could be disruptive to schooling, employment, and social encounters (Atkins and Ahmad 2001). Also disruptions make concealment more difficult and increase the potential for both enacted and self-perceived stigma. The resulting threat of unwanted disclosure could be a source of dysphoria for affected persons.

The dimension of “Responsibility” refers to the assumption that people are more likely to experience stigma if they are considered in some way personally responsible for acquiring the negative attribute. Although people affected by SCDs are clearly not responsible for acquiring the disorders, in communities where misinformation about the disorder is prevalent, affected persons may be blamed unfairly. The previous reference associating SCDs with malevolent spirits in Nigeria (Nzewi 2001) is a good example.

2.6. Evidence of stigma in sickle cell disease

Although Goffman’s seminal work on stigma was based mainly on mental disorders, the dimensions and characterisations of stigma have been successfully applied to a wide range of physical disorders such as epilepsy (Westbrook et al 1992) and stuttering (Blood et al 2003).

2.6.1. Literature search strategy

The literature on stigma in SCD is extremely limited. For the literature search, five electronic databases (Medline, Embase, PsycINFO, CINAHL, and Social Science Citation Index) were searched from their inception to June 2009. Specific MeSH terms were used for searching Medline and EMTREE terms were used to search
Embase. The search terms were divided into two groups. The first group had search terms related to sickle cell (sickle, sickle cell disease, sickle cell anaemia, sickle cell disorders, and haemoglobinopathy). The second group of search terms had terms related to stigma (stigma, discrimination, stereotype, negative attitude, prejudice, and disadvantage). Terms from each group were searched in combination using the OR function (for same group) and combined with the search outcome of terms from the other group using the AND function. The initial selection criteria focused on studies that:

- examined self-perceived stigma, and
- whose subjects had sickle cell disease

The search was filtered by age group (children) and language (English). The first search identified no study that met these criteria. The search criteria were therefore extended to include any study on any aspect of stigma or related subject on people with sickle cell disease of all ages (including carers), and in any language.

These new and very wide search criteria identified only four studies of direct relevance. The reference lists of the studies identified were also searched but this yielded no additional studies of relevance. Of the four studies identified, one involved mothers of children with SCDs (Burnes et al 2008), the second involved adults with SCDs (Sanker et al 2006) and the third and fourth studied young people with SCDs (Adedoyin 1992, Atkin and Ahmad 2001).

2.6.2. Discussion of identified studies

Burnes and colleagues conducted in-depth interviews with ten Canadian mothers of African and Caribbean origin whose children had SCDs (Burnes et al 2008). The mothers were recruited from a specialty SCDs clinic. The interviews explored the mother’s coping strategies as well as their perceptions of negative attitudes towards SCDs. All but one of the mothers had experienced SCDs-related stigma. The mothers were keen to keep their child’s sickle cell disorder secret for fear the children would be stigmatised. They reported negative public perceptions including beliefs that SCDs are infectious, or represent an ancestral curse on the family. Some of the mothers reported being blamed for knowingly conceiving an ill child. This study was limited
by the fact that it did not seek information about stigma from the children with SCD or their fathers. Also the findings are difficult to generalise given that respondents were all recruited from a specialist clinic for SCD.

In a study to explore the relationship between genetic aetiology and potential for stigma, Sankar and colleagues interviewed eighty American subjects made up of people with SCDs, cystic fibrosis, cancer and deafness (Sankar et al 2006). The respondents were recruited from support groups, clinics and through community meetings. The study found that contrary to common belief, genetic aetiology did not automatically or universally confer stigma on affected individuals. Instead, stigma appeared more related to the varied experiences of particular individuals. The authors compared people with cystic fibrosis with those with SCDs and noted that the latter group were more likely to report negative experiences or feelings about their condition even though the physical outlook is more favourable for SCDs in that setting. One limitation of this study is that it failed to account for confounding factors such as racism. Given that most people with SCD are of Black ethnicity and people from this ethnic group are at more risk of experiencing racism, the more negative experiences reported by the SCD group compared with the Cystic Fibrosis group may be in part due to racial discrimination.

Adedoyin (1992) explored the attitudes of Nigerian adolescents with SCDs towards having the disorder. The study found that the dysphoric adolescents attributed their unhappiness to a range of disease related life limitations including “a sense of shame in public”. The study was not designed to primarily examine stigma; hence the subject was not explored in any detail.

In a qualitative study of 26 young people with SCDs and 25 with thalassemia major, Atkin and Ahmad (2001) found that those in their mid teens resisted anything that marked them out as different including adherence to their treatment regimes. They report that young people with these conditions felt that due to ignorance, “disablism” and racism, people in their wider social network were insensitive to their concerns. Atkin and Ahmad did not explore stigma in detail as it was not the main focus of their study.
Other recent psychosocial reviews on SCD acknowledge the likely adverse impact of stigma (Anie 2005; Helps et al. 2003). Additional evidence comes from social-anthropological studies and everyday life examples. I explore these next.

2.7. Social anthropological evidence

Weiss and colleagues have highlighted the importance of cultural perspectives in considering stigma (Weiss et al. 2001). In many societies where SCD is prevalent, the condition and the associated high-infant-attrition rate is attributed to malevolent spirits (Onwubalili 1983; Nzewi 2001). In Nigeria – the country with the highest prevalence of SCD, local terms associated with the condition (e.g. Ogbanje) connote malevolent spirits linked with reincarnation (Nzewi 2001). Historically, such demonologic views tend to attract harsh and punitive responses (Hinshaw 2005). In Nigeria for example, the evil-spirit link with SCD results in practices that includes amputation and mutilation of newborn babies suspected of possessing the attribute (Nzewi 2001). The mutilation marks are intended to assist with identifying the child with the malevolent spirit in the event of his/her reincarnation.

In my personal experience, (which includes childhood, medical training and working as a doctor in Nigeria), the local term stated earlier (Ogbanje) is well recognised as deeply discrediting. In fact, ordinary children use the term as “swear word” to put-down peers who are non-sickle cell sufferers. Also the association with malevolent spirit of reincarnation is so pervasive and powerful that surviving children in affected families gradually acquire names with themes of death (Nzewi 2001). In the study by Sankar and colleagues, some respondents with SCD reported similar childhood experiences in which their peers used SCD to “say the cruellest things” about the respondents’ families (Sankar et al. 2006).

The presence of even one obvious sign of SCDs could be associated with a disproportionate impact on the life of affected individuals. For example, Alleyne and colleagues assessed the psychological, social and economic impact of leg ulceration on people with SCDs in Jamaica. Compared with a control group with no leg ulceration, the ulcer-affected group experienced wide ranging adverse psychosocial effects including on education, employment, and marriage (Alleyne et al. 1976).
However, it is also possible that leg ulcer is a marker for severity and or other socio-economic disadvantages that could explain the adverse impact on education, employment and other psychosocial variables. Also the social circumstances in Jamaica may have changed substantially in the past 30 years as to question the current relevance of this finding.

Another evidence of stigma in SCDs comes from everyday language. People with SCD are sometimes referred to as “Sicklers” both in general language and in published literature (Akuse 1996). This description, which identifies the individual with their disorder, increases stigma (Slovenko 2001). Although intended to describe a person with SCD, the term “Sickler” also unwittingly conveys the impression of someone who is frequently ill. Coincidentally, this can be the reality for some people with severe forms of SCDs.

2.8. Sickle cell disease stigma and disadvantage.

Stigma may be related to some of the physical and psychosocial disadvantages associated with SCD. Access to pain control may be a good example. Although I am not aware of any good evidence to suggest malicious practice or deliberate discrimination in the clinical care of people with SCD, Anionwu (1996) has suggested that some hospital staff view patients with SCD as “difficult” and that staff’s stereotypical beliefs can unwittingly result in inadequate management of SCD patients’ painful crisis. While acknowledging the legitimate risk of iatrogenic opiate dependence, concerns about less optimal opiate prescribing for pain management in SCD have been noted (Bevan et al. 1996).

Despite well-intentioned legislative frameworks aimed at protecting people with disabilities from discrimination, many examples of discriminatory practices against people with SCD are still noted in access to jobs and health insurance in the United States (Kass et al. 2004) and the UK (Atkins and Ahmad 2001). These may be indicative of persisting high levels of prejudice against people with SCD. Also if, as is sometimes reported to be case, people with SCDs have to literally fight for necessary services and allowances, the resulting frustration could accentuate the perception of stigmatisation by affected individuals.
There is recent evidence suggesting inappropriate use of SCD by Criminal Justice Systems to explain death of Black people in custody. Dyson and Boswell (2006) reviewed ten cases of sudden deaths among Black people in custody in United States and United Kingdom. Their findings suggested that SCD was misused to explain all ten deaths. The authors reviewed another seven deaths of people with SCD while in custody and suggested that inadequate SCD-care while in custody contributed to these fatalities. It is however uncertain whether these adverse events were borne out of the law enforcement officers’ ignorance of the needs of this client group or actual prejudice.

Sickle cell disease stigma may also interact with racism. Just like racism, stigma processes are thought to be consistent with concepts of class and command (Scambler 2004). Parker and Aggleton (2003) point out that stigmatisation functions at the point of intersection between culture and power. As a result, they highlight the importance of understanding the framework that promotes the interests of dominant groups and differential understanding of values and worth, which are the processes that facilitate stigmatisation (Parker and Aggleton 2003). Interaction of stigma and racism could be particularly important in SCD as the disorder affects predominantly people of Black African origin – a minority group who are already at significant risk of racial stereotyping (Kushnick 1988). Explicit and deprecatory racial attitudes are no longer widely expressed, but subtle and implicit racial stereotyping is still common (Hinshaw 2005). Racial factors may have contributed to the more negative experiences reported by SCD sufferers compared with cystic fibrosis patients in the study by Sankar and colleagues cited earlier (Sankar et al. 2006).

### 2.9. Courtesy stigma

Relatives of a person with the stigmatising attribute can experience “courtesy stigma” (Goffman 1963). Courtesy stigma is known to worsen the subjective burden of care on relatives. Fear of courtesy stigma results in concealment and secrecy, which limits access to family support (Hinshaw 2005).
In pure genetically determined conditions like SCD, parents and siblings are more likely to receive courtesy stigma (Hinshaw 2005). Parents may be unfairly blamed by their immediate community (Sankar et al. 2006; Burnes et al. 2008) or blame themselves resulting in high levels of guilt (Murray 1976). As siblings of people with SCD could be heterozygous carriers of the sickle cell gene, their peers may overlook them when considering long-term relationships (Bamisaiye et al. 1974).

2.10. Special consideration for adolescents with SCD

As this project focused mostly on adolescents with SCDs, it is important to consider the particular and special implications for SCDs stigma in this age group. For several reasons, there is concern that adolescents with SCD may be particularly vulnerable to stigma. It is already a major challenge for some adolescents to effectively negotiate the complex biological and social transitions associated with this age group (Dornbusch et al. 1991). The concern is that superimposing the demands of a serious and potentially stigmatising illness like SCD on such a complex system complicates an already difficult process (Hilton et al. 1997).

In some people, SCD is associated with pubertal delay (Pinckney and Stuart 2004). Many studies have shown that for boys in particular, delayed puberty is stigmatising and associated with low self-esteem (Alsaker 1996). For example, in a study of young adults with SCD in Jamaica, low body mass index was a feature in all the male subjects with a psychiatric disorder (Hilton et al. 1997). Because adolescence is a critical time for development of self-identity (Alsaker 1996), stigma engendered low self-esteem at this stage of development could have significant long-term adverse impact.

Individuating from families and formation of other long-term relationships is one of the key tasks of adolescence (Alsaker 1996). The genetic basis for SCD and the potential stigma engendered by this could make this task challenging for both people with SCD and their siblings (Sosan 2006). Concealment is often the stigma management strategy of choice for people with stigmatising conditions (Link et al. 1991). While this limits the risk of experiencing prejudice, it also results in isolation and missing of important social opportunities especially in adolescence. The Jamaican study cited previously (Hilton et al. 1997) found that compared with non-SCD
controls, young adults with SCDs were less likely to be in stable relationships or to have children.

2.11. Resilience
Although research on chronic health problems is often driven by deficit models (Reynolds 1992), which emphasise maladjustment and difficulties, it is important to recognize that negative outcome in SCD is not inevitable. There is evidence of resilience in both affected persons and their families (Robinson et al. 1995; Ohaeri and Shokunbi, 2002).

While SCD is a chronic illness, the course is characterised by episodes of acute illnesses separated by periods of relative well-being (Smith 1991). Many people with mild SCD disease can have lengthy periods of well being (Thomas et al. 1997). Also, the use of disease-modifying treatments such as Hydroxyurea is increasingly keeping many people with SCD free from acute events for longer periods (Atweh and Schechter, 2001). Research to document and highlight the course and positive outcomes in SCD could therefore help to challenge the therapeutic nihilism that might be contributing to stigma in SCD.

2.12. Methodological issues in the assessment of stigma
Enacted stigma can be assessed either from the perspective of the person with the stigmatising condition or from the perspective of people without the condition. From the former point of view, enacted stigma is commonly measured by enquiring from affected persons about actual experiences of discrimination and prejudice attributable to possessing the stigmatising condition. From the latter perspective, enacted stigma is assessed by measuring self reported attitudes such as social distance or behavioural rejection using fictional vignettes portraying or role playing the stigmatising condition. The Social Distance Scale originally developed by Borgadus is a commonly used example of this type of measurement. Versions of the original scale have been adapted to measure attitudes towards a wide range of situations and the measures generally show good reliability. However, there are two major threats to their validity. First, as the scenarios given to respondents are hypothetical, it is uncertain if they would give the same responses in similar but real situations.
Secondly, the validity is limited by the respondent’s tendency to provide socially desirable responses to controversial questions (van de Mortel 2008). For example, in an extensive review of over 14,000 studies measuring attitudes, van de Mortel (2008) found that only 31 studies (0.2%) attempted to identify socially desirable responding and of these, nearly half found that socially desirable responding influenced their results. Similarly, Hinshaw (2005) argues that changing social norms have made overt racist responses unlikely even though over-learned and hidden racial prejudices are still held by many individuals.

Thus the validity of Social Distance Scales can be enhanced by simultaneous measurement of the social desirability of the respondent’s answers. This measure can be used to eliminate respondents with unacceptably high socially desirable responses or social desirability can be controlled for using partial correlation or hierarchical regression techniques (Nederhof 1985). A commonly used reliable measure of social desirability is the Marlowe-Crowne Social Desirability Scale (Crowne and Marlowe 1960) and its shorter versions (e.g. Loo and Thorpe 2000).

Another method to minimise the threat of socially desirable responding in the assessment of enacted stigma by people without the stigmatising condition is the use of measures that are less overt than self reported behavioural rejection. The more subtle, less conscious and implicit measures (e.g. Implicit Association Test) (Teachman et al 2006) are recognised to be less prone to eliciting socially desirable responses.

The assessment of self perceived stigma typically involves eliciting the beliefs and perceptions of affected persons on how non-affected persons would behave in different interactional situations. This methodology is subject to limitations including that any perceived negative attitude reported by affected persons is likely to be simultaneously influenced by a myriad of other personal (e.g. pre-existing depressive illness) and ecological (e.g. social support) factors. Such reports are also likely to be prone to recall bias whereby a person with the stigmatising condition that has experienced discrimination would be more likely to interpret and report other neutral behaviours as discriminatory. Being a measure of attitude, the difficulties already noted regarding socially desirable responding would also apply.
2.13. Social model of disability applied to SCDs

The social model of disability argues that for disabling conditions like SCDs, the real cause of disability are social barriers and negative attitudes rather than the actual physical impairments that may be associated with the condition. The social disability model argues against explanatory models, which they see as unwittingly victimising affected persons by locating the difficulties associated with their condition entirely within the affected person. For example, in relation to stigma and negative attitudes, the social model of disability would argue that instead of people with SCDs adjusting to cope with negative attitudes, it is other people who should be adjusting their negative attitudes towards people with SCDs (Atkin and Ahmad 2001). Proponents of the social disability model attempt to distinguish this model from paradigms that focus on the individual impairments or deficits associated with the condition – sometimes referred to as the medical model. They argue that discourses of SCDs should not stop at the level of the individual but encompass social and political aspects. However, in reality, both the social disability model and so called medical model offer useful and practical understanding of the difficulties experienced by people living with chronically impairing and disabling conditions like SCDs. A good example is the frequent association between greater severity of disease and greater disability.

2.14. Summary

I have shown in this chapter that models of stigmatisation can be applied to SCDs. Specifically, several stigma dimensions show a good fit with the bio-medical and psychosocial aspects of SCDs. Despite paucity of direct empirical research, some evidence from everyday life and socio-anthropological studies suggest that people affected by SCDs may be at risk of stigmatisation.

Although I have argued that stigma dimensions developed in studies of mental and other physical illnesses are applicable to SCD, this remains to be tested. There is inadequate research into the perception and or actual experiences and impact of stigmatisation on people affected by SCDs and their families. This is a crucial gap given the evidence from other stigmatising physical conditions (e.g. epilepsy)
indicating both high prevalence and adverse impact of stigma. The project completed for this thesis is designed to start filling this gap by exploring the prevalence and psychosocial associations between SCDs and self-perceived stigma.

It is appropriate to recognise the valid critique of the concept of self-perceived stigma by proponents of the social model of disability (e.g. Atkin and Ahmad 2001), who argue that while SCDs are physical disorders, self-perceived stigma is a socially constructed concept that could imply that people with SCDs are victims. However, there is also good evidence from conditions like epilepsy (Westbrook et al 1992) to suggest that self-perceived stigma is a valid phenomenon that is associated with direct psychological distress to affected persons. This association justifies studying and where appropriate intervening directly against self-perceived stigma.
Chapter 3.

Literature review (2): Psychosocial adjustment and psychopathology in sickle cell disorders

3.1. Introduction

This chapter is focused on exploring the factors that contribute to psychological adjustment in children with SCDs. As the focus is on children, the evidence examined to quantify the level of psychological difficulties is limited to studies involving this age group. Studies assessing levels of general psychopathology in children with SCDs are discussed first. Due to hypothesised differences in levels of psychopathology between children with SCDs living in developing and developed countries, the evidence is examined separately for these two regions. In addition to general measures of psychopathology, I have included an extended discussion of depression. This is because of the seriousness of depression in terms of impairment and risk of suicide. Risk factors and the prevalence of depression in children with SCDs are explored. Evidence is drawn from studies of both children and adults with SCDs to discuss general principles or mechanisms. However, evidence used to quantify the problem is limited to studies of children.

3.1.1. Literature search strategy

Four electronic databases (Medline, Embase, PsycINFO, and CINAHL) were searched from their inceptions to June 2009. Specific MeSH terms were used for searching Medline and EMTREE terms were used to search Embase. The search terms were divided into two groups. The first group had search terms related to sickle cell (sickle, sickle cell disease, sickle cell anaemia, sickle cell disorders, and haemoglobinopathy). The second group had search terms related to psychopathology (psychological, psychosocial, psychiatric, mental health, mental illness, depression, anxiety, and self esteem). Terms from each group were searched in combination using the OR function (for same group) and combined with the outcome of search with terms from the other group using the AND function. The search was filtered with language (English) and age group (children). Although only publications in English
language were sought, one publication in French was included because it came from Africa where studies on SCD are very limited.

For purposes of quantifying the burden of psychological difficulties and depression in studies were selected if:

- subjects were children with SCD
- used an explicit measure of psychopathology or psychological function
- had a control group or used measures with established population norms

The reference lists of identified studies were searched for additional relevant publications.

3.2. Determinants of psychosocial adjustment in children with SCDs

Several factors are known to contribute to the psychosocial well-being of young people with SCDs. These factors could be explored using either the bio-psychosocial model or ecological models of aetiology (Bronfenbrenner, 1979). Using the bio-psychosocial approach, the factors could be categorised into three domains – biological (e.g. disease process), psychosocial (e.g. personal attitude to disease), and social (e.g. family function) (see Figure 1). The ecological approach (Figure 2) would consider individual factors (e.g. disease severity, temperament), factors in the immediate family environment (e.g. socio-economic status, family function) and factors in the wider environment (e.g. societal attitudes, availability of treatments). Although these models are primarily heuristic and illustrative, they have good face validity and are supported by empirical data.

While it is helpful to consider both models separately, in reality, they overlap. For example, the “social/environmental” component of the bio-psycho-social model incorporates the family and environmental aspects of the ecological model. It is also important to recognise that the relationships within the models are not linear and unidirectional. Instead, the components of both models have complex interactional and multidirectional properties. For example, parental relationship could be strained by the stress on parents supporting the needs of a child with a very severe form of SCD. On the other hand, parental marital problems not related to the child’s SCD
could nonetheless lead to a worsening of the child’s health if parents become too preoccupied by the conflict between them and neglect to administer the child’s treatments or keep hospital appointments. Another factor to consider in evaluating these models is that each domain contains not only risk factors but also resilience and protective factors. Thus the actual likelihood of psychopathology is dependent on the balance between risk and protective factors. For example, a child with a severe form of SCD may thrive psychologically in a very supportive and nurturing family and school environment.

**Figure 3.1. Bio-psycho-social framework**

Risk of psychopathology increases with additional factors in each domain

-Most risk for psychopathology
3.2.1. Biological

It is recognised that children with more severe indices of SCDs are at more risk of psychological complications. However, this association is not inevitable as some young people with SCDs show resilience and hardiness. For example, in a longitudinal study of children with SCDs, Getzoff (2005) found that disease severity did not contribute significantly to long-term emotional outcomes.

Apart from overall severity, the presence of specific physical complications are known to increase the risk of maladjustment in SCDs. For example, in a study in Jamaica, Alleyne and colleagues found a significant positive association between the presence of leg ulceration in people with SCDs and psychological distress (Alleyne et al. 1976). The neurological complications of SCDs are now well recognised. Up to 11% of affected persons suffer overt strokes and up to 20% have evidence of ischaemic brain damage on MRI by the age of 20 years (Pegelow et al 2002). Given that brain ischemia is a recognised independent precursor of psychological and cognitive difficulties, people with SCDs and ischaemic complications would be at increased risk.
3.2.2. Psychological
Several psychological and temperamental factors are known to contribute to adjustment in people with chronic physical disorders such as SCD. As part of the Cooperative Multi-site Study of SCD in United States of America, Burlew and colleagues assessed 90 American adolescents with SCD to determine the relative contributions of psychosocial and biomedical factors to the adolescents’ adaptation to SCD. The subjects were randomly selected from among patients with HbSS genotype enrolled in the Cooperative Study of SCD. The study showed that psychological factors such as self-esteem and assertiveness predicted adjustment while biomedical factors (indices of medical severity) did not (Burlew et al 2000). However, this conclusion needs to be considered cautiously as the finding may have arisen from use of inadequately sensitive biomedical indices in the study. The study was also limited by reliance on self-report psychological measures. As already highlighted, people with SCD are at a substantially increased risk of ischaemic strokes, which could lead to a wide range of neuro-cognitive deficits and behavioural difficulties (Bonner et al 1999).

3.2.3. Social/environmental factors
Families, peers and schooling experiences play an important role in children’s adjustment to SCDs. In a study of 182 young people with SCD, Barbarin and colleagues found that the best predictors of the affected child’s psychological adjustment included parental psychological function and relationships with parents and siblings (Barbarin et al 1999). The study cited earlier (Burlew et al 2000) also found that social ecological factors such as family relationships and social support were better predictors of adjustment compared with biomedical factors.

An important aspect of the social environment that could mediate disease adjustment in SCDs is area of residence. Even in the same country or region, urban or rural residence could make a difference to people with SCDs. For example, in a recent study in Jamaica, Asnani and colleagues found that living in a rural location compared with an urban area was associated with better self-rated quality of life among people with SCDs (Asnani et al 2008). This finding was contrary to the researchers’ expectation.
Although people with SCDs living in developed countries have obvious advantage over those in poorer countries with respect to access to advanced treatments, this does not always translate into psychological advantages. For example, when Thomas and colleagues (Thomas et al 2001) compared 30 patients with SCDs in London with an equivalent number in Jamaica, they found that those in Jamaica were less anxious and had lower levels of perceived pain and emotional response to pain. While this finding is useful, it is possible that the difference arose from inadequate baseline matching between the two groups. It is also possible that relative lack of pain relief had conditioned the Jamaican cohort to perceive pain less readily.

One important aspect of the social environment which is increasingly being recognised is the attitude of unaffected persons towards people with SCDs. There is good evidence that stigma contributes to psychosocial difficulties among children with chronic physical conditions such as epilepsy (Westbrook et al 1992 and Adeweya et al 2006). Although a similar impact could be hypothesised for SCDs, the evidence is very limited (see chapter 2). The study undertaken for this thesis aims to explore this evidence.

3.3. Psychopathology in children with SCDs

Epidemiological studies of childhood mental disorders such as the Isle of Wight study (Rutter et al 1976) and the British Survey of the mental health of children and adolescents (Meltzer et al 2000) consistently find increased risk of psychopathology among children with chronic physical conditions particularly disorders that involve the brain. In fact, Meltzer and colleagues found that having any physical health complaint increased the odds of a having a mental disorder by 82% (Meltzer et al 2000). Research data suggest that these findings may be applicable to SCDs (Anie 2005). Some studies of children with SCDs have shown increased rates of emotional difficulties, disruptive behaviour, and low self esteem among children with SCD (Helps et al 2003). However, Helps and colleagues suggested that recent improvements in the physical care of people with SCDs might have resulted in reduced rates of psychopathology in this group. While this may be the case in developed countries where advanced treatments are more widely available, a recent review found no evidence of reducing psychopathology in SCDs in developing
countries (Ani and Hodes 2006). Giving the differences between developed and developing countries in terms of access to treatments, the literature on psychopathology will be discussed separately for both parts of the world.

More recent studies in developed countries appear to consistently show closer to normative levels of psychopathology in SCDs. For example, a recent study of children with SCDs in USA (Noll et al 2007) found no differences in measures of emotional wellbeing compared with their unaffected peers. In this questionnaire based which compared 43 children with SCD and an equal number of healthy controls, teachers described the children with SCDs as more prosocial and less aggressive. These results are similar to what the authors found 10 years earlier (Noll et al 1996). These studies have many strengths including use of muti-informant data collection. However, the positive outcome described might be related to the fact that they included only children with SCDs who had not had an overt stroke. Also the studies recruited children with HbSS, HbSC, and HbBthal but the varying severity associated with these gentotypes was not accounted for in the analysis. The two studies (Noll et al 1996 and Noll et al 2007) were both limited by small sample sizes. Also the representativeness of the sample is limited by the fact that all the children with SCD were selected from a reputable nationally funded Centre with a comprehensive SCD service. Finally, the authors included five white children in the control group because there were no suitable black controls.

Another study of 36 adolescents with SCDs in USA (McElligott 2006) found that the young people did not record higher than normative scores in different measures of self esteem, anxiety, depression and behavioural difficulties. Further, a longitudinal study in USA, which followed up 48 children with SCDs and 49 healthy controls for up to 10 years found no differences in measures of depression, self worth and internalising symptoms between the two groups either cross-sectionally or longitudinally (Getzoff 2005). In an earlier study in the UK involving 39 children with SCD and 24 controls, Midence and colleagues found no significant differences between the two groups on depression and self esteem (Midence et al 1996). However, the SCD group had a significant reduction in intellectual ability and an increase in behaviour problems. While these studies benefitted from multi-informant data collection, they were all limited by sole reliance of questionnaires to ascertain psychopathology.
Unlike the above studies from developed countries, studies in developing countries consistently show increased rates of psychopathology among children with SCDs. Most studies of psychopathology in developing countries have been conducted in Nigeria. An early case-control study of 84 children with SCD matched with 84 healthy controls found parent and teacher-rated psychiatric morbidity using the Rutter’s scale in 27% of the children with SCD compared with 5% of the controls (Iloeje 1991). Another Nigerian study which compared 100 children with SCD with 75 children with asthma and 75 children with other acute non-SCD medical conditions also using the Rutter scales found higher rates of psychiatric morbidity in children with SCD (30%) compared with children with asthma (25%) or acute medical illnesses (20%) although these differences were not statistically significant (Ayinmode and Adelekan 2005). The most recent study of 135 children (45 with SCD, 45 with Juvenile-onset Insulin Dependent Diabetes Mellitus and 45 healthy controls) also from Nigeria (Bakare et al 2008) found increased rates of DSM-IV emotional disorders in the SCD group (38%) compared with healthy controls (11%). This is the only study of psychopathology in SCDs from Africa based on structured interview for DSM-IV diagnoses. Incidentally the children with SCDs had lower rates of emotional disorders compared with children with juvenile-onset Insulin Dependent Diabetes Mellitus (42%). However, the authors noted that 20% of the children with SCDs had experienced suicidal ideation in the previous year compared with 11% of the diabetics and none of the healthy controls. The high level of psychopathology among children with SCD in this study may be partly explained by the fact that only children with homozygous HbSS genotype were included. This genotype is the most severe form of SCDs. Also the authors did not assess for, hence could not exclude children with cerebral involvement.

In summary, recent evidence from developed countries suggest improving psychological adjustment for children with SCDs compared with non-affected children or siblings. It has been suggested that this improvement might be secondary to the hugely advanced physical care available in these regions, which has in turn led to improved physical, social and psychological well-being and longevity. Unfortunately, the opposite is the case in many developing countries where advanced physical care is lacking. It is not surprising that increased risk of psychological
distress continues to be demonstrated in these regions albeit in common with other chronic conditions.

3.3.1. Depression in sickle cell disease

In addition to considering studies of general psychopathology in SCD, it is important to give specific consideration to depression because of its association with significant impairment and risk of mortality from suicide. Given the similarity between SCDs and epilepsy in terms of chronic disease model, the significance of depression in SCDs could parallel that in epilepsy where life-time prevalence of depression is up to 30% making it the commonest co-morbid mental illness in that condition (Kanner 2003). In this section, I will explore difficulties with recognising depression in SCDs, the potential risk factors, and review the evidence for depression in children with SCDs. The literature on adults with SCDs is drawn on where appropriate to illustrate general principles and mechanisms. However only literature on children with SCDs is considered in the examination of evidence for depression in this age group.

3.3.2. Diagnostic issues for depression in SCDs

Before exploring studies of depression in SCDs, it is helpful to contextualise the disorder to SCDs. In particular, an understanding of the difficulties in diagnosing depression in people with SCDs is essential.

Depression is typically diagnosed in the presence of a combination of psychological and physical symptoms that are sustained over a period of time – usually two weeks or more. The psychological symptoms include low mood, anhedonia, inappropriate guilt, low self worth, poor concentration and suicidal ideation, while physical symptoms include lethargy, poor sleep and appetite and reduced libido.

The first difficulty in assessing depression in SCD is the overlap between some of the physical symptoms of depression and SCDs (e.g. lethargy) (Alao and Cooley 2001). A study by Yang and colleagues illustrates this well (Yang et al 1994). These researchers administered the Child Depression Inventory (CDI) to children with SCDs and controls and compared the risk of depression from CDI scores with the outcome of a diagnostic clinical interview for both groups of children (Yang et al 1994). They found high rates of depressive symptoms on the CDI among children with SCDs.
(29%) compared with controls (12%). However, clinical interview found no difference in rates of depressive disorder between the two groups. Further analysis found that questions relating to fatigue and physical complaints in the CDI accounted for the high false positive rates for depression among children with SCDs.

These findings also raise a general need for caution in interpreting studies where depression is measured with self-report questionnaires, which are designed to screen for but are not diagnostic of depression. The use of questionnaires also raises issues about what is appropriate cut-off. This difficulty is illustrated by Schaeffer and colleagues who used the Center for Epidemiologic Studies-Depression Scale (CES-D) to screen for depression in adults with SCDs. They noted that the percentage of patients categorised as depressed dropped from 43% to 18% following a minor change in the cutoff used (Schaeffer et al 1999).

However, despite the limitations in the use of screening questionnaires, their simplicity and ease of application to a large number of subjects make them attractive. Also, when properly applied and validated, and when appropriate cut-off is chosen, questionnaires can have good agreement with diagnoses made with clinical interview in SCDs (Grant et al 2000). The vast majority of studies discussed in this chapter used questionnaires to assess depression.

The second difficulty with diagnosing depression in SCDs is the transitory association between depressive symptoms and acute episodic complications of SCDs such as painful ischaemic crises. Although the symptoms may be very distressing and impairing, the transitory nature of the experience may not meet the duration criteria for a depressive disorder. For example, in a qualitative study of adolescents with SCDs and Thalassaemia, Atkin and Ahmad (2001) found that although most young people go through periods of feeling low and despondent, such periods were generally transitory and often triggered by life circumstances. They found no evidence of the sort of sustained withdrawal from family and peer relationships that characterise established depressive disorder. Following cessation of the stressor such as a hospital admission, the young people were able to successfully rebuild their coping strategies and sense of normalcy.
The transitory nature of depressive symptoms in SCDs raises another concern that genuine depressive disorder may not be accorded the appropriate significance it deserves in SCDs. The risk is that both clinicians and patients could see intermittent depressive symptoms as part of the “normal course” of having serious life-threatening disorders like SCDs. As a result, patients may not seek help even when their depressive symptoms are sustained as in genuine depressive disorder. Similarly, clinicians may be reluctant to enquire or to pursue a fuller enquiry when patients volunteer history suggestive of a depressive disorder. Thus for some people with SCDs, unrecognised and untreated depressive disorder could impair their quality of life over and above the direct physical complications of SCDs. Again using epilepsy as a model, it has been shown that for people with refractory seizures, depression was a more important variable in their quality of life than the seizure frequency or severity (Kanner 2003).

3.3.3. Why people with SCDs could be at increased risk of depressive disorder.
Using the bio-psycho-social model of adjustment to chronic diseases, depression in SCD could result from the disease process (e.g. severity, specific complications, and the unpredictable nature of some complications), psychological aspects of the individual (e.g. lack of acceptance or negative attitude to SCDs) and social factors (e.g. lack of social support, the need to make frequent adjustments in life-style, and negative attitude of others). These are explored in more details next.

3.3.3.1. Pain and depression in SCDs
Painful crisis is the hallmark physical complication in SCDs (Wethers 2000). Bearing this in mind, there is evidence from other disorders indicating that chronic pain is an independent risk factor for depression (Wolfe and Michaud 2009). A link between pain and depression is also supported by evidence that treatment for one improves the other (Kanai and Okamoto 2007). Evidence from studies of pain and mood in SCDs supports these conclusions.

