

**Health-Related Quality of Life and Health Behaviours in Children and Adolescents
with Sickle Cell Disease**

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Abstract

Sickle cell disease (SCD) is an inherited blood disorder. Physical health problems include painful vaso-occlusive crises. Children and adolescents with SCD may also experience psychological distress, social isolation and an impaired health-related quality of life (HRQL). Physical health problems and psychosocial factors may be managed, to a certain degree by engaging in daily health behaviours and exacerbated by engaging in risky behaviours. However, children's ability to engage in healthy behaviours may also be undermined by their condition, social influences, their environment and psychological factors. These behaviours have rarely been studied in paediatric SCD populations and both health behaviours and HRQL have rarely been examined in SCD by drawing on a theoretical approach. Therefore, the overall aim of this thesis was to gain a better understanding of HRQL by applying Gap theory (GT; which suggests that HRQL is based on the discrepancy between current and ideal self) and of health behaviours drawing on constructs from the Theory of Planned Behaviour (TPB). It also aimed to examine whether there is a relationship between health behaviours and HRQL. The research comprised two qualitative studies (one with children and healthy siblings using drawings and interviews and one with adolescents and healthy siblings using focus groups) and two quantitative studies using questionnaires focusing on children, adolescents and their parents. The findings from Study 1 suggested that children with SCD and healthy siblings had some discrepancies in different areas of their HRQL (physical vs. psychosocial domains respectively) but that this did not seem to manifest in adolescence (Study 2). Health behaviours were especially influenced by people in their lives and children and adolescents used some health behaviours to cope with their condition (Studies 1 and 2). In Study 3 GT was not applied and instead current perceived HRQL was predicted by greater disease

severity (i.e. fewer days missed from school as well as crises only in adolescents) and increased moderate exercise only in children. There was no difference in HRQL between adolescents and children but there was some underestimation of their HRQL among their parents. In Study 4 the TPB constructs were more consistent at predicting behaviours in children than adolescents. Children's weekly exercise was predicted by children's TPB beliefs (although unexpectedly, more perceived barriers) and parental subjective norms (SN), whereas in adolescents, exercise was predicted by male gender and parental attitude. Child and parent attitude were also the key predictors of child/adolescent water consumption. Adolescents alcohol use was predicted by their attitude, older age, and unexpectedly, more perceived barriers. The combined findings of the thesis contribute towards a more comprehensive understanding of HRQL and health behaviours in SCD.

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In loving memory of Dad

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Preface

Sickle Cell Disease (SCD) is one of the most prevalent genetic haemoglobinopathies which, due to migration, has become widespread in England, with the majority of sufferers living in London (Streetly, Latinovic, Hall, & Henthorn, 2009; Streetly, Latinovic, & Henthorn, 2010). During childhood common clinical manifestations include severe pain syndromes such as vaso-occlusive crisis, stroke, infection and growth failure (Bennett, 2011; Kanter & Kruse-Jarres, 2013), whilst in adolescence additional symptoms may include delayed puberty and leg ulcers (Knight-Madden et al., 2011; Serjeant & Serjeant, 2001). A plethora of quantitative research has consistently reported that children and adolescents with SCD have a lower perceived current health-related quality of life (HRQL) in comparison to their healthy peers (Panepinto & Bonner, 2012). However, with a few exceptions, this research rarely uses qualitative methodology (e.g. Panepinto, Torres, & Varni, 2012; Stegenga, Ward-Smith, Hinds, Routhieaux, & Woods, 2004) or healthy siblings as a comparison group (Hijmans et al., 2010). Furthermore, it is not based on a theoretical approach to HRQL. Gap theory (GT; Calman, 1984) suggests that HRQL is the discrepancy between individuals' perceived expectations and their perceived current experience. GT has been applied in a single study in paediatric SCD (Constantinou, Payne & Inusa, 2015).

SCD is generally managed at home and children/adolescents continue to attend school/college but clinical manifestations and impaired HRQL could potentially be controlled and improved to some extent through daily health behaviours like drinking more water and avoiding strenuous exercise and alcohol use (Brown, 2012; NHS Choices, 2019). There has been little research on health behaviours in SCD (e.g. Dyson et al., 2010a; Karlson et al., 2017; Melo et al., 2018; Omwanghe et al., 2017) and even fewer

studies have explored the association between health behaviours and HRQL in SCD (e.g. Ahmed et al., 2015; Wrotniak, Schall, Brault, Balmer, & Stallings, 2014; Zempsky et al., 2013). Furthermore health behaviours in children/adolescents with SCD have never been considered drawing on a theoretical perspective like the Theory of Planned Behaviour (TPB) which aids understanding of the beliefs that determine behaviours (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975).

Therefore, the aim of this thesis was to gain a better understanding of HRQL using a theoretical approach (GT; Calman, 1984) and of health behaviours (diet, water consumption and exercise levels) and also risky behaviours (alcohol use, cigarette smoking and drug use) by drawing on the TPB (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975). Both qualitative and quantitative methodology was adopted. The qualitative research also included a comparison group of healthy siblings, who themselves may suffer from psychosocial problems (e.g. Hijmans et al., 2009; Juanita Lee, Phoenix, Brown, & Jackson, 1997; Treiber, Mabe, & Wilson, 1987) and the quantitative research also included parents as they play a pivotal role in children's/adolescents' lives and in making decisions about their health and care.

Chapter 1 will begin by reviewing the literature on children's and adolescents' HRQL, health behaviours and risky behaviours. More specifically, this chapter will present a comprehensive review of quantitative and qualitative HRQL literature in children/adolescents with SCD and healthy siblings including factors that may determine HRQL, such as disease severity, and provide support for the use of GT (Calman, 1984). Further, it will provide a detailed account of how children's daily health behaviours can affect SCD (e.g. drinking water may help to avoid or treat a vaso-occlusive crisis) but, in turn, how their condition can influence their engagement in health behaviours (e.g. children may participate in fewer activities and exercise). This chapter will then describe

how the TPB has been applied to help understand children's and adolescents' health/risky behaviours (e.g. healthy eating, beverage consumption, exercise and alcohol use) as well as providing an overview of other determinants of these behaviours such as demographic factors. Next, there will be a brief examination of literature that has linked better management of health behaviours and avoiding risky behaviours to HRQL. Finally, the methodological approach of this thesis (i.e. exploratory sequential mixed methods) will be described as well as research to support the choice of qualitative methods.

The first two studies in this thesis used qualitative methods. Chapter 2 presents the first of these studies focusing on children aged 5 to 12 years old with SCD and healthy siblings (Study 1). The first study used a drawing task and semi-structured interviews with a sample of 18 children with SCD and 14 healthy siblings. This explored HRQL and health behaviours (diet, water consumption and exercise), including any discrepancies between their current and ideal self (based on GT) of children with SCD compared to healthy siblings.

Chapter 3 presents the second qualitative study focusing on adolescents aged 13 to 17 years old (Study 2). This study used focus groups with a sample of 23 adolescents with SCD and 21 healthy siblings to explore the same areas as Study 1, but also looked at adolescents' risky behaviours (alcohol use, cigarette smoking and drug use).

The final two studies in this thesis involved quantitative research. Chapter 4 describes a questionnaire based study with four groups of participants; 106 children with SCD and 106 of their parents and also 96 adolescents with SCD and 96 of their parents. The main aim of this study was to examine the predictors (e.g. demographic indicators, coping, disease severity measures, health/risky behaviours) of child/adolescent self-reports and parent proxy reports of current perceived HRQL. The third study also explored

differences between child self-reports, adolescent self-reports and parent proxy-reports of current perceived HRQL.

Chapter 5 describes another questionnaire based study (Study 4) using the same sample as Study 3. The fourth study primarily investigated the predictors (e.g. demographic indicators, disease severity measures and child and parent health beliefs) of children's and adolescents' water consumption and exercise levels and also adolescents' alcohol use. This study also explored similarities and differences between child, adolescent and parental reports of health beliefs and health behaviours.

The final chapter of this thesis begins by summarising the four studies (Chapter 6). The main findings and contributions of the research are then discussed in the context of the wider literature. Reflexivity in relation to the qualitative research is presented, followed by the limitations and strengths of the four studies. The chapter concludes by discussing the implications of the present research for practice and suggests areas for future research.

Chapter 1. Literature review

1.1 Health-related quality of life (HRQL)

1.1.1 Introduction

The first section of this chapter reviews sickle cell disease (SCD) and how children (aged 12 or below) and adolescents (aged 13 to 18) with this condition may suffer from physical and psychosocial problems as a result of their condition. There is evidence that shows that these problems are related to lower perceived health-related quality of life (HRQL) reports from children and parents, and therefore this is the main focus of this section. At present, questionnaires are the most prevalent method of ascertaining the HRQL of children and adolescents with SCD; however these measures generally examine their perceived current-self and they are not based on a theoretical framework. Adopting a gap approach, by examining perceived current-self in relation to perceived-ideal self, will be shown to be an appropriate approach. Gap approaches have been applied to chronically ill children and adolescents; although there is no validated multidimensional measure. In contrast to the proliferation of questionnaire studies, there are a limited number of qualitative HRQL studies in SCD. Qualitative methodology may be useful in identifying different areas of HRQL that are considered to be important to children and adolescents with SCD, to better understand their experiences and also to gauge the usefulness of applying a gap approach to exploring HRQL in this population. Furthermore, siblings of children and adolescents with SCD may also suffer from psychosocial problems, and have a comparable HRQL to their affected sibling, although they do not have SCD. They also reflect their socio-demographic characteristics (ethnicity and socio-economic status; SES). Therefore, healthy siblings may act as a comparison group and this will also be discussed.

1.1.2 Sickle cell disease (SCD)

SCD is one of the most common inherited haemoglobinopathies (blood disorders) (Bennett, 2011). The condition is the result of a single-point mutation in the haemoglobin molecule where a single amino acid is substituted in the sixth position of the beta chain valine for glutamic acid (e.g. Connes, Machado, Hue, & Reid, 2011) as first demonstrated by Pauling, Itano, Singer and Wells (1949). Affected alleles are referred to as the sickle haemoglobins S (HbS) (Kanter & Kruse-Jarres, 2013). Haemoglobin proteins are found in red blood cells and are responsible for carrying oxygen from the lungs to tissues and for carrying carbon dioxide from the tissues to the lungs e.g. gaseous exchange (Thomas, Wilson-Barnett, & Goodhart, 1998). HbS polymerises (combines) in deoxygenated conditions (low oxygen) leading to deformed rigid, sticky crescent shaped red blood cells; thus the label *sickle* cell (Ilesanmi, 2013).

Individuals inherit different haemoglobin genotypes from their parents (Brown, 2012). Haemoglobin A (HbA) is the most common genotype found in healthy individuals (Brown, 2012). Some individuals may inherit one normal HbA gene from one parent and abnormal HbS e.g. sickle gene from the other parent. These individuals would be referred to as having sickle cell trait (SCT) e.g. phenotypes HbAS. They may not generally have any symptoms of the condition themselves but there is a chance that their child could inherit SCT (2:4) or sickle cell anaemia (SCA) (1:4) if their partner also had the HbAS gene or they could inherit the normal haemoglobin phenotype e.g. HbAA (1:4). Therefore, families affected by SCD are likely to include children who do not have any variant of the condition.

There are four major phenotypes of SCD: HbSS, HbSC, HbS β^+ , and HbS β^0 , although many more exist (Bennett, 2011). Phenotype HbSS (SCA) is the most common and severest form of SCD (Frenette & Atweh, 2007; Weatherall, 2010). HbS β^0 is also a

severe form of SCD whereas HbS β + may be moderate or mild; both phenotypes are significantly less common (Frenette & Atweh, 2007; Weatherall, 2010). HbSC disease is a mild form of the condition but the second most common variant of SCD (Frenette & Atweh, 2007; Weatherall, 2010). Individuals with severe forms of SCD are more likely to experience more frequent disease complications like vaso-occlusive crises and also more severe anaemia (Dampier et al., 2010) which will be discussed further later. In England, SCD occurs in one in every two thousand live births (Streetly, Latinovic, Hall, & Henthorn, 2009; Streetly, Latinovic, & Henthorn, 2010). Moreover, over three quarters of children with SCD born in England reside in London (Streetly et al., 2008; Streetly et al., 2010). It is therefore a prevalent condition. The four major phenotypes of SCD affect males and females equally, with over a third of children coming from single-parent or divorced families who are typically of lower SES (Ilesanmi, 2013). In England, SCD is most prevalent among individuals of Black African and Black Caribbean ethnicities who mostly have phenotypes HbSS and HbSC (Streetly et al., 2010).

Sickle red blood cells pathology leads to vaso-occlusive crises which are the most prevalent complication of the condition (Ballas et al., 2010). Vaso-occlusive crises are characterised by a sudden onset of severe, acute painful episodes that can occur throughout the body including bones, muscles and organs and are often described as continuous and throbbing in nature (Darbari, Ballas, & Clauw, 2014; Kanter & Kruse-Jarres, 2013). These episodes commonly last for hours to days, and in rarer instances weeks and can be extremely painful and debilitating (Ballas et al., 2010; Knight-Madden, Lewis, Tyson, Reid, & MooSang, 2011). Individuals with SCD frequently report recurrent vaso-occlusive pain in multiple body sites (commonly in the extremities and hips) which is often managed by taking medication at home (Dampier, Ely, Brodecki, & O'Neal, 2002a; 2002b). Vaso-occlusive crises may be caused by physical or mental stress, temperature extremes, poor

health behaviours and risky behaviours amongst other triggers (Brown, 2012; Knight-Madden et al., 2011). More recently, Dampier et al., (2016) found that being an adolescent and female was associated with greater sickle pain interference, for example, with engagement in social, physical and recreational activities.

Individuals with SCD may also experience other physical health problems some of which are more prevalent in different stages of development (Kanter & Kruse-Jarres, 2013; Knight-Madden et al., 2011). During childhood and adolescence they are more likely to suffer from vaso-occlusive crises, stroke, infection, acute chest syndrome, growth failure and developmental delay (Bennett, 2011; Kanter & Kruse-Jarres, 2013; Knight-Madden et al., 2011). Moreover, during adolescence other complications may arise including delayed puberty, priapism (i.e. persistent and painful erection of the penis) and leg ulcers (Knight-Madden et al., 2011; Serjeant & Serjeant, 2001). The condition is also associated with co-morbidities such as asthma (Anim, Strunk, & DeBaun, 2011) and pulmonary hypertension (Colombatti et al., 2010). Despite these complications, survival rates have improved with most children and adolescents with SCA or phenotype HbS β^0 and milder forms of the condition living into adulthood (93.9% and 98.4% respectively) (Quinn, Rogers, McCavit, & Buchanan, 2010). Today, there is even a cure for SCD; haematopoietic stem cell transplantation (HSCT; Chaturvedi & DeBaun, 2016) which has an overall survival rate of 93% to 97% (Khoury & Abboud, 2011). However, HSCT is not available for everyone because a fully matched sibling donor is needed and it can lead to increased morbidities and mortality (Oringanje, Nemecek, & Oniyangi, 2009).

Some of the major physical health problems related to SCD can be prevented or treated through immunization programs, routine out-patient clinic appointments, annual transcranial Doppler ultrasonography to assess risk of stroke (Adams et al., 1997) and chronic blood transfusions to reduce this risk (Abboud & Atweh, 2006), and also to

prevent recurrent episodes of acute chest syndrome (Miller et al., 2001). Children and adolescents are also prescribed medication, for example, penicillin prophylaxis is offered to prevent invasive or recurrent bacterial infections, stroke or death (Battersby, Knox-Macaulay, & Carrol, 2010) and hydroxycarbamide is prescribed because of frequent vaso-occlusive crises, episodes of acute chest syndrome or to those who are severely anaemic (Brandow, Jirovec, & Panepinto, 2010; Mueller, 2008). These complications often lead to hospitalisations. Salman and Hassan (2015) found that most hospitalisations in children and adolescents were because of vaso-occlusive crisis (73.84%), and to a lesser extent infections (9.28%) and acute chest syndrome (8.02%) and that they lasted for an average of four days.

1.1.3 Psychosocial impacts of paediatric SCD

1.1.3.1 Introduction

Paediatric populations with SCD are reported to suffer from psychosocial problems, for example, increased depression, low self-esteem, poor body image, fatigue and more social problems compared to healthy children (Anderson, Allen, Thornburg, & Bonner, 2015; Anie, 2005; Edwards et al., 2005; Sehlo & Kamfar, 2015; Ünal, Toros, Kütük, & Uyaniker, 2011). These are a result of more frequent or greater levels of sickle-related pain and hospitalisations and also in adolescents, growth failure and delayed puberty (Anie, 2005; Edwards et al., 2005; Ozer, Yengil, Acipayam, & Kokacya, 2014; Ünal et al., 2011). Furthermore, evidence suggests that children and adolescents with SCD have experienced perceived sickle-related stigmatisation (Adeyemo, Ojewunmi, Diaku-Akinwumi, Ayinde, & Akanmu, 2015; Ani, Garralda, Anie, Wilkey, & Hodes, 2015). SCD also affects children's education leading to poorer academic achievement and lower intellectual ability in comparison to their peers (Berkelhammer et al., 2007) which is

caused by frequent short and long term absence from school (Schwartz, Radcliffe, & Barakat, 2009), increased illness or pain frequency (Schatz, 2004; Schwartz et al., 2009) and growth failure (Puffer, Schatz, & Roberts, 2010). Social-cultural factors including the shared values and circumstances of families with SCD like a reluctance to discuss illness among their community, family or friends, their negative perceptions of healthcare professionals and fear of being seen as drug-seeking and also their often financial hardships may also exacerbate psychosocial issues and lead to poor disease adjustment and coping (Barbarin & Christian, 1999). Thus SCD is likely to have an impact on HRQL but coping strategies may also influence SCD. Coping will be discussed below before the concept of HRQL is introduced.

1.1.3.2 Coping

There are different strategies that children and adolescents with SCD may use to cope with the physical, psychosocial and educational problems related to their condition, especially during a vaso-occlusive crisis. Coping, at a basic level, is when an individual uses their thoughts and behaviours to manage situations that are considered stressful, and is often divided into problem- and emotion-focused coping styles (Lazarus & Folkman, 1984). Both coping styles have been examined in paediatric research (Compas, Connor-Smith, Saltzman, Thomsen, & Wadsworth, 2001; Snooks, 2009). Problem-focused coping is used to change or modify the principal cause of the stress (e.g. a vaso-occlusive crisis) thereby reducing or removing the cause of the stressor (Lazarus & Folkman, 1984; Lazarus, 1991), and may include taking medication and resting during a vaso-occlusive crisis. Emotion-focused coping on the other hand aims to manage a person's feelings when dealing with (actual or perceived) unchangeable stressors which may involve self-reflection and taking control over one's emotions (Snooks, 2009). Examples of emotion-

focused coping include maladaptive and avoidant coping. Maladaptive forms of emotional-focused coping include engaging in risky behaviours such as alcohol use to suppress emotions whereas avoidant coping may include denial of the problem. Emotion-focused coping can also include more adaptive strategies such as seeking social support which can be structural (i.e. networks of close ties like parents and connections) or functional support (i.e. financial help or affection) (Snooks, 2009). Another example of emotion-focused coping is adopting spiritual or religious approaches such as prayer (Snooks, 2009). There is some evidence that religion and spirituality may help children and adolescents with SCD and their parents to cope with the condition by improving pain or symptom management, improving quality of life (QoL), gaining a sense of control or support from pastors/spiritual leaders when sick or in hospital and providing comfort and hope (e.g. Clayton-Jones & Haglund, 2015; Clayton-Jones, Haglund, Belknap, Schaefer, & Thompson, 2016; Sanchez et al., 2015). There have been criticisms of problem-focused and emotion-focused coping because the definitions are considered too broad, that there are too many diverse types of coping styles included in these two categories (Coyne & Gottlieb, 1996) and also that some strategies applied by adolescents could be considered dual-focused coping i.e. include both problem- and emotion-focused intent (Compas, Ey, Worsham, & Howell, 1996). However, this nevertheless provides a useful way of categorising coping and some self-report coping measures in children use this. Reviews have found that there is an abundance of self-report coping measures which have been applied to healthy and chronically ill children and adolescents (Blount et al., 2008; Lemétayer & Chateaux, 2008; Sveinbjornsdottir & Thorsteinsson, 2008).

In children and adolescents with SCD, coping is commonly used as a predictor of pain and psychosocial adjustment (Gil, Abrams, Phillips, & Keefe, 1989) which is often assessed using the Coping Strategy Questionnaire (CSQ; Rosenstiel & Keefe, 1983),

although this is a lengthy measure which includes 80 items and is not suitable for very young children (Compas et al., 2001). Paediatric research in pain has found that maladaptive and avoidant emotion-focused coping (i.e. internalising/catastrophizing and externalising) predicted greater pain intensity, functional disability, depression and emotional distress whereas adaptive emotion-focused strategies such as seeking social support and problem-focused coping resulted in increased pain control and coping effectiveness in children and adolescents with SCD (Ludwig, Sil, Khowaja, Cohen, & Dampier, 2018; Sil, Dampier, & Cohen, 2016) and also in other paediatric populations affected by pain like arthritis and headaches (Blount et al., 2008; Reid, Gilbert, & McGrath, 1998). Some evidence has reported that pain coping strategies remain stable over time for younger children with SCD but that these are more inconsistent for adolescents with SCD (Gil, Thompson, Keith, Tota-Faucette, Noll, & Kinney, 1993; Gil, Wilson, & Edens, 1997b). A recent study established that lower HRQL in children and adolescents with SCD was associated to greater pain intensity and increased participation in emotion-focused coping strategies (Ludwig et al., 2018). Therefore, problem-focused coping strategies and seeking social support are more favourable than maladaptive and avoidant emotion-focused coping strategies in paediatric SCD, and this may be linked to HRQL.

1.3.3.3 Defining HRQL

There is no universal definition of HRQL or QoL (De Civita et al., 2005; Matza, Swensen, Flood, Secnik, & Leidy, 2004). They are both complex terms that are difficult to define (Eiser & Morse, 2001). Both terms are used interchangeably in the literature (Taylor, Gibson, & Franck, 2008); however this is not recommended in paediatric research (Davis et al., 2006), because HRQL is rated lower than overall QoL and different variables predict each concept (Feldman, Grundland, McCullough, & Wright, 2000). Such issues

have led researchers to conduct concept analyses of HRQL and QoL (e.g. Taillefer, Dupuis, Roberge and LeMay, 2003) and develop terms that are more appropriate for children and adolescents (e.g. Taylor et al., 2008). QoL is a broad, multidimensional concept of well-being which is commonly defined by the World Health Organization (WHO) as an; *“individuals’ perception of their position in life in the context of culture and value systems in which they live, and in relation to their goals, expectations, standards and concerns”* (WHOQOL Group, 1995, p.1405). However, the term QoL was derived from adult literature and therefore may not be applicable to children and adolescents with chronic conditions because it fails to consider developmental changes and how children and adolescents construct health and illness (Taylor et al., 2008). In medicine and associated peer-reviewed research, the term HRQL is preferred to QoL (Rapley, 2003). HRQL is a multidimensional type of patient-reported outcome that considers an individual’s health condition, treatment or medical decisions (King & Hinds, 2012; Panepinto, 2012; Taylor et al., 2008). It is widely accepted that HRQL assesses patients physical, social and emotional functioning (Panepinto, O’Mahar, DeBaun, Loberiza, & Scott, 2005; Varni, Seid, & Rode, 1999), although it is also suggested that it assesses school/work functioning (Panepinto & Bonner, 2012) and role functioning (Varni, Burwinkle, Seid, & Skarr, 2003) amongst other things. HRQL (and QoL) is also generally recognised as subjective i.e. an individual’s perception or appraisal of HRQL (Eiser & Morse, 2001; Matza et al., 2004). However, Eiser and Morse (2001) postulate that it can also be objective, i.e. what people can do, which may be important when exploring how a child’s life is affected by a life-long condition. From King and Hinds’ (2012) viewpoint, HRQL is also influenced by individuals’ cultural beliefs including their life goals, values and norms, nevertheless many definitions of HRQL fail to include this. Some researchers have argued that HRQL should examine the discrepancy between an individual’s perceived

current self and their perceived ideal self (Eiser, Cotter, Oades, Seamark, & Smith 1999; Panepinto & Bonner, 2012), and the WHO's definition of QoL also incorporates this, but the majority of research does not do this. A review found that the term HRQL is mostly adopted in paediatric SCD research (e.g. Panepinto and Bonner, 2012). This thesis will adopt the following approach which was concluded from a concept analysis of HRQL in young people with a chronic condition.

HRQL in young people with chronic illness is subjective, multidimensional and dynamic. It is unique to each individual young person and includes aspects of physical, psychological and social function. It is dependent upon not only the stage of development but also the illness trajectory. This involves the achievement of goals and aspirations and the constraints imposed through ill-health and treatment. (Taylor et al., 2008, p. 1831)

1.3.3.4 Gap theory approach to HRQL

A detailed review of research concluded that paediatric HRQL or QoL instruments such as questionnaires are rarely based on a theoretical framework (Davis et al., 2006). Davis and colleagues (2006) identified three theories of HRQL or QoL; utility theory (Torrance, 1987), Lindström's model of QoL (Lindström, 1992) and discrepancy/gap theory (Calman, 1984). The discrepancy theory or a gap approach which measure the discrepancy between individuals' perceived-self and their expectations (Eiser et al., 1999), is consistent with Taylor's et al., (2008) definition of HRQL (and the WHO's definition of QoL) and thus this is the approach that will be evaluated in the research in this thesis to conceptualise HRQL. Eiser, Vance and Seamark (2000) observed that children with chronic conditions often describe what they are able to do and what they would like to do

but are restricted because of their condition or treatment. Therefore, the authors conclude that the discrepancy theory is useful in assessing the affect a condition has on a child's current day-to-day life and how the child would like to change (their expectations) which will be unique for each child (Eiser et al., 2000). However, Davis and colleagues (2006) postulate that the discrepancy theory does not identify the factors that determine whether children are happy with themselves, although adopting a qualitative methodology may address this concern and help to ascertain whether this approach applies to children with SCD.

There are many 'Gap' approaches that focus on psychological issues including Gap theory (GT; Calman, 1984). GT is an extension of Maslow's (1954) Hierarchy of Needs Theory (HNT) which explores human motivation, management training and personal development. The HNT is concerned with allowing individuals to fulfil their potential or self-actualisation (Maslow, 1954). This is achieved when individuals meet their basic biological and survival needs, and then focus on their emotional and social needs (Maslow, 1954). However, GT postulates that in a developed society, perceptions of QoL are less likely to be associated with individuals' basic needs, which are to a large extent met (Calman, 1984). Instead, QoL is more likely to be related to their expectations in life, and also with social comparisons they make with their past achievements (Calman, 1984). Therefore, QoL is defined as what an individual has lost, or needs as opposed to what they have (Calman, 1984). Calman (1984) argued that QoL is the discrepancy, at a certain point in time, between individuals' perceived expectations and their perceived current experience. Calman (1984) postulates that an improvement in QoL occurs when individuals narrow the gap between their expectations or aspirations and perceived-self. According to Calman (1984) improving QoL is not about lowering individuals' expectations, rather it is about making goals realistic, ensuring the 'gap' reduction is

achievable and encouraging individuals' to develop and grow in other ways. For example, Calman (1984) suggests that an individual who is terminally ill cannot realistically hope that their condition will improve. A key idea of GT is that illness and treatment will alter individuals' goals (Calman, 1984). GT is not restricted by an individual's gender or age as it measures individuals' preferred-self (ipsative standard) which remains self-adjusted as opposed to against an objective standard (Bracken, 2009). Calman (1984) applied GT to individuals who suffered from chronic conditions such as cancer. More recently, GT has been used with children and adolescents with SCA in the author's previous research (Constantinou, Payne, & Inusa, 2015), chronic kidney disease (CKD) (Heath et al., 2011) and asthma (Eiser et al., 2000). As the majority of past research using this approach has focused on adults, much of the literature review that follows on HRQL in paediatric SCD does not use a discrepancy or gap measure of HRQL but focuses on perceived current HRQL.

1.1.4 HRQL in paediatric SCD

1.1.4.1 Quantitative research in paediatric SCD

In paediatric HRQL research and clinical practice, questionnaires are implemented with sickle cell populations (Panepinto & Bonner, 2012). HRQL is most commonly obtained from either child self-report or proxy-reports which are often sought from parents using questionnaires (Varni, Burwinkle, & Lane, 2005a). Proxy-reports are when an adult completes a HRQL measure as if they were the child (Theunissen et al., 1998). There are different explanations for obtaining HRQL reports from one or more informants. In some circumstances only adult proxy-reports are used when children are unable to provide useful HRQL self-reports because they are too young, physically ill, too cognitively impaired or fatigued (Wallander & Schmitt, 2001). It is also imperative to ascertain the degree to which

parents can assess children's HRQL because they help to choose the child's treatment and care in paediatric SCD populations along with healthcare professionals (Goldstein & Morewitz, 2011). Obtaining both child self- and parent proxy reports of HRQL is helpful because each informant offers valuable information for healthcare professionals to implement treatments, trial innovative interventions or adapt clinical practice (Wallander & Schmitt, 2001; Majnemer, Shevell, Law, Poulin, & Rosenbaum, 2008).

There are a variety of questionnaires used to assess the HRQL of children with SCD but the most prevalent questionnaire is the Pediatric Quality of Life Inventory (PedsQL™) (Panepinto & Bonner, 2012). The PedsQL™ Measurement Model has generic (Varni, Burwinkle, & Seid, 2005b) and disease specific versions including a SCD version (Panepinto et al., 2013). Generic measures are more appropriate in children with SCD because they attend school regularly and cope with sickle related pain at home and are therefore more likely to compare themselves to healthy children (Dampier et al., 2002b). Also, the disease-specific measure cannot be used to make comparisons with other healthy or chronically ill children. The PedsQL™ is a generic multidimensional HRQL measure assessing physical, emotional, social and school functioning and is available in parallel child and adolescent self-report and parent-proxy report formats and can be administered to healthy and chronically ill patient populations (Varni et al., 2005b). Additionally, the PedsQL™ is able to differentiate between children and adolescents with SCD and those without the condition, those with mild and severe SCD and is responsive to change over time (Brandow, Brousseau, Pajewski, & Panepinto, 2010; Panepinto, Pajewski, Foerster, & Hoffmann, 2008). A review of HRQL in SCD concluded that the PedsQL™ is a feasible, reliable and valid measure and that it is the most appropriate generic measure in this population (Panepinto & Bonner, 2012).

Most past research on HRQL uses measures of perceived current HRQL such as the PedsQL™, rather than a discrepancy measure. Using these measures in SCD, child self-reported HRQL and parent proxy-reported HRQL were lower compared to healthy populations on physical, psychosocial (which includes emotional, social and school functioning) and total HRQL (e.g. Bonner et al., 2010; Dale, Cochran, Roy, Jernigan, & Buchanan, 2011; Panepinto, Pajewski, Foerster, Sabnis, & Hoffmann, 2009). Parent-proxy and child self-reported HRQL agreement was found to be 42 to 49% using the PedsQL™ (Panepinto, Hoffmann, & Pajewski, 2010), with parent-proxy reports generally being lower (Bonner et al., 2010), except on the emotional functioning domain (Bonner et al., 2010; Panepinto et al., 2005; Panepinto et al., 2010). Discordance in child/adolescent-parent agreement in SCD may be related to increased symptoms of parenting distress (Panepinto et al., 2010), disease-related parenting stress (Barakat, Patterson, Daniel, & Dampier, 2008), lower self-reported HRQL reports from mothers (van den Tweel et al. 2008), greater disease severity in children/adolescents (Panepinto et al., 2010) and difficulty in gauging non observable domains of HRQL such as emotional wellbeing (Panepinto et al., 2005; Upton, Lawford, & Eiser, 2008). Furthermore, disagreement in adolescent-parent HRQL reports may be because adolescents are more likely to discuss psychosocial issues and school problems with their friends (Valenzuela et al., 2013). Therefore, researchers propose that both mean differences (using paired samples t-tests) and correlation statistics (often Pearson's correlations) should be implemented to obtain a more comprehensive picture of the relationship between child/adolescent-parent reports of HRQL (Cremeens, Eiser, & Blades, 2006; Panepinto et al., 2005; Upton et al., 2008).

A limited amount of research has applied the gap approach in assessing children's HRQL or QoL (i.e. using a discrepancy measure to examine the discrepancy between their perceived current and ideal selves) but much of this does not focus on children with SCD.

Previous studies using the Exeter HRQL scale (Eiser et al., 1999), Exqol (Eiser et al., 2000) and the ‘How Are You?’ (Le Coq, Colland, Boeke, Bezemer, & van Eijk, 2000) measures have shown that children with asthma had a greater HRQL discrepancy compared to healthy children which suggests that they have a worse HRQL. The authors speculate that this is because children with asthma have equally high expectations than healthy children but lower perceived current self HRQL and therefore there is a larger gap between their current perceived self and ideal self (Eiser et al., 1999).

Contrary to this, Heath et al., (2011) found that children with CKD had a higher perceived current self HRQL and a higher QoL (i.e. smaller discrepancy) compared to healthy children using a discrepancy measure called the Generic Children’s Quality of Life Measure (GCQ; Collier, MacKinlay, & Phillips, 2000). There was no difference between the perceived ideal selves of both groups of children. Therefore, Heath et al., (2011) concluded that children with CKD do not lower their ideal perception of themselves. Similarly, in the current author’s previous research, the self-reported discrepancy QoL of children and adolescents with SCA on the GCQ was not lower compared to a matched healthy sample (Constantinou et al., 2015). Moreover, parent proxy-reported QoL discrepancy scores were not significantly lower compared to child self-reports, although parent proxy-reported perceived current QoL scores were lower than child perceived current QoL scores as previously reported in e.g. Bonner et al.,’s (2010) study.

Thus taking a gap approach in assessing the HRQL of children and adolescents with SCD may be important because it could help explain how individuals with a chronic condition do not necessarily rate themselves to have poor HRQL (Albrecht & Devlieger, 1999). For example, research in adults has demonstrated that women with cancer had lower actual- and ideal-self ratings than women without cancer but were no different in terms of discrepancies between the two and in adjustment. Therefore, they appeared to

have lowered their expectations or had more realistic expectations thereby reducing the discrepancy between their actual and ideal selves (Heidrich & Ward, 1992). Indeed, Heidrich et al., (1994) established that a smaller discrepancy between actual and ideal selves led to better adaptation and less psychological distress in adults with cancer.

Unfortunately, measures of HRQL using a gap approach are limited. For example, current paediatric discrepancy measures do not ascertain multidimensional HRQL (Collier et al., 2000). The GCQ has been validated in children and adolescents with SCA; however it is a unidimensional measure that focuses on generic QoL rather than HRQL and with a few exceptions it has rarely been applied in paediatric populations with chronic conditions (Constantinou et al., 2015; Heath et al., 2011). The Exqol and the Exeter HRQL scale are pictorial computer-delivered measures of QoL that do not have parent proxy versions, are unidimensional measures, have moderate internal reliabilities for discrepancy scores ($\alpha = .50$ and $\alpha = > .64$ respectively) and have only been validated in children with asthma (Eiser et al., 1999; Eiser et al., 2000) and therefore may not be suitable for children and adolescents with SCD. Lastly, the ‘How Are You?’ measure was specially designed for, and validated in children with asthma and healthy children, and while there are generic and parent proxy versions, there has been no published literature to support the validity and reliability of these measures (Le Coq et al., 2000). Therefore, the PedsQL™, which is not a discrepancy measure, remains the most prevalent HRQL measure that has been validated as a child self-report measure and as a parent proxy measure in SCD populations.

In summary, when using a perceived current self measure, HRQL is lower in children/adolescents with SCD than healthy children/adolescents and parent proxy reported HRQL is lower than child self-reported HRQL but this is not the case when using a discrepancy measure of HRQL.

1.1.4.2 Determinants of HRQL

Much quantitative research on HRQL in paediatric SCD using the PedsQL™ examines determinants. A recent systematic review found that child and adolescent self-reports and parent proxy-reports of HRQL are predicted by different demographic indicators, disease related problems like vaso-occlusive crises and psychosocial factors (Ojelabi, Graham, & Ling, 2017). HRQL (total, physical and psychosocial) was lower if the child was older, female, from a single-parent family, had a low neighbourhood SES, low parental education and low or high family income (e.g. Allen et al., 2014; Amr, Amin, & Al-Omair, 2011; Barton-Gooden, Grindley, Knight-Madden, & Asnani, 2019; Constantinou et al., 2015; Palermo, Schwartz, Drotar, & McGowan, 2002; Palermo, Riley, & Mitchell, 2008b; Panepinto et al., 2005).

A review of HRQL literature in paediatric SCD also found that different disease severity measures were examined as determinants, the most common being the frequency of vaso-occlusive crises, hospitalisations (often related to experiencing a vaso-occlusive crisis) and the number of missed days from school related to SCD in the past year (Panepinto & Bonner, 2012). These disease severity measures are often ascertained via parent report or hospital records (Panepinto & Bonner, 2012). Child and adolescent self-report and parent proxy reported total, physical and psychosocial HRQL were lower if they had greater disease severity, such as those who were experiencing vaso-occlusive crises or had more frequent vaso-occlusive crises, had been hospitalised in the last year or had more frequent hospitalisations per year, had missed more days from school, received chronic blood transfusions and had a comorbidity such as asthma (Amr et al., 2011; Barton-Gooden et al., 2019; Brandow et al., 2010; Constantinou et al., 2015; Dale et al., 2011; Dampier et al., 2010; Ozer et al., 2014; Palermo et al., 2002; Panepinto et al., 2005; Schlenz et al., 2012; Sehlo & Kamfar, 2015). Dampier et al., (2010) and Sehlo and Kamfar (2015) also showed that children and adolescents with the most physical effects, had the more

severe forms of SCD (e.g. SCA or HbSB+) and both these children and their parents reported a lower HRQL compared to children with HbSC disease. Despite this, a child's sickle cell phenotype is considered to be a poor measure of disease severity because of the considerable variability within a phenotype. For example, some children with SCA may experience three vaso-occlusive crises a year, be hospitalised and have prolonged periods of missed school whereas other children with SCA may not (e.g. Panepinto et al., 2008; Panepinto et al., 2010). Also, Dampier et al., (2016) postulate that there may be varying disease severity because individuals with more severe forms of SCD (HbSS or HbS β^0) are more likely to receive treatments like hydroxyurea and blood transfusions than those with milder forms (HbSC or HbS β^+), thus improving their symptoms. Additionally, lower reports of HRQL were also associated with complications which are more prevalent with the onset of adolescence, for example, dermatological manifestations (xerosis of skin, loss of hair and yellowish discolouration of the skin) and priapism (Dampier et al., 2010; Pradhan & Nayak, 2014).

Psychosocial problems including internalizing, e.g. depressive and/or anxiety symptoms (which is more prevalent in females; Dampier et al., 2016) and externalizing symptoms, like fatigue and sleep disturbance, were also associated with lower total, physical and psychosocial HRQL reports (Anderson et al., 2015; Long et al., 2008; Palermo & Kiska, 2005; Yengil et al., 2014). Nevertheless, other studies have found that anxiety and depression are not related to HRQL (Ozer et al., 2014). In adolescents with SCD, sickle-related stigma was also associated with lower reports of HRQL (Adeyemo et al., 2015; Wakefield et al., 2017). Thus, there is a myriad of complex disease-related and psychosocial factors that contribute to impaired HRQL which could be taken into account when examining HRQL.

1.1.4.3 Qualitative research in paediatric SCD

The majority of research on HRQL in paediatric SCD (discussed so far) is quantitative, and qualitative methodology has rarely been adopted (Panepinto, Torres, & Varni, 2012; Stegenga, Ward-Smith, Hinds, Routhieaux, & Woods, 2004), although some studies have explored the experiences of children with SCD more generally (i.e. not using a HRQL framework). Qualitative research in this area often focuses on pain rather than the entire experience of living with SCD. In past studies, children and adolescents discussed that pain was unpredictable and recurrent and that it interfered with their daily lives like planning or engaging in social activities and exercise (Atoui et al., 2015; Panepinto et al., 2012; Stegenga et al., 2004). This pain led to psychological distress, fear of dying and social isolation from their peers but children and adolescents appeared to have a good understanding of SCD, including pain triggers such as overexertion and dehydration whilst playing sports (Atoui et al., 2015; Stegenga et al., 2004). In contrast, children's family and friends had poor disease knowledge (Stegenga et al., 2004) which some children described as frustrating (Panepinto et al., 2012). There are also some differences between how children and adolescents with SCD would like to be treated. This is not necessarily because of the condition but may be due to general age differences, whereby adolescents do not want to be singled out whereas younger children may enjoy the attention (Forrester, Barton-Gooden, Pitter, & Lindo, 2015; Stegenga et al., 2004).

Qualitative paediatric research in this area, as well as research on adults, has not adopted a theoretical approach, such as GT, so only investigates children's current perceived selves and not their ideal selves (Panepinto et al., 2012; Stegenga et al., 2004; Strickland, Jackson, Gilead, McGuire, & Quarles, 2001; Thomas & Taylor, 2002). This is the same for the quantitative HRQL research described in this Chapter (section 1.1.4.1). Adopting a gap approach may expand health professionals' and researchers' understanding

of children's experience of SCD. More generally, qualitative research may be useful in identifying different areas of HRQL that are considered to be important to children and adolescents with SCD and also to better understand their experiences. Some of these issues are not ascertained in quantitative research. Therefore, adopting a qualitative methodological approach may give a more complete picture of the variables that shape children's HRQL. Additionally, the qualitative HRQL research that does exist has not compared children and adolescents with SCD to healthy children. Quantitative, questionnaire based research which has done this generally demonstrates that children with SCD have a lower HRQL. It is important to also use qualitative methodology because some issues such as limited social involvement, not wanting to be treated differently from their peers and engaging in healthy behaviours may be prevalent in healthy children and therefore not unique to SCD populations. Healthy siblings of children and adolescents with SCD may act as a comparison group because despite their physical health they may also suffer from psychosocial problems.

1.1.5 HRQL in healthy siblings

A limited number of studies have explicitly examined HRQL among siblings of children and adolescents with SCD and therefore this section also draws on broader literature (i.e. children/adolescents with other chronic conditions). Past research has found that healthy siblings experience similar levels of psychosocial problems to their sibling with SCD and also more psychosocial problems (i.e. more severe externalizing problems like aggressive behaviours) compared to their healthy peers (Hijmans et al., 2009; Juanita Lee, Phoenix, Brown, & Jackson, 1997; Treiber, Mabe, & Wilson, 1987). These psychosocial problems may be related to more frequent emergency room visits with the sibling with SCD (Gold, Treadwell, Weissman, & Vichinsky, 2008b; Gold, Treadwell,

Weissman, & Vichinsky, 2011), greater depression or anxiety among mothers (Treiber et al., 1987) and poorer family functioning, for example, more family conflict (Gold et al., 2011). Therefore, there is some evidence to suggest that healthy siblings experience psychosocial problems and further explorative research in this area may provide a better understanding of their HRQL.

1.1.5.1 Quantitative research in healthy siblings

There are a limited number of quantitative studies investigating HRQL among healthy siblings. However, there was no significant difference between the HRQL of children and adolescents with SCD and healthy siblings in a study by Hijmans et al. (2010). Since there is substantial research reporting that children with SCD have an impaired HRQL (e.g. Panepinto & Bonner, 2012), it is of concern that healthy siblings' HRQL is comparable to that of their affected siblings. In addition, in Hijmans et al.'s (2010) study, both groups had significantly lower HRQL compared to a sample of healthy children, but these were in different areas; younger siblings had lower autonomy, parent relations and financial resources whereas adolescents had lower mood and emotions compared to the sample of healthy children (Hijmans et al., 2010). Hijmans et al., (2010) postulate that adolescents are more resilient and that they develop a better coping style with increasing age. Moreover, the authors suggest that an impaired HRQL in families affected by SCD may be because they are often from single-parent families which are of lower SES (measured by education and employment) (Hijmans et al., 2010), so it may not necessarily be specifically related to SCD. An earlier study in siblings of children with other chronic conditions has shown that the type of illness, its impact on daily living and healthy siblings' age affect their QoL. Havermans, De Croock, Vercruyssen, Goethals and Van Diest (2015) found that the QoL of healthy siblings of children with different chronic

conditions varied depending on the illness. For example, the QoL of siblings of children with cancer was lower compared to healthy siblings of children with cystic fibrosis (Havermans et al., 2015). In addition, Havermans et al., (2015) found that the time since diagnosis of a chronic condition did not affect healthy siblings' QoL. Therefore, a genetic condition such as SCD and a newly diagnosed condition may have similar effects on healthy siblings' QoL. Moreover, the authors postulate that it is the extent of the day-to-day demands of the illness and less obvious issues related to the illness that may affect healthy siblings, although this has rarely been investigated (Havermans et al., 2015). This is supported by Gundlach and colleagues (2006) who have suggested that the greater the impact a chronic illness has on the healthy sibling, such as causing them to worry or having to look after their affected sibling, the poorer their HRQL. A healthy siblings' age also affects their HRQL (Gundlach et al., 2006) or QoL (Houtzager, Grootenhuis, Caron, & Last, 2004; Houtzager, Grootenhuis, Caron, & Last, 2005). Finally, healthy siblings of children with cancer reported an impaired QoL in more areas than adolescents (Houtzager et al., 2004).

1.1.5.2 Qualitative research in healthy siblings

There has been no qualitative research exploring HRQL among siblings of children with SCD, even though there is evidence demonstrating they experience psychosocial problems. There have been a small number of studies in siblings with other chronic, or genetic conditions which have adopted qualitative methodology. Siblings of children and adolescents with inflammatory bowel disease described their fears about the disease and treatment, their parents keeping information about the illness from them and worries that their affected sibling would be bullied because of their appearance (weight gain from steroids or weight loss from disease) (Akobeng et al., 1999). Furthermore, healthy siblings

of children and adolescents with a genetic condition (e.g. haemoglobinopathies, cystic fibrosis, duchenne muscular dystrophy, familial adenomatous polyposis, huntington's disease and neurofibromatosis) were upset by the condition and its effect on them and their parents, although they were relieved that they themselves were unaffected (Plumridge, Metcalfe, Coad, & Gill, 2011). In other research on genetic conditions, they expressed their worry about their siblings' condition including recognising that they may die and that they also felt stigmatised because they were part of a family with a chronic genetic condition (Hutson & Alter, 2007). Moreover, they described feeling invisible since their parents were preoccupied with their affected sibling, whilst they were also concerned about how difficult life was for their parents and affected sibling and did not recognise their own needs (Hutson & Alter, 2007). In a recent study, Tregidgo and Elander (2019) found that siblings of adults with haemophilia described experiencing similar difficulties during their childhoods. Siblings felt as though they did not receive the same amount of attention from their parents as their sibling with haemophilia, whilst this left them feeling left out, it also meant that they experienced more independence and formed closer relationships with other siblings (Tregidgo & Elander, 2019). There were also feelings of resentment, anger and frustration because of their sibling's condition which led to fewer, or disruption, in their own activities or family days and some of these feelings continued into adulthood (Tregidgo & Elander, 2019). In contrast, healthy siblings suggested that their parents had high expectations of their behaviour and academic achievement and therefore healthy siblings may experience more pressure (Hutson & Alter, 2007). Therefore, qualitative evidence suggests that healthy siblings' lives are affected by their siblings' illness and that this may be overlooked by their parents (Akobeng et al., 1999; Plumridge et al., 2011). The experiences of healthy siblings of children/adolescents with SCD may be similar but no

research has explored their HRQL, so the research in this thesis will also include healthy siblings.

This section (1.1) described that SCD can affect the HRQL of children and adolescents with SCD (as well as potentially lead to psychosocial problems for healthy siblings), although some coping strategies may be helpful. The next section will explain how engaging in daily health behaviours, and for adolescents, avoiding risky behaviours, can also be helpful by alleviating some symptoms of SCD and thereby may have an impact on HRQL.

1.2 Health behaviours

1.2.1 Introduction

This section reviews health and risky behaviour research in children and adolescents with SCD. It will first discuss the evidence that engaging in some daily health behaviours such as water consumption and avoiding excessive exercise may be important to help manage SCD, whereas engaging in some risky behaviours such as alcohol use may exacerbate SCD. There are a variety of factors that may influence health and risky behaviours in children and adolescents, such as the influence of family, friends and school, and indeed, SCD itself may sometimes undermine children's ability to engage in certain health behaviours or encourage them to engage in risky behaviours. These determinants, including theoretical approaches (the Theory of Planned Behaviour; TPB) to examining determinants, which have not been used in previous research on health behaviours in children with SCD, will also be reviewed in this section. Finally, managing health behaviours and avoiding risky behaviours may be linked to improved HRQL, as SCD may be better controlled and the symptoms not exacerbated, but the relationship has rarely been investigated. This will also be discussed in this section.

1.2.2 The influence of health behaviours on SCD

1.2.2.1 Water consumption

Water consumption is a health behaviour that can help to manage SCD. There is little research that investigates the prevalence or benefits of water consumption in SCD. Children and adolescents with SCD should drink between one to one and a half times more fluids each day than their healthy peers (Lane et al., 2001); although research suggests they do not drink the recommended amount of water (Fowler et al., 2010). Current evidence found that children with SCD (aged 11 years and over) reported that they drank four glasses of water at home each day over a two week period (Karlson et al., 2017) but Dyson et al. (2010a) found that 46% of 569 children with SCD reported they were prevented from drinking water in class at school. Increased fluids are commonly taken by children with SCD to avoid or treat vaso-occlusive crises (Beyer et al., 1999). Dehydration in individuals with SCD slows down the movement of red blood cells, allowing sickle cells to stick together and blood capillaries to become blocked which triggers vaso-occlusive crises (Brown, 2012; and see section 1.1.2). Water consumption is also used to treat vaso-occlusive crises because it decelerates or stops the sickling process (Okomo & Meremikwu, 2012). Nevertheless, recent evidence substantiated that self-reported water and total fluid intake at home was not related to same day or next day pain in children and adolescents with SCD (Karlson et al., 2017).

1.2.2.2 Diet

There are a limited number of studies examining the daily diet of children or adolescents with SCD, even though poor dietary behaviour has been comprehensively assessed in adolescents with other chronic conditions (Ssewanyana, Nyongesa, van Baar, Newton, & Abubakar, 2017). In one study, parents revealed that their children who have

SCD ate too much junk food; nonetheless the authors did not report how this affected the condition (Ievers-Landis et al., 2001). There are, however many studies which investigate their nutritional intake. Evidence has consistently demonstrated that children and adolescents with SCD have macronutrient deficiencies including consuming fewer proteins, carbohydrates, fats and calcium which are often a consequence of an inadequate diet. For example, children and adolescents with SCD only consumed 20% to 31% of the recommended daily servings of nearly all of the food groups with the exception of the meat group (Williams et al., 1997) and consumed significantly less total calories than the recommended daily requirement (Mandese, Marotti, Bedetti, Bigi, Palazzi, & Iughetti, 2016). Additionally, 57% to 85% of children and adolescents with SCD have micronutrient deficiencies including less folic acid, zinc, magnesium and vitamins D, A and E which is again related to poor diet (Mandese et al., 2016; Martyres et al., 2016). Nutritional insufficiencies/deficiencies have led to more annual hospitalisations (Mandese et al., 2016) and increased vaso-occlusive crises (Martyres et al., 2016). Moreover, deficiencies in macro- and micronutrients are more problematic in children with SCD compared to healthy children because they have greater resting energy expenditure (REE) or resting metabolic rate (RMR). For example, Kopp-Hoolihan, Van Loan, Mentzer and Heyman (1999) concluded that children and adolescents with SCA had a 30% to 50% higher REE compared to healthy populations and Hibbert et al., (2006) found that children with SCD have a 52% higher protein turnover rate compared to their healthy peers. Therefore, pre-pubertal children with SCD need to consume more food than their healthy peers to compensate for their higher RMR (Singhal, Parker, Linsell, & Serjeant, 2002). This is supported by a review which concluded that the Recommended Dietary Allowance (RDA) for the general population is inadequate for the growth and development of individuals

with SCA (Hyacinth et al., 2010). Therefore, they should be consuming more, not less nutrients (Hyacinth et al., 2013).

Good health behaviours such as increasing dietary intake of macro- and micronutrients can also help to manage some aspects of SCD including impaired growth and decreasing the number of vaso-occlusive crises, infection and hospitalisations. According to Zemel (2009) growth impairment is a treatable consequence of SCD. Oral zinc supplements led to increases in height and weight gain (Zemel, Kawchak, Fung, Ohene-Frempong, & Stallings, 2002) and glutamine (e.g. an amino acid) led to a 6% reduction in REE and improved growth (Williams et al., 2004) in children and adolescents with SCA. Addressing delayed growth is imperative because Puffer et al., (2010) established an association between low height and cognitive impairment (e.g. cognitive ability, language ability and academic achievement) and that a higher BMI was related to higher academic achievement in children and adolescents with SCD which is problematic as they have educational problems (see section 1.1.3.1). Diet may help to alleviate some of the physical manifestations of SCD. Magnesium supplements and Vitamins D and A therapy led to fewer acute painful days (De Franceschi et al., 2000; Osunkwo et al., 2012) and a decrease in the number and length of hospitalisations (Brousseau et al., 2004; Schall et al., 2004) in children and adolescents with SCD.

1.2.2.3 Strenuous, moderate and mild levels of exercise

The prevalence of exercise in children and adolescents with SCD is relatively unknown, although a review of health risk behaviours found that reduced levels of exercise has been measured extensively in adolescents with other chronic conditions (Ssewanyana et al., 2017). Health organisations recommend that healthy children and adolescents should participate in 60 minutes of moderate (e.g. walking to school or cycling on level ground) to

vigorous (e.g. running or football) exercise every day which on three days a week should include exercise for strong muscles and bones, such as hopping and skipping and gymnastics (NHS, 2018; World Health Organization, 2019). Recent evidence substantiated that fewer children and adolescents with SCD participated in daily vigorous or vigorous to moderate/moderate exercise for 60 minutes compared to healthy children using self-report questionnaires (Melo et al., 2018; Omwanghe et al., 2017) and an accelerometer (an objective monitor which was worn by participants for seven consecutive days) (Melo et al., 2018). However, Omwanghe et al., (2017) reported that the majority of participants with SCD (90%) attended physical education classes (although 42% reported sometimes being excused because of SCD) and participated in sports clubs (48%) or a sports team (61%) in the last year, although the authors did not specify the exact types of exercise. In contrast, Melo et al., (2018) reported that participants with SCA participated in significantly less physical education classes and sports at break/lunchtime, afterschool, in the evenings and at the weekends compared to healthy children, which may reflect the greater degree of disease severity in the sample. The NHS recommends that children with SCA should exercise regularly to maintain good health and for improved HRQL (NHS Choices, 2019). A recent study which evaluated the safety of a home aerobic exercise training program in adolescents and young adults with SCA found that participants did not suffer from any major adverse health effects related to exercise (Liem, Akinosun, Muntz, & Thompson, 2017). Therefore, it is important that SCD does not undermine their participation in exercise.

Nevertheless, children and adolescents with SCD should avoid overexerting themselves e.g. avoid prolonged or intense exercise, becoming out of breath or dehydrated and extreme or moderate temperature changes, exercising during illnesses and participating in contact sports (Connes et al., 2011; NHS Choices, 2019). This is because excessive

levels of exercise as well as strenuous and unsuitable exercise may negatively affect their condition. Vigorous exercise has been found to lead to dehydration in children and adolescents and trigger a vaso-occlusive crisis (Beyer et al., 1999) and also massive splenic infarction (Jama et al., 2002) and intravascular haemolysis (Platt, 1982) in adults with SCD. Furthermore, Platt (1982) found that the SCA group exhibited exercise-induced intravascular haemolysis (red cell damage) and had more dense cells, whereas adults in the HbSC group did not (Platt, 1982). This is expected as individuals with severe sickling phenotype generally have lower haemoglobin concentration. Therefore, individuals with milder sickling phenotype would be anticipated to have better exercise tolerance. Similarly, Machado et al., (2007) concluded that exercise led to higher blood pressure and decreases in oxygen saturation resulting in pulmonary hypertension. In adults with SCD, mild to moderate exercise was well tolerated and had no effect on haematocrit, blood viscosity and did not trigger a vaso-occlusive crisis or other clinical complications during or after exercise (Balayssac-Siransy et al., 2011; Faes et al., 2014). Dyson et al., (2010a) showed that unsuitable exercise at school triggered vaso-occlusive crises and Omwanghe et al., (2017) found that 41% of children reported sometimes experiencing sickle cell pain during or after exercising. However, studies in adults with SCA demonstrated that acute exercise (i.e. for a shorter amount of time) may be beneficial (Waltz et al., 2012) because acute exercise improves red blood cell disaggregation, thereby promoting microcirculatory blood flow (Waltz et al., 2012). However, it is unknown whether this will have similar effects in developing children and adolescents. Therefore, it is important to gauge the type, intensity and amount of exercise that children with SCD engage in.

Different methodologies are used to gauge the type, frequency and intensity of exercise and also where exercise takes place (Sallis & Saelens, 2000). Questionnaires have poor to good reliability and validity and are time efficient to administer, whereas

interviews, recall questionnaires and diaries/logs are typically completed over a few days to three months (Sallis & Saelens, 2000) and may be demanding for children. In children and adolescents with SCD there are limited validated self-report and parent proxy-report health behaviour questionnaires; two have been used to broadly examine health behaviours (Dyson et al., 2010a; Palermo, Lewandowski, Long, & Burant, 2008a) and different versions of the Physical Activity Questionnaire (PAQ) have been used to ascertain exercise levels (Melo et al., 2018; Omwanghe et al., 2017). These questionnaires do not gauge the types of exercises (e.g. rugby), intensity (e.g. strenuous), provide a definition of vigorous and moderate exercise and the average amount of weekly exercise. Also, Dyson et al.'s questionnaire (2010a; 2010b) focuses on health behaviours only in a school environment and the CALI-21 measure (Palermo et al's., 2008a) centres on general activity limitations during pain but not in everyday life. In contrast, the Godin-Leisure-Time Exercise Questionnaire (Godin, 2011; Godin & Shephard, 1985; 1997) assesses individuals' average weekly total, strenuous, moderate and mild exercise as well as how often, on average, they participate in regular leisure activities. It has been validated in children and adolescents, demonstrating good test-retest reliabilities ($\alpha = .81$) (Sallis, Buono, Roby, Micale, & Nelson, 1993) and with adult and paediatric populations with chronic conditions like cancer (Amireault, Godin, Lacombe, & Sabiston, 2015), although not in SCD. It may be the most appropriate measure for ascertaining the different types, intensity and frequency of exercise and will therefore also be used in the research in this thesis.

1.2.2.4 Prevalence of risky behaviours in SCD

Engaging in risky behaviours may aggravate symptoms of SCD. Such behaviours often develop during adolescence (Viner & Barker, 2005) and are prevalent in healthy adolescent populations and to the same extent, or to a greater extent, in chronically ill

populations (Sawyer, Drew, Yeo, & Britto, 2007; Surís, Michaud, Akre, & Sawyer, 2008). Surís et al., (2008) postulate that greater engagement in risky behaviours serves to convey that adolescents with chronic conditions are 'normal' regardless of their illness, that the higher prevalence of emotional distress is a risk factor for more engagement in these behaviours and that perhaps their potentially shortened lifespans mean that they are more impulsive and want to live life to the full. A recent review found that prevalent risky behaviours often investigated in adolescents with chronic conditions include; alcohol use, smoking or tobacco use and illegal drug or other substance use (Ssewanyana et al., 2017). Self-report measures of risky behaviours are frequently ascertained from adolescents, although these may not always be reliable and under-reporting occurs (Masson et al., 1992). However, reports from parents may not be useful as they are unlikely to be aware of their child's risky behaviours and objective measures are difficult to obtain. Therefore, the majority of research in this area generally obtains adolescent self-reports (Ssewanyana et al., 2017).

The prevalence of participating in risky behaviours in adolescents with SCD has rarely been determined (e.g. Asnani et al., 2014; Britto et al., 1998; Britto et al., 1999). Asnani et al., (2014) found that over three quarters (77.7%) of adolescents with SCD had consumed alcohol in the past which was significantly greater compared to their healthy peers (60.7%) (Asnani et al., 2014). Conversely, Britto et al., (1998) reported that adolescents with SCD were less likely to ever consume alcohol compared to their healthy peers and to adolescents with cystic fibrosis, although this was a younger sample. Nevertheless, this is concerning because alcohol use can lead to dehydration, increasing plasma viscosity (protein present in the plasma part of the blood) which restricts the movement of red blood cells, thereby allowing sickle shaped red blood cells to stick

together causing a vaso-occlusive crisis (Brown, 2012; as previously described in section 1.1.2).

The prevalence of other risky behaviours such as cigarette smoking is less common in adolescents with SCD compared to their healthy peers. Britto et al., (1998), for example, showed that adolescents with SCD smoked cigarettes less frequently compared to their healthy peers with 6.5% being regular smokers compared to 13.1% of their healthy peers. Also, fewer adolescents with SCD (30.0%) had ever smoked a cigarette compared to their healthy peers (42.9%). A more recent study demonstrated that there was no difference in adolescents with SCD and their healthy peers who had ever tried smoking with similar rates of 28.7% (Asnani et al., 2014). Not smoking is important as adolescents with SCD have lower oxygen saturations in comparison to their healthy peers (Young Jr, Rachal, Hackney Jr, Uy, & Scott, 1992). Carbon monoxide found in cigarette smoke reduces the oxygen-carrying capacity of red blood cells (hypoxia) (Aronow, 1978) and increases the risk of vaso-occlusive crises (Brown, 2012). Cigarette smoking has also been related to an increased rate of acute chest syndrome among adults with SCD (Cohen, DeBaun, Blinder, Strunk, & Field, 2010) and slower growth in adolescents who were healthy, had wheezing or asthma and smoked five or more cigarettes a day compared to those who had never smoked (Gold et al., 1996).

Finally, prevalent illegal drugs often investigated in adolescent research include; cannabis, marijuana, cocaine and intravenous drugs (Asnani et al., 2014; Britto et al., 1998; Suris et al., 2008). Asnani et al., (2014) found that some adolescents with SCD had smoked marijuana (17.4%), although this was no different from their healthy peers and Britto et al., (1998) concluded that adolescents (regardless of ill-health) rarely used cocaine or intravenous drugs. Adolescents with SCD are advised to avoid illegal drug use because it exacerbates their condition in similar ways to cigarette smoking. Also, illegal drug use

even in healthy adolescents causes chest pain due to myocardial ischemia (reduced blood flow to the heart) (Veeram, Reddy, Harinder, Singh, & Reddy, 2010).

In summary, this section has demonstrated that poor health behaviours like dehydration, poor diet and vigorous/strenuous or inappropriate exercise and risky behaviours can exacerbate SCD, potentially leading to a vaso-occlusive crisis. Since SCD may also impact participation in health behaviours, such as having to be careful about the amount and intensity of exercise undertaken, the following section addresses this further.

1.2.3 The influence of SCD on engaging in health behaviours

Children's ability to engage in health behaviours may be undermined by their condition. For example, research has suggested that children and adolescents with SCD consume less food during episodes of acute illness and hospitalisation (Ievers-Landis et al., 2001; Jacob et al., 2006; Malinauskas et al., 2000; Mitchell et al., 2004) and during painful episodes at home (Walco & Dampier, 1990), so may have difficulty taking in the nutrients they require. In addition, half of parents disclosed that their child did not want to hydrate to minimize sickle-related pain episodes (Ievers-Landis et al., 2001). A similar pattern emerged with diet, where infants with SCA consumed two-thirds less food than recommended during acute illness episodes such as pain, acute chest syndrome and infection (Malinauskas et al., 2000). Studies have also demonstrated that children and adolescents participate in fewer activities and exercise and spend less time outdoors during vaso-occlusive crises (Fuggle, Shand, Gill, & Davies, 1996; Jacob et al., 2006; Omwanghe et al., 2017; Shapiro, Dinges, Orne, Ohene-Frempong, & Orne, 1990; Walco & Dampier, 1990). It is not surprising that health behaviours alter when experiencing pain but daily health behaviours may help to control some symptoms of SCD. However, they have rarely been examined and never at both school and home (e.g. Dyson et al., 2010a; Karlson et al.,

2017; Omwanghe et al., 2017). Therefore, these behaviours will be examined in the research in this thesis. Furthermore, as children and adolescents can exert some control over these behaviours and they are amenable to modification, it is important to understand their determinants.

1.2.4 Determinants of health behaviours

1.2.4.1 Theoretical models of health behaviours

Health behaviours are also affected by aspects that are not related to the child's condition, for example, demographic, social, cultural, psychological and environmental factors. Cognitions (e.g. beliefs and attitudes) have been at the centre of much research on the determinants of health behaviours because they are thought to be key determinants and they are amenable to modification. There are different social cognition models that have been developed and applied to predict and explain health behaviours and these may ultimately be used as the basis of designing interventions to help change behaviours. Models include the Health Belief Model (Rosenstock, 1966; Becker & Maiman, 1975), Protection Motivation Theory (Rogers, 1975; Rogers, 1985), Social Cognitive Theory (Bandura, 1986; Bandura, 1977), the Health Action Process Approach (Schwarzer, 1992) and the Theory of Planned Behaviour (TPB)/Theory of Reasoned Action (TRA; Fishbein & Ajzen, 1975). The TPB is an extension of the TRA (Fishbein & Ajzen, 1975). The models assess an individual's intention to perform certain behaviours. This intention is the result of two determinants; their attitude towards the behaviour i.e. their overall evaluation of performing the behaviour and the perceived subjective norm (SN) i.e. belief about whether others think the individual should engage in the behaviour (Ajzen, 1988; Fishbein & Ajzen, 1975). An individual will have strong intentions to engage in, for example, regular water consumption if they evaluate it positively and believe that important

individuals in their lives believe they should engage in it (Ajzen, 1988; Fishbein & Ajzen, 1975). The TPB differs from the TRA because it also includes perceived behavioural control (PBC) (Ajzen, 1991). PBC refers to the perceived ease or difficulty of performing the behaviour. The three components of the TPB (attitude, SN and PBC) are determined by different types of beliefs. Firstly, attitudes are determined by an individual's salient behavioural beliefs concerning the outcome of the behaviour and their evaluation of that outcome. Secondly, SNs are determined by an individual's normative beliefs where important individuals or groups think they should, or should not, perform the behaviour and their motivation to comply with these individuals or groups. The TPB is the only social cognition model that includes a social component as a specific variable which makes it pertinent for use with children and adolescents whose behaviours may be particularly influenced by others. The last component, PBC, is determined by control beliefs about the presence of resources or barriers to performing the behaviour and the power of these to facilitate or inhibit performance of the behaviour. The TPB is one of the most popular and well supported theories which will be demonstrated in the following sections. In order to help understand health behaviours in children and adolescents with SCD, the present research will adopt the basic principles of the three 'belief' components of the TPB in determining behaviours, rather than applying the theory. In other words the research in this thesis will explore children's/adolescents' and parents' attitudes towards the child participating in behaviours, how social influences affect the child's engagement in behaviours (SN) and identifying what resources or barriers influence the child performing behaviours (PBC). Other research has also used the TPB in this way (e.g. Purewal & van den Akker, 2010).

There is a dearth of research investigating the determinants of health behaviours in children and adolescents with SCD and what little research there is, has not taken a

theoretical approach. However, much research has applied the TPB to examining health behaviours in children more generally. The next section will examine research on the determinants of health behaviours in children and adolescents with SCD, including more general research using the TPB, due to the absence of TPB studies in paediatric SCD. This section will also cover similarities and differences between child/adolescent and parental beliefs and the impact of parental beliefs on child/adolescent health behaviour.

1.2.4.2 Healthy eating and beverage consumption

Limited research in children and adolescents with SCD has identified some factors that may determine health behaviours. For example, inadequate dietary intake of proteins and micronutrients is more prevalent with increasing age and is especially poor in adolescents (Kawchak et al., 2007). Social factors such as the influence of family and friends on children's health behaviours have rarely been examined in children and adolescents with SCD. In SCD, the rate of food insecurity (i.e. reliable access to a sufficient quantity of food for an active, healthy life) was nearly twice as much as the national average in America (Gruntorad, Jones, Lapping-Carr, Peddinti, & Darlington, 2018). This is not surprising given the prevalence of lower SES in SCD (Ilesanmi, 2013), and therefore food insecurity may contribute to a poor diet or lack of nutrients in SCD. However, in healthy children, determinants of eating behaviours include SES, environment, parental influence (which can be counterproductive because they pressurise children to eat and offer them rewards) and dissatisfaction regarding body image (Scaglioni, Arrizza, Vecchi, & Tedeschi, 2011). A review conducted by Darmon and Drewnowski (2008) found that children of higher SES had better quality diets including more lean meats, fish, fruits and vegetables and micronutrients whereas the diets of children of lower SES included more fatty meats, refined grains and added fats. The

authors suggested that this could be related to lack of nutrition knowledge, lack of motivation and a general disinterest in cooking (Darmon & Drewnowski, 2008). A review of determinants of healthy eating among children and adolescents found that the types of foods available in different environments (i.e. home, school and fast-food establishments) and, to a lesser extent the media, particularly television, were the predominant influences on children's eating habits (Taylor, Evers, & McKenna, 2005). Taylor and colleagues (2005) also identified that individual factors influence healthy eating with food preferences being the strongest predictor and, to a lesser extent knowledge and attitude.

A systematic review and meta-analysis found that the TPB is the most prevalent model used in children and adolescents to predict their healthy eating, sugary snack and beverage consumption (Riebl et al., 2015) but reviews have identified different predictors of different health behaviours. Riebl et al., (2015) established that attitude was the strongest and most frequent predictor of behavioural intention to eat healthily (i.e. fruit, vegetable and water consumption) and that all constructs of the TPB were greater in female participants. This was supported by McDermott et al. (2015). However, when predicting adolescents' intention to limit their sugar-sweetened beverage consumption, the strongest predictor of intention for adolescents was SN and for parents was PBC (Riebl et al., 2016). Contrary to this, some studies have proposed that the intention of healthy children and adolescents to eat healthily is influenced more by other external factors and is therefore constantly changing (Fila & Smith, 2006).

The TPB has also been applied to understand parental feeding practices in healthy children and preadolescents. Hewitt and Stephens (2007) found that children's attitude, SNs and PBC from the TPB explained 51% of the variance in children's healthy eating intentions. However, the authors also showed that parental feeding practices, for example, restriction or responsibility was not useful in predicting any further variance in children's

intention to eat healthy foods. Using the Attitude-Social Influence-Self-Efficacy model (which is similar to the TPB), Melbye, Øverby and Øgaard (2012) reported that only one of twelve parental feeding practices (i.e. environment) predicted children's intention to eat vegetables but not fruit, whereas the model was able to explain a large portion of variance in children's intentions and consumption of fruit and vegetables. Family beliefs have also been explored in adults. A study reported some positive relationships between adult women, and their mothers' and grandmothers' beliefs, attitudes and intentions towards the consumption of twenty foods, although the study did not include all of the components of the TPB (Stafleu, Van Staveren, De Graaf, Burema, & Hautvast, 1995). These studies did not examine the relationships between child/adolescent and parental beliefs and how parental beliefs may predict children's dietary behaviours (Hewitt & Stephens, 2007; Melbye et al., 2012; Stafleu et al., 1995). However, Sumodhee and Payne (2016) found that mothers' and their adult child's TPB beliefs were related, and that children's intention to eat healthily were predicted by their mothers' intentions and PBC.

1.2.4.3 Exercise

There is a dearth of research investigating the determinants of exercise in paediatric SCD. However, Omwanghe et al. (2017) examined children's vigorous and vigorous to moderate exercise levels (the authors did not provide a definition of this) and found that factors associated with less participation in exercise included older age, perceptions that SCD negatively impacts their enjoyment of exercise and finally perceptions of poor physical functioning. Dyson et al., (2010a) did not ascertain the types of exercise participants did at home and how their peers may influence them but the authors reported that at school, 36.3% of children were made to do unsuitable exercise and also 43.2% of children had a vaso-occlusive crisis that was triggered by unsuitable exercise. In healthy

children, Sallis, Prochaska and Taylor's (2000) review found they exercised more if they enjoyed it, had a healthy diet, access to facilities and spent more time outdoors, whereas in adolescents, those who exercised more were of white ethnicity, not depressed, had greater perceived activity competence and had family members who were active or supported them. Furthermore, male gender and intention to be active were contributing factors in both groups (Sallis et al., 2000).

A systematic review and meta-analysis of TPB studies reported that attitude was the strongest construct for explaining exercise intention in healthy adolescents (Plotnikoff, Costigan, Karunamuni, & Lubans, 2013). In healthy children, SN and PBC predicted intention to participate in leisure-time exercise (Rhodes, Macdonald, & McKay, 2006). In contrast, Wang and Zhang (2016) applied an extended TPB (which included self-efficacy and past behaviour) to predict adolescents' intention to participate in moderate to strenuous exercise and found that the strongest predictor was self-efficacy followed by PBC. Therefore, different components of the TPB predict participation in exercise in younger children compared to adolescents. In children of African ethnicity, attitude and SN predicted moderate to vigorous exercise (Martin et al., 2005b) whereas in adolescents of low SES, attitude and PBC predicted behaviour towards exercise (Duncan, Rivas, & Jordan, 2012). The TPB has also been applied in adolescent chronically ill populations, where attitude was the strongest predictor of intention to exercise in cancer survivors (Keats, Culos-Reed, Courneya, & McBride, 2007).

The relationship between parental TPB beliefs and child/adolescent TPB beliefs towards exercise has not been investigated in the literature. However, parenting practices, behaviours and beliefs are important and can influence children's/adolescents' participation in exercise (Trost & Loprinzi, 2011). A longitudinal study that examined exercise behaviours of parents and their children found that physically active fathers

positively influenced their sons' participation in sports and that inactive mothers negatively influenced their daughters' engagement in sports (Martin, Dollman, Norton, & Robertson, 2005a). In addition, a review by Gustafson and Rhodes (2006) found that parental support (primarily encouragement, involvement and facilitation i.e. part of PBC, a component of the TPB) can predict exercise in children and adolescents and that this is often more pronounced in younger children. The authors established that greater parental support predicted increased exercise (Gustafson & Rhodes, 2006).

1.2.4.4 Risky behaviours

A limited number of studies have examined the determinants of risky behaviours in adolescents with SCD. They are more likely to begin smoking cigarettes or marijuana at an older age compared to healthy adolescents (13 to 14 years vs. 11 to 12 years) and also reported having their first alcoholic drink at an older age (15 years old or above) (Britto et al., 1998). Asnani et al., (2014) found that being male was the only predictor of ever having drunk alcohol in adolescents with SCD (age, currently in school, living with a parent and 'durable goods', which is an alternative measure to income, were not significant predictors). Adolescents also described smoking marijuana because they wanted to try it out and also because their friends were smoking, although the authors did not report whether this was because of peer pressure or because, being adolescents, they were seeking peer acceptance (Asnani et al., 2014). Moreover, Asnani and colleagues (2014) found that engaging in one type of risky behaviour increased the chances of engaging in another, for example, older adolescents who had consumed alcohol were more likely to have smoked cigarettes.

In healthy adolescents, demographic factors associated with regular drinking included being from a single-parent family, being in a higher year group, higher rates of

psychological morbidity and greater depressive symptoms (Viner et al., 2006). There were also some consistent factors that were related to less engagement in risky behaviours such as being of Black African ethnicity, in a higher Year group, being religious and greater family social support (Viner et al., 2006).

TPB research, including a systematic review and meta-analysis, found that attitude is consistently the strongest predictor of intention to consume alcohol in healthy adolescents and adults (Cooke, Dahdah, Norman, & French, 2016; López-Cisneros et al., 2013), despite the fact that adolescents are aware that it is unhealthy, not safe and may be detrimental to their health (López-Cisneros et al., 2013). Therefore, adolescents' intention to drink alcohol may be based more on their affective attitude (emotional reaction toward drinking i.e. enjoyment) rather than their cognitive attitude (knowledge regarding alcohol use). Cooke et al., (2016) noted that there were stronger TPB relationships reported in studies which specified units of alcohol consumption rather than vague definitions, for example, "*getting drunk*". In contrast, Marcoux and Shope (1997) found that external factors such as friends' experience with alcohol, peer pressure, normative beliefs of parents and availability of alcohol were more important in predicting intention to use alcohol compared to internal factors such as attitude in healthy children and adolescents.

In summary, health behaviours are determined by different factors in children and adolescents, which is not surprising given their differing developmental stages in life and levels of autonomy, but such behaviours have not been fully investigated in SCD. Similarly, predictors of adolescents' risky behaviours have not been examined in SCD. These issues will be examined in the research in this thesis, as well as similarities and differences between parental and child/adolescent beliefs. Whether health and risky behaviours are linked to HRQL will also be examined and research on this will be covered in the following section.

1.2.4.5 HRQL and health behaviours

Relatively few studies have examined health behaviours and HRQL in children and adolescents with SCD. Individuals experiencing greater pain were found to have increased activity limitations and also a lower HRQL (Zempsky et al., 2013). Wrotniak, Schall, Brault, Balmer and Stallings (2014) concluded that parent proxy-reported HRQL was lower in children with SCA at baseline in comparison to healthy children, and after a nutritional supplementation intervention the physical health summary score and emotional impact were comparable to the healthy population. Therefore, adherence to diet regimens improved parent proxy-reported HRQL, although child self-reports and the views of older adolescents were not ascertained (Wrotniak et al., 2014). This is in contrast to Barakat, Lutz, Smith-Whitley and Ohene-Frempong (2005), who reported that better medication adherence and family engagement in SCD-related care activities was associated with lower overall QoL, as opposed to higher QoL, as one might postulate. They explained this inverse relationship by suggesting that standard treatment recommendations in SCD reduce activity engagement meaning that adhering to treatment interferes with some aspects of QoL (Barakat et al., 2005). The same may apply to other aspects of health behaviours. In other words, increasing health behaviours, such as ensuring children stay hydrated, and reducing risky behaviours, such as limiting alcohol in adolescents, may, contrary to expectation, have an inverse relationship with HRQL. Furthermore, one might expect that limiting exercise would be related to increased HRQL, if it helps to avoid a vaso-occlusive crisis, but such limitations on, for example, sports may actually be linked to reduced HRQL. Indeed, in adults with SCD, regular exercise tended to improve some aspects of QoL including general health, bodily pain, social functioning and vitality (Ahmed et al., 2015).

In summary, there is a plethora of paediatric SCD research on HRQL but not on health and risky behaviours, despite the fact that these everyday behaviours could be modified at home and at school to help better control some symptoms of SCD. In addition, health and risky behaviours may influence HRQL. Since there is limited research on such behaviours and also including healthy siblings (as previously highlighted in section 1.1.5), an initial qualitative approach will help to understand the experiences of children and adolescents with SCD and their healthy siblings and identify areas that will then be studied further using quantitative methodology. Thus the research in this thesis will use a mixed methods approach, which is discussed below in the final section of this chapter.

1.3 Methodologies used in research on paediatric populations with chronic conditions

1.3.1 Introduction

The two previous sections have covered HRQL, and health and risky behaviours in children and adolescents with SCD. Since the present research adopts a mixed methods approach to examine these factors, this section briefly outlines the necessity for this approach. This is followed by a discussion of the types of qualitative methodology used when exploring issues in different age groups (children and adolescents) with and without chronic conditions. This includes describing the use of semi-structured interviews and drawings to engage children in discussions. Drawings are often used as a projective technique or as a communication tool, and the latter will be described to be the most appropriate method. The use of focus groups for exploring these issues in adolescents will also be discussed. These methodologies have not been adopted in SCD populations to explore HRQL, health behaviours or risky behaviours.

1.3.2 Mixed-methods

There are different designs in the mixed methods field, although Creswell (2014) identifies four primary models; convergent parallel, transformative, explanatory sequential and exploratory sequential mixed methods. Firstly, convergent parallel mixed methods are where the researcher converges or combines quantitative and qualitative data (usually collected simultaneously) in order to provide a comprehensive analysis of the research problem (Creswell, 2014). Secondly, transformative mixed methods uses a theoretical lens drawn from social justice or power as an overarching perspective within a design that contains both quantitative and qualitative data (Creswell, 2014). Thirdly, explanatory sequential mixed methods is where the researcher initially collects quantitative data, analyses the results, and then uses these results to plan the qualitative research which follows (Creswell, 2014). The present research will use exploratory sequential mixed methods, meaning that qualitative methods will be used to explore the views among children and adolescents with SCD and healthy siblings. The qualitative data will then be analysed and inform the development of two quantitative studies with a larger sample so that the results may be more generalisable (Creswell, 2014).

Quantitative methodology (usually using questionnaires such as the PedsQL™) has been used to investigate HRQL in SCD in past research; but the use of qualitative methodology is rare and also this method has not been based on a theoretical approach to HRQL (as previously shown in section 1.1.4). Moreover, research has rarely examined health behaviours of children and adolescents with SCD, especially from a theoretical perspective such as the TPB (which was highlighted in section 1.2.4). Therefore, qualitative methodology is considered useful in initially exploring the different areas of HRQL, health behaviours and risky behaviours in children and adolescents with SCD. The following sections will focus on the qualitative design of the first part of the research in

this thesis and describe qualitative methods used in healthy and chronically ill children and adolescents.

1.3.3 Drawings and interviews

Art-based techniques have been used to engage children in research and consultations (Coad, 2007). Coad (2007) discusses three types of art-based techniques; photography, drawings/posters or collages and mapping, although, many more exist which have been used to facilitate research conversation or informal interviews. A systematic survey of arts-based techniques used in health-related research found that drawing was the most prevalent technique (Driessnack & Furukawa, 2012). Drawing can be used as a projective technique or as a communication tool with children (Bekhit, Thomas, & Jolley, 2005; Driessnack & Furukawa, 2012). Projective techniques use verbal or visual stimuli where an individual 'projects' or reveals their unconscious feelings, attitudes, experiences and other characteristics (Will, Eadie, & MacAskill, 1996). They involve the researcher attributing behaviour to the participant rather than asking an individual directly about their own feelings and beliefs (Will et al., 1996). As a projective technique, drawing has been found to inform researchers and allied health professionals about a child's intellect (Goodenough, 1926; Harris, 1963; Naglieri, 1988), personality (Machover, 1949), current emotional state (Koppitz, 1966; Koppitz, 1968; Koppitz, 1984) and emotional attitudes towards topics drawn (Burkitt, Barrett, & Davis, 2003a; Burkitt, Barrett, & Davis, 2003b; Thomas & Jolley, 1998). There are a variety of projective techniques including Draw-a-Man (DAM) (Goodenough, 1926), Goodenough-Harris Draw-a-Man Test (GHDT; Harris, 1963), Draw-a-Person test (DAP; Machover, 1949), Emotional Indicators (EIs) (Koppitz, 1966) and Kinetic Family Drawing (KFD) (Burns & Kaufman, 1970). Most projective techniques involve a child drawing one or two pictures of a human figure (Goodenough, 1926; Harris, 1963; Koppitz, 1966; Machover, 1949). The KFD technique also requires a

child to 'draw your family doing something' emphasising that the child should draw 'some kind of action' (Burns & Kaufman, 1970) rather than just human figure drawings. The pictures are analysed using different objective scoring systems, for example, in the DAM technique, the presence of 51 human figure details such as the trunk are scored (Goodenough, 1926) and scoring is also done through subjective interpretations such as which gender the individual decides to draw first, particular body parts included, their size and shape and differences between the two drawings (Machover, 1949). Projective techniques have been applied to different populations including hospitalised children (Nyman, Baluch, & Duffy, 2011; Stefanatou, 2008), children with learning disabilities (Perets-Dubrovsky, Kaveh, Deutsh-Castel, Cohen, & Tirosh, 2010), children with behavioural difficulties (Catte & Cox, 1999) and healthy children (Burkitt et al., 2003b; Catte, & Cox, 1999). There have been inconsistencies using this methodology. Nyman et al., (2011) established that EIs (Koppitz, 1968) in self-drawings could not be used to detect emotional disturbance experienced by hospital school children. In contrast, Perets-Dubrovsky et al., (2010) found that analysing human figure drawings using Koppitz' cognitive development and EIs (Koppitz, 1968) was reliable and moderately valid when assessing children with Attention-Deficit Hyperactivity Disorder (ADHD) and learning disabilities. Projective techniques have also been applied to children with SCA, where the DAP test (Harris, 1963) and Koppitz's (1968) EIs were used to analyse their perceptions of pain by asking them to draw two pictures; one of themselves and one of themselves in pain and then the researcher discussed each picture with them (Stefanatou & Bowler, 1997). In the first picture no child drew themselves in pain which according to Stefanatou and Bowler (1997) indicated that pain was not an integral part of their self-concept. In the second picture children drew the site of pain, their mood state (e.g. sad) and them doing something about the pain (e.g. lying down or having a glass of water). Children's drawings

were viewed as indicative of their feelings, for example, children who discussed having pain in their legs also omitted their legs from their pain drawing (Stefanatou & Bowler, 1997). Although they used drawings as a projective technique, the methodology adopted by Stefanatou and Bowler (1997) involved drawings and discussion, which was a useful approach in engaging with children with SCA, who were as young as five years old, about their condition and how it affects their lives.

Projective measures have been criticised because of the lack of valid and reliable scoring systems (Klopfer & Taulbee, 1976; Suinn & Oskamp, 1969; Swenson, 1957); they do not describe personality, emotion and behaviour (Motta, Little, & Tobin, 1993; Smith & Dumont, 1995); some scoring systems were derived from American populations and have not been validated in other populations (La Voy et al., 2001; Skybo, Ryan-Wenger, & Su, 2007); interpretations of drawings are subjective, for example, a particular feature of a drawing could invoke several interpretations (Thomas & Jolley, 1998); and they cannot distinguish between clinical and non-clinical groups (Briccetti, 1994). Furthermore, drawings may not be useful in clinical settings because psychologists may 'see' things in human figure drawings that they may already be aware of in a patient's case history (Smith & Dumont, 1995). Thomas and Jolley (1998) have suggested that collating drawings from matched control children may help to interpret drawings from children in a clinical setting.

Drawing has been effectively applied as a communication tool to facilitate conversation during semi-structured interviews with children (Driessnack & Furukawa, 2012; Driessnack, 2005; Gross & Hayne, 1998; Macleod, Gross, & Hayne, 2013; Patterson & Hayne, 2011; Woolford, Patterson, Macleod, Hobbs, & Hayne, 2013), including children with SCD (Cotton, Grossoehme, & McGrady, 2012), hospitalised children (Boyd & Hunsberger, 1998) and children with mental health problems (Woolford et al., 2013). It has also been applied to healthy children to ascertain their views about issues that are

important to them surrounding their HRQL (White, Bushin, Carpena-Méndez, & Laoire, 2010) and starting school (Einarsdottir, Dockett, & Perry, 2009). In paediatric research, drawings are used in different ways, for example, as an ‘icebreaker’ during semi-structured interviews to reduce chronically ill children’s anxiety and build a rapport with the researcher (Boyd & Hunsberger, 1998), as a catalyst to focus and guide interviews (Driessnack & Furukawa, 2012) and also in conjunction with semi-structured interviews to facilitate conversation (Cotton et al., 2012). Drawing as a communication tool has proved effective in exploring sensitive topics in children with SCD who were asked to draw things that help them cope with being sick (Cotton et al., 2012). Moreover, evidence has found that children across age-groups have reported that drawing helps them to speak in clinical settings such as hospitals (Davies & Wright, 2008; Woolford et al., 2013) and they report enjoying drawing, although a minority of preadolescent children found it embarrassing (Patterson & Hayne, 2011).

Drawings may be effective alongside interviews because they may elicit more information from children. Children with mental health problems discussed their presenting problems through ‘drawing and telling’ or ‘telling’ alone with twice as much clinically relevant information obtained when they drew and told as opposed to told alone (Woolford et al., 2013). Drawings have been used to gauge young children’s past and current views about starting school (Einarsdottir et al., 2009) which is similar to a Gap Theory approach in HRQL where a child might be asked to draw something current vs. something ideal.

There are some disadvantages to drawing. For example, healthy children look at, and copy each other’s drawings and they produce peer-acceptable drawings such as including a PlayStation 2 (White et al., 2010). In addition, while 95% of healthy children liked drawing, 24% of healthy children felt it was embarrassing, most of whom were aged

11 to 12 year olds (Patterson & Hayne, 2011). It is therefore important, that children should not draw pictures surrounded by other children (White et al., 2010).

1.3.4 Focus groups

Studies have adopted different types of qualitative methodologies when exploring issues surrounding HRQL in adolescents with chronic conditions such as SCD and healthy adolescents. Prevalent qualitative methodologies used include semi-structured interviews (Atoui et al., 2015; Cotton et al., 2009; Forrester et al., 2015), in-depth and cognitive interviews (Panepinto et al., 2012) and focus or discussions groups (Jones & Broome, 2001; Porter, Graff, Lopez, & Hankins, 2014; Valenzuela et al., 2013). Panepinto et al., (2012) initially adopted focus group methodology which was unsuccessful as participants (parents and children and adolescents with SCD) failed to attend sessions and therefore they chose to conduct individual in-depth interviews. In a pilot study, Valenzuela et al., (2013) sought to understand the lives of children and adolescents with SCD through three photo group discussions using photographs that participants had taken. The authors found that these groups gave children and adolescents an opportunity to socialise and engage with their peers who also have SCD, as well as learning more about themselves and sharing this with others (Valenzuela et al., 2013). In health research, focus groups are often used to ascertain the views, perspectives and experiences of adolescents (Rabiee, 2004; Wong, 2008). Porter et al., (2014) conducted focus groups with families including adolescents with SCD, their siblings and parents to explore the challenges of transitioning from paediatric to adult care. Focus groups have also been used successfully to explore the views of healthy adolescents with regards to healthy eating (Stevenson, Doherty, Barnett, Muldoon, & Trew, 2007) and exercise (Whitehead & Biddle, 2008) and also their risky behaviours including smoking, alcohol use and drug use (Lavery, Robinson, & Holdsworth, 2015).

Focus groups are a useful method because they allow for the interaction and stimulation of thought during the group process (Peterson-Sweeney, 2005). Such interactions develop participants' attitudes and perceptions (Wong, 2008). Thus focus groups are advantageous in exploratory research, such as exploring adolescents HRQL and health and risky behaviours (Heary & Hennessy, 2002). Also, group discussions are relaxed and conversational because group members share a common experience. This allows the researcher to obtain rich data that may not be ascertained from one-to-one interviews and adolescents may not feel comfortable discussing their true feelings with an adult researcher or may be nervous to talk within such an interview (Peterson-Sweeney, 2005). Heary and Hennessy (2002) also suggest that focus groups place less emphasis on the adult-child relationship. Therefore, focus groups present a more natural environment because group members influence each other as would occur in normal conversations (Peterson-Sweeney, 2005). In addition, group discussions allow for different forms of communication that occur in day-to-day life such as joking, anecdotes, teasing and arguments producing richer data (Wong, 2008).

There has been a lot of debate about how to construct focus groups in adolescent populations. Peterson-Sweeney (2005) proposed that to promote group discussion and interaction participants should have similar backgrounds and experiences related to the topic. For example, as children with SCD and healthy siblings have different experiences from each other it may benefit them to be in separate groups. Heary and Hennessy (2002) suggest that single-sex groups of males or females facilitate more open discussions, especially when topics are sensitive as is the case with HRQL, health behaviours and risky behaviours. Moreover, Davis and Jones (1996) found that mixed gender groups were counterproductive because 13 to 14 year old males and females had conflicting agendas. It is not surprising that adolescent females and males are affected by different issues, for

example, Davis and Jones (1996) found that girls had more restrictions about how late they stayed out than boys. Moreover they found that eight single-sex focus groups produced rich data (Davis & Jones, 1996). The single-sex groups therefore appear to be most appropriate in adolescent populations. The size of focus groups is also important with between four to six participants being optimum (which is less than recommended for adult focus groups) because larger focus groups produced more ideas in adults (Heary & Hennessy, 2002), whereas in adolescent focus groups, four to six participants ensures that there are at least three ‘talkers’ (Heary & Hennessy, 2002). Furthermore, having larger adolescent focus groups may mean that they talk simultaneously, interrupt each other, and can be more intimidating, which therefore inhibits the nature of organic group discussions (Heary & Hennessy, 2002).

As was shown above, qualitative methodologies are not often used in paediatric SCD (or with healthy siblings; see sections 1.1.4.3 and 1.1.5.2) but have proved useful in health-related research and when exploring health and risky behaviours. In the qualitative research in this thesis, drawings will be used alongside a semi-structured interview to facilitate conversation in children, whereas in adolescents, focus groups will be implemented, as they have been demonstrated in this section to be more age-appropriate.

1.4 The present programme of research

The present programme of research aimed to integrate HRQL, health behaviour and risky behaviour research in paediatric SCD. This was achieved by adopting two theoretical approaches: GT to examine the discrepancy between current and ideal HRQL of children and adolescents (Studies 1 and 2) and some principles of the TPB to examine the factors that influence children’s and adolescents’ health behaviours (diet, water consumption and exercise) and also adolescents’ risky behaviours (alcohol use, cigarette smoking and drug

use). These theoretical approaches, with one exception (Constantinou et al., 2015), have not previously been applied in children and adolescents with SCD. The present programme of mixed methods research seeks to explore the HRQL, health behaviours and risky behaviours of children and adolescents with SCD. In the qualitative part of the research a representative sample of healthy siblings was also used to see if the issues identified are unique to SCD and also to explore how having a sibling with SCD is experienced by healthy siblings. Children are the focus of Study 1 and adolescents are the focus of Study 2. In the quantitative part of the research, parents' reports of their child's or adolescent's HRQL (Study 3) and health behaviours (Study 4) were also explored along with the self-reports of children/adolescents with SCD.

Study 1 explored the discrepancy between current and ideal HRQL and health behaviours (diet, water consumption and exercise) of children with SCD and healthy siblings aged 5 to 12 years old. The study used a drawing task and a semi-structured interview lasting between 21 and 69 minutes with 18 children with SCD and 14 healthy siblings. Thematic analysis was used to analyse the data. Therefore, the aim of Study 1 was to explore the HRQL and health behaviours (diet, water consumption and exercise levels) of children with SCD, including any discrepancies between their current and ideal self, compared to that of their healthy siblings.

Study 2 explored the same factors (HRQL and health behaviours) as well as risky behaviours (alcohol use, cigarette smoking and drug use) in adolescents with SCD and healthy siblings aged 13 to 17 years old. The study used eight focus groups involving 23 adolescents with SCD and 21 healthy siblings. Each single-sex group of males or females contained 5 to 6 participants and lasted for approximately one hour to one and a half hours. The data were analysed using thematic analysis. Thus, the aim of Study 2 was to explore the HRQL, health behaviours (diet, water consumption and exercise levels) and risky

behaviours (alcohol use, tobacco cigarette smoking and illegal drug use) of adolescents with SCD, including any differences between their current and ideal self, compared to that of their healthy siblings.

Study 3 examined predictors of children's and adolescents' self-reported and parent proxy reported current perceived HRQL. There were four groups of participants: 106 children with SCD aged 5 to 12 years old and 106 of their parents (this term will be used throughout this thesis to indicate either a parent or carer) and also 96 adolescents with SCD aged 13 to 18 years old and 96 of their parents. All four groups of participants completed the PedsQLTM and a health behaviour questionnaire. Children and adolescents with SCD also completed The Kids Coping Scale (KCS) and finally, adolescents with SCD also completed a risky behaviour questionnaire. Study 3 had two main aims. First, to explore if there was a relationship between child self-reports, adolescent self-reports and parent proxy-reports of HRQL. Second, to determine the predictors (e.g. demographic indicators, coping, disease severity measures, health/risky behaviours) of child/adolescent self-reports and parent proxy reports of HRQL.

Study 4 investigated the predictors of children's and adolescents' health behaviours (water consumption and exercise levels) and also an aspect of adolescents' risky behaviours (alcohol use). The same four groups of participants and measures were used as for study 3 but beliefs about the health and risky behaviours were also assessed measured. These belief measures were developed from the findings of the qualitative studies and also guided by the TPB. Study 4 had two main aims. The first aim was to examine the relationships between children's and adolescents' self-reported health beliefs and health behaviours and parental reports of children's and adolescents' health beliefs and health behaviours. The second aim consisted of two parts; the first was to determine the predictors (e.g. demographic indicators, disease severity measures and child and parent health beliefs)

of self-reported health behaviours in children and adolescents with SCD. The second part was to determine the predictors (e.g. demographic indicators, disease severity measures and health beliefs) of self-reported risky behaviours in adolescents with SCD.

Chapter 2. Exploring HRQL and Health Behaviours in Children with SCD and Healthy Siblings (Study 1)

2.1 Introduction

As discussed in Chapter 1, questionnaires are the most prevalent method of measuring health-related quality of life (HRQL) in paediatric sickle cell disease (SCD) (Panepinto & Bonner, 2012), where child and parent-proxy reports of perceived current HRQL are generally found to be lower than in healthy children (Bonner et al., 2010; Panepinto et al., 2005). In contrast, using measures based on a Gap Theory (GT; Calman, 1984) approach, where HRQL is the gap between children's current and ideal selves, children generally do not seem to have a lower quality of life (QoL) compared to healthy children, including in a study of SCA (Constantinou et al., 2015). There are some reservations with using current questionnaires because those based on a GT approach do not measure the multidimensional concept of HRQL and they are not available in both child self-report and parent proxy measures (Collier et al., 2000; Eiser et al., 1999; Eiser et al., 2000). Similarly, measures of perceived current HRQL such as the Pediatric Quality of Life Inventory (PedsQL™) are not based on a theoretical approach.

Adopting a qualitative approach in exploring HRQL in children with SCD may address some of the shortcomings of quantitative research, especially since the present study will be adopting a GT approach and thus examining children's perceived current and ideal selves. HRQL has rarely been explored using qualitative methods in children with SCD (e.g. Panepinto et al., 2012; Stegenga et al., 2004), and has only been explored once using a Gap approach, but this was in a quantitative study (Constantinou et al., 2015). Past evidence supports the use of drawings as a communication tool alongside semi-structured interviews (Driessnack & Furukawa, 2012), which has been used with chronically ill

children including children with SCD (Cotton et al., 2012) and healthy children. This method facilitates conversation in young children (as discussed in Chapter 1 section 1.3.3). In the present study general themes from drawings will be identified such as what the child has chosen to draw, and drawings will also be used to illustrate themes identified from the interview data and especially to help explore children's current and ideal selves.

As discussed in Chapter 1 (section 1.1.5), having a representative comparison group (in terms of age, gender, ethnicity and SES) such as healthy siblings who themselves may experience some psychosocial problems (Hijmans et al., 2009; Juanita Lee et al., 1997) could help identify what issues are unique to children with SCD and also how the healthy siblings' HRQL is affected by having a chronically ill sibling. This has never been explored using qualitative methods and has only been examined in a single quantitative study (Hijmans et al., 2010). It has also not been explored by taking a theoretical approach such as GT, so healthy siblings will also be included in the present study.

SCD is often managed at home and some factors such as children's health behaviours have been found to help manage symptoms associated with their condition. The behaviours that have been found to be more important for children with SCD are participating in regular and suitable exercise (NHS Choices, 2019), drinking water (Brown, 2012) and a healthy diet (Hyacinth et al., 2010). However, children's daily engagement in these behaviours has rarely been investigated quantitatively or qualitatively and when they have, different environments like their home and school have not been considered (Dyson et al., 2010a; Karlson et al., 2017) and the types of exercise were not reported (Dyson et al., 2010a; Karlson et al., 2017; Melo et al., 2018; Omwanghe et al., 2017). Thus the present study will explore these health behaviours. Determinants of such behaviours may be better understood by adopting the basic principles of the three 'belief' components of the Theory of Planned Behaviour (TPB); attitude, subjective norm (SN) and perceived

behavioural control (PBC) (Ajzen, 1988; Fishbein & Ajzen, 1975). The TPB has proved useful in gauging the determinants of health behaviours in healthy (Martin et al., 2005b; Riebl et al., 2015; Rhodes et al., 2006) and chronically ill (Keats et al., 2007) children. In addition, having SCD may also influence their health behaviours but this has rarely been studied in this population and will therefore be explored in the current study. Management of health behaviours may be linked to improved HRQL if children are able to have better control over their symptoms, for example, by keeping hydrated and avoiding excessive exercise and therefore not be hospitalised, miss school or suffer from psychosocial problems as described in Chapter 1 (section 1.1.3).

The aim of the current study was to explore the HRQL and health behaviours (diet, water consumption and exercise levels) of children with SCD, including any discrepancies between their current and ideal self, compared to that of their healthy siblings.

2.2 Method

2.2.1 Design

As discussed in Chapter 1 (section 1.3.3), the current study adopted qualitative methodology using drawing tasks with semi-structured interviews. Children with SCD and healthy siblings were compared.

2.2.2 Participants

Children with SCD were not invited to participate if they had any co-morbidities that were not related to their condition as this may have affected their HRQL. Healthy siblings were invited to participate if they had a sibling who was diagnosed with SCD and did not have any chronic, acute, physical, neurological, psychiatric or psychological health problems that may have affected their HRQL.

A convenience sample of 18 children with SCD and 14 healthy siblings who met the inclusion criteria were recruited to the study from a London hospital. No participants withdrew from the study. Participant recruitment ceased when data saturation had been reached and no new issues were raised by children. Genders were equally distributed, and their ages ranged from 5 to 12 years old, with children with SCD ($M = 8.56$, $SD = 2.18$) not significantly older than healthy siblings ($M = 8.36$, $SD = 2.44$; $t(30) = 0.24$, $p = .810$). The majority of children with SCD had the most prevalent and severe phenotype; 15 (83%) had HbSS and 3 (17%) had HbSC. Sixteen (89%) children with SCD and 12 (86%) healthy siblings were of Black African ethnic origin, with the remaining 2 (11%) children with SCD and 2 (14%) healthy siblings of Black Caribbean ethnic origin. Table 2.1 provides a comprehensive description of sample characteristics.

Table 2.1

Descriptive Statistics for Socio-Demographic Characteristics of Children with SCD and Healthy Siblings

<u>Participants</u>	Children with SCD M(SD) or % n	Healthy siblings M(SD) or % n
<u>Child variable</u>		
Age	8.56(2.18)	8.36(2.44)
Gender	50% Male	50% Male
Ethnicity	89% Black African, 11% Black Caribbean	86% Black African, 14% Black Caribbean
SCD phenotype	78% HbSS, 22% HbSC	100% HbAA
<u>Parent variable</u>		
Age	41.56(6.52)	39.79(4.58)
Gender	11% Male	7% Male
Marital status	56% Married/cohabiting, 44% Single/separated/ divorced	57% Married/cohabiting, 43% Single/separated /divorced
Employment status	67% Employed, 33% Unemployed/ /unable to work/ retired	71% Employed, 29% Unemployed/unable to work
Highest educational attainment	44% Degree, 22% Foundation degree/diploma, 33% A Levels or below	43% Degree, 29% Foundation degree/diploma, 29% A Levels or below

2.2.3 Materials

A semi-structured interview schedule (Appendix A) was used by the researcher as a guide to what issues were to be explored. Each participating child with SCD, healthy sibling and their parent received an information sheet, consent/assent form and a debriefing form (Appendix B). In addition, the parent also received a demographic information sheet (Appendix B).

2.2.3.1 Interview schedule

The interview schedule provides an idea of what was explored but not necessarily the exact manner or sequence in which the questions were asked; this was dependent on

what was raised by the children and also what was important to them. The interview schedule was developed and piloted with 5 children with SCD ($M = 8.20$, $SD = 2.86$) and 5 healthy children ($M = 8.60$, $SD = 2.70$) aged 5 to 12 years to ensure it was not too demanding for them, and that they understood what they were being asked.

The interview schedule consisted of six stages; stage 1 – ‘gaining informed consent’, stage 2 – ‘establishing a rapport’, stage 3 – ‘introducing the research’, stage 4 – ‘beginning the interview’, stage 5 – ‘ending the interview’ and stage 6 – ‘after the interview’.

Children with SCD and healthy siblings were first asked to draw two pictures. The first picture explored their current self “*Here is a piece of paper and some coloured pencils and crayons. Please can you draw a picture of you doing something you normally do? This may be with your family, friends or other people? You can use any colours that you like!*” and the second picture explored their ideal self “*Here is another piece of paper. There may be some things that you can’t normally do, maybe things that your friends can do but that you can’t. So this time please can you draw a picture of you doing something you wish you could do? This might be with family, friends or maybe some other people? Remember this should be something that you normally wouldn’t do but would like to do? You can use any colours that you like!*” The drawings were then used to explore different aspects of children’s HRQL by asking them what they are doing, how they are feeling and who they are with in their current and ideal pictures. These initial questions were used to extend conversations beyond the drawings and explore these areas in more depth. For example, children were then asked “*What kinds of things do you normally do outside of school?*”, “*Do you often worry or feel sad?*”, “*Can you tell me about your friends and family?*” and “*How do you feel about school?*” Children with SCD were asked about what it is like having SCD and about disclosing it (e.g. “*Have you told your friends that you*

have SCD? Why/why not?” and *“How did they react?”*) and healthy siblings were also asked to share their experiences of SCD (e.g. *“How do you feel about having a brother or sister with SCD?”* and *“How does it affect you?”*).

Children took approximately 5 to 20 minutes to draw their two pictures. Drawing materials included two HB pencils, twelve coloured pencils, twelve coloured crayons, a rubber, a sharpener and two sheets of plain A4 paper. There were more A4 sheets of plain paper available if the children wanted to start their drawing again. Each child was asked what they were going to draw to ensure they had understood the task and that they were going to draw a realistic picture, for example, they should not draw going to the moon.

The interview schedule also explored children’s health behaviours including daily exercise and sport activities and their diet and hydration. Children were asked about their daily health behaviours (e.g. *“Can you tell me about any types of exercise and sport you do every day?”*), preferred health behaviours (e.g. *“What do/don’t you like eating and drinking?”*), health behaviours that they cannot do (e.g. *“Are there things you can’t do (things that maybe your friends can do)?”*), health behaviours that they would like to try (e.g. *“Are there things you would like to eat and drink?”*) and how their condition affects their health behaviours (e.g. *“If you do a lot of exercise or sport do you get tired/feel ill/pain?”* and *“When you’re ill or tired or have pain what can’t you eat and drink?”*).

2.2.3.2 Demographic information sheet

The parent of any child with SCD and/or healthy sibling completed a demographic information sheet. The ‘information regarding the parent’ section consisted of six questions; 1. gender, 2. age, 3. marital status, 4. employment status, 5. occupation/job title and 6. highest level of educational attainment. An ‘information regarding the child’ section consisted of five questions; 1. gender, 2. age, 3. ethnic group, 4. education information and 5. type of sickle cell for children with SCD only.

2.2.4 Procedure and ethical considerations

The study was registered as a research project in line with NHS Trust policy (Research Ethics Committee reference number 14/LO/1548 and Research and Development; see Appendix C) as well as with the Middlesex Psychology Department's Ethics Committee, which adheres to the British Psychological Society (BPS) code of ethics (The British Psychological Society, 2009). The study was approved by both ethics committees.

All potential children with SCD had their diagnosis confirmed through routine NHS pathways. In total, 150 parents and their children who were 5 to 12 years old, who had no co-morbidities that were not related to their condition and received care for SCD at the hospital, as well as their healthy siblings, were invited to participate. Eligible children were identified by the lead consultant at the London hospital from hospital records. Children and their parent were initially posted age-appropriate information sheets to their home address one week before data collection was due to commence providing them with ample time to decide whether they wanted to participate in the study. The initial postal invitation was made by the lead consultant. Therefore, participants had time to read through and fully understand the study before providing informed consent or assent.

Children and their parent were then approached by the researcher during their next routine outpatient clinic appointment. The researcher read through the age-appropriate information sheets with all of the participants including parents (if requested) before they decided whether to participate or not. The researcher fully described the study, their involvement, allowed sufficient time to consider their/their child's participation and sought to resolve any questions or concerns any participant raised. For example, any queries they had with regards to the purpose of the study or their involvement in the study. In addition, the researcher explained how the child's anonymity would be maintained. Firstly, parents

and children were reassured (on the information sheets and consent/assent forms) that all of the data (i.e. interview transcripts and drawings) collected would remain anonymous. Furthermore, each participant was allocated a reference number and therefore was not identifiable. Secondly, all data were stored on password protected computers and in secured cupboards that were only accessed by individuals directly involved in the study. Thirdly, interview transcripts and drawings would not contain any identifiable data i.e. names or names of schools. The researcher also spoke with healthy siblings over the phone or in person, after gaining the parent verbal consent to do so and discussed the research and any concerns healthy siblings raised.

If a child and their parent agreed to participate they were given two choices of locations. The first location was a private room in the outpatient clinic at the London hospital as there would be sufficient time while their child waited to be seen by the consultant and the nursing staff. The second location was their home which may be more convenient if more than one of their children (e.g. one or more healthy children and/or another child with SCD) were going to participate. Before the semi-structured interview began, the parent was asked to provide informed written consent for each participating child on the day of their involvement in the study. If they were not accompanied they were invited to participate in the study at a later date convenient to them. On completion of the parental consent form, the child was also asked to provide written assent before they had any involvement in the study. Interpreting facilities were available within the London hospital if any participant was unable to speak English but all participants could speak English. The parent was also given a demographic information sheet to complete before the drawing tasks and interview began.

Prior to commencing the interview it was important to inform the children of their rights. Firstly, children were informed that they could have a snack or drink, use the toilet,

could withdraw from the study at any time with no explanation required and with no effect on the care they received from the NHS staff and were made to feel comfortable i.e. they were able to explore the room and asked whether they would like the door opened or closed. Secondly, the children were informed that in special situations, if the researcher was a bit worried about them, she would talk to a doctor, after speaking to them first, and that if they did not want to draw or talk about something, they did not have to do so. Thirdly, the children were given the opportunity to look at the digital recorder and shown how it worked and told when the digital recording had begun. It was clearly stated to parents (in the information sheet and consent form) and all participating children (in the consent form) that the interviews would be digitally recorded.

Only the researcher and child were present during the drawing tasks and interview which took approximately 45 minutes to complete. This was to ensure that the child's responses were not influenced by their parents or any other person. Throughout the interview, the researcher generally followed the interview schedule previously discussed, although the children were encouraged to speak openly, discuss their pictures and expand on issues that were important to them. As some sensitive questions were asked with regards to a child's feelings towards their family, friends, school and condition, it was possible that children may have discussed some of their worries or concerns relating to those areas with the researcher. If a child became distressed, anxious or upset or the researcher was concerned about their psychological well-being or about the drawings they had produced, then clinical psychologists who work in the paediatric psychology department (based within the London hospital) would offer immediate support and also support at a later date, if requested. Lastly, at the end of the interview the child was given the opportunity to ask anything else and the researcher also ensured that they were happy

that she either kept or photocopied their drawings (children would retain the original drawings if they wished).

After the interview had finished, the researcher verbally debriefed all of the participants. In addition, the researcher read through the debriefing forms with all of the children, answered any queries they had and they were given a copy of the form to retain. After their child had been debriefed, their parent was also given a debriefing form to retain and they were given an opportunity to discuss this with the researcher who answered any queries they had.

2.2.5 Analysis

Thematic analysis (e.g. Braun & Clarke, 2006) was used primarily to explore the interview transcripts and secondly, to explore the drawings, although analysing the drawings themselves was not the focus of the research as they were mainly used as a tool to aid communication. Drawings were also used to illustrate themes identified from the interviews. Thematic analysis was chosen because it is a flexible technique which is not set to any pre-existing theoretical framework and consequently can be used within different theoretical frameworks such as an essentialist, realist or a constructionist paradigm (Braun & Clarke, 2006). Furthermore, thematic analysis is well-suited to large datasets, providing a detailed description of the dataset (Braun & Clarke, 2006). Braun and Clarke (2006) suggest that thematic analysis can highlight similarities and differences across the dataset which will help in the present research when exploring discrepancies in HRQL between children's current and ideal self and also when drawing out similarities and differences between children with SCD and healthy siblings.

The analysis of interview transcripts was conducted with the assistance of QSR NVivo 10. The researcher took an inductive approach, identifying semantic themes across

the dataset. The researcher's epistemological position primarily focussed on the reality of the child rather than attempting to understand how participants' context shaped how they derived meaning from their experience (i.e., a 'realist' perspective as coined by Braun and Clarke (2006)). Also, participants in the present study were as young as five years old and therefore basing the analysis on what the child had directly discussed or drawn rather than searching for hidden meaning from their experiences seemed more appropriate.

Thematic analysis was implemented following Braun and Clarke's (2006) six phases of thematic analysis. The first phase of analysis involved familiarisation with the data i.e. the interview transcripts and drawings from all of the children. The researcher firstly transcribed all of the digital recordings and then actively read, and re-read through the data and repeatedly viewed the drawings to become familiar with issues arising from the children. During this process the researcher noted down initial ideas, being careful not to disregard anything. The second stage involved the researcher generating initial codes i.e. coding interesting features of the data and then identifying data extracts to support these codes. The third phase involved the researcher beginning to arrange the initial codes into potential themes and collating the supporting data extracts. Some initial codes formed themes or subthemes whereas others were merged together or disregarded. During the fourth phase the researcher reviewed potential themes by firstly viewing them alongside the coded data extracts and secondly viewing them with regards to the entire data set i.e. children with SCD and healthy siblings. Two of the researcher's supervisors also reviewed the themes and subthemes to ensure that the themes generated represented the children's discussions and pictures. The fifth phase involved defining and naming themes, and the most important aspect of this stage was ensuring that the themes were representative of the data extracts and children's drawings. The last stage involved assigning all participants pseudonyms (to ensure their anonymity), writing up the study and ensuring that there was a

coherent story across each theme and subtheme and selecting data extracts and drawings that reflect the story.

2.4 Results

Four themes were identified; “limitations of SCD and adjusted expectations”, “awareness but secrecy surrounding SCD”, “coping with SCD” and “influences on health behaviours”. Each theme contained subthemes as shown in Figure 2.1.