In a 6 months prospective study of 308 adults with SCDs, Levenson and colleagues found that 28% of the subjects were depressed. Compared with the non-depressed subjects, the depressed respondents had higher mean pain rating on more days and more distress and interference from pain (Levenson et al 2008). However, it is
important to note that although this study was prospective, it does not prove a causative link between pain and depression in SCD. Hasan and colleagues found in their study of 50 adults with SCDs, that 44% were depressed and the depressed subjects were more likely to have poor pain control and frequent ischaemic crises (Hasan et al 2003). Another study of 440 adults with SCDs found that patients who reported more frequent painful episodes were more likely to report depressive symptoms (Schaeffer et al 1999). Although this study had an impressively large sample size, it was limited by the fact that depression was not confirmed with structured clinical interviews. A study of the relationship between mood, pain and sleep in 20 children with SCDs found that mood mediates the relationship between pain and poor sleep (Valrie et al 2008). A general limitation of all the studies reviewed is the absence of firm evidence of causal association between pain and depression in SCDs.

In summary, evidence from SCDs suggest that as is the case with other chronic painful conditions, pain as a specific physical complication in SCDs could have a depressogenic effect although its causal role is unclear.

3.3.3.2. Depression and other physical markers of severity
The evidence is generally suggestive that people with more severe forms of SCDs are at more risk of depression. Segbena and Sangare (1994) used the Hamilton Depression Rating Scale to assess 30 adult patients with SCDs and 31 heterozygous carriers of the sickle gene. Although no subject in either group scored above the threshold for moderate depression, the level of anaemia and the number of sickle-cell crises per year was associated with depressive symptoms in the SCDs group. The study by Hasan and others cited earlier also showed that patients who made more frequent use of accident and emergency department and had more frequent blood transfusions (both surrogate markers of severity) were more likely to be depressed (Hasan et al 2003). However, it is worth noting that disease severity alone is not a sole determinant of mood or function in SCDs. For example, Grant et al (2000) investigated depression in 44 patients with SCDs using Structured Clinical Interview and found that disease severity alone did not explain the level of patient’s mood or level of impairment.
3.3.3.3. Depression and psychosocial factors

Carpentier et al (2009) have shown that personal psychological characteristics such as behavioral inhibition are associated with depression in SCDs. They examined behavioral inhibition and depression among 30 adolescents with SCDs and found that the adolescents who rated themselves high on behavioral inhibition displayed higher levels of depression than those with low behavioral inhibition. The study by Hasan and others cited earlier also found that adult patients with SCDs were more likely to be depressed if they had low family income (<$ 10,000), less than high school education, were female, or had inadequate social support (Hasan et al 2003). Similarly, Schaeffer and colleagues found in their study of 440 adults with SCDs that female gender and low family income were positively and significantly associated with depressive symptoms (Schaeffer et al 1999). This is an important study as it had one of the largest samples of people with SCD.

3.3.3.4. Depression and neurological complications

There is ample evidence that cerebral ischaemia increases the risk of depression (Hackett et al 2008). The risk of cerebral ischemia is now well recognised in SCDs and it is estimated that up to 11% of affected persons suffer overt strokes and up to 20% have evidence of ischaemic brain damage on MRI by the age of 20 years (Pegelow et al 2002). The impact of brain infarction on intellectual decline in children with SCDs is also well demonstrated (Schatz et al 2002). Given the high prevalence of ischaemic brain pathology in SCDs and the association between brain ischaemia and depression, it is reasonable to hypothesise increased rates of depression in people with SCD who have brain ischemia. However, I am not aware of any studies that have examined for depression in children with SCDs and brain infarction.

3.3.4. Paradoxical depression

Paradoxical depression is known to occur in epilepsy in response to a phenomenon called “forced normalisation”. This phenomenon refers to patients with epilepsy who develop psychiatric disorders when their seizures cease (Robertson 1998). While I am not aware of any studies of depression in SCDs following cure with successful bone marrow transplantation, this phenomenon of “forced normalisation” needs to be borne in mind. A colleague recently worked with a patient with SCDs who became depressed and suicidal after returning to her place of origin in Africa following
successful bone marrow transplantation in the UK. She described feeling guilty for being cured while many of her peers in Africa were still suffering from complications of SCDs with no hope of a cure or effective treatment.

3.3.5. Could any relationship between SCDs and depression be bidirectional?
While there are risk factors for depression in SCDs as outlined above, the available evidence is limited and mostly drawn from cross-sectional data. Bearing this in mind, it is possible to hypothesise an opposite direction of association whereby depression increases the risk of complications in SCDs. It is conceivable that depression in SCDs could lead to poorer treatment adherence; hence increasing the likelihood of more physical complications. For example, in a study of 46 adults with SCDs, Belgrave and Molock (1991) found that depression was associated with increased likelihood of emergency treatment and hospital admissions. However as this study was also cross-sectional, the opposite hypothesis could be true. Longitudinal study designs would be the most appropriate method to resolve the clarity about direction of association although structural equation modeling techniques could be helpful when applied to cross-sectional data.

3.3.6. Prevalence of depression in children with SCDs
This section aims to quantify the burden of depression in children with SCDs. As previously indicated (see 3.1.1), studies were included if the respondents were children, had a specific quantitative or categorical measure for depression that is compared with a controlled group or an established norm. The discussion was not separated into developing and developed countries because only one study from a developing country met the above inclusion criteria (Bakare et al 2008).

Benton et al (2007) and Kelch-Oliver et al (2007) have recently conducted reviews of depression in SCDs. Both reviews also included studies of other emotional disorders such as anxiety and other psychosocial difficulties. The reviews both noted that the data on the psychological aspects of children with SCDs are limited, of poor quality and conflicting. My examination of the literature on depression in this patient group came to similar findings.
3.3.6.1. Studies showing increased depressive symptoms in SCD

Three studies (Key et al 2001; Brown et al 1993; Morgan and Jackson 1986) found higher levels of depression in children with SCDs compared with control groups.

Key et al (2001) compared 125 adolescents (13-18 years) with chronic illnesses including SCDs, Cystic Fibrosis, Insulin Dependent Diabetes Mellitus, Spina Bifida, and Asthma with 21 healthy controls and the normative population on self-reported symptoms of depression. They found that in general, a higher proportion of the adolescents with chronic illnesses reported symptoms of moderate to severe depression compared with both the control group and normative data. However, among the group with chronic illnesses, the adolescents with SCDs and those with Asthma had the highest frequency of caseness for depression symptoms. The severity of depression among the children with chronic illnesses correlated positively with their own rating of the severity of their physical illness. However, this study was limited by reliance on questionnaire to determine depression caseness.

Brown et al (1993) compared 61 young people with SCDs (6-17 yrs) with their healthy siblings as controls (6-26 yrs). They found that the group with SCDs had more depressive symptoms. As in the previous study by Key et al (2001), severity of depression correlated with severity of SCDs. Another study by Morgan and Jackson (1986) in which 24 adolescents with SCDs were compared with the same number of controls matched for age, race, gender and socio-economic group found more depressive symptoms in the SCDs group. This study assessed depression with the Children’s Depression Inventory, which has four questions on somatic symptoms of depression (e.g. fatigue) which overlap with physical symptoms of SCD. Thus the authors checked if the excess depressive symptoms in the SCD group could be attributed to their scores on these four somatic items. Incidentally, a secondary analysis in which the four somatic items were removed found that the SCD group still had statistically significant more depressive symptoms than controls albeit with a smaller p value (Morgan and Jackson 1986).
3.3.6.2. Studies showing no increase in depression in SCD

The other seven studies that met the inclusion criteria did not find higher levels of depression in children with SCDs. One of the studies was conducted in a developing country (Bakare et al 2008) and the remaining six (Simon et al 2009, Lee et al 1997, and Yang et al 1994, Getzoff 2005, McElligott 2006, Midence et al 1996) in developed countries.

In a study from Nigeria, Bakare et al (2008) compared the rate of emotional disorders in 45 children with SCDs with equal numbers with Juvenile Diabetes Mellitus and healthy controls all aged 9-17 years. The Diagnostic Interview Schedule (DISC) was used to generate DSM-IV diagnoses. They found low levels of Major Depressive disorder in all three groups (2.2%, 6.7% and 2.2% respectively). However, they also found that 20% of the children with SCDs and 11% of those with Juvenile Diabetes Mellitus but none of the healthy controls expressed suicidal ideation in the past year. The low level of Major Depressive Disorder in this SCDs group in a developing country is a significant finding. However, the disproportionately higher levels of suicidal ideation among the children with SCDs suggest that this group may have had higher levels of depression but the assessment process was not sensitive enough to pick this up. In support of this possibility, the study also found that when all emotional disorders were pooled, children with SCDs and Juvenile Diabetic Mellitus both had significantly higher levels than controls (38%, 42% and 11% respectively).

The main strength of this study is the use of structured diagnostic interview rather than questionnaire to assess for depression. A second strength is the use of a homogenous sample of children with HbSS genotype. This strategy limited the variations associated with disease severity that arise from different genotypes of SCDs.

Simon et al (2009) compared 44 American adolescents with SCDs with 15 healthy siblings and found that depression scores for the SCDs group did not differ significantly from their siblings and were not in the clinical range of normative data. Similarly, Lee et al (1997) compared American young people with SCDs with healthy siblings. Surprisingly and contrary to expectation, these researchers found higher depression scores among the non-diseased siblings than in the SCDs group. These studies both had relatively small sample sizes. So the absence of a difference in the
study by Simon and colleagues (Simon et al 2009) could be due to Type II error. However, Type II error would not explain the findings of Lee and colleagues (Lee et al 1997) which found less depressive symptoms in the SCD group.

Yang et al (1994) compared 38 children with SCDs (aged 6-18 years) with 34 age, gender and race-matched healthy controls. Depression was measured with both a self report questionnaire (Children’s Depression Inventory) and Clinical Psychiatric interview. Yang and colleagues found a significant difference in the groups’ scores on the Children's Depression Rating Scale (27.1 and 22.1 respectively P = 0.007). Twenty-nine percent of the children with SCDs and 12% of the controls had scores suggesting a high risk for depression. However, clinical assessment by child psychiatrists found 13% of the SCDs group and 15% of controls to have a depressive disorder. Further analysis showed that items in the Children's Depression Rating Scale such as excessive fatigue and physical complaints contributed to the high false-positive rate. An important strength of this study is that DSM-IV diagnosis of depression was reached through two independent psychiatric interviews and agreement by consensus.

A longitudinal study, which followed up 48 children with SCDs in USA for up to 10 years and compared them with 49 healthy controls found no differences in depression (measured with Children’s Depression Inventory and Beck Depression Inventory) either cross-sectionally or longitudinally (Getzoff 2005). Another study of American adolescents with SCDs (McElligott 2006) found normative scores in depression. An earlier study in the UK involving 39 children with SCD and 24 controls (Midence et al 1996) found no significant differences between the two groups on depression. However, all the foregoing studies were limited by small samples sizes which raise concerns that their negative findings might be due to Type II error.

3.3.7. Summary of depression in children with SCDs
The evidence for depression in children with SCDs is limited. For example, of the ten studies reviewed, seven (including the only study from sub-Saharan Africa) did not show increased prevalence of depression in children with SCD. Helps and colleagues have suggested that the psychosocial adjustment of people with SCD has improved probably as a result of vastly improved physical care in SCDs (Helps et al 2003) in
this region. The finding of no increased depression in SCD could also be due to increasing levels of psychological resilience and hardiness among children with SCDs (Mckellop 2001). Another reason is the possibility that the evidence is limited because of methodological problems including the difficulties described earlier in relation to diagnosis of depression in children with SCDs. Another difficulty is the heterogeneous nature of SCDs. For example, children the HbSS genotype are generally more severely affected compared with those with HbSC. However, few studies (e.g. Bakare et al 2008) used homogenous samples of HbSS while others had a mixture of children with both genotypes, which makes interpreting their findings difficult.

3.3.8. Conclusion
There are theoretical models based on the biology and psycho-social contexts of children with SCDs that suggest increased risk of general psychopathology and depression. However, the research evidence is limited and conflicting. There are several putative reasons for this (Molock and Belgrave 1994). Some symptoms of depression and SCDs overlap to make diagnosis of the former difficult. Many studies were underpowered, and included children with heterogeneous genotypes of SCDs. For example, children with HbSS genotype are generally more severely affected compared with those with HbSC. Some studies had a mixture of children with both genotypes, which would make interpreting their findings difficult. The episodic nature of complications in SCDs probably makes associated psychological distress transitory, which increases the difficulty in recognising discreet depressive episodes. Although the evidence is limited, it appears that for children with SCDs in developed countries, the historical trend is towards less psychopathology. The situation in developing countries remains difficult with most studies indicating increased psychopathology. This distinctive regional difference appears to do with access to improved physical care in developed countries.

While acknowledging that the current evidence does not support increased prevalence of depression in children with SCD especially in developed countries, there could be sub-groups of children at risk. Identifying possible characteristics of such sub-groups could be helpful in targeting screening and interventions. This study aims to contribute to the identification of such predictive factors.
Chapter 4.
Aims and Methodology

This chapter is discussed under the following subheadings.

- Reasons for study
- Aims of study
- Hypotheses
- Study design
- Sample size
- Recruitment
- Response rate
- Measurements and reliability
- Analytical strategy
- Difficulties with recruitment

4.1. Reasons for study

Chapter 2 of this Thesis argued that stigma theory could be applicable to SCDs. Evidence from studies of children with other chronic physical conditions such as epilepsy suggests high prevalence of perceived stigma and a contribution of stigma to psychological distress in affected children (Westbrook et al 1992). Given the similarity between SCDs and epilepsy in terms of chronic disease model, a similar association between stigma and psychological distress can be hypothesised for SCDs. However, unlike in epilepsy where there is empirical support, there is no good data in SCDs to test this hypothesis. Thus the main aim of this study is to quantify the level of perceived stigma in children with SCDs and to explore any links between self-perceived stigma and psychological distress.

Chapter 3 of this Thesis found inconsistent evidence for increased levels of psychopathology in children with SCDs in developed countries with a tendency for less psychopathology in more recent studies. Given the inconsistency of the evidence, it is possible that there are some children with SCDs in developed countries who could be at increased risk of psychopathology. Bearing this in mind, there is a need to
continue to identify new and more precise predictors of psychological distress in children with SCDs. Such predictors could be used to identify high risk sub-groups or become incorporated into screening questions and algorithms to ease the identification and treatment of children with SCDs who have comorbid mental disorders. This study aims to explore self-perceived stigma and other factors as new potential predictors of psychopathology in children with SCDs. Although results of psychological interventions in SCD are encouraging (Anie 2005), the evidence base is still limited (Anie and Green 2002). Thus identification of more specific predictors of psychological distress in children with SCD could lead to new and more effective targeting of psychological interventions.

Most research on stigma has often focused on enacted stigma by people who do not have the stigmatising condition. This is usually assessed with measures of social distance or behavioural rejection towards people affected by the stigmatising condition. These studies have been criticised for not focusing on the experience of people with the stigmatising condition and for potentially reinforcing stereotypes by privileging the perspective of people without the stigmatising condition over those bearing the burden of the condition (Sankar et al 2006). In order to address this imbalance in stigma research, this study is focused on stigma as perceived by children with SCDs themselves.

4.2. Aims of the study

1. To estimate the prevalence of self-perceived stigma in young people with SCD.
2. To explore associations between self-perceived stigma and psychosocial, socio-demographic, and SCD-related variables.

4.3. Study hypotheses

The study hypotheses to be tested are:

1. Measures of disruptiveness (e.g. frequency of admissions) and visibility (e.g. presence of leg ulcer) will significantly and independently predict levels of self perceived stigma.
2. Self perceived stigma will significantly and independently predict scores on the Total difficulties scale of the Strengths and Difficulties Questionnaire (SDQ)
3. Self perceived stigma will significantly and independently predict Depressive symptoms measured by the short Mood and Feelings Questionnaire (SMFQ)
4. Self perceived stigma will significantly and independently predict Self Esteem measured by Rosenberg Scale

The process of exploring the above primary hypotheses led to a secondary objective, which was to explore additional associations between each dependent variable and other psychological, illness, and social predictor variables over and above self-perceived stigma.

4.4. Study design
The study design is a cross sectional questionnaire survey of young people aged 11-19 years with SCDs. A cross-sectional survey design was chosen because it is appropriate for achieving the primary objective of this study, which is to estimate the prevalence of self-perceived stigma among children with SCDs. The limitation of the design in exploring associations between stigma and other psychosocial variables is recognised in so far as a cross-sectional design would not be able to identify causal relationships. A longitudinal study design was considered as it would both achieve the main objective of the study and facilitate causal inferences by identifying the temporal direction of association between stigma and other variables. However, the expense and huge logistics that would be engendered by longitudinal design made it impractical for this study.

4.5. Sample size / power calculations
Two sample size calculations were carried out - the first based on the main objective and the second based on one of the secondary objectives.

4.5.1. Sample size required to estimate the prevalence of self-perceived stigma
EPI-INFO Statistical package Version 6, was used to estimate the sample size required to identify a prevalence of 40% for self-perceived stigma in SCD with 95% confidence and 10% margin of error. The estimated sample was 92 children with
SCDs. The expected prevalence of self-perceived stigma of 40% was based on two previous studies of epilepsy (Westbrook et al 1992) and stuttering (Blood et al 2003) which used similar methodology to assess self-perceived stigma as in this study.

**4.5.2. Sample size required to identify significant difference in mean scores on the SDQ**

The minimum sample size to detect a significant difference in psychological difficulty (Total difficulties score on the SDQ) between participants who are classified as having self-perceived stigma AND those not so classified is given by the Formula below.

\[ n \geq \frac{2F\sigma^2}{d^2} \]  

(Wade 1997)

Where:
- \( n \) = size of each of the two groups (i.e. total sample = 2n if the groups are equal)
- \( \sigma \) = standard deviation
- \( d \) = smallest difference expected between the groups.

With respect to the Total Difficulties Score on the self-report version of the SDQ, \( \sigma = 5.2 \) for UK norm [http://www.sdqinfo.com/bba1.pdf](http://www.sdqinfo.com/bba1.pdf) (accessed 5/2/10)

Assuming \( d = 2.6 \) (i.e. half a standard deviation difference between the two groups1), \( F = 7.85 \) (a constant based on 80% power and 5% level of significance) (Wade 1997)

The minimum sample size required to identify 0.5 (half) standard deviation difference in Total Difficulties Score on the SDQ with 80% power and 5% level of significance is:

\[ n \geq \frac{2 \times 7.85 \times 5.2^2}{2.6^2} = 62 \] (i.e. a total of 133 taking into account the hypothesised group ratio of 40:60).

Assuming a 50% response rate2, the target sample to be approached was doubled to 266 with the aim of recruiting 133.

The final sample of 93 recruited for the study was about the same as the first sample size of 92, which was calculated (above) as required to identify a 40% prevalence of stigma with 95% confidence level and an error margin of 10%. Incidentally, given

1 A difference of half a standard deviation between the two groups is possible given that Iloeje (1991) found differences of up to one standard deviation between children with SCD and healthy controls on the Rutter Scale (forerunner of the SDQ) albeit in Nigeria. Also, among a UK community sample (comparing child mental health clinic attendees and a general population sample) (Goodman et al 2003), the two groups differed on the SDQ by as much as 1.4 standard deviations.

2 A recent postal survey of people with rheumatoid arthritis (another chronic medical condition) in the UK reported a response rate of 57.3% (Neame and Hammond 2005).
that the actual prevalence of self-perceived stigma in the study was 15% (compared with the hypothesised level of 40%), a sample as low as 52 would have been sufficient to identify this prevalence (15%) with 95% confidence and 10% error margin.

A sample of 133 was calculated as required to identify 0.5 or more standard deviation difference in measure of psychological difficulty (Total Difficulties Score on the SDQ) between the respondents who perceived stigma and those who did not. As explained above, this sample size was based on 80% power and 5% level of significance with an assumption of 40% prevalence of stigma. However, the final sample size of 93 proved sufficient to identify significant differences in SDQ Total Difficulties Score (see Table 5.34) between the respondents who perceived stigma and those who did not. Thus, this smaller sample size was still adequately powered. The reason for this is because the actual difference between the respondents who perceived stigma and those who did not was twice higher than expected (more than 1.0 standard deviation compared with 0.5 standard deviation used for the sample size calculations).

In summary, although the final study sample size of 93 was less than planned (133), the study was still sufficiently powered to answer the main research questions.

4.6. Inclusion criteria:

1. Children and young people with all forms of SCD (HbSS, HbSC, HbSThal). Despite the recognised difficulty with including heterogeneous genotypes in psychological research on SCDs, I was advised that this study was unlikely to be able to recruit the desired sample size if inclusion was limited to children with homozygous SCDs (HbSS). Even with including all types of SCDs, it was still not possible to recruit the target sample size of 133.

2. Aged 11-18 years. We targeted this age group because the development of self-identity and peer relationships is important during this period; hence self-perceived stigma could have serious impact on psychosocial function and self esteem if it is experienced in this age group. One respondent who was 18 years old at the time of collecting the research pack turned 19 years by the time of
completing the questionnaire. Also one ten year old completed the questionnaire; hence the final age range was 10-19 years.

3. Consent by child and parent (for ages 10 - 15 years) and by young person (for 16 years and above).

4. Not currently acutely unwell (e.g. not admitted to the ward due to acute illness)

5. Adequate command of English language to complete the questionnaires.

4.7. Recruitment procedure

Subjects were recruited through the Sickle Cell Society and from three paediatric haematology clinics in London (Central Middlesex, North Middlesex, and St Mary’s Hospitals).

Postal questionnaires were sent to families who were members of the Sickle Cell Society who had an affected child within the age range of the study. A pack was sent to the family containing an invitation letter, separate information sheets for children and parents, consent forms for parents and assent forms for children under 16 years and consent forms for young people 16 years or older, the study questionnaire, telephone slip and a prepaid-addressed-envelope. Having consented, the child was invited to complete and return the questionnaire in the prepaid reply envelope enclosed in the pack. Children were also given the choice of having the questionnaire completed on their behalf by me through a telephone interview. However, no child took up this offer. The full content of the research pack are attached to this Thesis as Appendix 1.

The information sheet for children encouraged them to discuss the research with their families. Also the information sheet for parents/guardians contained the sentence “we expect that your child will complete the questionnaire himself/herself although he/she could ask you for help in remembering factual information to help them answer the questions”. It is therefore likely that some of the children may have had help from parents in completing the questionnaire. However, the data collection made no provision to identify if a parent helped or not.
The postal questionnaire of participants recruited through the Sickle Cell Society met with considerable difficulties. Two hundred questionnaires were mailed out as described earlier. However, it subsequently emerged that the membership database of the Sickle Cell Society was not up to date and the filtering mechanism was not accurate. As a result, some of the questionnaires were mailed to members who did not have SCDs or who had SCDs but were outside the study age criterion. Membership of the Sickle Cell Society is open to everyone including people without SCDs. A total of 25 young people responded to the postal questionnaires mailed out by the Sickle Cell Society. However, due to the technical difficulties described above, it was not possible to ascertain an accurate response rate from this source.

Recruitment from the three haematology clinics was carried out by an assistant psychologist or a research nurse who were trained and supervised by me. Families were approached as they attended routine out-patient appointments. The families were given an explanation about the research and those who showed interest were given the research pack, which had the same content as the postal questionnaire (i.e. invitation letter, separate information sheets for children and parents, consent form for parents and children 16 years and older, assent form for children younger than 16 years, study questionnaire, prepaid and addressed return envelope, and telephone slip). The family were informed that the children could consent and complete the questionnaire in the clinic or take the pack home to complete the consent forms and questionnaire at their convenience and return them in a prepaid reply envelope included in every pack. Most children opted for the latter. Families who decided to participate in the study during their clinic attendance completed the consent and assent forms and the child completed the questionnaire in the clinic waiting room either before or immediately after seeing the clinician. The assistant psychologist and research nurse were available to clarify any questions, but their assistance was rarely sought. The questionnaire pack was also posted to patients who were not due a clinic appointment soon. Each respondent was given £10 worth of shopping vouchers for completing the questionnaire. The vouchers were given in person to the children who completed the questionnaire in clinic and posted to others on receipt of their questionnaire. The number of children recruited from each clinic is outlined below (Table 4.1).
Table 4.1. Number of respondents recruited from each site

<table>
<thead>
<tr>
<th>Centre</th>
<th>Number of subjects recruited</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central Middlesex Hospital Park Royal/Brent</td>
<td>36</td>
</tr>
<tr>
<td>Sickle Cell and Thalassaemia</td>
<td></td>
</tr>
<tr>
<td>North Middlesex Hospital Edmonton</td>
<td>19</td>
</tr>
<tr>
<td>St Mary’s Hospital Paddington</td>
<td>13</td>
</tr>
<tr>
<td>Sickle Cell Society</td>
<td>25</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>93</strong></td>
</tr>
</tbody>
</table>

Chapter 5 will explore differences between respondents recruited from the Sickle Cell Society compared with those recruited from the three haematology clinics.

### 4.8. Reliability

For quality control, the medical records of 10 randomly selected participants at Central Middlesex Hospital were reviewed to check the accuracy of their responses for age, presence of leg ulcers, and prescription for hydroxyurea. These three questions were chosen because they represent “hard data” such that any discrepancy between the questionnaire and the medical records is not likely to be attributable to natural variability in the illness. This reliability check found 100% concordance between the questionnaire responses and medical records.

Also in Central Middlesex Hospital, basic demographics and sickle cell variables of 10 randomly selected respondents were compared with 10 randomly selected non-respondents. The comparison is shown below.

Table 4.2. Comparison of responders and non-responders

<table>
<thead>
<tr>
<th>Variable</th>
<th>Respondents</th>
<th>Non respondents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (mean)</td>
<td>15.0</td>
<td>15.1</td>
</tr>
<tr>
<td>Gender (M:F)</td>
<td>6:4</td>
<td>5:5</td>
</tr>
<tr>
<td>Admissions in past year (mean)</td>
<td>2.0</td>
<td>0.7</td>
</tr>
<tr>
<td>Leg ulcer</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Hydroxyurea</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>
As shown in Table 4.2, the respondents had three times more admissions in the previous year compared with non-respondents. This suggests the respondents may have been more unwell. Also there were more males than females in this sample of respondents (6:4). However, note that the gender proportion in the full sample is almost equal (M:F = 51:49% – see Table 5.2).

Central Middlesex Hospital was also the site with records that could be examined to ascertain response rate. Five children who were registered with the haematology clinic at Central Middlesex Hospital had responded to the postal questionnaire through the Sickle Cell Society. If these five children are added to the 36 children who were recruited directly from the Central Middlesex Hospital, then 45% of the eligible young people in that site participated in the study.

I planned to administer the questionnaire twice (two weeks apart) to 20 participants to assess the Test-Retest reliability of the component measures. However, due to the huge difficulty we had with recruitment, no respondent agreed to participate twice. A second £10 shopping voucher was offered for anyone completing the questionnaire again but this was not taken up.

My general experience with this client group suggests that they are over-researched. Many young people and their families commented that they are always being asked to take part in a research project. While many did not mind, others found this burdensome. This accounted for the major challenge I had with recruitment. Even the offer of £10 shopping voucher was not sufficient motivation for some children and their families.

The information sheet for the study indicated that families can register their refusal to participate in the study by returning the uncompleted questionnaire in the prepaid reply envelope enclosed in each research pack. Only 4 families registered their refusal with this method. Two of the families did not give a reason for their refusal. One family commented that the questionnaire was too long. Another stated that the questions were too negative and could undo the work the family had done to improve the self-confidence of their child. As a result of the latter comment and additional
feedback by a sickle cell counsellor regarding the wording of the “Study Title” in the Information Sheets and other documents in the research pack, the “Study Title” was changed from:

“Research project to examine whether young people with sickle cell disease feel shame and embarrassment as a result of their health problems”

to

“Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems”

The amendment was approved by the Ethics Committee (See Appendix II).

The study and all subsequent amendments were approved by the South West Multicentre Research Ethics Committee. Approvals were also obtained from the Research and Development Departments of the three clinical sites where children were recruited from (Appendix II).

4.9. Measurements and data collection

A questionnaire was constructed by assembling several standardised and validated instruments measuring the variables essential to achieve the objectives of the study. The questionnaire was designed to obtain information from the young people themselves. A copy of the questionnaire is attached as part of Appendix 1. The specific measures in the questionnaires and their validity and reliability are discussed below. The introduction to the questionnaire reassured the young people of the confidentiality of their responses. They were encouraged to answer truthfully and that there were no right or wrong answers. Data collection started in October 2006 and ended in April 2009. Before the study commenced, the questionnaire was reviewed and considered appropriate by clinicians working with children with SCDs.

4.9.1. Questionnaire Reliability

As stated above, ten of the respondents whose answers were checked against their medical records showed 100% concordance indicating good reliability. The instruments included in the questionnaire all showed good internal consistency (Cronbach’s Alpha) (Cronbach, 1960). These are reported below for each instrument. Another evidence to support reliability of the responses is that the variables correlated in a sensible manner.
4.9.2. Socio-demographic details

Standard background socio-demographic information was obtained. Gender was coded as 1=Male, and 2=Female. The occupation of the head of household was used to assign an OPCS socio-economic class. Given that multiple indicators for socioeconomic assessment have been used reliably among ethnic minorities in London (Stansfeld et al 2004), I gathered additional information on ownership of a land telephone, and a car. However, this data was not sufficiently discriminatory in analysis probably due to ceiling effect as most respondents came from families with high ownership of these items.

4.9.3. Self-perceived stigma

This was assessed with items adapted from a previous study of stigma in young people with epilepsy (Westbrook et al 1992). This study, which was based on stigma theory, presented a methodology for studying stigma in adolescents with chronic conditions. A subsequent study in adolescents who stutter demonstrated the adaptability of the methodology and confirmed the original factor structure (Blood et al 2003). The adapted questions are shown Table 4.3 below.

Respondents were categorised as having high self perceived stigma if they answered “Often” or “Sometimes” to one or more of the three “stigma questions” shown below (Table 4.3). Four additional questions on “disclosure of stigma” were used to measure avoidance behaviour, which is an indirect index of self-perceived stigma (Westbrook et al 1992, Blood et al 2003).

To maximise the analytical power of the measures of stigma, a dimensional stigma variable was created from the sum of the stigma questions. The resulting “stigma rating scale” had a good internal consistency (Cronbach Alpha 0.8). As predicted, Factor Analysis of the stigma and disclosure questions in the present study extracted two factors, which provides further support for the reliability of the questions. Higher scores on the stigma rating scale indicates more self-perceived stigma.
Table 4.3. Stigma and disclosure questions

<table>
<thead>
<tr>
<th>Stigma Questions</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Do you think that having Sickle Cell affects whether people want to be friends with you?</td>
</tr>
<tr>
<td>2. Do you think that having Sickle Cell affects whether people like you or not?</td>
</tr>
<tr>
<td>3. Do you think that having Sickle Cell affects whether or not you are invited to people’s homes or to parties?</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Disclosure Questions</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. When you can, do you keep your sickle cell a secret from others?</td>
</tr>
<tr>
<td>2. How frequently do you talk to people about your sickle cell?</td>
</tr>
<tr>
<td>3. Do any of your friends know that you have sickle cell?</td>
</tr>
<tr>
<td>4. When people find out you have sickle cell, it is usually because</td>
</tr>
<tr>
<td>a. You tell them</td>
</tr>
<tr>
<td>b. They see you have a sign of sickle cell and then you explain</td>
</tr>
<tr>
<td>c. Someone else tells them about it</td>
</tr>
</tbody>
</table>

4.9.4. Emotional and behaviour problems (psychological difficulty)

The self-report form of the Strengths and Difficulties Questionnaire (SDQ) was used to measure psychological difficulty. This is a well validated 25-item screening instrument for childhood psychopathology used widely in routine clinical practice and research in the UK and internationally (Goodman et al 2003). The SDQ produces a Total Difficulties Scale in addition to five other subscales (Emotional, Hyperactivity, Conduct, Peer problems, and Prosocial). The questionnaire has versions for parents, teachers and self-report by young people. The self-report version was standardised on young people aged 11-16 years (Goodman et al 1998), however, for pragmatic reasons, we extended its use to 18 years in this study. SDQ was used in a large national survey of child and adolescent mental health in the UK (Meltzer et al 2000), which included 4228 self reports. The UK norms from the Self report SDQ are given below (http://www.sdqinfo.com/bba1.pdf) (accessed 5/2/10). Except for the prosocial scale, higher scores in the other SDQ subscales indicate more difficulties.
Total Difficulties scale = 10.3 (SD 5.2). Borderline = 16-19, Abnormal ≥ 20
Emotional subscale = 2.8 (SD 2.1). Borderline = 6, Abnormal ≥ 7
Conduct subscale = 2.2 (SD 1.7). Borderline = 4, Abnormal ≥ 5
Hyperactivity subscale = 3.8 (SD 2.2). Borderline = 6, Abnormal ≥ 7
Peer problems subscale = 1.5 (SD 1.4). Borderline 4-5, Abnormal ≥ 6
Prosocial subscale = 8.0 (SD 1.7). Borderline = 5, Abnormal ≤ 4

In the standardisation study for the self report version (Goodman et al 1998), 5% of the community sample and 31% of a clinic sample (children attending a mental health service) scored within the abnormal range (≥ 20) on the Total Difficulties Scale, 18% and 28% respectively scored within the borderline range while 77% and 41% scored within the normal range respectively. In the national survey of child and adolescent mental health in the UK (Meltzer et al 2000), 5.3% of the total sample scored above ≥ 20 on the self report version of the SDQ.

Although a cut-off score of 20 or more on the Total Difficulties Scale is recommended for the self-report version of the SDQ to define caseness in clinical practice http://www.sdqinfo.com/bba1.pdf (accessed 5/2/10) (Goodman et al 1998), in this study, a cut-off of 18 and above was used for the self report SDQ to define caseness. This is because this cut-off gave prevalence figures in the British Child and Adolescent Mental Health Survey (Meltzer et al 2000) that were equivalent to the prevalence found using data from multiple sources including parents and teachers. Given that the same cut-off of 18 and above was used in a study of school children in East London (Stansfeld et al 2004), adopting this cut-off means that my data could be compared not only with UK-wide prevalence but also with a more ethnically appropriate sample. In fact Stansfeld and colleagues (Stansfeld et al 2004) provided specific data on SDQ caseness for black boys and girls in East London, which in the context of SCD makes it the most appropriate data to be compared with my sample.