2.4.1 Theme: Limitations of SCD and adjusted expectations

The first theme “limitations of SCD and adjusted expectations” includes three subthemes: exercise and recreational activities, psychological well-being and education and ambitions, and highlights some limitations in children’s lives in these three areas which are partly related to SCD, and creates discrepancies between their current and ideal selves.

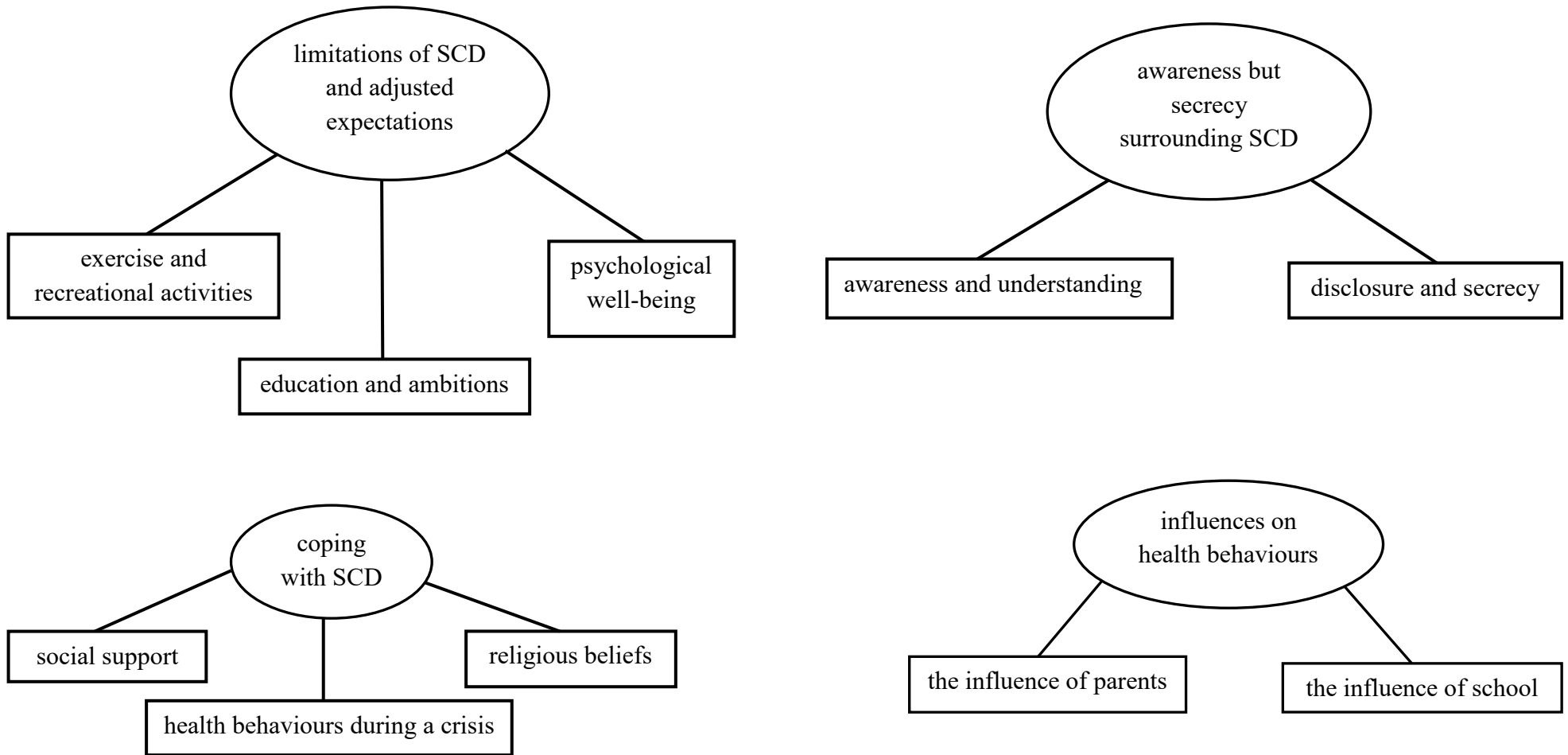


Figure 2.1. Thematic map showing final four themes and corresponding subthemes for children with SCD and healthy siblings

2.4.1.1 Subtheme: Exercise and recreational activities

Both groups of children (those with SCD and healthy siblings) reported that they participated in some types of exercise during physical education lessons or at break times; individually or as part of a team sport, but as shown by the two quotes below, healthy siblings were more likely than children with SCD to also participate in exercise afterschool or during the weekend:

Afterschool I do lots of things!...Running, juggling, jumping, skipping, dance, basketball, having fun and play time...Dance, jumping, skipping, tennis, walking, hockey, then um the one I really like is dancing...I go dance classes on Saturday at 10 and tennis classes on Sunday at 11 (Kanina, sibling, female, aged 5).

One time I had swimming lessons with my sister but I always play skipping and um hula hooping games with my friends at play time and I have P.E. sometimes (Zalika, SCD, female, aged 7).

Many children with SCD, unlike healthy siblings, also described being unable to participate in exercise to the same level as their peers, as a consequence of their condition, which made them feel different from their friends and, at times, meant that they could not be part of school teams.

I only play on Fridays [afterschool] because of sickle cell I can't every day like [redacted] [his friend]. Basically, a lot of sports I do are in P.E. that's it (Emmanuel, SCD, male, aged 11).

This is also demonstrated, for example, by Nailah's (SCD, female, aged 5) current self drawing where she was drawing princesses by herself and in her ideal self drawing where she was "running really fast with [redacted] [her friend]! I'm slow ((sighs)) because, um I'm sick." (Figures 2.2 and 2.3) and also by Jaheem's (SCD, male, aged 12) current self drawing, which was of him playing football on his PlayStation, while in his ideal self drawing he was outdoors playing football with his friends (Figures 2.4 and 2.5). Jaheem described how his condition affects his participation in exercise:

I don't mind...I still play it but I just kick the ball around but I can't run a lot because I get tired and I'm not in the football team. I just want to play the sports like my friends do them and be in the football team (Jaheem, SCD, male, aged 12).



Figure 2.2. Nailah's current self drawing (SCD, female, aged 5).

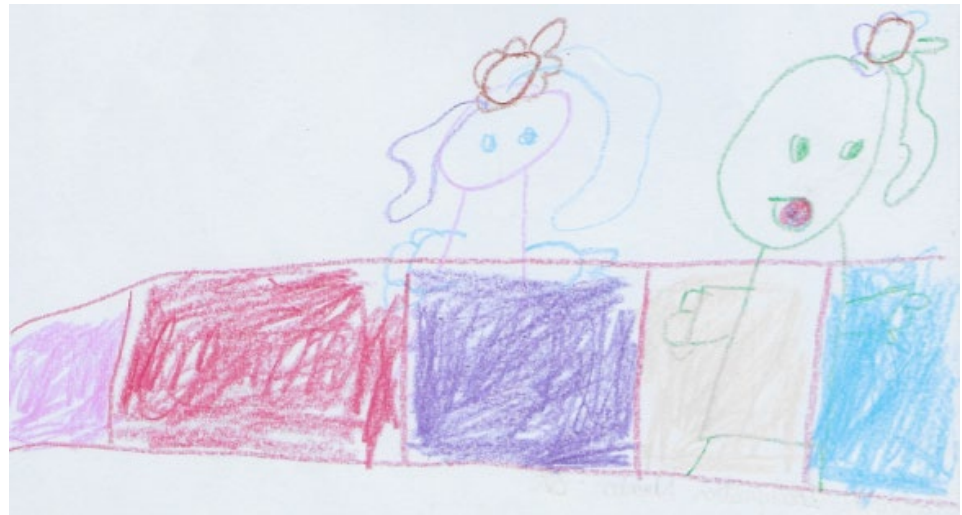


Figure 2.3. Nailah's ideal self drawing (SCD, female, aged 5).

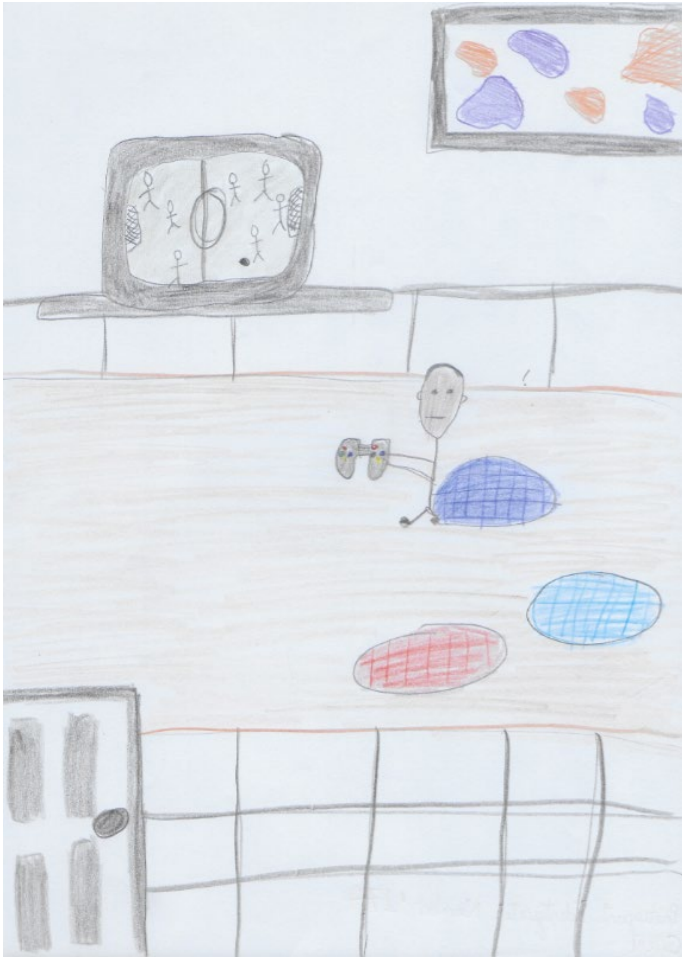


Figure 2.4. Jaheem's current self drawing (SCD, male, aged 12).



Figure 2.5. Jaheem's ideal self drawing (SCD, male, aged 12).

This discrepancy was evident in most children with SCD, who would like to participate in more types of exercise, mostly with their friends, like Daren (SCD, male, aged 11), who in his current self drawing was in his maths class, whereas his ideal self drawing was of himself and his friends playing football, tennis and basketball (Figures 2.6 and 2.7).

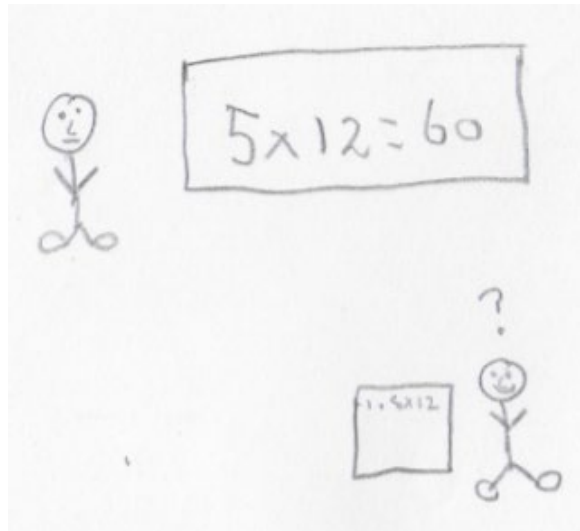


Figure 2.6. Daren's current self drawing (SCD, male, aged 11).



Figure 2.7. Daren's current self drawing (SCD, male, aged 11).

Children with SCD also discussed wanting to be better at sports, again often drawing comparisons with other children and even with sporting professionals:

No. I can't run fast, I get tired. I'm too short, slow and weak but I wish I wasn't. It's all because I have sickle cell! Alexis Sanchez isn't, he's like a machine! I can't run fast, I'm shorter than my friends, have weaker bones than my friends - all because of sickle cell, I can't play outside in the cold, I have to wear lots of clothes, um that's it and have medication (Kasim, SCD, male, aged 9).

However, some children with SCD, explained that they were apprehensive about trying a new type of exercise and would prefer to watch their friends first. For example, in her ideal self drawing Zenna watched her friend do a cartwheel before she would attempt it (Figure 2.8):

No, it's my friend doing cartwheels and I'm watching her.....So I can learn, then then I can do cartwheels (Zenna, SCD, female, aged 8).



Figure 2.8. Zenna's ideal self drawing (SCD, female, aged 8).

In other cases, children stated that they would prefer to be by themselves when trying a new type of exercise as Oria's (SCD, female, aged 6) ideal self drawing highlights where she is doing a cartwheel by herself (Figure 2.9) and is sad "because I'm trying but I still can't do it. Yeah ((hesitation)) because I know it's dangerous so so I shouldn't do it. It's ok, I can do other things!" This shows that children are aware of their physical limitations and do not imagine themselves without SCD, so they are perhaps realistic that these limitations will always be part of their lives.

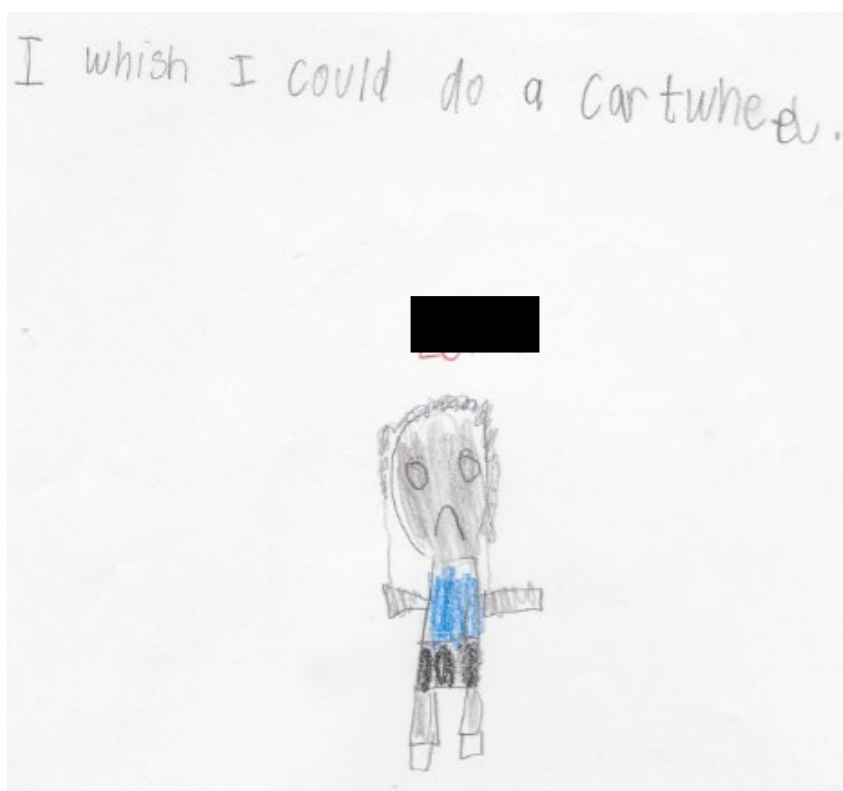


Figure 2.9. Oria's ideal self drawing (SCD, female, aged 8).

Children's apprehension may be because they are afraid that they may not be as good as their peers at sports because of their condition, as discussed earlier by Kasim. It may also be that some children with SCD are afraid that participating in contact sports or doing sports and exercising in cold environments may trigger a vaso-occlusive crisis, and some children have experienced this in the past:

I like swimming but then I get sick and then the water's very cold and brings on my crisis (Aiyana, SCD, female, aged 12).

Both children with SCD and healthy siblings also reported other barriers to participating in exercise, such as their parents are not able to afford sports lessons or are too busy to take them to sports activities, especially if caring for the child with SCD. Healthy siblings reported that they are sometimes limited by their sibling's condition. John's drawings reflect this. His current self drawing was of himself revising in his bedroom while his ideal self drawing was of himself, his brother and best friends playing football in the park (Figures 2.10 and 2.11) as he revealed:

Because my little brother can't play football outside because he has Sickle Cell and because he can't be in the cold and because I have to be with my mum or dad if I'm outside and they're busy working, cleaning, cooking (John, sibling, male, aged 12).



Figure 2.10. John's current self drawing (sibling, male, aged 12).



Figure 2.11. John's ideal self drawing (sibling, male, aged 12).

As suggested by John, healthy siblings also reported that they would like to do activities more with other people, especially family, and they described their current routines as *“suffocating and boring and just same thing all the time”* (Effia, sibling, female, aged 11). For example, in Rihana's (sibling, female, aged 6) current self drawing she was playing hide and seek with her best friend while in her ideal self drawing she was going to Disneyland with her family (Figures 2.12 and 2.13).



Figure 2.12. Rihana's current self drawing (sibling, female, aged 6).



Figure 2.13. Rihana's ideal self drawing (sibling, female, aged 6).

As highlighted by Rihana above, healthy siblings often spoke about wanting to do more exciting recreational activities than children with SCD, for example, in Oban's (sibling, male, aged 9) ideal self drawing he was going to LEGOLAND with his brother (Figure 2.14) and in Elizabeth's (sibling, female, aged 12) ideal self drawing she was on a rollercoaster at Alton Towers with her siblings (Figure 2.15).



Figure 2.14. Oban's ideal self drawing (sibling, male, aged 9).



Figure 2.15. Elizabeth's ideal self drawing (sibling, female, aged 12).

Furthermore, some healthy siblings described being lonely and having no-one to play with when their sibling is hospitalised or ill:

No but it makes me feel left out....Because then I have no one to play with but I can play with my brother but I like playing with my sister [who has SCD] (Jamal, sibling, male, aged 8).

Therefore, both groups of children reported some discrepancy between their current and ideal participation in exercise and recreational activities, which was reflected in their drawings. For children with SCD the discrepancy was due to wanting to participate in more exercise, especially with, and to be like, their friends, and the main barrier to participating in more exercise was their condition. For healthy siblings' the discrepancy was about wanting to participate more in activities with their family or to engage in more exciting recreational activities, and one of the barriers to this was their sibling's condition. Thus while there is some discrepancy, the nature of this differs between children with SCD and healthy siblings. Furthermore, the discrepancy may be less for children with SCD because they appear to be more realistic about the types of exercise they can participate in and are aware that their condition is life-long (ideal self) whereas healthy siblings have higher expectations (ideal self) akin to their classmates or peers because they are not restricted by a physical condition. Having SCD influences PBC (from the TPB) over participating in exercise, either by making certain activities too risky or via children being apprehensive about engaging in activities. However, other control factors, such as parents' lack of financial resources and busy lives, also have an influence.

2.4.1.2 Subtheme: Psychological well-being

Most children with SCD described their current self drawings and especially their ideal self drawings using positive adjectives such as “*happy*” and “*good*” because they were able to take part in a sport or activity that they normally could not do. For example, in Amma’s (SCD, female, aged 8) current self drawing she was smiling while attending Saturday swimming lessons (Figure 2.16), proudly explaining “*I have a green swimming top at the end. I’m doing Stage 4! Look at me! I’ll get my Docklands certificate at Christmas!*” and in her ideal self drawing she was again smiling while attending gymnastics classes (Figure 2.17), although in her description she also used negative adjectives:

Happy at the same time and scared because I might fall off the gymnastics beam and it might hurt me because its high ((pauses)) and because I get hurt more than my friends. My bones are weak and my friends don’t ((pauses)) don’t get hurt so much (Amma, SCD, female, aged 8).



Figure 2.16. Amma's current self drawing (SCD, female, aged 8).



Figure 2.17. Amma's ideal self drawing (SCD, female, aged 8).

This illustrates that children with SCD are aware that they will always have the condition and are being realistic about their expectations. However, they generally do not allow their condition to monopolise their lives and some even discuss some positive aspects of receiving treatments/attending hospital:

I don't think about it [sickle cell disease] because it's not a big deal. I take my medicine every day. I go hospital every 4 weeks which isn't a lot and I like going!...And the nurses are nice to me and they play with me and sometimes I see the same children and we make paper animals! (Keshia, SCD, female, aged 9)

Sickle cell doesn't come in my thinking. I don't go to sleep ((pauses)) or wake up and think about it. It's just there in the background (Tano, SCD, male, aged 8).

Therefore, there does not appear to be a large discrepancy between the current and ideal selves of children with SCD in terms of their psychological well-being. In fact, with one exception, in their ideal self drawings children did not wish that they did not have SCD. This may be because they struggle to see beyond their condition or this may be because they have begun to normalise their condition and have, to some degree, accepted it:

It's just one part of who I am. I find it normal. It's nothing; I have a normal life (Emmanuel, SCD, male, aged 11).

Similarly, most healthy siblings also described themselves using positive adjectives such as “happy” but, in contrast to children with SCD, most healthy siblings used

exaggerated positive adjectives to describe themselves in their ideal self drawings. For example, Hassana (sibling, female, aged 10) was “*happy in this one [current self drawing] and in this one very very very very very happy and crazy excited [ideal self drawing] because I’m on holiday with all my family in Jamaica and not just with my baby sister like in that one*” (Figures 2.19 and 2.20). The contrast between this and Amma’s ideal self drawing (discussed and shown in Figure 2.17), which still involved some concerns about her SCD, suggests that healthy siblings have greater expectations for positive well-being than children with SCD.

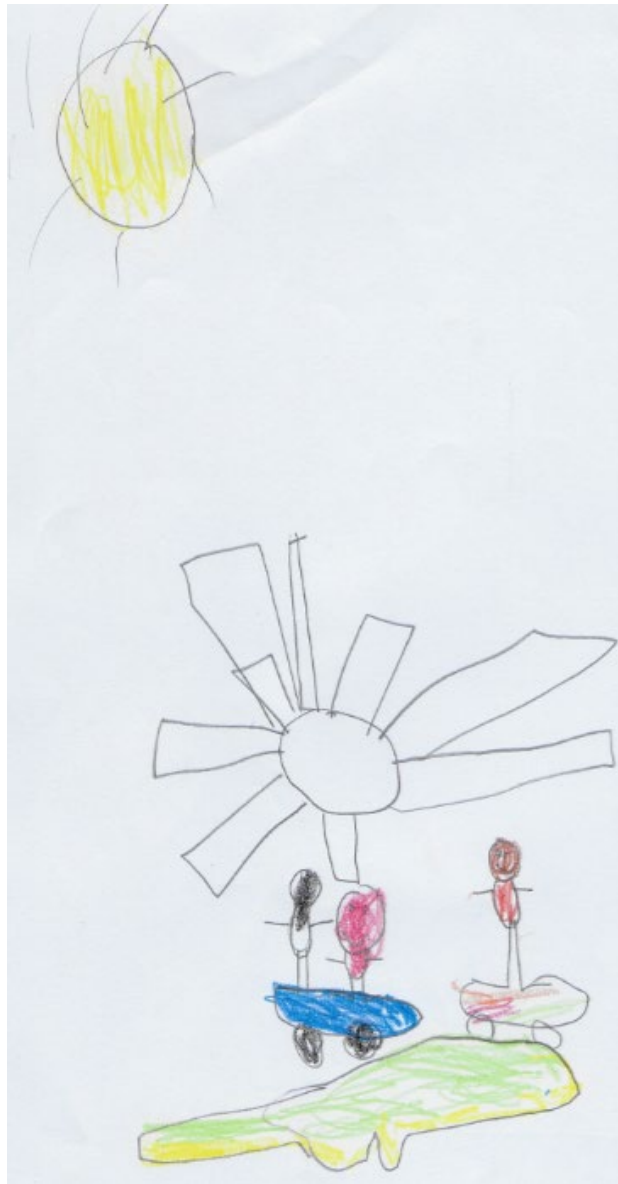


Figure 2.18. Zarif ideal self drawing (sibling, male, aged 7).



Figure 2.19. Hassana's current self drawing (sibling, female, aged 10).



Figure 2.20. Hassana's ideal self drawing (sibling, female, aged 10).

Similarly to children with SCD, most healthy siblings described SCD as “*um normal it’s just normal that he has sickle cell, like that it rains here a lot or I have to go school. Our normal*” (Effia, sibling, female, aged 11), and some even explained that their sibling does not get sick. Nevertheless, unlike children with SCD, most healthy siblings described feeling worried about their sibling with SCD:

I feel worried about my sister going to hospital ((hesitation)) err ((pauses)) and um ((pauses)) it makes me upset, no not really, yeah it does, but going through that pain and saying ((hesitation)) and she’s crying all the time and when I have to come back home ((hesitation)) and and leave her in the hospital that’s hard ((pauses)) and like is she gonna come back the next day um um ((hesitation)) not knowing when she’s gonna come back home and mum saying ‘I’m not sure, maybe’ and then she doesn’t and stuff (Ibrahim, sibling, male, aged 10).

Another issue that may affect psychological well-being for children with SCD is body image. Most children with SCD spoke with strong emotion about how their bodies and physiques are affected by their condition, often describing themselves as small, short and weak and comparing themselves to their friends. This is reflected in David’s (SCD, male, aged 9) current self drawing where he drew a picture of himself and his friend playing basketball where his friend was taller than him while in his ideal self drawing David was taller than his friend and he was leaping over him displaying his strength (Figures 2.21 and 2.22).

My friend’s really tall ((pauses)) his legs are really long. Look, my friends arms are up, really really high [current self drawing] ((whispers)) I can’t do that because I’m

short and weak. I have that thing, begins with s [sickle cell disease] and he don't
(David, SCD, male, aged 9).

*This is my favourite picture [ideal self drawing]! It's me jumping really high, over
my friend ((giggles)) I'm really really really really really really really strong in this
one!* (David, SCD, male, aged 9)

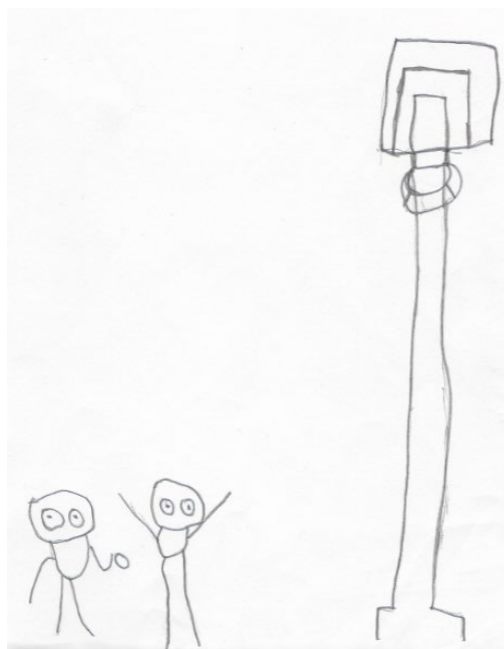


Figure 2.21. David's current self drawing (SCD, male, aged 9).



Figure 2.22. David's ideal self drawing (SCD, male, aged 9).

In contrast, healthy siblings are more positive when they discuss their body image ideals, such as expressing that a change in their physique would lead to a career. For example, Adebayo (sibling, male, aged 6) and Malik (sibling, male, aged 5) explained that if they had muscles then they could become a policeman or a fireman.

Therefore, children with SCD do not appear to have a large discrepancy between their current and ideal selves in terms of their psychological well-being, although some were dissatisfied with their body image. In their ideal pictures children generally did not wish that they did not have SCD. This may be because children have begun to normalise their condition and to accept it and thus have adjusted their expectations accordingly. In contrast, there appears to be some evidence of a discrepancy between healthy siblings' current and ideal selves in terms of their psychological well-being. This may be because they have some higher expectations (ideal self) compared to their sibling with SCD. It may also be that some healthy siblings worry about their siblings' condition (current self).

2.4.1.3 Subtheme: Education and ambitions

Children's school attendance seemed to be affected to varying degrees because of SCD; most children with SCD and healthy siblings reported that the condition had little impact on their education besides experiencing the occasional day of disruption in their school routines and education. This was generally every six months because of their outpatients' appointments and, in the case of transfused patients, approximately every month, for their blood tests and blood transfusions:

Every five weeks I go to hospital, I get a blood test and the next day come for blood transfusion and every six months just to see doctor (Tano, SCD, male, aged 8).

I didn't miss any school this year ((pauses)) and I always do my writing and reading tasks ((pauses)) my favourites the pirate game and the tricky Trevor words! (Nailah, SCD, female, aged 5)

However, a few children with SCD described missing substantial periods of school due to feeling sick, vaso-occlusive crisis and hospitalisations:

School told me at the end of the Year that I missed thirty per cent because of crisis and when I go to hospital (Essien, SCD, male, aged 10).

These disruptions in their school routines may explain why relatively few children with SCD discussed current successes and their future ambitions, but, again, this may be because they could not see beyond their condition. Where they did discuss ambitions, they had adjusted their expectations in light of their SCD:

I don't know, different jobs like like ((hesitation)) well first I wanted to be a dancer ((pauses)) but but I don't think I can because of my bones um I want to be someone nice, who helps (Oria, SCD, female, aged 6).

In contrast, most healthy siblings reported that they were ambitious and had already achieved some successes in their lives which may help them in the future: *"I played the trumpet and then won a scholarship from the Major of London"* (Aisha, sibling, female, aged 9). This is also reflected in their current self drawings. For example, John (sibling, male, aged 12) drew himself revising for an exam (Figure 2.10). In their ideal self drawings most healthy siblings, but no child with SCD, initially chose to draw themselves

doing something challenging, adventurous or ambitious, that it would not be possible for them to do as a child, like Adebayo (sibling, male, aged 6) who wanted to “*climb up a real mountain!*”, or Zarif (sibling, male, aged 7) who wanted to “*go on a motorbike!*” and in some cases it was something that is unrealistic such as Nala (sibling, female, aged 7) who wanted to “*fly!*”. However, the researcher asked them to think of something else to draw that their friends can normally do and they wish they could do.

Healthy siblings reported they were facilitated in their achievements. For example, their parents helped them with their homework, and provided them with private tutors. Children with SCD did not report this. Parents’ encouragement may explain why healthy siblings also reported having higher ambitions compared to children with SCD. Healthy siblings said with strong emotion and self-determination that they would be successful and have a good occupation, attain assets, earn a high income and amass wealth in the future:

Assessment week is easy! I do extra violin assessments and the spelling, reading, writing and maths assessments! I will do the best in my Year so I can get the best job and lots of money and a massive triple house! (Oban, sibling, male, aged 9)

Nah I never miss school because then I can be a a policeman and save people and buy a house and buy a car and buy lots of toys!... I’ll be a really really really good policeman and help everybody and live somewhere with lots of rooms and a big big garden! (Malik, sibling, male, aged 5)

This was also illustrated in their ideal self drawings where Aisha (sibling, female, aged 9) drew how she would like to see what doctor’s do (Figure 2.23).

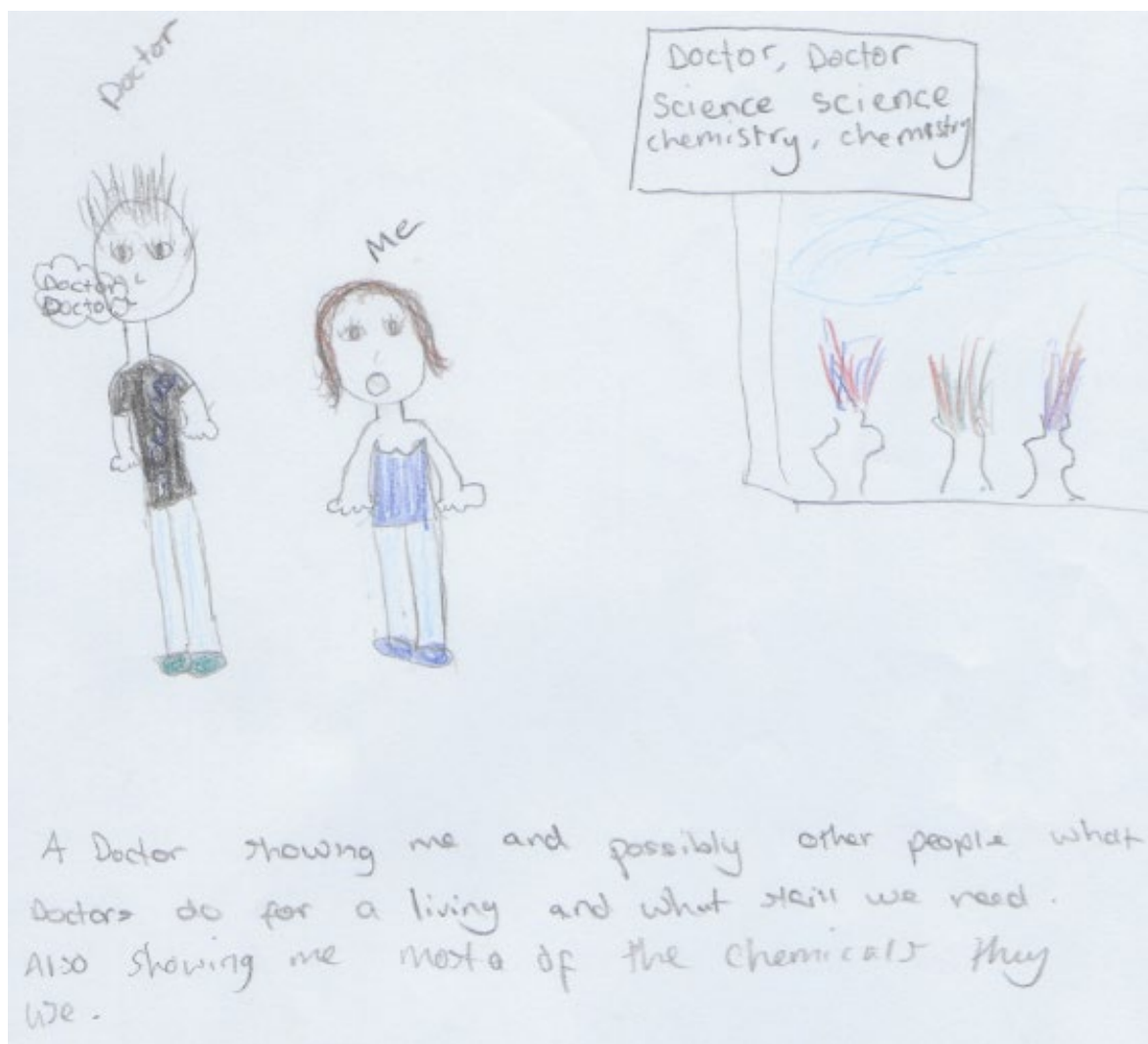


Figure 2.23. Aisha's ideal self drawing (sibling, female, aged 9).

Therefore, children with SCD view their education as mostly unaffected by SCD but they do not tend to report high ambitions and expectations for the future, perhaps because they approach life on a day-to-day basis rather than focusing on their futures. Where they do consider the future, children with SCD appear to consider their ambitions within the constraints of their condition and may just have a more realistic view of what they can achieve. This could suggest that there is more of a discrepancy between the current and ideal selves of healthy siblings who often reported high ambitions and expectations for the future. However, many healthy siblings are also currently higher

achieving (current self) and some are given more opportunities, which may also motivate them to be more ambitious in their day-to-day lives and contemplate their future.

2.4.2 Theme: Awareness but secrecy surrounding SCD

The second theme, “awareness but secrecy surrounding SCD” describes two subthemes; children’s awareness and understanding of SCD and issues surrounding disclosure and secrecy, including some concerns about stigma. Children’s explanations for nondisclosure were mostly influenced by their mothers.

2.4.2.1 Subtheme: Awareness and understanding

All children were aware that they have or their sibling has SCD or at least, are “*sick*”, even if they could not identify the condition. However, ‘sickness’ was only explicitly illustrated by two children in their drawings; Keshia (SCD, female, aged 9) drew a current picture of herself with her mum drinking Exjade (her medication) (Figure 2.24) and Aiyana (SCD, female, aged 12) drew an ideal picture of herself where she was no longer sick (Figure 2.25). This may be because they do not see the condition as the central element in their lives, perhaps because, as discussed previously, they have normalised it, so it is just part of life.

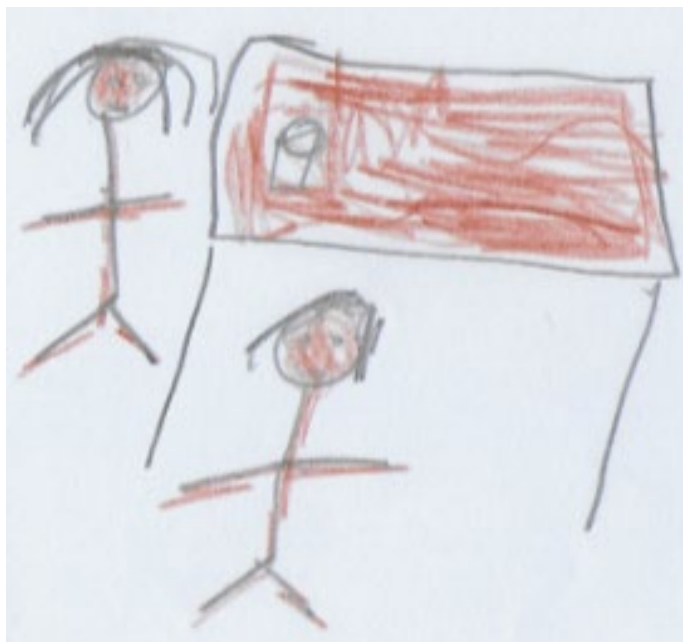


Figure 2.24. Keshia's current self drawing (SCD, female, aged 9).



Figure 2.25. Aiyana's ideal self drawing (SCD, female, aged 12).

Children with SCD and healthy siblings generally had a reasonable understanding of the condition and were able to describe some of the biological mechanisms related to the condition:

I got sickle cell from my mum and dad because that's how people get it and that it's a blood problem because my red blood is banana um moon shape and it's not supposed to be and when they stick together I get something called crisis which hurts (Jaheem, SCD, male, aged 12).

Well well she was born with it because she got it from my mum and dad's blood and my mum had to make sure that I wasn't born with it ((pauses)) she had a DNA test to find out. And she gets crisis sometimes and like it makes her bones weak and eyes yellow (Jamal, sibling, male, aged 8).

Both groups of children also demonstrated a good understanding of what they should be doing e.g. drinking water and taking medication and what they should not be doing e.g. being in the cold and how their actions could trigger a vaso-occlusive crisis:

I take medicine every day before I go to bed to stop me getting sick and I can't play outside in the cold because that makes me sick and I have to wear lots of jumpers and I drink water all the time and and I have to go hospital to stop me getting sick (Nailah, SCD, female, aged 5).

■ *[my affected sibling] can't be in the cold, wear jumpers, drink water, medicine, his penicillin and folic thingy ((hesitation)) go hospital, yeah (Zarif, sibling male, aged 7).*

Some children, especially healthy siblings, compared SCD to other physical health conditions, for example, cancer which they believe is more serious than SCD, or perhaps they make this comparison to reassure themselves:

Um hearing about it [sickle cell disease], I have heard that it's not that serious and that it's not the worst condition. It's not like cancer because people die from that and they look really ill (Aisha, sibling, female, aged 9).

My sister's not sick not like sick people....Like people who are always in the hospital, who look sick, people who have cancer ((pauses)) who die sometimes ((pauses)) that's really sad (Nala, sibling, female, aged 7).

Therefore, all children appeared to be aware of SCD or at least some type of sickness and they also have a reasonable understanding of the condition and the necessary measures to help prevent exacerbating it. Children's drawings generally did not explicitly represent 'sickness' which may be because, as some reported, they see SCD as a life-long condition that can be managed rather than a life-threatening condition like cancer, and, as already discussed in the "limitations of SCD and adjusted expectations" theme, most children see SCD as part of them and their lives.

2.4.4.2 Subtheme: Disclosure and secrecy

Children generally reported that they were secretive about SCD and decided not to tell their extended family and friends, and in one case, the school was not even informed: *“Mum told me not to tell anybody in school about my disease ((pauses)) no friends, teachers, dinner ladies”* (Mark, SCD, male, aged 7). Children with SCD suggested that they were secretive because they did not want to draw attention to themselves or be treated differently:

Because I could be different ((hesitation)) not like my friends ((pauses)) [her teacher] would be really nice to me and my friends wouldn't like that (Zalika, SCD, female, aged 7).

They were also afraid that their classmates may gossip about them, be mean or react badly:

My friends they'll spread a rumour, they'll start a rumour and say oh guess what [redacted] has some sickle problem (Essien, SCD, male, aged 10).

These attitudes and behaviours appeared to be influenced by their parents, especially their mothers. Children reported that their parents provided them with different explanations for not disclosing their condition to their friends; most often suggesting that they would be different from their friends and that their friends would gossip about them and a few even suggested that their condition is contagious. This may suggest they are concerned about stigma or they just consider it a private matter:

Mum said that my friends can get my disease and they would find out I was different, not like them (Mark, SCD, male, aged 7).

Because because ((pauses)) mum says that my friends will tell it to other children and they'll tell it to other children and be nasty to me (Safiya, female, SCD, aged 7).

There may also be some cultural and social influences where people from ethnic minorities believe that any type of 'disease' should not be discussed, even amongst their own family, like Aiyana described:

Eh huh she doesn't really tell people that I have Sickle Cell and she doesn't tell people that she has diabetes....Sickle Cell is embarrassing in Ghana. It puts you lower than aunties, cousins ((pauses)) no one in my family has it ((sarcastically)) (Aiyana, SCD, female, aged 12).

Children also discussed occasions where their friends or classmates confronted them about their condition because, for example, they had noticed that the child was not allowed to play outside or had noticed a symptom of SCD. In some but not all cases, they then decided to disclose. Disclosure led to a mixture of outcomes from acceptance to indifference:

Some of them [friends] just noticed, like on the first day I came, um they saw that my eyes were a bit yellow and so I thought I'd tell em about sickle cell and they said they learnt about it before at school. They were nice about it, just treated me like normal (Daren, male, SCD, aged 11).

There were also occasions where children with SCD believed that they had experienced some form of stigmatisation by their peers or adults. These negative experiences may lead to nondisclosure in the future:

Um I don't know, um. You know the dinner lady in school, I went to get some water and the dinner lady said sit down because ((pauses)) because they'll get it for you and then she looked at my eyes and said why does this girl have yellow eyes and the football coach heard. I did tell the dinner lady why um, and after and then I told my mum but I was a bit sad and um I don't want to tell other people (Zenna, SCD, female, aged 8).

Healthy siblings also reported that they were secretive about the condition, and again this was generally influenced by their mothers. Healthy siblings provided similar explanations for their secrecy; for example, they feared that people would be mean towards their sibling but some also reported that they did not want to be reminded of SCD:

Some of my friends sisters have sickle cell....Because if I talk about it with them because I don't like to remind myself of it because it makes me feel sad when I remind myself about it....Some of my family know she has sickle cell but mum doesn't tell all of them because they gossip and they tell everyone and can be mean to us so mum doesn't want them to know, just the nice family like my auntie but not all (Jamal, sibling, male, aged 8).

There appeared to be secrecy surrounding SCD, with most parents, in particular mothers, encouraging children not to disclose SCD to other people. They may be

concerned about stigma and want to protect their child, or they may see it as a personal issue that they do not want to discuss with anyone, in some cases, possibly due to cultural influences. Nevertheless, disclosure did not generally lead to what children and their mothers were concerned about but there were a few children with SCD who experienced negative reactions and therefore these experiences may have informed their decisions to not disclose their condition in the future.

2.4.3 Theme: Coping with SCD

The third theme “coping with SCD” includes three subthemes; social support, health behaviours during a crisis and religious beliefs. The theme captures the different strategies used to cope with the condition. Mothers provide support in coping with SCD which includes providing children with their favourite food when they are in pain. Children with SCD also described health behaviours that help when they are in pain, for example, reducing their levels of exercise. Healthy siblings report coping with SCD through their religious beliefs.

2.4.3.1 Subtheme: Social support

Children generally reported that they sought general and sickle cell related social support from their mothers. Their social circle from whom to seek support appears limited, as they described having difficult or fragmented relationships with some of their friends or family, including, in a number of cases, their fathers, several of whom were absent: “*My da da daddy didn’t, he wasn’t here, ((hesitation)) I don’t have a daddy*” (Zenna, SCD, female, aged 8). Their mothers supported them in different ways relating to their physical health, such as providing them with medicine during a vaso-occlusive crisis and managing

their day-to day care and treatment, and also supporting their psychological well-being when they are upset:

My mum gives me medicine [during a vaso-occlusive crisis] and I go to sleep. My mum hugs me and lays in bed with me and waits for me to sleep and um sometimes she reads to me (Mark, SCD, male, aged 7).

My mum, my mum helps me because she's with me and cuddles me and she makes me happy when I feel a bit worried about my friends playing with me and my mum helps me take Exjade [medicine] and she brings me my spotty pink and purple blanket (Zeena, SCD, female, aged 8).

However, children with SCD and healthy siblings generally did not report discussing the condition or their feelings relating to the condition with family members, including their mothers, even though they discussed other issues with them such as concerns about arguing with their siblings and friends. Children did not provide any explanation for this in their interviews but again it may be due to having begun to normalise the condition, meaning that it is not something out of the ordinary that requires discussion.

There were relatively few circumstances where children with SCD reported receiving support for their condition from their friends but often, those friends who were supportive, would deflect unwanted attention:

Yeah, so they knew that. And then like when, when um, say like if I was to do certain sports and I said no, some of my friends would ask me and then um my other friends

would say um, 'It's not really your business and not worry about it.' And then they'll keep on asking that person who told them and finally they'd say what I have (Daren, SCD, male, aged 11).

However, as discussed under the “awareness but secrecy surrounding SCD” theme, many of their friends are not aware of their condition and therefore could not provide support. This lack of support may also be because some children reported that they do not have friends or have difficult relationships with them and therefore are not able to discuss their feelings although this may not be related to having SCD:

My friend um ((hesitation)) I have one friend sometimes and she is nine. She makes me sad when she plays with her friends and doesn't want to play with me ((hesitation)) and my teacher plays with me but I feel lonely and sometimes I cry and my teacher is nice to me (Rosi, SCD, female, aged 10).

Mothers were the main providers of social support and the relative lack of support from others with regards to SCD may be somewhat related to non-disclosure as discussed under the previous theme “awareness but secrecy surrounding SCD”.

2.4.3.2 Subtheme: Health behaviours during a crisis

There was no universal health behaviour that children with SCD reported implementing to cope with their sickle-related pain, with the exception of reducing their physical activity, whereby children did not exercise, slept or laydown and participated in relatively sedentary activities such as watching television:

When I have pain I sleep and wait for it to hurt more and then I tell my mum that my belly hurts and she gives me medicine and then I wait for it to stop and I sleep and I might watch TV (Safiya, SCD, female, aged 7).

When I've got pain because of sickle cell, I don't go outside and I don't play football and tennis ((pauses)) no bike, not even basic football dribbling skills. Nothing (Kasim, SCD, male, aged 9).

Children with SCD also reported that there is no common food or drink that helped to cope with, or alleviate their pain. Different coping strategies when in pain seemed to help different children, for example, some children ate normally, some preferred to eat plain food like bread and others preferred their favourite food or drink:

I can't really eat something solid if I have pain in my tummy. Mum makes juices with fruits for me but I don't eat anything solid, I can't, it hurts. But I like to have hot chocolate, my favourite, that helps err it makes me feel happy (Tano, SCD, male, aged 8).

I like to have different drinks at home, like fresh juices or hot tea, especially if I have pain, I wouldn't have water but I do like it, I'd just want something like a home comfort ((hesitation)) that's like a hug ((laughter)) (Emmanuel, SCD, male, aged 11).

Children with SCD also identified more specific foods and drinks that helped their pain, although there was no food or drink consistently reported, and these were often influenced by their mothers' and grandmothers' beliefs and attitudes:

Granny and mummy say that a little bit of coke, ((hesitation)) a little at a time, helps a bad tummy go away and and it actually works! (Oria, SCD, female, aged 6)

However, whilst children with SCD were aware that drinking water may prevent and help them to cope with a vaso-occlusive crisis (as discussed in the “awareness but secrecy surrounding SCD” theme), some children, such as Emmanuel (discussed above) reported avoiding water in favour of a more comforting drink, and only Rosi reported that she drank water when she was in pain:

Well, ((hesitation)) carrots helps my belly hurt [a vaso-occlusive crisis] but I don't like it when it's cooked and water helps my belly hurt (Rosi, SCD, female, aged 10).

Similarly to the “social support” subtheme, children’s mothers provided practical support with coping with SCD. Although children like Oria and Emmanuel reported that specific food or drinks helped, this is most likely due to feeling comforted by having a treat or to the placebo effect. Water is the only drink that would actually help when children experience a vaso-occlusive crisis, although this strategy was rarely mentioned by children, even though they are aware that this helps whilst experiencing pain. Children also reported reducing their levels of physical activity including exercise which helped alleviate their pain. Thus there are some positive attitudes towards health behaviours as being helpful during a crisis, often influenced by mothers’ attitudes, but this does not always translate into behaviour, as children may prefer more comforting and less healthy alternatives during a crisis (e.g. a fizzy drink rather than water).

2.4.3.3 Subtheme: Religious beliefs

Healthy siblings generally spoke with strong emotion about how their religious beliefs provide them with an opportunity to socialise and are a support when coping with their sibling's condition. This was not discussed by children with SCD but as shown in the previous two ("social support" and "health behaviours during a crisis") subthemes, children with SCD receive other support in coping with SCD so religion may not be an important part of their coping mechanism. In contrast, healthy siblings may need to find more emotion-focused ways of coping with their sibling's condition, as problem-focused strategies, such as staying hydrated and resting during a crisis, used by children with SCD are not relevant to them. Most healthy siblings explained that they were of Christian or Muslim faiths, attended Church/Mosque with their family and also attended Sunday school with friends, so it was something that brought them together:

I'm Christian! I have friends in Sunday school and my mum sings in Church err that's it and my sister sings and ((pauses)) and we all go together! (Malik, sibling, male, aged 5)

This was also sometimes reflected in their current self drawings where Kanina (aged 5) drew herself and her family attending Church (Figure 2.26).



Figure 2.26. Kanina's current self drawing (sibling, female, aged 5).

Many healthy siblings also reported that they used their religious beliefs to cope with their sibling's illness which was often influenced by their parents and grandparents:

No she [her mother] saw that I was upset and told me not to be because God is with us and will protect us and my baby sister?... No not really just upset and that I pray a lot and ask God to make us stay here for a long time... Yeah not to leave us and to protect her...It's comforting (Hassana, sibling, female, aged 10).

They discussed how their religious beliefs were a source of comfort in coping with SCD, for example, stating that God watches over and protects their family and in particular

their sibling with SCD on a daily basis and when they are hospitalised and that they believe that God will make them better:

Well it's hard seeing him [sibling with SCD] like um sick but I know that he's getting better from it and that God will like watch after him and help him get better but obviously it's sad and horrible to see him sick and stuff like that (John, sibling, male, aged 12).

In summary, children with SCD are able to use some problem-focused coping where they use different health behaviours such as reducing their physical activities including exercise, eating their favourite foods and taking medicine (often with their mother's assistance) to alleviate their pain, alongside some emotional support from their mothers. In contrast, healthy siblings appear to use emotion-focused coping by seeking solace through their religious beliefs, or through socialising at church. As healthy siblings do not have the condition they may be not be able to focus on some practical coping strategies as children with SCD seem to do.

2.4.4 Theme: Influences on health behaviours

The final theme, "influences on health behaviours" includes two subthemes: the influence of parents and the influence of school. This theme discusses how children's environments (home and school) affect different health behaviours. Children's parents are the most influential, encouraging children to eat healthily, but children's schools influence them to drink water. Parents and sometimes schools discourage children with SCD from participating in exercise because of their condition.

2.4.4.1 Subtheme: The influence of parents

According to both groups of children, parents have the greatest influence on different health behaviours. Participating in exercise was a recurrent issue for children with SCD, as already discussed in the “limitations of SCD and adjusted expectations” theme. Children with SCD generally reported that their mothers discouraged them from participating in exercise because of their physical limitations, for example, their weaker bones and the possibility that they would be harmed and become exhausted more easily than their peers if they participated in particular types of exercise like doing cartwheels or playing rounders and especially contact sports like rugby:

Because I'm sick they say don't do running blah blah basketball ((pauses)) no rugby the worst because you get knocked down and ((shouts)) I'll get very hurt (Shafiq, SCD, male, aged 5).

My mum, you know ((pauses)) says no cartwheels and running because because my mum says my neck and bones is not good (Zeena, SCD, female, aged 8).

Children with SCD also reported that most of their parents do not allow them to participate in outdoors activities because of the adverse effect cold weather has on their condition:

Mum lets me play PlayStation and cards and inside games really ((pauses)) mm because of the cold and because it hurts me (Mark, SCD, male, aged 7).

These restrictions on exercise were spoken of with intense feelings by most children with SCD because they did not like it. While parents' protectiveness is understandable (and based on medical advice), being overprotective could reduce the amount of exercise needed to be healthy and also exacerbate the discrepancy between their current and ideal selves, thus undermining HRQL, so it is important to strike a balance. There were occasions where compromises were reached between the child and their parent:

I want ((pauses)) I really really want to do gymnastics but my mum said I can do swimming lessons or I can do gymnastics on Saturday because my bones are weak to do two and I wanted to do swimming lessons because I can't until Year 4. But ((pauses)) mum said to do one only on Saturday (Amma, SCD, female, aged 8).

In contrast, healthy siblings reported that their parents encouraged them to participate in exercise because it was beneficial to their physical and mental health:

Yeah my mum and dad know that I really like football so they encourage me to do that because I really like it and because it's good exercise because you're always running about so you can stay a good weight and not get like diabetes or something and after it I'm always happy and like more alert (John, sibling, male, aged 12).

Yeah but I do, she [his mother] does, but most of the time I'm active and most of the time she tells me to go outside, don't sit inside, you sack of potatoes and get outside and do some sports or go for a walk or something so yeah (Ibrahim, sibling, male, aged 10).

Therefore, parents discourage children with SCD from exercising whilst encouraging healthy siblings to exercise. In contrast, it appeared that children were treated more similarly by parents when it came to healthy eating and drinking water. For example, both groups of children reported that they are given some autonomy in their food choices such as being provided with options for meals. However, children were also encouraged to eat a healthy diet which included limiting the amount of some foods with high fat or salt content and sugary drinks because they could adversely affect their general physical health and appearance and also may reduce their concentration in school:

Not all the time because my mum said that if I drink fizzy stuff at weekends then my teeth will get rotten and I won't be able to sleep and I'll get hyper and not concentrate at school (Essien, SCD, male, aged 10).

I can't eat chocolate because my mum says, well I do gets spots and get fat, so I don't eat chocolate (Hassana, sibling, female, aged 10).

In contrast to healthy eating, children generally reported that they were not encouraged to drink water whether they had SCD or not, and, on occasions, when they were encouraged, this was mostly to benefit their general health rather than related to SCD:

Sometimes mum says to me and my brother and my sister to drink water because we're made up of water and then my teeth won't go black and fall out! But I only drink apple juice at home, it's my favourite! (Kasim, SCD, male, aged 9)

Thus, healthy eating appeared to be an important issue for parents because of general health benefits but this was unrelated to SCD and children (and parents) did not seem to be aware that their condition could be somewhat managed through their diet. Moreover, parents generally did not appear to encourage children to drink water. Parents may not be aware of the benefits of drinking water for preventing and helping to manage vaso-occlusive crises, but this would have been discussed with them when their child was diagnosed with the condition and also raised on occasions during their child's routine outpatient appointments. It may be that parents do not consider drinking water to be as important or effective as taking medication (which they do encourage as touched on under the "coping with SCD" theme) or having hospital treatments such as chronic blood transfusions. However, they did appear to be aware of being careful when it comes to their child engaging in strenuous exercise or dangerous sports, especially in cold weather. Parent's protectiveness as a result of SCD necessarily reduces the amount of exercise children with SCD participate in but reducing this too much may impair their general health, so it is important to strike a balance. With parents, especially mothers, having the major influence on children's health behaviours, the influence of SNs from the TPB is clearly important. This influence on behaviour may be direct but also indirect, via parents' influence on children's own beliefs. For example, telling children that a health behaviour is good or bad for them may influence their own attitudes towards that behaviours.

2.4.4.2 Subtheme: The influence of school

Children also reported that their schools had some influence on their health behaviours by encouraging all children, regardless of SCD, to have a water bottle in class:

Yeah everyone in my class has their own water bottle on the desk (Jaheem, SCD, male, aged 12).

Well we can only drink water at school and we have water bottles on us everywhere ((pauses)) even on the side of the green at P.E. (Effia, sibling, female, aged 11).

Some children with SCD also reported that their school made allowances for their condition; some teachers reminded them to drink water or refill their water bottle during class:

Um no only I can refill my water bottle during class, my friends have to wait for break or lunch because not, not everybody in my class has a medical condition; there's only one person who I know in my class who has a medical condition (Daren, SCD, male, aged 11).

However, there were also some children with SCD who explained that they did not want to draw attention to themselves by asking for water or to be seen to receive special treatment from the teacher, as also discussed under the “awareness but secrecy surrounding SCD” theme.

If I get water at any time, like, when we can't refill in lessons then the children will know something. I'll be different. Getting special ((pauses)) you know. I don't want that, so I don't ask for water (Tano, SCD, male, aged 8).