4.9.5. Depressive symptoms
Depressive symptoms were measured with the Short Mood and Feelings Questionnaire (SMFQ) (Angold et al 1995). This is a brief (13-item) self-report rating scale with good psychometrics. The child responds to each directly framed statement with “True”, “Sometimes”, or “Not True” (scored 2, 1, 0 – such that higher scores
indicate more depressive symptoms. The SMFQ was developed from the highly reliable 30-item Mood and Feelings Questionnaire (MFQ) by selecting the 13 items that performed well in a variety of psychometric analyses on the MFQ. The selected items were predominantly affective and cognitive, which are known to be the best predictors of depression. It also included physical symptoms like tiredness. The SMFQ correlated well with the Children’s Depression Inventory and discriminated well between children diagnosed with depression by clinical interview and those without diagnosis. The Internal Consistency of the SMFQ (0.85) reported by the developers is almost the same as in the present study (Cronbach Alpha 0.83). The questionnaire is usable with children from 6 years. The original study in USA found sensitivity of 60% and specificity of 85% with a cut off of 8. A community-based twin study in the UK (Thapar and McGuffin 1998) found that twins who met DSM-III-R diagnosis for depression had a mean score of 8.76 (SD = 4.19) compared with twins with no diagnosis of depression (mean = 4.46, SD = 5.24). Receiver-Operator-Curves showed that a cut-off score of 8.0 on the self rated Short MFQ achieved optimum sensitivity (0.75) and specificity (0.74) for DSM-III-R depression (Thapar and McGuffin 1998). Another UK study involving ethnic minorities in London found a mean of 4.4 for boys and 5.7 for girls (Stansfeld et al 2004). Using a cut-off of 8.0, this study classified 17.2% of Black boys and 29.5% of Black girls as depression-positive cases. These figures were also comparable to the proportion of cases among White boys and girls in the study (Stansfeld et al 2004).

4.9.6. Self esteem

Self-esteem was assessed with the Rosenberg Self-Esteem Scale (Rosenberg 1965). This is a reliable and widely used Self Esteem Scale. The scale is a summative 10-item Likert scale with items answered on a four point scale - from strongly agree to strongly disagree (scored 1-4). Higher scores indicate higher self esteem (range 10-40). The Internal Consistency in the present study was excellent (Cronbach Alpha 0.86). A cross national comparison among students in USA, Canada and New Zealand (Rusticus et al 2004) found the following Means (SD) U.S.A = 31.9 (4.97), Canada = 31.0 (4.82), New Zealand = 29.9 (4.52), which are comparable to the only study I found that used the same questionnaire on adolescents with SCD in America (Mean 31.9 (SD 5.0) (Burlew et al 2000).
4.9.7. **Family function**

Family function was measured with the 12-item General Functioning (GF) subscale of the Family Assessment Device (FAD) (Epstein et al, 1983). This is a self-report questionnaire designed to evaluate family functioning according to the McMaster Model of Family Functioning (Epstein et al, 1978). The FAD (Epstein et al, 1983) and the GF subscale (Byles et al 1988) have good psychometrics. This instrument was used in the Survey of Mental Health of Children and Young People in Britain 1999 (Meltzer et al 2000). It is a Likert scale score 1-4. However, the total score is divided by 12 to create a scale range of 1-4. On this scale, scores of 2 or less are considered “healthy” family functioning, while scores from 2.01 – 4.0 are considered “unhealthy” family functioning. The FAD performed reliably in the present study with Internal Consistency (Cronbach Alpha) of 0.86.

4.9.8. **Peer network**

Peer social network was measured by asking the young people to identify how many friends they have frequent mutual engagement with and whether they had a best friend (Fang et al 2003). A composite rating “peer network scale” was developed by combining the total number of friends and having a best friend. A weighting of three times was given to having a best friend.

4.9.9. **Sickle cell severity**

Although various measures and validated scales exist for measuring “illness severity”, it is well recognised that intrinsic markers of illness severity can be difficult to identify. Thus, so called “severity scales” are often designed to tap into indices of impairment (e.g. frequency of hospital admission), which are surrogates of illness severity rather than intrinsic measures of severity. One problem with using surrogate markers of severity is possible confounding by other factors such as treatment adherence or the impact of other unrelated diseases or environmental factors. Despite this limitation, surrogate measures of severity can be useful where intrinsic measures of severity do not exist or are difficult to obtain.

SCD is one of the few conditions for which there is an intrinsic biologically determinable measure of illness severity. It is well recognised that different genotypes
in SCD confer different severity profiles to affected persons. For example, the genotype HbSS, which results from replacement of glutamic acid by valine in position six of the Beta Globin chain confers a more severe illness than HbSC genotype, which results from substitution with lysine in the same position (Castro et al 1994, Peak 2008).

Despite the limitations noted above, surrogate measures of severity were adopted for this study. Consideration was given in the study design to obtaining the children’s genotype as an intrinsic measure of severity. However, it was felt that children may not be reliable informants for such information and that accessing medical records for all participants would be the reliable means of obtaining the data. Unfortunately, the resources available for the study would not have coped with the logistics of accessing medical records for all participants. Also the conditions of the study ethics approval allowed only limited access to ten medical records for purposes of checking reliability of responses. However, since the conclusion of the study, I have become aware that children with SCDs and their parents can be reliable informants about their genotype. The absence of genotype information is an important limitation of this study, and consideration is being given to seeking further ethics approval to obtain this data. However, the additional data if obtained would not form part of this Thesis.

Illness severity measures used in this study were adapted from surrogate measures used reliably in a previous study by Hurtig et al (1989) based on:

1. Frequency of ward admissions (defined as an overnight or longer stay in hospital).
2. Number of Accident and Emergency Department visits not resulting in admission.
3. Frequency, intensity, and duration of painful crises.
4. Frequency of school absence

Each of the above measures was presented as incremental ordinal scales and coded such that higher scores indicate more severity. A composite rating of illness severity was calculated by combining the above measures. However, as frequency of ward admissions proved most discriminatory than all the other measures of severity including the composite measure, the former was chosen as the measure of severity in subsequent analyses.
4.9.10. Visible signs of SCDs
The questionnaire prompted respondents to indicate if they currently had jaundice and or leg ulcer. Both variables were coded as 1=Yes/Present, 2=Not present.

4.9.11. Treatment Adherence
Medications commonly prescribed for people with SCD (penicillin V, hydroxyurea, and Folic Acid) were listed in the questionnaire and respondents asked to indicate which medication, if any, was prescribed for them and how often they remembered to take it. The number of subjects prescribed hydroxyurea was so small (N = 7) that this was not included in the adherence scale. The medication adherence scale was therefore developed by combining adherence to penicillin and folic acid. Higher score on this scale indicates better adherence. Subjects were also asked how often they kept out-patient appointments.

4.9.12. Attitude toward illness
This was measured with an adaptation of the Child Attitude Toward Illness Scale (CATIS) (Heimlich et al 2000, Austin and Huberty 1993). CATIS is a 13-item summative scale that measures children’s feelings towards having a chronic illness. The scale is designed for children from the age of 8 years old. A higher score indicates a more favourable attitude towards the illness. Initial psychometric assessment of the scale (Austin and Huberty 1993) with children suffering from epilepsy or asthma, and further evaluation (Heimlich et al 2000) with children suffering from epilepsy found excellent internal consistency, test-retest reliability, and construct validity. I found no studies with CATIS in children with SCDs. However, the scale performed very well in the current study with a Cronbach Alpha of 0.91.

4.9.13. Receipt of Counselling
Respondents were asked if they were currently engaged in a regular counselling with a therapist. Responses were coded as 1=Yes, 2=No.
4.10. Analysis and hypotheses testing
Data entry and analysis was conducted with SPSS Version 15. The data entry was independently checked for accuracy. Frequencies and charts were used to further identify inconsistencies, which were checked against the original questionnaire. Questionnaires used were tested for internal consistency using - Cronbach’s alpha. Interval and ordinal data which were normally distributed were summarised with means and standard deviations, while categorical data were described with Numbers and Percentages. Bi-variate comparisons to determine group differences were conducted with chi-square, Fishers exact, and t-tests. To test the study hypotheses, independent variables associated with the outcome variables for each hypothesis were identified and included as predictors in separate multiple regression models with self-perceived stigma, Total Difficulties Scale on the SDQ, depressive symptoms, and self esteem as the dependent variables. All confidence intervals presented are based on 95%.

All the variables were explored for missing data. On average, most variables had 9% missing data (N = 85). The variable with the most missing data (29%) was the “Severity rating scale” (N = 66). This is a composite scale which suffered because of cumulative effect of missing data in the constituent variables. Incidentally, this “severity scale” was not used in the multivariate analyses because frequency of ward admission proved to be a better index of severity and was used instead. The pattern of missing data appeared random, which means the main consequence is loss of sample size and “Power” in the analyses. As recommended by Pallant (2007), subsequent analyses used the SPSS Option for “Exclude Cases Pairwise” which excludes cases only if they are missing the data required for that particular analysis. This option has a less severe limiting effect on the sample size compared with other alternative strategies for handling missing data.

4.11. Difficulties encountered during data collection.
I encountered major difficulties with recruitment of subjects. Thus, I was only able to recruit 93 subjects instead of my target of 133 (i.e. 70% of target) although the study remained well powered due to larger-than predicted group differences. Secondly, the period of recruitment lasted almost 2½ years (instead of one year that was set aside for recruitment). The main difficulty was that people with SCDs in the UK appear to be an over-researched group. Many young people and their families commented that they are frequently being approached to take part in research projects, which they found
burdensome. Recruitment remained difficult despite having a research nurse on site at North Middlesex and St Mary’s Hospitals. Even the offer of £10 shopping voucher was not sufficient motivation for some children and their families.

Due to difficulties with the membership database of the Sickle Cell Society, the response to the postal questionnaires mailed by the organisation was not optimum. It also made it difficult to calculate an accurate response rate from the mail out. The difficulties included sending questionnaires to members without SCDs or who had the condition but were outside the study age range. The Sickle Cell Society is based in Brent and not far from Central Middlesex Hospital. Thus there was an overlap between the two catchments and some young people recruited in the hospital acknowledged receiving but not responding to the previous mail out by the Sickle Cell Society.

The accuracy of the patient database at North Middlesex Hospital was also problematic; hence it was not possible to accurately ascertain a response rate from that site. St Mary’s Hospital Paddington is a Tertiary referral centre that sees patients from a large catchment and sometimes for consultation rather than for ongoing care. As a result, it was not feasible or appropriate to calculate response rate from this site. Central Middlesex Hospital was therefore the only site with defined catchment and sufficiently accurate and accessible database to calculate a response rate (45%).
Chapter 5.
Analysis and results

This chapter describes the data collected for this thesis. The chapter is divided into four Sections:

- Section 1 provides a description of the variables in the study.
- Section II explores the bivariate relationship between stigma and other variables.
- Section III compares the respondents recruited from the Sickle Cell Society and those recruited from haematology clinics on variables such as self-perceived stigma and measures of psychosocial function.
- Section IV deals with multivariate analyses, which include the testing of the four specific hypotheses proposed in the study.

For each Section, the results are presented as tables and or figures with accompanying text highlighting the main findings in the relevant table.

Chapter 5, Section 1

5.1. Descriptive statistics

This section on Descriptive statistics is presented under the following headings:

- Statistics related to data collection
- Sociodemographic variables
- Physical health variables
- Mental health and adversity variables
- Stigma and disclosure variables
Statistics related to data collection

Table 5.1. Source of respondents

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Where questionnaire was completed:</td>
<td>(n = 88)</td>
</tr>
<tr>
<td>At home</td>
<td>52 (59.1)</td>
</tr>
<tr>
<td>In clinic</td>
<td>36 (40.9)</td>
</tr>
<tr>
<td>Source of recruitment</td>
<td></td>
</tr>
<tr>
<td>Central Middlesex Hospital</td>
<td>36 (38.7)</td>
</tr>
<tr>
<td>North Middlesex Hospital</td>
<td>19 (20.4)</td>
</tr>
<tr>
<td>St Mary’s Hospital Paddington</td>
<td>13 (14.0)</td>
</tr>
<tr>
<td>Sickle Cell Society</td>
<td>25 (26.9)</td>
</tr>
<tr>
<td>Total</td>
<td>93 (100)</td>
</tr>
</tbody>
</table>

Some of the young people who took part in the study were sent postal questionnaires (see Chapter 4) but others were approached through the haematology clinics. Young people approached in the clinics were given the choice of completing the study questionnaires in the clinic or taking the questionnaire home for completion and return by prepaid post. Table 5.1 shows that more respondents (59.1%) completed the questionnaires at home than in the clinics.

Recruitment of subjects took place in the four centres shown in Table 5.1. Most respondents (38.7%) were recruited from Brent Sickle Cell and Thalassaemia Service (based at Central Middlesex Hospital). Sickle Cell Society was the only centre where respondents had only the choice of postal questionnaire.

Section III of this chapter will compare self perceived stigma between respondents who completed their questionnaire in the clinic and those who completed theirs at home. Comparison will also be made between respondents recruited from Sickle Cell Society and the haematology clinics.

Summary of statistics on data collection

In summary, this section showed that most respondents completed the questionnaire at home and of the four study sites, most subjects (38.7%) were recruited from Brent Sickle Cell and Thalassaemia Service (based at Central Middlesex Hospital).
Sociodemographic variables

Table 5.2. Age and gender

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%) or Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender:</td>
<td>(n= 90)</td>
</tr>
<tr>
<td>Males</td>
<td>46 (51.1)</td>
</tr>
<tr>
<td>Females</td>
<td>44 (48.9)</td>
</tr>
<tr>
<td>Age – Mean (SD)</td>
<td>14.2 (2.1)</td>
</tr>
<tr>
<td></td>
<td>Range = 10-19</td>
</tr>
</tbody>
</table>

The respondents were almost evenly split in gender with a slight preponderance of males (51%) (Table 5.2). An even split is expected given that SCDs are autosomal disorders with no genetic gender preponderance. The young people ranged in age from 10-19 years with a Mean of 14.2 years. The age of the respondents was normally distributed (Figure 5.1).

Figure 5.1. Age distribution of respondents
Table 5.3. Ethnicity and UK birth.

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ethnicity:</strong></td>
<td>(n=87)</td>
</tr>
<tr>
<td>Black British</td>
<td>43 (49.4)</td>
</tr>
<tr>
<td>Black African</td>
<td>31 (35.6)</td>
</tr>
<tr>
<td>Black Caribbean</td>
<td>9 (10.3)</td>
</tr>
<tr>
<td>Mixed ethnicity</td>
<td>1 (1.1)</td>
</tr>
<tr>
<td>Asian</td>
<td>3 (3.4)</td>
</tr>
<tr>
<td><strong>Born in UK</strong></td>
<td>(n=91)</td>
</tr>
<tr>
<td>Yes</td>
<td>69 (75.8)</td>
</tr>
<tr>
<td>No</td>
<td>22 (24.2)</td>
</tr>
</tbody>
</table>

The vast majority of respondents described themselves either as Black British (49%) or Black African (35.6%). Three-quarter of the young people were born in the UK (Table 5.3). Parts II and III of this chapter will explore associations between non-UK birth and stigma and depression respectively.

Table 5.4. Family composition and living arrangements

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%) or Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Who lives with:</strong></td>
<td>(n=91)</td>
</tr>
<tr>
<td>Both biological parents</td>
<td>40 (44.0)</td>
</tr>
<tr>
<td>Mother only</td>
<td>42 (46.2)</td>
</tr>
<tr>
<td>Father only</td>
<td>2 (2.2)</td>
</tr>
<tr>
<td>Others</td>
<td>7 (7.7)</td>
</tr>
<tr>
<td><strong>Sibling has Sickle cell disease:</strong></td>
<td>(n=90)</td>
</tr>
<tr>
<td>Yes</td>
<td>26 (28.9)</td>
</tr>
<tr>
<td>No</td>
<td>64 (71.1)</td>
</tr>
<tr>
<td><strong>Number of biological siblings – Median (Range)</strong></td>
<td>1.5 (0-7)</td>
</tr>
</tbody>
</table>

A high proportion of respondents lived with one parent (48.4%) who was most often a mother (46.2%) (Table 5.4). This high proportion of single parent living arrangement is consistent with the demographics of many parts of inner London (e.g. 48% lone parent families in Lambeth [http://www.statistics.gov.uk/CCI/nugget.asp?ID=1166](http://www.statistics.gov.uk/CCI/nugget.asp?ID=1166) (accessed 5/2/10). The median number of siblings is 1.5 (range 0-7); hence if the index child is considered, the median number of children per family in my sample (i.e.2.5) is more than the mean number of children in UK families (1.8 children per family) [http://www.statistics.gov.uk/cci/nugget.asp?id=1163](http://www.statistics.gov.uk/cci/nugget.asp?id=1163) (accessed 5/2/10). About a quarter
of the young people had siblings who also have SCD. This figure is consistent with an autosomal recessively inherited condition where the natural frequency of occurrence is a quarter of all conceptions.

Table 5.5. Socio-economic status

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Socio-economic status (OPCS):</strong></td>
<td>(n=80)</td>
</tr>
<tr>
<td>I = Professional</td>
<td>15 (18.8)</td>
</tr>
<tr>
<td>II = Managerial-technical</td>
<td>10 (12.5)</td>
</tr>
<tr>
<td>III = Skilled</td>
<td>27 (33.8)</td>
</tr>
<tr>
<td>IV = Partly skilled</td>
<td>18 (22.5)</td>
</tr>
<tr>
<td>V = Unskilled</td>
<td>6 (7.5)</td>
</tr>
<tr>
<td>VI = Unemployed</td>
<td>4 (5.0)</td>
</tr>
<tr>
<td><strong>Ownership of car and landline telephone:</strong></td>
<td></td>
</tr>
<tr>
<td>Car</td>
<td>72 (80.0)</td>
</tr>
<tr>
<td>Landline telephone</td>
<td>83 (91.2)</td>
</tr>
</tbody>
</table>

Socio-economic status was assessed in two ways (Table 5.5). First the OPCS occupational classification was used. In the OPCS system, most of the respondents lived in households headed by someone in a skilled occupation (33.8). In this cohort, the proportion of respondents from household headed by someone in professional or managerial occupations (31.3%) is comparable to the figure for black Caribbean women in professional and managerial occupations (30.2%) in the UK (NOS 2005). However, the proportion of heads of household in managerial, professional, and intermediate/skilled occupations in this study (65.1%) is considerably higher than the proportion of Black people in the UK in the same combined occupation bands (45.1% for Black Caribbean and 37.1% for Black Africans) (NOS 2005).

I supplemented the occupation-based OPCS-SES classification with additional information on whether respondent’s family owned a car and or a landline telephone. The vast majority of respondents lived in families that owned a car (80%) and landline telephone (91%).

OPCS-SES category, ownership of a car or landline telephone, and a composite wealth scale that combined these two variables did not correlate with many of the measures of
psychological wellbeing or adversity (e.g. SDQ, SMFQ) (Figures not shown). This observation may be due to low variability resulting from ceiling effect (e.g. nearly all families had a land telephone).

In order to explore for associations between extremes of SES and psychological difficulty as observed in the National Child Mental Health Survey (Meltzer et al 2000), the OPCS-SES categories were further dichotomised into extreme groups by combining Classes 1 and 2 into one category and Classes 4, 5, and 6 into another category. Apart from Child Attitude Toward Illness Scale, the extreme OPCS-SES categories did not correlate with other measures of psychological wellbeing or adversity.

**Summary of socio-demographic data**

In summary, the results in this section of the analysis showed that the respondents were evenly split in gender. Their age was normally distributed with a mean of 14 years. As expected, nearly all were of Black ethnicity (95%). Most were born in the UK, more than half lived with a lone parent, and a quarter had another sibling with SCD. More than two thirds of the cohort came from households headed by someone in a skilled, managerial or professional occupation, which is considerably better than average Black families in the UK.

**Physical health variables**

**Table 5.6: Frequency of admissions**

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Frequency of ward admissions in past year:</strong></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>34 (38.6)</td>
</tr>
<tr>
<td>Once</td>
<td>15 (17.0)</td>
</tr>
<tr>
<td>2-4 times</td>
<td>25 (28.4)</td>
</tr>
<tr>
<td>5-6 times</td>
<td>8 (9.1)</td>
</tr>
<tr>
<td>7 or more times</td>
<td>6 (6.8)</td>
</tr>
<tr>
<td><strong>Frequency of A&amp;E attendance in past year:</strong></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>42 (48.3)</td>
</tr>
<tr>
<td>Once</td>
<td>29 (33.3)</td>
</tr>
<tr>
<td>2-4 times</td>
<td>11 (12.6)</td>
</tr>
<tr>
<td>5-6 times</td>
<td>3 (3.4)</td>
</tr>
<tr>
<td>7 or more times</td>
<td>2 (2.3)</td>
</tr>
</tbody>
</table>
I obtained data on both admissions to the ward and to Accident and Emergency Departments. The ward admissions include patients who had planned admissions, for example, for blood transfusion as well as those admitted to the ward after a period in A&E. Data on A&E attendance is meant to capture more acute complications. The data shows that two-thirds and more than half of the respondents had had at least one ward or A&E admission in the past year respectively (Table 5.6).

**Table 5.7. Experience of pain**

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Frequency of experience of pain:</strong></td>
<td>(n=76)</td>
</tr>
<tr>
<td>More than once a week</td>
<td>7 (9.2)</td>
</tr>
<tr>
<td>Once a week</td>
<td>10 (13.2)</td>
</tr>
<tr>
<td>Twice a month</td>
<td>23 (30.3)</td>
</tr>
<tr>
<td>Once in 1-5 months</td>
<td>14 (18.4)</td>
</tr>
<tr>
<td>Twice a year or less frequent</td>
<td>22 (29.0)</td>
</tr>
<tr>
<td><strong>Intensity of pain:</strong></td>
<td>(n=82)</td>
</tr>
<tr>
<td>Mild or no pain</td>
<td>14 (16.4)</td>
</tr>
<tr>
<td>Moderate</td>
<td>21 (24.7)</td>
</tr>
<tr>
<td>Intense</td>
<td>29 (34.1)</td>
</tr>
<tr>
<td>Very intense</td>
<td>18 (21.2)</td>
</tr>
</tbody>
</table>

Pain was assessed as an additional measure of severity and disruptiveness in SCD among the respondents. More than a fifth of the subjects experienced pain once a week or more often (Table 5.7). However about half (47.4%) experienced infrequent pain (once a month or less frequently). More than half (55.3%) reported intense or very intense pain.

**Table 5.8 School absence**

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Number of days absent from school in past year:</strong></td>
<td>(n=88)</td>
</tr>
<tr>
<td>None</td>
<td>8 (9.3)</td>
</tr>
<tr>
<td>Less than 7 days</td>
<td>26 (30.2)</td>
</tr>
<tr>
<td>7-14 days</td>
<td>21 (24.4)</td>
</tr>
<tr>
<td>15-21 days</td>
<td>11 (12.8)</td>
</tr>
<tr>
<td>22-28 days</td>
<td>4 (4.7)</td>
</tr>
<tr>
<td>More than 28 days</td>
<td>16 (18.6)</td>
</tr>
</tbody>
</table>
School absence is another index of severity and disruptiveness of SCD. Like previous measures of severity, the school absence data illustrates the high level of variability in SCD with some (9.3%) having had no school absence while 18.6% had more than 28 days (Table 5.8). The school absence rate for the children in this study is worse than the average data for secondary school students in England in the year 2007/2008, which showed that 6.1% had no absence, while 8.9% had more than 25% days of absence [http://www.dcsf.gov.uk/rsgateway/DB/SFR/s000832/SFR03_2009NationalTablesv2.xls](http://www.dcsf.gov.uk/rsgateway/DB/SFR/s000832/SFR03_2009NationalTablesv2.xls). (accessed 5/2/10)

Table 5.9 Visual signs of SCD (jaundice and leg ulcer)

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jaundiced:</td>
<td>(n=89)</td>
</tr>
<tr>
<td>Yes</td>
<td>41 (46.1)</td>
</tr>
<tr>
<td>No</td>
<td>48 (53.9)</td>
</tr>
<tr>
<td>Leg ulcer:</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Presence of jaundice and or leg ulcer:</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>No</td>
<td></td>
</tr>
</tbody>
</table>

Jaundice and leg ulcer were used as indices of visual manifestations of SCD. Nearly half of the respondents were jaundiced but only 5.6% had leg ulcer (Table 5.9). The proportion with either jaundice or leg ulcer was 46.1%. This proportion is the same as the proportion with jaundice because all the subjects with leg ulcer were also jaundiced.
Table 5.10 Treatment adherence

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>(n=69)</td>
</tr>
<tr>
<td><strong>Adherence to folic acid</strong></td>
<td></td>
</tr>
<tr>
<td>Usually remembers</td>
<td>34 (49.3)</td>
</tr>
<tr>
<td>Sometimes forgets</td>
<td>18 (26.1)</td>
</tr>
<tr>
<td>Often forgets</td>
<td>17 (24.6)</td>
</tr>
<tr>
<td><strong>Adherence to penicillin</strong></td>
<td></td>
</tr>
<tr>
<td>Usually remembers</td>
<td>27 (33.3)</td>
</tr>
<tr>
<td>Sometimes forgets</td>
<td>31 (38.3)</td>
</tr>
<tr>
<td>Often forgets</td>
<td>23 (28.4)</td>
</tr>
<tr>
<td><strong>Clinic attendance:</strong></td>
<td></td>
</tr>
<tr>
<td>Usually attends</td>
<td>83 (91.2)</td>
</tr>
<tr>
<td>Sometime or often forgets</td>
<td>8 (8.8)</td>
</tr>
</tbody>
</table>

In addition to clinic attendance, adherence to two medications commonly prescribed for people with SCD were used to assess treatment adherence. Half of the respondents prescribed folic acid usually remembered to take the medication (Table 5.10). Adherence to penicillin was more limited with only a third of respondents usually remembering to take the medication. Unlike medications, most respondents reported they usually attended their clinic appointments. The fact that respondents had much better clinic attendance than medication adherence may be related to the fact that clinic appointments are infrequent but more tangible events; hence easier to remember compared with medications which have to be taken daily.

**Summary of physical health variables**

In summary, the results in this section of the analysis showed that there was significant variability in the physical health of the respondents. Half were jaundiced but only 6% had leg ulcer. Two thirds had had at least one ward admission and half had had one or more admissions to Accident and Emergency department in the past year. A fifth experienced frequent pain (weekly or more often) and 18% had been absent from school for month or longer in the previous year.
Mental health and adversity variables

Table 5.11 Strengths and Difficulties Questionnaire (SDQ)

Table 5.11 displays the Total Difficulties scale of the SDQ and all the five subscales. The Total Difficulties Scale is derived from a sum of the subscales (excluding the prosocial subscale). Table 5.11 shows the mean scores for subjects in this study and the proportion scoring above the cut-off for the Total difficulties scale and the five subscales.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Mean (SD) or N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Total Difficulties score – Mean (SD) (n=78)</strong></td>
<td></td>
</tr>
<tr>
<td>Abnormal (≥ 18) N(%)</td>
<td>10.8 (5.4)</td>
</tr>
<tr>
<td>Abnormal (≥ 18) N(%)</td>
<td>12 (15.4)</td>
</tr>
<tr>
<td><strong>Conduct subscale – Mean (SD) (n=88)</strong></td>
<td></td>
</tr>
<tr>
<td>Abnormal ≥ 5 N(%)</td>
<td>2.2 (1.6)</td>
</tr>
<tr>
<td>Abnormal ≥ 5 N(%)</td>
<td>10 (11.4)</td>
</tr>
<tr>
<td><strong>Emotional subscale – Mean (SD) (n=86)</strong></td>
<td></td>
</tr>
<tr>
<td>Abnormal ≥ 7 N(%)</td>
<td>3.6 (2.2)</td>
</tr>
<tr>
<td>Abnormal ≥ 7 N(%)</td>
<td>9 (10.3)</td>
</tr>
<tr>
<td><strong>Peer problems subscale – Mean (SD) (n=83)</strong></td>
<td></td>
</tr>
<tr>
<td>Abnormal ≥ 6 N(%)</td>
<td>1.7 (1.9)</td>
</tr>
<tr>
<td>Abnormal ≥ 6 N(%)</td>
<td>5 (6.0)</td>
</tr>
<tr>
<td><strong>Hyperactivity subscale – Mean (SD) (n=84)</strong></td>
<td></td>
</tr>
<tr>
<td>Abnormal ≥ 7 N(%)</td>
<td>3.3 (2.0)</td>
</tr>
<tr>
<td>Abnormal ≥ 7 N(%)</td>
<td>4 (4.7)</td>
</tr>
<tr>
<td><strong>Prosocial subscale – Mean (SD) (n=85)</strong></td>
<td></td>
</tr>
<tr>
<td>Abnormal ≤ 4 N(%)</td>
<td>7.6 (1.9)</td>
</tr>
<tr>
<td>Abnormal ≤ 4 N(%)</td>
<td>3 (3.5)</td>
</tr>
</tbody>
</table>

In this study, a cut-off of 18 and above was used for the self report SDQ because this cut-off gave prevalence figures in the British Child and Adolescent Mental Health Survey (Meltzer et al 2000) that were equivalent to the prevalence found using data from multiple sources including parents and teachers (Stansfeld et al 2004). Using this cut-off, the percentage of young people classified as SDQ-cases was 15.4%. This percentage was higher but not statistically significantly different when compared with the percentages of SDQ-caseness among young people in London including Black boys (9.2%) or Black girls (10.9%) (Stansfeld et al (2004) (comparison done with the StatCalc function of Epi Info statistical package, $\chi = 3.1$, $p = 0.078$ which is not significant).
With respect to the subscales, the mean scores for the young people with SCD in this study are compared with UK-normative data in Table 5.12 below.

### Table 5.12 SDQ subscale scores compared with UK norms

<table>
<thead>
<tr>
<th>SDQ Subscales</th>
<th>Young people with SCD (this study) (Mean SD) (see each scale for N)</th>
<th>*UK norms (Mean SD) (n=4228)</th>
<th>T-Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total difficulties scale</td>
<td>10.8 (5.4)</td>
<td>10.3 (5.2)</td>
<td>T=0.88 CI-1.6, 0.6</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>P=0.4</td>
</tr>
<tr>
<td>Conduct subscale</td>
<td>2.2 (1.6)</td>
<td>2.2 (1.7)</td>
<td>equal</td>
</tr>
<tr>
<td>Emotional Subscale</td>
<td>3.6 (2.2)</td>
<td>2.8 (2.1)</td>
<td>T=3.47 CI 0.35, 1.25</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>P=0.0008</td>
</tr>
<tr>
<td>Peer problems subscale</td>
<td>1.7 (1.9)</td>
<td>1.5 (1.4)</td>
<td>T=1.0 CI -0.18, 0.59</td>
</tr>
<tr>
<td>Hyperactivity subscale</td>
<td>3.3 (2.0)</td>
<td>3.8 (2.2)</td>
<td>T=-2.4 CI -0.91, -0.09</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>P=0.019</td>
</tr>
<tr>
<td>Prosocial subscale</td>
<td>7.6 (1.9)</td>
<td>8.0 (1.7)</td>
<td>T=-2.0 CI -0.79, -0.01</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>P=0.047</td>
</tr>
</tbody>
</table>


The differences in mean scores and standard deviations were compared for statistical significance using a free online statistical package available at [http://www.quantitativeskills.com/sisa/statistics/t-test.htm](http://www.quantitativeskills.com/sisa/statistics/t-test.htm) (accessed 2/10/09). The comparison showed that the young people with SCD score significantly higher than the norm on the emotional subscale and significantly less on the hyperactivity and Prosocial subscales (Table 5.12). While these findings are of interest, it is worth noting that the differences may not be clinically significant given that in all subscales, the Mean scores for children with SCDs are below the cut-off for abnormal scores.
(See Section 4.9.4). However, the increase in emotional symptoms over expected norm is in line with the literature on paediatric chronic illness. The reduced hyperactivity and prosocial scores are of additional interest as this has not been examined sufficiently in the literature on chronic illness in children.

**Table 5.13. Gender comparison on the SDQ**

<table>
<thead>
<tr>
<th></th>
<th>Male Mean (SD)</th>
<th>Female Mean (SD)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>SDQ Total Difficulties scale (n=77)</td>
<td>10.3 (4.6)</td>
<td>11.3 (6.0)</td>
<td>T = -0.84, df=72.2, CI -3.4,1.4, p=0.4</td>
</tr>
<tr>
<td>Conduct subscale (n=87)</td>
<td>2.3 (1.5)</td>
<td>2.0 (1.7)</td>
<td>T = 0.98, df=81.1, CI -0.3,1.0, p=0.3</td>
</tr>
<tr>
<td>Emotional subscale (n=86)</td>
<td>3.3 (2.2)</td>
<td>4.2 (2.2)</td>
<td>T=-1.9, df=83.9, CI -1.8,0.4, p=0.06</td>
</tr>
<tr>
<td>Peer problem subscale (n=83)</td>
<td>1.5 (1.8)</td>
<td>2.0 (1.8)</td>
<td>T=-1.2, df=81.0, CI -1.3,0.3, p=0.2</td>
</tr>
<tr>
<td>Hyperactivity subscale (n=84)</td>
<td>3.3 (1.8)</td>
<td>3.2 (2.0)</td>
<td>T=0.3, df=81.8, CI -0.7,1.0, p=0.8</td>
</tr>
<tr>
<td>Prosocial subscale (n=85)</td>
<td>7.7 (2.0)</td>
<td>7.6 (1.8)</td>
<td>T=0.1, df=82.8, CI -0.7,0.9, p=0.9</td>
</tr>
</tbody>
</table>

Unlike previous studies (e.g. Stansfeld et al 2004), which showed gender differences in the SDQ, Table 5.13 shows no significant gender differences in Total Difficulties scale or any of the five subscales of the SDQ in this study.
Table 5.14 Depressive symptoms

<table>
<thead>
<tr>
<th>Variable</th>
<th>N (%) or Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Short Mood and Feeling Questionnaire (SMFQ) score – Mean (SD) (n=88)</td>
<td>4.5 (4.0)</td>
</tr>
<tr>
<td>Depression-positive cases N(%)</td>
<td>(n=88)</td>
</tr>
<tr>
<td>Depression-negative cases N(%)</td>
<td>16 (18.2)</td>
</tr>
<tr>
<td></td>
<td>72 (81.8)</td>
</tr>
</tbody>
</table>

The mean score on the SMFQ by respondents in the current study (4.5) is similar to scores by non-depressed children in other UK studies (4.6) (Thapar and McGuffin 1998) and (4.4) (Stansfeld et al 2004). I used a cut off score of 8.0 and above on the SMFQ to define caseness for depression. This cut-off was used in the original validation study for the self report version of the SMFQ in USA (Angold et al 1995) and in two other UK studies (Thapar and McGuffin 1998, Stansfeld et al 2004). In the current study, the cut-off $\geq 8.0$ identified 18.2% of respondents as depression-positive (Table 5.14).