Yet, some children reported that their school restricted when they were allowed to drink water and use the toilet, even though they are aware of their condition and most likely have an individual healthcare plan in place. It may be that schools do not fully understand the importance of drinking water for children with SCD:

We're not allowed to drink in the classroom, they only let us drink at the playground ((pauses)) break time and if we ask for water we're not allowed to go to the toilet so can't go to the toilet. They have three rules in school, we can't eat, drink or go to the toilet in the classroom we have to wait (Safiya, SCD, female, aged 7).

As with drinking water, some children with SCD reported that their school was aware that participating in contact sports, overexertion or playing outdoors when it is cold may adversely affect their condition, whereas other children with SCD reported that their school lacked knowledge about this, or at least did not reinforce it during break or lunch times or during physical education lessons:

My teachers know [about the condition]. I can't play outside at break time and lunch time like my friends because it's really really cold ((hesitation)) but my teachers don't know that I can't do lots of running and dangerous things err dodgeball and rugby like mum says because I play them in P.E. (David, SCD, male, aged 9).

Unlike parents, schools did not generally influence healthy eating; in fact, both groups of children often reported eating school dinners that were high in calories, saturated fat and sugar:

At home there's more seasoning, flavour in the food and it's more balanced meal like chicken breast, bit of jerk rice and peas n other nice vegetables in nice healthy sauce and never pudding ((pauses)) sometimes fruit pods and at school it's a bit bland and oily and fatty like chips, wedges, pizza, burgers and always some bland cake and custard (Elizabeth, sibling, female, aged 12).

Overall, children reported that their school encouraged them to drink water, whereas parents influenced their healthy eating and exercise. Parents' influence on children is partly related to their condition, for example, avoiding strenuous or dangerous exercise, but also to their general health, for example, eating healthily and engaging in exercise for healthy siblings. The influence of school is mostly in relation to their general health. This may be because schools lack awareness of the dangers of children with SCD overexerting themselves and some parents may not provide the school with sufficient information about the role of health behaviours in controlling SCD (perhaps because they are not aware themselves) when they work with the school to put an individual healthcare plan in place.

2.4.5 Summary of findings

The first theme "limitations of SCD and adjusted expectations" described different limitations in children's lives. While children with SCD participate in some exercise (generally at school), they would like to participate in more, suggesting a discrepancy between their current and ideal selves in this area. In contrast, healthy siblings would like to participate in more exciting activities such as outings or holidays abroad. However, both groups of children described having a limited social life; children with SCD would like to see their friends more whilst participating in some type of exercise or sport and healthy siblings would like to spend more time with their family whilst engaging in recreational

activities. The exercise and recreational activities of children with SCD and healthy siblings are limited because of the influence of SCD but also due to other circumstances e.g. low financial resources and parents' busy lives. In terms of psychological well-being, healthy siblings may have a larger discrepancy between their current and ideal selves compared to affected siblings. This is likely to be because children with SCD have lower or more realistic expectations and ideals in the context of the limits of their condition. This is further demonstrated because healthy siblings' expectations and ideals and hopes for the future are more ambitious than children with SCD. Therefore, the discrepancy between the current and ideal self of children with SCD is not as pronounced as might be expected. Children try to normalise their condition. As it is all they have ever known they may accept it to some extent and thus their expectations and ideals may be adjusted accordingly.

The second theme "awareness but secrecy surrounding SCD" revealed that children were aware of SCD and had a reasonable understanding of the condition. However, most children were secretive about SCD which was generally influenced by their mothers. The main reasons for nondisclosure were that children feared stigma and bullying and also that children would be different from their friends.

The third theme "coping with SCD" discussed the different strategies that children used for coping with SCD. Children with SCD, via their parents, adopted some problem-focused coping, for example, trying to reduce pain by reducing exercise levels and taking medication and they also received support from their mothers. In contrast, healthy siblings used more emotion-focused coping, especially through their religious beliefs, possibly because it was difficult for them to seek support from their mothers, who were providing this to their affected sibling, but also because the secrecy surrounding SCD reduced other avenues of potential support.

The final theme “influences on health behaviours” captured the main sources of influence on their health behaviours. Parents encouraged all children to have a healthy diet and encouraged healthy siblings to exercise but they discouraged children with SCD from exercising. Children’s schools did not generally encourage healthy eating, but they did encourage children to drink water and also discouraged some children with SCD from participating in exercise. However, schools may not be fully aware of the importance of engaging in some health behaviours and the dangers of engaging in others, perhaps due to a lack of communication between parents and schools.

2.5 Discussion

This qualitative study provided an insight into the HRQL and daily health behaviours, including any discrepancies between the current and ideal self, of children with SCD compared to healthy siblings. This study adopted a Gap approach (GT; Calman, 1984) and the TPB (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975). There are a number of key findings and contributions. Firstly, there was some, albeit limited, discrepancy or ‘gap’ between the current and ideal selves of children with SCD and healthy siblings which was unexpectedly perhaps more pronounced in the latter group of children. Second, both groups of children are secretive about SCD, often encouraged by their mothers. Third, the two groups of children adopted different coping strategies to deal with SCD; broadly, children with SCD used some problem-focused coping (like reducing exercise) and healthy siblings used emotion-focused coping (their religious beliefs). Lastly, parents and schools influence children’s health behaviours but while most parents restrict participation in exercise for children with SCD, they do not encourage hydration which may help manage SCD. There were four main themes generated from the analysis of the interview transcripts and children’s drawings which will be discussed in turn below.

2.5.1 Limitations of SCD and adjusted expectations

The theme on “limitations of SCD and adjusted expectations” largely reflected four areas of children’s lives that are part of multi-dimensional HRQL; their physical functioning, social lives, psychological well-being and education. In the current study, children with SCD appeared to have little discrepancy between their current and ideal self, besides in the physical functioning aspect of their HRQL. Children with SCD often described and drew themselves in their ideal pictures participating in more physical activities, especially with their friends, which they mostly currently only participate in during school time. There is a dearth of research in this area and therefore it is difficult to discern whether children’s interviews in this study may reflect a common experience. Dyson et al., (2010a) did not ascertain the level of participation in compulsory or extracurricular sports at school. In a recent study, Melo et al., (2018) found that children and adolescents with SCA are significantly more likely to be sedentary and participate in less afterschool, evening and weekend activity (i.e. exercise or sports) than their healthy peers. The findings of the present study also revealed that children with SCD would like to be better at sports. This issue has not been explored in children with SCD and therefore it is unknown whether these findings reflect the views of this group as a whole.

In this study, some children with SCD described being apprehensive about participating in new physical activities that they would ideally like to partake in. In previous research, most children with SCD limited their activities to prevent a vaso-occlusive crisis (Panepinto et al., 2012) and some described that unsuitable exercise at school led to a vaso-occlusive crisis (Dyson et al., 2010a). It may be that children with SCD are more realistic regarding their physical abilities and the limitations of their condition. Past evidence has found that children with SCD highlighted that sickle cell pain

is unpredictable and recurrent and therefore planning and participating in any type of activity can be challenging (Panepinto et al., 2012).

In contrast, healthy siblings in the present study did not have a discrepancy between their current and ideal selves in the physical functioning domain of their HRQL. Rather than ideally wanting to participate in fairly basic physical activities with their friends like children with SCD, they instead wanted to participate in more exciting recreational activities with their families, so there was still discrepancy in this respect. This is not surprising given that siblings of children with genetic chronic conditions sometimes described feeling as though they did not receive attention from their parents or being left out as their parent cared for their sibling (Hutson & Alter, 2007; Tregidgo & Elander, 2019). Additionally, some adults recalled that during their childhoods they missed out on family days or activities (Tregidgo & Elander, 2019). In the current study, healthy siblings also explained that part of the reason that they do not currently participate in more recreational activities was because of low financial resources and their parents being too busy (sometimes because they were looking after their sibling). This is in line with Hijmans et al., (2010) study and some of these issues may be because families affected by SCD are often single-parent families, and of lower socio-economic status (SES; Hijmans et al., 2010).

Children's interviews in this study also focused on their day-to-day psychological well-being. Children with SCD did not seem greatly affected by their condition. This is consistent with earlier studies that have taken a gap approach, for example, the current author found that children and adolescents with SCA did not have a lower self-reported discrepancy QoL compared to their healthy peers (Constantinou et al., 2015). There is, however, some literature indicating that many children with SCD worry about attending hospital and getting further complications of SCD (Panepinto et al., 2012), although some

children in the current study reported there were also positive aspects of attending hospital/receiving treatment. Bhagat, Baviskar, Mudey and Goyal (2014) found that role limitation due to emotional problems and fatigue in children and adolescents with SCD was higher than in their healthy peers. The authors postulate that such emotional burden may be associated with children's smaller physical size as well as chronic fatigue (Bhagat et al., 2014). In the present study, some children with SCD expressed feeling dissatisfied with their body image or appearance which may be a consequence of growth failure in this population (Bennett, 2011). Despite this, in previous research, children and adolescents with SCD did not report a lower body image or self-esteem compared to their healthy peers even though they were a smaller weight and shorter (Cepeda, Allen, Cepeda, & Yang, 2000). This may reflect the age range in Cepeda et al's., (2000) study, which included adolescents, as even most healthy adolescents have some dissatisfaction with their body or physical appearance (Jain, Prasuna, & Khandekar, 2015).

In contrast to children with SCD, healthy siblings seemed to have a larger discrepancy in their psychological well-being, partly because they have higher expectations (ideal self) but also because they worry about their siblings' condition (current self). This is supported by previous research (Plumridge et al., 2011). These findings may also provide an explanation for the higher prevalence of depression reported by healthy siblings in comparison to children and adolescents with SCD (Juanita Lee et al., 1997). Healthy siblings' younger age in the present study, which did not include adolescents, may also result in greater anxiety or worry. Past evidence has shown that siblings of children with cancer had a lower QoL compared to adolescents (Houtzager et al., 2004). The present study also found that healthy siblings were more ambitious than children with SCD which was supported by their parents. Similarly, Hutson and Alter

(2007) established that siblings of children with a chronic genetic condition felt as though their parents expected them to excel academically.

The findings of this study provide some support for Gap Theory (GT; Calman, 1984) and suggest that there may be less discrepancy in the HRQL of children with SCD in comparison to healthy siblings (except, unsurprisingly, in the physical domain). In addition, sickness was not obvious in most children's current or ideal drawings in this study and some spoke of the condition being 'normal', which suggests that children are trying to normalise their condition and see it as part of their life rather than the focal point of their life. It is difficult to discern whether the findings in the present study are unique to this sample, children with SCD or chronic conditions more generally. A gap approach has been rarely applied in child populations and when it has, the findings have been inconsistent. Constantinou et al., (2015) found that children with SCA had the same discrepancy QoL as healthy children and Heath et al., (2011) reported that children with chronic kidney disease (CKD) had a higher perceived current QoL and smaller discrepancy QoL compared to healthy children. In contrast, Eiser et al., (1999) found that children with asthma had higher ideal and discrepancy HRQL (i.e. worse HRQL) than healthy children. Children in the current study may have developed good coping strategies (which will later be discussed under the 'coping with SCD' theme) which may provide an explanation for the relative lack of discrepancy in HRQL. It may also be that children with chronic conditions have different priorities, and have adapted accordingly and therefore do not necessarily have a lower QoL (Eiser et al., 2000). In this study, healthy siblings have more ambitions (higher ideals) thus making their discrepancy larger whereas children with SCD have lower ideals and are more realistic about what they can achieve. Heidrich et al., (1994) found that adults with cancer who have smaller discrepancies between their current and ideal self (regardless of their disease severity) have higher levels of purpose in life.

Children with SCD appear to have adapted their expectations or have more realistic expectations and therefore the 'gap' between their current and ideal self may be smaller in some domains of their HRQL than healthy siblings. Furthermore, they are learning to live with SCD which may be due to trying to normalise their condition, having good coping strategies, having positive influences which encourage healthy behaviours and also due to their awareness and the depth of their understanding of SCD. These issues will be discussed in the following themes.

2.5.2 Awareness but secrecy surrounding SCD

Children's interviews revealed that they are aware of SCD/being sick and have a reasonable understanding of the condition for their age, including the precautions that need to be taken to prevent a vaso-occlusive crisis. This is supported by past research (Stegenga et al., 2004). There are no studies that have explored healthy siblings' disease knowledge; although Plumridge et al., (2011) found that siblings of children with genetic conditions sought greater knowledge about their siblings' condition and that some parents had not considered discussing some specific aspects of the condition with them. This may also be the case in SCD but it has not been explored in previous literature. Furthermore, some children in the present study, in particular healthy siblings, viewed SCD as a manageable condition, as opposed to a life-threatening condition, and therefore, even at their young age, they have some perspective, or perhaps they also make this comparison to reassure themselves. This is important because Sharpe and Rossiter (2002) found that chronic conditions that are perceived as life-threatening have a greater effect on children. Thus providing children with greater disease knowledge may alleviate some of their anxieties.

Secrecy surrounding SCD was a common thread throughout children's interviews; they generally do not discuss the condition with their extended family members or friends.

Unsurprisingly, children with SCD did not want to be treated differently from their peers and were afraid of the repercussions of disclosure; namely being the subject of gossip or bullying. Panepinto et al., (2012) reached similar conclusions. However, even without disclosure, children with SCD have some visible symptoms of their condition which their peers may notice, such as jaundice (yellowing pigmentation of the skin and whites of eyes) and a smaller physical stature (Kanter & Kruse-Jarres, 2013; Knight-Madden et al., 2011).

Similarly, in this study, healthy siblings were afraid that children would be mean toward their sibling with SCD. This is in line with past research involving siblings of children and adolescents with inflammatory bowel disease (Akobeng et al., 1999). Moreover, in the present study, healthy siblings did not want to be reminded of SCD and therefore most did not disclose it at school. As highlighted in the previous theme; healthy siblings worry about their sibling and therefore may see school as an opportunity to forget about it or as a distraction from their home life.

Children raised issues around their fear of stigmatisation. In this study, some children with SCD disclosed that they feared experiencing stigmatisation from their classmates or adults. In past research, more disruption to children's lives (i.e. frequent hospitalisations and greater school absenteeism) led to increased self-perceived stigma in children and adolescents with SCD (Ani, Garralda, Anie, Wilkey, & Hodes, 2015). Therefore, the current sample may have included some children with SCD who have missed more school or were more regularly admitted to hospital. The authors also found that more visible symptoms of SCD was not related to greater self-perceived stigma, or perhaps the sample had less disease severity as past studies oppose this viewpoint (e.g. Panepinto et al., 2012; Patel & Pathan, 2005; Stegenga et al., 2004).

In the current study, healthy siblings were also concerned about stigmatisation from members of their family. Plumridge et al., (2011) found that siblings of children with

genetic conditions did not discuss their condition with extended family members or their peers (Plumridge et al., 2011). There may also be a reluctance to disclose chronic diseases in some African communities because of reprisals or cultural beliefs (de-Graft Aikins et al., 2010), for example, in Nigeria, SCD is seen as ‘divine retribution’ (Anie, Egunjobi, & Akinyanju, 2010). In the present study, children mentioned that some members of their families live in Nigeria or Ghana and also that many of their parents or grandparents were raised in African countries and therefore they may be influenced by their cultural views.

Children’s secrecy regarding SCD was somewhat influenced by their mothers in the findings of this study. Mothers’ secrecy may be related to their own difficulty accepting SCD or feelings of guilt for passing the condition to their child, which young adults with SCD recalled occurred during their childhoods in research by Thomas and Taylor (2002). Mothers’ secrecy may also be because culturally or socially there is more responsibility placed on mothers than fathers for SCD (Marsh, Kamuya, & Molyneux, 2011), or perhaps mothers are concerned about stigma which has been found in past evidence (e.g. Burnes, Antle, Williams, & Cook, 2008).

Secrecy surrounding SCD needs to be addressed at individual, family, school/education and societal levels, but firstly it is important to raise awareness among parents and in these communities to tackle any stigma regarding SCD. Part of this process will involve educating people about the condition to help to eliminate stereotypes and cultural blame which is often placed on the mothers who play a pivotal role in helping children cope with SCD.

2.5.3 Coping with SCD

In order to deal with the physical and psychological consequences of SCD, the findings of this study suggest that, mainly with support from their mothers, children with

SCD have adopted specific coping mechanisms to alleviate pain related to their condition (problem-focused coping) whereas healthy siblings tended to ease the psychological burden of SCD using their religious beliefs (emotion-focused coping). Research has suggested that older children with SCD develop their own individual coping techniques for their pain (Panepinto et al., 2012) which could mean that, with increasing age, they learn to better cope with their condition. During a vaso-occlusive crisis, children with SCD explained how, with the support of their mothers, they used health behaviours; they reduced any type of exercise (which is in line with medical advice; NHS Choices, 2019), and consumed their favourite food (which probably helped by providing comfort or possibly acting as a placebo) but they did not generally drink more water, despite the importance of drinking water, which they appeared to understand as shown in the “awareness but secrecy surrounding SCD” theme. This is concurrent with past evidence which found that children with SCD never hydrate during a vaso-occlusive crisis but instead were more likely to use medication or diversional activities (Maikler, Broome, Bailey, & Lea, 2001). Similarly, adults with SCD also drink less water when experiencing a vaso-occlusive crisis (Okomo & Meremikwu, 2012), perhaps suggesting that childhood behaviours are transferred into adulthood.

This study found that children with SCD also sought some social support (both practical and emotional, in the form of comfort) from their mothers but did not report seeking emotional support in terms of discussing their condition or their feelings about it. Patel and Pathan (2005) revealed that children and adolescents with SCA were concerned that their condition was a burden for their immediate family members, including siblings, and therefore they may intentionally not seek this kind of emotional support from them. The social support options available to children with SCD and their healthy siblings may also be limited because, as discussed in the “awareness but secrecy surrounding SCD”

theme, most of their extended family members and friends are not aware of their condition and therefore cannot provide practical or emotional support. Moreover, some children had either absent or fragmented relationships with their fathers and thus could not receive any form of support from them, or indeed their paternal family. A significant proportion of children in the current study are from single-parent families which is fairly common in families affected by SCD (Ilesanmi, 2013). Thus children's mothers have time- and financial demands and also have to care for one or multiple children with SCD, which in turn may especially reduce the time or capacity they have to provide support to healthy siblings. However, it should also be noted that the young children in this study may not have the emotional maturity to recognise or disclose the extent of their mother's emotional support during the interview for this study or they may simply have forgotten to mention it, as demonstrated by Forrester et al., (2015).

Healthy siblings adopted different coping mechanisms to children with SCD because they do not have to focus on dealing with the physical symptoms of the condition, and perhaps also, as discussed above, because their mothers are concentrating on providing support to their sibling with SCD. Religion is an important aspect of many African people's lives and identity (Ilesanmi, 2013) and it provided healthy siblings with comfort in the current study. Royal et al., (1999) found that healthy siblings who had stronger faiths and whose families were fragmented sought emotional support (i.e. sympathy from others), although the authors did not report who provided this support to them. Previous research has also shown that children with SCD use religion as a coping mechanism but that some participants believed that their condition was a punishment from God (Cotton et al., 2012). Attending church/mosque also provided healthy siblings with an opportunity to socialise outside of school, which, as described in the "limitations of SCD and adjusted expectations" theme, was a rarity for them. This may provide an explanation for healthy

siblings' discussions around religion and church/mosque, whereas this was not a subject matter raised by children with SCD who had other concerns or priorities.

Involving healthy siblings in the daily management of SCD may help them to feel more included and help them to spend more time with their family (which was an issue they raised in the "limitations of SCD and adjusted expectations" theme). Evidence examining different coping strategies suggests that where possible strategies should focus on problem-focused coping for both groups of children, which is supported by past literature in SCD (e.g. Ludwig et al., 2018; Sil et al., 2016), and this approach could also be discussed or monitored during routine clinical appointments for children with SCD. An example of problem-focused coping is children's daily health behaviours which can help to exert some control over symptoms of SCD. However, emotion-focused coping strategies for children are also important because practical solutions may not always work even for those with SCD, and both groups of children should be encouraged to discuss their concerns and feelings regarding SCD more.

2.5.4 Influences on health behaviours

There are two principal influences on children's daily health behaviours; their parents and school, which is not unexpected given the age of the children in the sample. Findings in this study revealed that parents discouraged children with SCD from participating in exercise because of SCD, whereas healthy siblings were encouraged to be active to improve their health. This is in line with retrospective studies focusing on adults with SCD where parent or family overprotection during their childhoods limited their involvement in exercise/sports and meant that some participants did not discuss SCD with their parents (Forrester et al., 2015; Thomas & Taylor, 2002). Parental protectiveness is understandable given the potential physical risks associated with SCD. However,

children's condition should not undermine their participation in moderate exercise, which is good for general health (NHS Choices, 2019) and may even improve some aspects of their lives (Ahmed et al., 2015). Retrospective quantitative and qualitative studies in adults with SCD revealed that parental (mostly mothers) protective behaviours during their childhood years negatively affected their self-care behaviours, health outcomes and QoL (Jenerette & Murdaugh, 2008; Jenerette & Valrie, 2010). Moreover, parents discouraging exercise in children with SCD may increase the discrepancy in their HRQL as seen in the "limitations of SCD and adjusted expectations" theme. Thus a balance is needed and it is possible that parents need clearer guidelines when it comes to exercise. In contrast, parents encouraged healthy siblings to participate in exercise. One might expect this as parents' attitudes may simply reflect the predicament of their children; healthy siblings are not restricted by a physical condition, so parents may feel that exercise is good for their health, whereas for children with SCD, they may feel that exercise may be detrimental to their health due to their condition.

Children received similar treatment from their parents regarding healthy eating (i.e. limiting fats and salt) and they did not encourage them to drink water at home irrespective of SCD. This is not consistent with past research which showed that parents of children with SCD encouraged their child to remain hydrated (mostly by drinking water or caffeinated beverages) especially when experiencing a vaso-occlusive crisis or when they have a poor appetite (Mitchell et al., 2004). In the present study it may be that children with SCD did not want to drink water and that parents do not want to pressure them, as also highlighted in previous research (Ievers-Landis et al., 2001), but instead they want to focus on providing something that the child finds comforting. Parents in the current study may also believe that some health behaviours (like reducing exercise levels and taking medication) are more effective than drinking water (as suggested in the "coping with SCD"

theme) or their knowledge regarding SCD may be limited in this area. Panepinto et al., (2012) found that children with SCD would like their family to have a better understanding of SCD, although as discussed under the “awareness but secrecy surrounding SCD” theme, their mothers do not openly discuss SCD which may impair their capacity to learn more about it, including healthy behaviours that may improve their child’s condition.

School also influences children’s daily health behaviours but they seem to have an inconsistent approach in applying and reinforcing them in children with SCD. Primary school children can generally drink water during class, break times and lunch times which is part of a health initiative in England (Department for Education, 2016). During their interviews some children with SCD revealed that their teachers reminded them to drink water or refill their water bottle because of SCD, whereas other children were prohibited from doing so. However, some children did not want to be treated differently from their peers (this is in line with Panepinto’s et al., (2012) study which highlighted that children did not like receiving special treatment), or perhaps this is to ensure that SCD remains a secret (as described under the “awareness but secrecy surrounding SCD” theme). In contrast, Stegenga et al., (2004) found that children with SCD spoke positively about being treated differently by their teachers and peers (although the authors did not provide examples of this or explain what the children meant by ‘different’). This present study also showed that some children were unable to drink water and use the toilet when needed, which past research also found e.g. Dyson et al., (2010a; 2010b). Schools may have limited information regarding the importance of water consumption in children with SCD, although this area has not been explored in previous literature, and they may not know that children with SCD need to pass urine more regularly because they cannot concentrate it (Dyson et al., 2010a).

Schools also have different approaches to exercise and SCD. Some children with SCD disclosed that their schools were aware that some types of exercise may impair their physical health, whereas other children revealed that their schools had limited knowledge of this, or did not enforce it. Dyson et al., (2010a) reported that approximately a third of participants with SCD in their study were '*made to do exercise that was unsuitable*' in school, which in 43% of cases led to a vaso-occlusive crisis. As part of the school admissions process in England, parents are asked to disclose whether their child has a medical condition (Department for Education, 2015). Parents, children and schools (and sometimes healthcare professionals) would then devise an individual healthcare plan (Department for Education, 2015). It may be that some parents in the current study decided not to disclose their child's medical condition (as suggested in the "awareness but secrecy surrounding SCD" theme), or for those that did, Dyson et al., (2010b) postulate this information may not always be communicated to all staff or between school years, or perhaps staff forget about it, or do not have the time to action it.

Children's health behaviours in this study may be further illuminated by considering the three constructs of the TPB (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975); attitude towards the behaviour, subjective norm (SN; perceived social pressure regarding engagement in the behaviour) and perceived behavioural control (PBC; perceived ease or difficulty of carrying out the behaviour). This theme ('influences on health behaviours') shows the importance of children's SNs relating to parents, especially mothers. Thus children's SNs may be a fundamental determinant of their health behaviours, which is not unexpected given their age. This study revealed that children with SCD are influenced by their parents to limit their exercise levels outside of school and both groups of children are influenced by parents to consume a healthy diet at home and influenced by their school to drink water when at school. In past research, SNs predicted

leisure-time exercise (Rhodes et al., 2006) and also moderate to vigorous exercise (Martin et al., 2005b) in healthy children. In past systematic reviews and meta-analyses that have applied the TPB in healthy children, attitude and PBC were the strongest predictors of healthy eating and beverage consumption intentions (Riebl et al., 2015) and healthy dietary patterns (McDermott et al., 2015), rather than SNs as suggested by the current study. However, these studies did not include the influence of parents' own beliefs on children's health behaviours. While Hewitt and Stephens (2007) found that children's TPB beliefs, not parental TPB beliefs, predicted healthy dietary behaviour, in the present study children's health behaviours may be influenced by parental TPB beliefs, which one might expect given the age of the children in this study. For example, children's interviews suggested that parents' attitude toward their health behaviours meant that they either encouraged or discouraged that behaviour (for example, children with SCD participate in less exercise because of their parents' concerns about its effect on their condition). There is little research investigating the effect of parental TPB beliefs on young children's health behaviours but a review of determinants of healthy eating in children showed that positive parental attitude and knowledge about nutrition had an impact on nutrient intakes, pleasant mealtime practices and fewer eating problems (Taylor et al., 2005). Bazillier, Verhiac, Mallet and Rouëssé, (2011) found that parental SNs and conformity to these predicted healthy eating intentions in children which they postulate is because they are still dependent on their parents to purchase and prepare their food (Bazillier et al., 2011). A review of qualitative studies reported that children and adolescents were motivated to participate in exercise when they perceived parental support (Allender, Cowburn, & Foster, 2006). Of course parents' beliefs may not only influence children's health behaviour but may also influence children's beliefs. For example, parents' attitudes towards exercise

being potentially bad for the health of children with SCD may have a direct influence on how much exercise they do but may also mean children hold similar attitudes.

It is imperative to improve parents' knowledge about health behaviours in relation to SCD and general health as they have the most influence on children and shaping their future behaviours and lifestyle choices. For example, the importance of drinking water and striking a balance when it comes to exercise. This may also encourage parents to disclose SCD during the school admissions process and provide the school with a better understanding about the importance of these behaviours in managing, and not exacerbating SCD. Further discussion of practical implications, as well as consideration of limitations of this research study will be presented in Chapter 6.

2.6 Conclusions

Children's interviews and drawings revealed that there was some discrepancy between their current and ideal selves but in different domains of their HRQL; children with SCD appeared to have some discrepancy in their physical HRQL, whereas healthy siblings appeared to have some discrepancy in the psychological and social domains of their HRQL. This provides some support for GT (Calman, 1984) in this population and age group. The current study also highlighted the differences in children's daily health behaviours; healthy siblings participated in more exercise than children with SCD but all children generally had a healthy diet at home and drank water at school regardless of SCD. Engagement in health behaviours was mainly influenced by children's SNs and also parental TPB beliefs which is not unexpected given the young age of participants. Therefore, the next chapter will explore the HRQL and health behaviours of adolescents with SCD compared to healthy siblings to see whether discrepancies in HRQL and some less healthy behaviours (i.e. not drinking water at home) remain during adolescence and

also what and who influences these behaviours. Furthermore, during puberty, some adolescents may engage in more risky behaviours like alcohol use or tobacco cigarette smoking. This is relatively unknown in this population and has not been explored using qualitative methodology, so will also be covered in the next chapter.

Chapter 3: Exploring HRQL, Health Behaviours and Risky Behaviours in Adolescents with SCD and Healthy Siblings (Study 2)

3.1 Introduction

The previous chapter (i.e. Study 1) explored the health-related quality of life (HRQL) and health behaviours of children with sickle cell disease (SCD) compared to healthy siblings. Study 1 supported the use of Gap theory (GT; Calman, 1984) and concepts from the Theory of Planned Behaviour (TPB; Ajzen, 1988; Fishbein & Ajzen, 1975) were also useful but it is unknown whether discrepancies in HRQL and the influences on health behaviours identified in Study 1 also manifest in adolescence. Therefore, these theoretical approaches and a comparison group of healthy siblings were also used when exploring the HRQL and health behaviours of adolescents with SCD in the present study. Furthermore, as the sample in Study 2 will be considerably older, risky behaviours were also explored and a more age-appropriate methodological approach was taken; focus groups rather than the use of drawings.

Past studies have shown that healthy adolescents (Hibell et al., 2012) and adolescents with chronic conditions (Ssewanyana et al., 2017) participate in some risky behaviours like alcohol use, tobacco cigarette smoking and illegal drug use. The prevalence of risky behaviours among adolescents with SCD has not been investigated in the UK (as previously discussed in Chapter 1, section 1.2.2.4). In other countries, alcohol use was the most prevalent risky behaviour in adolescents with SCD (77.7% and 70.1% had ever consumed alcohol) but some also smoked tobacco cigarettes (28.7% and 42.9%) and used illegal drugs (17.4% and 25.4% had ever smoked marijuana) (Asnani et al., 2014; Britto et al., 1998). In healthy adolescents studied across 36 European countries, 87% had tried alcohol, 54% had tried tobacco cigarettes and 29% had used illegal drugs (Hibell et

al., 2012). Hibell et al., (2012) reported that adolescents living in the United Kingdom consumed alcohol; 65% reported drinking alcohol in the past 30 days and 52% reported drinking heavily in the past 30 days.

Engagement in risky behaviours can exacerbate symptoms of SCD, for example, alcohol use can cause dehydration, thus increasing the risk of a vaso-occlusive crisis (Brown, 2012; see Chapter 1, section 1.2.2.4). Compared to children in Study 1, adolescents may be seen as taking greater responsibility for their behaviours, and there may be different factors that facilitate or prevent their engagement in healthy behaviours which have rarely been examined in this population and have never been explored qualitatively (Asnani et al., 2014; Britto et al., 1998). Focus or discussion groups have been used effectively to explore different issues experienced by adolescents with SCD and healthy adolescents', such as Jones and Broome (2001), Porter et al., (2014) and Valenzuela et al., (2013) (as highlighted in Chapter 1, section 1.3.4). However, they have not been used to explore the HRQL and health and risky behaviours of adolescents with SCD.

The aim of the present study was therefore to explore the HRQL, health behaviours (diet, water consumption and exercise levels) and risky behaviours (alcohol use, tobacco cigarette smoking and illegal drug use) of adolescents with SCD, including any differences between their current and ideal self, compared to that of their healthy siblings.

3.2 Method

3.2.1 Design

As discussed in Chapter 1 (see section 1.3.4), the current study adopted qualitative methodology using focus groups. Adolescents with SCD and healthy siblings were compared.

3.2.2 Participants

In line with the study on children in Chapter 2, adolescents who had any co-morbidities that were not associated with SCD were not approached to participate in this study because these may have impaired their HRQL. Healthy siblings were asked to participate if they had a sibling with SCD and did not have any other physical or psychological health condition that may have affected their HRQL.

A total of 44 adolescents participated in eight same sex focus groups. A convenience sample of 23 adolescents with SCD and 21 healthy siblings were recruited to the study from two London hospitals; approximately half of the participants were recruited from each hospital. A further five participants were recruited but did not participate; three participants were not able to attend (a female adolescent with SCD experienced a vaso-occlusive crisis, a female healthy sibling experienced a bereavement in her family and a male healthy sibling had a job interview) and two healthy siblings did not participate because no further groups were being organised. Gender was equally distributed, and their ages ranged from 13 to 17 years old, with adolescents with SCD ($M = 14.87$, $SD = 1.39$) not significantly older than healthy siblings ($M = 14.81$, $SD = 1.44$; $t(42) = 0.14$, $p = .889$). The majority of adolescents with SCD had the most prevalent and severe phenotype; 18 (78%) had HbSS and the remaining 5 (22%) had HbSC. The majority of adolescents with SCD, 19 (83%) and healthy siblings, 17 (74%) were of Black African ethnic origin, with the remaining 4 (17%) adolescents with SCD and 4 (19%) healthy siblings of Black Caribbean ethnic origin. A complete description of sample characteristics is shown in Table 3.1.

Adolescents with SCD and healthy siblings aged 13 to 15 years old were also required to be accompanied by a parent as they needed to provide informed consent on behalf of their child which was made clear to them before they attended the focus group.

Adolescents with SCD and healthy siblings aged 16 to 18 years old did not need to be accompanied by a parent as they were able to provide informed consent for themselves. Adolescents with SCD and healthy siblings aged 13 to 18 years old had to speak and understand English to participate in the focus groups.

Table 3.1

Descriptive Statistics for Socio-Demographic Characteristics of Adolescents with SCD and Healthy Siblings

<u>Participants</u>	Adolescents with SCD M(SD) or % n	Healthy siblings M(SD) or % n
<u>Adolescent variable</u>		
Age	14.87(1.39)	14.81(1.44)
Gender	52% Male	48% Male
Ethnicity	82.6% Black African, 17.4% Black Caribbean	81% Black African, 19% Black Caribbean
SCD phenotype	78% HbSS, 22% HbSC	100% HbAA
<u>Parent variable</u>		
Age	43.52(6.93)	45.29(7.46)
Gender	9% Male	14% Male
Marital status	44% Married/cohabiting, 57% Single/separated/divorced	48% Married/cohabiting, 52% Single/separated/ divorced/widowed
Employment status	74% Employed, 4% In education, 22% Unemployed	76% Employed, 5% In education, 19% Unemployed
Highest educational attainment	39% Higher degree or degree, 13% Foundation degree or diploma, 48% A Levels or below	38% Higher degree or degree, 24% Foundation degree or diploma, 39% A Levels or below

3.2.3 Materials

There was a focus group schedule (Appendix A) which was used by the researcher as a guide to what issues were to be explored. Each participating adolescent with SCD, healthy sibling and their parent received an information sheet, consent form and a

debriefing form (Appendix B). There were two sets of information sheets and debriefing forms; one for adolescents with SCD and healthy siblings and one for parents or carers. There were also four sets of consent forms; one for adolescents with SCD, one for healthy siblings, one for parents of adolescents with SCD and one for parents or carers of healthy siblings. In addition, the parent or adolescent if aged 16 to 18 years old also received a demographic information sheet (Appendix B). All of the materials described, except for the focus group schedule, were available in two hospital site specific versions i.e. including the hospital logo and contact details of the consultant responsible for caring for patients with SCD.

3.2.3.1 Focus group schedule

The focus group schedule gives an indication of the issues that were explored but not necessarily the exact manner or order in which the questions were asked; this was dependent on what was raised by, and considered to be important to adolescents. The focus group schedule was developed and piloted with five adolescents with SCD ($M = 15.12$, $SD = 2.10$) and five healthy siblings ($M = 14.94$, $SD = 1.92$) aged 13 to 17 years from one of the London hospitals. Adolescents with SCD and healthy siblings were asked to make suggestions about what could be explored (in relation to their HRQL, health behaviours and risky behaviours), whether the questions were relevant to them and whether they understood what they were being asked.

The focus group schedule consisted of four stages; stage 1 – ‘gaining informed consent’, stage 2 – ‘establishing a rapport’, stage 3 – ‘introducing the research’ and stage 4 – ‘beginning the focus group’. Under stage 4 – ‘beginning the focus group’ there were three main parts; ‘part one: health-related quality of life’, ‘part two: health behaviours’ and ‘part three: risky behaviours’ and a total of twenty-one questions which were followed by

'probes' which facilitated discussions amongst group members. The first part of the focus group schedule included 11 questions which focused on adolescents' HRQL such as their daily lives (for example "*What kinds of things do you normally do in your free time like evenings and weekends?*"), important people in their lives (for example, "*Is there anything you'd like to change about the people in your lives or the relationships you have with people?*"), education, training or work (for example "*What are the positives and negatives of going to school, college, training or work?*"), emotional well-being and physical health (for example, "*How do you feel physically in your day-to-day lives?*") and SCD (for example, "*Who knows that you (your brother or sister*) has SCD?*"). Participants were asked about their current self and ideal self and the researcher explored any differences. The second part of the focus group schedule included five main questions regarding adolescents' health behaviours such as their daily exercise (for example "*How do you feel after participating in different types of exercise?*") and their diet and hydration (for example "*Are there things that you can't eat and drink and would like to? Why?*"). Lastly, the third part of the focus group schedule also included five main questions which explored different aspects of adolescents risky behaviours including their knowledge (for example "*What do you know about alcohol, smoking and other substances?*"), participation (for example, "*Has anyone ever tried alcohol, smoking or other substances?*") and influences on their behaviours (for example "*Who or what influenced your decision to try/not try alcohol, smoking or other substances?*") and also the importance of engaging in behaviours (for example, "*Why do you think it's important to be careful about how much you engage in the health behaviours we have spoken about today like exercise, what you eat and drink, and drinking alcohol and smoking?*"). Similar questions were explored for adolescents with SCD and healthy siblings with the exception of exploration regarding how engagement in different health and risky behaviours can affect their condition.

3.2.3.2 Demographic information sheet

The parent of any adolescent with SCD or healthy sibling aged 13 to 15 years old or adolescents with SCD or healthy siblings aged 16 years old and over completed a demographic information sheet as was provided to parents of children in Study 1. The demographic information sheet consisted of two sections; 'information regarding the parent or carer' and 'information regarding the child'. The 'information regarding the parent or carer' section consisted of six questions; 1. gender, 2. age, 3. marital status, 4. employment status, 5. occupation/job title and 6. highest level of educational attainment. The 'information regarding the child' section consisted of five questions; 1. gender, 2. age, 3. ethnic group, 4. education/employment information and 5. type of sickle cell (which was only applicable for adolescents with SCD).

3.2.4 Procedure and ethical considerations

The study was registered as a research project in line with NHS Trust policy (Research Ethics Committee reference number 14/LO/1548 and it was also registered with the Research and Development departments of the two London hospitals; see Appendix C) as well as with the Middlesex University Psychology Department's Ethics Committee, which adheres to the British Psychological Society (BPS) code of ethics (The British Psychological Society, 2009). The study was approved by all of the ethics committees.

All potential adolescents with SCD had their diagnosis confirmed through routine NHS pathways and were identified from hospital records by the lead consultant at each NHS hospital. Two-hundred and seventy adolescents with SCD aged 13 to 18 years and their parent were initially posted age-appropriate information sheets to their home address one week before data collection was due to commence. The initial postal invitation was made by the lead Consultants at each participating hospital. Therefore, all adolescents had

time to read through and fully understand the study before providing assent or informed consent.

Adolescents and their parent were then approached by the researcher during their next routine outpatient clinic appointment. However, not all of the two-hundred and seventy potential participants were approached due to time restraints of the research and practical issues (e.g. adolescents were returning to school or college after the summer holidays and participating in focus groups may have interrupted their education and personal plans or activities). Additionally, in previous research involving adolescents between six and nine focus groups were conducted (Laverty et al., 2015; Porter et al., 2014; Whitehead & Biddle, 2008) and therefore eight focus groups in the current study was considered sufficient and no further participants were approached once these had been conducted.

The researcher provided all participants with the appropriate information sheet to read through, discussed the study with them and their involvement, including having to attend the hospital on a day that they would not normally attend, and sought to resolve any questions or concerns any participant raised. It was also clearly stated how their anonymity would be maintained. Firstly, each participant would be allocated a random reference number to maintain their anonymity. Secondly, that all focus group transcripts were stored on password protected computers and in secured cupboards that were only accessed by responsible individuals directly involved in the study. Thirdly, focus group transcripts would not contain any identifiable data i.e. names or the area where they live. Lastly, adolescents were informed in the information sheets that they would participate alongside their peers of the same gender and also that family members could not participate in the same focus groups to ensure they felt at ease discussing sensitive topics and also that their anonymity would be maintained. Moreover, the researcher asked parents or carers to

provide healthy siblings with a copy of the information sheet, describe the study to them and, if they expressed any interest, ask whether they would be willing to discuss this further with the researcher.

After a week, the researcher contacted the parent by telephone or email (depending on their preferred form of communication) to enquire whether healthy siblings would like to discuss the research further, and where this was the case, the researcher answered any queries they had. All participants who agreed to take part in the research were contacted by the researcher via telephone or email to arrange a convenient day and time for the focus group. Focus groups were conducted on a Saturday or a Sunday afternoon in a private room near the outpatient clinic at the two participating London hospitals. Adolescents attended the hospital that they or their sibling were a patient of, as this was most familiar and convenient to them. It was clearly stated to parents or carers and all participating adolescents that the focus group would be digitally recorded.

Before the focus group began, the parent was asked to provide informed written consent for each participating adolescent aged 13 to 15 years old on the day of their involvement in the study. On completion of the parental consent form, the adolescent aged 13 to 15 years old was also asked to provide written assent. Adolescents aged 16 years or older provided written informed consent on their own behalf and their parent was aware that they did not have to attend on the day. All participants were made aware that they could change their minds with no effect on the care they or their sibling received at the hospital they attend. Before commencing the focus groups, the parent or adolescent (if aged 16 years and over) were asked to complete a demographic information sheet which was about the adolescent participating in the focus group and the parent providing informed consent. The researcher asked older adolescents to complete information regarding a parent that they live with; it was their choice whether they chose their mother

or father. The researcher made it clear that everything discussed in the focus group would be confidential and that *“only in exceptional circumstances I may become concerned about your life or well-being is at risk would I speak to a consultant about what you have discussed with me. This is very unlikely and I would tell you before I speak to a consultant”* (this was stated in the information sheets).

At the beginning of the focus group adolescents were asked how they would like the chairs arranged i.e. circle, rows etc., they were informed that they could have a snack, drink or use the toilet at any time and that they could withdraw from the study at any time with no explanation required and with no effect on the care they, or their sibling received from NHS staff. They were clearly advised that *“If I ask you something that you don’t want to discuss in the group, please just tell me that you’d rather not go into that. It is your choice”*. Then introductions and icebreakers were used to introduce the adolescents to the researcher and to one another and to create a more relaxed, open and safe environment, for example, they were asked to state their age, favourite band or singer and something interesting about themselves. Since first names were used during these initial introductions the digital recording did not begin until after this to help maintain confidentiality. The researcher clearly informed adolescents when the digital recording commenced. The discussions were facilitated by the researcher and broadly followed the focus group schedule, depending on what issues or areas were of importance to the adolescents in their particular focus group. Moreover, participants were encouraged to speak openly and also to listen and respect other group members.

During the focus group, the researcher asked sensitive questions regarding adolescents’ feelings towards important people in their lives, their education, condition and their participation in health behaviours and also risky behaviours. Therefore, adolescents may have shared some of their concerns regarding those areas with the researcher. If an

adolescent became distressed, anxious or upset or the researcher was concerned about their psychological well-being, then she would contact a consultant or clinical psychologist who worked with the paediatric sickle cell and thalassaemia team (based within the hospitals) who would offer telephone support and if requested, or deemed necessary by the health professionals, support at a later stage. Toward the end of the focus group, adolescents were given the opportunity to discuss other areas that they believed were relevant, or should have been considered during the group discussions regarding their lives, health behaviours and risky behaviours.

There were separate same sex focus groups of between five and six adolescents with SCD or healthy siblings. The four focus groups involving adolescents with SCD lasted between 81 and 96 minutes and the four focus groups involving healthy siblings lasted between 73 and 87 minutes. All focus groups took place in a private outpatient clinic room where only the researcher and adolescents involved in them were present. The door where the focus groups took place was closed and remained closed until the research finished maintaining confidentiality.

After the focus group had finished, the researcher verbally debriefed all of the participants individually. They were also provided with a 13 to 18 year old debriefing form to retain. Moreover, a debriefing form was also provided to their parents or carers to retain. All participants were given the opportunity to discuss the debriefing form with the researcher and raise any questions they had. The researcher also asked participants whether they would like to receive a simple age appropriate leaflet regarding the overall findings of the research and those who agreed provided their contact details. Before leaving the hospital, parents or carers (of adolescents aged 13 to 15 years old) and adolescents signed a form acknowledging that they had received a financial reimbursement to cover costs of

travel and as an acknowledgement of their time; parents or carers received ten pounds and adolescents received twenty pounds.

3.2.5 Analysis

The focus group transcripts were analysed using thematic analysis as described by Braun and Clarke (2006). Thematic analysis was chosen to analyse the focus group transcripts for the reasons discussed in Chapter 2; it is a flexible technique which is not set to any pre-existing theoretical framework, it is appropriate for larger datasets, provides a detailed description of the dataset and can highlight similarities and differences across the dataset, for example, between adolescents current and ideal selves, and between adolescents with SCD and healthy siblings (Braun & Clarke, 2006).

The analysis of focus group transcripts was conducted with the assistance of QSR NVivo 10. As described in Chapter 2, the researcher took an inductive approach, identifying semantic themes across the datasets. The researchers' epistemological position predominantly focussed on reporting the reality, experiences and meaning for adolescents who participated in the focus groups i.e., a 'realist' perspective (Braun & Clarke, 2006). This position assumes that through language (adolescents' discussions) participants can convey their meanings and experiences (Braun & Clarke, 2006) which in Study 2 includes living with SCD. Moreover, this type of analysis is not fixed to a pre-existing theoretical framework (Braun & Clarke, 2006) which may be more appropriate when exploring the HRQL and health behaviours of adolescents with SCD and healthy siblings, which is relatively unknown using this methodological approach.

Braun and Clarke's (2006) six phases of thematic analysis (as described in Chapter 2) were also applied when analysing the focus group transcripts. In summary, in the first phase, the researcher became familiar with all of the focus group transcripts, transcribed

them, actively read and re-read them and wrote down initial ideas. Phase two involved generating initial codes and then identifying data extracts to support these codes. In phase three, the researcher began to arrange the initial codes into potential themes and collated the supporting data extracts searching for themes. During stage four the researcher reviewed themes in relation to the coded extracts and each dataset and in stage five ensured that there were clear definitions and names for each theme. The last stage involved presenting a written story that was supported by compelling data extracts.

The themes and subthemes were also reviewed by two of the researcher's supervisors to ensure that the final themes and conclusions drawn represented the adolescents' discussions (see Appendix A).

3.3 Results

The analysis of four verbatim focus group transcripts from adolescents with SCD and of four verbatim focus group transcripts from healthy siblings led to the identification of four themes across groups; "awareness and disclosure of SCD", "coping with SCD", "influences on health behaviours" and "education and beyond". Each theme explored the similarities and differences between adolescents with SCD and healthy siblings. Themes and subthemes are shown in Figure 3.1.

3.3.1 Theme: Awareness and disclosure of SCD

The first theme "awareness and disclosure of SCD" includes two subthemes: "awareness and understanding" and "disclosure of SCD". This theme shows that all adolescents are aware of, and that most have a good understanding of the condition. There are various explanations among adolescents' for their disclosure of SCD and healthy siblings gave differing explanations for their decision to not tell their friends.

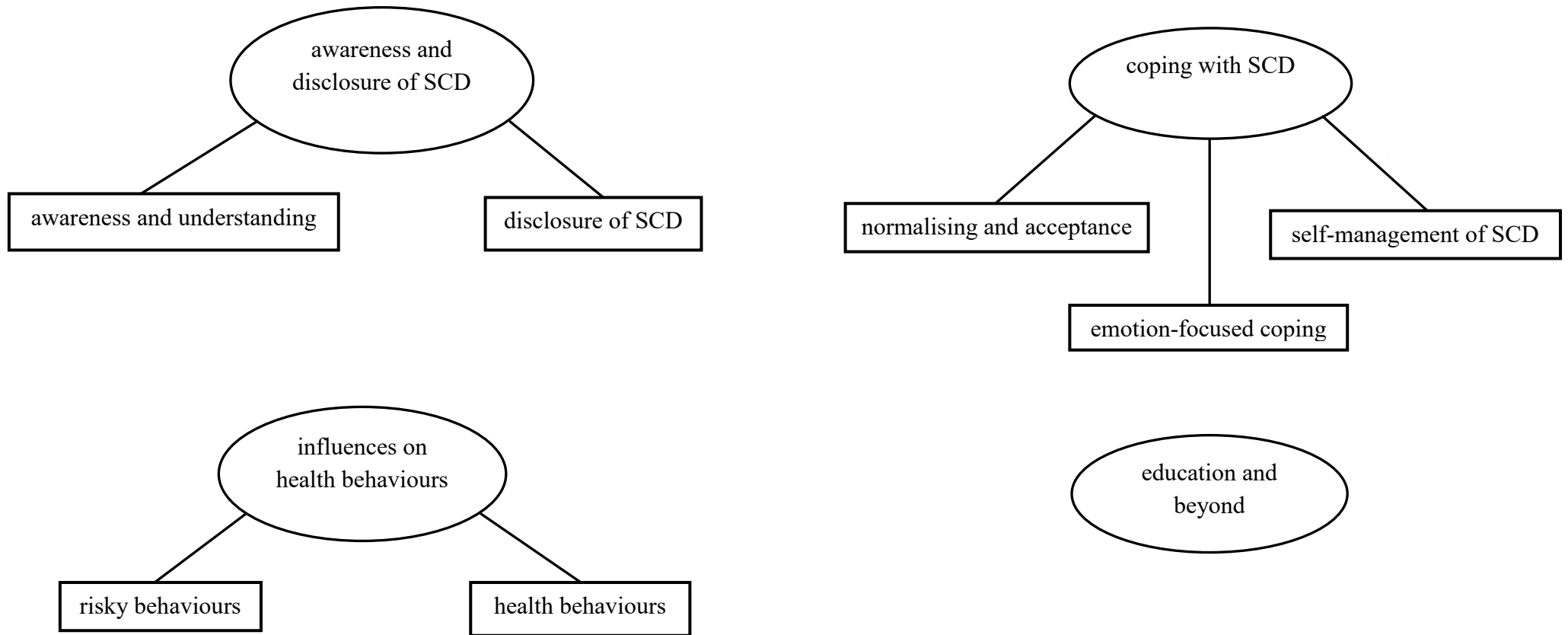


Figure 3.1. Thematic map showing final four themes and corresponding subthemes for adolescents with SCD and healthy siblings.

3.3.1.1 Subtheme: Awareness and understanding

All adolescents were aware that they or their sibling has SCD. Adolescents with SCD recalled attending hospital from a young age, taking medication and being aware that they were unwell, although they did not report when they were first aware because they were unable to remember:

I've always known that I'm sick, that I've got something but I don't remember exactly when I knew that it was called sickle cell disease. Obviously I've been going to hospital consistently my whole life and I've always had medicine, you know hydroxyurea or penicillin. But I've always known that I have something but I couldn't have said that I had sickle cell disease when I was 5 or 6 because, well, who would know that? (Akeem, SCD, male, aged 15, group 5)

Similarly, healthy siblings also recalled being aware of their siblings' condition from a young age and certainly by the time they began primary school and they tended to view it as just part of life. This is demonstrated through Bolade's (sibling, female, aged 13, group 8) narrative: *"I've always known about my brother's sickle cell ((agreement)) practically from birth, it's just always been a small part of our lives"*.

Both adolescents with SCD and healthy siblings were generally able to explain the aetiology and clinical manifestations related to SCD and preventative measures such as taking penicillin (which is an antibiotic to prevent infection) as described below by Adewale. They were even aware of some protective elements of the condition, as highlighted below by Balafama, suggesting that they were also able to see beyond the negative elements of the condition:

I know they can't get malaria. Because in West Africa there's a high amount of people with sickle cell disease, because there's a high amount of malaria. So the cells evolved with them and they can't get it. My point is that it's not all doom and gloom (Balafama, sibling, female, aged 15, group 4).

I understand all about sickle cell disease. It's inherited from your parents, one in four chance, they have to have a sickle gene but not necessarily sickle cell disease and you have moon shaped red blood cells that can stick together and that's when a crisis happens and you get infections more easily and you have low oxygen in your body (Adewale, SCD, male, aged 14, group 5).

Adolescents with SCD and healthy siblings also had a good understanding of what they should do e.g. mostly drinking water and taking pain medication and should not do e.g. playing outdoor sports in the cold or being exposed to cold environments as this may trigger a vaso-occlusive crisis:

Yeah, they're kind of sickle-shaped, like a banana shape. So, anyway, then they clog the artery and cause a crisis and that's why they need to drink a lot of water, not do too much exercise especially in cold weather and take medicine, so it don't happen, so they don't get a crisis (Bron, sibling, male, aged 16, group 3).

Adolescents with SCD and healthy siblings explained that they learnt about the condition during school lessons and online, although they generally did not mention approaching their parents or medical professionals when they wanted to learn more about SCD. Moreover, adolescents with SCD and healthy siblings did not feel that it was

necessary to discuss the condition with their family because it is just part of their life. Additionally, the tendency to go online for information probably reflects a generation that has grown up in an internet era:

Sickle has always been part of our family. There's no need for big conversations about it, if I want to learn about it I surf the net like with anything I want to learn about and my siblings will do the same ((pauses)) my mum would probably go to the library! ((laughter)) (Amara, SCD, female, aged 17, group 6)

Therefore, all adolescents were aware of SCD and most had a good understanding of the condition and also what measures they should take to manage the condition irrespective of whether they had SCD or not. Their knowledge of SCD has been acquired by chance, during school lessons and also sometimes intentionally, where they have sought information about the condition online. Yet, it is also likely that adolescents with SCD learnt about their condition during their hospital appointments even though they did not discuss this. This may be because they cannot recall these initial conversations with medical professionals from when they were very young and see SCD as being something that has always been part of their lives. Moreover, families who live with SCD may not feel that it is necessary to discuss it, again, because it has always been part of their lives, so they have normalised it.

3.3.1.2 Subtheme: Disclosure of SCD

Adolescents, especially healthy siblings were selective about to whom and under what circumstances they disclosed SCD. Most adolescents with SCD chose to disclose their condition to their family and close friends and in some cases they decided to tell all of

their friends. However, a few male adolescents had not told any of their friends about their condition because, for example, *“telling your friends doesn't change anything”* (Abidemi, SCD, male, aged 15, group 1) or they *“didn't want to be different from other people”* (Akono, SCD, male, aged 15, group 5).

Adolescents with SCD provided different explanations for disclosing their condition; some adolescents did not see the need to hide it, some disclosed it so that they could receive support if needed and others felt that it is important to raise awareness of SCD:

I tell people just because it makes them aware, and not a lot of people know about it, so it's just like spreading the word (Ashanti, SCD, female, aged 15, group 6).

There was often a trigger to disclosing their condition such as they were hospitalised because of SCD, their friends also have a sibling with SCD or their friends have another health problem meaning they could sympathise or relate to the experience:

They haven't got sickle cell, but some of them have got asthma. I don't know how to explain it. They might not have the same condition but they kind of get it and it made me think, yeah, I'll tell them about sickle cell (Afram, SCD, male, aged 16, group 5).

Moreover, adolescents believed that their peers with health problems were less likely to hassle them with constant questions regarding SCD:

Because once I was with friends and they overheard something and then suddenly they stop you and are like 'Oh, what's this, what's wrong?', 'Are you OK' ((sighs)) then

constant questions about it and telling you to take some water. Gets on my nerves. My good friends wouldn't do that and the ones who get it, because they have ((pauses)) diabetes (Atu, SCD, male, aged 13, group 1).

Adolescents also reported that their school or college but not necessarily their individual subject teachers were aware that they have a medical condition, which their parent most likely disclosed during the school admissions process. Most adolescents explained that their school or college issued them with a medical pass which allows them to be excused during lessons without having to provide the teacher with an explanation, although some adolescents do not like this because they do not want to be given special treatment (some chose not to disclose to friends for this same reason). This special treatment is a reminder of having SCD and may mean they miss out on something as a result of having SCD:

I hate special treatment. I hated in primary school how the teacher would treat me. I hate how when I first went to secondary school I had a teacher with me and they gave me a card to get past the lunch queue and I purposely lost that card because I hated it because you could see everyone having fun in the lunch queue, even though they were pushing each other, it looked like something I wouldn't want to miss. So, it kind of does interfere with your life – ((pauses)) I wouldn't say as much as other things, but it's still like having a little burden on you (Amber, SCD, female, aged 13, group 6).

Unlike adolescents with SCD, most healthy siblings did not disclose their sibling's condition. They only discussed SCD with their immediate family members i.e. parents and siblings and also sometimes close friends. There was no single explanation for

nondisclosure of SCD. The most prevalent explanations were that it is private, it does not affect their friends, that disclosing the condition would not change anything and also concerns about gossip and people breaking their confidence on social media:

I get on with everyone. I don't really have any problems. I have got some very good friends. But gossip is King and with social media everything spreads quickly. You have to be careful what you say and to who you say it. My close friends know about my sister's condition. I can trust them but it doesn't mean I'd tell anyone because you might as well announce it on Instagram then ((agreement)) (Bina, sibling, female, aged 17, group 4).

Amongst female healthy siblings there was a general agreement that it was not necessary for their condition to be disclosed whilst some male healthy siblings also felt as though there needed to be a trigger for disclosure, otherwise it was not necessary:

Only a few family know, like, we wouldn't go round telling everyone because it's not necessary now, if something happened or yeah then maybe, yeah but it's just not necessary now (Bem, sibling, male, aged 13, group 3).

In summary, adolescents with SCD were generally open about their condition, especially when they find the person to be trustworthy or that they can relate to their situation. In contrast, healthy siblings were more likely not to disclose. Healthy siblings may feel that they are respecting their sibling's privacy, as some were aware that people may potentially gossip about SCD, although they, and adolescents with SCD did not suggest that any family member encouraged them to be secretive about the condition.

3.3.2 Theme: Coping with SCD

The second theme “coping with SCD” includes three subthemes: “normalising and acceptance”, “emotion-focused coping” and “self-management of SCD”. This theme discusses how adolescents with SCD and healthy siblings appear to have normalised and accepted the condition. It also describes the different strategies that adolescents with SCD and healthy siblings implemented to cope with SCD, including some emotional support, their religious beliefs and that they have learnt effective strategies to manage SCD.

3.3.2.1 Subtheme: Normalising and acceptance

Both adolescents with SCD and healthy siblings seemed to have accepted that SCD is part of their identity: *“Sickle cell is and always will be a small part of me. It’s normal. Like I have black hair, brown eyes, a mole here, scar there. I have sickle cell. It makes me, me”* (Ada, SCD, female, aged 15, group 6) and also Baakir who explained that: *“My sisters and brother might have sickle cell but that’s not what I see, it’s only a bit of them ((agreement)) They’re normal. Like they don’t let it define or control them”* (Baakir, sibling, male, aged 17, group 3).

Adolescents with SCD and healthy siblings explained that their social lives were generally unaffected on a daily basis by the condition or related pain. They reported that they partake in a variety of social activities, mostly with their friends, such as going to the cinema or shopping, playing sports and attending parties, and that they are generally satisfied with what they do and how much time they spend with their family or friends. This acceptance of SCD was also described by adolescents when coping with specific aspects of the condition such as pain. Male adolescents were generally determined that their condition would not dictate any aspect of their lives. They appeared to have learned to accept and tolerate pain, having lived with it for many years:

Not really ((pauses)) because if I'm in pain, I just get through it. I'm still going out, doing my thing, doing whatever sport I want. I always think like the pain is another person, saying 'Oh, don't let the pain get better of me. I should always be stronger than the pain, just get through it' (Azi, SCD, male, aged 17, group 1).

When I was younger I used to get pain and even if it was a little bit of pain I'd stop, not be able to do anything and whine! A lot! But now ((pauses)) now I'm older, I deal with pain better and I can basically take more extreme pain better ((general agreement)) (Adom, SCD, male, aged 14, group 1).

Of course another possible explanation for these quotes is that male adolescents are conforming to social expectations about masculinity. For example, Adewale (SCD, male, aged 14, group 5) said *"I don't want to show weakness if my friends are around"*. There was evidence of this in most male healthy siblings too, like Bashir (sibling, male, aged 16, group 7) who said: *"I get knackered playing football, basketball because I push myself to my limits. No one's getting one up on me. Fact"*. Also, it may be the case that male adolescents were trying to assert their masculinity while participating in a focus group.

In contrast, female adolescents reported coping with their pain by distracting themselves and engaging in solitary activities such as reading books, watching television programmes or listening to music, but similarly to males, they spoke of 'learning' to deal with their pain:

I've learnt to deal with my pain, in bed, listening to music or watching Netflix in my own little world until it passes. But I do things I enjoy so I don't see it as a negative.

It's a minor obstacle. It works for me ((agreement)). I'm lucky because my pain is passing (Abena, SCD, female, aged 16, group 6).

Referring to SCD as normal and having learned to cope with the pain suggests that adolescents with SCD and healthy siblings have normalised the condition. Furthermore, when they make social comparisons they do not feel dissimilar to their peers, many of whom also have some type of 'problem':

You know I have sickle cell, but some people in my class have diabetes and this one girl is dyslexic and is allergic to nuts. Oh he left in the summer, but a boy in my class has anorexia but it got ((shouts)) that bad, he left, he was a skeleton. My point is that most people have something. It's common (Agnes, SCD, female, aged 13, group 2).

Adolescents with SCD and healthy siblings also made social comparisons to people with more serious or life threatening diseases such as cancer to provide some perspective regarding SCD. Making downward social comparisons is a defensive tendency that helps people to feel better about themselves or their personal situation:

Sickle's just a small part of me and my life. It not ((hesitation)) not like a serious, life threatening disease like cancer. So really what do I have to moan about? People with cancer have a death sentence. I do feel sorry for them (Ashanti, SCD, female, aged 15, group 6).

A family member had cancer, that's really bad ((pauses)) not like sickle cell at all, you can't even compare the two, the stuff they go through, the treatments and then the likely outcome for them: death ((sighs)) (Bobo, sibling, male, aged 17, group 7).

Adolescents' lives are not monopolised by the condition. For example, both groups of adolescents generally did not report any discrepancy between their current and ideal selves in relation to social activities. Also, adolescents seemed to have normalised SCD; especially with increasing age they have learnt to accept it and developed effective strategies to cope with their pain. Furthermore, they drew social comparisons with some of their peers who suffer from health problems and have daily medical treatments and therefore do not feel different to them. Both adolescents with SCD and healthy siblings also drew downward social comparisons to people with more serious conditions such as cancer.

3.3.2.2 Subtheme: Emotion-focused coping

Adolescents with SCD and healthy siblings both described themselves in their day-to-day lives using positive adjectives, for example, "*happy*", "*motivated*" or "*lucky*". Furthermore, both groups of adolescents suggested that they would not change anything in their lives to improve their psychological well-being besides their typical adolescent behaviours (e.g. laziness) which were unrelated to SCD. They also described that they had 'typical' adolescent problems (e.g. friendship troubles). However, they reported limited emotional support with SCD from people in their lives, although most of them (besides male adolescents with SCD) sought solace through their religious beliefs. The limited support was mainly because some of their fathers, siblings or extended family members were no longer living in the family home, lived outside of London or lived abroad. Relying

on only one or two family members for support led to some anxieties about over-burdening them:

I don't have much to do with dad and I don't see a lot of my family ((pauses)) they're in Africa. My mum brought me up, by herself and sorted sickle stuff ((pauses)) that's why I don't like putting anything else on her. She's a typical woman gets emotional easily, so I wouldn't go into sickle with her ((hesitation)) she's stressed enough ((general agreement)) (Adegoke, SCD, male, aged 13, group 1).

Some adolescents with SCD and healthy siblings also reported feeling more comfortable discussing their problems with people who have SCD that they are close to, or who are in their age group because they are more likely to relate to their experiences and emotions. However, this limits the supportive network:

Support from anyone, even my Mum is tricky, she doesn't have it. Who can really understand it? Only people who have it and I don't know ((pauses)) I'm not close to anyone with sickle cell, let alone anyone my age, so I don't have that many options for conversations. But I've learnt to deal with my emotions myself. I don't like negative people (Awa, SCD, female, aged 17, group 2).

Adolescents, especially females, described having complex and, at times difficult relationships with their friends but this was due to typical adolescent grievances rather than SCD. Adolescents did not generally describe receiving emotional support from their friends regarding their condition. For healthy siblings this may not be unexpected because, as previously shown under the “disclosure of SCD” subtheme, they rarely disclosed.

However, this may be more surprising among adolescents with SCD because some of their friends were aware of their condition. It may be that their friends lack the emotional maturity to cope with it. For example, Abigail (SCD, female, aged 14, group 2), suggested that they would “*panic all the time about sickle cell ((agreement)); they’re emotional wrecks ((laughter))*”. Additionally, as discussed above in relation to family, it may again be that adolescents with SCD only disclose emotional problems that they believe their friends can relate to, or they do not wish to burden them with their problems:

I wouldn’t talk to the close ones [friends] about sickle cell, say if I was having pain or something happened, because, everyone has their own shit, some have low money, family arguments ((general agreement)) so I’m going put mine on them? Unnecessary (Afram, SCD, male, aged 16, group 5).

Finally, most female adolescents with SCD and healthy siblings spoke with strong emotion about how their religious beliefs provided them with strength, comfort and hope, which is helpful in coping with SCD, especially during an acute situation such as a vaso-occlusive crisis or hospital admission:

The way I see it, when I was sick, with crisis, I was always like ‘Oh God, help me’. I was praying every day, looking for His comfort, His guidance. I would sleep in church at nights sometimes which was empowering. And then when I got better I was like ‘God helped me through my struggle. I am here. Stronger!’ (Afua, female, aged 15, group 2)

Therefore, both adolescents with SCD and healthy siblings do not appear to have a discrepancy between their day-to-day current and ideal perceptions of their psychological well-being. Both groups of adolescents also reported that they sought emotional support relatively rarely and from a limited number of people but most in both groups found comfort in their religious beliefs, especially when experiencing ill-health, although this was not reported by male adolescents with SCD. Adolescents with SCD preferred to discuss their problems with people that could relate to their problems or shared similar experiences to them, although there were not many people with SCD that they could confide in. They also reported feeling that they did not want to burden their mothers who have raised them alone, or their friends who have their own problems.

3.3.2.3 Subtheme: Self-management of SCD

Most adolescents with SCD experienced less vaso-occlusive crises than when they were younger. One reason for this is the effectiveness of medical interventions, such as blood transfusions and different medication. For example, Akeem (SCD, male, aged 15, group 5) explained: *“I was put on hydroxyurea a few years back and everything changed. I had more energy, crises stopped”*. However, with age, adolescents have also learnt preventative strategies to cope with their condition more successfully, including avoiding triggers, adapting their routine and improving their medication adherence. Thus they are attempting to exert some control over the management of their SCD, and as such, using some problem-focused coping strategies.

A key potential trigger was engaging in outdoor sports. Whilst most adolescents with SCD, especially males, discussed participating in regular exercise, they avoided participating in outdoor sports that have triggered a vaso-occlusive crisis in the past. This

is because the physical nature of some sports and a cold environment can be problematic for SCD:

I used to play rugby and dodgeball when I was younger but then I'd get crisis ((general agreement)). I think ((pauses)) they were rough which isn't good with sickle cell but, but ((pauses)) sometimes when it was cold, that didn't help with sickle cell ((overtalking)) but I just don't do them now (Ayo, SCD, male, aged 14, group 1).

Despite this, adolescents with SCD and also healthy siblings generally reported that they were content with their current participation in exercise. Asir (SCD, male, aged 15, group 5) explained: *"I do and try whatever sport I want ((agreement)). It's not like when you're a kid and you need permission"*. Furthermore, adolescents with SCD have also changed aspects of their daily routines so that they do not overexert themselves and are not exposed to cold weather for prolonged periods:

I use to walk, for about 20 minutes to school in the freezing cold and get sickle pain ((general agreement)) so I decided to stop and catch the bus and then I stopped getting sickle pain. Simple changes really help (Ada, SCD, female, aged 15, group 6).

Another area in which adolescents with SCD report self-management skills is medication adherence. Most adolescents with SCD became more responsible for taking their medication once they reached secondary school by implementing different approaches that helped them remember to take their medication:

I stopped using mine [medicine] for about a year, and then, last week I got this thing where I put my medication, that organises it. And actually, it helps me remember to take it. It's this box where you just put your medication (Adegoke, SCD, male, aged 13, group 1).

I've got an app on my phone to remind me to take my medication which I've been using for about ten months. I downloaded it because I was like, in secondary, so I can't expect my mum to remind me and I sometimes forgot to take it. I was thinking of things I could do so I set an alert but then I found the app ((laughter)) I love my phone! (Anna, SCD, female, aged 13, group 2)

While secondary school age adolescents with SCD are taking responsibility for the management of their condition, younger children still need to be reminded to take medication. Thus, older healthy siblings often reminded their younger sibling with SCD to take their medication, again, often using their smartphones:

My brother's only six so I have an app on my phone so I can tell him when it's time to take his medicine because my mum forgets! (Bacia, sibling, female, aged 14, group 4)

Some female adolescents with SCD reported consuming their favourite food when they experienced pain as it was comforting. However, even though adolescents with SCD are aware of the benefits of drinking water (as discussed earlier under the “awareness and understanding” subtheme), they generally do not report using food or drink to help manage SCD.

The theme “coping with SCD” described that adolescents with SCD and healthy siblings have normalised their condition and generally did not report any discrepancy between their current and ideal selves in relation to exercise, social activities and psychological well-being, which may suggest that they have accepted SCD and, with age have learnt to manage it more successfully. Adolescents have taken an independent role in self-managing their condition, in some cases in part to remove the burden from their mother (see the “emotion-focused coping” subtheme). Adolescents with SCD reported implementing different strategies when coping with pain; male adolescents avoid some exercise or outdoor sports and female adolescents often use a mixture of problem-focused coping (e.g. improving medication adherence) and also emotion-focused coping (e.g. their religious beliefs). It may be expected that adolescents with SCD would also seek emotional support because, as discussed under the “disclosure of SCD” subtheme, most people that are close to them are aware of their condition. However, they have a limited support network and they reported that they do not want to burden their family and friends and feel that many people in their network cannot relate to SCD. Adolescents with SCD seem to have independently positive attitudes towards exercise and medication adherence and they are taking more control of their health behaviours as part of self-managing their condition, suggesting that have increased perceived behavioural control (PBC from the TPB). Furthermore, healthy siblings seemed to find solace through their religious beliefs and some also adopted problem-focused coping strategies, especially to support younger siblings with SCD e.g. reminding them to take their medication.

3.3.3 Theme: Influences on health behaviours

The third theme, “influences on health behaviours” includes two subthemes; “risky behaviours” and “health behaviours”. This theme highlights peer or social influences on

alcohol use. It also suggests different influences between adolescents with SCD and healthy siblings on their participation in exercise and on their diet.

3.3.3.1 Risky behaviours

The most prevalent risky behaviour that adolescents, especially those with SCD have tried, and continue to participate in is alcohol use; sixteen adolescents with SCD compared to six healthy siblings have tried alcohol. However, they did not report drinking excessively nor more frequently than on a weekly basis, which therefore may not be considered 'risky', but is not advisable for SCD. Most male adolescents reported that their alcohol use was influenced by their peers and social situations and many female adolescents reported drinking occasionally with their parents. Some adolescents with SCD and healthy siblings spoke of how they first tried alcohol when attending a special occasion or celebration with their parents. For example, Babatu (sibling, male, aged 14, group 3) explained: *"it was alcohol because we must have been watching a football match; I think it was the Champions League final and dad poured some champagne into my cup"*. Alcohol use appears to be the most acceptable substance because some of their parents also drink:

Drugs and probably smoking, nah, my parents wouldn't allow but with alcohol, yeah, ((pauses)) because they drink. Let's say they were having a glass wine and I said I wanted some, they would say yes because that's not every day (Asir, SCD, male, aged 15, group 5).

Adolescents disclosed that their peers mostly influenced them to drink alcohol when they were socialising, for example, at house parties or in the park. Most adolescents with SCD and healthy siblings had mixed feelings about drinking alcohol, but were afraid

that they may become socially isolated from their friends if they did not join in, so the influence of subjective norms (SN) from the TPB appeared to be important:

I've always had that temptation to try drinking ((general agreement)), like my friends are, 'You should come to the party. There will be drinks there'. Even though I want to go and just talk to my friends, there's always that feeling that they're going to pressure you to try drinks, that's where I first tried cider and southern comfort. So maybe it's best if you just don't go but then you miss out and end up distancing yourself from your friends. It's difficult (Ada, SCD, female, aged 15, group 6).

Adolescents generally did not describe participating in other risky behaviours, besides four older adolescents with SCD who had tried cannabis or shisha and a healthy sibling who smoked tobacco cigarettes. There was no single explanation for engaging in these behaviours besides curiosity or to feel 'different':

Because my best friend did it and told me that it would just make me feel different. So, I just wanted to experience it. But it was my decision. Nothing to do with him or Sickle. I wanted to try it...Weed was ok at the time but it's like the next day....crash....No, I've only done weed two times because it makes my throat hurt. Don't know what the big deal was about it at least when you have shots you have a laugh, I'd recommend that (Akono, SCD, male, aged 15, group 5).

A number of reasons why adolescents with SCD and healthy siblings may be more likely to drink alcohol rather than take drugs emerged. They believed that alcohol is more visible and easier to obtain for their age group:

I'm more likely to take alcohol because it's in shops, school, outside school gates, park, friends' houses, so for example, I or anyone my age could easily access that compared to drugs (Alberta, SCD, female, aged 16, group 2).

Additionally, most adolescents with SCD and healthy siblings spoke of more serious physical and mental health consequences related to drug use and cigarette smoking compared to alcohol use. Therefore, adolescents' negative attitudes towards the consequences of risky behaviours may have influenced their participation:

Drugs and smoking are the worse but alcohol isn't really unless you become an alcoholic, that's the only one I would try but take drugs once and you're dead ((agreement)) smoking too in time and they give you the serious diseases like cancer and mental issues that make you suicidal (Baina, sibling, female, aged 13, group 8).

Moreover, adolescents with SCD generally believed that drug use and cigarette smoking were more likely to trigger a vaso-occlusive crisis than alcohol use. These opinions tended to be based on observations of other adolescents with SCD drinking alcohol with no ill effects:

The effects of these things are bad for everyone but I think, like he said, sickle cell mixed with drugs, smoking will bring on crisis, one-hundred percent ((general agreement)) but taking alcohol wouldn't. Around me people with sickle cell take alcohol regularly and medication. No problem (Afram, SCD, male, aged 16, group 5).

In contrast to reports of peer pressure in relation to alcohol use, for a few adolescents, having SCD may actually facilitate greater self-assurance to resist peer pressure. This was especially the case where adolescents also have negative attitudes towards a risky behaviour such as drug use, and it was also especially reported by female adolescents with SCD. For example, Awa suggested that growing up with SCD provides strength and maturity:

I think most people with sickle cell tend to have a lot of self conf ((hesitation))... not self-confidence, but they don't kneel down to peer pressure. And I feel like we get stronger and stronger every time we grow up, every year (Awa, SCD, female, aged 17, group 2).

Therefore, more adolescents with SCD reported drinking alcohol than healthy siblings but there is no explanation from the focus groups about why this may be the case. Adolescents with SCD and healthy siblings explained that they were most likely to drink alcohol because of peer pressure, so SNs are important, and because it is the most accessible substance for their age group, so factors that make a behaviour easier or more difficult to perform (i.e. PBC) may also be important. However, they do not generally participate in other risky behaviours because they believe that these will have more serious consequences for their general health and SCD, so attitudes are also important.

3.3.3.2 Health behaviours

Adolescents' explained that health behaviours were to varying degrees influenced by their parents, school and the media. For example, parents had an influence on healthy siblings' participation in exercise. One of the adolescents who spoke of this was Bello

whose father encouraged him to be active for his general health and also by funding extracurricular sporting activities but does not treat his sibling who has SCD the same:

Dad pays for football training and basketball lessons. He wants me to keep moving, for my stamina, strength and health but he doesn't tell my brother because he's got sickle cell and dad says his day-to-day focus is that, not sports (Bello, sibling, male, aged 14, group 7).

Adolescents with SCD reported that their parents were less likely to encourage them to exercise, at least in part, because they were concerned about the negative consequences of exercise for SCD. For example, Agnes (SCD, female, aged 13, group 2) reported: *"mum thinks that every time I swim or go gym I'll end up in A&E or have a crisis!"* However, there was some evidence that they were not especially bothered by this lack of encouragement: *"Mum doesn't tell me to do sports or ask about the game but I do them for myself. It's all good ((agreement))"* (Atu, SCD, male, aged 13, group 1). Additionally, as discussed under the 'self-management of SCD' subtheme, adolescents tend to moderate their own exercise due to SCD and generally appear content with their current level of participation in exercise.

Nevertheless, the media has some positive influence on male adolescents' participation in exercise. Most male adolescents, especially those with SCD described watching sports on television and have then tried to imitate these sportsmen and achieve their physiques or sporting abilities:

I watch every Arsenal match to reproduce the tricks, movement and skill of Alexis Sánchez. I do specific exercises that I read he does ((pauses)) to get his physique, but

obviously I'm not at his level, he trains a few hours every day! (Akono, SCD, male, aged 15, group 5)

Several male healthy siblings also reported more negative influences, explaining that they were influenced to purchase unhealthy food because of advertising, food channels and supermarket promotions. However, adolescents with SCD, especially males, generally reported having a healthier diet as part of wanting to improve their physiques, perhaps as a means of showing strength or masculinity:

I eat good food for my sports. Everything I do is to get the physique I want, to be strong, a leader. I do it every day. No excuses. Lean protein for muscles, carbs for energy to keep me going at training and some fresh fruit. Repeat (Abidemi, SCD, male, aged 15, group 1).

This may be influenced by viewing their condition as a form of weakness and therefore their 'improved' physiques show that they are strong or it may be a means to maintain some control over their physical appearance and health when it is hard to control the consequences of SCD. For example, Abdel (SCD, male, aged 17, group 5) explained: *"I can control what my body looks like, my basic health ((pauses)) not crisis, jaundice, ulcers ((agreement)) and still be on par with my friends"*. Indeed, some adolescents with SCD choose to consume a healthier diet because they are aware that poor eating habits and not drinking enough water can have an adverse effect on their physical and mental health. In particular, as suggested by Afua, the discipline and maturity developed through having SCD may facilitate making more healthy choices (this was also discussed under the "risky behaviours" subtheme in relation to saying no to peer pressure to drink alcohol):

You are what you eat. So eat junk food and you'll feel moody, gain weight and get diabetes ((pauses)) people with sickle cell have more self-control because we've had to mature earlier ((general agreement)). For example, my friends don't know what to eat and when to stop and then cry about it! (Afua, SCD, female, aged 15, group 2)

Adolescents with SCD do not report being encouraged to drink water more than healthy siblings. However, both groups of adolescents reported that school had an influence in this respect because it is compulsory for all pupils to only drink water at school, whereas at home there is more variety so sugary drinks may also be consumed:

Same as other people I can only drink water at school but at home there's more choice. I'll have tea for breakfast, juice or lemonade afterschool, hot chocolate before bed, mm just depends what I'm doing and the time of the day (Amber, SCD, female, aged 13, group 6).

In summary, adolescents' risky behaviours, namely their alcohol use, were mostly influenced by their peers or social situations and SCD, as they believed that drug use may cause a vaso-occlusive crisis. Healthy behaviours, for example, exercise and diet were influenced by the media, their parents and to some extent SCD. Thus SNs from the TPB are important but personal attitudes towards behaviour and factors that hinder behaviours (such as SCD hindering exercise), or facilitate behaviour (such as easier access to alcohol), are also important for adolescents (i.e. PBC from the TPB). An example of the influence of attitudes is that some male adolescents with SCD believe exercise will help them develop a good physique to demonstrate their strength and masculinity, so this influences their health

behaviour. However, unexpectedly, in this respect, male adolescents seemed more concerned with their body image than female adolescents. They have found role models in the media such as sportsmen because they receive little encouragement from their parents to engage in exercise, mainly due to parental concerns about the impact this may have on their condition. Finally, some female SCD adolescents believe that they have more strength and maturity for their age because of growing up with their condition, which influences their behavioural choices.

3.3.4 Theme: Education and beyond

The final theme “education and beyond” describes the perceived impact of SCD on adolescents’ education, as well their beliefs about their futures and who encourages them to contemplate their future prospects.

Adolescents’ mothers arranged their hospital appointments or blood transfusions at times or on days that do not interfere with their education:

If I have an appointment, my mum always tells that, if the hospital books me an appointment and it's on a school day, my mum won't allow me to go. She'll say they can rebook it on the weekend, like my blood transfusion on a Saturday, or when I go into the operating room. So my mum makes sure that I don't miss school. Then when I, say, have my blood transfusion I'll bring schoolwork, so it's easy (Alberta, SCD, female, aged 16, group 2).

Whilst discussing their education, adolescents with SCD and healthy siblings also spoke with self-determination about their futures. There were similarities between the types of industries, careers or lifestyles that adolescents with SCD and their healthy

siblings were hoping to achieve and they did not appear to feel that their futures were restricted by SCD:

The thing I desperately want is to have a proper career not just a job, like I said, a theoretical physicist or a games developer. I'm not 100%. That will lead onto a good life, smokin house, holidays, those watches. I'm determined to get there. I have as much chance as anyone else – there's no colour, there's no sickle cell, no negativity (Ayo, SCD, male, aged 14, group 1).

Education comes first right now because I need to get somewhere in life, in terms of my career. I am obsessed. I will be a professional, in finance ((pauses)) and live in a nice house, have a nice car, stability. I'm the oldest so I have to set a good example to my brother and sister but I like that responsibility. We're all capable of success in our future; sickle cell will not hold us back ((general agreement)) (Basma, sibling, female, aged 16, group 8).

This is most likely to be because, as already described in the “coping with SCD” theme, they have normalised their condition and feel that it does not dictate their lives, and with age, they have learnt effective methods of preventing and coping with a vaso-occlusive crisis. Some older adolescents with SCD have even thought as far ahead as to consider their retirement:

I want know what area of work would give me the best wage ((pauses)) pension so I can plan my long-term future and have a retirement house in Jamaica! (Amara, SCD, female, aged 17, group 6)

Adolescents with SCD and healthy siblings were encouraged to consider their futures by different people in their lives. Adolescents with SCD described that various staff in school encouraged them more than their parents. For example, Abigail (SCD, female, aged 14, group 2) revealed *“I think people like the librarian, teachers, careers advisors push me the most, a lot more than my mum ((agreement)) to think about my future”*. Moreover, many adolescents with SCD appreciated the opportunities afforded to them by staff in their school:

It is nice when people go beyond to help you, for example, my English teacher put me forward for a writing competition last month and our librarian gave me information and contacts about becoming a general practitioner just because she saw me looking it up (Amber, SCD, female, aged 13, group 6).

In contrast, healthy siblings described that their family (namely, parents, siblings and cousins) had the greatest influence by encouraging them to revise, find reliable occupations and have aspirations in life. For example, Barika (sibling female, aged 15, group 8) described: *“My parents say I should do something better than what they’ve done; I should go further”*. Most healthy siblings also spoke about having personal tutors and that their parents encouraged them to think of their long-term future:

Parents influence me more than teachers. They always, always tell me to revise, get me the best kit, get me a tutor three times a week and talk about the type of job I should get, you know, good salary, they think about my pension, my long-term future (Bashir, sibling, male, aged 16, group 7).

Although, as discussed earlier, adolescents with SCD consider their long-term future, they did not refer to their parents encouraging them to do so. This may be because their parents were more focused on their current health as opposed to their future:

My mum worries more about my health, right now and doesn't think about in five, ten years you're going to be this. It's not her priority (Abdel, SCD, male, aged 17, group 5).

Therefore, there did not appear to be a discrepancy between the current and ideal self of adolescents with SCD and healthy siblings in relation to their education and futures. Families go to efforts to limit the effect of SCD on adolescents' education and adolescents with SCD and healthy siblings both consider their futures. However, they appeared to be influenced by different people in their lives. In the case of adolescents with SCD their parents may be more focused on their current physical health, whereas this is not an issue for healthy siblings who reported that their parents provided them with tutors and encouraged them to prosper academically and consider their long-term futures.

3.3.5 Summary of findings

The first theme "awareness and disclosure of SCD" described that adolescents were aware of, and had a good understanding of SCD and also that adolescents with SCD generally reported that they were more likely to disclose their condition compared to healthy siblings. Some adolescents with SCD see disclosure as an opportunity to raise awareness of SCD and receive support if needed, whereas healthy siblings fear that

disclosure may lead to gossiping or breaking confidence on social media and they also believe that they are respecting their siblings' privacy.

The second theme "coping with SCD" revealed that adolescents with SCD and healthy siblings have accepted and normalised the condition, often drawing downward social comparisons with people who have more serious physical health conditions which provides some perspective. Neither group of adolescents reported a discrepancy between their current and ideal self in relation to their psychosocial well-being. With age, adolescents have taken an independent role in self-managing SCD (perhaps to alleviate some of their mothers' burden) and have successfully adopted strategies to cope with their condition, especially with pain. For example, male adolescents with SCD reported that they avoid participating in exercise that has triggered a vaso-occlusive crisis and female adolescents with SCD and many healthy siblings reported that improved medication adherence and also their religious beliefs provided comfort. It appears that few adolescents sought emotional support because in the case of most adolescents with SCD their social network is small, there were few people that could relate to their problems and they did not want to burden their mothers or friends.

The third theme "influences on health behaviours" identified differences between the two groups of adolescents' in their behaviours and also the influences on these behaviours. Adolescents with SCD were more likely to drink alcohol than healthy siblings, although there is no explanation for this from the focus groups. Adolescents with SCD and healthy siblings both reported that their peers and also their social environment influenced alcohol use. In contrast, their health behaviours were influenced by different people; adolescents with SCD were sometimes influenced to participate in exercise by the media but not by their parents, because of their condition, unlike healthy siblings who were encouraged by their parents. Neither group reported a discrepancy between their current

and ideal participation in exercise. Male adolescents were more body conscious than female adolescents and they described consuming a healthier diet and participating in more sports.

The final theme “education and beyond” described that adolescents’ education was relatively unaffected by SCD and thus there did not appear to be a discrepancy between their current and ideal self in relation to their education and futures. Adolescents reported different influences on their education and futures; adolescents with SCD were influenced by staff in school, whereas healthy siblings were influenced by their family members, especially their parents. It appeared that parents of adolescents with SCD may be more focused on their current physical health, which is also supported by the fact that they did not want them to participate in exercise, as they are afraid this may trigger a vaso-occlusive crisis.

3.4 Discussion

This study built on Study 1 to explore the HRQL and daily health behaviours, including any differences between current and ideal self, in an older sample of adolescents with SCD compared to healthy siblings. This study also explored their risky behaviours. As in Study 1, this study drew on GT (Calman, 1984) and the TPB (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975). The current study provided a better understanding regarding the lives of adolescents affected by SCD and contributed to knowledge about their health/risky behaviours. There are a number of key findings and contributions. Firstly, there appeared to be no discrepancy or ‘gap’ in adolescents’ current and ideal selves regardless of whether they had SCD or not, which was different from the findings in Study 1, where there was evidence of a gap in some areas like limited participation in exercise or recreational activities and socializing with family or friends. Second, during adolescence they appear to have further normalised and adapted to SCD and taken greater

responsibility for self-managing the condition through, for example, moderating their exercise and reminder strategies to take their medication. Self-managing SCD could be a contributing factor to eliminating the ‘gap’ between adolescents’ current and ideal selves, although it is likely that there were also other reasons that were not explored in the present study. Third, in contrast to Study 1 adolescents with SCD are less secretive about their condition, although they would rather discuss it with someone who has similar experiences, whereas healthy siblings are less likely to disclose SCD. Fourth, again in contrast to Study 1, adolescents with SCD consider their future life aspirations as much as healthy siblings, although the latter are more likely to be encouraged by their parents and the former by staff at school. Fifth, health behaviours were influenced by various individuals or factors. Adolescents’ parents had some influence on their participation in exercise; encouraging healthy siblings but not adolescents with SCD, which in contrast to Study 1, did not appear to bother them. Also, the media (which was not discussed by children in Study 1) encouraged male adolescents to exercise but also influenced poor diet choices in male healthy siblings. Perhaps this difference between male healthy siblings and male adolescents with SCD is because the latter aspire for improved physiques as a result of their condition or maybe their condition has provided them with greater discipline and maturity to make healthier choices. Sixth, unexpectedly, only male adolescents with SCD have body/appearance issues, not both genders as revealed in Study 1. Lastly, the present study also showed that adolescents with SCD consumed or had tried alcohol more than healthy siblings and that this is mainly influenced by peer pressure, ease of access and the erroneous belief that drug use and cigarette smoking may affect SCD, whereas drinking alcohol does not. The analysis of the focus group transcripts is next discussed theme by theme in the context of the wider literature.

3.4.1 Awareness and disclosure of SCD

Adolescents disclosed that they were aware of SCD from a young age which supports the findings of Study 1. Adolescents' discussions also provided insight into the depth of their understanding regarding SCD, including the aetiology, clinical manifestations and helpful health behaviours, which is consistent with past evidence (Forrester et al., 2015; John-Olabode, Awodele, & Oni, 2015). There was no evidence in the present study of gender differences, but previous research found disease knowledge to be greater in female adolescents with SCD (Asnani, Barton-Gooden, Grindley, & Knight-Madden, 2017; Barton-Gooden, Grindley, Knight-Madden, & Asnani, 2019; Bhatt, Reid, Lewis, & Asnani 2011). Those with greater disease knowledge perceived greater personal and treatment control but it was not related to their HRQL (Asnani et al., 2017). Therefore, in the current study adolescents' in-depth understanding of SCD may provide an explanation for their increasingly independent self-management of the condition which, in turn, may have improved some aspects of their HRQL, which will be discussed further in the "coping with SCD" theme.

Unsurprisingly, there appears to have been an advancement in disease knowledge from the transitional period of childhood into adolescence (i.e. shown through children's interviews in Study 1 to adolescents' discussions in Study 2) which is in line with past research (Metcalf, Plumridge, Coad, Shanks, & Gill, 2011). An increased acquisition of disease knowledge in the current study may have been facilitated by the myriad of sources (e.g. school lessons and online) from which adolescents obtain information, compared to when they were younger, which is similar to previous research in the UK (Dyson et al., 2010a). Adolescents tend to use technology to seek health-related information online and to acquire information relating to specific medical conditions (Gray, Klein, Noyce, Sesselberg, & Cantrill, 2005; Skinner, Biscope, Poland, & Goldberg, 2003). In the present

study adolescents also reported that do not obtain knowledge about SCD from parents or medical professionals, despite Kemp et al., (2015) reporting that adolescents with SCD described health professionals in London clinics as friendly and helpful during their outpatient clinic appointments. Perhaps adolescents with SCD would like to avoid awkward or embarrassing conversations with their parents and medical professionals (i.e. males with SCD may suffer from priapism; Knight-Madden et al., 2011). This may also be part of adolescents asserting their independency, as they are approaching adulthood and thus taking greater responsibility for their health, as a recent review concluded (Poku, Caress, & Kirk, 2018). Additionally, as SCD has always been part of their lives they may not feel that it is necessary to discuss it with their families.

In the present study, adolescents with SCD were generally open about their condition but in Study 1 they suggested that their mothers encouraged them to be secretive about it, a likely indicator of adolescents' maturity with increasing age. In this study, there was also no suggestion of gender differences in disclosure of SCD even though past studies have reported that female adolescents were less likely to disclose SCD than male adolescents (Barton-Gooden et al., 2019). In the current study, some adolescents disclosed SCD so that they could receive support if needed. Similarly, Dyson et al., (2010b) showed that some participants disclosed SCD to their peers for emotional, social or academic support, although others did not disclose it because of fear of, or actual bullying, teasing and gossiping. Adolescents' discussions in the present study also suggested that they disclosed SCD to friends who they felt were more likely to understand their predicament and how to treat them. This is also supported by past research (Atoui et al., 2015).

Disclosure of SCD in this study was also related to adolescents raising awareness of the condition. SCD has become a worldwide problem because of migration (Chakravorty & Williams, 2015; Modell & Darlison, 2008). The condition is most prevalent in Africa

but it is increasing in Europe, the United States of America (USA), the Eastern Mediterranean and South-East Asia (Modell & Darlison, 2008). In 2008, the General Assembly of the United Nations introduced an annual World Sickle Cell Awareness Day (June 19) in order to raise public knowledge and awareness of the condition, although the findings of this study would suggest that adolescents feel that some people living in England may not be aware of SCD. In the current study, adolescents with SCD discussed that their schools were aware of their condition, as Dyson et al., (2010b) also concluded. Adolescents explained that their schools had issued them with a medical pass but some chose not to use it as they saw it as receiving 'special treatment'. This is in line with past studies (Panepinto et al., 2012) where some adolescents expressed their annoyance or sadness when they felt as though their teachers were more lenient on them because of SCD (Forrester et al., 2015). They may not want to be singled out in front of their peers or it may be a reminder of their condition.

In the current study, healthy siblings were less likely to disclose and generally only discussed SCD with their immediate family members. Healthy siblings revealed that SCD is not their condition to disclose and that it is private. They also discussed concerns about gossip, similar to issues that were raised by children in Study 1. This is a prevalent issue in research involving healthy siblings of adolescents with a chronic condition (Akobeng et al., 1999; Gundlach et al., 2006; Houtzager et al., 2004; Hutson & Alter, 2007; Plumridge et al., 2011). Healthy siblings also expressed concerns in this study that people may break confidence on social media. A review found that 11% to 43% of adolescents (often girls) had experienced bullying on social media, including girls criticising popularity or appearance and boys receiving homophobic messages or comments about their physical abilities, which in some studies led to depression, self-harm or suicidality (Hamm et al., 2015).

Fears of stigmatisation did not seem to be evident in the present study among adolescents with SCD, although healthy siblings did appear to be more worried about people's reactions, as already discussed. In contrast, Adeyemo et al., (2015) found that 70% of adolescents with SCD have experienced moderate to very high perceived sickle-related stigmatisation, and also it has been reported that greater perceived health-related stigma was related to a lower HRQL (Adeyemo et al., 2015; Martin et al., 2018; Wakefield et al., 2017) and also more pain interference and more loneliness (Martin et al., 2018). These studies used American and Nigerian samples and therefore may not reflect the beliefs of adolescents with SCD living in London, or perhaps adolescents in the present study had milder symptoms of SCD, less pain and felt more supported and therefore had lower levels of health-related stigma. Differences between adolescents with SCD and healthy siblings regarding fear of disclosure may be because adolescents with SCD can focus more on the practical day-to-day tasks of having a chronic condition, for example, taking medication, avoiding cold temperatures and attending hospital appointments. On the other hand, healthy siblings do not have this focus and therefore may overthink how other people may react to SCD.

The issues discussed in this theme have implications for practice. Adolescents with SCD are more open about their condition (than children in Study 1 and healthy siblings in this study) and are also aware of the importance of raising awareness about SCD amongst their peers. This awareness raising is needed among families and in the wider community (a suggestion also made in Study 1). The popularity and possible pitfalls of social media among adolescents affected by SCD should be examined; technology can help to promote understanding of SCD but it may also facilitate bullying or perceived health-related stigma thus undermining adolescents' ability to cope with SCD.

3.4.2 Coping with SCD

Adolescents affected by SCD have many challenges; from physiological, emotional and social changes during puberty to coping with a chronic condition and the implications that this may have for their lives and their HRQL as well as their family's lives. Adolescents' discussions in this study revealed the different coping strategies they use to exert some control over, and help to manage SCD; for example, males avoid triggers which can cause a vaso-occlusive crisis (problem-focused coping). Similarly, Atoui et al., (2015) found that adolescents with SCD also adapted parts of their daily routine or exercise at home. Findings of this study also suggest that adolescents with SCD (and older healthy siblings) have taken greater responsibility for managing their SCD (or helping to manage the SCD of their sibling), for example, by using their smartphones to remind them to take their medication, which is in contrast to the findings in Study 1 where children relied on their mothers for practical support with SCD. Badawy, Thompson and Liem (2016) reported that 85% of adolescents and young adults with SCD owned a smartphone and used apps including a daily medication reminder, education about SCD and adherence text prompts which improve medication adherence in this population (Creary, Gladwin, Byrne, Hildesheim, & Krishnamurti, 2014). Thus there is some use of problem-focused coping strategies.

In the present study, adolescents (except for male adolescents with SCD) also revealed that they drew strength and comfort from their religious beliefs (emotion-focused coping), although in Study 1 this coping strategy was only adopted by healthy siblings. This may be because children with SCD received other forms of support from their mothers, which adolescents do not and therefore they turn to religion, or perhaps children with SCD simply did not mention religion in Study 1. Past evidence found that both male and female adolescents valued their religious beliefs and that they prayed during a vaso-

occlusive crisis which provided them with support (Clayton-Jones, Haglund, Belknap, Schaefer, & Thompson, 2016). Adolescents discussions may have also been influenced by the focus groups which involved their peers who were of the same sex and either all had SCD or were all healthy siblings. For example, male adolescents with SCD did not discuss their religious beliefs as that was the direction that those particular focus groups took, whereas this topic was raised in female adolescents and healthy sibling focus groups because one of the participants mentioned religion.

In this study adolescents did not discuss seeking emotional support for SCD. This may be because the main person in their lives who provides support is their mother (the same as in Study 1). Atoui et al., (2015) found that adolescents with SCD do not want to overburden their mothers who worry about them, care for them and their siblings and are under financial pressure and therefore they may seek other sources of support. In the present study overburdening their mothers was discussed by adolescents. In contrast to Study 1 where a lack of emotional support may not be surprising because they were secretive about the condition, in this study it is more surprising because, as described in the previous theme “awareness and disclosure of SCD”, people in their lives are generally aware of their SCD. Adolescents also talked of over burdening their friends, who have problems of their own, so this may be one explanation. They also discussed feeling that their friends cannot relate to SCD and that they would prefer to speak to someone who has similar experiences. Previous studies support these findings (Valenzuela et al., 2013).

There are other coping strategies that were emphasised by adolescents with SCD. During a painful vaso-occlusive crisis female adolescents preferred to engage in solitary activities and consumed their favourite food but similarly to children in Study 1, adolescents with SCD did not drink water. This is in line with most research (Clayton-Jones et al., 2016; Karlson et al., 2017) but not Maikler et al., (2001) who reported that

hydration was the second most prevalent non drug based coping strategy implemented. It is surprising that adolescents with SCD do not drink water when they are in pain because they have demonstrated an in-depth knowledge of their condition as shown in the “awareness and disclosure of SCD” theme. Drinking water can help to prevent, or cope with a vaso-occlusive crisis (see Chapter 1, section 1.2.2.1).

The issues discussed in this theme have implications for practice. Future studies could identify the most useful adherence and educational apps for SCD populations and there could be an online support group or forum for adolescents to share self-management techniques with their peers and provide the opportunity for interaction with people who understand their experiences. Families affected by SCD could also receive support since adolescents expressed some concerns about their parents. Moreover, guidance on the importance of engaging in healthy behaviours for general health and SCD could be delivered using technology and support services.

3.4.2.1 Normalising and acceptance

Adolescents in this study do not allow sickle-related pain to dominate their lives. This is in contrast to research indicating that adolescents and young adults with SCD felt that the unpredictability or fear of a vaso-occlusive crisis can undermine their daily activities and meant that they were not ‘taking part’ in life (Thomas & Taylor, 2002). In fact, in the present study adolescents with SCD explained that, with age, they experience fewer vaso-occlusive crises, even though past evidence has shown that pain and hospitalisations increase during adolescence (Blinder et al., 2013; Theodore, Barry, Quarmyne, Dampier, & Lane, 2015), perhaps these findings are because of the advancement in effective medical interventions (see Chapter 1, section 1.1.2). Further, findings of this study suggest that adolescents have accepted that SCD is part of their lives

and they have incorporated small changes into their daily routines that take their condition into consideration, thus adapting to SCD. Hijmans et al., (2010) and Panepinto et al., (2012) concluded that with older age adolescents have developed their own or better coping styles. This was a particularly strong message from male adolescents who feel they have learnt to cope with pain. Of course, it may be that male adolescents are conforming to social expectations or asserting their masculinity (especially when participating in a male focus group) as they do not want to show weakness or be different from their peers, or perhaps they have developed a higher pain threshold over the years. In previous research, adolescents with SCD discussed downplaying their pain (Atoui et al., 2015). It may also be that, with age, greater disease knowledge and life experience, adolescents have a more mature attitude to SCD and its severity. Indeed, they talked of developing maturity as a result of SCD under the “influences on health behaviours” theme in relation to making healthy behavioural choices.

Furthermore, adolescents with SCD and healthy siblings made downward social comparisons to people with more life-threatening health conditions or other problems that their peers and family members are coping with. This may help them have some perspective or they may make such comparisons to reassure themselves. Making downward social comparisons in relation to another individual or group is a defensive tendency that helps people to feel better about themselves or their personal situation (Wills, 1991) and may be used especially by people with chronic conditions in response to the threat of illness (Tennen, McKee, & Affleck, 2000). These comparisons are escalated when there is some type of actual threat (Tennen et al., 2000; Wills, 1991), for example, in SCD this may be whilst experiencing a vaso-occlusive crisis or being hospitalised. In adults with SCD, fewer depressive symptoms were related to downward social comparisons whereas greater depressive symptoms were linked to upward social comparisons (Wilson, Gil, &

Raezer, 1997). Thus in the present study these downward comparisons may be considered helpful.

Contrary to the findings of Study 1, it appears that adolescents with SCD and healthy siblings have little discrepancy between their current and ideal self in regards to their psychological well-being and physical or social activities. Previous studies that have adopted a gap theory approach (Calman, 1984) have similarly found little evidence of discrepancy, even among children. Constantinou et al., (2015) found that the discrepancy QoL of children and adolescents with SCA was comparable to a healthy sample. Furthermore, Heath et al., (2011) found that children and adolescents with chronic kidney disease (CKD) had a smaller discrepancy QoL compared to a healthy sample. Therefore, the current study suggests that, especially with age, adolescents with SCD (and healthy siblings) have learnt to cope with the condition and have successfully adapted to it. This is further supported by Albrecht and Devlieger (1999) who postulate that adults with chronic illness or disability are able to adapt socially and emotionally to illness and therefore do not experience a lower QoL. Heidrich and Ward (1992) obtained similar findings among women with cancer. The findings in this study support this interpretation, for example, adolescents with SCD talk of it being normal and just a small part of who they are. Thus they seem to have normalised SCD and accepted it as part of their identity. Whilst children's interviews in Study 1 suggest there is evidence of normalising in childhood, this seems to be more pronounced among the adolescents in Study 2.

3.4.3 Influences on health behaviours

This theme covered engagement in, and influences on adolescents' health behaviours. In the present study, alcohol use was more prevalent among adolescents with SCD compared to healthy siblings. While this was only a small sample, Asnani et al.,

(2014) also substantiated these findings. Adolescents' discussions do not provide an explanation for this difference. It may be because chronic conditions no longer serve as a protective factor against engagement in risky behaviours (Sawyer et al., 2007), or perhaps because adolescents with SCD have had to lead more controlled lives during their childhood they have become more rebellious or curious during puberty. For example, children (in Study 1) described restrictions in their participation in exercise and opportunities to socialise with their friends. These issues have been raised in past studies (Atoui et al., 2015; Forrester et al., 2015; Thomas & Taylor, 2002). It may also be that adolescents with SCD have a greater desire to live life to the full, including having different experiences like trying alcohol. In a retrospective study, adolescents and young adults with SCD explained being aware from an early age that their life expectancy was shortened and that death or dying was part of their reality (Thomas & Taylor, 2002). Group differences in alcohol use may also have been influenced by discussions in the focus groups. Some adolescents may have felt pressure to boast or exaggerate their experience with alcohol in front of their peers once this had been raised and it just happened that this was raised more frequently in focus groups involving adolescents with SCD and not healthy siblings.

Findings of the current study indicate that adolescents' alcohol use was mainly influenced by their peers and social situations, which is not surprising given their age and the greater opportunities they have to independently socialise with their friends at parties. Some adolescents also explained that they drank alcohol so that they would not become socially isolated from their friends. This is in line with a systematic review in healthy adolescents (Leung, Toumbourou, & Hemphill, 2014) and may also be a cause in adolescents with SCD. In contrast, some female adolescents with SCD described being able to resist peer pressure because their condition provides them with more self-

confidence. Forrester et al., (2015) revealed that older adolescents with SCD had a positive self-concept during their adolescence, did not feel different from their peers and have never allowed SCD to define them.

Adolescents' discussions also revealed that alcohol use appeared to be the most acceptable risky behaviour. Previous evidence has concluded that alcohol use was the most prevalent risky behaviour among adolescents with SCD (77.7%); although, in contrast to the findings of this study, adolescents had also tried tobacco cigarette smoking (28.7%) and marijuana smoking (17.4%) (Asnani et al., 2014). In the present study, adolescents explained that some of their parents also consumed alcohol and had allowed them to try their first alcoholic drink on special occasions. Jackson, Henriksen and Dickinson (1999) found that adolescents whose parents allowed them to drink alcohol at home were more likely to continue drinking alcohol two years later.

Adolescents also explained in the present study that alcohol was the most visible and accessible substance for their age group and they perceived it to be the least harmful substance for their health. Furthermore, adolescents with SCD believed that alcohol was less likely to trigger a vaso-occlusive crisis compared to cigarette smoking or drug use which is surprising given their in-depth knowledge of SCD shown in the "awareness and disclosure of SCD" theme. In past research, older adolescents and young adults with SCD understood that it was important to avoid alcohol and tobacco use (John-Olabode et al., 2015). Adolescents' misunderstanding may be because they have fewer opportunities to have open and confidential discussions about risky behaviours with healthcare professionals as they attend outpatient appointments accompanied by their parent (Surís et al., 2008). Kemp et al., (2015) reported that older adolescents with SCD (aged 16 years old and over) were more likely to have the opportunity to speak with a doctor or nurse without a parent present but the sample in this study included adolescents as young as thirteen

years old. These discussions are important because adolescents with chronic conditions that engage in risky behaviours are more likely to have poor disease management and treatment adherence (Scaramuzza et al., 2010).

In the current study, there were multiple influences on adolescents' health behaviours, some of which were the same as in Study 1. According to adolescents' discussions, their parents had some influence on their participation in exercise; healthy siblings were encouraged to exercise whereas adolescents with SCD were not, mostly because this may trigger a vaso-occlusive crisis. However, unlike children with SCD in Study 1, this did not seem to bother them. In contrast, Atoui et al., (2015) reported that these restrictions left adolescents feeling socially isolated from their friends and frustrated. Irrespective of parents' lack of encouragement, adolescents with SCD in this study do appear to participate in exercise, unlike in Study 1, where children wanted to participate in more sports. This is consistent with some previous studies (Omwanghe et al., 2017) whilst other evidence suggests that adolescents with SCA participate in exercise less than their healthy peers (Melo et al., 2018), although the participants in Melo et al.'s study may have had greater disease severity than the present study and therefore were unable to exercise as much. Adolescents are able to make more independent choices and are less reliant on their parents (e.g. taking them to, and from activities). They were also proactive in moderating their own exercise levels, which may have provided more of a sense of control.

The present study also suggested that there was some influence of the media on positive health behaviours like consuming a healthier diet and exercising in male adolescents with SCD. In contrast, a review of healthy eating in healthy children and adolescents concluded that the media, especially television and food advertising promoted more frequent consumption of less healthy foods like higher-fat snacks (Taylor et al.,

2005). This does not appear to be the case in male adolescents with SCD, perhaps because they are more health conscious due to their condition and the health advice they would have received from healthcare professionals over their lifetime. In this study, adolescents with SCD also discussed being more mature for their age, as a result of SCD, and having to be more disciplined (e.g. taking daily medication) which may have led to making healthier behavioural choices. Alternatively they may make healthier choices because they are challenged by issues related to body image (see Chapter 1, section 1.1.2) and thus they are exerting some control over their physiques by exercising and eating healthily. In support of this, evidence has demonstrated that adolescents with SCD have higher levels of negative body satisfaction compared to a healthy sample (Bhatt-Poulose, James, Reid, Harrison, & Asnani, 2016).

The present study found that male adolescents appeared to be more aware of their body image than female adolescents. Erskine (2011) established that male adolescents with SCD were self-conscious about their physiques, general outward appearance and viewed themselves as deficient in comparison to their healthy peers. Older adolescents have a more positive self-concept (Forrester et al., 2015) and therefore male adolescents' body image may improve with age. Traditional gender differences in body image (where healthy female adolescents have greater body image issues than males, e.g. Golan, Hagay, & Tamir, 2014) appear to be different in adolescents with SCD according to the findings of the present study. This may be because delayed growth in children with SCD is more pronounced in males, as females experience a growth spurt with the onset of puberty (Bennett, 2011). Children and male adolescents with SCD have a lower fat mass and also males with SCD have delayed skeletal maturation (Barden, Kawchak, Ohene-Frempong, Stallings, & Zemel, 2002). Perhaps it is more acceptable for female adolescents to be small and slim but male adolescents are expected to be tall and have muscular physiques. Gender

differences in body image found in this study may also be because of the direction of discussion that the male focus groups took. Alternatively, male body image (the rise of television programmes like Love Island and men's health magazines) and health matters are now increasingly prevalent in the media.

Making healthy behavioural choices did not seem to apply to drinking water. In this study both groups of adolescents reported drinking more water at school than at home. This could be a phase during adolescence, as young adults with SCD explained that they drink more water when in pain (Matthie, Hamilton, Wells, & Jenerette, 2016), which was not discussed by adolescents in the present study. It may also be that adolescents simply have more drink options available at home.

Adolescents' engagement in health behaviours may be better understood by exploring the three TPB constructs; attitude towards the behaviour, SN and PBC (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975). The most influential belief in terms of alcohol use was adolescents' SNs; they perceived that there was social pressure from their friends to drink alcohol, which is not surprising for their age group, and felt that it was acceptable because their parents drink too or because they had their first drink with their parents. In addition, adolescents' attitude to alcohol use (for example, it is the least harmful substance for general and SCD-related health) and PBC (most visible and accessible substance for their age group) contributed to adolescents' alcohol use. In past research that has applied the TPB, attitude was the strongest predictor of adolescents and young adults' intention to consume alcohol and alcohol use (Cooke et al., 2016; López-Cisneros et al., 2013). However, in line with the current study, Marcoux and Shope (1997) reported that alcohol use in healthy children and adolescents was influenced more by external (friend's alcohol consumption, peer pressure and availability of alcohol) rather than internal factors (attitude).

The present study also found that adolescents' health behaviours were influenced by their attitude toward the behaviour (for example, exercise and healthy eating leads to an improved physique) and PBC (for example, having to moderate exercise due to SCD). This is consistent with earlier studies that have applied the TPB in examining health behaviours in children and adolescents (Duncan et al., 2012; Keats et al., 2007; McDermott et al., 2015; Plotnikoff et al., 2013; Riebl et al., 2015). In systematic reviews and meta-analyses, healthy eating/dietary patterns and water consumption in healthy children and adolescents were predicted by attitude and PBC, whereas adolescents' SNs was the weakness predictor (McDermott et al., 2015; Riebl et al., 2015). Similar to the findings of this study, past evidence demonstrated that the strongest predictor of exercise is attitude in healthy adolescents (Plotnikoff et al., 2013), British adolescents of lower SES (Duncan et al., 2012) and adolescent cancer survivors (Keats et al., 2007). More recently, Wang and Zhang (2016) showed that self-efficacy and PBC were the strongest predictors of adolescents' intention to participate in moderate to strenuous exercise using an extended version of the TPB.

Once again, there are implications for practice here. Adolescents with SCD and their families would benefit from more information about the effect of alcohol use on SCD. For example, NHS hospitals could provide leaflets about this matter during outpatient clinic appointments or further research could investigate the effectiveness of a smartphone app for relaying information about risky behaviours. There needs to be further research exploring perceptions of body image in adolescents with SCD (especially males) and their motivation to exercise and eat healthily. It may also be important to emphasise the importance of drinking water in families affected by SCD, who may encourage better health behaviours from a younger age in all family members.

3.4.4 Education and beyond

Adolescents felt that their education was unaffected by SCD. In fact, this study indicates that there is less disruption in their education with older age, as some children with SCD in Study 1 described having fragmented school routines and extended school absences. This does not support earlier studies, which have consistently shown that adolescents with SCD have some significant school absenteeism and general disruption in their learning as a result of SCD, for example, because of extended hospitalisations (Atoui et al., 2015; Day & Chismark, 2006; Forrester et al., 2015; Schwartz et al., 2009; Thomas & Taylor, 2002). The findings in the current study may be because adolescents with SCD described having fewer vaso-occlusive crises with increasing age (in the “coping with SCD” theme). Frequent vaso-occlusive crises and a low HRQL were generally not evident among the adolescents with SCD in the present study, perhaps reflecting the lack of disease severity in the sample.

The present study showed that both groups of adolescents are focused on their education and achieving future success. This is reassuring because in Study 1 children with SCD (but not healthy siblings) rarely spoke of their future ambitions. The findings of the present study are in line with past studies that have found that some adolescents with SCD and adults recalled putting in more effort in school to achieve their academic potential because they were driven to succeed (Atoui et al., 2015; Thomas & Taylor, 2002). Adolescents’ commitment to achieving future success also extends to participating in extracurricular activities and planning on attending university (Crosby et al., 2015). In the current study, adolescents with SCD did not appear to feel suppressed by their condition and, unlike in Study 1, there was no evidence of any discrepancy between their current and ideal self. This may be because they have developed better coping strategies with

increasing age and have normalised the condition, so they are not constrained by SCD as already discussed in the “coping with SCD” theme.