Table 5.15 Depression caseness and gender

<table>
<thead>
<tr>
<th>Gender</th>
<th>Depression Positive</th>
<th>Depressive negative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male N(%)</td>
<td>(n=16)</td>
<td>(n=71)</td>
</tr>
<tr>
<td>4 (9.1)</td>
<td>40 (90.9)</td>
<td></td>
</tr>
<tr>
<td>Female N(%)</td>
<td>12 (27.9)</td>
<td>31 (72.1)</td>
</tr>
</tbody>
</table>

Fishers Exact test p = 0.024

The association between depression and gender was explored (Table 5.15). Female respondents were statistically significantly more likely than male respondents to be classified as depression-positive on the SMFQ. This is consistent with previous studies of the SMFQ in the UK including among black children (Stansfeld et al 2004).
Table 5.16 Depression and receipt of counselling from a therapist

<table>
<thead>
<tr>
<th></th>
<th>Depression Positive (n=16)</th>
<th>Depressive negative (n=71)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Receiving Counselling</td>
<td>N(%)</td>
<td>N(%)</td>
</tr>
<tr>
<td>Yes</td>
<td>3 (18.8)</td>
<td>7 (9.9)</td>
</tr>
<tr>
<td>No</td>
<td>13 (81.3)</td>
<td>64 (90.1)</td>
</tr>
</tbody>
</table>

Fishers Exact test p=0.27

Overall, 11.5% of the young people in the study were receiving counselling at the time of completing the questionnaire. Table 5.16 shows that only 18.8% of the children who were classified as depression-positive were in receipt of counselling. Also there was no statistically significant difference in receipt of counselling between the depression-positive and depression-negative cases.

Table 5.17. Self esteem

<table>
<thead>
<tr>
<th></th>
<th>Rosenberg Self Esteem Scale – Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Young people with SCD (this study) (n=80)</td>
<td>31.9 (5.0)</td>
</tr>
<tr>
<td>USA* (n=543)</td>
<td>31.9 (5.0)</td>
</tr>
<tr>
<td>Canada* (n=1443)</td>
<td>31.0 (4.8)</td>
</tr>
<tr>
<td>New Zealand* (n=300)</td>
<td>29.9 (4.5)</td>
</tr>
</tbody>
</table>

* Data from a cross national comparison among students in USA, Canada and New Zealand (Rusticus et al 2004).

Table 5.17 shows the mean score (and standard deviation) of the young people with SCD on the Rosenberg Self Esteem Scale. The scores are same or better than scores from young people in three other developed countries.
Table 5.18 Child Attitude to Illness Scale (CATIS)

<table>
<thead>
<tr>
<th></th>
<th>CATIS Mean (SD)</th>
<th>T-tests comparing SCD separately with diabetes, severe, and mild epilepsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Young people with SCD (this study) (n=88)</td>
<td>3.1 (0.8)</td>
<td></td>
</tr>
<tr>
<td>Children with Type I Diabetes* (n=31)</td>
<td>3.6 (0.8)</td>
<td>T=-2.99</td>
</tr>
<tr>
<td></td>
<td></td>
<td>CI -0.83, -0.17</td>
</tr>
<tr>
<td></td>
<td></td>
<td>P=0.004</td>
</tr>
<tr>
<td>Children with severe epilepsy** (104)</td>
<td>3.2 (0.8)</td>
<td>T=-0.86</td>
</tr>
<tr>
<td></td>
<td></td>
<td>CI -0.33, 0.13</td>
</tr>
<tr>
<td></td>
<td></td>
<td>P=0.39</td>
</tr>
<tr>
<td>Children with mild epilepsy** (n=41)</td>
<td>3.9 (0.6)</td>
<td>T=-6.3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>CI -1.11, -0.55</td>
</tr>
<tr>
<td></td>
<td></td>
<td>P=0.000</td>
</tr>
</tbody>
</table>


Using the free online statistical package at [http://www.quantitativeskills.com/sisa/statistics/t-test.htm](http://www.quantitativeskills.com/sisa/statistics/t-test.htm) (accessed 2/10/10), it was shown that young people with SCD had significantly worse attitude toward illness than children with Type I Diabetes Mellitus and mild epilepsy. Young people with SCD scored similar to children with severe epilepsy (Table 5.18).

Table 5.19 Family function

<table>
<thead>
<tr>
<th></th>
<th>Raw score – Mean (SD) (n=79)</th>
<th>Standardised Scale – Mean (SD)</th>
<th>Proportion with “healthy” family function N (%)</th>
<th>Proportion with “unhealthy” family function N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><a href="#">Family Assessment Device (FAD):</a></td>
<td>20.8 (5.1)</td>
<td>1.7 (0.4)</td>
<td>63 (79.7)</td>
<td>16 (20.3)</td>
</tr>
</tbody>
</table>

A fifth of the subjects in the study (20.3%) were classified as living in families with unhealthy function (Table 5.19). This is comparable to 19% found in the British Child and Adolescent Mental Health Survey (Meltzer et al 2000), which used a similar definition. Several studies including Meltzer et al (2000) implicate unhealthy family
function in a range of psychological difficulties. Bearing this in mind, I will use FAD as a covariate in Part IV for multivariate analyses and testing of hypotheses.

**Summary of mental health and adversity variables**

In summary, the results of this section of the analysis showed that 15.4% of the respondents were classified as SDQ-cases. In line with the literature on paediatric chronic illness, this cohort scored significantly higher than UK norm on the emotional subscale of the SDQ. No differences were observed in other subscales. Also no gender differences were seen in any SDQ subscale. In relation to depression, 18.2% of respondents were classified as depression-positive cases and female respondents were significantly more likely to be so classified than males. However, only 18.8% of the children who were classified as depression-positive were in receipt of counselling. The young people’s attitude towards SCD was similar to children with severe epilepsy. However, they had normative levels of self esteem. The proportion living in families with unhealthy function (20%) was comparable to UK norm.

**Stigma and disclosure questions**

**Table 5.20. Stigma questions**

Self-perceived stigma was assessed with three questions, which are shown in Table 5.20 below. These questions were adapted from two previous studies of self-perceived stigma among children with epilepsy (Westbrook et al 1992) and stuttering (Blood et al 2003). For ease of comparison, data from the latter two studies are displayed in Table 5.20 against data from the current study on young people with SCD.
### Stigma questions:

<table>
<thead>
<tr>
<th>Stigma questions:</th>
<th>Sickle cell N(%)</th>
<th>Epilepsy (Westbrooke et al 1992) %</th>
<th>Stuttering (Blood et al 2003)%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Do you think that having Sickle Cell affects whether people want to be friends with you?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never</td>
<td>65 (73.0)</td>
<td>66</td>
<td>65</td>
</tr>
<tr>
<td>Rarely</td>
<td>14 (15.7)</td>
<td>12</td>
<td>10</td>
</tr>
<tr>
<td>Sometimes</td>
<td>7 (7.9)</td>
<td>16</td>
<td>15</td>
</tr>
<tr>
<td>Often</td>
<td>3 (3.4)</td>
<td>6</td>
<td>10</td>
</tr>
<tr>
<td><strong>Do you think that having Sickle Cell affects whether people like you or not?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never</td>
<td>76 (83.5)</td>
<td>60</td>
<td>63</td>
</tr>
<tr>
<td>Rarely</td>
<td>8 (8.8)</td>
<td>14</td>
<td>12</td>
</tr>
<tr>
<td>Sometimes</td>
<td>6 (6.6)</td>
<td>20</td>
<td>10</td>
</tr>
<tr>
<td>Often</td>
<td>1 (1.1)</td>
<td>6</td>
<td>15</td>
</tr>
<tr>
<td><strong>Do you think that having Sickle Cell affects whether or not you are invited to people’s homes or to parties?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Never</td>
<td>75 (82.4)</td>
<td>69</td>
<td>60</td>
</tr>
<tr>
<td>Rarely</td>
<td>8 (8.8)</td>
<td>5</td>
<td>15</td>
</tr>
<tr>
<td>Sometimes</td>
<td>6 (6.6)</td>
<td>17</td>
<td>10</td>
</tr>
<tr>
<td>Often</td>
<td>2 (2.2)</td>
<td>9</td>
<td>15</td>
</tr>
<tr>
<td><strong>Prevalence of Self-perceived Stigma</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>N (%)</td>
<td>14 (15.1)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Stigma scale – Mean (SD)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>4.0 (1.8)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Range 3-10</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

A comparison of the proportion of young people answering “Never” to all the three stigma questions suggest that young people with SCD experience less self-perceived stigma compared with peers with epilepsy and stuttering (Table 5.20).

In order to maximise the analytical power of the stigma questions, responses to the three stigma questions were combined into a Stigma Scale (Mean 4.0, Range 3-10) (higher scores = more self perceived stigma).

---

3 In this study, I defined respondents as having self–perceived stigma if they endorsed a positive answer (i.e. sometimes or often) to at least one of the three stigma questions. Based on this definition, the proportion of young people with self-perceived stigma was 15.1%.
Direct questions about perceived discrimination may be open to socially desirable responding. This is a common liability for questions assessing social attitudes. To gain additional insight into self-perceived stigma with less risk of interference from socially desirable responding, questions on disclosure were introduced (Table 5.21).

**Table 5.21 Disclosure of stigma questions**

Four additional questions on “disclosure of stigma” were used to assess avoidance behaviour. These are used as indirect measures of self-perceived stigma that may be less vulnerable to socially desirable responding. The questions shown in Table 5.21 were also adapted from previous studies of epilepsy (Westbrook et al 1992) and stuttering (Blood et al 2003). The data from these two studies are displayed for comparison against the data from the current study of young people with SCD.

<table>
<thead>
<tr>
<th>Disclosure questions:</th>
<th>Sickle Cell N (%)</th>
<th>Epilepsy (Westbrook et al 1992)%</th>
<th>Stuttering (Blood et al 2003)%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Do you keep your sickle cell a secret from others?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Often</td>
<td>19 (20.9)</td>
<td>20</td>
<td>21</td>
</tr>
<tr>
<td>Sometimes</td>
<td>30 (33.0)</td>
<td>33</td>
<td>19</td>
</tr>
<tr>
<td>Rarely</td>
<td>13 (14.3)</td>
<td>16</td>
<td>21</td>
</tr>
<tr>
<td>Never</td>
<td>29 (31.9)</td>
<td>31</td>
<td>39</td>
</tr>
<tr>
<td><strong>How often do you talk to people about your sickle cell?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Often</td>
<td>7 (7.7)</td>
<td>10</td>
<td>21</td>
</tr>
<tr>
<td>Sometimes</td>
<td>32 (35.2)</td>
<td>20</td>
<td>19</td>
</tr>
<tr>
<td>Rarely</td>
<td>34 (37.4)</td>
<td>50</td>
<td>46</td>
</tr>
<tr>
<td>Never</td>
<td>18 (19.8)</td>
<td>20</td>
<td>14</td>
</tr>
<tr>
<td><strong>Do any of your friends know that you have sickle cell?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>All</td>
<td>27 (29.7)</td>
<td>33</td>
<td>83</td>
</tr>
<tr>
<td>Some</td>
<td>35 (38.5)</td>
<td>45</td>
<td>10</td>
</tr>
<tr>
<td>Few</td>
<td>20 (22.0)</td>
<td>19</td>
<td>2</td>
</tr>
<tr>
<td>None</td>
<td>9 (9.9)</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td><strong>When people find out you have sickle cell, is it usually because:</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>You tell them?</td>
<td>62 (68.9)</td>
<td>65</td>
<td>15</td>
</tr>
<tr>
<td>They see you have a sign of sickle cell and then you explain?</td>
<td>20 (22.2)</td>
<td>19</td>
<td>15</td>
</tr>
<tr>
<td>Someone else tells them about it?</td>
<td>7 (7.8)</td>
<td>16</td>
<td>22</td>
</tr>
</tbody>
</table>
Unlike the direct questions on self-perceived stigma, more of the respondents with SCD positively endorsed the disclosure questions (Table 5.21). For example, 54% often or sometimes keep their having SCD secret and 57% rarely or never talk to other people about having SCD. This pattern and proportion of responses are also evident from the data on children with epilepsy and stuttering. Thus “disclosure questions” suggest that the true prevalence of self-perceived stigma could be more than indicated by the direct measures.

Summary of stigma and disclosure

A summary of this section of the analysis showed that 15% of the subjects were classified as having self-perceived stigma based on direct questions on stigma. However, more respondents positively endorsed indirect questions on stigma based on avoidance and disclosure behaviour. The latter suggest that the prevalence of self-perceived stigma among young people with SCD could be more than indicated by the direct questions. Overall, young people in the current study endorsed less self-perceived stigma compared with peers with epilepsy and stuttering.
Chapter 5: Section II

5.2. Unadjusted bivariate associations between stigma and other variables

Self-perceived stigma is the main focus of this study. This section will therefore focus on exploring the association between stigma and other variables. The analyses shown in this section are bivariate (between self-perceived stigma and other variables individually). The purpose of this Section is to identify variables that have significant associations with self-perceived stigma, which would be considered later in Section IV for multivariate analyses. The associations explored here in Section II are not adjusted for confounding or interactions. Confounding and interactions will be considered in Section IV (multivariate analyses).

In order to present comparisons between stigma and other variables as proportions, which are more readily appreciated in tables, I used the categorical definition of self-perceived stigma (i.e. proportion of respondents who endorsed a positive answer to at least one of the stigma questions) in this Section. However, where the analysis shows no statistically significant association, I conducted a further analysis using the dimensional measure of stigma (Stigma Scale) to ensure that no statistically significant association was missed by the penalisation of the data that could result from using categorical data where a dimensional alternative is available. The result of the additional dimensional analysis is shown only in cases where the two analytical strategies yield different results. Otherwise, only the categorical result is shown.

5.2.1 Associations between stigma and socio-demographic variables

The study gathered a considerable amount of socio-demographic data. However, in the section that follows, associations are explored only for socio-demographic variables with theoretical or putative relationships with self-perceived stigma, comparing the 14 participants with perceived stigma according to the above categorical definition with the 77 children with no or low perceived stigma.
Table 5.22. Perceived stigma and gender

<table>
<thead>
<tr>
<th>Gender:</th>
<th>Perceived Stigma (n=14)</th>
<th>No or low perceived stigma (n=76)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male N (%)</td>
<td>4 (8.7)</td>
<td>42 (91.3)</td>
<td>Fishers Exact</td>
</tr>
<tr>
<td>Female N (%)</td>
<td>10 (22.7)</td>
<td>34 (77.3)</td>
<td>p = 0.085</td>
</tr>
</tbody>
</table>

Table 5.22 shows that a higher percentage of females than males were classified as having self-perceived stigma (22.7% vs. 8.7%) but, the difference was not statistically significant.

Table 5.23 Perceived stigma and age

<table>
<thead>
<tr>
<th>Age - Mean (SD)</th>
<th>Perceived Stigma (n=14)</th>
<th>No or low perceived stigma (n=77)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>13.4 (2.5)</td>
<td>13.4 (2.5)</td>
<td>14.4 (2.1)</td>
<td>T=-1.5, df 86, CI -2.16, 0.29, p = 0.13</td>
</tr>
</tbody>
</table>

Table 5.23 shows no statistically significant difference in mean age between the respondents classified as having self-perceived stigma and those without perceived stigma.

Table 5.24 Perceived stigma and having a sibling with SCDs

<table>
<thead>
<tr>
<th>Sibling with SCD:</th>
<th>Perceived Stigma (n=13)</th>
<th>No or low perceived stigma (n=77)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes N (%)</td>
<td>4 (15.4)</td>
<td>22 (84.6)</td>
<td>Fisher’s Exact</td>
</tr>
<tr>
<td>No N (%)</td>
<td>9 (14.1)</td>
<td>55 (85.9)</td>
<td>p = 1.0</td>
</tr>
</tbody>
</table>

The above table shows that there was no statistically significant difference in the percentage of young people with a sibling with SCD who were classified as having self perceived stigma compared with those with no affected sibling Table 5.24).
The above table shows that there was no statistically significant difference in the percentage of children with SCD born in the UK who were classified as having self-perceived stigma compared with those born outside the UK (Table 5.25).

Table 5.25 Perceived stigma and UK birth

<table>
<thead>
<tr>
<th>Born in UK:</th>
<th>Perceived Stigma (n=14)</th>
<th>No or low perceived stigma (n=77)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes - N (%)</td>
<td>9 (13.0)</td>
<td>60 (87.0)</td>
<td>Fishers Exact p = 0.31</td>
</tr>
<tr>
<td>No – N (%)</td>
<td>5 (22.7)</td>
<td>17 (77.3)</td>
<td></td>
</tr>
</tbody>
</table>

Table 5.25 shows no statistically significant difference in the percentage of children with SCD born in the UK who were classified as having self-perceived stigma compared with those born outside the UK (Table 5.25).

Table 5.26 Self-perceived stigma and Socio-economic status

To reduce the number of cells with small numbers, the six OPCS-SES categories were collapsed into three groups by combining Groups 1 and 2, 3 and 4, and 5 and 6 as shown in Table 5.26.

<table>
<thead>
<tr>
<th>OPCS-SES category:</th>
<th>Perceived Stigma (n=12)</th>
<th>No or low perceived stigma (n=68)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 &amp; 2 - N (%)</td>
<td>5 (20.0)</td>
<td>20 (80.0)</td>
<td>χ² = 3.4 df= 2 p = 0.17</td>
</tr>
<tr>
<td>3 &amp; 4 - N (%)</td>
<td>4 (8.9)</td>
<td>41 (91.1)</td>
<td></td>
</tr>
<tr>
<td>5 &amp; 6 – N (%)</td>
<td>3 (30)</td>
<td>7 (70.0)</td>
<td></td>
</tr>
</tbody>
</table>

Table 5.26 shows no statistically significant association between OPCS-SES category and classification as having self-perceived stigma. The analysis was repeated with other measures of socio-economic status (ownership of a car and land telephone line) which also showed no statistically significant association (Figures not shown).

Summary of stigma and Sociodemographic variables

In summary, the results of this section of the analysis shows that no socio-demographic variable (gender, age, whether or not born in the UK, having a sibling with SCD, and SES) was statistically significantly associated with self-perceived stigma.
5.2.2. Associations between stigma and illness variables

Table 5.27 Self perceived stigma and presence of jaundice

<table>
<thead>
<tr>
<th>Perceived Stigma (n=14)</th>
<th>No or low perceived stigma (n=75)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jaundice:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes N(%)</td>
<td>6 (14.6)</td>
<td>35 (85.4)</td>
</tr>
<tr>
<td>No N(%)</td>
<td>8 (16.7)</td>
<td>40 (83.3)</td>
</tr>
<tr>
<td></td>
<td>$\chi^2 = 0.07$</td>
<td>df = 1</td>
</tr>
<tr>
<td></td>
<td>p = 0.8</td>
<td></td>
</tr>
</tbody>
</table>

Jaundice is one of the physical manifestations of SCD. Stigma theory predicts that physical signs that increase visibility increase the stigmatising potential of a condition. However, Table 5.27 shows no significant association between stigma and presence of jaundice.

Table 5.28 Self-perceived stigma and leg ulcer

<table>
<thead>
<tr>
<th>Perceived Stigma (n=14)</th>
<th>No or low perceived stigma (n=75)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leg ulcer:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes N(%)</td>
<td>3 (60.0)</td>
<td>2 (40.0)</td>
</tr>
<tr>
<td>No N(%)</td>
<td>11 (13.1)</td>
<td>73 (86.9)</td>
</tr>
<tr>
<td></td>
<td>Fishers Exact test</td>
<td>p = 0.03</td>
</tr>
</tbody>
</table>

The presence of leg ulcer is another physical feature seen in SCD. It is a less common feature than jaundice (5.6% vs. 46.1% in this study). However, unlike jaundice, Table 5.28 shows that the presence of leg ulcer is statistically significantly associated with self-perceived stigma. This therefore provides preliminary support for the application of stigma theory to SCD, which suggests that having a visible sign increases the stigmatising potential of a condition. This association is preliminary because it has not been subjected to multivariate analysis to partial out the effect of other variables that may be potential confounders. This will be addressed in Section IV of this chapter.
Self-perceived stigma and measures of severity of SCD

Five surrogate variables were used to assess severity in SCD: frequency of ward admissions, frequency of admissions to Accident and Emergency department, frequency of school absence, frequency of experience of pain, and intensity of pain. These measures of severity were included as indices of disruptiveness because stigma theory proposes that disruptiveness increases the stigmatising potential of a condition. Given that the above measures of severity were determined in categories with up to five or more options, I decided that for purposes of bivariate and multivariate analyses, it would not be appropriate to assess their relationships with self-perceived stigma with cross-tabulation as the loss of power would increase the risk of Type II errors. Instead, I treated the categories in the measures of severity as ordinal scales and calculated correlation coefficients (Spearman rho) between them and the dimensional measure of stigma (Table 5.29).

Table 5.29 Correlation coefficients between self-perceived stigma and measures of severity

<table>
<thead>
<tr>
<th></th>
<th>Stigma (dimensional)</th>
</tr>
</thead>
<tbody>
<tr>
<td>School absence</td>
<td>R .09 N 86</td>
</tr>
<tr>
<td>Pain frequency</td>
<td>R .04 N 74</td>
</tr>
<tr>
<td>Pain intensity</td>
<td>R .30** N 83</td>
</tr>
<tr>
<td>Frequency of ward admissions</td>
<td>R .34** N 86</td>
</tr>
<tr>
<td>Frequency of A&amp;E admissions</td>
<td>R .31** N 85</td>
</tr>
</tbody>
</table>

$r$ = Spearman rho correlation  
** Correlation is significant at the 0.01 level (2-tailed).

Table 5.29 shows that self-perceived stigma had statistically significant correlation with only three of the five measures of severity (pain intensity, frequency of ward admissions and frequency of Accident and Emergency admissions). Based on the coding scheme of the data, the direction of the correlations indicates that young people who experienced more of these three measures of severity had more self-perceived stigma. This finding provides further preliminary support for the
application of stigma theory to SCD, which predicts that the more disruptive a condition is the more stigmatising it is likely to be. The independence of this association will be explored further with multivariate analysis in Section IV of this chapter.

Table 5.30 Self-perceived stigma and clinic attendance:

<table>
<thead>
<tr>
<th>Clinic attendance:</th>
<th>Perceived Stigma (n=14)</th>
<th>No or low perceived stigma (n=77)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usually remembers to attend. N(%)</td>
<td>12 (14.5)</td>
<td>71 (85.5)</td>
<td>Fisher’s Exact p = 0.6</td>
</tr>
<tr>
<td>Sometimes or often forgets to attend. N(%)</td>
<td>2 (25.0)</td>
<td>6 (75.0)</td>
<td></td>
</tr>
</tbody>
</table>

The association in Table 5.30 was explored because of the possibility that young people who perceive stigma may be less likely to associate with activities that highlight the saliency of SCD such as attending appointments at sickle cell clinics. The Table shows no significant association between self-perceived stigma and clinic attendance.

Tables 5.31 (a,b,c) Self-perceived stigma and medication adherence:
As with clinic attendance, the association between self-perceived stigma and medication adherence was explored on the basis that young people with increased self-perceived stigma may be less likely to adhere to their SCD medication because the latter makes the disorder more salient for them. More data was available on adherence to two medications commonly prescribed for people with SCD (folic acid and penicillin). The analysis was based on adherence to penicillin as this had more complete data compared with folic acid. One-way ANOVA was used to compare the Mean scores on the dimensional stigma scale among the three categories of adherence to penicillin. The outputs are shown in Tables 5.31 a-c below.
Table 5.31a. Descriptive statistics for self-perceived stigma at different three levels of adherence to penicillin

<table>
<thead>
<tr>
<th>Adherence to penicillin</th>
<th>N</th>
<th>Mean</th>
<th>Std. Deviation</th>
<th>Std. Error</th>
<th>95% Confidence Interval for Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>Often forgot</td>
<td>21</td>
<td>4.80</td>
<td>2.18</td>
<td>0.47</td>
<td>3.81 to 5.80</td>
</tr>
<tr>
<td>Sometimes forgot</td>
<td>31</td>
<td>3.90</td>
<td>1.83</td>
<td>0.32</td>
<td>3.23 to 4.57</td>
</tr>
<tr>
<td>Usually remembers</td>
<td>27</td>
<td>3.62</td>
<td>1.49</td>
<td>0.28</td>
<td>3.03 to 4.22</td>
</tr>
<tr>
<td>Total</td>
<td>79</td>
<td>4.05</td>
<td>1.86</td>
<td>0.021</td>
<td>3.63 to 4.46</td>
</tr>
</tbody>
</table>

Table 5.31b. Test of Homogeneity of Variances

<table>
<thead>
<tr>
<th>Levene Statistic</th>
<th>Df1</th>
<th>Df2</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.42</td>
<td>2</td>
<td>76</td>
<td>.096</td>
</tr>
</tbody>
</table>

Table 5.31c. ANOVA

<table>
<thead>
<tr>
<th></th>
<th>Sum of Squares</th>
<th>Df</th>
<th>Mean Square</th>
<th>F</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Between Groups</td>
<td>17.55</td>
<td>2</td>
<td>8.77</td>
<td>2.62</td>
<td>.08</td>
</tr>
<tr>
<td>Within Groups</td>
<td>254.24</td>
<td>76</td>
<td>3.34</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>271.79</td>
<td>78</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The ANOVA showed that young people who often forgot to take their penicillin scored higher on the stigma scale than those who sometimes forgot. The latter group also scored higher than those who usually remember to take their penicillin. However the differences in mean did not reach statistical significance.

Summary of associations between stigma and illness variables

In summary, the results of this section of the analysis show that there are statistically significant associations between self-perceived stigma and presence of leg ulcer, and three indices of severity (pain intensity, frequency of ward admissions and frequency
of A&E attendance). No significant associations were found with keeping of clinic appointments or medication adherence.

5.2.3. Associations between stigma and emotional and behavioural adjustment

Table 5.32. Self perceived stigma and scores on the Strengths and Difficulties Questionnaire

<table>
<thead>
<tr>
<th>Perceived Stigma</th>
<th>No or low perceived stigma</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Difficulties Score</td>
<td>(Mean, SD)</td>
<td></td>
</tr>
<tr>
<td>(n=13)</td>
<td>16.1 (6.5)</td>
<td>t = 4.3, df = 76</td>
</tr>
<tr>
<td>(n=65)</td>
<td>9.7 (4.5)</td>
<td>p = 0.0001</td>
</tr>
<tr>
<td>Conduct subscale</td>
<td>(n=14)</td>
<td>t = 0.4, df = 86</td>
</tr>
<tr>
<td>2.4 (2.0)</td>
<td>2.1 (1.5)</td>
<td>p = 0.7</td>
</tr>
<tr>
<td>Emotional subscale</td>
<td>(n=14)</td>
<td>t = 3.5, df = 85</td>
</tr>
<tr>
<td>5.6 (2.2)</td>
<td>3.4 (2.1)</td>
<td>p = 0.003</td>
</tr>
<tr>
<td>Peer problems subscale</td>
<td>(n=13)</td>
<td>t = 5.6, df = 85</td>
</tr>
<tr>
<td>4.0 (2.2)</td>
<td>1.3 (1.5)</td>
<td>p = 0.0001</td>
</tr>
<tr>
<td>Hyperactivity subscale</td>
<td>(n=14)</td>
<td>t = 0.9, df = 83</td>
</tr>
<tr>
<td>3.7 (2.2)</td>
<td>3.2 (1.9)</td>
<td>p = 0.3</td>
</tr>
<tr>
<td>Prosocial subscale</td>
<td>(n=13)</td>
<td>t = 0.6, df = 84</td>
</tr>
<tr>
<td>7.9 (2.1)</td>
<td>7.6 (1.9)</td>
<td>p = 0.6</td>
</tr>
</tbody>
</table>

Compared with those with no or low self-perceived stigma, young people classified as having self-perceived stigma had statistically significantly higher Total Difficulties scores as well as higher scores on the emotional and peer problems subscales (Table 5.32). The group means for each of these three subscales of the SDQ differed by more than one standard deviation.

Table 5.33 Self-perceived stigma and depression scores

<table>
<thead>
<tr>
<th>Perceived Stigma</th>
<th>No or low perceived stigma</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Short MFQ score</td>
<td>(n = 13)</td>
<td>(n = 75)</td>
</tr>
<tr>
<td>(Mean, SD)</td>
<td>7.6 (4.7)</td>
<td>3.9 (3.6)</td>
</tr>
<tr>
<td></td>
<td>t = 3.3</td>
<td>df = 86</td>
</tr>
<tr>
<td></td>
<td>p = 0.001</td>
<td></td>
</tr>
</tbody>
</table>
Table 5.33 shows that young people classified as having perceived stigma had statistically significant more depressive symptoms than those with no or low self-perceived stigma. In fact, the mean depressive symptoms for two groups differed by almost one standard deviation.

**Table 5.34 Self-perceived stigma and self esteem**

<table>
<thead>
<tr>
<th>Perceived Stigma (n = 11)</th>
<th>No or low perceived stigma (n = 69)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rosenberg Self Esteem Rating Scale (Mean, SD)</td>
<td>Rosenberg Self Esteem Rating Scale (Mean, SD)</td>
<td>t = 4.3 df=78 p = 0.07</td>
</tr>
<tr>
<td>29.4 (5.6)</td>
<td>32.3 (4.8)</td>
<td></td>
</tr>
</tbody>
</table>

Table 5.34 shows that young people with self perceived stigma had lower self esteem than those with no or low stigma. However, the mean difference between the two groups was not statistically significant. On the other hand, when the dimensional measure of self-perceived stigma was correlated with self esteem scores (data not shown), the coefficient of correlation was statistically significant ($r = -0.29, p = 0.01$) showing that young people with self-perceived stigma scored less on the self esteem scale. The fact that the T-test in Table 5.34 was not statistically significant (but the correlation coefficient was significant) demonstrates the loss of statistical power associated with categorisation of continuous dimensional measures.

**Table 5.35 Self-perceived stigma and family function**

<table>
<thead>
<tr>
<th>Perceived Stigma (n = 12)</th>
<th>No or low perceived stigma (n = 67)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family Assessment Device (raw scores) Mean (SD)</td>
<td>Family Assessment Device (raw scores) Mean (SD)</td>
<td>t=0.5 df=77 p=0.6</td>
</tr>
<tr>
<td>21.4 (5.9)</td>
<td>20.6 (4.9)</td>
<td></td>
</tr>
</tbody>
</table>

Young people with self-perceived stigma did not differ significantly in their family function compared with those without perceived stigma (Table 5.35).
Table 5.36 Self–perceived stigma and attitude toward illness

<table>
<thead>
<tr>
<th>Child Attitude toward Illness Scale (raw scores) Mean (SD)</th>
<th>Perceived Stigma (n = 13)</th>
<th>No or low perceived stigma (n = 75)</th>
<th>Test and p value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>35.1 (10.6)</td>
<td>41.3 (10.3)</td>
<td>t=-2.0</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>df=86</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>p=0.049</td>
</tr>
</tbody>
</table>

Young people with self-perceived stigma had statistically significant poorer attitude towards SCD than peers without self-perceived stigma (Table 5.36).

**Summary of associations between stigma and emotional and behavioural adjustment**

In summary, the results of this section of the analysis show that in unadjusted bivariate comparisons, young people classified as having self-perceived stigma had significantly worse scores than those not so classified on most of the measures of emotional and behavioural adjustment. In some cases, the two groups differed by more than one standard deviation. The group with self-perceived stigma had statistically significantly higher Total Difficulties scores as well as higher scores on the emotional and peer problems subscales of the SDQ. Also, they had significantly more depressive symptoms, lower self esteem, and poorer attitude towards SCD. The only measure with no significant difference between the two groups was family function. Multivariate analysis will be employed later to explore if these significant associations between stigma and emotional and behavioural variables remain after controlling for confounding variables.
Chapter 5: Section III

5.3. Unadjusted bivariate comparisons between respondents recruited from Sickle cell society and those recruited from haematology clinics

Table 5.37. Age of respondents recruited from Sickle Cell Society compared with those recruited from haematology clinics

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=24)</th>
<th>Haematology clinics (n=64)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age. Mean (SD)</td>
<td>14.2 (2.4)</td>
<td>14.2 (2.1)</td>
<td>t=0.13</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>df=86</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>p=0.90</td>
</tr>
</tbody>
</table>

Table 5.37 shows no difference in mean age between subjects recruited from the Sickle Cell Society or haematology clinics.

Table 5.38. Gender of respondents recruited from Sickle Cell Society compared with those recruited from haematology clinics

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=24)</th>
<th>Haematology clinics (n=66)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male N(%)</td>
<td>9 (19.6)</td>
<td>37 (80.4)</td>
<td>χ=2.4</td>
</tr>
<tr>
<td>Female N(%)</td>
<td>15 (34.1)</td>
<td>29 (65.9)</td>
<td>df=1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>p=0.15</td>
</tr>
</tbody>
</table>

Table 5.38 shows more males were recruited from clinics and more girls from the Sickle Cell Society but the percentage differences are not statistically significant.
Table 5.39. Association between Socioeconomic status (OPCS) and whether respondents were recruited through a haematology clinic or Sickle Cell Society

To test this association, the six categories of the OPCS were collapsed into three categories by combining categories 1 and 2, 3 and 4, 5 and 6 in order to reduce the number of cells with small or no numbers. As shown in Table 5.39 below, there was no statistically significant association between these OPCS-SES categories and source of recruitment.

<table>
<thead>
<tr>
<th>OPCS-SES categories</th>
<th>Sickle cell society (n=20) N(%)</th>
<th>Haematology clinics (n=60) N(%)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>1&amp;2</td>
<td>3 (15)</td>
<td>22 (36.7)</td>
<td>$\chi = 3.8$ df=2 p=0.15</td>
</tr>
<tr>
<td>3&amp;4</td>
<td>13 (65)</td>
<td>32 (53.3)</td>
<td></td>
</tr>
<tr>
<td>5&amp;6</td>
<td>4 (20)</td>
<td>6 (10)</td>
<td></td>
</tr>
</tbody>
</table>

However, given that the six OPCS-SES categories represent an ordinal scale, the association was also tested by calculating the Spearman rho correlation coefficient between the full six-category OPCS-SES (coded as in Table 5.5) and the two categories of source of recruitment (coded as haematology clinics = 1, Sickle Cell Society = 2). The Spearman rho correlation coefficient (n=80, r = 0.3) was statistically significant (p=0.006). The coding frame indicates that respondents recruited from haematology clinics were more likely to come from households headed by someone in a higher professional status compared with the children recruited from Sickle Cell Society.

Table 5.40 Comparison between respondents recruited from Sickle Cell Society and those recruited from haematology clinics on whether or not they were born in the UK

<table>
<thead>
<tr>
<th>Born in the UK</th>
<th>Sickle cell society (n=24)</th>
<th>Haematology clinics (n=67)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes N(%)</td>
<td>15 (21.7)</td>
<td>54 (78.3)</td>
<td>$\chi=3.1$ df=1 p=0.10</td>
</tr>
<tr>
<td>No N(%)</td>
<td>9 (40.9)</td>
<td>13 (59.1)</td>
<td></td>
</tr>
</tbody>
</table>
Table 5.40 shows that a higher percentage of the subjects born in the UK were recruited from the haematology clinics compared with respondents who were not born in the UK. However, the difference was not significant.