Adolescents’ discussions suggested that different people influenced their futures; people at school encouraged adolescents with SCD, whereas family members encouraged healthy siblings to succeed. This is in contrast to past studies, where some adolescents with SCD discussed receiving little support from teachers at times and more generally the educational system (Atoui et al., 2015; Crosby et al., 2015; Thomas & Taylor, 2002). In Studies 1 and 2 healthy siblings were more likely to have private tutors, perhaps because family members are more focused on the current physical health of adolescents with SCD, as suggested in their discussions. The findings of Studies 1 and 2 also indicate that as children have grown older they have found encouragement and role models in different people and that, most importantly, in the long-term their ambitiousness has not been undermined by SCD. It is important that adolescents with SCD are given the same opportunities as healthy siblings because Thomas and Taylor (2002) showed that adolescents and young adults with SCD found it difficult to find or remain in employment because of their condition.

Adolescents with SCD appear to be supported in school and, with increasing age have found role models who encourage them to persevere with their education and aspirations in life. It may be advantageous for family members to be included in setting these goals during adolescents’ schooling so that they can provide continued support when they are seeking employment which may be more difficult for people with SCD.

3.5 Conclusions

This study provided some insight into adolescents’ health behaviours. Adolescents with SCD generally report having similar, if not better, health behaviours compared to

healthy siblings, as they have a healthy diet, and participate in exercise. However, neither group reported drinking much water at home, or to help cope with sickle-related pain, and there were more reports of alcohol use among adolescents with SCD than healthy siblings. Alcohol use is mainly influenced by SNs. Health behaviours appear to be influenced by adolescents' attitude and PBC, but in Study 1 SNs and parents' beliefs appeared to have more influence on health behaviours, which is not surprising at the children's younger age.

The findings from this study also revealed that adolescents with SCD and healthy siblings do not appear to have a discrepancy between their current and ideal selves in their HRQL, as suggested by GT, which is in contrast to the findings of Study 1 where there was evidence of some discrepancies. This may be because SCD has become more normalised and accepted with age (i.e. in Study 1 there were indications of children normalising SCD which became more pronounced in adolescents' discussions in Study 2). Living with the condition for longer, developing more effective coping strategies and greater maturity with age may all contribute to reducing any discrepancy between adolescents' current and ideal self in the current study. It may be the case that people are driven to reduce discrepancy and there are a number of examples of this in psychology. For example, Rogers (1959) argued that the 'self' includes all experiences available at a given moment, both conscious and unconscious, and that the closer a person's self-image and ideal-self are to each other the more congruent or consistent they are, and the higher the sense of self-worth. According to Rogers (1959) a person strives for congruence (i.e. bringing their self-image and ideal-image closer to each other). Another example is cognitive dissonance theory (Festinger, 1957). According to Festinger (1957), people tend to seek a consistency among their cognitions (i.e. their beliefs and opinions and behaviour) and avoid disharmony (i.e. they seek cognitive consistency). Cognitive dissonance occurs when there are conflicting attitudes, beliefs or behaviours, producing a feeling of mental discomfort and an imbalance

which a person restores by changing their attitudes, beliefs or behaviours to reduce the dissonance (Festinger, 1957).

If it is the case that ultimately humans are driven to reduce discrepancy and hence there is little evidence of HRQL discrepancy for adolescents and adults, then this may question whether a gap approach is a useful way to consider HRQL. Indeed, the small amount of previous quantitative research that has taken a gap approach (i.e. Calman's (1984) Gap Theory; GT) to examine HRQL has not found much evidence of a gap, not just in adolescents and adults but also in children with chronic conditions including SCA and chronic kidney disease (CKD) (Constantinou et al., 2015; Heath et al., 2011). Furthermore, the vast majority of research in paediatric HRQL in SCD does not take a gap approach but focuses on current perceived HRQL (e.g. Bonner et al., 2010; Dale et al., 2011; Panepinto & Bonner, 2012; Panepinto et al., 2005; 2009) Consequently, the remaining research in this thesis will not be taking a gap theoretical approach to HRQL and will focus on current perceived HRQL.

The next two studies in the thesis will focus on the HRQL and health behaviours (participation in exercise and water consumption) in both children and adolescents with SCD and also adolescents' risky behaviours (alcohol use, smoking and drug use). Diet will not be examined further because studies 1 and 2 suggested it had little impact on SCD. Data from a large, cross-sectional quantitative survey will be presented. Study 3 examines some of the determinants of HRQL. Study 3 will also draw together HRQL and health behaviours to determine whether there is a relationship between them i.e. whether increased water consumption and more or less exercise and alcohol consumption predict HRQL. Study 4 examines the predictors of health behaviours (participation in exercise and water consumption) in children and adolescents, and the predictors of adolescents' alcohol use. The previous qualitative studies drew on the TPB (Ajzen, 1988; Fishbein & Ajzen,

1975) and study 4 will also be adopting the three constructs of the TPB. Studies 3 and 4 will obtain child, adolescent and parent reports, as the qualitative studies in this thesis suggest that parents have some influence over children's/adolescents' health behaviours and they are able to provide an insight into their child's HRQL.

Chapter 4: Examining and Predicting HRQL in Children and Adolescents with SCD (Study 3)

4.1 Introduction

The findings of Study 2 suggested that the application of Gap theory (GT) in paediatric sickle cell disease (SCD) may not meaningfully contribute to understanding the health-related quality of life (HRQL) of these children/adolescents and so instead, the present quantitative study examined current perceived HRQL, like the majority of previous research in SCD (e.g. Bonner et al., 2010; Dale et al., 2011; Panepinto & Bonner, 2012; Panepinto et al., 2005; 2009). The most prevalent measure used to ascertain HRQL in this previous research is the Pediatric Quality of Life Inventory (PedsQLTM; as discussed in Chapter 1, section 1.1.4.1) which is available as child and adolescent self-report and a parent proxy measure (Panepinto & Bonner, 2012). There is also a SCD specific measure (Panepinto et al., 2012) but as Studies 1 and 2 suggested that children/adolescents with SCD see it as only a small part of their identity and so consider themselves the same as other people of their age, the generic PedsQLTM seems to be more appropriate and was used in the current study.

Using this measure, parents often reported that their child had a lower HRQL than their child self-reports (Bonner et al., 2010) and lower parent proxy reports of HRQL have been associated with more severe symptoms of SCD (Panepinto et al., 2010). Additionally, there are reports of reduced current perceived HRQL in paediatric SCD compared to healthy children/adolescents (Bonner et al., 2010; Dale, et al., 2011; Panepinto et al., 2009), so it is important to understand the determinants of HRQL. A recent systematic review found that socio-demographic variables (older age and female gender), low socio-economic status (SES; measured by neighbourhood or parental education) and more disease severity (i.e. vaso-occlusive crises or days missed from school) predicted lower

self-reported HRQL in children and adolescents with SCD (Ojelabi et al., 2017). In addition, Constantinou et al., (2015) reported that parents' single status predicted lower parent-proxy reported QoL in children/adolescents with sickle cell anaemia (SCA). The current study also investigated these determinants of HRQL.

Additionally, whether children's/adolescents' coping styles to deal with sickle-related problems (like pain or tiredness) were also predictors of HRQL was examined. This is because a theme (i.e. "coping with SCD") in Studies 1 and 2 suggested that problem-focused coping, emotion-focused coping and, to a much lesser extent, seeking social support were potentially important.

Finally, whether healthy behaviours, and also risky behaviours in adolescents, were predictors of HRQL was examined. In the previous qualitative studies, health behaviours appeared to be used as a coping strategy, for example, male adolescents with SCD moderated their exercise to avoid a vaso-occlusive crisis. Indeed, health behaviours such as drinking water and participating in less strenuous exercise may help to alleviate some physical symptoms of SCD (e.g. Brown, 2012; Knight-Madden et al., 2011), maintain general health and possibly improve HRQL, and importantly these can be managed at home by children, adolescents and their parents. Conversely, participating in strenuous or unsuitable exercise (Machado et al., 2007; Moheeb et al., 2007) and risky behaviours (Beyer et al., 1999; Brown, 2012; Okomo & Meremikwu, 2012) may lead to vaso-occlusive crises and may undermine HRQL. However, healthy/risky behaviours and HRQL have rarely been brought together in previous paediatric SCD research except by Wrotniak et al., (2014) who examined the impact of a nutritional supplementation intervention on proxy parent reported HRQL in children with SCA.

The current study investigated two research aims. The first aim was to explore if there was a relationship between child self-reports, adolescent self-reports and parent

proxy-reports of HRQL. The second was to determine the predictors (e.g. demographic indicators, disease severity measures, coping, health/risky behaviours) of child/adolescent self-reports and parent proxy reports of HRQL.

4.2 Method

4.2.1 Design

The current cross-sectional study adopted an inter-dependent groups, correlational design using questionnaires. Children and adolescents with SCD as well as their parent completed the same primary outcomes questionnaire (PedsQL™) and a health behaviour questionnaire and adolescents also completed a risky behaviour measure. The primary outcome measures were the HRQL scores generated from the PedsQL™. The predictors were demographic indicators, disease severity measures, as well as coping and healthy/risky behaviours.

4.2.2 Participants

The convenience sample included four groups of participants, comprising two pairs of inter-dependent groups: children with SCD and their parent and adolescents with SCD and their parent. Children and adolescents were recruited from three London hospitals. Child and adolescent sample characteristics are fully described in Table 4.2.

The study included a convenience sample of 106 children with SCD: 54 (51%) were male, ranging in age from 5 to 12 years old ($M = 8.75$, $SD = 2.31$) and the majority, 83 (78%), of children had the most prevalent and severest phenotype HbSS (i.e. SCA).

There were 106 parents or carers of children with SCD: 20 (19%) were male, ranging in age from 27 to 54 years old ($M = 40.39$, $SD = 6.19$). Parent marital status was fairly equally distributed; 51 (48%) were “married” (i.e. married or cohabiting) and 55

(52%) were “single” (i.e. single, separated, divorced or widowed). Parents had varying educational attainment levels; 38 (36%) had higher degree or degree, 23 (22%) had foundation degree or diploma and 45 (42%) had A Levels or below.

The study also included a convenience sample of 96 adolescents with SCD with equally distributed genders, whose ages ranged from 13 to 18 years old ($M = 15.29$, $SD = 1.61$) and 77 (80%) had phenotype HbSS.

Finally, the study included the 96 parents of the adolescents with SCD. The majority, 80 (83%), were female, whose ages ranged from 37 to 68 years old ($M = 48.05$, $SD = 6.24$). Parent marital status was fairly equally distributed, with 46 (48%) “married” and 50 (52%) “single”, although their highest educational attainment level was unequally distributed; 39 (41%) had higher degree or degree, 23 (24%) had foundation degree or diploma and 34 (35%) had A Levels or below.

4.2.3 Materials

Each participating child and adolescent with SCD and parent received an information sheet, assent or consent form, the PedsQL™, a health behaviour questionnaire and a debriefing form. There were three sets of age-appropriate information sheets (Appendix D), health behaviour questionnaires (Appendix E) and debriefing forms (Appendix D); one for children aged 5 to 12 years old, one for adolescents aged 13 to 18 years old and one for parents and also four sets of assent or consent forms (Appendix D); one for children aged 5 to 12 years old, one for adolescents aged 13 to 15 years old, one for adolescents aged 16 to 18 years old and one for parents or carers. There were also two sets of age-appropriate PedsQL™ for children and their parent; a version for 5 to 7 year olds and a version for 8 to 12 year olds and also an age-appropriate PedsQL™ for adolescents aged 13 to 18 years old and their parent (Appendix E). In addition, adolescents with SCD

received a risky behaviour questionnaire (Appendix E) and children and adolescents with SCD also received a modified version of The Kids Coping Scale (KCS) (Appendix E). The parent also received a demographic information sheet (Appendix D).

4.2.3.1 Pediatric Quality of Life Inventory (PedsQL™)

The PedsQL™ is a validated generic multidimensional HRQL questionnaire which is available in parallel child self-report for children aged 5 to 18 years and parent-proxy formats for children aged 2 to 18 years (Varni et al., 2005b; Varni et al., 2003). It consists of 23 items assessing four dimensions of HRQL; physical functioning (eight items), emotional functioning (five items), social functioning (five items) and school functioning (five items). The PedsQL™ can also generate a HRQL total score (i.e. including all 23 items), a physical health summary score (i.e. only including items in the physical dimension), and a psychosocial health summary score (i.e. including items in the emotional, social and school dimensions). The questionnaire asks children and parents to think about how much of a problem the child has had with these areas in the past month. For example, in the physical functioning domain, examples of questions are as follows; ‘it is hard for me to lift something heavy’ and ‘it is hard for me to take a bath or shower by myself’; in the emotional functioning domain, ‘I feel afraid or scared’ and ‘I have trouble sleeping’; in the social functioning domain; ‘is it hard for you to get along with other kids’ and ‘can other kids do things that you cannot do’ and in the school functioning domain; ‘it is hard to pay attention in class’ and ‘I miss school because of not feeling well’. There is a 3-point Likert scale for children aged 5 to 7 years (e.g. 0 is ‘not at all’, 2 is ‘sometimes’ and 4 is ‘a lot’) and there is a 5-point Likert scale for all other child, adolescent and parent versions of the PedsQL™ (e.g. 0 is ‘never’, 1 is ‘almost never’, 2 is ‘sometimes’, 3 is ‘often’ and 4 is ‘almost always’).

The HRQL scores were calculated by transforming responses on a scale which ranged from 0 to 100, where 0 was 100, 1 was 75, 2 was 50, 3 was 25 and 4 was 0, and computing a mean score, i.e. for the HRQL total score, all of the items were summed and divided by 23 (Varni et al., 2005b; Varni et al., 2003). HRQL scores were from 0 to 100, with a higher score indicating a higher HRQL (Varni et al., 2005b; Varni et al., 2003).

In the present study, the analyses included the total HRQL score and also physical and psychosocial health summary scores for each group of participants; children, adolescents and their parents respectively. These subscales have been used in previous studies e.g. Engelke, Guttu, Warren and Swanson (2008); Jackson et al., (2014) Palermo et al., (2008b); Panepinto et al., (2009).

The self-reported and proxy-reported total HRQL scores (23 items; $\alpha = .90$ to $.93$), physical health summary scores (8 items; $\alpha = .81$ to $.89$) and psychosocial health summary scores (15 items; $\alpha = .86$ to $.90$) demonstrated good reliability ($\alpha = .70$; DeVellis, 2003) as shown in Table 4.1.

4.2.3.2 The Health Behaviour Questionnaire

The questionnaire measured two health behaviours: exercise and water consumption. Children and adolescents provided their own opinion regarding frequency of these behaviours and parents or carers also provided their own opinion about frequency of their child's health behaviours. This questionnaire also measured health beliefs developed based on the TPB and the interviews and focus groups in Studies 1 and 2. However, these measures were not used in the current study and will be discussed in the next chapter.

4.2.3.2.1 Exercise behaviour

This was measured based on the Godin-Leisure-Time Exercise Questionnaire (Godin, 2011; Godin & Shephard, 1985; 1997) which ascertained how many times in an average week children engaged in strenuous, moderate and mild types of exercise for at least 15 minutes. Examples of strenuous, moderate and mild exercise were provided. A total score was calculated comprising the number of times a week reported for each of strenuous, moderate and mild exercise multiplied by nine, five and three respectively, which are the metabolic equivalents for each intensity category (i.e. a measure of exercise intensity based on oxygen consumption (Godin, 2011; Godin & Shephard, 1985; 1997)) and then summed:

$$\text{Total weekly leisure activity score} = (9 \times \text{strenuous}) + (5 \times \text{moderate}) + (3 \times \text{mild})$$

In addition to the total score, scores for strenuous exercise (nine multiplied by times of exercise per week) and moderate exercise (five multiplied by times of exercise per week) were also included separately. This is because children and adolescents with SCD should try not to engage in strenuous exercise as this may exacerbate their condition but should engage in moderate exercise for their general health. Mild exercise does not have the same health benefits (such as improving cardiorespiratory fitness, bone health and cardiovascular biomarkers; World Health Organization, 2019) so was not included separately but was part of the total exercise score.

4.2.3.2.2 Water consumption

This measure was developed because there was no pre-existing measure to ascertain weekly water consumption. This was measured by asking three questions about

the number of glasses of water consumed in an average day: 1) at school, 2) afterschool and 3) at the weekend, and responses were on a 7-point Likert scale from zero (0 glasses of water) to six (6 or more glasses of water). A weekly water consumption score was then calculated by multiplying number of glasses consumed at school by five, afterschool by five and at the weekend by two and then summing:

$$\text{Total weekly water consumption} = (\text{school day} \times 5) + (\text{afterschool} \times 5) + (\text{weekend} \times 2)$$

4.2.3.3 *The Risky Behaviour Questionnaire*

This questionnaire focused on alcohol use because few adolescents with SCD discussed engaging in drug use or smoking tobacco cigarettes in Study 2, although the prevalence of drug use and smoking was still ascertained in the present study. The questionnaire also measured beliefs about alcohol use developed based on the TPB and the interviews and focus groups in Studies 1 and 2. However, these measures were not used in the current study and will be discussed in the next chapter.

The questionnaire was for adolescents (aged 13 to 18) only and was not completed by their parents because past research has found that approximately two thirds of parents are unaware of adolescents' alcohol use (Bogenschneider, Wu, Raffaelli, & Tsay, 1998; Williams, McDermitt, Bertrand, & Davis, 2003) and only 6% to 26% of parents are aware of adolescents' excessive alcohol use (Berge, Sundell, Öjehagen, Höglund, & Håkansson, 2015). Furthermore, Study 2 found that some adolescents drink alcohol with their friends or whilst attending parties, which their parents may not be fully aware of, as also found in previous literature e.g. Borawski, Ievers-Landis, Lovegreen and Trapl (2003).

There were five questions in total, three of which ascertained how *often* adolescents engaged in alcohol use, and with one question on each on substance use (like cannabis and

shisha) and cigarette smoking. ‘How often do you drink alcohol?’, ‘I take a substance like cannabis, shisha etc. (not including tobacco cigarettes)’ and ‘how often do you smoke tobacco cigarettes?’ had a 7-point Likert scale including ‘never’, ‘a few times a year’, ‘once or twice a month’, ‘once a week’, ‘2-3 times a week’, ‘4-5 times a week’ and ‘almost every day’ (scored from 0 to 6). There was a question which asked adolescents how many tobacco cigarettes they would smoke in an average day. There was also a question which asked adolescents the number of alcohol units that they consumed in an average week. Adolescents were provided with images depicting the number of units in different beverages e.g. two units in a can of lager, beer or cider. For the purposes of analysis, the number of units were turned into a dichotomous variable: ‘never drink’ vs. ‘drink alcohol’.

4.2.3.4 *The Kids Coping Scale (KCS)*

The KCS is a brief self-report nine-item generic measure which assesses three coping styles in children and adolescents; problem-focused coping, emotion-focused coping and seeking social support (Maybery, Steer, Reupert, & Goodyear, 2009). While the KCS has low (emotion focused-coping) to moderate reliability (problem-focused coping) in healthy children (Maybery et al., 2009), it overcomes criticisms of many other measures, such as the Coping Strategies Questionnaire (CSQ; Rosenstiel & Keefe, 1983). For example, the KCS does not represent too many different types of coping strategies, it uses more than one item to represent each coping style and is not lengthy, which is a key problem with the CSQ (Blount et al., 2008; Compas et al., 2001; Sveinbjornsdottir & Thorsteinsson, 2008).

In the present study, children and adolescents with SCD completed a revised version of the KCS which was piloted using three children ($M = 8.48$, $SD = 2.75$), and three adolescents with SCD ($M = 15.32$, $SD = 2.08$) where they were asked ‘when your

sickle cell disease is bad, like when you're in pain, have a crisis or are tired, what do you normally do?' The modified version of the KCS consisted of eight items which assessed a child's problem-focused coping (three items; e.g. 'you tried your best to make things better'), emotion-focused coping (three items; e.g. 'you did not want to think about it') and seeking social support (two items; e.g. 'you asked someone to help'). However, one emotion-focused item ('you did things to stop thinking about it') was removed from all analyses to improve reliability. The KCS is scored on a 3-point Likert scale ('never', 'sometimes' and 'a lot'), where a higher score is greater use of that coping style.

In the current study, child and adolescent self-reported problem-focused coping (three items; $\alpha = .65$ to $.74$), emotion-focused coping (two items; $\alpha = .52$ to $.56$) and seeking social support (two items; $\alpha = .42$ to $.66$) demonstrated reasonable to good reliability with the exception of seeking social support on child report ($\alpha = .70$; DeVellis, 2003) as shown in Table 4.1.

Table 4.1

Reliabilities Across the PedsQLTM and The Kids Coping Scale for Children with SCD (n=106), Adolescents with SCD (n=96) and their Parents

	Children α	Parent α	Adolescents α	Parent α
<u>PedsQLTM</u>				
HRQL total score	.90	.93	.93	.93
Physical health summary score	.81	.89	.89	.89
Psychosocial health summary score	.86	.88	.90	.90
<u>The kids coping scale</u>				
Problem-focused coping	.65		.74	
Emotion-focused coping	.56		.52	
Seeking social support	.42		.66	

4.2.3.5 Demographic and disease severity information sheet

The demographic information sheet included basic information regarding the parent and their child e.g. gender, age, phenotype, child's religion and ethnic group and parents marital status, occupation, highest educational attainment level and questions that assessed the child's disease severity e.g. how many crises and how many days the child has missed from school in the last twelve months.

Those found to be related to HRQL or health behaviours in previous research were included for analysis (e.g. Darmon & Drewnowski, 2008; Dampier et al., 2010; Jackson et al., 2014; Kawchak et al., 2007; Panepinto et al., 2005; Sallis et al., 2000) and will be described. Continuous demographic indicators included the child's or adolescent's age. Discrete demographic indicators included the child's or adolescent's gender, the parent's or carer's marital status (married/cohabiting or single) and highest educational attainment (higher degree or degree, foundation degree or diploma and A Levels or below). Disease severity questions enquired about the total number of crises their child had encountered in the last twelve months. Parents or carers were also asked how many times their child or adolescent received different types of treatments for their crises e.g. 'treated at home', 'GP', 'medication required', 'A&E attendance', 'hospital attendance' or 'other'. The final measure asked how many days the child had missed from school due to illness related to SCD in the last twelve months.

4.2.4 Procedure and ethical considerations

The study was registered as a research project in line with NHS Trust policy (Research Ethics Committee reference number 17/LO/0322; see Appendix F). In addition, it was registered with the Research and Development departments of the three participating hospitals as well as with the University Psychology Department's Ethics Committee,

which adheres to the British Psychological Society (BPS) code of ethics (The British Psychological Society, 2009). Approval was granted by all of the ethics committees.

All children and adolescents with SCD were identified by the local collaborator at the respective London hospitals, their medical teams and the researcher. Children and adolescents with SCD and their parent were posted age-appropriate information sheets to their home address one week before data collection was due to commence which provided them with ample time to carefully consider their participation in the study.

Following this, the researcher approached potential participants who were accompanied by their parent at their routine outpatient clinical appointment. However, children and adolescents with SCD who were currently experiencing symptoms such as a vaso-occlusive crisis and had any other comorbidities that were not related to SCD were not invited to participate because these factors may have affected their HRQL. At a convenient time to participants and hospital staff, the researcher read through information sheets with parents or carers (if they asked) and also children and adolescents before they gave consent. This provided an opportunity for participants to discuss any questions or concerns that they had regarding the study with the researcher before they agreed to take part. In addition, participants were informed that all of the data collected would remain confidential, that each participant was allocated a reference number and therefore was not identifiable and that all data were stored on password protected computers and were only accessed by individuals directly involved in the study. Hard copies of data e.g. consent forms were stored in a secured cupboard in one of the participating London hospitals throughout the study, and demographic information sheets and questionnaires were stored in a secured cupboard at Middlesex University during data collection (only accessible to the researcher) and at one of the participating London hospitals in the long-term (only accessible by the paediatric sickle cell team). It was also made clear to all participants that

they could withdraw from the study at any time up to data analysis with no explanation required and with no impact on the care they received from the NHS staff.

Participants who agreed to participate were then provided with an assent or consent form to complete; the parent was asked to provide written informed consent for their own participation and for their child's participation (if they were aged 5 to 15 years old) and the child also completed a very simple written assent form. Older adolescents aged 16 to 18 years old provided written informed consent for their own participation.

The parent was then asked to complete a demographic information sheet. Children, adolescents and their parent were asked to independently complete the PedsQL™ and the health behaviour questionnaire and adolescents were also asked to complete the risky behaviour questionnaire. Additionally, children and adolescents were asked to complete the KCS. The PedsQL™, health behaviour questionnaire, risky behaviour questionnaire and KCS were distributed in a different order to participants to control for order effects. For example, if a child, adolescent and parent were first given the PedsQL™ questionnaire to complete, then the health behaviour questionnaire and then the KCS, then the following participant (child, adolescent and parent) would be first given the health behaviour questionnaire to complete, then the KCS and then the PedsQL™.

Children and adolescents (with the researcher's assistance) and also parents or carers completed the questionnaires in separate private clinic rooms near the waiting room of the outpatient clinic away from one another (so that their responses were not influenced by each other or hospital staff). The researcher made parents aware of where their child would be and also that they were able to ask for the researchers' assistance, if needed. Due to the focus of the questions it was possible that children or adolescents may have discussed some of their worries or concerns with the researcher. If a child became distressed, anxious or upset, then members of the Sickle Cell and Thalassaemia team who

work at each hospital offered immediate support and also support at a later date, if requested.

Once all participants had completed the questionnaires they were verbally debriefed. Furthermore, participants were also provided with age-appropriate debriefing forms which discussed the study in layman's terms and provided them with the contact details of the researcher, academic supervisors and support services, if needed. The debriefing form stated that participants could contact the researcher (whose details were clearly on the form) to provide an email or postal address to receive the anonymous results of the study after a prolonged period of time.

4.3 Results

4.3.1 Introduction

This section examines cross-informant agreement regarding HRQL and predictors of HRQL. First, descriptive statistics relating to all variables are presented. The variables include; sample characteristics, HRQL (total, psychosocial and physical), health behaviours (total, strenuous and moderate exercise and weekly glasses of water consumption) and risky behaviours (alcohol use, tobacco cigarette smoking and substance/drug use).

4.3.2 Descriptive statistics

4.3.2.1 Sample Characteristics

Sample characteristics in Table 4.2 include child and adolescent demographic indicators and disease severity measures.

Table 4.2

Descriptive Statistics for Socio-Demographic Characteristics of Children (n=106) and Adolescents (n=96)

	Children M(SD) or % n	Adolescents M(SD) or % n
Age	8.75(2.31)	15.29(1.61)
Gender	51% Male	50% Male
Ethnicity	75% Black African, 21% Black Caribbean, 3% Black African and Black Caribbean, 1% Black African and Asian, 1% Black African and Greek- Cypriot	76% Black African, 21% Black Caribbean, 3% Black African and Black Caribbean
Religion	84% Christianity, 8% Islam, 4% Atheist, 3% Catholic, 2% Jehovah Witness	88% Christianity, 8% Islam, 2% Jehovah Witness, 1% Catholic, 1% Atheist
SCD phenotype	78% HbSS, 22% HbSC	80% HbSS, 20% HbSC
Crises	1.96(2.37), range 0 to 12	1.95(2.23), range 0 to 14
Total number of crises	31% no crises 25% 1crisis 15% 2 crises 8% 3 crises 14% 4 crises 1% 5, 6, 7, 8,10 and 12 crises	28% no crises 24% 1crisis 22% 2 crises 6% 3 crises 9% 4 crises 3% 5 crises 5% 6 crises 1% 9 and 14 crises
Treatment for crises	31% Not applicable 21% Treated at home and medication 14% Hospital attendance and medication 12% Treated at home and hospital attendance 8% Other treatment combinations 7% Treated at home 7% Treated at home and medication and A&E attendance	30% Treated at home and medication 27% Not applicable 17% Other treatment combinations 12% Hospital attendance and medication 8% Treated at home and hospital attendance 6% Treated at home and A&E attendance and hospital attendance and medication
Missed days from school	7.75(9.93), range 0 to 45	12.17(22.80), range 0 to 200

Table 4.2 shows that most children (78%) and adolescents (80%) had the most severe form of SCD (i.e. SCA), that both groups had on average two crises a year but that adolescents had missed on average more days of school than children (12 vs. 8 respectively).

The demographic indicators relating to parents of children and adolescents are presented in Table 4.3.

Table 4.3

Descriptive Statistics for Socio-Demographic Characteristics of Parents of Children (n=106) and Parents of Adolescents (n=96)

	Parents of Children M(SD) or % n	Parents of Adolescents M(SD) or % n
Age	40.39(6.19)	48.05(6.24)
Gender	19% Male	17% Male
Marital status	48% Married/cohabiting, 52% Single/separated/divorced /widowed	48% Married/cohabiting, 52% Single/separated /divorced
Employment status	63% Employed, 7% In education, 30% Unemployed	84% Employed, 16% Unemployed/unable to work/retired
Highest educational attainment	36% Higher degree or degree 22% Foundation degree or diploma 43% A Levels or below	43% Higher degree or degree, 24% Foundation degree or diploma 33% A Levels or below
SCD phenotype	7% HbSS, 5% HbSC, 84% Sickle cell trait, 5% HbAA	2% HbSS, 93% Sickle cell trait, 5% HbAA

The table highlights that the majority of parents were female with mostly sickle cell trait and that half of them were single. Around two fifths of parents had a degree and there was a higher employment rate among parents of adolescents than children (84% vs. 63% respectively).

4.3.2.2 HRQL

Child/adolescent self-reported and parent proxy-reported descriptive statistics for HRQL (total, physical and psychosocial) are shown in Table 4.4.

Table 4.4

Mean HRQL Scores Reported on the PedsQLTM by Children (n = 106), Adolescents (n=96) and their Parents

	Child self-report	Parent proxy	Adolescent self-report	Parent proxy
	M(SD)	M(SD)	M(SD)	M(SD)
<u>HRQL</u>				
HRQL total score	73.16(16.40)	71.08(17.16)	73.05(16.99)	71.68(17.16)
Physical health summary score	72.49(19.48)	68.96(20.62)	68.20(21.42)	67.71(20.81)
Psychosocial health summary score	73.52(16.74)	72.22(16.70)	75.64(16.51)	73.80(16.92)

4.3.2.3 Health behaviours

Child/adolescent self-reported and parent reported descriptive statistics for health behaviours (total, strenuous and moderate exercise and weekly glasses of water consumption) can be found in Table 4.5.

Table 4.5

Descriptive Statistics for Health Behaviours of Children (n=106) and Adolescents (n=96) and Parents' Reports of their Child's Health Behaviours

	Child self-report	Parent report	Adolescent self-report	Parent report
	M(SD)	M(SD)	M(SD)	M(SD)
<u>Health Behaviour</u>				
Weekly exercise	134.37(58.89)	136.10(62.83)	108.22(68.48)	102.61(69.03)
Strenuous exercise	68.86(46.64)	68.77(51.04)	50.44(48.06)	47.63(48.53)
Moderate exercise	33.16(18.26)	34.95(17.61)	27.50(19.64)	27.08(20.15)
Water consumption	31.95(13.62)	34.55(12.88)	43.07(16.18)	42.22(15.99)

4.3.2.4 Risky behaviours

Adolescent self-reported risky behaviours in Table 4.6 included alcohol use, tobacco cigarette smoking and substance/drug use. As found in Study 2, the prevalence of drug use and smoking was very low.

Table 4.6

Descriptive Statistics for Risky Behaviours of Adolescents (n=96)

<u>Participants</u>	<u>Adolescents</u> M(SD) or % n
Units of alcohol in a week	1.65(0.48)
Frequency of alcohol use (a)	35% Never drink, 65% Drink alcohol
Frequency of alcohol use (b)	35% Never, 28% A few times a year, 23% Once or twice a month, 9% Once a week, 4% 2 to 3 times a week
Tobacco cigarette smoking	97% Never, 2% A few times a year, 1% Once or twice a month
Substance/drug use	93% Never, 3% A few times a year, 3% Once or twice a month, 1% Once a week

Further analyses will not include tobacco and substance/drug use because these behaviours were not prevalent amongst adolescents with SCD, so alcohol consumption will be the only risky behaviour examined.

4.3.3 Research Aim One: To explore if there is a relationship between child self-reports, adolescent self-reports and parent proxy-reports of HRQL.

To address aim one, cross-informant agreement between child and parent and adolescent and parent reports of HRQL were first examined and then differences between child and adolescent reports were examined. Paired-samples t-tests were conducted to

compare child and adolescent self-reported total HRQL, physical HRQL and psychosocial HRQL with parent proxy-reports on the PedsQL™ as shown in Table 4.7.

Table 4.7

Mean HRQL Scores for Different Dimensions of the PedsQL™ for Children (n = 106), Adolescents (n=96) and their Parents

	Child self-report	Parent proxy	<i>T</i>	Adolescent self-report	Parent proxy	<i>t</i>
	M(SD)	M(SD)		M(SD)	M(SD)	
HRQL						
HRQL total score	73.16(16.40)	71.08(17.16)	2.23*	73.05(16.99)	71.68(17.16)	2.31*
Physical health summary score	72.49(19.48)	68.96(20.62)	2.76**	68.20(21.42)	67.71(20.81)	.53
Psycho-social health summary score	73.52(16.74)	72.22(16.70)	1.24	75.64(16.51)	73.80(16.92)	2.85**

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

The table shows that children reported higher total HRQL and physical HRQL and also adolescents reported higher total HRQL and psychosocial HRQL compared to parents. Although there were some significant mean differences, Pearson's correlations for the relationships between child-parent and adolescent-parent reports for total HRQL, physical HRQL and psychosocial HRQL showed strong significant positive correlations ranging from $r = 0.64$ to 0.94 . Therefore, higher child and adolescent reported HRQL were associated with higher parent proxy-reported HRQL.

Independent-samples t-tests comparing child self-reported and adolescent self-reported HRQL on the different dimensions of the PedsQL™, showed no significant differences as shown in Table 4.8.

Table 4.8

Mean HRQL Scores for Different Dimensions of the PedsQL™ for Children (n = 106) and Adolescents (n=96)

	Child self-report	Adolescent self-report	<i>t</i>
	M(SD)	M(SD)	
<u>HRQL</u>			
HRQL total score	73.16(16.40)	73.05(16.99)	.05
Physical health summary score	72.49(19.48)	68.20(21.42)	1.49
Psychosocial health summary score	73.52(16.74)	75.64(16.51)	-.91

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

In summary, both child and adolescent and parent reports of HRQL were significantly related but there were mean differences; parents underestimated children's total and physical HRQL and parents underestimated adolescents' total and psychosocial HRQL. However, there were no differences between child and adolescent reports of HRQL.

4.3.4 Research Aim Two: To determine the predictors (e.g. demographic indicators, coping, disease severity measures, health/risky behaviours) of child self-reports, adolescent self-reports and parent proxy reports of HRQL.

To address aim two preliminary univariate analyses were first performed to examine relationships between HRQL (the outcome variable) and the predictors; health and risky behaviours, coping, demographics and disease severity measures. Then multiple regression analyses were implemented to predict HRQL from these variables. First analyses were conducted to ascertain there were no noteworthy violations of the assumptions of normality, linearity, multicollinearity and homoscedasticity which was carried-out for the analyses in relation to research aim two.

4.3.4.1 Preliminary analyses

These analyses were initially executed to help identify statistically significant predictors of HRQL to be used in multiple regression analyses. As these are preliminary analyses and for the sake of brevity, only statistically significant findings are reported in relation to coping, demographic indicators and disease severity measures. However, findings are fully reported in Appendix G.

First, Pearson's correlation analyses were used to explore the relationship between children's health behaviours (weekly total, strenuous and moderate exercise and also weekly water consumption) and HRQL (total, physical and psychosocial HRQL) as reported by children, adolescents and parents. There were no significant relationships between water consumption and HRQL (ranging from $r = -0.01$ to -0.12) and no relationship between adolescent reports of psychosocial HRQL and moderate exercise ($r = 0.20$). All other correlations between HRQL and exercise were significant and positive ranging from $r = 0.23$ to 0.51 . Therefore, overall higher reports of HRQL (total, physical

and psychosocial) were associated with greater levels of child, adolescent and parent-reports of the child's/adolescent's participation in weekly total, strenuous and moderate exercise.

Independent-samples t-tests were also performed to explore alcohol consumption and HRQL (total, physical and psychosocial HRQL). As shown in Table 4.9 there were no significant differences in HRQL between adolescents who drink alcohol and those who do not drink alcohol.

Table 4.9

Mean HRQL Scores for Different Dimensions of the PedsQLTM and Alcohol Use in Adolescents (n=96)

	Adolescent self-reported alcohol use		<i>t</i>
	Drink M(SD)	Never drink M(SD)	
<u>HRQL</u>			
HRQL total score	74.81(15.23)	69.85(19.63)	-1.37
Physical health summary score	69.96(19.71)	64.98(24.22)	-1.09
Psychosocial health summary score	77.39(15.06)	72.45(18.68)	-1.41

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Second, the relationships between child and adolescent self-reported coping styles (problem-focused coping, emotion-focused coping and seeking social support) on the Kids Coping Scale and self- and parent proxy-reported HRQL were also assessed using Pearson's correlation analyses. No correlations were statistically significant.

Finally, the relationships between demographic indicators (child's gender and age and also parent's marital status and highest educational attainment level), disease severity

measures (number of crises and days missed from school in the last twelve months) and HRQL using child, adolescent and parent-proxy reports were examined. Independent-samples t-tests found that there were no statistically significant differences in HRQL between male and female children or adolescents. Also, one-way ANOVAs found no statistically significant differences in HRQL when comparing parents with different levels of educational attainment. However, independent-samples t-tests showed there was one statistically significant difference on the psychosocial dimension of HRQL between married and single parents. Adolescents who had single parents ($M = 72.23$, $SD = 18.31$) reported lower psychosocial HRQL compared to adolescents whose parents were married ($M = 79.35$, $SD = 13.55$; $t(94) = -2.15$, $p = .034$).

Pearson's correlation analyses were used to explore the relationship between child or adolescent age and HRQL. There were two significant negative correlations (significant at $p < .05$) between age and parent proxy-reported total HRQL ($r = -.20$) and age and parent proxy-reported psychosocial HRQL ($r = -.21$) for children, with parents of older children reporting lower HRQL respectively. Pearson's correlations to examine the relationships between disease severity measures (i.e. the number of crises and the number of days missed from school in the last twelve months) and HRQL showed significant negative correlations (significant at $p < .001$) ranging from $r = -0.32$ to -0.59 , with higher HRQL on all dimensions being associated with fewer crises or missed days from school.

4.3.4.2 Multiple regression analyses

Hierarchical multiple regression analyses were used to assess the ability of demographic indicators, disease severity measures and health behaviours to predict HRQL. There were 3 separate regression analyses conducted for each HRQL outcome (i.e. total HRQL, physical HRQL and psychosocial HRQL) and for each of child self-report,

adolescent self-report and parent proxy-reports (separately for children and adolescents), so there were twelve regression analyses in total. Based on the significant findings from the preliminary analyses, the regression analyses for child, adolescent and parent reports included two demographic indicators entered on the first step (child's age and parent's marital status), two disease severity measures entered on the second step (crises and days missed from school in the last twelve months) and two health behaviour measures entered on the third step (strenuous and moderate exercise). In hierarchical regression analyses it is acceptable to have five cases for every independent variable or predictor (Tabachnick & Fidell, 1989, pp. 128-129). The multiple regressions predicting child and parent reports of HRQL are shown in Tables 4.10 and 4.11. R^2 square change is reported for each step but the betas are reported only for the final step.

Table 4.10

Summary of Hierarchical Multiple Regression Analysis predicting Child Self-Reported HRQL on the PedsQL™ (n = 106)

	HRQL total score			Physical health summary score			Psychosocial health summary score		
	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change
STEP 1		.023	1.21		.035	1.87		.012	.65
Child's age	.02			-.00			.02		
Parents marital status	.04			.05			.02		
STEP 2		.264	18.74***		.290	21.73***		.195	12.43***
Crises	-.05			-.14			.01		
Days missed from school	-.40**			-.33**			-.40*		
STEP 3		.046	3.44*		.067	5.43***		.027	1.74
Child reported strenuous exercise	.08			.12			.04		
Child reported moderate exercise	.21*			.23**			.16		

Note. Betas only in the final model are reported.

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

F change degrees of freedom are (2,103) at step 1, (2,101) at step 2 and (2,99) at step 3.

Table 4.11

Summary of Hierarchical Multiple Regression Analysis predicting Parent Proxy-Reported Child HRQL on the PedsQL™ (n = 106)

	HRQL total score			Physical health summary score			Psychosocial health summary score		
	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change
STEP 1		.039	2.11		.022	1.16		.046	2.49
Child's age	-.07			-.02			-.09		
Parents marital status	-.00			-.01			.00		
STEP 2		.241	16.87***		.237	16.18***		.208	14.08***
Crises	-.11			-.19			-.05		
Days missed from school	-.30*			-.23			-.32*		
STEP 3		.082	6.36**		.069	5.05**		.079	5.86**
Parent reported strenuous exercise	.10			.13			.07		
Parent reported moderate exercise	.26**			.22*			.27**		

Note. Betas only in the final model are reported.

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

F change degrees of freedom are (2,103) at step 1, (2,101) at step 2 and (2,99) at step 3.

The final models predicting total HRQL and physical HRQL reported by children explained 33% ($F(6,99) = 8.27, p < .001$) and 39% ($F(6,99) = 10.64, p < .001$) of the variance respectively and fewer days missed from school followed by increased moderate exercise were the statistically significant predictors as shown in Table 4.10. R^2 change at steps 2 (disease severity) and 3 (exercise) were statistically significant. The final model predicting psychosocial HRQL explained 23% ($F(6,99) = 5.05, p < .001$) of the variance and only fewer days missed from school was a statistically significant predictor. Only R^2 change at step two (disease severity) was statistically significant.

The final models predicting parent proxy-reported total HRQL and psychosocial HRQL for children explained 36% ($F(6,99) = 9.36, p < .001$) and 33% ($F(6,99) = 8.24, p < .001$) of the variance respectively as indicated in Table 4.11. The statistically significant predictors in these final models were fewer days missed from school followed by increased moderate exercise. The final model predicting physical HRQL explained 33% ($F(6,99) = 8.05, p < .001$) of the variance and the only statistically significant predictor was increased moderate exercise. R^2 change at steps 2 (disease severity) and 3 (exercise) was statistically significant in all three models.

The multiple regressions predicting adolescent and parent reports of HRQL are shown in Tables 4.12 and 4.13.

Table 4.12

Summary of Hierarchical Multiple Regression Analysis for Adolescent Self-Reported HRQL on the PedsQL™ (n = 96)

	HRQL total score			Physical health summary score			Psychosocial health summary score		
	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change
STEP 1		.070	3.49*		.030	1.45		.089	4.52*
Child's age	-.08			-.05			-.10		
Parents marital status	.07			.00			.11		
STEP 2		.324	24.28***		.244	15.32***		.311	23.58***
Crises	-.23*			-.24*			-.20*		
Days missed from school	-.40***			-.30**			-.43***		
STEP 3		.026	2.03		.033	2.15		.019	1.42
Adolescent reported strenuous exercise	.18			.18			.15		
Adolescent reported moderate exercise	-.02			.03			-.05		

Note. Betas only in the final model are reported.

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

F change degrees of freedom are (2,93) at step 1, (2,91) at step 2 and (2,89) at step 3.

Table 4.13

Summary of Hierarchical Multiple Regression Analysis for Parent Proxy-Reported Adolescent HRQL on the PedsQL™ (n = 96)

	HRQL total score			Physical health summary score			Psychosocial health summary score		
	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change
STEP 1		.058	2.89		.037	1.80		.063	3.14*
Child's age	-.05			.00			-.09		
Parents marital status	.06			.05			.06		
STEP 2		.299	21.18***		.252	16.15***		.275	18.88***
Crises	-.22*			-.24*			-.18		
Days missed from school	-.34***			-.24*			-.37***		
STEP 3		.077	6.06**		.113	8.41***		.045	3.25*
Parent reported strenuous exercise	.27**			.31**			.22*		
Parent reported moderate exercise	.05			.09			.01		

Note. Betas only in the final model are reported.

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

F change degrees of freedom are (2,93) at step 1, (2,91) at step 2 and (2,89) at step 3.

The final models predicting total HRQL, physical HRQL and psychosocial HRQL reported by adolescents explained 42% ($F(6,89) = 10.74, p < .001$), 31% ($F(6,89) = 6.60, p < .001$) and 42% ($F(6,89) = 10.67, p < .001$) of the variance respectively as presented in Table 4.12. The statistically significant predictors were days missed from school followed by number of crises in all three final models. R^2 change at steps one (demographics) and two (disease severity) were statistically significant for total HRQL and psychosocial HRQL whereas R^2 change was only statistically significant at step two for physical HRQL.

The final models predicting parent proxy-reported total HRQL and physical HRQL for adolescents explained 44% ($F(6,89) = 11.40, p < .001$) and 40% ($F(6,89) = 9.99, p < .001$) of the variance respectively as shown in Table 4.13. The statistically significant predictors were number of crises, days missed from school and strenuous exercise and R^2 change was statistically significant at steps two (disease severity) and three (exercise). The final model predicting psychosocial HRQL explained 38% ($F(6,89) = 9.21, p < .001$) of the variance as demonstrated in Table 4.13. Days missed from school and strenuous exercise were statistically significant predictors and R^2 change was statistically significant at all three steps.

In summary, fewer days missed from school and increased moderate exercise were key predictors of HRQL reported by children and their parents. However, for adolescents, fewer days missed from school and fewer crises were the key predictors and for their parents, increased strenuous exercise was an additional predictor.

4.4 Discussion

Study 3 examined the predictors of HRQL in children and adolescents with SCD. These findings were in line with past studies as well as providing some new insights, especially regarding the relationship between exercise and HRQL. In children with SCD, fewer days missed from school and increased moderate exercise predicted child and parent reported HRQL whereas in adolescents with SCD, fewer days missed from school and fewer crises predicted adolescent and parent reported HRQL as well as increased strenuous exercise also predicting parent reported HRQL. This study also explored the relationship between child/adolescent self-reported HRQL and parent-proxy reported HRQL. The findings of Study 3 therefore supported previous research; parents underestimate children's total and physical HRQL and also adolescents' total and psychosocial HRQL. However, there were no differences between children and adolescents in self-reports of HRQL.

4.4.1 Research Aim One: To explore if there is a relationship between child self-reports, adolescent self-reports and parent proxy-reports of HRQL.

The current study illuminated that the relationship between child/adolescent self-reported HRQL and parent proxy-reported HRQL differed depending on the domain of HRQL (i.e. total, physical and psychosocial) being assessed. Parents underestimated their child's/adolescents' total HRQL. This is in line with previous research which has consistently reported that parents of children/adolescents with SCD report lower total HRQL (e.g. Bonner et al., 2010; Dale et al., 2011; Panepinto et al., 2005; Panepinto et al., 2010). Poor HRQL agreement between children/adolescents and parents has also been found in other chronic conditions such as cerebral palsy (White-Koning et al., 2007) and congenital heart disease (Goldbeck & Melches, 2005) so perhaps parents of a child with any chronic condition may have difficulty gauging their child's HRQL. The fact that the

majority of proxy informants in the present study were the mothers may also have influenced the findings. Few studies in paediatric HRQL research report whether the proxy informant is the mother or father, or have examined the differences between these proxy informants (Upton et al., 2008). It may be that mothers provide lower HRQL reports than fathers which may provide an explanation for the lower proxy HRQL reports in the current study, as most informants were mothers.

The present study also found differences between parent proxy reports for children and adolescents; parents underestimated children's physical HRQL and adolescents psychosocial HRQL. Again, these findings are generally supported by past research (e.g. Bonner et al., 2010; Dale et al., 2011; Panepinto et al., 2005; Panepinto et al., 2010). The severity of SCD may provide an explanation for lower child physical HRQL reported by parents in this study. Panepinto et al., (2010) demonstrated that greater disease severity categorised by a history of stroke, acute chest syndrome, three or more hospitalisations in the last three years or recurrent priapism led to worse parental physical HRQL reports but not child self-reported physical HRQL on the PedsQLTM. The present sample did include a large proportion of children with severe SCD, for example, three quarters of children had suffered from at least one crisis in the past year, many of whom were hospitalised, so perhaps parents seeing their child suffering and in debilitating physical pain means that they imagine their physical HRQL is worse than it is, or at least, worse than their child's own perception. Moreover, Eiser and Varni (2013) argue that parent proxy reports may be affected by their daily involvement in their child's hospital appointments and treatments as well as all of the time they spend caring for them. Once again, witnessing their child's daily struggles and SCD related symptoms may provide an explanation for lower parent proxy reported HRQL found in this study. However, it may also be that children with SCD have more realistic expectations of their physical capabilities (Panepinto et al., 2012) whereas

their parents may compare them to their healthy peers who they believe have a higher HRQL (Dale et al., 2011).

In contrast, there was agreement in adolescent-parent physical HRQL, despite adolescents facing many additional challenges than children because of their condition, such as, delayed puberty, priapism (i.e. persistent and painful erection of the penis) and leg ulcers (Knight-Madden et al., 2011; Serjeant & Serjeant, 2001). Discordance in child-parent HRQL reports may be related to the child's age. A review found that the largest discrepancies in physical HRQL were in younger children (aged 5 to 7 years) and the largest discrepancies in psychosocial HRQL were in adolescents (aged 13 to 17 years) (Eiser & Varni, 2013). Therefore, adolescent-parent agreement in physical HRQL observed in this study is not unexpected and is supported by previous literature. Maybe adolescents are less vocal and more independent in coping with their SCD related pain and the physical restrictions of the condition than children. Also, from a parent's perspective, these additional symptoms of SCD that manifest during adolescence may have more effect on adolescents' psychosocial rather than physical HRQL.

Indeed, this study showed that parents provided lower psychosocial HRQL reports for adolescents but not children. This may be because adolescents tend to suffer from emotional and behavioural changes during puberty (Kaltiala-Heino, Marttunen, Rantanen, & Rimpelä, 2003) and may struggle to disclose their emotions during this period to their parents, which is a typical issue for their age group rather than specifically related to SCD. This is supported by Upton et al., (2008) who showed that there was discordance in adolescent-parent psychosocial reports in healthy and chronically ill adolescents, again demonstrating that perhaps their age has more impact than SCD on psychosocial HRQL. Thus, these age related changes and difficulties disclosing their feelings, may make it more

challenging for parents to accurately gauge adolescents' psychosocial state, and perhaps provide an explanation for lower reports because parents imagine the worst.

Some researchers also postulate that it is easier to assess observable domains of HRQL e.g. physical functioning, bodily pain, and school functioning rather than emotional well-being (Upton et al., 2008; Panepinto et al., 2005). This may be especially apt in adolescents who are more likely to be out of the family home and have many social environments that are part of their everyday lives in which their parents do not observe them (Eiser & Varni, 2013). Consequently, parents of adolescents may be less informed about the nuances of their emotional functioning and social relationships which may account for lower parent proxy reports (Eiser & Varni, 2013). It may also be that adolescents with SCD do not want to overburden their mothers with their emotional problems as they have their own personal, family and financial worries to cope with (Atoui et al., 2015) which they may recognise more than children. Therefore, adolescents may not discuss their emotional problems or worries and again, this may mean parents imagine the worst.

As discussed there were differences found between some child/adolescent-parent reports of HRQL, although the differences in scores were relatively small (except perhaps for child and parent reports of physical HRQL). A smaller effect size may indicate that the discrepancy in child/adolescent-parent HRQL reports is relatively unimportant and that there is a stronger relationship between these reports (Field, 2013). In other words, there is little disagreement in reports of HRQL between children/adolescents and their parents. Thus the findings should be interpreted with caution. However, studies in HRQL have reported similar findings (e.g. Panepinto et al., 2010; Upton et al., 2008).

The findings of this study showed that there was no difference between child and adolescent self-reported HRQL suggesting that children as young as five years old and

adolescents as old as 18 years old are no worse off than each other. Therefore, there may not be such a difficult transition from childhood to adolescence in those with SCD. The advancement in medical interventions and treatments such as immunization programs in England (NHS Choices, 2019) and medication to prevent symptoms of SCD (Battersby et al., 2010; Brandow et al., 2010) as well as the support provided to patients with SCD by hospital staff in London (Kemp et al., 2015) may have eased the transition into adolescence, or perhaps has meant that children and adolescents have received consistent care leading to a stable HRQL. Current literature in paediatric SCD has rarely examined HRQL overtime besides Brandow et al's., (2010) study which was only over a seven-day period, suggesting longitudinal research is needed.

In summary, there is a longstanding disparity between child/adolescent self-reports and parent proxy reports of HRQL which needs to be addressed as parents make fundamental health decisions which affect their child's medical treatment and their future opportunities. There is also a distinct shift from childhood to adolescence in this respect; parents underestimate children's physical HRQL but then underestimate adolescents' psychosocial HRQL. Improving communication between children/adolescents and their parents may allow parents to gain a better insight into their child's HRQL, and the areas that need addressing. Children's HRQL appears to remain stable into adolescence which is reassuring, although longitudinal research is needed. Contributing factors may include regular hospital care, treatments and monitoring, so longitudinal research also needs to explore this.

4.4.2 Research Aim Two: To determine the predictors (e.g. demographic indicators, coping, disease severity measures, health behaviours) of child self-reports, adolescent self-reports and parent proxy reports of HRQL.

The findings of the present study demonstrated that disease severity measures were the key predictors across child/adolescent self-reported and parent proxy reported total, physical and psychosocial HRQL; days missed from school (except for parent-proxy reported child physical HRQL) and number of crises for adolescent and parent-proxy reports (except for parent-proxy reported adolescent psychosocial HRQL). In addition, increased moderate exercise predicted most domains of HRQL in children by self-report and parent proxy-report and increased strenuous exercise predicted parent proxy-reports of adolescent HRQL. The variance in HRQL explained ranged from 23% to 44% with slightly more explained in adolescent and parent proxy reports (31% to 44%) compared to child and associated parent proxy reports (23% to 39%).

This study showed that disease severity impacts the HRQL of children and adolescents with SCD. Fewer days missed from school predicted child/adolescent self-reported and parent proxy reported HRQL. This is supported by previous research where fewer days missed from school led to higher HRQL reports (i.e. Constantinou et al., 2015; Dampier et al., 2010). Furthermore, past evidence has consistently shown that lower reports of HRQL from children/adolescents with SCD and their parents are related to frequent hospital admissions (Dale et al., 2011; Dampier et al., 2010; Palermo et al., 2002) which would mean that they are likely to be absent from school. Frequent short and long-term school absenteeism in children/adolescents with SCD (Schwartz et al., 2009) may lead to poorer academic attainment and lower intellect compared to their peers (Berkelhammer et al., 2007). School absenteeism may also affect their social relationships and psychological well-being. It has been documented that children and adolescents with

SCD may suffer from increased anxiety, social withdrawal, poor relationships and social interaction because of their condition (Anie, 2005; Edwards et al., 2005; Pradhan & Nayak, 2014). Therefore, it is not surprising that in the current study, more days missed from school was associated with a lower HRQL.

In the current study, fewer crises predicted higher adolescent self-reported HRQL on all domains of HRQL and on the majority of parent proxy reported HRQL domains but this disease severity measure did not affect HRQL reports in children and their parents. Past research has found that more vaso-occlusive crises led to lower HRQL reports in both children and adolescents with SCD (i.e. Amr et al., 2011; Brandow et al., 2010; Ozer al., 2014; Panepinto et al., 2005; Schlenz et al., 2012). The findings of this study may therefore reflect a difference in disease severity in the sample; parents reported that five or more crises in the past year were experienced by a greater number of adolescents compared to children (9% vs. 1%). This is consistent with previous studies that have demonstrated that the frequency of vaso-occlusive crises increases during adolescence (Theodore et al., 2015). Therefore, the greater frequency of vaso-occlusive crises in adolescents may provide an explanation for why this predicted reduced HRQL in this group but not in children.

While disease severity explained 20-32% of the variance in HRQL, exercise added 5-11%. In the present study, increased moderate exercise predicted higher HRQL in most domains of child self-reported and parent proxy reported HRQL (with the exception of psychosocial HRQL reported by children) but it did not affect any informant reports of adolescents' HRQL. Earlier studies have not examined the association between exercise and HRQL in children/adolescents with SCD, but in adults with SCD regular exercise tended to improve some domains of HRQL including general health, social function and vitality and reduced pain (Ahmed et al., 2015), which may provide some support for the

fact that moderate exercise predicted increased HRQL in children in the present study. This is also supported by NHS medical advice, where engagement in regular moderate levels of exercise is an important factor in maintaining good physical and mental health and also for improved HRQL in children and adolescents with SCD (NHS Choices, 2019).

In the current study, exercise of any intensity did not predict HRQL in adolescents but increased strenuous exercise predicted parent proxy reports of adolescents' HRQL. Exercise participation may not be such a pivotal issue for adolescents who may be more preoccupied with other matters such as socialising with their friends, preparing for GCSEs or A Levels and their futures (Atoui et al., 2015; Crosby et al., 2015). Therefore, participating in exercise may have little impact on adolescents' HRQL, whereas missing school or having recurrent vaso-occlusive crises (i.e. greater disease severity) may have more of a negative effect on areas of their lives that are important to them. On the other hand, parents may believe that participation in strenuous exercise is important to adolescents. This misconception may be because of poor adolescent-parent communication or perhaps parents assume that adolescents will rebel against them i.e. past research has found that parents deter exercise participation because of the potential ill effects that it may have on SCD (Forrester et al., 2015; Thomas & Taylor, 2002).

It is important that SCD should not undermine participation in exercise. This has been found in past studies where children's physical activities are limited to prevent a vaso-occlusive crisis (Panepinto et al., 2012) or they participate in significantly less sports at school and in their leisure time compared to their healthy peers (Melo et al., 2018). Participating in moderate exercise is good for their general health and SCD but they need to be more careful with strenuous exercise which induces metabolic and physiological changes that may have health consequences in SCD (Beyer et al., 1999; Jama et al., 2002;

Platt, 1982), such as triggering a vaso-occlusive crisis (Dyson et al., 2010a; Omwanghe et al. 2017).

In summary, this study provided some new insights regarding the predictors of HRQL, which include disease severity and exercise. However, preliminary analyses suggested that HRQL reports were not related to other health behaviours such as water consumption or alcohol use or to demographic indicators such as gender. It should be noted that while disease severity was a predictor of a lower HRQL, the measures of days missed from school and number of crises were self- or parent-proxy reports, so were not objective reports of disease severity. Nevertheless, parents, healthcare professionals and schools (who have a long-term daily involvement in their lives) should work together when a child/adolescent has a vaso-occlusive crisis or is absent from school (because of SCD) to reduce any adverse effect on the child's HRQL. Exercise was also an important predictor of HRQL, especially in children. Education about exercise is important because this can improve general health and HRQL in children (as found in this study) who may feel they are missing out and experience a reduced HRQL if their parents are too protective when it comes to exercise. However, only moderate exercise was found to be important for children's HRQL, so although parents may feel that strenuous exercise is important for adolescents' HRQL, adolescents do not appear to share this view themselves, which is positive for parents wishing to moderate the amount of strenuous exercise they do.

4.5 Conclusions

This study clearly identified differences in parent proxy reported HRQL among children and adolescents with SCD; parents underestimated children's total and physical HRQL and adolescents total and psychosocial HRQL. There were also some different predictors of paediatric HRQL between the two groups. Less disease severity in terms of

fewer days missed from school in the past year predicted HRQL in children and adolescents, whereas fewer crises was only important for adolescents' HRQL. In addition, increased moderate exercise predicted better HRQL reports in children but only increased strenuous exercise predicted parent proxy reports in adolescents. Health behaviours, especially exercise are important in paediatric SCD and there are different factors that may influence engagement in these (as shown in Studies 1 and 2), but these have not been examined in paediatric SCD. The next chapter will examine the predictors of health behaviours (exercise and water consumption) in children and adolescents with SCD, as well as examining the predictors of adolescents' alcohol use.

Chapter 5: Examining and Predicting Health Behaviours in Children and Adolescents with SCD (Study 4)

5.1 Introduction

The qualitative studies (1 and 2) in this thesis used a theoretical approach; the Theory of Planned Behaviour (TPB; Ajzen, 1988; Fishbein & Ajzen, 1975) to help understand the health behaviours of children and adolescents with sickle cell disease (SCD). Using the TPB as a framework may be useful as it helps to identify determinants of unhealthy behaviours that may be modified. This is especially important if such behaviours contribute to children's/adolescents' HRQL, as was found for exercise in Study 3. The three constructs of the TPB (Ajzen, 1988; Fishbein & Ajzen, 1975) used in Studies 1 and 2 were helpful in providing insight. However, a larger quantitative study is needed to examine whether these beliefs may determine behaviour. Thus, these three TPB constructs (Ajzen, 1988; Fishbein & Ajzen, 1975); attitude toward the behaviour, SN and PBC, as well as the findings from Studies 1 and 2, were used in the current study to devise health and risky behavioural beliefs questionnaires. The TPB was used as a framework, so the principles of the three components of the TPB were adopted to help understand health behaviour. However, the present research did not seek to directly test the TPB in explaining children's behaviours. This quantitative study investigated children's and adolescents' health beliefs and behaviours (exercise and water consumption) and an aspect of adolescents' risky behaviour (alcohol use). Alcohol use was the only risky behaviour chosen because of the low prevalence of other risky behaviours in Studies 2 and 3.

Research regarding health behaviours is not often conducted in children and adolescents with SCD (especially from a theoretical perspective like the TPB), does not take into account both child and parent reports, does not consider the influence of both the

school and home environment, does not obtain average water consumption and rarely obtains the types (e.g. strenuous, moderate and mild) and quantities of exercise (Dyson et al., 2010a; Karlson et al., 2017; Melo et al., 2018; Omwanghe et al., 2017). Thus the present study incorporated all of these elements. Furthermore, determinants of these behaviours in SCD have rarely been considered, with the exception of a few studies. For example, pain intensity and duration were associated with exercise and hydration (Karlson et al., 2017) and older age, disease impact, negative personal beliefs and perceptions of poor physical functioning were related to exercise (Omwanghe et al., 2017). Therefore, the current study investigated demographics and disease severity, as well as health beliefs, as predictors of exercise and water consumption. As discussed, health beliefs were based on the TPB's attitude, SN and PBC. The TPB has not been used in research on health behaviours in SCD before but it has been used to explain health behaviours in children and adolescents (e.g. McDermott et al. 2015; Martin et al., 2005b; Plotnikoff et al., 2013; Riebl et al., 2015; 2016; Wang & Zhang, 2016).

The qualitative studies in this thesis established that children and adolescents with SCD had some unhealthy behaviours. Although the samples in these studies were small, the findings suggested that environmental and social influences were contributing factors to children's/adolescents' unhealthy behaviours, where parents provided them with sugary drinks rather than water and were less likely to encourage them to participate in exercise due to concerns that this may trigger symptoms of SCD. However, parents' views regarding health behaviours have not been ascertained in prior studies in this area which is important because as revealed in Study 1, they have an influence on whether children engage in healthy behaviour, although they had less of an influence on adolescents in Study 2. Past research on healthy adult children using the TPB shows similarities in beliefs between parents and adult children (Stafleu et al., 1995; Sumodhee & Payne 2016) and this

is likely to begin in childhood. Previous studies have also found that parents' participation in, and support for exercise, predicted a more positive child attitude and increased participation in exercise (Gustafson & Rhodes, 2006; Martin et al., 2005a).

Alcohol use was also examined in the present study. Past evidence in SCD has rarely considered determinants of alcohol use besides Asnani et al., (2014) who found that male gender predicted ever having drunk alcohol and Britto et al., (1998) who reported that the first alcoholic drink was at an older age (i.e. 15 years old or above) for adolescents with SCD and also that they were less likely to have consumed alcohol in the past 30 days. This is important because alcohol use can exacerbate symptoms of SCD as well as leading to psychosocial problems (Brown, 2012). The findings of Study 2 revealed that adolescents with SCD living in London may be more likely to drink alcohol than healthy siblings (contrary to the findings of Britto et al., 1998) and that they drink alcohol with family and socially with their friends. Therefore it is important to ascertain the prevalence of alcohol use in a larger sample and also what factors may facilitate or discourage this behaviour. Thus the current study also investigated demographics and disease severity, as well as health beliefs, as predictors of alcohol use. Once again, health beliefs were based on TPB constructs, which have not been used in adolescents with SCD but have been used to explain alcohol use in healthy adolescents and adults (Cooke et al., 2016; López-Cisneros et al., 2013).

This study had two research aims:

- The first was to examine the relationships between children's and adolescents' self-reported health beliefs and health behaviours and parental reports of children's and adolescents' health beliefs and health behaviours.

The second aim had two parts;

- the first part was to determine the predictors (e.g. demographic indicators, disease severity measures and child and parent health beliefs) of health behaviours in children and adolescents with SCD.
- the second part was to determine the predictors (e.g. demographic indicators, measures of disease severity and health beliefs) of risky behaviours in adolescents with SCD.

5.2 Method

5.2.1 Design

The current cross-sectional study adopted an inter-dependent groups, correlational design using questionnaires. Children and adolescents with SCD as well as their parents or carer completed the same health behaviour questionnaire and adolescents also completed a risky behaviour measure. The primary outcome measures were the health/risky behaviours. The predictors were demographic indicators, disease severity measures as well as health beliefs.

5.2.2 Participants

The convenience sample consisting of 106 children and their parents and 96 adolescents and their parents is described in Chapter 4.

5.2.3 Materials

The materials were outlined in detail earlier in Chapter 4. This includes the measures of exercise (moderate, strenuous and total), water consumption and alcohol use (never drink v drink alcohol), so only the health belief measures will be described below.

The belief items were developed based on the semi-structured interviews and focus groups with 18 children and 23 adolescents with SCD described in Studies 1 and 2 (see Chapters 2 and 3). The items were also developed based on the TPB (Ajzen, 1988; Fishbein & Ajzen, 1975).

5.2.3.1 *The Health Behaviour Questionnaire*

Children and adolescents provided their own beliefs regarding their health behaviours and parents or carers also provided their own beliefs regarding their child's health behaviours. Both groups also answered a question about who had the most influence on each behaviour (e.g. parents, friends, media). The health behaviour questionnaire was piloted with three children with SCD ($M = 8.48$, $SD = 2.75$), three adolescents with SCD ($M = 15.32$, $SD = 2.08$) and also their parents or carers who were identified by the consultant at one of the participating hospitals. Participants were asked whether they had fully understood the questions and were also given the opportunity to raise any concerns about the nature of the questions and the questions were adapted accordingly.

5.2.3.1.1 Beliefs about exercise

The questionnaire assessed the three TPB beliefs; attitude (questions two to five e.g. 'I like doing exercise' and 'I think that doing exercise might make me get a crisis'), SN (questions six to eight e.g. 'important people in my life (e.g. family, friends, teachers, doctors) encourage me to exercise' and 'important people in my life (e.g. family, friends, teachers and doctors) think that doing exercise might be bad for my sickle cell disease') and PBC (perceived behavioural control; questions 10 to 14 e.g. 'it's difficult to exercise because we don't have much money' and 'it's difficult to exercise because sickle cell disease makes me tired'). These were scored on a 7-point Likert scale ('1 Disagree' to '7

Agree’). A total mean score was calculated for each of the TPB beliefs, with a higher score illustrating more positive beliefs. Negatively phrased items were reversed, including all the PBC items. Exercise self-reported and parent-reported TPB components had reasonable to good reliability (DeVellis, 2003) as detailed in Table 5.1; attitude (four items; $\alpha = .61$ to $.79$), SN (three items; $\alpha = .54$ to $.70$) and PBC (five items; $\alpha = .59$ to $.66$).

5.2.3.1.2 Beliefs about water consumption

The three components of the TPB were also evaluated; attitude (questions two to four e.g. ‘I think that drinking water helps me be healthy’ and ‘I think that drinking water helps my sickle cell disease’), SN (questions five to seven e.g. ‘important people in my life (e.g. family, friends, teachers and doctors) drink water’ and ‘important people in my life (e.g. family, friends, teachers and doctors) think that drinking water will help my sickle cell disease e.g. help stop me from getting a crisis’) and PBC (questions nine and 10 e.g. ‘it’s difficult to drink water during lessons at school like maths class’ and ‘it’s difficult to drink water at school as I don’t like getting special treatment because I have sickle cell disease’). Items were scored on a 7-point Likert scale (‘1 Disagree’ to ‘7 Agree’). A mean score was calculated for each TPB component, with a higher score relating to more positive beliefs. There was reasonable reliability for most water consumption TPB components (DeVellis, 2003) with the exception of parental reports of attitude; attitude (three items; $\alpha = .34$ to $.63$), SN (three items; $\alpha = .60$ to $.75$) and PBC (two items; $\alpha = .54$ to $.75$) as reported in Table 5.1. One PBC item ‘it’s difficult drinking water when I’m busy doing other things’ was removed from all analyses to improve reliability.

5.2.3.2 *The Risky Behaviour Questionnaire*

The risky behaviour questionnaire was piloted with three adolescents with SCD ($M = 15.32$, $SD = 2.08$) to ensure that the questions were clearly understood and were nonintrusive. Items measured the three TPB principles in relation to drinking alcohol; attitude (questions one to three e.g. ‘I like drinking alcohol’ and ‘I think that drinking alcohol might make me get a crisis’), SN (questions four to six e.g. ‘important people in my life (e.g. family, friends, teachers and doctors) approve of me drinking alcohol’ and ‘important people in my life (e.g. family, friends, teachers and doctors) think that drinking alcohol might make me get a crisis’) and PBC (questions eight to 10 e.g. ‘it’s difficult for me to drink alcohol because it’s expensive’ and ‘when I’m with my friends it’s hard for me to say ‘no’ to drinking alcohol’). There was a separate question which assessed who had most influence on adolescents’ alcohol use (e.g. friends, parents). Items were scored on a 7-point Likert scale e.g. ‘1 Disagree’ to ‘7 Agree’, and items for each TPB component were summed, where a higher score is a more favourable belief. Negatively worded items were reversed scored. Adolescent self-reported TPB components in Table 5.1 showed good reliability ($\alpha = .70$; DeVellis, 2003) for attitude (three items; $\alpha = .73$), SN (three items; $\alpha = .71$) and PBC (three items; $\alpha = .68$).

Table 5.1

Reliabilities Across the Behavioural Belief Measures for Children with SCD (n=106), Adolescents with SCD (n=96) and their Parents

<u>Health behaviour questionnaire</u>	Children	Parents of children	Adolescents	Parents of adolescents
<u>Exercise TPB beliefs</u>				
Attitude	.70	.77	.79	.61
SN	.60	.61	.54	.70
PBC	.66	.59	.62	.64
<u>Water consumption TPB beliefs</u>				
Attitude	.63	.34	.58	.36
SN	.65	.60	.75	.71
PBC	.54	.58	.75	.68
<u>Risky behaviour questionnaire</u>				
<u>Alcohol TPB beliefs</u>				
Attitude			.73	
SN			.71	
PBC			.68	

5.2.4 Procedure and ethical considerations

The procedure is described in Chapter 4.

5.3 Results

5.3.1 Introduction

This section reports on the health beliefs and health behaviours of children and adolescents with SCD and their parents and also factors that predict children's and adolescents' healthy/risky behaviours.

5.3.2 Descriptive statistics

The descriptive statistics relating to TPB health beliefs (attitude, SN and PBC) and the people who most influence the behaviours are reported below. Descriptives for demographics and disease severity (Tables 4.2 and 4.3), health behaviours (Table 4.5) and alcohol use (Table 4.6) can be found in Chapter 4.

5.3.2.1 TPB health beliefs

Child/adolescent self-reported and parent reported descriptive statistics for the TPB health beliefs (attitude, SN and PBC) are shown in Table 5.2.

Table 5.2

Descriptive Statistics for Health Beliefs of Children (n=106) and Adolescents (n=96) and Parents' Reports of their Child's Health Beliefs

	Child self-report	Parent report	Adolescent self-report	Parent report
	M(SD)	M(SD)	M(SD)	M(SD)
<u>Exercise TPB beliefs</u>				
Attitude	5.41(1.26)	5.11(1.56)	4.98(1.35)	5.30(1.31)
SN	3.46(1.60)	4.43(1.64)	4.63(1.42)	4.18(1.67)
PBC	3.73(1.39)	4.28(1.25)	5.02(1.11)	4.73(1.14)
<u>Water consumption TPB beliefs</u>				
Attitude	5.14(1.26)	5.57(1.04)	5.75(1.12)	5.98(1.01)
SN	5.71(1.10)	6.29(0.77)	6.11(1.04)	6.40(0.84)
PBC	4.36(1.88)	4.27(1.92)	4.56(2.04)	4.01(2.00)

5.3.2.2 People who most influence health/risky behaviours

Children/adolescents and parents identified the most influential person (parent, friend) or organisation (media) on their health behaviours (exercise and water consumption) and also adolescents disclosed who most influenced their alcohol use as indicated in Table 5.3.

Table 5.3

Influences on Children's (n=106) and Adolescents (n=96) Health/Risky Behaviours

	Child self-report	Parent report	Adolescent self-report	Parent report
	% n	% n	% n	% n
Exercise	78% Parent 12% Sibling 5% Teachers 2% Best friend 2 % Doctor 1% Friends	83% Parent 11% Sibling 5% Teachers 1% Friends	33% Parent 28% Friends 17% Media 6% Sibling 6% Best friend 3% Teachers 3 % Doctor 2% Grandparents 1% Classmates	38% Parent 32% Friends 10% Sibling 10% Media 4% Best friend 3% Teachers 2% Grandparents
Water consumption	80% Parent 9% Teachers 6% Grandparents 3% Sibling 1% Best friend 1% Friends	83% Parent 6% Teachers 6% Grandparents 3% Sibling 1% Best friend 2% Friends 1% Doctor	69% Parent 8% Grandparents 6% Friends 5% Sibling 4% Doctor 3% Media 3% Teachers 1% Best friend	70% Parent 7% Sibling 6% Grandparents 6% Friends 6% Teachers 2% Doctor 1% Media 1% Best friend
Alcohol Use			57% Friends 32% Parent 3% Sibling 3% Best friend 2% Teachers 1% Grandparents 1% Classmates	

Children, adolescents and parents generally reported that parents have the greatest influence on water consumption. Similarly, the majority of children and their parents believed that parents influence participation in exercise, whereas there was greater variability in adolescents' and their parents' responses; with beliefs that parents, friends and to a lesser extent the media influence adolescents' participation in exercise. Finally, nearly two-thirds of adolescents reported that their friends or best friend had the most influence on their alcohol use.

5.3.3 Research Aim One: To examine the relationships between children's and adolescents' self-reported health beliefs and health behaviours and parental reports of children's and adolescents' health beliefs and health behaviours.

To address aim one cross-informant agreement between child and parent and adolescent and parent reports of the child/adolescent's health behaviours and then health beliefs were examined. Finally, differences between child and adolescent reports of health behaviours and then health beliefs were also examined.

5.3.3.1 Comparing child/adolescent and parent reports of health behaviours and health beliefs

Initial analyses examined child-parent and adolescent-parent reports of health behaviours (weekly total, strenuous and moderate exercise and weekly water consumption) and also TPB health beliefs (attitude, SN and PBC) which were related to participating in exercise and consuming water using paired t-tests. Cross-informant agreement was statistically significantly different only for child-parent informants for weekly water consumption and also for adolescent-parent informants for weekly total exercise as shown in Table 5.4. Therefore, children reported that they consumed less water than their parents

believed they did, while adolescents reported participating in more weekly total exercise compared to parent reports.

Paired-samples t-tests were also conducted to compare child and adolescent self-reported health beliefs with parent reported health beliefs as shown in Table 5.4.

Cross-informant agreement was statistically significantly different for all beliefs except child and parent reports of PBC over water consumption. Children's attitudes toward exercise were more positive, and adolescents' attitudes were less positive than their parents, whereas adolescents' SNs and PBC were greater than their parents but children's SNs and PBC were lower than their parents. The PBC findings suggest that children perceived more, and adolescents perceived fewer barriers to participating in exercise compared to their parents. For water consumption, children and adolescents had a less positive attitude and lower SNs towards drinking water compared to their parents. However, adolescents had greater PBC than their parents, with the perception that there were fewer barriers to water consumption compared to their parents.

Table 5.4

Mean Health Behaviour Scores and Health Belief Scores for Children (n = 106), Adolescents (n=96) and Parents

	Child self-report	Parent report	<i>t</i>	Adolescent self-report	Parent report	<i>t</i>
	M(SD)	M(SD)		M(SD)	M(SD)	
<u>Health Behaviour</u>						
Weekly exercise	134.37(58.89)	136.10(62.83)	-.52	108.22(68.48)	102.61(69.03)	2.09*
Strenuous exercise	68.8(46.64)	68.77(51.04)	.03	50.44(48.06)	47.63(48.53)	1.38
Moderate exercise	33.16(18.26)	34.95(17.61)	-1.16	27.50(19.64)	27.08(20.15)	.34
Water consumption	31.95(13.62)	34.55(12.88)	-2.83**	43.07(16.18)	42.22(15.99)	.78
<u>Exercise TPB beliefs</u>						
Attitude	5.41(1.26)	5.11(1.56)	2.32*	4.98(1.35)	5.30(1.31)	-3.32**
SN	3.46(1.60)	4.43(1.64)	-3.20**	4.63(1.42)	4.18(1.67)	3.76***
PBC	3.73(1.39)	4.28(1.25)	-2.25*	5.02(1.11)	4.73(1.14)	3.14**
<u>Water consumption TPB beliefs</u>						
Attitude	5.14(1.26)	5.57(1.04)	-4.18***	5.75(1.12)	5.98(1.01)	-2.85**
SN	5.71(1.10)	6.29(0.77)	-6.12***	6.11(1.04)	6.40(0.84)	-2.73**
PBC	4.36(1.88)	4.27(1.92)	.49	4.56(2.04)	4.01(2.00)	4.04***

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

5.3.3.2 Comparing child and adolescent health behaviours and health beliefs

Independent-samples t-tests were conducted to compare child self-reported and adolescent self-reported health behaviours and also health beliefs as shown in Table 5.5.

Table 5.5

Comparing Mean Health Behaviour and Health Belief Scores of Children (n = 106) and Adolescents (n=96)

	Child self-report	Adolescent self-report	
	M(SD)	M(SD)	t
<u>Health Behaviour</u>			
Weekly exercise	134.37(58.89)	108.22(68.48)	2.90**
Strenuous exercise	68.86(46.64)	50.44(48.06)	2.76**
Moderate exercise	33.16(18.26)	27.50(19.64)	2.12*
Water consumption	31.95(13.62)	43.07(16.18)	-5.30***
<u>Exercise TPB beliefs</u>			
Attitude	5.41(1.26)	4.98(1.35)	2.36*
SN	3.46(1.60)	4.63(1.42)	-0.41
PBC	3.73(1.39)	5.02(1.11)	-4.21***
<u>Water consumption TPB beliefs</u>			
Attitude	5.14(1.26)	5.75(1.12)	-3.61***
SN	5.71(1.10)	6.11(1.04)	-2.60*
PBC	4.36(1.88)	4.56(2.04)	-0.74

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Table 5.5 shows that children reported participating in greater levels of exercise (total, strenuous and moderate) compared to adolescents but adolescents reporting consuming more glasses of water in a week than children. The table also indicates that children had a more positive attitude and lower PBC (i.e. more perceived barriers) toward participating in exercise than adolescents. In contrast, adolescents had a more positive attitude toward water consumption and higher SNs than children.

In summary, children reported they consumed less water than their parents believed they do and adolescents reported participating in more weekly total exercise compared to their parents' reports. Children also had less positive attitudes and SNs relating to water consumption than their parents. Adolescents had less positive attitudes to exercise but higher SNs and PBC, whereas children had more positive attitudes to exercise but lower SNs and PBC. There were also differences between child and adolescent reports of health behaviours (i.e. children report they exercise more but drink less water than adolescents) and health beliefs (i.e. children had a more positive attitude towards exercising but perceived more barriers, yet a less positive attitude towards drinking water and lower SNs than adolescents).

5.3.4 Research Aim Two Part I: To determine the predictors (e.g. demographic indicators, disease severity measures and child and parent health beliefs) of health behaviours in children and adolescents with SCD.

To address aim two (part I) preliminary univariate analyses were first conducted to investigate within person relationships between health behaviours (the outcome variable) and the predictors: health beliefs, demographics, and disease severity measures. Then multiple regression analyses were conducted predicting health behaviours from these variables. Analyses were initially conducted for research aim two (parts I and II) to establish there were no noteworthy violations of the assumptions of normality, linearity, multicollinearity and homoscedasticity.

5.3.4.1 Preliminary analyses

Initial univariate analyses were carried-out to help ascertain the statistically significant predictors of health behaviours in children and adolescents to be used in

multiple regression analyses. The relationships between demographic indicators (child's gender and age and also parent's marital status and highest educational attainment level), disease severity measures (number of crises and days missed from school in the last twelve months), health beliefs and health behaviours using child, adolescent and parent reports were examined. As these are preliminary analyses and for the sake of brevity, only statistically significant findings are reported in relation to demographic indicators and disease severity measures. However, findings are fully reported in Appendix G.

Firstly, pearson's correlation analyses were used to explore the within person relationships between reports of TPB health beliefs (attitudes, SN and PBC) and reports of health behaviours (weekly total, strenuous and moderate exercise and weekly water consumption) for children, adolescents and parent reports, as shown in Table 5.6.

Table 5.6

Correlations between Health Beliefs and Health Behaviours in Children (n=106) and their Parents and Adolescents (n=96) and their parents

	Child self-report			Parent report			Adolescent self-report			Parent report		
	Attitude	Subjective norm	PBC	Attitude	Subjective norm	PBC	Attitude	Subjective norm	PBC	Attitude	Subjective norm	PBC
<u>Health Behaviour</u>												
Weekly exercise	.53***	-.40***	-.54***	.47***	.51***	.49***	.54***	-.03	.38***	.59***	.29**	.38***
Strenuous exercise	.47***	-.47***	-.59***	.43***	.51***	.45***	.47***	-.01	.35**	.52***	.24*	.33**
Moderate exercise	.40***	-.13	-.22*	.36***	.31**	.35***	.46***	.03	.29**	.55***	.31**	.32**
Water consumption	.59***	.43***	-.01	.60***	.40***	-.03	.66***	.33**	.37***	.51***	.21*	.15

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Most beliefs and behaviours were significantly positively correlated as predicted by the TPB. However, there were a few exceptions. Adolescent reports of SNs were not correlated with exercise (total, strenuous and moderate) and child and parent reports of PBC were not correlated with water consumption. Furthermore, relationships between exercise (total, strenuous and moderate) and child reports of SNs and PBC were in the opposite direction to anticipated, such that high PBC and high SNs were related to lower reported participation in exercise.

Secondly, independent-samples t-tests were conducted to compare reports of health behaviours between male and female children and adolescents as shown in Table 5.7.

Table 5.7

Gender Differences in Mean Health Behaviour Scores for Children (n=106) and Adolescents (n=96)

	Self-report			Parent report		
	Male M(SD)	Female M(SD)	<i>T</i>	Male M(SD)	Female M(SD)	<i>t</i>
<u>Child health behaviour</u>						
Water consumption	29.37(12.70)	34.63(14.13)	-2.02*	32.06(12.69)	37.13(12.69)	-2.06*
<u>Adolescent health behaviour</u>						
Weekly exercise	140.65(62.62)	75.79(58.38)	5.25***	133.92(63.27)	71.31(60.22)	4.97***
Strenuous exercise	70.50(49.13)	30.38(37.85)	4.48***	65.63(49.29)	29.63(40.84)	3.90***
Moderate exercise	33.65(18.44)	21.35(19.04)	3.21**	35.10(20.33)	19.06(16.62)	4.23***

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

There was a statistically significant difference for child self-reported and parent proxy-reported water consumption where males consumed fewer glasses of water in a week compared to females. There was also a statistically significant difference for adolescent self-reported and parent reported weekly total, strenuous and moderate exercise where males participated in more exercise than females.

Pearson's correlation analyses were used to explore the relationship between child or adolescent age and their health behaviours. There were negative correlations (significant at $p < .05$) between age and participation in weekly total exercise by child self-report ($r = -.22$) and parent report ($r = -.20$) and also participation in moderate exercise by child self-report ($r = -.23$), adolescent self-report ($r = -.22$) and parent proxy-report ($r = -.22$). Therefore, children who participate in more weekly total exercise and also children and adolescents who participate in more moderate exercise are of younger age.

Independent-samples t-tests found that there was no statistically significant difference on health behaviour reports between children and adolescents of parents that are single or married. However, there were differences when comparing parents with different levels of educational attainment using one-way ANOVAs as shown in Table 5.8. There was a statistically significant difference between the 3 educational attainment groups in child self-reported and also parent reported weekly water consumption. Post-hoc comparisons using the Tukey HSD test indicated that children whose parents have a higher degree or degree (group 1) consume more water in a week compared to those who have A Levels or below (group 3) based on child and parent reports. There was also a statistically significant difference between the 3 educational attainment groups in parent reported strenuous exercise for adolescents. Post-hoc comparisons using the Tukey HSD test indicated that parents who had attained A Levels or below (group 3) reported that their child participated in more weekly strenuous exercise compared to parents who attained a

foundation degree or diploma (group 2). There was also a similar trend towards greater adolescent participation in weekly strenuous exercise for group three compared to group two, $F(2, 93) = 3.06, p = .052$.

Table 5.8

One-Way ANOVAs of Health Behaviours in Children (n=106) and Adolescents (n=96) by Highest Parental Educational Attainment

	Self-report			Parent proxy		
	Group 1 M(SD)	Group 2 M(SD)	Group 3 M(SD)	Group 1 M(SD)	Group 2 M(SD)	Group 3 M(SD)
<u>Child health behaviour</u>						
Water consumption	35.95(14.32)	32.22(11.24)	28.44(13.44)	39.58(13.57)	33.70(11.05)	30.73(11.95)
<u>Adolescent health behaviour</u>						
Strenuous exercise	51.69(47.52)	30.91(45.64)	62.21(47.44)	45.23(46.01)	30.52(42.54)	61.94(52.09)

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Group 1 = higher degree or degree. Group 2 = foundation degree or diploma. Group 3 = A Levels or below.

Finally, Pearson's correlations to examine the relationships between disease severity measures (i.e. the number of crises and the number of days missed from school in the last twelve months) and health behaviours are shown in Table 5.9.

Table 5.9

Correlations between Disease Severity Measures and Mean Health Behaviours in Children (n=106) and Adolescents (n=96)

	Crises				Days missed from school			
	Child self-report	Parent proxy	Adolescent self-report	Parent proxy	Child self-report	Parent proxy	Adolescent self-report	Parent proxy
<u>Health Behaviour</u>								
Weekly exercise	-.41***	-.45***	-.27**	-.27**	-.48***	-.50***	-.32**	-.33**
Strenuous exercise	-.39***	-.44***	-.25**	-.24*	-.44***	-.46***	-.26*	-.25*
Moderate exercise	-.17	-.20*	-.19	-.21*	-.31**	-.30**	-.28**	-.27**
Water consumption	.08	.11	-.26**	-.10	.00	.05	-.20	-.06

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

There were significant negative correlations ranging from $r = -0.20$ to -0.50 between disease severity measures and weekly total, strenuous and moderate exercise, with fewer crises or days missed from school related to greater participation in exercise (the only exceptions were that the correlations between child and adolescent self-reports of crises and moderate exercise were not significant). There was only one significant negative correlation ($r = -0.26$) between weekly water consumption and crises on adolescent self-report, where adolescents who consumed more water during the week had fewer crises. Thus overall, disease severity measures were not related to water consumption.

5.3.4.2 Multiple regression analyses

Hierarchical multiple regression analyses were performed to examine the contribution of demographic indicators, disease severity measures and health beliefs in the explanation of children's and adolescents' health behaviours. The relationships between child/adolescent and parent reports of health behaviours were all strong ($> r = .74$) meaning that there was good child/adolescent-parent agreement. Therefore, it was deemed only necessary to use reports from one informant so child/adolescent reports of health behaviours were used. However, parental beliefs were included as predictors to examine whether they had an influence over and above the child's/adolescent's own beliefs. There were four separate four step regression analyses conducted for child self-reported and also adolescent self-reported health behaviours (weekly total, strenuous and moderate exercise and also weekly water consumption), so eight in total. Each regression included a total of 12 predictors; four demographic indicators entered on step one (child's age and gender and also parent's educational attainment which was dummy coded), two disease severity measures entered on step two (number of crises and days missed from school in the last twelve months), three child TPB beliefs entered on step three and three parent TPB beliefs

entered on step four (attitude, SN and PBC). Demographic and disease severity variables included in the regressions were found to be statistically significant in at least one of the preliminary univariate analyses reported above. As noted previously, five participants for each predictor or independent variable is satisfactory in hierarchical regression analyses (Tabachnick & Fidell, 1989, pp. 128-129). The multiple regressions predicting child reports of health behaviours are shown in Table 5.10 and those predicting adolescent reports are shown in Table 5.11. R square change is reported for each step but the betas are reported only for the final step.

Table 5.10

Summary of Hierarchical Multiple Regression Analyses for Child Participation in Health Behaviours (n = 106)

	Weekly exercise			Strenuous exercise			Moderate exercise			Water		
	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change
STEP 1		.068	1.84		.041	1.07		.080	2.19		.106	2.99*
Age	-.10			-.02			-.17			.07		
Gender	.05			.04			-.01			-.01		
Foundation	.08			.05			.05			.11		
Degree	-.13			-.06			-.16			.19*		
STEP 2		.213	14.63***		.194	12.52***		.082	4.87**		.007	.42
Crises	-.03			-.04			.16			-.05		
Days missed	-.10			-.08			-.18			.18		
STEP 3		.229	14.95***		.261	16.54***		.075	3.14*		.312	17.37***
<u>Child TPB</u>												
Attitude	.24*			.15			.28*			.35**		
Sub norm	.47**			.37*			.36			.07		
PBC	-.22*			-.34**			.03			-.03		
STEP 4		.109	8.84***		.100	7.68***		.046	2.00		.092	5.94**
<u>Parent TPB</u>												
Attitude	.03			.04			-.01			.43***		
Sub norm	.64***			.66***			.30			-.09		
PBC	.18			.04			.25			.00		

Note. Betas only in the final model are reported.

* $p < .05$. ** $p < .01$. *** $p < .001$. Sub norm = SN.

F change degrees of freedom are (4,101) at step 1, (2,99) at step 2, (3,96) at step 3 and (3,93) at step 4.

The final model predicting weekly total exercise for children explained 62% ($F(12, 93) = 12.56, p < .001$) of the variance. Child attitudes, PBC and SNs as well as parental SNs were statistically significant predictors. The final model predicting strenuous exercise explained 60% ($F(12, 93) = 11.40, p < .001$) of the variance. Child PBC and SNs as well as parental SNs were statistically significant predictors. The final model predicting moderate exercise explained 28% ($F(12, 93) = 3.07, p = .001$) of the variance. Child attitude was the only significant predictor. The final model predicting water consumption for children explained 52% ($F(12, 93) = 8.32, p < .001$) of the variance as reported in Table 5.9. A parent having a higher degree/degree and child attitude as well as parental attitude were the significant predictors.

R^2 change in Table 5.10 indicates that the inclusion of disease severity measures explained a significant proportion of the variance in weekly total, strenuous and moderate exercise but not in water consumption. Ultimately children's own TPB beliefs regarding each behaviour explained the largest amount of the variance, with parental beliefs also adding a significant, although much smaller, proportion to explaining total and strenuous exercise (although not moderate) and water consumption.

Table 5.11

Summary of Hierarchical Multiple Regression Analyses for Adolescents' Participation in Health Behaviours (n = 96)

	Weekly exercise			Strenuous exercise			Moderate exercise			Water		
	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change	β from last step	R^2 change at each step	F change
STEP 1		.259	7.96***		.214	6.20***		.120	3.10*		.061	1.49
Age	.03			.06			-.02			.11		
Gender	-.27**			-.23*			-.15			-.06		
Foundation	-.08			-.15			.09			-.18		
Degree	-.09			-.08			-.01			-.06		
STEP 2		.062	4.05*		.048	2.91		.039	2.07		.088	4.61*
Crises	-.01			-.07			.05			-.09		
Days missed	-.03			.01			-.02			.01		
STEP 3		.101	4.99**		.073	3.14*		.100	3.85*		.364	21.40***
<u>Adolescent TPB</u>												
Attitude	.10			.11			.09			.41**		
Sub norm	-.12			-.04			-.10			.03		
PBC	.07			.04			.06			.28*		
STEP 4		.068	3.69*		.041	1.83		.072	2.98*		.025	1.47
<u>Parent TPB</u>												
Attitude	.39*			.33			.42*			.27*		
Sub norm	.07			-.03			.05			-.06		
PBC	-.01			.04			-.00			-.09		

Note. Betas only in the final model are reported.

* $p < .05$. ** $p < .01$. *** $p < .001$. Sub norm = SN.

F change degrees of freedom are (4,91) at step 1, (2,89) at step 2, (3,86) at step 3 and (3,83) at step 4.

The final model predicting weekly total exercise for adolescents explained 49% ($F(12, 83) = 6.64, p < .001$) of the variance. Adolescents' male gender and parental attitude were statistically significant predictors. The final model predicting strenuous exercise explained 38% ($F(12, 83) = 4.18, p < .001$) of the variance. Adolescents' gender (male) was a statistically significant predictor. The final model predicting moderate exercise explained 33% ($F(12, 83) = 3.42, p < .001$) of the variance. Parental attitude was a statistically significant predictor. However, adolescent attitude was also a statistically significant predictor at step three in all three exercise models. The final model predicting water consumption for adolescents explained 54% ($F(12, 83) = 8.04, p < .001$) of the variance as described in Table 5.11. Adolescent attitude and PBC as well as parental attitude were statistically significant predictors.

R^2 change in Table 5.11 demonstrates that demographic indicators explained the largest amount of the variance in weekly total, strenuous and moderate exercise, followed by adolescent TPB beliefs. Parental TPB beliefs (for total and moderate exercise) and disease severity measures (for total exercise) also explained a significant, but small amount of variance in exercise participation. R^2 change in Table 5.11 shows that adolescent TPB beliefs and to a much lesser extent disease severity measures explained a significant proportion of the variance in water consumption.

In summary, for children’s total and strenuous exercise, parental SNs and child SNs and PBC were the key predictors (although contrary to expectation, lower PBC i.e. perception of more barriers predicted exercise). For moderate exercise, child attitude was the only predictor. Disease severity measures explained a substantial proportion of the variance but the variables were not significant predictors. In contrast, adolescent’s gender (male) and parental attitude were the key predictors of increased exercise; with the former only predicting total and strenuous exercise and the latter predicting total and moderate exercise. Predictors of child water consumption were child and parental attitude and a parent having a higher degree/degree. Predictors of adolescent water consumption were attitude and PBC as well as parental attitude. Thus attitude is a key variable for water consumption.

5.3.5 Research Aim Two Part II: To determine the predictors (e.g. demographic indicators, measures of disease severity and health beliefs) of risky behaviours in adolescents with SCD.

To address aim two (part II) preliminary univariate analyses were first conducted to examine relationships between alcohol use (outcome variable) and the predictors health beliefs, demographics and disease severity measures. Then logistic regression analyses were conducted predicting alcohol use from these variables.

5.3.5.1 Preliminary analyses

Preliminary univariate analyses were conducted to determine statistically significant predictors of alcohol use in adolescents.

Initial analyses were conducted to examine the relationship between demographic (child's gender and age and also parents marital status and educational attainment) and disease severity measures (child's crises and days missed from school in the last twelve months), health beliefs and alcohol use by comparing those who reported zero units of alcohol in an average week to those who reported consuming some alcohol in an average week.

The relationship between adolescents' health beliefs and their alcohol use was investigated. Independent-samples t-tests were conducted to examine differences in adolescents' TPB beliefs (attitudes, norms and PBC) between those who did and did not drink alcohol as reported in Table 5.12.

Table 5.12

Health Belief Scores and Alcohol Use in Adolescents (n=96)

	Adolescent self-reported alcohol use		<i>t</i>
	Drink M(SD)	Never drink M(SD)	
<u>Alcohol TPB beliefs</u>			
Attitude	4.61(1.01)	2.50(1.09)	-9.51***
SN	5.03(1.27)	2.71(1.07)	-9.06***
PBC	3.30(1.49)	5.36(1.58)	6.36***

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Table 5.12 shows that, as expected from the TPB, adolescents who drank alcohol have a more positive attitude and greater SNs towards alcohol use. However, they also had lower PBC (i.e. believed that there are more barriers to alcohol use) than those who did not drink alcohol, which is contrary to what would be expected according to the TPB.

Independent-samples t-tests were conducted to compare age, number of crises and days missed from school between adolescents that have never drunk alcohol and those that

have drunk alcohol. There was a statistically significant difference in the age at which adolescents drank alcohol, $t(94) = -3.03, p = .003$ (two-tailed) but not the number of crises, $t(47) = 0.65, p = .518$ (two-tailed) or days missed from school, $t(94) = 1.22, p = .224$. Adolescents who did not drink alcohol ($M = 14.65, SD = 1.45$) were younger than those who drink alcohol ($M = 15.65, SD = 1.59$).

Chi-square tests for independence (with Yates Continuity Correction) were conducted for child's gender, parent's marital status and parent's educational attainment in relation to whether adolescents reported drinking or not drinking alcohol. This indicated a statistically significant association between alcohol use and gender, $\chi^2(1, n = 96) = 5.51, p = .019, phi = .010$ but no significant association between alcohol use and parent's marital status $\chi^2(1, n = 96) = 0.27, p = .606, phi = -.074$ or educational attainment, $\chi^2(1, n = 96) = 3.38, p = .185, phi = .188$. Therefore, alcohol use is significantly different between male and female adolescents; 77% of males compared to 52% of females drank alcohol whereas 23% of males and 48% of females never drank alcohol.

5.3.5.2 Logistic regression analysis

Binary logistic regression was performed to assess the impact of a number of factors on the likelihood that adolescents would report drinking alcohol or not as shown in Table 5.13. The three step model contained nine independent variables in total; step one included four demographic indicators (adolescents' age and gender and parent's educational attainment, which was dummy coded); step two included two disease severity measures (number of crises and days missed from school in the last twelve months) and step three included three self-reported TPB health beliefs; adolescents' attitude, SN and PBC. Although preliminary analyses showed that disease severity measures and parent marital status and education were not likely to be important in predicting alcohol

consumption, a decision was made to include the same variables in this logistic regression as the previous multiple regressions predicting health behaviours (in relation to aim four, part I). It is generally accepted that when performing a logistic regression there should be at least 10 events per variable (EPV) for reliable results (Peduzzi, Concato, Kemper, Holford, & Feinstein, 1996), however other authors have suggested that five EPV are sufficient (Vittinghoff & McCulloch, 2007).

Table 5.13

Summary of Logistic Regression Analysis for Variables Predicting Adolescent Alcohol Use (n = 96)

	<i>B</i> from last step	<i>SE</i> β	Wald	e^B	95% CI	
					Lower	Upper
STEP 1						
Age	.74	.29	6.43*	2.09	1.18	3.71
Gender	-.75	.86	.76	.47	.09	2.56
Foundation	-.52	1.18	.19	.60	.06	6.00
Degree	.17	1.02	.03	1.18	.16	8.64
STEP 2						
Crises	.09	.22	.15	1.09	.71	1.68
Days missed	-.03	.02	2.27	.97	.94	1.01
STEP 3						
Attitude	1.17	.47	6.23*	3.21	1.29	8.03
SN	.57	.38	2.26	1.77	.84	3.75
PBC	-.56	.28	3.87*	.57	.33	.998

Note. Betas only in the final model are reported.

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

e^B = exponentiated *B*

F change degrees of freedom for adolescent reports are (4,91) at step 1, (2,89) at step 2 and (3,86) at step 3.

A test of the full model containing all predictors was statistically significant, χ^2 (9, $n = 96$) = 78.85, $p < .001$, indicating that the model was able to distinguish between

adolescents who reported that they did or did not drink alcohol. The model as a whole explained between 56% (Cox & Snell R square) and 77% (Nagelkerke R Squared) of the variance in alcohol use, and correctly classified 92% of cases. Three independent variables made a unique statistically significant contribution to the final model; age, attitude and PBC. Adolescents who were older and had a more positive attitude toward drinking alcohol were more likely (two and three times more likely respectively) to report drinking alcohol when controlling for all other factors in the model. Also, adolescents with lower PBC (i.e. more perceived barriers) were .57 times less likely to report drinking alcohol.

In summary, older age and attitude predicted alcohol use. Also, contrary to expectation, lower PBC i.e. perception of more barriers also predicted alcohol use.

5.4 Discussion

The primary aims of Study 4 were to investigate the predictors of daily health behaviours (exercise and water consumption) in children and adolescents with SCD and also the predictors of alcohol use in adolescents with SCD. This study also examined whether parent reports and child/adolescent reports of the TPB beliefs i.e. attitude, SN and PBC (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975) and health behaviours were different. There were several key findings regarding the predictors of health behaviours. First, children's weekly total and strenuous exercise were predicted by child and parental SNs and also lower child PBC, whereas children's weekly total and moderate exercise was predicted by their attitude. Second, in contrast to children, there were fewer predictors of adolescents' exercise; increased weekly total and strenuous exercise were predicted by male gender and increased weekly total and moderate exercise were predicted by parental

attitude. Third, attitude was a key predictor in both children's and adolescents' water consumption along with a parent having a higher degree/degree (for children) and PBC (for adolescents). Finally, adolescents' alcohol use was predicted by older age and attitude and, unexpectedly, lower PBC. The present study also provided insights regarding differences in child/adolescent and parent reports of health beliefs and health behaviours. First, children report drinking less water, and adolescents report participating in more weekly total exercise than their parents report. Second, children and adolescents had less positive attitudes and SNs to water consumption than their parents. Children also had more positive attitudes to exercise but lower SNs and PBC, whereas adolescents had less positive attitudes to exercise but higher SNs and PBC. Lastly, the present study also found differences in children's and adolescents' health behaviours and health beliefs. For example, children report that they exercise more than adolescents but do not drink as much water as them. Therefore, the findings of Study 4 established that the TPB beliefs (i.e. attitude, SN and PBC) did not consistently predict health behaviours, especially in adolescents.

5.4.1 Research Aim One: To examine the relationships between children's and adolescents' self-reported health beliefs and health behaviours and parental reports of children's and adolescents' health beliefs and health behaviours.

The findings of the present study highlighted the differences between child/adolescent reported health behaviours and beliefs and parent reported health behaviours and health beliefs. It is not surprising that this study also showed that there were some differences between child and adolescent reports of health behaviours (i.e. children report that they exercise more but drink less water than adolescents) and health beliefs (i.e.

children had a more positive attitude towards exercising but a less positive attitude towards drinking water than adolescents).

This study established that parent and child reports of the amount of weekly exercise (i.e. total, strenuous and moderate) that the child participated in were similar whereas adolescents reported participating in more weekly total exercise than their parents believe that they do. This is not unexpected as primary school children are likely to rely on their parents to take, and collect them from school or afterschool clubs and weekend sports activities. In contrast, adolescents are at an age where they are asserting more independency in different areas of their lives, making more choices about what activities they do and are more likely to travel unaccompanied by an adult, so it may be the case that parents are less aware of what they are doing. Similarly, past research has found that many adolescents with SCD participate in physical education classes, sports clubs or team sports (Omwanghe et al., 2017) and they strive for a 'normal' adolescent life (Poku et al., 2018).

There were differences between child/adolescent and parent health beliefs in relation to exercise. Children had a more positive attitude towards exercise, lower SNs (i.e. less pressure) and perceived more barriers than their parents, whilst adolescents' beliefs about exercise compared to their parents were the opposite (i.e. a less positive attitude, more pressure and perceived fewer barriers). Some of these differences may be related to age more than SCD, for example, children may enjoy exercise more than adolescents, and indeed, in the present study children had more positive attitude than adolescents. In contrast, perhaps some adolescents, may, for example, find more pleasure spending time with their friends or attending parties. Therefore, children have a more positive attitude than their parents, whereas exercise may not be as important to adolescents and so they are less enthused than their parents. In addition, children may have lower SNs because they are aware of not being encouraged to exercise due to having SCD, which also means they

perceive more barriers, but adolescents have higher SNs and perceive fewer barriers because perhaps teachers encourage them to exercise which parents may not know.

Children with SCD reported that they drank less water than their parents believe they do whereas parent and adolescent reports of water consumption are similar. Perhaps children are not accurate in their perceptions or may be parents believe that their children drink more at school than in reality. Evidence has shown that adolescents with SCD report that they drink a few glasses of water at home each day (Karlson et al., 2017) although Dyson et al., (2010a) found that approximately half of students were prevented from drinking water during class. The fact that adolescents may be less encouraged to drink water at school than younger children may explain why parents and adolescents provide similar reports of water consumption but children provide lower reports compared to parents who may not be aware of how much they are drinking at school. This is further supported by the finding in the present study that children reported drinking less water than adolescents.

The present study also showed that there were differences in child/adolescent and parent beliefs about water consumption. Parents report a better attitude and higher SNs (i.e. more pressure) regarding drinking water than children and adolescents do. Therefore, parents feel that drinking water is more important than children/adolescents do and even though parents feel they encourage them, this is not felt so much by the child/adolescent themselves. Parents more positive attitudes and higher SNs regarding water consumption are not unexpected as they should be aware that hydration may prevent (Brown, 2012), and help to treat vaso-occlusive crises (Okomo & Meremikwu, 2012) and would feel a sense of responsibility for their child's health. Children/adolescents are also aware of the SCD related benefits of increased water consumption (Poku et al., 2018) which parents encourage (Mitchell et al., 2004), but perhaps they view it as less important or effective

than medical remedies for preventing and treating a vaso-occlusive crisis (Maikler et al., 2001) and therefore feel less pressure to drink water. It may also simply be that children/adolescents attitude to water is based on its plain taste and preference for other beverages.

The present study also illuminated the differences in children's and adolescents' health behaviours and health beliefs. Children participated in more weekly total, strenuous and moderate exercise than adolescents and also had a better attitude towards exercise yet lower PBC (i.e. perceived more barriers) than adolescents. This is in line with Omwanghe et al. (2017) who found that less exercise in children and adolescents with SCD was related to older age, fear of poor physical functioning and negative beliefs about disease impact. It may be that adolescents have other priorities, for example, preparing for their examinations and choosing their subjects (for GCSEs and A Levels) and career paths, as well as socialising with their friends and beginning relationships, whereas children do not have these concerns. The fact that children had more positive attitudes may also help explain the difference in behaviour. In other words, children may exercise more because they enjoy it more (e.g. Sallis et al., 2000), even though they also perceive more barriers, perhaps because they are reliant on their parents who may be reluctant for them to engage in too much exercise due to the possible risk of a vaso-occlusive crisis (e.g. Dyson et al., 2010a; Omwanghe et al., 2017).

This study also concluded that adolescents drank more water than children and had a better attitude and higher SNs (i.e. more pressure) towards drinking water than children, despite the fact that younger children may be more likely to be encouraged to drink water at school. This may be because adolescents are more aware of the health benefits of drinking water for their condition and also general health including helping with their physical appearance, for example, alleviating or helping with acne, shiny hair and strong

nails, which may be more important to them than to younger children. Previous research has shown that adolescents with SCD have in-depth disease knowledge including understanding helpful health behaviours for their condition (Forrester et al., 2015; John-Olabode et al., 2015), which was greater in female adolescents with SCD (Asnani et al., 2017; Bhatt et al., 2011) and that, unsurprisingly there has been an advancement of disease knowledge with age (Metcalfe et al., 2011).

In summary, there are many differences in beliefs between children/adolescents and parents, which may suggest the need to educate both together. Educational interventions should occur at different points during their childhoods as engagement in, and views on health behaviours change with age. For example, highlighting the general and SCD related health benefits of drinking water may address any children's/adolescents' negative attitudes, and perhaps provide some motivation to regularly drink water and also ease some of their parents' burden.

5.4.2 Research Aim Two Part I: To determine the predictors (e.g. demographic indicators, disease severity measures and child and parent health beliefs) of health behaviours in children and adolescents with SCD.

This part of the study examined the predictors of children's and adolescents self-reported health behaviours (weekly total, strenuous and moderate exercise and also weekly water consumption). Different variables predicted children's and adolescents' participation in exercise (e.g. their own TPB beliefs and parental SNs vs. parental attitude and male gender respectively) whereas similar variables (i.e. their own and parental attitude) predicted their water consumption.

In the present study, children's weekly total and strenuous exercise was predicted by parental SNs and child SNs and PBC and their weekly total and moderate exercise was

predicted by a more positive child attitude. These predictors explained around 60% of the variance in total and strenuous exercise but less than half of this (28%) in moderate exercise. It is not clear why the TPB beliefs were less effective in predicting moderate exercise but in all cases, children's own beliefs explained more of the variance in exercise than their parents' beliefs. This study selected a number of variables based on preliminary analyses and other factors that are more relevant to children's moderate levels of exercise may have been overlooked.

Similarly to the present study, past literature has found that SNs and PBC predicted leisure-time physical activity in healthy children (Rhodes et al., 2006) and that SNs and attitude predicted moderate to vigorous physical activity in children of African ethnicity (Martin et al., 2005b). Allender et al., (2006) reviewed qualitative papers which discussed that perceived parental support encouraged children and adolescents to participate in sports and physical activities. However, unexpectedly for child reports in this study, lower PBC i.e. perception of more barriers predicted increased exercise. This may be because in families affected by SCD, parents tend to be protective, especially in relation to reducing the amount of exercise that children were able to participate in (e.g. Forrester et al., 2015; Thomas & Taylor, 2002). It may be that the unpredictable and recurrent nature of sickle cell pain has meant that children have become accustomed to the challenges of their condition which includes planning any type of physical activity despite barriers (Panepinto et al., 2012).

The current study also found that disease severity measures (i.e. vaso-occlusive crises or days missed from school in the past twelve months) explained a substantial proportion of the variance in weekly total and strenuous exercise in children with SCD, and to a lesser extent moderate exercise, although the individual predictors were not significant. A child's level of disease severity is likely to impact the amount of, and

intensity level of exercise a child with SCD can participate in. During a vaso-occlusive crisis, children and adolescents participate in fewer activities including less sports (Fuggle et al., 1996; Jacob et al., 2006; Omwanghe et al., 2017; Shapiro et al., 1990; Walco & Dampier, 1990) which is in line with medical advice (NHS Choices, 2016). Furthermore, participating in strenuous or prolonged exercise can trigger a vaso-occlusive crisis (Connes et al., 2011; NHS Choices, 2019).

Adolescent predictors explained less of the variance in weekly total (49%) and strenuous (38%) exercise than for children, but explained similar variance in moderate exercise (33%). Perhaps the complexities of puberty including the physiological, psychological and social changes effect adolescents' participation in weekly total and strenuous exercise (i.e. they are embarrassed by sweating and are more conscious of their appearance) more than the TPB beliefs. However, this study did establish that male gender predicted increased participation in weekly total and strenuous exercise in adolescents. This is in contrast to the findings of Omwanghe et al. (2017) who did not report any gender differences in the daily physical activity levels and physical education participation in older children and adolescents with SCD. However, literature involving healthy adolescents has consistently shown that males were less sedentary, and that they participated in more moderate/vigorous activity, sports and organised sports than females (Butt, Weinberg, Breckon, & Claytor, 2011; Jago, Anderson, Baranowski, & Watson, 2005; Sallis et al., 2000; Slater & Tiggemann, 2011). This may be because female adolescents reported that they experienced more teasing related to sport and did not enjoy some aspects of physical exertion (e.g. getting sweaty and out of breath), whereas these issues were not as important to adolescent males (Butt et al., 2011; Slater & Tiggemann, 2011). In SCD, some evidence has suggested that male adolescents are body conscious, especially of their smaller physiques and appearance (Erskine, 2011), and this may be an

incentive to exercise. Nevertheless, male adolescents participating in more strenuous exercise is potentially concerning as this may cause a vaso-occlusive crisis (Dyson et al., 2010a; Omwanghe et al. 2017).

Findings in this study also concluded that a positive parental attitude predicted increased weekly total and moderate exercise. This is consistent with a systematic review which found that parents' attitude, overall support and own participation in sports led to greater levels of adolescent participation in overall physical activities but that frequency of physical activity was most affected by parents' encouragement (Edwardson & Gorely, 2010). While parental subjective norms did not predict adolescents' exercise, they did predict children's weekly total and strenuous exercise.

In contrast to the findings in children with SCD, disease severity measures explained little of the variance in adolescents weekly total, strenuous or moderate exercise. This is unexpected because previous studies have reported that sickle related pain and hospitalisations increase with age and are most prominent during adolescence (Blinder et al., 2013; Theodore et al., 2015). It may be that, with age adolescents have developed an ability not to allow SCD and the complications associated with the condition to dominate their lives (Panepinto et al., 2012) and to adapt parts of their daily routine to reduce overexertion (Atoui et al., 2015).

Few health beliefs besides child and parent attitude explained the variance in children's/adolescents' water consumption, and the predictors explained around 50% of the variance, so there are other factors that effect this. The current study found that a more positive attitude in all informants predicted child and adolescent water consumption. A systematic review and meta-analysis also showed that attitude was the strongest predictor of behavioural intention for children and adolescents to eat healthily, which included consuming water. Positive attitudes to water consumption may be because from a young

age they have only been allowed to drink water in schools in England (Department for Education, 2016).

In this study, children's water consumption was also predicted by a parent having a higher degree/degree. Increased hydration can help to manage SCD or treat a vaso-occlusive crisis by decelerating or stopping the sickling process (Beyer et al., 1999; Okomo & Meremikwu, 2012). Perhaps parents who have a higher level of education are more aware of, and therefore are more likely to encourage their child to drink water at home. Similarly, a study found that healthy children of mothers with a higher educational level (a university education) had healthier eating behaviours, for example, eating breakfast and more pieces of fruit and vegetables every day than those with a lower educational level (primary school and lower secondary education) (van Ansem, Schrijvers, Rodenburg, & van de Mheen, 2014). The current parent/carer sample comprised mostly mothers, with around one third or so educated to A level or below. Families affected by SCD tend to be of lower SES (Ilesanmi, 2013).