**Table 5.41. Self-perceived stigma among respondents recruited through haematology clinics or Sickle Cell Society**

Given that the Sickle Cell Society is an advocacy organisation, people with SCD who are members could be a self select group that have a more positive attitude toward SCD and be less likely to perceive stigmatisation. This premise justifies exploring whether fewer children from among those recruited through the Sickle Cell Society were classified as having self perceived stigma compared with those recruited from haematology clinics.

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=24)</th>
<th>Haematology clinics (n=67)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Self perceived stigma</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes N(%)</td>
<td>3 (21.4)</td>
<td>11 (78.6)</td>
<td>Fisher’s Exact p=0.75</td>
</tr>
<tr>
<td>No N(%)</td>
<td>21 (27.3)</td>
<td>56 (72.7)</td>
<td></td>
</tr>
</tbody>
</table>

Table 5.41 above shows that there is no significant association between source of recruitment and classification as having self-perceived stigma.

**Table 5.42. Association between stigma and whether respondents completed questionnaire in clinic or at home**

Theoretically, young people completing questionnaires in clinic may be more likely to report self-perceived stigma as their attendance to the clinic while their peers are at school or engaging in other activities could become a symbolic reminder of the disruptive effect of having SCD.
<table>
<thead>
<tr>
<th>Where questionnaire was completed</th>
<th>Perceived Stigma (n=13)</th>
<th>No or low perceived stigma (n=74)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Homes N (%)</td>
<td>6 (11.8)</td>
<td>45 (88.2)</td>
<td>$\chi^2 = 1.0$</td>
</tr>
<tr>
<td>Clinic N (%)</td>
<td>7 (19.4)</td>
<td>29 (80.6)</td>
<td>df=1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>$p=0.4$</td>
</tr>
</tbody>
</table>

Table 5.42 shows no significant association between stigma and whether respondents completed questionnaires in haematology clinics or at home.

**Table 5.43. Comparison of depressive symptoms between respondents from Sickle Cell Society and haematology clinics**

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=23)</th>
<th>Haematology clinics (n=65)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Depressive symptoms (SMFQ) Mean (SD)</td>
<td>3.4 (2.8)</td>
<td>4.8 (4.2)</td>
<td>$t=1.5$</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>df=86</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>$p=0.08$</td>
</tr>
</tbody>
</table>

Table 5.43 shows that the children recruited from haematology clinics scored higher on the SMFQ than those recruited from Sickle Cell Society but the difference was not statistically significant.

**Table 5.44. Comparison of Total Difficulties Scale of the SDQ between respondents from Sickle Cell Society and haematology clinics**

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=23)</th>
<th>Haematology clinics (n=55)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Difficulties Scale of the SDQ. Mean (SD)</td>
<td>11.0 (5.1)</td>
<td>10.6 (5.5)</td>
<td>$t=-0.26$</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>df=76</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>$p=0.79$</td>
</tr>
</tbody>
</table>

Table 5.44 shows that the children recruited from haematology clinics did not differ in their score on the Total Difficulties scale of the SDQ compared with those recruited from
Sickle Cell Society. Also they did not differ significantly on any of the five SDQ subscales (data not shown).

Table 5.45. Comparison of self esteem between respondents from Sickle Cell Society and haematology clinics

<table>
<thead>
<tr>
<th>Rosenberg Self Esteem Scale Mean (SD)</th>
<th>Sickle cell society (n=23)</th>
<th>Haematology clinics (n=57)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>33.7 (3.7)</td>
<td>31.2 (5.3)</td>
<td>$t=-2.1$ df=78 $p=0.04$</td>
</tr>
</tbody>
</table>

Table 5.45 shows that the children recruited from haematology clinics scored lower on the Rosenberg Self Esteem scale compared with those recruited from Sickle Cell Society and the difference is statistically significant ($p=0.04$).

Table 5.46. Comparison of Attitude towards illness between respondents from Sickle Cell Society and haematology clinics

<table>
<thead>
<tr>
<th>Attitude toward illness (CATIS score) Mean (SD)</th>
<th>Sickle cell society (n=23)</th>
<th>Haematology clinics (n=65)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>40.6 (11.5)</td>
<td>40.3 (10.3)</td>
<td>$t=-0.12$ df=86 $p=0.91$</td>
</tr>
</tbody>
</table>

Table 5.46 shows that the children recruited from haematology clinics did not differ in their attitude towards SCD compared with those recruited from Sickle Cell Society.
Table 5.47. Receipt of counselling among respondents recruited from Sickle Cell Society compared with those recruited from haematology clinics

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=24)</th>
<th>Haematology clinics (n=76)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Counselling</td>
<td></td>
<td></td>
<td>Fisher’s Exact p=1.0</td>
</tr>
<tr>
<td>Yes N(%)</td>
<td>2 (20.0)</td>
<td>8 (80.0)</td>
<td></td>
</tr>
<tr>
<td>No N(%)</td>
<td>22 (27.5)</td>
<td>58 (72.5)</td>
<td></td>
</tr>
</tbody>
</table>

Table 5.47 shows no significant difference in the percentage of respondents recruited from Sickle Cell Society who were receiving counselling compared with those recruited from haematology clinics.

Table 5.48. Comparison of family function between respondents from Sickle Cell Society and haematology clinics

<table>
<thead>
<tr>
<th></th>
<th>Sickle cell society (n=22)</th>
<th>Haematology clinics (n=57)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family Assessment Device (FAD) – General Function Scale (raw score) Mean (SD)</td>
<td>20.5 (4.5)</td>
<td>20.9 (5.3)</td>
<td>t=0.28 df=77 p=0.78</td>
</tr>
</tbody>
</table>

Table 5.48 shows that the children recruited from haematology clinics did not differ in family function compared with those recruited from Sickle Cell Society.

Table 5.49. Association between frequency of admission and whether respondents were recruited through a clinic or Sickle Cell Society

<table>
<thead>
<tr>
<th>Frequency of ward admission in past year</th>
<th>Sickle cell society (n=23)</th>
<th>Haematology clinics (n=65)</th>
<th>Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>N(%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No admission</td>
<td>11 (47.8)</td>
<td>23 (35.4)</td>
<td>χ = 4.7 df=5 p=0.5</td>
</tr>
<tr>
<td>Once</td>
<td>3 (13.0)</td>
<td>12 (18.5)</td>
<td></td>
</tr>
<tr>
<td>2-4 times</td>
<td>4 (17.4)</td>
<td>21 (32.3)</td>
<td></td>
</tr>
<tr>
<td>5-6 times</td>
<td>3 (13.0)</td>
<td>5 (7.7)</td>
<td></td>
</tr>
<tr>
<td>7-10</td>
<td>0</td>
<td>2 (3.1)</td>
<td></td>
</tr>
<tr>
<td>&gt;10 times</td>
<td>2 (8.7)</td>
<td>2 (3.1)</td>
<td></td>
</tr>
</tbody>
</table>
Table 5.49 shows no statistically significant association in frequency of ward admission (a measure of severity) between respondents recruited through Sickle Cell Society and those recruited from haematology clinics.

Summary of unadjusted bivariate comparisons between respondents recruited from Sickle cell society and those recruited from haematology clinics

In summary, the results of the analysis for this section show that the subjects recruited from the Sickle Cell Society differed only on two items (OPCS-SES status and Self-esteem) from those recruited from haematology clinics. Respondents recruited from the Sickle Cell Society were more likely to come from families with lower SES. However, they were more likely to have higher self-esteem. The two groups did not differ significantly on age, gender, whether or not born in the UK, levels of self-perceived stigma, whether they completed the questionnaire at home or in the clinic, depressive symptoms, Total Difficulties scale or subscales of the SDQ, receipt of counselling, family function, or frequency of ward admissions (measure of severity).
Chapter 5. Section IV.

5.4. Multivariate analysis and hypotheses testing

This section describes the procedure and outcome of the tests of the four hypotheses proposed for this thesis. The analytical strategies employed are multivariate, which allow for confounders to be controlled and for interactions to be explored.

The study hypotheses to be tested are:

1. Measures of disruptiveness (e.g. frequency of admissions) and visibility (e.g. presence of leg ulcer) will significantly and independently predict levels of self perceived stigma.

2. Self perceived stigma will significantly and independently predict scores on the Total difficulties scale of the SDQ.

3. Self perceived stigma will significantly and independently predict Depressive symptoms measured by SMFQ

4. Self perceived stigma will significantly and independently predict Self Esteem measured by Rosenberg Scale

Incidentally, the process of exploring the above primary hypotheses gave opportunities to explore additional associations between each dependent variable and other psychological, illness, and social predictor variables over and above self-perceived stigma.
First hypothesis

Measures of disruptiveness (e.g. frequency of admissions) and visibility (e.g. presence of leg ulcer) will significantly and independently predict levels of self perceived stigma.

This hypothesis is based on stigma theory, which suggests that certain attributes of a condition (referred to as stigma dimensions) predict whether the condition is likely to be stigmatising. The common dimensions described in the literature are; Visibility, Threat or Peril, Chronicity, Responsibility, and Disruptiveness (Katz 1981, Jones et al 1984). Stigma dimensions were explored in detail in Chapter 2. The stigma dimensions considered measurable for this study on SCD are visibility and disruptiveness. The hypothesis is that if stigma theory is applicable to SCD, then measures of visibility and disruptiveness would predict self-perceived stigma.

The first step in exploring this hypothesis is to establish if measures of visibility and disruptiveness have statistically significant associations with self-perceived stigma.

Measures of visibility

In this study, Visibility was measured with presence of jaundice and presence of leg ulcer. As shown in Table 5.27, jaundice was not significantly associated with self perceived stigma. Thus this measure of visibility was not considered further in the analysis. However presence of leg ulcer had a statistically significant association with self-perceived stigma (Table 5.28) in bivariate analysis. The latter therefore shows preliminary support for this hypothesis. However, multivariate analysis will be used next to ascertain if this association is independent of other explanatory variables.

Measures of disruptiveness

Disruptiveness was assessed in this study with five variables: frequency of ward admissions, frequency of admissions to Accident and Emergency department, frequency of school absence, frequency of experience of pain, and intensity of pain. A composite measure “severity” was also created. As shown in Table 5.50 below, only the combined scale “severity”, pain intensity, frequency of ward admission and frequency of A&E admissions correlated significantly with stigma. It was not
appropriate to consider including all the three measures of disruptiveness in multivariate analysis for several reasons (Field 2005). First is to avoid problems of multicollinearity. Secondly, to avoid having redundant independent variables, as each extra covariate reduces the degree of freedom for error by one (the goal being to obtain maximum adjustment for the dependent variable with minimum loss of degrees of freedom for the error term).

http://www.psych.umn.edu/courses/spring05/federicoc/psy8815/lectures/stats_lecture9.pdf (accessed 03/01/2010). Third reason is to reduce the total number of predictors. Although many authors suggest that a ratio of 10 subjects per predictor in a regression model is adequate (Pallant 2007), Field (2005) recommends a more stringent ratio. He suggests a ratio based on the formula 50 + 8K, where K is the number of variables. Given that the sample size of this study is 93, this more stringent ratio will allow the inclusion of a maximum of five predictor variables (50 + 8x5) = 90. Taking account of missing data, four variables per model would be the most stringent. However, I took a pragmatic approach in the following multivariate analyses by using varying stringency of number of variables while keeping to a total of 4 - 7 predictors per model.

Bearing the above considerations in mind, I decided to choose one of the three measures of disruptiveness for the multivariate analysis. Of the four variables that correlated significantly with self-perceived stigma, frequency of ward admissions was chosen as the best measure of disruptiveness as it correlated strongest with self-perceived stigma and also with other measures of disruptiveness (Table 5.50).
Thus, this hypothesis will be tested using presence of leg ulcer as the measure of visibility and frequency of ward admissions as the measure of disruptiveness. The Hypothesis can therefore be re-stated as “Presence of leg ulcer (visibility) and frequency of ward admissions (disruptiveness) will significantly and independently predict levels of self perceived stigma”.

Using the **dimensional measure of stigma as the dependent variable**, this hypothesis could be tested with one of three multivariate techniques: hierarchical linear regression, standard linear regression, or Analysis of Covariance.

In practice I availed the data the opportunity to be explored with all three techniques, which gave similar results. I have presented the analysis using hierarchical linear regression as I considered it theoretically more appropriate for this hypothesis.

<table>
<thead>
<tr>
<th></th>
<th>School absence</th>
<th>Pain frequency</th>
<th>Pain Intensity</th>
<th>Frequency of ward admissions</th>
<th>Frequency of A&amp;E admissions</th>
<th>Stigma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain frequency</td>
<td>r</td>
<td>.14</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>72</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pain intensity</td>
<td>r</td>
<td>.18</td>
<td>.09</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>80</td>
<td>74</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency of ward admissions</td>
<td>r</td>
<td>.27*</td>
<td>.28*</td>
<td>.37**</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>83</td>
<td>73</td>
<td>82</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency of A&amp;E admissions</td>
<td>r</td>
<td>.16</td>
<td>.18</td>
<td>.38**</td>
<td>.54**</td>
<td></td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>83</td>
<td>73</td>
<td>81</td>
<td>85</td>
<td></td>
</tr>
<tr>
<td>Stigma (dimensional)</td>
<td>r</td>
<td>.09</td>
<td>.04</td>
<td>.30**</td>
<td>.34**</td>
<td>.31**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>86</td>
<td>74</td>
<td>83</td>
<td>86</td>
<td>85</td>
</tr>
<tr>
<td>Severity scale</td>
<td>r</td>
<td>.17</td>
<td>.44**</td>
<td>.56**</td>
<td>.66**</td>
<td>.63**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>66</td>
<td>69</td>
<td>69</td>
<td>69</td>
<td>69</td>
</tr>
</tbody>
</table>

$r = $ Spearman rho

** Correlation is significant at the 0.01 level (2-tailed).
* Correlation is significant at the 0.05 level (2-tailed).
Hierarchical linear regression is appropriate where there is a theoretically valid reason for deliberately entering predictor variables in separate blocks in the regression model. As a general rule, known predictors or confounders are entered first, and the new predictors entered last.

Thus hierarchical multiple regression was used to test the ability of two stigma dimensions (presence of leg ulcer and frequency of admissions to predict) to predict levels of self-perceived stigma (dimensional stigma scale) after controlling for potential confounders (Age and Gender).

Age and Gender were entered together in Step 1, and they explained 6% of the variance in self-perceived stigma. Presence of leg ulcer was entered alone in Step 2 because visibility has more saliency, proximity, and immediacy in eliciting or inducing self-perceived stigma than disruptiveness. The entry of Leg ulcer alone in Step 2 increased the total variance explained by the model to 15% $F(3, 81) = 4.7$, $p = 0.004$. Thus leg ulcer alone explained an additional 8.4% of the variance in self-perceived stigma after controlling for Age and Gender, $R$ squared change = 0.085, $F$ change $(1, 81) = 8.1$, $p = 0.006$. Frequency of ward admission was entered alone in Step 3. The total variance explained by the model as a whole was 19% with Frequency of admission alone explaining an additional 4.3% of the variance in self-perceived stigma after controlling for Age, Gender, and leg ulcer, $R$ squared change = 0.43, $F$ change $(1, 80) = 4.3$, $p = 0.42$. In the final model, only the two stigma dimensions (leg ulcer and admissions) were statistically significant, with leg ulcer recording a slightly higher beta value ($\beta = -0.23$, $p = 0.031$) than frequency of admissions ($\beta = 0.22$, $p = 0.042$). The regression outputs are shown in Tables 5.51 (a,b,c) below. Thus these findings provide support for the hypothesis “Presence of leg ulcer (visibility) and frequency of ward admissions (disruptiveness) will significantly and independently predict levels of self perceived stigma”.

The analysis was checked to ensure that the assumptions for multiple regression: normality, linearity, multicollinearity and homoscedasticity (Field 2005) were met. For example, the Durbin-Watson statistic of 1.9 in Table 5.51a indicates no autocorrelation in the residuals (values close to 2.0 are considered normal). Tolerance
is the percentage of the variance in a given predictor that cannot be explained by the other predictors. The Tolerance values between 0.8-0.99 in Table 5.51c are considered very good (meaning that less than 20% of the variance in the predictors are explained by other predictors. Tolerance of less than 0.1 suggests multicollinearity). The Variance Inflation Factors (VIF) in the range of 1.0-1.1 in Table 5.51c are also good because only values greater than 2 are considered abnormal. Finally, the Normal P-P plot of residuals (Figure 5.2) follow a 45° angle indicating that the assumption of normality was not violated.

Table 5.51a. Model Summary

<table>
<thead>
<tr>
<th>Mode</th>
<th>R</th>
<th>R Square</th>
<th>Adjusted R Square</th>
<th>Std. Error of the Estimate</th>
<th>R Square Change</th>
<th>F Change</th>
<th>df1</th>
<th>df2</th>
<th>Sig. F Change</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>.25(a)</td>
<td>.06</td>
<td>.04</td>
<td>1.74</td>
<td>.06</td>
<td>2.81</td>
<td>2</td>
<td>82</td>
<td>.066</td>
</tr>
<tr>
<td>2</td>
<td>.38(b)</td>
<td>.15</td>
<td>.11</td>
<td>1.67</td>
<td>.08</td>
<td>8.06</td>
<td>1</td>
<td>81</td>
<td>.006</td>
</tr>
<tr>
<td>3</td>
<td>.43(c)</td>
<td>.19</td>
<td>.15</td>
<td>1.64</td>
<td>.04</td>
<td>4.26</td>
<td>1</td>
<td>80</td>
<td>.042</td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Gender, Age
b Predictors: (Constant), Gender, Age, Ulcer
c Predictors: (Constant), Gender, Age, Ulcer, Frequency of ward admissions
d Dependent Variable: Self-perceived stigma (dimensional measure)

Table 5.51b. Model Summary

<table>
<thead>
<tr>
<th>Model</th>
<th>Sum of Squares</th>
<th>Df</th>
<th>Mean Square</th>
<th>F</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Regression</td>
<td>17.15</td>
<td>2</td>
<td>8.57</td>
<td>2.81</td>
</tr>
<tr>
<td>Residual</td>
<td>249.94</td>
<td>82</td>
<td>3.04</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>267.10</td>
<td>84</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Regression</td>
<td>39.79</td>
<td>3</td>
<td>13.26</td>
<td>4.72</td>
</tr>
<tr>
<td>Residual</td>
<td>227.30</td>
<td>81</td>
<td>2.80</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>267.10</td>
<td>84</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Regression</td>
<td>51.30</td>
<td>4</td>
<td>12.82</td>
<td>4.75</td>
</tr>
<tr>
<td>Residual</td>
<td>215.79</td>
<td>80</td>
<td>2.69</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>267.10</td>
<td>84</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Gender, Age
b Predictors: (Constant), Gender, Age, Ulcer
c Predictors: (Constant), Gender, Age, Ulcer, Frequency of ward admissions
d Dependent Variable: Self-perceived stigma (dimensional measure)
<table>
<thead>
<tr>
<th>Model</th>
<th>Unstandardised Coefficients</th>
<th>Standardised Coefficients</th>
<th>Collinearity statistics</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>B</td>
<td>Std. Error</td>
<td>Beta</td>
</tr>
<tr>
<td>1</td>
<td>(Constant)</td>
<td>4.28</td>
<td>1.32</td>
</tr>
<tr>
<td></td>
<td>Age</td>
<td>-.11</td>
<td>.09</td>
</tr>
<tr>
<td></td>
<td>Gender</td>
<td>.85</td>
<td>.38</td>
</tr>
<tr>
<td>2</td>
<td>(Constant)</td>
<td>8.58</td>
<td>1.97</td>
</tr>
<tr>
<td></td>
<td>Age</td>
<td>-.11</td>
<td>.08</td>
</tr>
<tr>
<td></td>
<td>Gender</td>
<td>.90</td>
<td>.37</td>
</tr>
<tr>
<td></td>
<td>Ulcer</td>
<td>-2.24</td>
<td>.79</td>
</tr>
<tr>
<td>3</td>
<td>(Constant)</td>
<td>6.99</td>
<td>2.08</td>
</tr>
<tr>
<td></td>
<td>Age</td>
<td>-.09</td>
<td>.08</td>
</tr>
<tr>
<td></td>
<td>Gender</td>
<td>.72</td>
<td>.37</td>
</tr>
<tr>
<td></td>
<td>Ulcer</td>
<td>-1.77</td>
<td>.80</td>
</tr>
<tr>
<td></td>
<td>Frequency of admission</td>
<td>.28</td>
<td>.14</td>
</tr>
</tbody>
</table>

a Dependent Variable: Self-perceived stigma (dimensional measure)
Figure 5.2

Normal P-P Plot of Regression Standardized Residual

Dependent Variable: STIGMAD
Second hypothesis

*Self perceived stigma will significantly and independently predict Total difficulties score on the Strengths and Difficulties Questionnaire (SDQ)*

This hypothesis explores whether self-perceived stigma contributes uniquely to psychological difficulty (assessed with The Total Difficulties Scale of the SDQ) in young people with SCD. The Total Difficulties Scale of the SDQ is the sum of four of the five subscales of the SDQ (emotional, conduct, peer problems, and hyperactivity subscales).

In the ensuing analysis **the Total Difficulties subscale of the SDQ is considered the dependent variable.** The first step in exploring this hypothesis is to establish if The Total Difficulties subscales of the SDQ is significantly associated with self-perceived stigma. This association was established in Section II (Table 5.32) and in Table 5.52 below. The next step is to identify other independent variables significantly associated with Total Difficulties Scale of the SDQ that could confound the relationship with self-perceived stigma. Table 5.52 shows a correlation matrix of the Total Difficulties scale of the SDQ and potential independent variables. Self-perceived stigma was included in the correlation matrix (Table 5.52) to ease the identification of potential confounders, which are variables that correlate with both Total Difficulties Scale of SDQ and self-perceived stigma. Table 5.52 shows that Frequency of ward admissions and attitude towards SCD meet this criterion for confounding; hence they will be included as covariates in the multivariate analysis. Given that leg ulcer had shown significant association with stigma in bivariate analysis (Table 5.28), and that previous studies have shown associations between, age, gender, family function and psychological difficulty in children, these variables were also included as covariates. Although depressive symptoms and self esteem correlated strongly with SDQ (data not shown), the former two variables were not considered for inclusion because they measure overlapping aspects of psychological function or wellbeing. Overall, the number of predictor variables included in this regression analysis (i.e. 7) is within the range of stringency described earlier for the sample size of this study.
Table 5.52. Bivariate correlation coefficients between SDQ and potential predictor variables.

<table>
<thead>
<tr>
<th></th>
<th>SDQ</th>
<th>Stigma (dimensional)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>R</td>
<td></td>
</tr>
<tr>
<td>SDQ</td>
<td>R</td>
<td>.52(**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>76</td>
</tr>
<tr>
<td>Stigma (dimensional)</td>
<td>R</td>
<td>.51**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>76</td>
</tr>
<tr>
<td>Age</td>
<td>R</td>
<td>-.15</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>76</td>
</tr>
<tr>
<td>Gender</td>
<td>r*</td>
<td>.04</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>76</td>
</tr>
<tr>
<td>OPCS</td>
<td>r*</td>
<td>.21</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>77</td>
</tr>
<tr>
<td>Born in UK</td>
<td>r*</td>
<td>.05</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>70</td>
</tr>
<tr>
<td>Frequency of ward admissions</td>
<td>r*</td>
<td>.30**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>76</td>
</tr>
<tr>
<td>Ulcer</td>
<td>r*</td>
<td>-.14</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>76</td>
</tr>
<tr>
<td>Receipt of counselling</td>
<td>r*</td>
<td>-.14</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>78</td>
</tr>
<tr>
<td>Family Function</td>
<td>R</td>
<td>.341(**)</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>70</td>
</tr>
<tr>
<td>Attitude towards Illness</td>
<td>R</td>
<td>-.542(**)</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>75</td>
</tr>
</tbody>
</table>

r = Pearson correlation coefficients, r* = Spearman rho correlation coefficients
** Correlation is significant at the 0.01 level (2-tailed).
* Correlation is significant at the 0.05 level (2-tailed).
Coding: Gender- male = 1, female = 2; Born in UK – yes = 1, no = 2, leg ulcer – yes = 1, no = 2

This hypothesis could be tested with either multiple regression or ANCOVA. The data was explored with both methods and similar results were obtained. The ANCOVA showed no significant interactions; hence the output from multiple regression is presented here. Standard “simultaneous” regression method was used as there was no theoretical justification to adopt either a hierarchical or a stepwise method. The results are shown in Tables 5.53(a,b,c).
Table 5.53a. Model Summary

<table>
<thead>
<tr>
<th>Model</th>
<th>R</th>
<th>R Square</th>
<th>Adjusted R Square</th>
<th>Std. Error of the Estimate</th>
<th>Durbin-Watson</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>.69(a)</td>
<td>.48</td>
<td>.42</td>
<td>4.07</td>
<td>2.23</td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Family function, Age, Leg ulcer, Frequency of ward admissions, Gender, Stigma, Attitude toward illness
b Dependent Variable: SDQ

Table 5.53b. ANOVA

<table>
<thead>
<tr>
<th>Model</th>
<th>Sum of Squares</th>
<th>Df</th>
<th>Mean Square</th>
<th>F</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Regression</td>
<td>947.06</td>
<td>7</td>
<td>135.29</td>
<td>8.15</td>
<td>.000(a)</td>
</tr>
<tr>
<td>Residual</td>
<td>1028.58</td>
<td>62</td>
<td>16.59</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>1975.65</td>
<td>69</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Family function, Age, Leg ulcer, Frequency of ward admissions, Gender, Stigma, Attitude toward illness
b Dependent Variable: SDQ

Table 5.53c. Coefficients

<table>
<thead>
<tr>
<th>Mode 1</th>
<th>Unstandardised Coefficients</th>
<th>Standardised Coefficients</th>
<th>Collinearity Statistics</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>B</td>
<td>Std. Error</td>
<td>Beta</td>
</tr>
<tr>
<td>1</td>
<td>(Constant)</td>
<td>14.02</td>
<td>6.91</td>
</tr>
<tr>
<td></td>
<td>Stigma</td>
<td>.99</td>
<td>.31</td>
</tr>
<tr>
<td></td>
<td>Frequency of ward admission</td>
<td>.47</td>
<td>.40</td>
</tr>
<tr>
<td></td>
<td>Leg ulcer</td>
<td>-.22</td>
<td>2.29</td>
</tr>
<tr>
<td></td>
<td>Attitude towards Illness</td>
<td>-.17</td>
<td>.05</td>
</tr>
<tr>
<td></td>
<td>Age</td>
<td>-.28</td>
<td>.23</td>
</tr>
<tr>
<td></td>
<td>Gender</td>
<td>-.87</td>
<td>1.08</td>
</tr>
<tr>
<td></td>
<td>Family Function</td>
<td>.21</td>
<td>.10</td>
</tr>
</tbody>
</table>

a Dependent Variable: SDQ
In summary, a standard simultaneous multiple regression was used to test the ability of self-perceived stigma to predict scores on the Total Difficulties Scale of the SDQ independent of other potential confounding variables (Age, Gender, leg ulcer, frequency of admissions, family function, and attitude towards SCD).
The model as a whole explained 48% of the variance in the Total Difficulties Scale of the SDQ $F(7, 62) = 8.2, p < 0.001$. Only self-perceived stigma and attitude towards SCD were statistically significant and independent predictors of Total Difficulties Scale of the SDQ. Attitude towards SCD recorded a slightly higher beta value ($\beta = -0.34, p = 0.02$) than self-perceived stigma ($\beta = 0.33, p = 0.02$). Thus these findings provide support for the hypothesis “Self perceived stigma will significantly and independently predict Total difficulties score on the Strengths and Difficulties Questionnaire (SDQ)”.

The analysis was checked to ensure that the assumptions for multiple regression (Field 2005) such as Durbin-Watson statistic of 2.2 in Table 5.53a, which indicates no autocorrelation in the residuals, Tolerance values between 0.77-0.94 in Table 5.53c and Variance Inflation Factors (VIF) in the range of 1.2-1.3 in Table 5.53c. Also, the Normal P-P plot of residuals (Figure 5.3) follow a 45° angle indicating that the assumption of normality was not violated.

**Third hypothesis**

*Self perceived stigma will significantly and independently predict Depressive symptoms as measured by SMFQ*

In addition to psychological difficulty, which was assessed with the SDQ, the study also obtained data on a specific psychopathology (depression). This hypothesis therefore explores whether self-perceived stigma contributes uniquely to depressive symptoms (assessed with the Short Mood and Feelings Questionnaire – SMFQ).

This hypothesis was assessed with a standard multiple regression with the **Short Mood and Feelings Questionnaire score as the dependent variable**. The first step in the analysis is to establish if SMFQ is significantly associated with self-perceived stigma. This association was established in Section II (Table 5.33) and in Table 5.54 below. The next step is to identify other independent variables significantly associated with SMFQ that could confound the relationship with self-perceived stigma. Table 5.54 shows a correlation matrix of SMFQ and potential independent variables. Potential confounders are variables that correlate significantly with both SMFQ and
self-perceived stigma. Table 5.54 shows that attitude towards SCD, and frequency of ward admission met this criterion to be included as covariates in the regression analysis. However, given that gender, birth in UK, and family function correlated significantly with SMFQ (Table 5.54) and leg ulcer showed statistical significance in bivariate comparison with depressive symptoms (data not shown), these four variables were included as additional covariates. Also as previous studies have shown association between age and depression, age was also included as a covariate.

The analysis presented here is standard multiple regression. However, the analysis was repeated with ANCOVA to explore for interactions. No significant interactions were seen and the ANCOVA model was similar to the regression model; hence only outputs from the latter are presented (Tables 5.54a,b,c).
Table 5.54. Correlation matrix between depression, self-perceived stigma and other predictor variables

<table>
<thead>
<tr>
<th></th>
<th>Depression (SMFQ)</th>
<th>Stigma (dimensional)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Depression (SMFQ)</td>
<td>R .39**</td>
<td></td>
</tr>
<tr>
<td>N 86</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stigma (dimensional)</td>
<td>R .39**</td>
<td></td>
</tr>
<tr>
<td>N 86</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td>R .05</td>
<td>.09</td>
</tr>
<tr>
<td>N 85</td>
<td>86</td>
<td></td>
</tr>
<tr>
<td>Gender</td>
<td>R* .32**</td>
<td>.18</td>
</tr>
<tr>
<td>N 87</td>
<td>88</td>
<td></td>
</tr>
<tr>
<td>OPCS-SES</td>
<td>R* .13</td>
<td>.07</td>
</tr>
<tr>
<td>N 77</td>
<td>78</td>
<td></td>
</tr>
<tr>
<td>Sibling has SCD</td>
<td>R* .003</td>
<td>-.04</td>
</tr>
<tr>
<td>N 87</td>
<td>88</td>
<td></td>
</tr>
<tr>
<td>Born in UK</td>
<td>R* .23*</td>
<td>.20</td>
</tr>
<tr>
<td>N 88</td>
<td>89</td>
<td></td>
</tr>
<tr>
<td>Leg ulcer</td>
<td>R* -.19</td>
<td>-.19</td>
</tr>
<tr>
<td>N 86</td>
<td>87</td>
<td></td>
</tr>
<tr>
<td>Family function</td>
<td>R .43**</td>
<td>.21</td>
</tr>
<tr>
<td>N 78</td>
<td>77</td>
<td></td>
</tr>
<tr>
<td>Attitude towards Illness</td>
<td>-.52**</td>
<td>-.33**</td>
</tr>
<tr>
<td>N 85</td>
<td>86</td>
<td></td>
</tr>
<tr>
<td>Frequency of ward admissions</td>
<td>R* .42**</td>
<td>.33**</td>
</tr>
<tr>
<td>N 85</td>
<td>86</td>
<td></td>
</tr>
</tbody>
</table>

r* = Spearman rho correlation coefficient. r = Pearson correlation coefficient

** Correlation is significant at the 0.01 level (2-tailed).

* Correlation is significant at the 0.05 level (2-tailed).

Coding: Gender- male = 1, female = 2; sibling has SCD – yes = 1, no = 2; Born in UK – yes = 1, no = 2; leg ulcer – yes = 1, no = 2

Table 5.55a. Model Summary

<table>
<thead>
<tr>
<th>Model</th>
<th>R</th>
<th>R Square</th>
<th>Adjusted R Square</th>
<th>Std. Error of the Estimate</th>
<th>Durbin-Watson</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>.69(a)</td>
<td>.47</td>
<td>.41</td>
<td>3.04</td>
<td>2.00</td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Frequency of admission, Age, Family function, Born in UK, Leg ulcer, Gender, Stigma, Attitude towards illness,
b Dependent Variable: Depression
Table 5.55b. ANOVA

<table>
<thead>
<tr>
<th>Model</th>
<th>Sum of Squares</th>
<th>df</th>
<th>Mean Square</th>
<th>F</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Regression</td>
<td>559.42</td>
<td>8</td>
<td>69.93</td>
<td>7.54</td>
<td>.000(a)</td>
</tr>
<tr>
<td>Residual</td>
<td>621.46</td>
<td>67</td>
<td>9.28</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>1180.88</td>
<td>75</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Frequency of admission, Age, Family function, Born in UK, Leg ulcer, Gender, Stigma, Attitude towards illness, b Dependent Variable: Depression

Table 5.55c. Coefficients

<table>
<thead>
<tr>
<th>Model</th>
<th>Unstandardised Coefficients</th>
<th>Standardised Coefficients</th>
<th>Collinearity Statistics</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>B</td>
<td>Std. Error</td>
<td>Beta</td>
</tr>
<tr>
<td>1</td>
<td>(Constant)</td>
<td>1.40</td>
<td>5.03</td>
</tr>
<tr>
<td>Gender</td>
<td>.98</td>
<td>.79</td>
<td>.12</td>
</tr>
<tr>
<td>Leg ulcer</td>
<td>-.81</td>
<td>1.64</td>
<td>-.05</td>
</tr>
<tr>
<td>Attitude towards illness</td>
<td>-.11</td>
<td>.04</td>
<td>-.29</td>
</tr>
<tr>
<td>Age</td>
<td>.07</td>
<td>.17</td>
<td>.04</td>
</tr>
<tr>
<td>Born in UK</td>
<td>.03</td>
<td>.89</td>
<td>.003</td>
</tr>
<tr>
<td>Family Function</td>
<td>.19</td>
<td>.08</td>
<td>.24</td>
</tr>
<tr>
<td>Stigma</td>
<td>.28</td>
<td>.22</td>
<td>.13</td>
</tr>
<tr>
<td>Frequency of admission</td>
<td>.71</td>
<td>.29</td>
<td>.24</td>
</tr>
</tbody>
</table>

a Dependent Variable: Depression
In summary, a standard simultaneous multiple regression was used to test the ability of self-perceived stigma to predict depressive symptoms (scores on SMFQ) independent of other potential confounding variables (Age, Gender, leg ulcer, frequency of admissions, family function, birth in or outside UK, and attitude towards SCD).

The model as a whole explained 47% of the variance in SMFQ scores $F(8, 67) = 7.5$, $p < 0.001$. The statistically significant and independent predictors of SMFQ scores
were; attitude towards SCD (beta = -0.30, p = 0.006), family function (beta = 0.24, p = 0.017, and frequency of ward admissions (beta = 0.24, p = 0.019.

Thus these findings **DO NOT** provide support for the hypothesis “*Self perceived stigma will significantly and independently predict Depressive symptoms as measured by SMFQ*”.

The analysis met the assumptions for multiple regression (Field 2005) such as Durbin-Watson statistic of 2.0 in Table 5.55a, which indicates no autocorrelation in the residuals, tolerance values between 0.73-0.92 in Table 5.55c and Variance Inflation Factors (VIF) in the range of 1.1-1.4 in Table 5.55c. Also, the Normal P-P plot of residuals (Figure 5.4) follow a 45º angle indicating that the assumption of normality was not violated.

**Fourth hypothesis**

*Self perceived stigma will significantly and independently predict Self Esteem measured by Rosenberg Scale*

This hypothesis explores whether self-perceived stigma contributes uniquely to self esteem (assessed with Rosenberg Scale).

This hypothesis was assessed with a standard multiple regression with **Self esteem as the dependent variable**. Section II (Table 5.34) and Table 5.56 below show statistically significant association between self esteem and self-perceived stigma. The next step is to identify other independent variables significantly associated with Self esteem that could confound the relationship with self-perceived stigma. Table 5.56 shows a correlation matrix of Self-esteem and potential independent variables. As previously explained, potential confounders are variables that correlate significantly with both Self esteem and self-perceived stigma. Table 5.56 shows that frequency of ward admissions and attitude towards SCD met this criterion to be included as covariates. Gender, family function, receipt of counselling and source of recruitment were included as additional covariates as they correlated significantly with self esteem.
The analysis presented here is standard multiple regression. When the analysis was repeated with ANCOVA, no significant interactions were seen and the ANCOVA model was similar to the regression model; hence only outputs from the latter are presented (Tables 5.57a,b,c).

Table 5.56. Bivariate Correlations between self esteem and potential predictors

<table>
<thead>
<tr>
<th></th>
<th>Self esteem</th>
<th>Stigma (dimensional)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Self esteem</td>
<td>R</td>
<td>-.29**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>78</td>
</tr>
<tr>
<td>Stigma (dimensional)</td>
<td>R</td>
<td>-.29**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>78</td>
</tr>
<tr>
<td>Gender</td>
<td>R*</td>
<td>-.33**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>.18</td>
</tr>
<tr>
<td>OPCS</td>
<td>R*</td>
<td>-.12</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>.07</td>
</tr>
<tr>
<td>Age</td>
<td>R</td>
<td>.02</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>-.09</td>
</tr>
<tr>
<td>Sib has SCD</td>
<td>R*</td>
<td>-.11</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>-.04</td>
</tr>
<tr>
<td>Born in UK</td>
<td>R*</td>
<td>-.12</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>.20</td>
</tr>
<tr>
<td>Frequency of admission</td>
<td>R*</td>
<td>-.35**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>.33**</td>
</tr>
<tr>
<td>Leg ulcer</td>
<td>R*</td>
<td>.04</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>-.19</td>
</tr>
<tr>
<td>Family function</td>
<td>R</td>
<td>-.48**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>.21</td>
</tr>
<tr>
<td>Attitude towards SCD</td>
<td>R</td>
<td>.48**</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>-.33**</td>
</tr>
<tr>
<td>Receipt of counselling</td>
<td>R*</td>
<td>.24*</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>-.17</td>
</tr>
<tr>
<td>Source of recruitment</td>
<td>R*</td>
<td>.22*</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>.02</td>
</tr>
</tbody>
</table>

r* = Spearman rho correlation coefficient. r = Pearson correlation coefficient
** Correlation is significant at the 0.01 level (2-tailed).
* Correlation is significant at the 0.05 level (2-tailed).
Coding: Gender- male = 1, female = 2; sibling has SCD – yes = 1, no = 2; Born in UK – yes = 1, no = 2; leg ulcer – yes = 1, no = 2; receipt of counselling – yes = 1, no = 2; source of recruitment – haematology clinic = 1, Sickle Cell Society = 2
Table 5.57a. Regression Model Summary

<table>
<thead>
<tr>
<th>Model</th>
<th>R</th>
<th>R Square</th>
<th>Adjusted R Square</th>
<th>Std. Error of the Estimate</th>
<th>Durbin-Watson</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>.69(a)</td>
<td>.48</td>
<td>.42</td>
<td>3.80</td>
<td>1.7</td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Stigma, Source of recruitment, Family function, Receipt of counselling, Frequency of admission, Gender, Attitude towards illness.
b Dependent Variable: Self esteem

Table 5.57b. ANOVA

<table>
<thead>
<tr>
<th>Model</th>
<th>Sum of Squares</th>
<th>df</th>
<th>Mean Square</th>
<th>F</th>
<th>Sig.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Regression</td>
<td>859.11</td>
<td>7</td>
<td>122.73</td>
<td>8.47</td>
</tr>
<tr>
<td></td>
<td>Residual</td>
<td>926.45</td>
<td>64</td>
<td>14.47</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Total</td>
<td>1785.56</td>
<td>71</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

a Predictors: (Constant), Stigma, Source of recruitment, Family function, Receipt of counselling, Frequency of admission, Gender, Attitude towards illness.
b Dependent Variable: Self esteem

Table 5.57c. Regression Coefficients

<table>
<thead>
<tr>
<th>Model</th>
<th>Unstandardised Coefficients</th>
<th>Standardised Coefficients</th>
<th>Collinearity Statistics</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>B Std. Error Beta T Sig.</td>
<td>B VIF</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>(Constant) 29.57 4.51 .24 6.55 .000</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Source of recruitment 2.75 1.03 .24 2.66 .010 .96</td>
<td>1.04</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Gender -2.32 .98 -23 -2.36 .021 .84</td>
<td>1.19</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Frequency of admissions -32 .37 -09 -.86 .391 .79</td>
<td>1.26</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Family function -29 .09 -29 -2.99 .004 .83</td>
<td>1.19</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Attitude towards SCD .14 .05 .29 2.78 .007 .75</td>
<td>1.33</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Receipt of counselling 2.06 1.49 .13 1.38 .172 .92</td>
<td>1.09</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Stigma -09 .28 -03 -.32 .747 .79</td>
<td>1.27</td>
<td></td>
</tr>
</tbody>
</table>

a Dependent Variable: Self esteem
In summary, a standard simultaneous multiple regression was used to test the ability of self-perceived stigma to predict self-esteem independent of other potential confounding variables (Source of recruitment, Gender, frequency of admissions, family function, attitude towards SCD, and receipt of counselling).

The model as a whole explained 48% of the variance in Self-esteem scores $F(7, 64) = 8.5$, $p < 0.001$. The statistically significant and independent predictors of self-esteem scores were; source of recruitment ($\beta = 2.8$, $p = 0.01$), gender ($\beta = -2.3$, $p =$
0.02), family function (beta = -0.29, p = 0.004, and attitude towards SCD (beta = 0.14, p = 0.007).

Thus these findings **DO NOT** provide support for the hypothesis “Self perceived stigma will significantly and independently predict Self Esteem measured by Rosenberg Scale”.

Depression and self esteem are closely related and low self esteem is a criterion diagnostic symptom for depressive disorder (WHO 1992). Bearing this in mind, the above regression analysis on self-esteem was repeated with depressive symptoms (SMFQ) included as a covariate (in place of stigma). Stigma was removed from the model when SMFQ was added in order to limit the number of covariates to a maximum of seven as previously explained. The inclusion of depression in the model is meant to explore if the independent predictors of self esteem identified earlier remained predictors even after taking depressive symptoms into account. The inclusion of SMFQ in the model made some difference in that gender and attitude towards SCD ceased to be significant predictors. The new model explained 56% of the variance in self esteem scores F(7, 64) = 11.7 p < 0001. The new statistically significant and independent predictors of self esteem scores were; depression (beta = -0.51, p = 0.001, source of recruitment (beta = 2.0, p = 0.047), and family function (beta = -0.20, p = 0.038).

The above finding suggests that exploring self-esteem in its own right rather than just as a symptom of depression is worthwhile. This significance is demonstrated by the fact that recruitment from Sickle Cell Society and healthy family function predicted self-esteem over and above the presence of depressive symptoms.

The finding that attitude towards SCD ceased to predict self-esteem when depression was included in the model suggest that negative attitudes towards SCD may be more relevant as a contributor to active psychological and emotional difficulties (e.g. as measured by SDQ and SMFQ) rather than to self-esteem (which is a background psychological state or trait).
Also, nullification of the predictive ability of attitude towards SCD on self-esteem in the presence of depression is not entirely surprising given that, intuitively, young people with SCD who have depressive symptoms are more likely to have a negative attitude towards the disorder they see as being responsible for their depression. This view is supported by a large correlation between depressive symptoms (SMFQ) and attitude towards SCD (CATIS), ($r = -0.52$, $p = 0.001$).

**Summary of multivariate analyses**
The multivariate analyses found support for two of the four study hypothesis. Hierarchical regression provided support for the first hypothesis that presence of leg ulcer (visibility) and frequency of ward admissions (disruptiveness)) will predict levels of self perceived stigma. Regression analysis also provided support for the second study hypothesis that self perceived stigma will predict SDQ Total difficulties score. The hypotheses that self perceived stigma will predict depressive symptoms was not supported. Instead, more depressive symptoms were predicted by poorer attitude towards SCD, unhealthy family function, and frequent ward admissions. Similarly, the hypothesis that self perceived stigma will predict self esteem was not supported by multivariate analysis. Instead, higher self esteem was predicted by recruitment from Sickle Cell Society, male gender, healthy family function and positive attitude towards SCD. Recruitment from Sickle Cell Society and healthy family function remained significant predictors of better self-esteem even when depressive symptoms were included as a covariate.
Chapter 6
Discussion

This chapter is discussed under the following subheadings:

6.1. Introduction and summary of main findings

6.2. Stigma: prevalence and correlates in SCD

6.2.1. Stigma prevalence
6.2.2. Application of study findings to stigma theory
6.2.3. Significance of leg ulcer in SCD

6.3. Prevalence and predictors of psychological difficulties (SDQ) in SCD

6.3.1. Prevalence of psychological difficulty
6.3.2. Association between stigma and psychological difficulty
6.3.3. Relationship between attitude to illness and psychological difficulty

6.4. Prevalence and predictors of depressive symptoms

6.4.1. Prevalence of depressive symptoms
6.4.2. Family function and depression
6.4.3. Illness severity and depression
6.4.4. Attitude to SCD and depression
6.4.5. Depression and stigma
6.4.6. Depression and receipt of counselling

6.5. Level and predictors of self esteem in SCD

6.5.1. Level of self esteem in SCD
6.5.2. Association between source of recruitment and stigma
6.5.3. Gender and self esteem
6.5.4. Family function and self esteem
6.5.5. Attitude to illness and self esteem
6.5.6. Stigma and self esteem

6.6. Methodological issues and limitations

6.6.1. Reliability
6.6.2. External validity
6.6.3. Limitations

6.7. Summary
6.1 Introduction and summary of main findings.

The aims of this study were to estimate the prevalence of self-perceived stigma in young people with SCD and to explore associations between self-perceived stigma and illness, psychosocial, and socio-demographic variables. These aims were achieved with a cross-sectional questionnaire survey of 93 young people with SCD aged 10-19 years. To my knowledge some of the findings of this study are unique and have not been previously reported among people with SCD. The main findings are summarised next.

- Only 15% of the respondents met the study criteria for directly measured self-perceived stigma. However, indirect assessment of self perceived stigma based on disclosure practices (e.g. keeping SCD secret) suggests a much higher level of perceived stigma (57%) in the current study.

- Consistent with stigma theory, the study found that high self-perceived stigma was independently and significantly predicted by presence of leg ulcer (measure of visibility), and increased frequency of ward admissions (measure of disruptiveness). As far as I am aware, this is the first time stigma theory has been successfully applied to SCD.

- Compared with SDQ UK norms, young people with SCD in this study scored significantly higher on the emotional subscale of the SDQ (p=0.0008). The prevalence of SDQ-caseness (caseness for psychological difficulty) among young people with SCD (15.4%) is slightly higher compared with young black boys and girls in London (9.2% and 10.9% respectively) but the difference was not statistically significant (p=0.078).

- More psychological difficulty was independently and significantly predicted by high self-perceived stigma and negative attitude to illness. The independent contribution of self-perceived stigma to psychological difficulty in SCD is another unique finding of the study.
• The prevalence of depressive symptoms and caseness for depression (depression-positive) in the young people with SCD were similar or lower compared with young black people in London

• Contrary to my hypothesis, depressive symptoms were not predicted by self-perceived stigma. However, increased depressive symptoms were independently and significantly predicted by unhealthy family function, increased frequency of ward admissions (index of severity), and negative attitude towards SCD.

• Only 18.8% of the children who were classified as depression-positive were in receipt of counselling. Also there was no statistically significant difference in receipt of counselling between the depression-positive and depression-negative cases.

• The young people with SCD achieved self esteem scores that were similar to young people without SCD in three other developed countries

• Contrary to my hypothesis, self-perceived stigma was not an independent predictor of self-esteem among young people with SCD. However higher self-esteem was significantly predicted by source of recruitment (respondents from Sickle Cell Society had higher self esteem than those from haematology clinics, gender (males higher than females), healthy family function, and positive attitude towards SCD. The predictive ability of gender and attitude towards SCD was nullified when depression was included in the regression model for self-esteem. However, source of recruitment and family function remained predictors of self-esteem independent of depressive symptoms.

• Given that the Sickle Cell Society is an advocacy organisation, the children recruited through the organisation were compared with those recruited from haematology clinics on self-perceived stigma and all psychological measures to see if they had different responses. Apart from self-esteem, there were no statistically significant differences between the two groups on psychological measures.
6.2. Stigma: prevalence and correlates in SCD

6.2.1. Stigma prevalence

Only 15% of the young people with SCD were classified as having self-perceived stigma in this study. These were the respondents who gave positive endorsement to one or more of the three direct stigma questions. To put this in perspective, the responses of the young people with SCD in this study were compared with those of young people with epilepsy (Westbrook et al 1992) and stuttering (Blood et al 2003), which used similar methodology (Table 5.20).

In response to the question “Do you think that having Sickle Cell affects whether people want to be friends with you?” 73% of the children with SCD in this study stated that this was “never” the case. The proportions of children with epilepsy and stuttering who gave the same answer are 66% (Westbrook et al 1992) and 65% (Blood et al 2003) respectively. Similarly, in response to the question, “Do you think that having Sickle Cell affects whether people like you or not?” 84% of the children with SCD stated that this was “never” the case. The proportions for children with epilepsy and stuttering are 60% and 63% respectively. These comparisons show that children with SCD in this study consistently endorsed less direct perception of stigma compared with children with epilepsy or stuttering.

Several reasons can account for the differential responding to these direct questions assessing the prevalence of self-perceived stigma (with respect to being liked by other children or not) by children with SCD in this study compared with children with epilepsy and stuttering.

The first possibility is that children with SCD may genuinely not perceive stigma as much as children with epilepsy or stuttering. This possibility has some support from stigma theory. As outlined in Chapter 2, conditions that are more visible tend to be more stigmatising than those with less visible features. Although SCD, epilepsy and stuttering all have visible features, it could be argued that for most affected persons in developed countries like the UK (with access to advanced medical care), SCD may be
less visible than epilepsy and stuttering. While some children with epilepsy may have infrequent seizures, each fit; especially grand mal seizures can be very dramatic and highly visible. Similarly, children with stuttering demonstrate evidence of their difficulty with almost every verbal communication.

Another possible explanation for the different prevalence is socially desirable responding by the children with SCD. However, there is no good evidence that children with SCD are more prone to socially desirable responding than children with epilepsy or stuttering. Neither this study nor the studies by Westbrook and colleagues or Blood and colleagues incorporated measures of socially desirable responding. So this hypothesis cannot be tested directly at present.

Differences in context could be another reason for the observed differences in prevalence of self-perceived stigma between children with SCD and those with epilepsy or stuttering. The latter two studies were conducted in the United States among predominantly white children, whereas this study was conducted in the UK among predominantly black children. I am not aware of any data on the differential perception of stigma among different ethnic groups or countries, but such a difference is possible. However, given the possible association between stigma and racism (Scambler 2004), a higher prevalence of self-perceived stigma among black children and young people could be expected. Instead, I found the opposite.

We also used questions on disclosure behaviour as indirect measures of self-perceived stigma. Unlike the direct questions on self-perceived stigma, more of the children and young people with SCD in this study positively endorsed the disclosure questions. For example, 54% reported that they keep their SCD secret “sometimes” or “often”. This compares with 53% for children with epilepsy (Westbrook et al 1992) and 40% for children with stuttering (Blood et al 2003). Also 57% of children with SCD in this study reported that they “rarely” or “never” talk to other people about having SCD. This figure compares with 70% of children with epilepsy and 60% of children with stuttering. Thus, although the prevalence of directly measured self-perceived stigma (with regards to being liked by other children) was lower among children with SCD,
indirect evidence from “disclosure questions” suggests that the prevalence of self-perceived stigma is closer to children with epilepsy or stuttering.

In summary, when self-perceived stigma is assessed directly by measures of being liked by other children, fewer children with SCD appear to perceive stigma compared with children with epilepsy or stuttering. However, when measured indirectly, similar proportions of children in all three disorder groups appear to perceive stigma. This observation provides some indirect support for the suggestion in an earlier paragraph (above) that socially desirable responding by children with SCD may account for their lower endorsement of direct questions on perceived stigma compared with the other two groups. One way to explore this further is a head-to-head study of self-perceived stigma among children with SCD and the other disorders with an instrument that incorporates a measure of socially desirable responding.

6.2.2. Application of study findings to stigma theory

The findings of this study provide support for the application of stigma theory to SCD. As outlined in Chapter 2, stigma dimensions are the characteristics that determine the stigma potential of conditions like SCD (Katz 1981, Jones et al 1984). The stigma dimensions that are relevant here are “visibility” and “disruptiveness”.

Visibility refers to the extent that SCD has attributes that are obvious, not concealable, or aesthetically challenging to others. In general, stigma theory predicts that the more visible and disfiguring an attribute the more stigmatising it is likely to be. Some people with SCD have easily recognisable physical manifestations such as jaundice, leg ulcers, and delayed physical development (Dick 2008). Severe cases, especially in developing countries where effective treatments are not widely available, may be associated with gross physical signs such as gnathopathy (Wessberg et al. 1980), and bossing of the forehead (Acquaye et al. 1985). While widespread access to advanced medical treatment in the UK makes such gross physical signs unlikely, many affected persons in the UK still have obvious signs of the disease. In this study, almost half of the subjects (46.1%) were jaundiced but very few (5.6%) had leg ulcer. Jaundice was not associated with self-perceived stigma in bivariate analysis but leg
ulcer was; hence the latter was used as the measure of visibility in multivariate analysis.

The stigma dimension of “Disruptiveness” describes the extent to which having SCD interferes with personal functioning and interpersonal relationships of affected persons. The findings of this study support the well recognised fact that the natural course of SCDs is variable such that while many affected persons live relatively healthy undisrupted lives, others require frequent hospitalisation as a result of different acute illness episodes particularly pain (Wethers 2000). For example, 38.6% of the respondents in this study had had no ward admissions in the past year while 16% had five or more admissions. In addition, some affected persons may experience even more frequent but less severe episodes of pain not requiring hospital admission but nonetheless necessitating rest at home. The limitations imposed by these illness episodes could be disruptive to schooling, employment, and social encounters (Atkins and Ahmad 2001). For example, 23.3% of the respondents in this study had had four weeks or more of school absence in the previous year. The disruptiveness engendered by these frequent admissions, pain and school absence make concealment of having SCD difficult and increase the threat of unwanted disclosure.

In this study, visibility was assessed with presence of leg ulcer, while disruptiveness was assessed with frequency of ward admissions. As predicted by stigma theory, both measures were independent and significant predictors of self-perceived stigma. This is the first study that I am aware of to show a direct application of stigma theory to SCD.

6.2.3. Significance of leg ulcer in SCD
As already highlighted above, this study found that leg ulcer independently predicted self perceived stigma. Leg ulceration is a severely disabling complication of SCD associated with the most severe forms of the disease (Halabi et al 2008). The ulcers are chronic with up to 40% having open wounds for over a year (Briggs and Flemming 2007) and a median duration of 29 months even in developed countries (Halabi et al 2008). In a study of risk factors for leg ulcers in people with SCD in Jamaica, Cumming and colleagues found the predictors of ulceration to include low socio-economic status and biological variables such as high lactate dehydrogenase and venous incompetence (Cumming et al 2008). The prevalence of leg ulcer in that
study was 29%, which is considerably higher than the 6% found in the current study. This difference may reflect access to advanced treatment for SCD in the UK compared with Jamaica.

In addition to being a marker for severity, leg ulceration has been directly associated with social difficulties in SCD in other studies. For example, in an early study in Jamaica, Alleyne and colleagues found that compared with a control group with no leg ulceration, the ulcer-affected group experienced wide ranging adverse psychosocial effects on education, employment, and marriage (Alleyne et al. 1976).

Given the specific association shown in this study between leg ulcer and self perceived stigma, leg ulceration could be seen as a specific physical indicator of increased psychosocial vulnerability. Incidentally, in the current study, leg ulcer had statistically significant bivariate associations with the emotional subscale of the SDQ, depressive symptoms, and poor attitude towards SCD. These findings would suggest that more effective treatment of leg ulcer could contribute to improved psychological well-being in SCD. There is in fact evidence that intensive combination of several treatment modalities can achieve rapid healing of chronic leg ulcer in people with SCD (Schleucher et al 2007).

6.3.  Prevalence and predictors of psychological difficulty (SDQ)

6.3.1. Prevalence of psychological difficulty

In this study, psychological difficulty was assessed with the self-report version of the Strengths and Difficulties Questionnaire (SDQ). The SDQ produces a Total Difficulties Scale in addition to five other subscales (Emotional, Hyperactivity, Conduct, Peer problems, and Prosocial). I used the cut-off of \( \geq 18 \) on the Total Difficulties Scale to define SDQ-caseness as this cut-off gave prevalence figures in the British Child and Adolescent Mental Health Survey (Meltzer et al 2000) that were equivalent to the prevalence found using data from multiple sources including parents and teachers. The same cut-off was also used in a more recent UK epidemiological study (Stansfeld et al 2004). Based on this cut-off, the proportion of young people with SCD classified as SDQ-cases (15.4%) was higher compared with black boys or girls in East London schools (9.2% and 10.9% respectively) (Stansfeld et al 2004). However, the
difference was not statistically significant (p=0.078). I am not aware of any other SCD-related study that has used the SDQ as a measure of psychological difficulty.

In relation to the SDQ subscales, a comparison of the mean scores by young people with SCD in this study and the UK norms showed statistically significant differences in three subscales (emotional, hyperactivity and prosocial scales). Conduct and peer problems subscales were not significantly different.

For the three subscales with significant differences, young people with SCD scored higher on the emotional subscale and lower on both the hyperactivity and prosocial subscales.

Large epidemiological studies of childhood mental disorders (e.g. Meltzer et al 2000) consistently find increased risk of psychopathology among children with chronic physical. Children with disorders that involve the brain (like SCD) are particularly at higher risk. For example, Pegelow and colleagues found that up to 20% of people with SCD have evidence of ischaemic brain damage on MRI by the age of 20 years (Pegelow et al 2002). Thus, compared with UK norms, the higher score on the SDQ-emotional subscale by young people with SCD may be partly related to overt or covert ischaemic brain damage. In addition to bio-medical causes, other psychosocial factors (e.g. cognitive and educational, family and wider interpersonal relationships) may be contributing to the observed difference.

As discussed in Chapter 2, studies of psychopathology in SCD tend to give different results depending on whether the samples are from developed or developing countries. In general, the trend suggests that more recent studies in developed countries find no increased rates of psychopathology while studies in developing countries continue to show higher rates compared with unaffected control groups. Helps and colleagues suggest that this differential trend might be due to easy access to advanced physical care for people with SCDs in developed countries (Helps et al 2003). Bearing this in mind, the prevalence of psychopathology in this study will only be compared with previous studies in developed countries.
Thus, on the whole, recent studies in developed countries show closer to normative levels of psychopathology in SCDs. For example, a study of affected children in USA (Noll et al 2007) found no differences in measures of emotional wellbeing compared with their unaffected peers. In this study, teachers described the children with SCDs as more prosocial and less aggressive. These results are similar to what the authors found 10 years earlier (Noll et al 1996).

Another study of adolescents with SCDs in USA (McElligott 2006) found that affected young people did not score higher than the norms in different measures of self esteem, anxiety, depression and behavioural difficulties. Further, a longitudinal study, which followed children with SCDs in USA for up to 10 years found no differences in measures of depression, self worth and internalising symptoms compared with healthy peers either cross-sectionally or longitudinally (Getzoff 2005). In an earlier study in the UK involving 39 children with SCD and 24 controls, Midcence and colleagues (Midence et al 1996) found no significant differences between the two groups on depression and self esteem.

Thus, the finding in this study of a statistically significant difference in the SDQ-emotional subscale compared with UK norms is not in keeping with the trend of less psychopathology in recent studies in developed countries. There are six possible explanations for this difference. First, it is possible that the emotional subscale of the SDQ used in this study is a more sensitive measure compared with the questionnaires and methods of assessment used in the previous studies. Secondly, there could be relevant differences in the sampling of the young people with SCD and their reference groups. For example, this study compared the young people with SCD against UK national norms and reference groups, which were established 6-10 years earlier. This is different from the other studies in which the young people with SCD were compared with contemporary control or reference groups. Third, there could be cultural differences given that the three most recent comparison studies (Noll et al 2007, McElligot 2006, and Getzoff 2005) were conducted among American children, while this study is on UK children. Fourth, my study has both community and clinic samples compared with two of the other studies (Noll et al 2007, Getzoff 2005) which were community samples. Although the two samples in my study did not differ significantly in stigma or psychopathology, the trend was for more depressive
symptoms in the clinic sample and the clinic sample in my study had significantly lower self esteem. Fifth, whereas my sample did not exclude children with overt brain involvement, at least one of the comparison studies (Noll et al 2007) included only children with SCDs who had not had an overt stroke. Finally, this could be a false positive finding which has arisen by chance due to the multiple statistical tests in the Thesis. It is recognised that if multiple statistical tests are conducted at 5% level of significance, there is a 1 in 20 chance of a false positive finding.

6.3.2 Association between stigma and psychological difficulty
Consistent with my hypothesis, regression analysis (Table 5.53c) showed that self-perceived stigma independently and significantly predicted scores on the Total Difficulties scale of the SDQ. To my knowledge, this is the first study to demonstrate this association between stigma and psychological difficulty in SCD.

This finding is consistent with previous studies of self-perceived stigma and other physical disorders with recognised stigma potential. For example, several studies have shown associations between self-perceived stigma and higher levels of psychological and emotional distress in psoriasis (Richards et al 2001, Leary et al 1998), vitiligo (Kent 2000), and epilepsy (Westbrook et al 1992; Austin et al 2004; Adewuya et al 2006).

Several mechanisms can explain the association between self-perceived stigma and psychological difficulty in SCD. First, self-perceived stigma has been shown to increase a sense of demoralisation and alienation (Link and Phelan 2001). Applying this to SCD, it is possible that young people with SCD who perceive stigma may feel different from others, which further increases what Link and Phelan referred to as the “us and them” gap (Link and Phelan 2001), which in turn could lead to even more sense of alienation and a vicious negative cycle. For a young person who already has a serious, potentially life threatening illness, this sequence of events could lead to sustained psychological difficulty.

Another mechanism associating self-perceived stigma and psychological difficulty is pervasive fear of unwanted disclosure of their SCD status. The fear of disclosure
could lead to avoidance behaviour such as social withdrawal and self isolation. Consistent with the cognitive behavioural theory, this sequence of events could set up a vicious cycle resulting in progressive increase in self-perceived stigma and worsening distress and self isolation. Link and colleagues have shown that self isolation is a common stigma management strategy for people with stigmatising conditions (Link et al 1991). Unfortunately, this is a maladaptive coping strategy with costly penalties in terms of peer relationships. This study provides support for this proposition. For example, the study showed that of the four subscales of the SDQ, the peer problems scale recorded the most difference between the young people with SCD who perceived stigma and those that did not perceive stigma (Table 5.32). An even greater support for this hypothesis is provided by the observation that peer problem does not appear to be a pervasive problem for all young people with SCD but rather a specific difficult for those who have self-perceived stigma. For example, Table 5.12 shows that compared with UK norms, as a group, the young people with SCD did not differ from their peers on the peer problems scale of the SDQ. In fact, of the five SDQ subscales, the peer problem scale was one of the only two scales that did not differ between the young people with SCD and UK norms. Thus peer problems appear to be a specific difficulty only for the young people with SCD who perceive stigma.

The significance of the finding in this study that self-perceived stigma predicts psychological difficulty in SCD is the possibility that intervening directly against self-perceived stigma could reduce the risk of psychological difficulty in SCD.

6.3.3 Relationship between attitude to illness and psychological difficulty
Attitude towards SCD (measured by CATIS) was a statistically significant independent predictor of Total Difficulties Scale of the SDQ. This is another unique finding, which has not been shown previously among children with SCD.

This finding shows that young people’s attitude towards having SCD can influence their psychological adjustment or vice versa. This is consistent with studies of young people with other chronic physical conditions like epilepsy, asthma, and diabetes mellitus, which show association between negative attitude towards illness and a range of adverse psychological outcomes including depression, behaviour problems and low self esteem (Austin and Huberty 1993, Amer 2008).
It is possible that young people with negative attitude toward SCD could see the disorder and associated disruptions as frequent intrusions on daily life. This view of their circumstance could result in a perpetual sense of distress and inability to take up and maximise their periods of adequate health. On the other hand, a positive attitude toward SCD could help bolster young people’s resistance to risk factors for poor adaptation. For example, if a young person sees the limitations associated with having SCD as challenges that can be overcome, they would be more likely to actively seek and use opportunities to optimise their function and reduce impairment.

6.4. Prevalence and predictors of depressive symptoms

6.4.1. Prevalence of depressive symptoms
Depressive symptoms were assessed with the Short MFQ. Caseness for depression (depression-positive) was determined by converting the Short MFQ scores into categorical classification using a cut-off score of \( \geq 8.0 \). This cut-off was chosen because a previous UK study (Thapar and McGuffin 1998) found 8.0 to have optimum sensitivity and specificity for the self report version of the Short MFQ. Also the original psychometric study on the Short MFQ in USA found optimum sensitivity and specificity with a cut-off of 8.0 (Angold et al 1995).

In this study, the whole sample of children with SCD had a Mean score on the Short MFQ of 4.5 with a standard deviation of 4.0. These figures are comparable to the mean Short MFQ scores of healthy UK twins (Mean (SD) 4.5 (5.2). Incidentally, the mean score for the whole sample of children with SCD in this study was much lower compared with the scores of UK twins clinically diagnosed with a depressive disorder (Mean (SD) 4.5(4.0) Vs 8.8(4.2).

In this study, a cut-off of \( \geq 8.0 \) on the Short MFQ identified 18% of the children with SCD as depression-positive cases. However, given the evidence that questions about somatic symptoms in screening questionnaires like the Children Depression Inventory (which is similar to the Short MFQ) lead to overestimation of depression in SCD
(Yang et al 1994), the actual proportion of children identified in this study as depression-positive cases may be lower than 18%.

However, to put the prevalence of depression caseness in this study into perspective, I compared it with the prevalence of depression caseness reported in a community-based survey involving Black adolescent children in London using the same cut-off of ≥ 8.0 on the Short MFQ (Stansfeld et al 2004). These researchers reported a depression caseness of 17.2% for Black boys and 29.5% for Black girls (Stansfeld 2004). Thus the prevalence among young people with SCD in this study is comparable to the reported prevalence for Black boys but much lower than the prevalence for Black girls in the community. However, this comparison should be taken with caution because the young people in my study appeared to be socioeconomically more advantaged than the Black children in the Stansfeld et al (2004) study (e.g. unemployed head of household 5% vs. 25% and car ownership 80% vs 70%). Also my sample had a wider age range (10-19 years) compared with the sample in Stansfeld et al (2004) 11-14 years. I have focused the comparison with Black children because SCD affects predominantly people of African and Caribbean origin but the prevalence of depression caseness for White children in the same study was similar to Black children (Stansfeld et al 2004).

The finding of no increased rates of depression in this study is consistent with the trend since the late 1990s whereby people with SCDs in developed countries have less psychopathology compared with previous decades and compared with affected persons in developing countries (see chapter 3).

In summary, the findings from this study and the comparison with Black children in the UK suggest that with specific reference to depressive symptoms, children with SCD do not appear to have increased rates of depression caseness. However, as already discussed, unlike depression (measured by SMFQ), the young people with SCD in this study scored higher on the emotional subscale of the SDQ compared with UK population norms (Goodman et al 1998). These different findings indicate mixed results suggesting that the prevalence of some specific psychopathologies (e.g. depression) is not increased while the prevalence of more general emotional difficulty is increased in SCD. The difference in the findings could also be due to the different
symptoms targeted by the SDQ and SMFQ. For example, of the five items in the emotional subscale of the SDQ, four items are explicit measures of anxiety rather than depression. Thus, it is possible that anxiety related measures are more sensitive in SCD. In this study the emotional subscale of the SDQ proved more sensitive than the Total Difficulties scale and the other four subscales.

This study found that female children had a statistically significantly higher score on the Short MFQ compared with males. This finding is consistent with other studies of depression in adolescence (e.g. Stansfeld et al 2004).

Even though rates of depression were not increased against expected norms we found associations between depression and illness features which indicate that when depression is present in young people with SCD, it is shaped by the illness; hence depression may affect the management of SCD and its management is likely to require an understanding of the vicissitudes of SCD. In the following paragraphs I discuss the psychosocial and illness associations of depression in our sample with SCD.

6.4.2. Family function and depression

Family function was one of the three statistically significant and independent predictors of high SMFQ scores. The other predictors were negative attitude towards SCD and severity (increased frequency of ward admissions).

The association between family function and depression can be easily understood from an ecological perspective (Brofenbrenner 1979). Families can have a powerful influence on the well-being of their members, especially those already vulnerable from physical and mental disorders. For example, Silk and colleagues have recently shown that compared with a low risk control group, children with a current or previous episode of major depressive disorder had mothers with higher expressed emotion, especially criticism (Silk et al 2009).

With reference to SCD, other studies have shown a powerful influence of families on the adjustment of affected members. For example, in a study of 182 young people with SCD, Barbarin and colleagues found that the best predictors of the affected
child’s psychological adjustment included relationships with their parents and siblings (Barbarin et al. 1999). Similarly, Burlew et al. (2000) found that ecological factors such as family relationships were better predictors of adjustment than biomedical factors in SCD.

Thus, the finding in this study that family function predicts depressive symptoms is in keeping with theoretical models of adjustment and previous studies of children with SCDs.

Living with a child affected by SCDs could be challenging especially for more severe cases with frequent complications. Parents may have to make allowances for and give more attention to the affected child. In particular, parents’ capacity to set and enforce appropriate boundaries for their child with SCD may be compromised by this perceived need to make allowances for the child. This could lead to resentment by siblings and disagreements and feeling of alienation in the spouse or partner. Parents may have to give up work to provide more care for the affected child resulting in lost economic and social opportunities. Parents may be blamed by others or may blame themselves for having an affected child (Burnes et al. 2008). Parents and siblings could be at risk of courtesy stigma (Hinshaw 2005).

Given the importance of family in the psychosocial adjustment of children with SCD as shown in this and other studies, it is important that families are assessed and supported as part of caring for the child affected by SCDs.

6.4.3. Illness severity and depression
Consistent with the bio-psycho-social model of adjustment in chronic physical disorders, this study found that a surrogate index of severity of SCD (frequency of ward admissions) was one of the independent predictors of depressive symptoms.

This finding is in keeping with the general evidence that people with more severe forms of SCDs are at more risk of depression. For example, Segbena and Sangare (1994) used the Hamilton Depression Rating Scale to assess 30 patients with SCDs and 31 heterozygous carriers of the sickle gene. Although no subject in either group scored above the threshold for moderate depression, the level of anaemia and the
The number of sickle-cell crises per year was associated with depressive symptoms in the SCDs group. Similarly, Hasan and colleagues showed that SCD patients who make more frequent use of accident and emergency department and had more frequent blood transfusions (both good markers of severity) were more likely to be depressed (Hasan et al 2003).

Thus, although disease severity alone is not a sole determinant of mood or function in SCD (Grant et al 2000), evidence from this and other studies suggest that young people with more severe forms of SCD (e.g. frequent in-patient admissions) should be targeted for screening for depression.

6.4.4. Attitude to SCD and depression
Negative attitude to SCD was the third statistically significant independent predictor of depression in young people with SCD in this study. This is consistent with previous studies of young people with other chronic physical conditions like epilepsy, asthma, and diabetes mellitus, which all show association between negative attitude towards illness and psychological adversities such as depression, behaviour problems and low self esteem (Austin and Huberty 1993, Amer 2008). The mechanisms and implications explained in Section 6.3.3 will also be applicable to depression; hence will not be repeated.

6.4.5. Depression and self-perceived stigma
My hypothesis that self-perceived stigma would be an independent predictor of risk of depression in young people with SCD was not supported by the data. Although risk of depression and self perceived stigma were strongly associated in bivariate analysis, the relationship was not sustained in multivariate analysis. Once other relevant variables such as age, gender, leg ulcer, frequency of admissions, family function, birth in or outside UK and attitude towards SCD were controlled for, the apparent association between self-perceived stigma and depression ceased.

This finding that self-perceived stigma did not predict depression was surprising given that self-perceived stigma did predict SDQ, which is a measure of psychological difficulty. The difference may lie in the nature of the constructs measured by the two instruments used to assess depression (SMFQ) and psychological difficulty (SDQ). On
closer examination, two SDQ subscales (emotional and peer problems) had the strongest association with self-perceived stigma. The differences between these two subscales and the SMFQ may hold the key to why this difference occurred. Unlike the SMFQ which maps only onto depression, the emotional subscale of the SDQ maps mostly onto anxiety. It is therefore possible that measures of anxiety are more sensitive to self-perceived stigma than measures of depression. Also, in keeping with stigma theory, the peer problem scale of the SDQ is likely to be sensitive to self-perceived stigma compared with the SMFQ. In fact, the SMFQ has no question that directly taps into peer problems.

6.4.6. Depression and receipt of counselling

Although 18% of the children with SCD in the study were classified as depression-positive cases on the Short MFQ and 15.4% scored in the abnormal range on the Total Difficulties Scale of the SDQ, only 11% of the whole sample was receiving counselling from a therapist. More importantly, only 18.8% of the children who were classified as depression-positive were in receipt of counselling. Also there was no statistically significant difference in receipt of counselling between the depression-positive and depression-negative cases.

This finding suggests possible problems with identifying children with SCD who have significant psychological and emotional difficulty and targeting psychological therapy towards them. This mismatch could be due to patchy availability of assessment and counselling opportunities for children with SCD or lack of information to enable precision in targeting resources to those in greatest need. Thus the findings of this study could contribute towards targeting. For example, although the prevalence of depression is not increased in young people with SCD as a group, the following three subgroups; those with unhealthy family function, more severe forms of SCD, and more negative attitude toward SCD, should be targeted for screening for depression. The study also showed that the Short MFQ could be a useful screening instrument for depression in SCD although the overlap of physical symptoms between the questionnaire and SCD could lead to high false positive rates. It is important to resource local SCD services with paediatric liaison Child and Adolescent Mental Health staff to ensure that those identified have ready access to psychological
treatment as appropriate. Alternatively, cases identified could be referred to local Child and Adolescent Mental Health Services (CAMHS).

6.5. Level and predictors of self esteem in SCD

6.5.1. Level of self esteem in SCD

This study measured self esteem with the Rosenberg scale. Therefore in order to assess whether or not the self esteem of the young people in this study is normative, their scores have to be compared with a similar sample where the same instrument was used as a measure of self esteem. I found only one previous study of children with SCD where the Rosenberg scale was used to measure self esteem (Burlew et al 2000). The mean score on the Rosenberg Self Esteem scale by young people in my study was 31.9 (SD 5.0). These figures are almost the same as the mean score in the study by Burlew and colleagues (Mean = 31.2, SD = 5.5) (Burlew et al 2000). As there were no other studies of children with SCD in which Rosenberg self esteem scales was used, I compared my sample with other groups. I was unable to find a UK norm for the Rosenberg scale but I located a multinational study of self esteem among students in USA, Canada and New Zealand, which used the Rosenberg Self esteem scale (Rusticus et al 2004). Compared with the mean score of young people in my study (Mean, 31.9, SD 5.0), the American students scored exactly the same (Mean, 31.9, SD 5.0), the Canadian students score slightly less (Mean, 31.0, SD 4.8), while the New Zealand students scored the lowest (Mean 29.9, SD 4.5).

In general, the results of studies of self esteem in children with SCD are inconsistent just as the studies of psychopathology in children with SCD. As such, some studies show reduced self esteem (e.g. Brown et al 1993), while others (like my study) indicate normal self esteem (e.g. Midence et al 1996, Cepeda et al 2000). It is possible that this variation is due to use of different measurements for self-esteem in the studies (e.g. Brown et al 1993 used Harter’s scale, Cepeda et al 2000 used Peirs-Harris scale, while my study used the Rosenberg scale).

In this study, self-esteem was strongly correlated with depression, Total Difficulties scale and emotional and peer problems subscales of the SDQ, and attitude towards SCD. Good self esteem is thus a desirable characteristic especially in children with a chronic
disorder and is regarded as a protective factor in relation to the development of some psychological disorders (Kliewer and Sandler 1992). I will therefore consider here the specific associations with young people with SCD.

6.5.2. Association between source of recruitment and self esteem

An interesting finding of the study was that Self esteem was independently and significantly predicted by whether subjects were recruited from Sickle Cell Society or haematology clinics. Young people recruited from Sickle Cell Society scored higher on the self esteem scale than those recruited from haematology clinics. This finding remained even after controlling for depressive symptoms.

Consistent with this finding, several other studies have linked membership or participation in self help or support groups with improved self-esteem in a wide variety of disorders and settings (Yahne and Long 1988; Folgheraiter and Annalisa 2009; Castelein et al 2008).

Whilst it is not possible to rule out the possibility that better self-esteem and possibly confidence made membership of Sickle Cell Society more likely, it is also worth considering how the latter in turn could help improve self-esteem.

Crocker and Major (1989) have outlined possible mechanisms which could help vulnerable people to preserve their self esteem despite experiencing stigma. Some of these strategies could explain how membership or participation in self-help could improve self esteem in SCD. One protective mechanism is the availability of alternative positive attitudes from significant others. Sickle Cell Society is an advocacy organisation. They employ volunteers who provide active support and encouragement to people with SCD, which is likely to improve their self esteem. In fact, the Organisation runs a befriending service for young people with SCD with one of the stated aims being “to boost their confidence and self esteem.”


Another mechanism is what Crocker and Major (1989) referred to as “in-group comparison”. This strategy suggests that membership of an organisation like Sickle Cell Society provides the young people opportunities to compare themselves with
other young people with similar difficulties. They argue that this comparison is less exacting on self esteem than comparisons with other people with perceived advantages.

6.5.3. Gender and self esteem
Gender was one of the independent significant predictors of self esteem in this study. Male respondents scored higher than females on the self esteem scale. However, the robustness of this association is in question as it was nullified when depressive symptoms were included in the model.

Other studies have shown higher self esteem in males than females. For example, Kling and colleagues conducted a meta-analysis of studies involving over 90,000 respondents to examine this phenomenon (Kling et al 1999). They found a small overall effect size of 0.21 in favour of males. Incidentally, the authors found that the greatest difference between the genders emerged in adolescence, which is the age group of the respondents in my study. Kling and colleagues also analysed three large data sets of 48,000 young Americans and found that male students scored higher than females in measures of self esteem (Kling et al 1999). These authors offered several potential explanations. For example, they highlight that traditional male gender roles tend to be associated with better self esteem. Another factor they suggested is physical appearance, especially in adolescence. Whereas the masculine physique developed by males in adolescence is associated with improved self esteem, the weight gain associated with menarche in females results in body image dissatisfaction and low self esteem in some girls.

6.5.4. Family function and self esteem
Family function is one of the four statistically significant independent predictors of self esteem scores by young people with SCD in this study. Young people from families with “unhealthy function” scored lower on the Rosenberg self esteem scale compared with those from healthy functioning families. This finding remained even after controlling for depressive symptom. It is also consistent with other studies on the development of self esteem in children, which emphasise the importance of the family (Gecas and Schwalbe, 1986).
The family is one of the most important environmental influences on the development of children’s self esteem. This is not surprising given the critical role of the family in children’s early socialisation and identity formation. In the context of a child with SCD, certain aspects of family function could have potential adverse consequences on the children’s self esteem. Examples include inappropriate comparisons with non-affected siblings, and overprotection resulting in less than optimum challenging of the affected child’s abilities (e.g. inappropriate exclusion from all sporting activities). On the other hand, adequate parental warmth, support and encouragement that are frequently and positively communicated to the child could benefit the self esteem of children affected by SCDs (Gecas and Seff 1990).

6.5.5. Attitude toward illness and self esteem
Attitude to illness was a statistically significant predictor of self esteem in this study. Young people with negative attitude toward SCD had lower self esteem compared with those with positive attitude. Although this finding was not sustained when depression was included in the regression model, it is nonetheless illustrative of the importance of attitude towards illness in the adjustment of young people with chronic physical disorders. Other studies of young people with diabetes (Ho et al 2008, Amer 2008) and epilepsy (Heimlich et al 2000) have shown associations between attitude towards illness and self esteem.

The finding that attitude towards SCD ceased to predict self-esteem when depression was included in the model is not surprising. For example young people with SCD who have more depressive symptoms are likely to have a negative attitude towards the disorder they see as being responsible for their depression. This observation also suggests that negative attitudes towards SCD may be more relevant as a contributor to active psychological and emotional difficulties than to self-esteem, which is a background psychological trait.

6.5.6. Stigma and self esteem
Contrary to my hypothesis, in this study, self-perceived stigma was not an independent statistically significant predictor of self esteem among young people with SCD. This was a surprise and ran contrary to studies of other chronic physical conditions in children and young people, particularly epilepsy, which have shown significant associations between
self perceived stigma and self esteem (Westbrook et al 1992). However, the lack of association between self-perceived stigma and self esteem in this study could be because the measure (Rosenberg scale) was not sensitive enough. It is possible that a longer, multi-domain and child-specific measure (e.g. Pier-Harris or Harter scales) might have demonstrated a significant association (Piers and Harris 1969, Harter 1985).

Although my study did not find independent association between stigma and self esteem, it is worth exploring the possible mechanisms since these could explain the alternative findings in other studies (e.g. Westbrook et al 1992).

Crocker and Major have reviewed mechanisms by which stigma could lead to low self esteem (Crocker and Major 1989). Two examples are readily applicable to the association between self esteem and self-perceived stigma found in some studies. One example is the so called “reflected appraisals” theory. This theory proposes that an individual could develop low self esteem if he believes that other people have a low or negative evaluation of him and goes on to adopt the negative views he perceives are held about him. The second mechanism is called “efficacy-based” theory. According to this theory, self esteem is improved by someone’s ability to control and master their environment (Crocker and Major 1989). Thus, conditions like SCD, which could interfere with or limit a person’s ability to demonstrate some types of competence, ability or achievement, could interfere with self esteem. A third mechanism is the “discounting” theory (Crocker and Major 1989). This theory suggests that people with stigmatising conditions may attempt to protect their self esteem by diminishing the importance or significance of abilities they are unable to acquire. While this may help them cope in the short term, in the long term, they may become even more sensitive to differences between them and their peers resulting in a greater threat to their self-esteem (Yovetich et al 2000).

While the theoretical and empirical details outlined above provide a clear association between low self esteem and self-perceived stigma, this association is also not inevitable for every person with a stigmatising condition (Crocker and Major 1989). Some individuals protect their self esteem despite experiencing or perceiving stigma. As already pointed out, one protective mechanism is the availability of alternative positive attitudes from significant others such as family members, teachers, and
counsellors and the ability of the young person to accept and assimilate the positive views.

6.6. Methodological issues and limitations
The study has many strengths which include the sample being reasonably representative (e.g. even gender split and normally distributed age), the sample size which was large enough to examine the issues under study, the reliability of the instruments used and the acceptable response rate. Nevertheless it also has limitations. The strengths and limitations are discussed in detail next.

6.6.1. Reliability
Reliability refers to the level of consistency shown by a measuring instrument. There are five reasons to support the reliability of the measurements used in this study.

- First, measurements were selected only if they had shown good reliability in their development and previous publications.
- Second, the measurements all showed good to excellent internal consistencies (Cronbach’s Alpha) in this study.
- Third, the measurements correlated in a sensible and predictable manner with each other.
- Fourth, the data demonstrated the types of associations predicted by stigma theory.
- Fifth, there was a complete agreement when some responses by ten of the children were checked against their medical records.

6.6.2. External validity
This refers to how generalisable the findings could be to a wider population (Ebrahim and Sullivan 1995). The following considerations support the external validity of the findings.
• First, the response rate of 45% is respectable given that the target sample is an over researched and hard to reach group

• Second, there were no major differences between respondents and non respondents in the demographic measures we had access to.

• Third, the gender ratio of the respondents in the study is about equal and so reflective of the general population

• Fourth, about a quarter (29%) of the children had a sibling with SCD. This proportion is consistent with the expectation for a recessively inherited autosomal condition where a quarter of conceptions are likely to be homozygous.

• Fifth, the findings that females had more depressive symptoms and more self-perceived stigma than males are consistent with other studies.

• Sixth, of the several socio-demographic and psychosocial variables compared between the children recruited from sickle cell society and those recruited from haematology clinics, only two statistically significant differences were identified between the two groups (socioeconomic status and self esteem). The two groups did not differ significantly on age, gender, whether or not born in the UK, levels of self-perceived stigma, whether they completed the questionnaire at home or in the clinic, depressive symptoms, Total Difficulties scale or subscales of the SDQ, receipt of counselling, family function, or frequency of ward admissions (measure of severity).

6.6.3. Limitations
There are several limitations to this study.

• The first and important limitation of the study is the absence of data on the genotype of the young people who took part in the study. It is well recognised that different genotypes in SCD confer different severity profiles to affected persons. For example, people with HbSS genotype tend to have a more severe illness compared with those with HbSC genotype (Peak 2008). The study used surrogate
markers of severity such as frequency of admissions, whereas genotype would have been a more direct and valid biological measure of severity. Surrogate markers of severity are at risk of confounding by other factors such as treatment adherence or the impact of other unrelated diseases or environmental factors. Also the inclusion of all forms of SCDs in the study was likely to have introduced some variability which is difficult to quantify because of the absence of information on the young people’s genotype. Consideration was given to obtaining the children’s genotype in this study. However, it was felt that children may not be reliable informants for such information and that accessing medical records for all participants would be the reliable means of obtaining the data. Unfortunately, the study did not have the capacity to cope with the logistics of accessing medical records for all participants. Also the conditions of the study ethics approval allowed only limited access to ten medical records to assess reliability of responses. However, since the conclusion of the study, I have become aware that children with SCDs and their parents can be reliable informants about their genotype. Consideration is now being given to seeking further ethics approval to obtain this data. However, the additional data if obtained would not form part of this Thesis.

- The second limitation is that the respondents were volunteers who were willing to participate in research and thus might constitute an altruistic group who may have come to terms with SCD in a way that those who did not volunteer to take part might not have. This potential selection bias might account for the less than hypothesised prevalence of stigma and normative levels of depressive symptoms and self esteem.

- Third, socially desirable responding was not assessed in the study. This may have contributed to lower than hypothesised prevalence of self-perceived stigma.

- The study adopted a cross-sectional design. This was logistically more feasible to execute and methodologically adequate to test the study hypotheses. However, it has the limitation of ascertainment of risk factors at the same time as the outcome
measures (e.g. stigma). This makes temporal relationships and causal interpretations difficult.

- Despite the cross-sectional data collection, the interpretation of the study hypotheses was strengthened by the use of robust multivariate data analysis such as standard and hierarchical regression modelling. While not able to resolve the dilemma of temporality, the regression techniques produced more robust associations by controlling for known and measured confounders.

- Another limitation is the sample size. Although the sample was sufficient to support adequate exploration of key associations and hypothesis, missing data meant it was probably insufficient for a few associations that approached but did not reach statistical significance (probable Type II errors).

- As the study was conducted in the UK, the external validity may not apply to developing countries, where the vast majority of children with SCD live. Even within the UK, the external validity has to be considered with some caution. For example, the proportion of heads of household in managerial, professional, and intermediate occupations in this study (65.1%) is much higher than the proportion of Black people in the UK in the same occupation bands (45.1% for Black Caribbean and 37.1% for Black Africans) (NOS 2005). This suggests the respondents came from families that were more economically advantaged than average back families in the UK.

- Another threat to the external validity is the observation in Table 4.2 that the respondents experienced three times more admissions in the past year compared with non respondents. This suggests that the respondents may have been more unwell than typical children with SCD in the community.

- Lack of a control group for the psychological measures was a limitation but this was mitigated by good use of epidemiological and other comparative data, which allowed a meaningful comparison to be made.
• The use of questionnaires to assess psychological status is a limitation as they do not assess psychopathology but are very useful as screening instruments. Also the reliance on young people’s self report and lack of data triangulation from parents and teachers is a limitation. There was data triangulation with hospital records but this was limited.

Summary

Despite the limitations noted above, this study achieved its objectives and two of the four hypotheses were supported by the data. The findings provide robust evidence to support the application of stigma theory to SCD. The study showed that when asked directly, young people with SCD do not appear to perceive stigma as much as would be predicted but indirect questioning indicates higher levels of perceived stigma. The finding that self-perceived stigma predicts psychological difficulty in SCD is important as this has not been shown previously in SCD. On the whole, young people with SCD in the sample did not experience depressive symptoms more than peers in the community. However, children with SCD appear to be at risk of anxiety symptoms (emotional subscale of SDQ has mainly anxiety symptoms) when compared with general population norms and there appears to be a subgroup of young people with SCD who are at increased risk of depression. The latter include those with more severe disease, who easily perceive stigma, come from families with unhealthy function, and those with negative attitude towards SCD. Children with SCD who have these risk factors should be targeted for screening for psychological and emotional difficulties. Most children with SCD who were depression-positive were not receiving counselling. This suggests a need for an effective system to target psychological intervention towards children with SCD most in need. The characteristics outlined earlier could form the basis for targeting such services.
Chapter 7

Conclusions, clinical and research implications

The bio-medical, psychosocial, and economic burden of SCD on some affected persons and their families is already very high. This study has contributed to a better understanding of how self-perceived stigma could be adding to this burden.

This study set out to explore the extent to which young people with SCD perceive being stigmatised and the associations between this perception and psychosocial outcomes. The methodology proved successful in answering the research objectives. The measurements demonstrated good evidence of reliability in data collection. Also the evidence supports some external validity for the findings. The study supported two of the four hypotheses including a demonstration that the theoretical constructs in stigma theory are applicable to SCD. Some of the findings are unique as they have not been shown previously in SCD. The conclusions from the study are outlined below.

These are followed by clinical and research recommendations.

7.1. Conclusions.

The following hypotheses were supported by the data from this study:

1. Measures of disruptiveness (e.g. frequency of admissions) and visibility (e.g. presence of leg ulcer) will significantly and independently predict levels of self perceived stigma. This is the first time stigma theory has been successfully applied to SCD.

2. Self perceived stigma will significantly and independently predict Total difficulties score on the Strengths and Difficulties Questionnaire (SDQ)

Two hypotheses were not supported by the data:

1. Self perceived stigma will significantly and independently predict Depressive symptoms as measured by SMFQ
2. Self perceived stigma will significantly and independently predict Self Esteem measured by Rosenberg Scale

Other important findings include:

- Only 15% of the respondents met the study criteria for directly measured self-perceived stigma. However, indirect assessment of self perceived stigma based on disclosure practices (e.g. keeping SCD secret) suggests a much higher level of perceived stigma (57%) in the current study.

- When directly measured, fewer young people with SCD in this study appeared to perceive stigma compared with young people with epilepsy and stuttering. However, when indirectly measured, a similar proportion of young people in all three groups appear to perceive stigma.

- Compared with SDQ UK norms, young people with SCD in this study scored significantly higher on the emotional subscale of the SDQ. The prevalence of SDQ-caseness (caseness for psychological difficulty) among the group (15.4%) is slightly higher compared with young black boys and girls in London (9.2% and 10.9% respectively) but the difference was not statistically significant (p=0.078).

- Compared with Black children in London, the children with SCD in this study did not have increased risk of depressive disorder. However, based on regression analysis, a subgroup of children with SCD (e.g. those with unhealthy family function, increased frequency of ward admissions (surrogate index of severity), and who have negative attitude towards SCD) were found to be more vulnerable to risk of depression and need to be targeted for screening and interventions.

- The young people with SCD in the study had normal levels of self esteem but low self-esteem was associated with female gender, having a negative attitude to SCD, and coming from a family with unhealthy function. Association with Sickle Cell Society was linked with higher self esteem. Self esteem and depression were strongly associated and the inclusion of depression in the model nullified the predictive ability of gender and negative attitude towards SCD.

- The children with SCD had limited access to counselling as only 11.5% of the sample was in receipt of counselling. Also the availability of counselling did not reflect need. For example, only 18.8% of the children who were classified as depression-positive were in receipt of counselling. Also there was no statistically
significant difference in receipt of counselling between the depression-positive and depression-negative cases.

- The presence of leg ulcer appears to be a useful marker for both illness severity and increased risk of psychosocial distress.

- Negative attitude toward illness predicted all the three psychosocial outcomes variables (i.e. psychological difficulty (SDQ), depression, and to a less extent, self esteem). Unhealthy family function, predicted both depression and self esteem.

7.2 Recommendations.
I have made two sets of recommendations. First for clinical interventions, and secondly for further research needs.

7.2.1. Clinical
SCD is a chronic and potentially challenging illness; hence affected children require optimum support to adjust to living with the disorder. In developed countries, advances in physical care have led to tremendous improvement in life expectancy, quality of life, and to some extent improvement in psychosocial adjustment.

However, there appear to be sub-groups of children with SCD who are still at increased risk of psychological and emotional difficulties. These include young people who perceive SCD-related stigma, females, those with leg ulcer and more severe illness, who have negative attitude to having SCD, and or come from families with unhealthy function. Young people with these features need to be targeted for mental health screening and intervention.

There is need to resource more psychological services to support children with SCD. However, such services need to use the markers of increased risk of psychological and emotional difficulties identified in this study to screen children at risk and target their interventions more effectively and efficiently.

The evidence-base for the types of psychological interventions that are effective in SCD is still limited (Anie and Green 2002). Pending the availability of better evidence, it would be pragmatic and appropriate to consider interventions that first target those at
risk of psychiatric disorder (e.g. abnormal SDQ or SMFQ scores) who may be at more immediate need and may be keener for help. Secondly, it may be helpful to specifically address or incorporate the psychosocial risk variables identified in this study (e.g. self-perceived stigma, negative attitude towards SCD, and family dysfunction) as targets for intervention. For example, in relation to family dysfunction, it is likely that the benefit of a psychological intervention for a young person with SCD would be enhanced if his or her family were to be supported simultaneously.

In relation to self-perceived stigma, this study shows that not every young person with SCD experience self-perceived stigma. However, for those who do, it is associated with significant psychological difficulty. It is recognised that while some aspects of self-perceived stigma could originate from actual experience of discrimination, other components could be based on unfounded or exaggerated fears of discrimination. Indeed self-perceived stigma in SCD has the potential to become a self-fulfilling prophesy whereby the fear of discrimination leads the young person to avoid peers thereby missing opportunities to test out whether in fact their fears of discrimination would have happened. For this reason, it is possible for self perceived stigma to be modified with a cognitive behavioural intervention (CBT). CBT is an effective therapy where beliefs driving a particular distress are not founded on fact or reality. The technique could involve the use of behavioural experiments to disconfirm the fears driving self-perceived stigma. This type of intervention can help affected young people to develop resilience.

In addition, given that some self-perceived stigma may be driven by actual experience of enacted stigma caused by discriminatory or prejudicial behaviour by other people, the case is made for a public education and anti-stigma campaign to improve public understanding and attitude towards SCD. The campaign could initially be targeted at communities or places with many young people with SCD. Greater understanding of SCD by the public could lead to greater sensitivity in interactions with people with SCD, which could improve their psychosocial outcomes.

The finding that respondents recruited from the Sickle Cell Society had higher self esteem than those recruited from haematology clinics provides impetus for the advocacy and psychosocial support provided by the organisation to people with SCD. While bearing in mind the caveat of reverse causality, this finding could be seen as
providing some evidence that the organisation’s stated goal of improving the self-esteem of affected young people is being achieved. It may therefore be appropriate to young people with SCD to be encouraged to avail themselves of the opportunities provided by the Sickle Cell Society.

7.2.2. Research
This study has pioneered the exploration of stigma and related concepts and experiences in children with SCD. While the study has answered some important research questions with clinical and policy implications, it has also generated new questions worth further exploration.

This study found low endorsement of self perceived stigma by young people with SCD when asked directly and more admission to the phenomenon when asked indirectly (through disclosure practices). This suggests that the findings may have been confounded by socially desirable responding, which is a recognised limitation of attitudinal surveys. It would therefore be helpful to extend this study and incorporate measures of socially desirable responding and compare children with SCD with children with other conditions known to be potentially stigmatising (e.g. epilepsy).

Future studies of this nature would be strengthened if the assessment of psychopathology is based on standardised psychiatric interview rather than questionnaires. It would be further enhanced if in addition to self report; information on psychopathology is sourced from teachers and parents to allow triangulation.

This study found for the first time that self-perceived stigma independently predicted psychological difficulty in SCD. It is likely that this association operates through intermediate factors such as self-isolation by people with high perceived stigma. In order to understand this association fully and to exploit the potential therapeutic gains embedded in it, further studies are required to identify the intermediate factors linking self-perceived stigma and psychological difficulty in SCD.

Negative attitude towards SCD was a consistent predictor of psychological and emotional outcomes in this study. However, there is limited understanding of the determinants of attitude towards having SCD. This warrants further exploration to
understand how negative attitude towards illness is generated and maintained. Such studies could unlock hitherto unknown therapeutic strategies for young people with SCD.

7.3. Concluding remark
The physical and psychosocial outcomes for children with SCD in developed countries have improved significantly. However, the outlook in poor developing countries remains desperate as highlighted by a recent study from Kenya. Williams and colleagues showed that most of the 90% of children with SCD who die even before diagnosis could be saved if diagnosed early and given existing vaccines against the common bacterial infections responsible for their mortality (Williams et al 2009). Thus there is an urgent need for more investment in the care of children with SCD in poorer countries to reduce this avoidable waste of human life. However, more resources may not be the solution if the additional resources are not equitably and efficiently utilised. The situation of children with SCD in Cuba illustrates this. Despite limited resources, the organisation of health care in Cuba has enabled people with SCDs to achieve good outcomes compared with other poor countries (Aguila et al 2008). Finally, this study has shown that despite access to advanced medical treatment and better psychosocial support, a minority of young people with SCD in developed countries may still be at risk of psychological and emotional difficulties. It is therefore important to have systems for identifying and providing appropriate psychological interventions for this subgroup.
References


Smith JA (1991) What do we know about the clinical course of sickle cell disease? *Seminar in Hematology*, 28(3) 209-12

Sosan, O. 2006. Stigmatisation of Sickle Cell Anaemia: Young Women's willingness to consider a male Sickle Cell sufferer as a potential life partner. *Dissertation submitted in partial fulfilment of the requirement for a Masters Degree in Health Community and Development. London School of Economics and Political Science*


http://apps.who.int/gb/ebwha/pdf_files/WHA59/A59_9-en.pdf (accessed 5/2/10)


**ONLINE RESOURCES**


http://www.dcsf.gov.uk/rsgateway/DB/SFR/s000832/SFR03_2009NationalTablesv2.xls (accessed 5/2/10)


http://www.sdqinfo.com/bba1.pdf  (accessed 5/2/10)


http://www.statistics.gov.uk/CCI/nugget.asp?ID=1166 (accessed 5/2/10)

http://www.statistics.gov.uk/cci/nugget.asp?id=1163 (accessed 5/2/10)
Appendix 1

(Content of postal/research pack)

a. Invitation letters for Parents Version 003
b. Information Sheet for Parents/Guardians Version 003
c. Information Sheet for Young People Version 003
d. Consent Form for Parents/Guardian Version 003
e. Assent Form for Children Version 003
f. Slip for telephone interview
g. Study questionnaire
Appendix 1a - Invitation letters for Parents Version 003
Appendix 1e - Assent Form for Children Version 003
Appendix 1g – Study Questionnaire Version 004
Appendix II

Ethics approval

a. MREC Approval letter
b. R&D Approvals from Central Middlesex, North Middlesex, and St Mary’s Hospitals
c. Notice of Substantial Amendment
d. Information Sheet before “title” was amended.
Appendix IIa - MREC Approval letter
Appendix IIb - R&D Approvals from Central Middlesex, North Middlesex and St Mary’s Hospitals
Appendix IIc - Notice of Substantial Amendment
Appendix IIId - Information Sheet for Parents/Guardians Version 002 before “title” was amended.
Invitation letter for parents/guardians to be sent with questionnaire

Dear Sir/Madam,

Invitation to take part in a research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems.

The Sickle Cell Society or your Sickle Cell Clinic sent this letter to you on our behalf.

We would like to invite your child to take part in a research project. The purpose of the research is to see whether young people with sickle cell disease feel stigmatised (treated differently by other people) and whether this affects how they feel about themselves. The information we gather from the research could help improve the support we provide young people with sickle cell disease. Those who agree to take part will be asked to complete a questionnaire, which will take about 20 minutes.

Before you decide for your child to take part, it is important for you to understand why the research is being done and what it will involve. This is all explained in the enclosed information leaflets for you and your child. Please take time to read the information carefully and discuss it with other people if you wish. Also ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish for your child to take part. Please ask you child to read his or her own Information leaflet and to discuss it with you and other people if they wish to.

If having read the information leaflet and you and your child are happy to take part, please do the following:

1. Sign both copies of the Consent Form
2. You and your child should sign both Copies of the Assent Form.
3. Ask your child to complete the Questionnaire
4. Keep one copy each of the signed Consent Form and Assent Form for your records.
5. Put all the other signed Forms and the Questionnaire in the stamped and addressed envelope we provided (you do not need to put any more stamp)
6. Put the envelope in a Post Box

We appreciate the time involved in completing the questionnaire and we will offer £10 worth of shopping vouchers to every young person who completes one.

If you do not wish for your child to take part in the research, please do not complete the Forms and Questionnaire. Instead, return them (uncompleted) to us in the envelope we provided. We will not send you any reminders once we receive the uncompleted Forms and Questionnaires. Your decision not to take part will not affect the standard of care your child will receive.
The Organisation that sent you this letter on our behalf has **not** given us your name, address or clinical details. We will only know the name of people who return completed questionnaires to us.

Thank you for reading this.

Yours sincerely

Dr Cornelius Ani  
Honorary Lecturer and Specialist Registrar in Child and Adolescent Psychiatry
Information Sheet for Parents/Guardians

Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems.

What is this about?
Your child is being invited to take part in a research project. Before you decide it is important for you to understand why the research is being done and what it will involve.

Before you decide for your child to take part, it is important for you to understand why the project is being done and what it will involve. Please take time to read the following information carefully and discuss it with other people if you wish. Also ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish your child to take part. This research is being done for an educational project and will involve completing a questionnaire.

What is the purpose of the project?
Some people do not understand sickle cell disease and how it affects people who have it. It is possible that such people may treat people with sickle cell disease differently for having the disease. This is what we refer to as someone feeling stigmatised. The purpose of this research is to see whether young people with sickle cell disease feel stigmatised by other people and whether this affects how they feel about themselves.

Why have my child been chosen?
All young people with sickle cell disease aged between 11 and 18 years and who live in London are being invited to participate. We hope that about 133 young people will take part.

Does my child have to take part?
No, it is up to you and your child to decide whether or not to take part. You are both free to decide whether or not to take part. You are both free to decide to withdraw from the research at any time and without giving a reason. Your decisions about this will not affect the standard of care your child will receive.

What will happen to my child if we agree to take part?
If you are happy to for your child to take part, and are satisfied with our explanations, you will be asked to sign a consent form. If your child is able to understand the research and is happy to take part and can write their name, they will be asked to sign an “assent” form with you, if they want to. You will be given a copy of the signed information sheet and consent/assent forms to keep for your records.
What does my child have to do if we agree to take part?
Your child will be given a questionnaire to complete. The questionnaire will take about 20 minutes to complete. They can complete it on their own and return it to us in a stamped envelope, which we will give you. Alternatively, they can complete it in the clinic, where Dr Cornelius Ani will be available to help if he or she wants. Finally, he or she can choose for one of us to telephone him/her and complete the questionnaire for them over the phone. Dr Ani will also go through the medical records of 20 persons who take part to compare some of their responses to the information in their medical records. We appreciate the time involved in completing the questionnaire and we will offer £10 worth of shopping vouchers to every young person who completes one. Twenty participants will be asked to complete a second copy of the same questionnaire 2 weeks after completing the initial questionnaire. The second questionnaire will also take about 20 minutes to complete. A second offer of £10 worth of shopping vouchers will be made to the 20 participants who complete the second questionnaire. We expect that your child will complete the questionnaire himself/herself although he/she could ask you for help in remembering factual information to help them answer the questions. If your child prefers to complete the questionnaire over the phone, please return the enclosed Slip indicating a preferred phone number and time when we could contact you.

What are the possible disadvantages and risks of taking part?
Apart from the 20 minutes or so it will take to complete the questionnaire; we do not envisage any risks or disadvantages to your child.

What are the possible benefits of taking part?
The information we get might help improve our understanding of whether young people with sickle cell disease feel stigmatised and how this might be affecting them. This could help improve the support we provide young people with sickle cell disease.

What if something goes wrong?
If you are not happy and wish to complain about any aspect of the way you or your child have been approached or treated during the course of this research, the normal National Health Service complaints procedure should be available to you. It is unlikely, but if your child is harmed by taking part in this project, you may be entitled to compensation.

Will my child’s taking part in this study be kept confidential?
If you agree for your child to take part, his/her records may be inspected as part of the research. Your child’s name, however, will not be disclosed outside the hospital. All information, which is collected, about your child during the course of the research will be kept strictly confidential. The information will be kept securely at the Academic Unit of Child and Adolescent Psychiatry, Imperial College London for 15 years. Any such information, which leaves the hospital, will have your child’s name and address removed so that they cannot be recognised from it.
What will happen to the results of the research study?
We hope to complete the study within the next 2 years after which you will be informed about the results. The results will be published in a medical journal so that other professionals working with young people with sickle cell disease can learn from our research. The individuals who took part in the study will NOT be identified in any report/publication about the project.

Who is organising the research?
The research is organised by a team of doctors from Imperial College and Central and North West London Mental Health NHS Trust. None of the doctors involved in the research will benefit financially from your participation.

Who has reviewed the study?
This research study has been reviewed and approved the South West Multicentre Research Ethics Committee.

Contact for Further Information
If you would like any further information about the research, please contact Dr Cornelius Ani or Dr Matthew Hodes at Academic Unit of Child and Adolescent Psychiatry, Imperial College London, St Mary's Campus, Norfolk Place, London W2 1PG, Tel: 02078861145  Fax: 0207886 6299, e-mail: c.ani@imperial.ac.uk

Thank you for reading this and considering taking part in this study. You will be given a copy of this information sheet and a signed consent form to keep for your records.
Patient Identification Number:

**CONSENT FORM (for parents/guardians)**

**Title of Project:**
Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems.

Name of Researcher:

Please initial box

1. I confirm that I have read and understand the information sheet dated ............................ (version ............) for the above study and have had the opportunity to ask questions.

2. I understand that my child’s participation is voluntary and that we are free to withdraw at any time, without giving any reason, without my child’s medical care or legal rights being affected.

3. I understand that sections of any of my child’s medical notes may be looked at by Dr Cornelius Ani or responsible individuals from regulatory authorities where it is relevant to my child taking part in research. I give permission for these individuals to have access to my child’s records.

4. I agree that my child may take part in the above study.

_________________________ ________________ ____________________
Name of Parent/ guardian Date Signature

_________________________ ________________ ____________________
Name of Person taking consent Date Signature (if different from researcher)

_________________________ ________________ ____________________
Researcher Date Signature

1 for patient; 1 for researcher; 1 to be kept with clinical notes
ASSENT FORM FOR CHILDREN
(to be completed by the child and their parent/guardian)

Title of Project:
Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems.

Child (or if unable, parent on their behalf) /young person to circle all they agree with please:

Have you read (or had read to you) the Information Sheet about this project?     Yes/No
Do you understand what this project is about?                Yes/No
Have you asked all the questions you want?       Yes/No
Have you had your questions answered in a way you understand? Yes/No
Do you understand it’s OK to stop taking part at any time without giving reason? Yes/No
Are you happy to take part?                Yes/No

If any answers are ‘no’ or you don’t want to take part, don’t sign your name!

If you do want to take part, please write your name and today’s date

Your name       ___________________________
Sign                      ___________________________
Date                       ___________________________

Your parent or guardian must write their name here too if they are happy for you to do the project

Print Name  ___________________________
Sign               ___________________________
Date              ___________________________

The doctor who explained this project to you needs to sign too:

Print Name    ___________________________
Sign               ___________________________
Date              ___________________________

Thank you for your help.

1 copy for patient; 1 for researcher; 1 copy to be kept with clinical notes
Slip to be completed and returned by participants who prefer to be interviewed over the phone

Title:
Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems.

Dear Participant,

If you would prefer for Dr Ani to contact you and arrange for you to complete the research questionnaire over the phone, please provide the information requested below and return this slip in the stamped envelope provided.

I would like to complete the Questionnaire over the phone (tick)  

My name is ____________________________________________________________

My preferred telephone number to be contacted on is ________________________________

My preferred day of the week to be contacted is ________________________________

My preferred time to be contacted is ________________________________

Thank you.

Please put this Slip in the envelope provided and place it in a Post Box. Dr Ani will then contact you on the number and at the preferred day and time you indicated.
Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems.

Thank you for your help in completing this questionnaire. The questions measure a variety of attitudes, feelings, and behaviours about yourself and other people. There are no right or wrong answers so we would be grateful if you could answer as honestly as you can. The answers are completely confidential. Please contact Dr Ani or Dr Hodes on 02078861145 if you are unsure about how to complete the questionnaire. We will call you back and explain. It would help us a lot if you answered all questions even if you are not absolutely certain or the question seems daft!

Once again, thank you for your help.

Please tell us your name and address below. This will enable us to contact you to let you know the results of the research study. This first page, which identifies you, will be removed from the rest of the Questionnaire and kept secure in a safe place.

Name:________________________________________

Address (including post code): _________________________________
QUESTIONNAIRE

Date of completing questionnaire: __________________

Where was the questionnaire completed?        At Home □
                                                In Clinic (name) □
                                                Over the telephone □

What is your Date of Birth? ______________________________

Are you a boy or a girl?    Boy □    Girl □

Which of your parents live at home?  Both natural parents □
                                      Mother only □
                                      Father only □
                                      Mother + stepfather □
                                      Father + stepmother □
                                      None (I live on my own) □

What work do your parents’ do?

Father  _______________
Mother  _______________
Stepfather  _______________
Stepmother  _______________

How many brothers and sisters do you have?

Natural brothers and sisters________
Half brothers and sisters _________

Do any of your brothers or sisters also suffer from Sickle Cell Disease?

Yes □
No □

How many people live in your house or flat?  _________________

How many bedrooms does your house or flat have?  _________________
Does your family own a car?
- Yes ☐
- No ☐

Does your family have a BT or Cable telephone line (not mobile phone)?
- Yes ☐
- No ☐

How would you describe your Ethnic Group?
- Black British ☐
- Black African ☐
- Black Caribbean ☐
- Mixed Race ☐
- Black other ☐
- Mediterranean ☐
- Asian ☐

Were you born in the UK?
- Yes ☐
- No ☐

If you were not born in the UK, how long have you lived in the UK?
_______________

Are you still in school?
- Yes ☐
- No ☐

If you are in school, how many days have you had off school in the past school year?
- None ☐
- Less than 7 days ☐
- 7-14 days ☐
- 15-21 ☐
- 22-28 days ☐
- More than 28 days ☐

Do you have a best friend?
- Yes ☐
No ☐

How many other friends do you have that are so close to you that you sleep over at each other's house? _________________________________

How many other friends do you have that are so close to you that you can confide in him/her? _________________________________

Do you think that having Sickle Cell affects whether people want to be friends with you?

Never ☐
Rarely ☐
Sometimes ☐
Often ☐

Do you think that having Sickle Cell affects whether people like you or not?

Never ☐
Rarely ☐
Sometimes ☐
Often ☐

Do you think that having Sickle Cell affects whether or not you are invited to people's homes or to parties?

Never ☐
Rarely ☐
Sometimes ☐
Often ☐

Do you keep your sickle cell a secret from others?

Often ☐
Sometimes ☐
Rarely ☐
Never ☐

How often do you talk to people about your sickle cell?

Often ☐
Sometimes ☐
Rarely ☐
Never ☐
Do any of your friends know that you have sickle cell?
   All ☐
   Some ☐
   Few ☐
   None ☐

When people find out you have sickle cell, is it usually because:
   You tell them? ☐
   They see you have a sign of sickle cell and then you explain? ☐
   Someone else tells them about it? ☐
The following questions are to help us understand you and your family. By “Family” we mean those individuals with whom you usually live and have the strongest emotional ties. For each of the following statements, please tick the box that most applies to you and your family.

<table>
<thead>
<tr>
<th></th>
<th>Strongly Agree</th>
<th>Agree</th>
<th>Disagree</th>
<th>Strongly Disagree</th>
</tr>
</thead>
<tbody>
<tr>
<td>Planning family activities is difficult because we misunderstand each other</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>In times of crises we can turn to each other for support</td>
<td></td>
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<tr>
<td>We cannot talk to each other about the sadness we feel</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Individuals are accepted for what they are</td>
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</tr>
<tr>
<td>We avoid discussing our fears and concerns</td>
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<tr>
<td>We can express feelings to each other</td>
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<tr>
<td>There is lots of bad feelings in the family</td>
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<tr>
<td>We feel accepted for what we are</td>
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<td></td>
</tr>
<tr>
<td>Making decisions is a problem for our family</td>
<td></td>
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</tr>
<tr>
<td>We are able to make decisions on how to solve problems</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>We don’t get along well together</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>We confide in each other</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Thank you for getting this far in completing the questionnaire. Please remember that there are no right or wrong answers and all answers are completely confidential to the researchers. Your answers will not affect you in any way whatsoever.

Over the past year, how often have you had sickle cell pain?
- More than once a week
- Once a week
- Two times a month
- Once a month
- Once every 2-4 months
- Two times in the year
- Once in the year
- No pain in the past year

If you had sickle cell pain in the past year, how would you describe the average intensity of the painful episodes?
- Mild
- Moderate
- Intense
- Very intense
- I did not have pain in the last year
Over the past year, how many times have you been admitted for one night or more in a hospital ward due to problem of sickle cell?
   No admission in the past year □
   Once □
   2-4 times □
   5-6 times □
   7-10 times □
   More than 10 times □

Over the past year, how many times have you been to Accident and Emergency (A & E) Department due to problem of sickle cell but not admitted to a hospital ward?
   No visit to A & E in the past year □
   Once □
   2-4 times □
   5-6 times □
   7-10 times □
   More than 10 times □

Which of the following signs of sickle cell do you have at present?
   Jaundice (yellow eyes) □
   Leg ulcer □

Are you prescribed Penicillin Tablets?
   Yes □
   No □

If you answered "Yes", how often do you forget to take Penicillin Tablets?
   I often forget to take the tablet □
   I sometimes forget to take the tablet □
   I usually remember to take the tablet □

Are you prescribed Folic Acid Tablets?
   Yes □
   No □
If you answered “Yes”, how often do you forget to take Folic Acid Tablets?
   I often forget to take the tablet ☐
   I sometimes forget to take the tablet ☐
   I usually remember to take the tablet ☐

Are you prescribed Hydroxyurea Tablets?
   Yes ☐
   No ☐

If you answered “Yes”, how often do you forget to take Hydroxyurea Tablets?
   I often forget to take the tablet ☐
   I sometimes forget to take the tablet ☐
   I usually remember to take the tablet ☐

How often do you remember to attend your appointments at the Sickle Cell Clinic?
   I usually remember to attend ☐
   I sometimes forget to attend ☐
   I often forget to attend ☐

Are you receiving any regular counselling from a therapist?
   Yes ☐
   No ☐

For each item, please mark an ‘x’ in the box for Not True, Somewhat True or Certainly True. Please give your answer on the basis of how things have been for you over the last 6 months.

<table>
<thead>
<tr>
<th></th>
<th>Not True</th>
<th>Somewhat True</th>
<th>Certainly True</th>
</tr>
</thead>
<tbody>
<tr>
<td>I try to be nice to other people. I care</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Statement</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>--------------------------------------------------------------------------</td>
<td>---</td>
<td>---</td>
<td>---</td>
</tr>
<tr>
<td>about their feelings</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am restless, I cannot stay still for long</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I get a lot of headaches, stomach-aches or sickness</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I usually share with others (food, games, pens etc)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I get very angry and often lose my temper</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am usually on my own. I generally play alone or keep to myself</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I usually do as I am told</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I worry a lot</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am helpful if someone is hurt, upset or feeling ill</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am constantly fidgeting or squirming</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I have one good friend or more</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Statement</td>
<td>Not True</td>
<td>Somewhat True</td>
<td>Certainly True</td>
</tr>
<tr>
<td>--------------------------------------------------------------------------</td>
<td>----------</td>
<td>---------------</td>
<td>----------------</td>
</tr>
<tr>
<td>I fight a lot. I can make other people do what I want</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>I am often unhappy, down-hearted or tearful</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other people my age generally like me</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am easily distracted, I find it difficult to concentrate</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am nervous in new situations. I easily lose confidence</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am kind to younger children</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am often accused of lying or cheating</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other children or young people pick on me or bully me</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I often volunteer to help others (parents, teachers, children)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I think before I do things</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I take things that are not mine from home, school, or elsewhere</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I get on better with adults than with people my own age</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I have many fears, I am easily scared</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I finish the work I'm doing. My attention is good</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Please place an ‘x’ in the column, which you think most nearly applies to you.

<table>
<thead>
<tr>
<th>Statement</th>
<th>Strongly Agree</th>
<th>Agree</th>
<th>Disagree</th>
<th>Strongly Disagree</th>
</tr>
</thead>
<tbody>
<tr>
<td>On the whole, I am satisfied with myself</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>At times I think that I am no good at all</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I feel that I have a number of good qualities</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I am able to do things as well as most people</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I feel I do not have much to be proud of</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I feel useless at times</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I feel that I am a person of worth, at least on an equal plane with others</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I wish I could have more respect for myself</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>All in all I am inclined to feel that I am a failure</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>I take a positive attitude towards myself</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Please place an ‘x’ in the box, which you think most nearly applies to you.

1. How good or bad do you feel it is that you have sickle cell disease?
   - Very good □
   - A little good □
   - Not sure □
   - A little bad □
   - Very bad □

2. How fair is it that you have sickle cell disease?
   - Very fair □
   - A little fair □
   - Not sure □
   - A little unfair □
   - Very unfair □

3. How happy or sad is it for you to have sickle cell disease?
   - Very sad □
   - A little sad □
   - Not sure □
   - A little happy □
   - Very happy □

4. How bad or good do you feel it is to have sickle cell disease?
   - Very good □
   - A little good □
   - Not sure □
   - A little bad □
   - Very bad □
5. How often do you feel that your sickle cell disease is your fault?
   Never □ 
   Not often □ 
   Sometimes □ 
   Often □ 
   Very often □ 

6. How often do you feel that your sickle cell disease keeps you from doing things you like to do?
   Very often □ 
   Often □ 
   Sometimes □ 
   Not often □ 
   Never □ 

7. How often do you feel you will always be sick?
   Never □ 
   Not often □ 
   Sometimes □ 
   Often □ 
   Very often □ 

8. How often do you feel that your sickle cell disease keeps you from starting new things?
   Very often □ 
   Often □ 
   Sometimes □ 
   Not often □ 
   Never □
9. How often do you feel different from others because of your sickle cell disease?
   - Never
   - Not often
   - Sometimes
   - Often
   - Very often

10. How often do you feel bad because you have sickle cell disease?
    - Very often
    - Often
    - Sometimes
    - Not often
    - Never

11. How often do you feel sad about being sick?
    - Never
    - Not often
    - Sometimes
    - Often
    - Very often

12. How often do you feel happy even though you have sickle cell disease?
    - Never
    - Not often
    - Sometimes
    - Often
    - Very often

13. How often do you feel just as good as other kids your age even though you have sickle cell disease?
    - Very often
    - Often
    - Sometimes
    - Not often
    - Never
This form is about how you might have been feeling or acting recently. For each item, please tick how much you have felt or acted this way in the past two weeks.

If a sentence was true about you most of the time, tick true. If it was only sometimes true, tick sometimes. If a sentence was not true about you, tick not true.

<table>
<thead>
<tr>
<th>Statement</th>
<th>True</th>
<th>Sometimes</th>
<th>Not True</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. I felt miserable or unhappy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. I didn't enjoy anything at all</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. I felt so tired I just sat around and did nothing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. I was very restless</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. I felt I was no good any more</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. I cried a lot</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. I found it hard to think properly or concentrate</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. I hated myself</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. I was a bad person</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10. I felt lonely</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11. I thought nobody really loved me</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>12. I thought I could never be as good as other kids</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13. I did everything wrong</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Please read each of the following statements carefully and indicate in the appropriate box how much this is like you.
I worry about what other people will think of me even when I know it doesn’t make any difference
  Not at all like me ☐
  Slightly like me ☐
  Moderately like me ☐
  Very much like me ☐
  Extremely like me ☐

I am unconcerned even if I know people are forming an unfavourable impression of me
  Not at all like me ☐
  Slightly like me ☐
  Moderately like me ☐
  Very much like me ☐
  Extremely like me ☐

I am frequently afraid of other people noticing my shortcomings
  Not at all like me ☐
  Slightly like me ☐
  Moderately like me ☐
  Very much like me ☐
  Extremely like me ☐

I rarely worry about what kind of impression I am making on someone
  Not at all like me ☐
  Slightly like me ☐
  Moderately like me ☐
  Very much like me ☐
  Extremely like me ☐
I am afraid that others will not approve of me
   Not at all like me □
   Slightly like me □
   Moderately like me □
   Very much like me □
   Extremely like me □

I am afraid that others will not approve of me
   Not at all like me □
   Slightly like me □
   Moderately like me □
   Very much like me □
   Extremely like me □

I am afraid that people will find fault with me
   Not at all like me □
   Slightly like me □
   Moderately like me □
   Very much like me □
   Extremely like me □

Other people’s opinions of me do not bother me
   Not at all like me □
   Slightly like me □
   Moderately like me □
   Very much like me □
   Extremely like me □

When I am talking to someone, I worry about what they may be thinking about me
   Not at all like me □
   Slightly like me □
   Moderately like me □
   Very much like me □
Extremely like me □

I am usually worried about what kind of impression I make
Not at all like me □
Slightly like me □
Moderately like me □
Very much like me □
Extremely like me □

If I know someone is judging me, it has little effect on me
Not at all like me □
Slightly like me □
Moderately like me □
Very much like me □
Extremely like me □

Sometimes I think I am too concerned with what other people think of me
Not at all like me □
Slightly like me □
Moderately like me □
Very much like me □
Extremely like me □

I often worry that I will say or do the wrong things
Not at all like me □
Slightly like me □
Moderately like me □
Very much like me □
Extremely like me □

In the last 12 months, have you been attacked for reasons to do with your race or colour?
Yes □
No □

If yes, was this verbal abuse
Yes □
No □

Have you experienced a direct physical attack?
Yes □
No □

Have you experienced destruction or vandalism of your property?
  Yes □
  No □

Do you think there are employers in Britain who would refuse a job to a person because of his or her race, or colour, religion or cultural background
  Yes □
  No □

If so, do you think this is true of
  Most employers □
  Half of employers □
  Fewer than half of employers □
  Hardly any employers □

THE END

THANK YOU VERY MUCH FOR YOUR TIME.
THIS IS THE END OF THE QUESTIONNAIRE

Now, please return the completed questionnaire to the person who gave it to you or in the return envelope provided. Your £10 shopping voucher will be posted to you once we receive your completed questionnaire.

If you found answering parts of the questionnaire made you feel worried or upset, you can discuss this with your parents, or arrange to discuss it with your Sickle Cell Counsellor or GP. You can also phone NHS Direct (08454647) or ChildLine (08001111) for advice.
South West Multi-centre Research Ethics Committee

11 April 2006

Dr Cornelius Ani
Specialist Registrar and Honorary Lecturer in Child and Adolescent Psychiatry
Central and North West London Mental Health NHS Trust
5 Collingham Gardens
London
SW5 0HR

The Lescaze Offices
Shinner's Bridge
Dartington
Devon
TQ9 6JE
Tel: 01803 861947
Fax: 01803 861914
Email: swmrec@sw-devon-ha.swest.nhs.uk

Dear Dr Ani,

Full title of study: Self-perceived Stigma and Fear of Negative Evaluation (FNE) in young people with sickle cell disease: Associations with psychosocial distress

REC reference number: 06/MRE06/10

Thank you for your letter of 04 April 2006, responding to the Committee’s request for further information on the above research and submitting revised documentation.

The further information has been considered on behalf of the Committee by members of the committee.

Confirmation of ethical opinion

On behalf of the Committee, I am pleased to confirm a favourable ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised.

Ethical review of research sites

The Committee has designated this study as exempt from site-specific assessment (SSA. There is no requirement for [other] Local Research Ethics Committees to be informed or for site-specific assessment to be carried out at each site.

Conditions of approval

The favourable opinion is given provided that you comply with the conditions set out in the attached document. You are advised to study the conditions carefully.

Approved documents

The final list of documents reviewed and approved by the Committee is as follows:

<table>
<thead>
<tr>
<th>Document</th>
<th>Version</th>
<th>Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>Application</td>
<td>5.0</td>
<td>12 January 2006</td>
</tr>
<tr>
<td>Investigator CV</td>
<td></td>
<td>16 January 2006</td>
</tr>
<tr>
<td>Protocol</td>
<td>002</td>
<td>16 January 2006</td>
</tr>
<tr>
<td>Covering Letter</td>
<td></td>
<td>16 January 2006</td>
</tr>
</tbody>
</table>
Research governance approval

You should arrange for the R&D department at all relevant NHS care organisations to be notified that the research will be taking place, and provide a copy of the REC application, the protocol and this letter.

All researchers and research collaborators who will be participating in the research must obtain final research governance approval before commencing any research procedures. Where a substantive contract is not held with the care organisation, it may be necessary for an honorary contract to be issued before approval for the research can be given.

Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees (July 2001) and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

06/MRE06/10 Please quote this number on all correspondence

With the Committee's best wishes for the success of this project

Yours sincerely

John Alexander
Chair

/\ Jeffrey Stark
(Assistant Administrator)
Enclosures:  Standard approval conditions SL-AC2

Copy to:  Ms Maria Tsappis  
West London R&D Consortium  
Research & Development Office  
Trust Headquarters, St Bernards Wing  
Uxbridge Road  
Southall  
UB1 3EU
NHS Management Approval

To:       Dr Cornelius Ani (cc Dr Jo Howard)
From:    Dr Alan Wames (R&D Manager)
Date:  18/10/2006

Project: Self-perceived Stigma and Fear of Negative Evaluation (FNE) in young people with sickle cell disease: Associations with psychological distress (Reference: 06/MRE06/10)

I understand that you have recently received a favourable ethical opinion for the above project, with the condition that you do not undertake research in an NHS organisation until relevant NHS management approval has been received. I am therefore writing on behalf of the North West London Hospitals Trust to inform you that the project has also been approved by the Trust and may now proceed.

To maintain Trust approval for the above project, all staff involved within it must adhere to Trust and Research Governance Framework requirements (see www.nwlh.nhs.uk/research). As Chief/Principal Investigator you are required to formally advise the R&D Office of ANY changes to the project including:

- Any changes to the status of the project, e.g. abandoned, completed etc
- Any changes to the protocol – however minor.
- Any changes to the funding arrangements

You are also required to:

- Notify, in a timely fashion, the R&D adverse Events relating to the Research and the appropriate urgent safety measures taken in line with ICH GCP requirements.
- Ensure that the R&D Office has copies of all annual and final progress reports.
- Ensure all researchers involved in the project hold the necessary expertise required and have Honorary Contracts should they need to.
- Ensure adequate and accurate reporting and monitoring of said project.
- Co-operate with all internal Trust monitoring and auditing procedures.
- Where the Trust has agreed to be sponsor to the project, you should be aware and comply with the required responsibilities.

Failure to comply with these conditions may result in Trust approval being rescinded.

Yours sincerely,
Dr Tracy Assari  
Research Governance Co-ordinator  
Research and Development Office  
Institute of Child Health (ICH) and  
Great Ormond Street Hospital for Children (GOSH) 30 Guilford Street  
London WC1N 1EH  
Ext: 2845  
Tel:+44 (0)20 7905 2845  
Fax:+44 (0)20 7905 2201  
Email: t.assari@ich.ucl.ac.uk  
Web: http://www.ich.ucl.ac.uk/
01 October 2008

Professor Irene Roberts
Professor of Paediatric Haematology
The Bays Building
St. Mary’s Hospital
London W2 1NY

Dear Irene,

**Project Title:** Self-perceived Stigma and Fear of Negative Evaluation (FNE) in young people with sickle cell disease: Associations with psychosocial distress

**R&D reference number:** 08/GD/011  **Ethics reference number:** 06/MRE06/10

**Principal Investigator:** Professor Irene Roberts,

I confirm that this project has now been approved by the Research & Development Department. The project may now start at Imperial College Healthcare NHS Trust sites. Please note that the start date of the project is the date of this letter and the duration is the same as that provided in your application form.

Before you commence your research, please note that you must be aware of your obligations to comply with the minimum requirements for compliance with the Research Governance indicators 17 (Data Protection); 25 (Health and Safety) and 22 (Financial Probit). Details of the requirements to be met can be found in the Trust Research Governance Policy in the current R&D Annual Report, R&D Management Office, or on the Trust Intranet under Research & Development. Please also refer to the Research Governance Framework available on [www.dh.gov.uk](http://www.dh.gov.uk).

Under the Research Governance regulations, Serious Adverse Event Reports, Adverse Reactions and amendments to the protocol or other supporting documents must be forwarded to the Research & Development Office and Ethics Committee.

In accordance with the Research Governance Framework, research projects carried out in the Trust will be randomly chosen by the R&D team for auditing. Please see the attached checklist for documentation that will be required during the audit.

I wish you well in your research.

Yours sincerely,

[Signature]

Professor Myra McClure
Research and Development Management Office
St Mary’s Hospital
NOTICE OF SUBSTANTIAL AMENDMENT

For use in the case of all research other than clinical trials of investigational medicinal products (CTIMPs). For substantial amendments to CTIMPs, please use the EU-approved notice of amendment form (Annex 2 to ENTR/CT1) at http://eudract.emea.eu.int/document.html#guidance.

To be completed in typescript by the Chief Investigator in language comprehensible to a lay person and submitted to the Research Ethics Committee that gave a favourable opinion of the research (“the main REC”). In the case of multi-site studies, there is no need to send copies to other RECs unless specifically required by the main REC.


Details of Chief Investigator:

Name: Dr Cornelius Ani
Address: Academic Unit of Child and Adolescent Psychiatry
         Imperial College London
         St Mary’s Campus
         Norfolk Place
         London W2 1PG

Telephone: 02078861145
Email: c.ani@imperial.ac.uk
Fax: 02078866299

Full title of study: Self-perceived stigma and Fear of Negative Evaluation (FNE) in young people with sickle cell disease: Associations with psychosocial distress

Name of main REC: South West Research Ethics Committee
REC reference number: 06/MRE06/10
Date study commenced: 31st October 2006
Type of amendment (indicate all that apply in bold)

(a) Amendment to information previously given on the NRES Application Form

   No

   If yes, please refer to relevant sections of the REC application in the “summary of changes” below.

(b) Amendment to the protocol

   No

   If yes, please submit either the revised protocol with a new version number and date, highlighting changes in bold, or a document listing the changes and giving both the previous and revised text.

(c) Amendment to the information sheet(s) and consent form(s) for participants, or to any other supporting documentation for the study

   Yes

   If yes, please submit all revised documents with new version numbers and dates, highlighting new text in bold (done)

Is this a modified version of an amendment previously notified to the REC and given an unfavourable opinion?

   No
Summary of changes

Briefly summarise the main changes proposed in this amendment using language comprehensible to a lay person. Explain the purpose of the changes and their significance for the study. In the case of a modified amendment, highlight the modifications that have been made.

If the amendment significantly alters the research design or methodology, or could otherwise affect the scientific value of the study, supporting scientific information should be given (or enclosed separately). Indicate whether or not additional scientific critique has been obtained.

In the course of recruitment, we have received feedback to suggest that the project title “Research project to examine whether young people with sickle cell disease feel shame and embarrassment as a result of their health problems”, which is on our recruitment documents were found to be “too negative” by some users because of the phrase “shame and embarrassment”. This may have contributed to our low response rate.

In order to address this, we propose to amend the project title by replacing the phrase “made to feel shame and embarrassment” with “treated differently by other people”. The new title will then read “Research project to examine whether young people with sickle cell disease feel they are treated differently by other people as a result of their health problems”.

We believe the amended title would be more acceptable to the young people and their families while still accurately conveying the essence of the project.

We do not believe that this amendment significantly alters the research design or methodology, or has the potential to affect the validity of our study.

Any other relevant information

Applicants may indicate any specific ethical issues relating to the amendment, on which the opinion of the REC is sought.

We do not believe that this amendment raises any specific ethical issues.

List of enclosed documents

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Notice of amendment (non-CTIMP), version 3.1, November 2005
Declarations

- I confirm that the information in this form is accurate to the best of my knowledge and I take full responsibility for it.
- I consider that it would be reasonable for the proposed amendment to be implemented.

Signature of Chief Investigator: ...........................................

Print name: Dr Cornelius Ani

Date of submission: 24/05/07
Information Sheet for Parents/Guardians

Research project to examine whether young people with sickle cell disease feel shame and embarrassment as a result of their health problems.

What is this about?
Your child is being invited to take part in a research project. Before you decide it is important for you to understand why the research is being done and what it will involve.

Before you decide for your child to take part, it is important for you to understand why the project is being done and what it will involve. Please take time to read the following information carefully and discuss it with other people if you wish. Also ask us if there is anything that is not clear or if you would like more information. Take time to decide whether or not you wish your child to take part. This research is being done for an educational project and will involve completing a questionnaire.

What is the purpose of the project?
Some people do not understand sickle cell disease and how it affects people who have it. It is possible that such people may behave in ways that make people with sickle cell disease feel shame for having the disease. This is what we refer to as someone feeling stigmatised. The purpose of this research is to see whether young people with sickle cell disease feel stigmatised by other people and whether this affects how they feel about themselves.

Why have my child been chosen?
All young people with sickle cell disease aged between 11 and 18 years and who live in London are being invited to participate. We hope that about 133 young people will take part.

Does my child have to take part?
No, it is up to you and your child to decide whether or not to take part. You are both free to decide whether or not to take part. You are both free to decide to withdraw from the research at any time and without giving a reason. Your decisions about this will not affect the standard of care your child will receive.

What will happen to my child if we agree to take part?
If you are happy to for your child to take part, and are satisfied with our explanations, you will be asked to sign a consent form. If your child is able to understand the research and is happy to take part and can write their name, they will be asked to sign an “assent” form with you, if they want to. You will be given a copy of the signed information sheet and consent/assent forms to keep for your records.
What does my child have to do if we agree to take part?
Your child will be given a questionnaire to complete. The questionnaire will take about 20 minutes to complete. They can complete it on their own and return it to us in a stamped envelope, which we will give you. Alternatively, they can complete it in the clinic, where Dr Cornelius Ani will be available to help if he or she wants. Finally, he or she can choose for one of us to telephone him/her and complete the questionnaire for them over the phone. Dr Ani will also go through the medical records of 20 persons who take part to compare some of their responses to the information in their medical records. We appreciate the time involved in completing the questionnaire and we will offer £10 worth of shopping vouchers to every young person who completes one. Twenty participants will be asked to complete a second copy of the same questionnaire 2 weeks after completing the initial questionnaire. The second questionnaire will also take about 20 minutes to complete. A second offer of £10 worth of shopping vouchers will be made to the 20 participants who complete the second questionnaire. We expect that your child will complete the questionnaire himself/herself although he/she could ask you for help in remembering factual information to help them answer the questions. If your child prefers to complete the questionnaire over the phone, please return the enclosed Slip indicating a preferred phone number and time when we could contact you.

What are the possible disadvantages and risks of taking part?
Apart from the 20 minutes or so it will take to complete the questionnaire; we do not envisage any risks or disadvantages to your child.

What are the possible benefits of taking part?
The information we get might help improve our understanding of whether young people with sickle cell disease feel stigmatised and how this might be affecting them. This could help improve the support we provide young people with sickle cell disease.

What if something goes wrong?
If you are not happy and wish to complain about any aspect of the way you or your child have been approached or treated during the course of this research, the normal National Health Service complaints procedure should be available to you. It is unlikely, but if your child is harmed by taking part in this project, you may be entitled to compensation.

Will my child’s taking part in this study be kept confidential?
If you agree for your child to take part, his/her records may be inspected as part of the research. Your child’s name, however, will not be disclosed outside the hospital. All information, which is collected about your child during the course of the research will be kept strictly confidential. The information will be kept securely at the Academic Unit of Child and Adolescent Psychiatry, Imperial College London for 15 years. Any such information, which leaves the hospital, will have your child’s name and address removed so that they cannot be recognised from it.
What will happen to the results of the research study?
We hope to complete the study within the next 2 years after which you will be informed about the results. The results will be published in a medical journal so that other professionals working with young people with sickle cell disease can learn from our research. The individuals who took part in the study will NOT be identified in any report/publication about the project.

Who is organising the research?
The research is organised by a team of doctors from Imperial College and Central and North West London Mental Health NHS Trust. None of the doctors involved in the research will benefit financially from your participation.

Who has reviewed the study?
This research study has been reviewed and approved the South West Multicentre Research Ethics Committee.

Contact for Further Information
If you would like any further information about the research, please contact Dr Cornelius Ani or Dr Matthew Hodes at Academic Unit of Child and Adolescent Psychiatry, Imperial College London, St Mary's Campus, Norfolk Place, London W2 1PG, Tel: 02078861145 Fax: 0207886 6299, e-mail: c.ani@imperial.ac.uk

Thank you for reading this and considering taking part in this study. You will be given a copy of this information sheet and a signed consent form to keep for your records.