Future research could explore female adolescents reduced participation in weekly total and strenuous exercise including obstacles such as body image issues. However, there was no difference in their participation in moderate exercise compared to males which is good and should be encouraged. In addition, the factors that influence male adolescents' increased participation in strenuous exercise should be examined. As this is potentially harmful for SCD, it is important that participation in this is safe. The positive aspects of water consumption for SCD and general health should be reinforced to children and adolescents by their parents (whose attitudes on this matter affects their behaviour). Putting this into practice, parents (especially of primary aged children) could be provided with a leaflet during hospital appointments about how water consumption helps SCD.

5.4.3 Research Aim Two Part II: To determine the predictors (e.g. demographic indicators, measures of disease severity and health beliefs) of risky behaviours in adolescents with SCD.

In the current study, adolescents' self-reported alcohol use was only predicted by a single demographic variable (older age) and no disease severity variable. This is in contrast to research indicating that being male was a predictor of adolescents with SCD ever having drunk alcohol (Asnani et al., 2014). In healthy adolescents, regular alcohol use was related to being from a single-parent family (which is common in SCD; Ilesanmi, 2013) and being in higher year groups, whereas less regular alcohol use was associated with being of Black African ethnicity (in which SCD is prevalent; Streetly et al., 2010) and religious observance at least once a week (most adolescents with SCD are religious and regularly attend a religious place of worship; Cotton et al., 2009) (Viner et al., 2006). Therefore, the wider literature provides some support for the findings in this study but it is conceivable that some other aspects of SCD (perhaps prevalence of depression, lower self-esteem or social problems; Edwards et al., 2005; Sehlo & Kamfar, 2015; Ünal et al., 2011) are likely to have some effect on adolescents' engagement in alcohol use but this was not examined in the present study.

In this study, a more positive attitude toward drinking alcohol predicted alcohol use. This is in line with past research in healthy adolescents and adults which also found that attitude was the strongest predictor of intention to use alcohol (Cooke et al., 2016; López-Cisneros et al., 2013). Perhaps adolescents' attitudes toward alcohol use are primarily based on their affective attitude rather than cognitive attitude as healthy adolescents (López-Cisneros et al., 2013) and older adolescents with SCD (John-Olabode et al., 2015) understand that alcohol use can adversely affect their health. For example, in SCD, dehydration (because of alcohol use) can trigger a vaso-occlusive crisis (Brown,

2012) but adolescents get pleasure from drinking socially (Hibell et al., 2012) and so some may engage in this behaviour regardless. It is perhaps surprising though that SNs did not predict alcohol use. Santor, Messervey and Kusumakar (2000) found that peer pressure (i.e. feeling pressured, urged or dared by others) and peer conformity (i.e. an adolescent adopting their peer groups actions) were the strongest predictors of risky behaviours including alcohol use in adolescents. However, the present study reported that, somewhat unexpectedly, lower PBC (i.e. perception of more barriers) predicted adolescents' alcohol use. It may be that adolescents with SCD are greater risk takers, opposing authoritative figures like their parents or doctors, or perhaps they are more rebellious as some aspects of their lives have always been controlled (Atoui et al., 2015; Forrester et al., 2015; Thomas & Taylor, 2002). Thus, if they perceive more barriers to alcohol use this only makes them more determined. Alternatively, simply having a chronic condition may no longer make someone less susceptible to participating in risky behaviours (Sawyer et al., 2007) which may be especially apt in SCD as it is a genetic condition and adolescents may have normalised it.

Adolescents' attitudes towards alcohol use need to be examined further, but perhaps early intervention (and education regarding the relationship between alcohol, dehydration and vaso-occlusive crisis) may prevent (or at the least reduce) excessive drinking at a later stage. A balance needs to be struck between education, advice and discouragement to drink alcohol (as perceived barriers increased drinking among adolescents in the present study), so perhaps these discussions should be led by young adults with SCD or people affected by alcohol misuse.

Ultimately, in the sample in Study 4 thirty-five per cent of adolescents reported that they had never drunk alcohol, and of those who did, this was not on a frequent basis (i.e. approximately only half of participants consumed alcohol a few times a year or once or

twice a month) or excessively (i.e. on average 1.65 units of alcohol a week which is the equivalent of a bottle of lager, beer or cider). Therefore, alcohol consumption in this sample of adolescents with SCD does not appear to be a significant problem.

5.5 Conclusions

The present study examined children's and adolescents' health behaviours and health beliefs. There were some discrepancies between child/adolescent and parent reports; parents believed that children drank water more frequently than children reported themselves and that adolescents participated in less weekly total exercise than adolescents reported themselves. There were many differences between parents and children/adolescents in beliefs about exercise and water consumption. The current study also highlighted the different variables that predict child and adolescent health behaviours. Increased weekly total and strenuous exercise in children was predicted primarily by their own as well as parental SNs, which is not unexpected given their young age, whereas in adolescents they were predicted by male gender and parental attitude. In contrast, there were fewer predictors of moderate exercise; children's own attitude and parental attitude in adolescents. Parental and child/adolescent attitude were also key predictors of increased water consumption in both groups, and adolescents' alcohol use was also primarily predicted by attitude, as well as older age.

This study provides inconsistent support for the TPB (Ajzen, 1988; Fishbein & Ajzen, 1975) in determining the predictors of health behaviours in children and adolescents with SCD. This is particularly the case in adolescents where none of their own beliefs predicted exercise (although their attitude predicted water consumption), whereas in children, their own beliefs explained more variance in exercise than their parents' beliefs. These findings may in part be due to the fact that the TPB was only used as a framework in

this study (and also in the qualitative studies). For example, the beliefs were not operationalised as recommended (e.g. Ajzen, 1988; Fishbein & Ajzen, 1975) and the beliefs were used to predict behaviour rather than to predict intention. This study did not aim to test the TPB but to use it as a framework to gain some insight into the beliefs and behaviours of children and adolescents with SCD.

The next and final chapter will draw together the findings from the qualitative and quantitative studies reported in the previous four chapters and consider strengths and limitations of the research in this thesis, as well as implications for theory and for practice.

Chapter 6. General discussion

6.1 Introduction

Sickle cell disease (SCD) is a life-long inherited chronic condition that has some debilitating features in children and adolescents, for example, painful vaso-occlusive crisis, stroke, infection, growth failure and delayed puberty (Bennett, 2011; Kanter & Kruse-Jarres, 2013; Knight-Madden et al., 2011). They are also known to suffer from psychosocial problems (Anderson et al., 2015; Anie, 2005; Edwards et al., 2005; Sehlo & Kamfar, 2015; Ünal et al., 2011). These clinical manifestations and psychosocial problems may impair their health-related quality of life (HRQL). A plethora of quantitative studies have often reported lower perceived current HRQL in children and adolescents with SCD (e.g. Panepinto & Bonner, 2012). Engagement in daily health behaviours and avoiding risky behaviours may help to manage some symptoms of SCD and improve HRQL. There has been relatively little research into the health/risky behaviours of children and adolescents with SCD, with a few exceptions (e.g. Asnani et al., 2014; Britto et al., 1998; Dyson et al. 2010a; Karlson et al., 2017; Melo et al., 2018; Omwanghe et al., 2017) or how these behaviours may be linked to HRQL (Wrotniak et al., 2014; Zempsky et al., 2013). Importantly, previous literature in paediatric SCD has not explored HRQL or health behaviours from a theoretical perspective, used qualitative methods and has rarely used healthy siblings to act as a comparison group (Hijmans et al., 2010). Thus, the qualitative part of the thesis applied Calman's (1984) Gap theory (GT) to explore the discrepancy between children's/adolescents' current and ideal selves in different areas of their lives. It also drew on the three constructs of the Theory of Planned Behaviour (TPB); attitude, subjective norms (SN) and perceived behavioural control (PBC) (Ajzen, 1988; Fishbein & Ajzen, 1975) to better understand children's/adolescents' health/risky behaviours.

The primary aim of this thesis was to gain a better understanding of the HRQL and health behaviours of children aged 5 to 12 years old and adolescents aged 13 to 18 years old with SCD. Adolescents' engagement in risky behaviours was also examined. This was achieved by adopting an exploratory sequential mixed-methods approach. First, the qualitative studies (i.e. Studies 1 and 2) explored the HRQL and health behaviours (diet, water consumption and exercise levels) of children and adolescents with SCD as well as adolescents' risky behaviours (alcohol use, cigarette smoking and drug use) compared to healthy siblings (who acted as a comparison group). Second, the findings of these studies informed the focus of, and the measures used in Studies 3 and 4 (the quantitative part of the thesis). Study 3 sought to establish the predictors of HRQL and Study 4 examined the predictors of health/risky behaviours.

This chapter presents a summary of the findings from each study, and discusses the overall findings in the context of the wider literature and highlights the contributions made. There is a reflexivity section, where the researcher reflects on how her pre-conceptions and position may have affected the findings of the qualitative studies. Limitations and strengths of the four studies are also outlined. Finally, implications of the research for theory, policy and practice and possible developments for future research are discussed.

6.2 Summary of findings

6.2.1 Study 1: Exploring HRQL and Health Behaviours in Children with SCD and Healthy Siblings

The aim of Study 1 was to explore the HRQL and health behaviours (diet, water consumption and exercise levels) of children with SCD, including any discrepancies between their current and ideal self, compared to that of their healthy siblings. Since the study sample included children aged 5 to 12 years, an appropriate methodological

approach was considered to be a drawing task with a semi-structured interview. In past research with children as young as 5 years old, drawings have been effectively used to engage them in conversation about different topics including coping with SCD (Cotton et al., 2012), issues that are important to them surrounding their HRQL (White et al., 2010) and their past and current views about starting school (Einarsdottir et al., 2009). This technique has also been found to facilitate conversation in clinical settings such as hospitals (Davies & Wright, 2008; Woolford et al., 2013), where many of the semi-structured interviews took place.

Findings of the thematic analysis of the interview transcripts and drawings identified four themes; “limitations of SCD and adjusted expectations”, “awareness but secrecy surrounding SCD”, “coping with SCD” and “influences on health behaviours”. The first theme suggested that there was some discrepancy or ‘gap’ between the current and ideal selves of both groups of children and that this was perhaps more prominent in healthy siblings psychosocial domains but more prominent for children with SCD in the physical domain as they wanted to engage in more physical activities. However, they tried to normalise their condition and so adjusted their expectations accordingly. The second theme “awareness but secrecy surrounding SCD” revealed that both groups of children were secretive about SCD and that this attitude often stemmed from their mothers. In the third theme, both groups of children disclosed that they used different coping strategies when dealing with the condition. For example, children with SCD generally favoured support from their mothers and used more problem-focused coping, sometimes involving health behaviours (such as limiting their exercise). Healthy siblings sought solace through their religious beliefs and thus adopted more emotion-focused coping. The final theme presented the key influences on children’s health behaviours. There was a tendency for parents of children with SCD to restrict their participation in exercise because of the

adverse consequences on their condition (e.g. triggering a vaso-occlusive crisis) but they did not encourage them to drink water, although this is encouraged at school.

6.2.2 Study 2: Exploring HRQL, Health Behaviours and Risky Behaviours in Adolescents with SCD and Healthy Siblings

The primary aim of Study 2 was to explore the HRQL, health behaviours (diet, water consumption and exercise levels) and risky behaviours (alcohol use, tobacco cigarette smoking and illegal drug use) of adolescents with SCD, including any discrepancies between their current and ideal self, compared to that of their healthy siblings. Qualitative methodology was again adopted, as with Study 1, but instead same-sex focus groups of adolescents with SCD and healthy siblings were used as this was considered a more appropriate approach for this age-group. Focus groups have often been successfully applied in health-related research with adolescents (Rabiee, 2004; Wong, 2008) and in SCD (Porter et al., 2014; Valenzuela et al., 2013) to obtain their views regarding healthy eating (Stevenson et al., 2007), exercise (Whitehead & Biddle, 2008) and also their risky behaviours (Lavery et al., 2015).

Thematic analysis of the focus group transcripts from adolescents with SCD and healthy siblings generated four themes across groups; “awareness and disclosure of SCD”, “coping with SCD”, “influences on health behaviours” and “education and beyond”. The first theme suggested that adolescents with SCD were less secretive about their condition than children in Study 1. However, healthy siblings were more reluctant to discuss it, partly out of respect for their sibling. In contrast to Study 1, the second theme, “coping with SCD” revealed that there seemed to be little discrepancy or ‘gap’ in the current and ideal selves of both groups of adolescents. This theme showed that adolescents had normalised SCD. This may partly be attributed to their adaptation to, and greater

responsibility for self-managing SCD, which was achieved through, for example, moderating their exercise and using technology to remind them to take their medication. The third theme identified the different individuals or factors that influenced adolescents' healthy behaviours, which were generally similar to Study 1. Parents encouraged healthy siblings to exercise but not adolescents with SCD and the media had some influence on male adolescents' exercise participation. Male adolescents with SCD discussed body/appearance issues, whereas these issues were more prevalent in girls and boys in Study 1. This theme also showed that alcohol use or experimentation occurred more in adolescents with SCD than healthy siblings and was generally influenced by peer pressure, accessibility and the (mis)perceived lack of effect on SCD. The final theme, "education and beyond" revealed that both groups of adolescents had future life aspirations, which was only found among healthy siblings in Study 1. However, similarly to Study 1, healthy siblings were encouraged more by their parents.

6.2.3 Study 3: Examining and Predicting HRQL in Children and Adolescents with SCD

Study 3 examined the predictors of self-reports of HRQL from children/adolescents with SCD and parent proxy reports of HRQL. It also explored the relationship between child self-reports, adolescent self-reports and parent proxy-reports of HRQL. This part of the research adopted quantitative methodology. A series of questionnaires were given to children and adolescents with SCD and their parents during their routine outpatient appointments across three London hospitals on one occasion (i.e. a cross-sectional study). In paediatric research questionnaires are the most prevalent method used to ascertain child/adolescent self-reported and parent proxy reported HRQL, and the most frequently used measure in SCD is the Pediatric Quality of Life Inventory (PedsQL™; Panepinto & Bonner, 2012). In the present study, questionnaires completed by all participants included

the PedsQL™ and a health behaviour questionnaire and adolescents also completed a risky behaviour measure. The health/risky behaviour measures were designed using the findings from Studies 1 and 2. The primary outcome measures were the HRQL scores generated from the PedsQL™ and predictors were demographic indicators, disease severity measures, coping and healthy/risky behaviours.

Results showed that disease severity was the most consistent predictor of HRQL and also that exercise had some importance (although preliminary analyses suggested that other health/risky behaviours were not linked to HRQL). Specifically, in children with SCD, fewer days missed from school and increased moderate exercise predicted child and parent reported HRQL. In adolescents with SCD, less disease severity (i.e. fewer days missed from school and fewer crises) predicted adolescent and parent reported HRQL, as well as increased strenuous exercise also predicting parent reported HRQL. Study 3 also found that parents provided lower total HRQL reports than children/adolescents as well as underestimating children's physical HRQL and adolescents' psychosocial HRQL. Lastly, it appeared that there were no differences between child and adolescent HRQL, perhaps indicating a smooth transition from childhood to adolescence in SCD.

6.2.4 Study 4: Examining and Predicting Health Behaviours in Children and Adolescents with SCD

The main aims of the final study were to determine the predictors of self-reported daily health behaviours (exercise and water consumption) in children and adolescents with SCD and of risky behaviours (alcohol use) in adolescents with SCD using self-reports. Study 4 also examined differences in child/adolescent reports and parent reports of the TPB beliefs i.e. attitude, SN and PBC (Ajzen, 1988; Ajzen, 1991; Fishbein & Ajzen, 1975) and health behaviours. This study adopted the same methodological approach as Study 3

i.e. using questionnaires (cross-sectional). All participants completed the same health behaviour questionnaire that also included beliefs about exercise and water consumption and adolescents additionally completed a risky behaviour measure that also included beliefs about alcohol consumption. The primary outcome measures were the health/risky behaviours and the predictors were demographic indicators, disease severity measures and health beliefs.

Study 4 found that TPB beliefs did not consistently predict health behaviours. In fact, predictors were more successful at explaining child rather than adolescent participation in different types of exercise (i.e. weekly total, strenuous and moderate). Weekly total and strenuous exercise were predicted by child and parental SNs and lower child PBC. Child attitude also predicted weekly total and moderate exercise. In contrast, in adolescents, only parental attitude predicted increased weekly total and moderate exercise. There were few other predictors of exercise. Male gender predicted increased weekly total and strenuous exercise and disease severity measures explained a substantial proportion of the variance in weekly total and strenuous exercise in children and adolescents with SCD, although no individual predictors were significant. Furthermore, this study found that water consumption was predicted by child and parent attitude in both groups as well as a parent having a higher degree/degree (for children) and PBC (for adolescents). Lastly, adolescents were more likely to drink alcohol if they were older, had more positive attitudes and, surprisingly, had lower PBC (i.e. perceived more barriers).

Another key finding of Study 4 was that children reported drinking less water, and adolescents reported exercising more each week compared to their parents' reports. Beliefs regarding health behaviours also differed. Children had more positive attitudes to exercise but lower SNs (i.e. less pressure) and PBC, (i.e. perception of more barriers) whereas adolescents held the opposing beliefs (i.e. less positive attitudes and higher SNs and PBC)

than their parents. In terms of beliefs about water consumption, children/adolescents had less positive attitudes and SNs than their parents and also parents perceived more barriers than adolescents. Finally, there were differences in children's and adolescents' health behaviours and health beliefs such as children reporting that they participate in more exercise than adolescents but that they drink less water.

6.3 Main discussion

The overarching aim of the thesis was to gain a better understanding of HRQL and health/risky behaviours in children and adolescents with SCD by adopting a mixed-methods approach. This section will bring together the findings of the four studies by first discussing the HRQL of children/adolescents with SCD (from their own, as well as their parents' viewpoints) as well as healthy siblings' HRQL. The relationship between disease severity, health behaviours (specifically exercise) and HRQL will then be discussed. Finally children's/adolescents' health behaviours and the relationship between health beliefs and health behaviours will be considered.

6.3.1 HRQL

The discussion in this section focuses on the contribution of the present research to knowledge about the differences in HRQL between children and adolescents with SCD and the utility of GT in SCD, followed by parents' views of children's/adolescents' HRQL and finally insights regarding healthy siblings.

6.3.1.1 Children and adolescents with SCD and the utility of GT

The qualitative studies based on GT (Calman, 1984) suggested that children and adolescents with SCD had different experiences of HRQL; children appeared to experience

more discrepancy between their current and ideal selves than adolescents. However, differences between these groups were not apparent in Study 3 where there was no difference between children's and adolescents' current perceived HRQL. This may be because this study included a much larger, more diverse sample or due to differences in methodology (i.e. qualitative versus quantitative). However, it is certainly due to the different approaches to HRQL used; there were different findings when using a GT approach (Studies 1 and 2) and when examining current perceived HRQL (Study 3). The findings of Study 3 may be indicative of a smooth transition from childhood into adolescence regardless of SCD and the additional clinical complications that arise during the teenage years, for example, delayed puberty and leg ulcers (Knight-Madden et al., 2011; Serjeant & Serjeant, 2001). Recent advancements in medical interventions and treatments and the level of care at London hospitals may have contributed to a more stable HRQL overtime (Kemp et al., 2015; NHS Choices, 2019), although this has rarely been investigated in earlier research (Brandow et al., 2010). Additionally, the findings of the qualitative studies suggest that, if anything HRQL may improve in adolescence, so again supporting that there may not be a problematic transition for those with SCD.

An explanation for this may be that with age, children with the condition have learnt to accept SCD and normalise it. In Study 1, there were some signs of normalisation such as the majority of children in their ideal drawings did not wish not to have the condition. In Study 2, normalisation was more pronounced. Adolescents spoke of SCD as being part of their identity, similar to any other physical characteristic, like having brown eyes, and also that they had developed more effective coping strategies, for example, avoiding known triggers of vaso-occlusive crises (like strenuous exercise) and using technology to remind them to take their medication. The use of smartphone apps has been found to improve medication adherence in SCD (Badawy et al., 2016; Creary et al., 2014).

This greater sense of self-sufficiency in controlling SCD, coupled with greater maturity and having had the condition for longer may have contributed to narrowing the ‘gap’ in adolescents’ current and ideal selves. Ultimately, it may be that humans are driven to reduce discrepancy as previous theoretical approaches have hypothesised. For example, Rogers (1959) believes that humans strive for congruence i.e. closing the gap between self-image and ideal-image, and cognitive dissonance theory (Festinger, 1957) argues that conflicting attitudes and behaviours lead to emotional discomfort and an imbalance which a person restores by changing said attitudes or behaviours. This may mean that taking a GT approach is not useful and this is supported by previous quantitative research that has applied a gap approach where little discrepancy was found, even in children (not just adolescents) with chronic conditions including sickle cell anaemia (SCA; Constantinou et al., 2015; Heath et al., 2011). It may also provide an explanation for the abundance of literature in paediatric SCD that focuses on current perceived HRQL (Bonner et al., 2010; Dale et al., 2011; Panepinto & Bonner, 2012; Panepinto et al., 2005; 2009).

Another important finding in Study 3 regarded the key predictors of child/adolescent self-reported and parent proxy reported HRQL; broadly disease severity and exercise (the latter will be discussed later in the “health behaviours and HRQL” section 6.3.2 but it should be noted that exercise only influenced children’s self-reports of HRQL and not adolescents’). Disease severity was the most consistent predictor of HRQL. This is in line with paediatric HRQL research (Amr et al., 2011; Brandow et al., 2010; Constantinou et al., 2015; Dampier et al., 2010; Ozer et al., 2014; Panepinto et al., 2005; Schlenz et al., 2012). Greater disease severity is associated with more hospitalisations (Dale et al., 2011; Dampier et al., 2010; Palermo et al., 2002), poorer academic attainment and lower intellect (Berkelhammer et al., 2007) and also poor social relationships and psychological well-being (Anie, 2005; Edwards et al., 2005; Pradhan & Nayak, 2014) and

therefore it is expected to have an adverse impact on HRQL. It is noteworthy that there were some differences between the groups of participants and different measures of disease severity. Both fewer crises and fewer days missed from school affected adolescent HRQL whereas only the latter predicted child HRQL. The number of crises increase during adolescence (Theodore et al., 2015) and the sample in Study 3 contained a greater number of adolescents who had more frequent crises (i.e. five or more) in the past year than children (9% vs. 1% respectively) which may provide an explanation for the difference in the influence of disease severity measures. In contrast, in Study 2, adolescents suggested that the number of crises they experienced had declined with age, and also that they coped with pain better with age, even suggesting that they had a higher pain threshold. Atoui et al., (2015) found that adolescents tend to downplay their pain. Their discussions in Study 2 may also have been influenced by participating in same sex focus groups where they (especially male adolescents) may be conforming to social expectations and asserting their masculinity.

6.3.1.2 Parental view

The findings of the qualitative studies led to the use of the PedsQL™ in Study 3, which is a prevalent current perceived HRQL measure which has been applied in children and adolescents with SCD (Panepinto & Bonner, 2012) but is not based on a specific theoretical approach to HRQL. Study 3 confirmed that in the plethora of HRQL research, parents underestimate the total HRQL of children and adolescents with SCD (Bonner et al., 2010; Dale et al., 2011; Panepinto et al., 2005; Panepinto et al., 2010). This study also illuminated the different domains of HRQL that parents underestimate; children's physical HRQL and adolescents psychosocial HRQL. These findings are again in line with most past studies (Bonner et al., 2010; Dale et al., 2011; Panepinto et al., 2005; Panepinto et al.,

2010). The results of Study 3 may not be unexpected given that in Study 1, children with SCD expressed that they wanted to participate in more exercise but that parents had concerns about how this may adversely affect SCD. Perhaps parents assume that children are less physically capable because of SCD i.e. they are physically smaller in stature (Bennett, 2011) and in past studies their smaller physical size has contributed to their emotional burden (Bhagat et al., 2014). Vigorous or prolonged exercise can exacerbate symptoms of SCD such as triggering a vaso-occlusive crisis (Beyer et al., 1999; Jama et al., 2002; Platt, 1982) and therefore parents fears are reasonable. However, in Study 2 adolescents appeared to have more autonomy and even moderated their own exercise to avoid vaso-occlusive crises but they were not fazed by their parents' apprehensive attitudes to exercise and perhaps this is the reason that parents underestimated physical HRQL for children but not adolescents.

In contrast, parents underestimated adolescents psychosocial HRQL but this was not found in children. These findings are consistent with previous studies where disagreement in adolescent-parent psychosocial HRQL is often reported in adolescents who are healthy and those with chronic conditions (Upton et al., 2008). This may be because of their age and not specifically related to SCD. Parents may recognise that puberty is a difficult time for adolescents who are going through emotional and behavioural changes (Kaltiala-Heino et al., 2003) and adolescents may be less likely to be open about their feelings, leaving parents to reach more dreadful conclusions about their psychosocial well-being. Additionally, as adolescents have more autonomy and are away from the family home more often, parents may have less insight into their social relationships and emotional well-being (Eiser & Varni, 2013). Adolescents' discussions in Study 2 also showed that they had developed greater maturity in different aspects of their lives (e.g. independent disease management) and may be this extends to emotional

awareness. Certainly adolescents said they avoided overburdening their mothers who have their own emotional troubles. However, this again may leave their mothers to imagine the worst, thus leading them to underestimate their psychosocial HRQL. Past evidence has shown that adolescents with SCD recognise that their mothers have their own problems, be these financial or balancing caring for their siblings and running the family home, and therefore seek other avenues of support (Atoui et al., 2015).

6.3.1.3 Healthy siblings

The qualitative studies provided some insights into the areas of HRQL that were more problematic for healthy siblings, especially children, and highlighted differences between children with SCD and healthy siblings. Study 1 revealed that both groups of children had discrepancies between their current and ideal selves but apparently in different domains of their HRQL. In children with SCD, this discrepancy was in their physical HRQL (i.e. they wanted to participate in more exercise) whereas in healthy siblings it was more in their psychological and social HRQL (i.e. they wanted to participate in more recreational activities especially with their family). Therefore, in childhood it appears that SCD also affects healthy siblings but just in different areas of their lives than children with SCD. This is partly because their parents are preoccupied with taking care of their sibling with SCD but also due to financial difficulties. Furthermore, healthy siblings worry about their sibling with SCD and they tend to use religion to help them cope, which may in part be due to them having less opportunities for problem-focused coping and support from their mothers than children with SCD. There have been reports of more psychosocial problems in healthy siblings of children with SCD than healthy children (Hijmans et al., 2009). In contrast to these findings, Study 2 showed that there appeared to be little discrepancy between the current and ideal selves of both groups of adolescents in

any of the domains of HRQL, perhaps with increasing age and maturity both groups of adolescents have developed better coping strategies to cope with SCD and how it can affect their lives (as discussed in section 6.3.1.1 above). For example, in Study 2 adolescents described adapting parts of their daily routine so that they avoided unnecessary overexertion and that they also used their smartphones to remind them to take medication. Also, adolescents have more autonomy than children and therefore healthy siblings would have more opportunities to attend activities and perhaps with older age they have other interests such as socialising with their friends and focusing on their education or exams. Thus issues that seemed more pertinent when healthy siblings were younger are less important to them in adolescence, although the long-term effect on a sibling's relationships with their parents and with their sibling with SCD has not been addressed in the current literature or thesis. Hijmans et al., (2010) found no difference in the current perceived HRQL of children/adolescents with SCD and healthy siblings, and also that both groups of participants had lower HRQL compared to a healthy comparison group. Therefore, the present research and the limited number of studies in this area (e.g. Hijmans et al., 2009; 2010) suggest that healthy siblings' HRQL is similar to children/adolescents with SCD.

6.3.2 Health behaviours and HRQL

This section focuses on the relationship between health behaviours (mainly exercise but also water consumption and alcohol use) and HRQL, which has not been examined in previous research on paediatric SCD. Study 3 found that in addition to disease severity predicting HRQL, increased exercise also predicted HRQL. A study by Ahmed et al., (2015) concluded that adults with SCD who participated in regular exercise (the type, intensity, duration or frequency of which were not specified) showed improvement in some domains of HRQL such as general health, social function and vitality as well as reduced

pain. This may provide some support for the findings of Study 3. More specifically, Study 3 demonstrated that increased moderate exercise (i.e. this is not exhausting but makes someone sweat, and examples include dancing and easy swimming) predicted child and parent reported HRQL and increased strenuous exercise (i.e. this makes someone's heart beat rapidly, breathe harder and faster such as running or playing football) predicted parent proxy reported HRQL in adolescents (Godin, 2011; Godin & Shephard, 1985; 1997; NHS, 2018) but exercise was not a predictor of adolescents' self-reports.

Engaging in moderate exercise on a regular basis is beneficial for a child's/adolescent's general health and is not problematic for SCD. In support of this, Liem et al., (2017) reported that participation in a home aerobic exercise training program did not lead to any significant adverse health effects that were associated with exercise in adolescents and young adults with SCA. Other research in adults with SCD also supports that mild to moderate exercise does not lead to vaso-occlusive crises or other SCD related complications (Balayssac-Siransy et al., 2011; Faes et al., 2014). However, in the qualitative studies (especially Study 1), children/adolescents suggested that their parents were protective and treated them differently to their healthy siblings, for example, taking them less frequently to weekend sporting activities or being more focused on their current physical health, which contributed to a discrepancy between their current and ideal selves. It is imperative that SCD does not interfere with children's/adolescents' participation in moderate exercise and activities.

In Study 3 parent proxy reported HRQL in adolescents was predicted by increased strenuous exercise. The NHS recommends that participation in strenuous forms of exercise or overexertion should be avoided as this may exacerbate symptoms of SCD (NHS Choices, 2019). Similar conclusions were made by Connes et al., (2011) who argued that intense and prolonged exercise can lead to dehydration and also that participating in

contact sports may be precarious for children and adolescents with SCD. In past research, Dyson et al., (2010a) found that unsuitable exercise at school triggered a vaso-occlusive crisis. Exercise can lead to higher blood pressure and a decline in oxygen saturation causing pulmonary hypertension (Machado et al., 2007) and dehydration resulting in a vaso-occlusive crisis due to the slower movement of red blood cells which causes sickle cells to stick together thereby blocking blood capillaries (Brown, 2012). Given these issues outlined, it is not surprising that parents would be protective when it comes to exercise (as discussed in Studies 1 and 2). The fact that for parents in Study 3, increased strenuous exercise predicted increased proxy reports of HRQL (or to put it another way, reduced strenuous exercise predicted reduced proxy reports of HRQL) suggests that they may consider this kind of exercise to be important to their child. However, increased strenuous exercise did not predict adolescents' own HRQL reports so it appears that this is not important to adolescents themselves, which is good given the possible risks of strenuous exercise for SCD.

In contrast to exercise, preliminary analyses in Study 3 suggested that neither water nor alcohol consumption were linked to HRQL. This is surprising as water consumption (and avoiding dehydration caused by drinking alcohol) may help to prevent and reduce the impact of vaso-occlusive crises (Brown, 2012; Okomo & Meremikwu, 2012). However, exercise was such a pertinent issue throughout the qualitative research in this thesis, whereas drinking water or alcohol seemed less of an issue for children and adolescents with SCD. Perhaps exercise has more of an impact in children/adolescents' lives more generally and it is not just about the health benefits, as drinking water is, but it is also about enjoyment, socialising and achievement. In terms of alcohol use, adolescents in Study 3 did not report drinking large quantities of alcohol, or drinking frequently enough for this behaviour to affect their HRQL.

6.3.3 Health behaviours and beliefs

Although the only daily health behaviour in this thesis that played an important role in HRQL among children and adolescents with SCD was exercise, other health behaviours can help to control or worsen the condition, so it is important to establish what may influence them. This association has not been examined in the existing literature. SCD may also prevent or reduce engagement in some health behaviours, such as strenuous exercise as discussed above in section 6.3.2. Few studies have ascertained the level of engagement in health behaviours in SCD (Dyson et al., 2010a; Karlson et al., 2017; Melo et al., 2018; Omwanghe et al., 2017) and no study has applied a theoretical approach to better understand motivations regarding these behaviours in SCD, despite the extensive application of, for example, the Theory of Planned Behaviour (TPB; Ajzen, 1988; Fishbein & Ajzen, 1975) to help to understand children's/adolescents' consumption of a healthy diet, including water intake, and participation in exercise (Keats et al., 2007; Martin et al., 2005b; Riebl et al., 2015; Rhodes et al., 2006). Furthermore, risky behaviours in adolescents may also affect SCD but there has been little research into their engagement in these, and also what influences such behaviours (Asnani et al., 2014; Britto et al., 1998). The dearth of research in these areas led to the application of qualitative methodology (Studies 1 and 2) to initially identify health/risky behaviours and beliefs among children/adolescents with SCD (using healthy siblings as a comparison group). The key constructs of the TPB; attitude, subjective norm (SN) and perceived behavioural control (PBC) (Ajzen, 1988; Fishbein & Ajzen, 1975) were drawn upon to aid understanding of health behaviours (in the qualitative studies and Study 4).

6.3.3.1 Health behaviours

This thesis showed that children/adolescents use exercise to cope with SCD. For example, children with SCD described limiting their exercise when in pain (Study 1) and adolescents discussed avoiding triggers, such as decreasing their exercise levels or reducing overexertion in their daily routines (Study 2), as also found in past studies (Atoui et al., 2015). Panepinto et al., (2012) suggested that better coping strategies develop with age. Furthermore, Study 4 showed that disease severity was potentially important in predicting weekly and strenuous exercise in children. This is not unexpected as past evidence has found that during vaso-occlusive crises, children and adolescents participate in less sports (Fuggle et al., 1996; Jacob et al., 2006; Omwanghe et al., 2017; Walco & Dampier, 1990) and that participation in strenuous exercise can cause a vaso-occlusive crisis (Brown, 2012).

Study 4 indicated that child and parent reports of weekly exercise (i.e. total, strenuous and moderate) were similar but that parents believed that adolescents participated in less weekly exercise than they reported. This is not surprising because parents are more likely to be aware of children's participation in sports as they would collect them from afterschool clubs and take them to weekend sports clubs, whereas adolescents are more likely to travel around London independently. Similarly, Omwanghe et al., (2017) reported that many adolescents with SCD participate in sports clubs or team sports.

Another factor that influenced increased exercise in adolescents with SCD in Study 4 was male gender. There were also some findings specific to males in Study 2, including that their exercise was influenced by the media and body/appearance concerns. The influence of the media in this age group is not surprising and furthermore, male adolescents reported having found role models in sportsmen, perhaps in part because their

parents do not encourage them to exercise (Study 2). Their body/appearance concerns led them to exercise and eat healthily. In contrast, body/appearance issues were prevalent in both girls and boys in children in Study 1. Having SCD may partly explain male adolescents' greater participation in exercise as they spoke of wanting to be big and strong. In SCD, children and male adolescents experience delayed growth whereas with the onset of puberty, females have a growth spurt (Bennett, 2011), which may provide an explanation for why only males shared these concerns in Study 2.

In addition to gender, age may have an influence. Although age was not a predictor of exercise within groups of children or adolescents, Study 4 found that children reported exercising more than adolescents over a week. Similar findings were reported by Omwanghe et al. (2017). Perhaps children get more enjoyment from exercising or may be adolescents are more self-conscious about sweating or embarrassing themselves in front of their peers whilst exercising, or they may use their free time to socialise with their friends or study.

Another health behaviour that may help children/adolescents avoid or manage some symptoms of SCD like vaso-occlusive crises is water consumption (Beyer et al., 1999). When people with SCD are dehydrated their red blood cells move slower which allows sickle cells to stick together and blood capillaries to become blocked triggering a vaso-occlusive crisis (Brown, 2012). The qualitative studies demonstrated that parents do not appear to encourage water consumption at home and that children/adolescents generally drink water at school, most likely because it is one of the few beverages allowed in English schools (Department for Education, 2016). In contrast, Study 4 found that children, adolescents and parents generally reported that parents had the greatest influence on water consumption, although, this study also found that children with SCD reported drinking less water than their parents reported. It may be that children's recall is not accurate.

Alternatively, parents may be reporting what they believe their children should be doing, or perhaps they simply overestimate the amount of water children drink at school or are allowed to drink (Dyson et al., 2010a). Karlson et al., (2017) found that adolescents with SCD drink a few glasses of water at home each day, which may explain why parents' and adolescents' reports of water consumption were similar, whereas children may have other drinks at home, as suggested in Study 1. Indeed, in Study 4, adolescents reported drinking more water than children. They may be more aware of the health benefits of drinking water for SCD and their general well-being or they may believe that drinking water may help improve their appearance (i.e. glossier hair, stronger nails, clearer complexion) whereas children do not have these concerns. However, when coping with sickle-related pain, both children and adolescents did not report drinking water to alleviate their symptoms despite having reasonable knowledge about how beneficial it can be for SCD (Studies 1 and 2). In contrast, they report having their favourite food or drink as a form of comfort. This is in line with other evidence where children would rather take medication or use diversional activities to cope with sickle-related pain (Maikler et al., 2001), although drinking water can help to decelerate or stop the sickling process i.e. treating vaso-occlusive crises (Okomo & Meremikwu, 2012).

Alcohol was the most common substance used by adolescents with SCD, who were more likely to try alcohol, or be regular drinkers than healthy siblings (Study 2). There has been little research into this area but one study reported that adolescents with SCD were more likely to have drunk alcohol than their healthy peers (Asnani et al., 2014), whereas research conducted two decades ago showed the inverse relationship i.e. less alcohol use in SCD (Britto et al., 1998). Therefore, it may be that alcohol experimentation or use in adolescents with SCD is continuing to rise which is concerning as engagement in risky behaviours leads to poor disease management and treatment adherence in adolescents with

chronic conditions (Scaramuzza et al., 2010). Study 4 showed that adolescents' alcohol use was not excessive but regardless of this it can still effect their condition. Alcohol use can cause dehydration which may lead to a vaso-occlusive crisis (Brown, 2012).

6.3.3.2 Health beliefs

This section will consider the three TPB beliefs; attitude, SNs and PBC in relation to children's/adolescents' participation in exercise, consumption of water and alcohol use. The TPB beliefs (Ajzen, 1988; Fishbein & Ajzen, 1975) were not consistent at predicting behaviour in Study 4, although this research only drew on these concepts and did not use them to predict intention.

A key belief in predicting children's exercise was both their own and their parents' SNs (i.e. perceived social pressure), which is expected given their young age and is in line with some past studies in healthy children where SN and PBC predicted intention to participate in leisure-time physical activity (Rhodes et al., 2006). The importance of SNs was also found in Study 1. However, Study 4 reported that in adolescents, only a positive parental attitude predicted increased weekly total and moderate exercise, even though a systematic review and meta-analysis found that adolescents own attitude was the strongest construct for explaining exercise intention (Plotnikoff et al., 2013). Perhaps parents' attitude effects adolescents more than they care to admit. Group discussions in Study 2 suggested that their own beliefs had more influence but participating in focus groups may have affected this, e.g. they may have wanted to appear in control and more independent of their parents.

The fourth study indicated that attitude (as well as parental attitude) was the key TPB belief that predicted increased water consumption in children and adolescents, as well as PBC for adolescents (i.e. fewer perceived barriers). Similarly, a systematic review and

meta-analysis concluded that attitude was the most frequent predictor of intention to consume a healthy diet including drinking water (Riebl et al., 2015). Therefore, strengthening children's/adolescents' positive attitude to water consumption is fundamental.

Adolescents' discussions in Study 2 revealed that they believe that alcohol is the least harmful substance for general and SCD-related health and that peer pressure influences their drinking. Whilst John-Olabode et al., (2015) found that older adolescents with SCD understood the importance of avoiding substance use, including alcohol, this was not evident in Study 2, perhaps reflecting the lack of opportunities for younger adolescents to speak with healthcare professionals without their parents present (Kemp et al., 2015; Surís et al., 2008). Study 4 found that older adolescents were more likely to drink, similar to Britto et al., (1998). However, adolescents' alcohol use was not influenced by their SNs in Study 4, which is surprising given their discussions of peer pressure in Study 2. Instead more positive attitude and surprisingly perception of more barriers were predictors. This may indicate that adolescents' risky behaviours are driven by their affective rather than cognitive attitudes (i.e. they drink because they enjoy it), and that they are more rebellious after having led controlled lives (hence rebelling in the face of barriers). This has been evident in past studies (Atoui et al., 2015; Forrester et al., 2015; Thomas & Taylor, 2002).

6.4 Reflexivity

The section provides a reflection of the qualitative research (Studies 1 and 2). This focuses on how my pre-conceptions and position affected the way I interacted with children/adolescents with SCD which may have influenced their discussions and interpretations and also my analysis and interpretations of the data (i.e. researcher bias).

I acknowledge that my past involvement working with children/adolescents with SCD, conducting interviews and focus groups with them and analysing the transcripts may have influenced my objectivity. I have past experience working with a patient population of the same age (5 to 17 year olds with SCA) in a London hospital for my Master's research whilst conducting quantitative HRQL research. Further, I have worked with adolescents with SCD, facilitating psychoeducation workshops and family days, as well as facilitating focus groups with other chronically ill adolescents and young people. These experiences meant that I already had some knowledge of certain aspects that affected the lives of children/adolescents with SCD.

I am conscious that my past experience shaped the development of the interview and focus group schedules that I used when conducting the research. In the schedules I was inclined to ask some leading questions due to my prior experience working with this population (e.g. "If you do a lot of exercise or sport do you get tired/feel ill/pain?" or "Do they treat you the same as their other friends?"). Consequently, my academic supervisors who have less experience working with children and adolescents with SCD reviewed the questions to ensure they were not closed questions or too leading. Further, whilst conducting the research I allowed participants the freedom to direct conversation.

My previous experience also has some benefits. Prior to focus group discussions, I was aware of the importance of establishing a rapport with group members and also ensuring that they felt comfortable with one another to ensure the richness of the data. Therefore, as part of the focus group schedule, I asked adolescents how they would like me to arrange the chairs, I introduced myself and the research and used icebreakers. In addition, I was aware that adolescents would be discussing sensitive areas and therefore, postulated from my previous experience, that they would feel more comfortable participating in smaller, same sex focus groups.

Conducting the pilot studies was a useful part of the research process because it allowed me to ensure that children understood the questions in the interview schedule and that they were not too demanding (Study 1). Moreover, it allowed me to ensure that the schedules reflected different aspects of their HRQL and health/risky behaviours that were important to the lives of children/adolescents rather than predetermined aspects I considered important because of my past experience with them (and also reduced the number of leading questions). Whilst conducting pilot studies at the hospital I was aware that some parents were waiting and that the interviews or focus group discussions were taking longer than stated in the information sheets. This was overcome when conducting subsequent interviews as I waited until after children had seen the doctor, which allowed more than an hour before their blood tests. I informed parents via telephone, email or in person that interviews and focus groups may take up to an hour and a half, that they were not obliged to wait after they had provided written informed consent and that I would remain in the waiting room to debrief them. The focus groups were conducted during the weekends for convenience of participants and parents. In my experience, this open approach made the parent and child/adolescent more likely to participate in the research and produced more rich discussions because participants were not concerned that their parents were waiting for them and allowed them to speak more freely as they were not afraid that they were listening (even though the door remained closed during interviews/discussions).

Whilst conducting the research, participants sometimes disclosed information I found distressing. In Study 1 some children spoke of experiencing racial abuse and bullying at school and in Study 2 a few adolescents revealed that their family members had recently passed away, that their teachers informed them that they had fewer options available to them because of their ethnicity and that they experienced peer pressure to

engage in risky behaviours. I was conscious that I felt concerned for them and so actively chose to not probe further, which may have meant the data was not as rich as it may have been but I did not want to cause distress.

My prior knowledge about SCD and previous experience working with children/adolescents with SCD could have led me to search for certain themes, for example, how they cope with pain and disruptions in their education because of SCD. However, my two academic supervisors were involved in the research design of both studies and development of the interview/focus group schedules and there was agreement between all parties on the themes and key messages of these themes. Furthermore, my supervisors read most transcripts, reviewed drawings and ensured that the themes were a good reflection of the data.

6.5 Strengths and limitations

There are some limitations that may have impacted the findings of the qualitative and quantitative studies, although the research has many strengths which will also be discussed in this section.

6.5.1 Study samples

The samples firstly underrepresented some children/adolescents with SCD, healthy siblings and their parents. Children and adolescents in all four of the studies were not ethnically diverse as many participants were of Black African or Black Caribbean origin. Studies have shown that SCD is prevalent in other ethnic groups such as those of Indian, Eastern Mediterranean and Arab descent (Serjeant & Serjeant, 2001) and in England occurs in individuals of mixed race (e.g. White and Black African), Bangladeshi, Asian, Indian and Pakistani ethnicities, although it is most common in Black African and Black

Caribbean ethnicities (Streetly et al., 2010). Second, some of the studies utilised a young adolescent population with a mean age of approximately fifteen years old (Studies 2, 3 and 4). Therefore, ascertaining self-report from a greater number of older adolescents may have been valuable as evidence from past research suggested that older adolescents are more likely to engage in risky behaviours (Viner et al., 2006) and also have adapted to SCD more than younger adolescents, which led to higher self-reported HRQL on the Miami Pediatric Quality of Life Questionnaire (Ziadni, Patterson, Pulgarón, Robinson, & Barakat, 2011). In the present research adolescents of all ages seemed to have adapted to SCD, so this may not have made a difference. Third, Studies 1 and 2 (qualitative research) could have included more children and adolescents with more severe conditions such as those who have more vaso-occlusive crises. Furthermore, in Studies 1 and 2 (qualitative research) the severity of children's condition (for example, number of crises, SCD related days missed from school or hospitalisations or children undergoing regular blood transfusions or chelation therapy) could have been ascertained in the demographic information sheet. Questionnaire-based research has found that greater disease severity (such as days missed from school) is related to lower child self-reported QoL (Constantinou et al., 2015; Panepinto et al., 2005; Yates et al., 2009). Fourth, in all of the studies a convenience sample was recruited during children's/adolescents' outpatient appointments. Children and adolescents with SCD and also their parents who attend regular hospital appointments may have a better understanding of SCD or are perhaps more likely to follow medical advice and consequently adopt healthier behaviours and fewer risky behaviours and have a better HRQL (or at least be monitored by hospital staff and have the opportunity to receive additional financial, psychological or social support). Healthy siblings in Studies 1 and 2 were also, initially, recruited through these outpatient appointments and therefore parents with a greater engagement in research, or with fewer

time-demands may have been more likely to participate. Thus the samples may not be representative.

6.5.2 Methodological approach in the qualitative studies

The methodological approach of the research could have led to some shortcomings in Studies 1 and 2. The interview and focus group schedules delved into a number of areas; HRQL (covering multiple aspects including family, friends, physical health/SCD, emotional well-being, school), health behaviours (exercise, diet and water consumption) as well as risky behaviours (alcohol use, tobacco cigarette smoking and substance/drug use) for Study 2. Therefore, in-depth exploration of each area was not fully achievable in a single interview lasting up to an hour (including 5 to 20 minutes to draw two pictures) or a single focus group which lasted between 73 and 96 minutes and so there were areas where findings are not clear because of not having time to probe further. In Study 1, interviews involving younger children were expectedly shorter, thereby leading to perhaps more superficial consideration of some areas that were explored. However, drawings were used as a communication tool in this study and proved successful in engaging all children, especially younger children, in the research and also served to focus children's attention.

Discussions may have been influenced by the different approaches taken in the qualitative studies. In Study 1, drawing tasks were used, where children were asked to draw two pictures, one of themselves doing something that they would normally do and another picture of something that they would like to do. This may have led children to discuss certain areas of their lives such as dissatisfaction with participating in exercise or attending recreational activities. In contrast, the drawing tasks were not included in Study 2 as this technique was not considered appropriate with an adolescent sample, for example,

Driessnack and Furukawa (2012) reported that drawing as a communication tool is used with children who are aged up to twelve years old.

Another issue is the 13 to 17 year old age range of adolescents in Study 2. This may have meant that adolescents did not share similar experiences (for example, in terms of GCSE/A Level exam stress or career choices), younger adolescents may have felt intimidated or dominated by older ones or, perhaps felt that they had to exaggerate their health/risky behaviours and experiences to conform to social expectations. Shared similar experiences can help to facilitate conversations amongst participants in focus groups (Heary & Hennessy, 2002). Heary and Hennessy (2002) postulate that focus groups should be separate for younger (aged 13 to 15 years old) and older (aged 16 to 18 years old) adolescents because they have different life experiences. In reality recruiting participants of similar ages and the same gender to commit to a specific day and time proved challenging, particularly in the case of healthy siblings where there were a limited number of potential participants. Conversely, during the focus groups adolescents gave each other the opportunity to speak and developed good rapport and therefore age distribution was not an obvious issue. In fact, adolescents' behaviours and life experiences were not dependent or reflected in their ages, for example, a male participant with SCD aged 13 years old had engaged in risky behaviour (and was open in expressing his experiences), whereas older adolescents aged 17 years old in the same group had not. Perhaps adolescents with SCD are more self-assured in expressing their own views as suggested in Study 2. Focus groups were also chosen because Peterson-Sweeney (2005) suggests that adolescents may feel more at ease and able to discuss their inner feelings in a group with their peers rather than in a one-to-one interview with an adult researcher.

The influence of discussions in a focus group is another potential limitation (Study 2). Group discussions may have changed how adolescents spoke about the different areas

of HRQL and health/risky behaviours because there were other adolescents around. For example, they may have been more guarded about voicing their insecurities, over- or underemphasised their engagement in, or experiences with risky behaviours, or perhaps, in the case of male adolescents, overstated how well they cope with pain as a means of showing off or asserting their masculinity in front of their male peers. Further, the groups comprised same sex participants who all had SCD or were all healthy siblings which again may have had some bearing on the direction of the discussions. An example of this is that only males with SCD discussed body image, perhaps because that is the direction that those particular groups took, rather than the issue being important to male and not female adolescents. Nevertheless, Peterson-Sweeney (2005) postulates that group member influence is an important aspect of focus groups as this would happen in everyday conversations in natural settings like school, and therefore, this method was on balance, considered appropriate.

The environment may have impacted the findings of the qualitative studies. In Study 1, most interviews involving children with SCD normally took place during their outpatient appointments whereas most healthy sibling interviews were conducted at their homes. In Study 2, all of the focus groups were conducted on hospital premises on a Saturday or Sunday, where members of the sickle cell teams were not normally working, so adolescents may have felt more comfortable to speak openly. Whilst conducting interviews and focus groups at the hospital, parents remained in the waiting room to maintain their child's privacy. Therefore, children with SCD and adolescents may have adapted their responses because of the environment (social desirability), even though interviews and focus groups were conducted away from hospital staff and parents in a private room. In contrast, healthy siblings' interviews in Study 1 were conducted in a more relaxed environment (their home) and because the interviews were conducted afterschool

or at the weekend there were no time constraints. However, healthy siblings may have felt as though their parents were more likely to overhear the interviews (i.e. they were in closer proximity than in a hospital) even though they were never present in the same room during the research.

Despite some possible limitations, the qualitative research provided a “voice” to children as young as 5 years old who live with a life-long chronic condition and healthy siblings who are also affected by SCD. These children/adolescents are often not afforded the chance to openly express their concerns (as the findings in the qualitative studies have suggested) and therefore this is a positive outcome of the research.

6.5.3 Methodological approach in the quantitative studies

In Studies 3 and 4, the questionnaires used may have affected children’s responses. It is possible that younger children (who were as young as five years) may have not fully understood the measures they were completing, although, there was a practice question in the PedsQL™ Young Child Report (for children aged 5 to 7) for the researcher to assess their level of understanding (Study 3). The application of the PedsQL™ in children, adolescents and parents has been extensively assessed (Varni et al., 2005b) and has been validated in paediatric SCD (Panepinto & Bonner, 2012). However, the health behaviour and beliefs measures have not, although they were piloted with a small number of children and adolescents with SCD to ensure that they understood what was being asked of them (Studies 3 and 4). The researcher was also in close proximity to all participants and offered them guidance when needed. Nevertheless, young children may have struggled with them. For example, in two analyses the PBC measure had the opposite relationship to expected with behaviour and it is possible that this was due to their understanding of the questions, although in one of these two analyses this was not with young children but with

adolescents. Additionally, it should be noted that some of the measures had a less than ideal reliability coefficient, although this was mainly only attitudes to water consumption in parents of children and adolescents.

In Study 4, the TPB was not operationalised using the measures recommended by Ajzen (1988) because these were considered too prescriptive and lengthy for a study that was focused on more than just these measures and to be inappropriate for young children. Additionally, Study 4 only measured whether the variables based on the TPB could predict behaviour, but not whether they predict intention to participate in a health behaviour which is the proposition of the model (Ajzen, 1988; Fishbein & Ajzen, 1975). Regardless of this, it was not the aim of the fourth study to test the TPB but just to draw on it as a framework to aid understanding about health behaviours in children and adolescents.

There may also be problems with the measures used to ascertain children's and adolescents' participation in exercise, and levels of water consumption (which was part of the health behaviour questionnaire). The water measure was new and used the researcher's own scoring system because at the time the third and fourth studies were designed there was no pre-existing water measure but since then Karlson et al., (2017) used children's self-reported diary entries to record beverage consumption. Also, the exercise measure used (i.e. the Godin-Leisure-Time Exercise Questionnaire; Godin, 2011; Godin & Shephard, 1985; 1997) has not often been implemented with children and adolescents, but when it has, it has shown good test-retest reliabilities ($\alpha = .81$) (Sallis et al., 1993). Furthermore, the exercise measure has not often been separated into different intensities (i.e. mild, moderate and strenuous) but this was considered useful as participation in strenuous exercise can cause a vaso-occlusive crisis but moderate exercise has general health benefits (NHS Choices, 2019).

The use of self-reports of health/risky behaviours in the quantitative studies may have affected the findings because such reports may not be reliable and socially desirable responding may occur, as found in previous studies that utilise self-report questionnaires/surveys (Sandvik, Diener, & Seidlitz, 2009). In SCD, past research has mostly used subjective measures of exercise or water consumption like self-report questionnaires (e.g. Dyson et al., 2010a; Melo et al., 2018; Karlson et al., 2017; Omwanghe et al., 2017) and therefore this is the most prevalent method. Karlson et al., (2017) used self-report diary entries to determine the beverage consumption of children (aged 11 years and over) who reported drinking four glasses of water at home each day over a two week period. There are a limited number of studies that have examined the frequency of water consumption in SCD and therefore it is difficult to gauge the accuracy of findings in the thesis. For example, as water consumption is integral for preventing and controlling their condition (Brown, 2012; Okomo & Meremikwu, 2012) there may have been some over reporting. In contrast, for exercise, Melo et al., (2018) assessed the physical activity levels of older children and adolescents with SCA using an objective measurement tool (an accelerometer) and also a self-report questionnaire, with both methods yielding the same results (the SCA group had lower activity levels than healthy controls). Nevertheless, objective measures may have been more reliable. It would also have made it possible to see whether differences between children's and parents' reports were due to one or the other under- or over-estimating.

Disease severity measures (i.e. the number of days missed from school and crises in the past year) used in Studies 3 and 4 were also reported, mostly by parents (except for adolescents aged 16 or over). Using objective measures of disease severity may have provided more accurate reports as parents may have had difficulty recalling the exact number of days missed from school due to SCD. However, Constantinou et al., (2015)

used both hospital records and parent reports of disease severity measures; none of which predicted reports of Quality of Life (QoL) by children/adolescents, parents or healthcare professionals and therefore this may not have been an issue.

Studies 3 and 4 implemented many univariate analyses to examine one of the research aims, which may have led to a type 1 error and a bonferroni correction was not used. However, as relatively few analyses were significant, this seems unlikely. Additionally, the key aims in these studies (i.e. predicting HRQL and health and risky behaviours) were examined using multivariate analyses.

Finally, Studies 3 and 4 adopted a cross-sectional design and so changes in HRQL over time were not evaluated using the PedsQLTM and also changes in health behaviours over time were not ascertained. Past research has substantiated that the PedsQLTM is responsive to changes in HRQL over time in children and adolescents with SCD (Brandow et al., 2010). Nevertheless, this means the present study cannot show evidence of causality between variables. For example, it was hypothesised that health behaviours impact on HRQL and this was tested in multiple regression analysis. However, it is also possible that HRQL affects engagement in health behaviours and in fact, the relationship is likely to be reciprocal.

6.6 Implications for practice and future research

The series of studies has identified areas in HRQL and health/risky behaviours that need further investigation and practical implications that may improve the lives of children and adolescents with SCD as well as alleviating the psychosocial effects SCD may have on healthy siblings.

6.6.1 Improving HRQL

6.6.1.1 Communication and support

Parents are responsible for making health decisions on behalf of children/adolescents, yet they have difficulty gauging their HRQL. Parents underestimated children's physical HRQL and adolescents' psychosocial HRQL in Study 3. In practice, there may be some communication issues between children/adolescents and their parents that could be addressed by healthcare professionals in the interim, and in the longer-term, future research should investigate how to improve lines of communication. While child/adolescent-parent communication is important, children and adolescents may also benefit from, and feel more comfortable joining a peer support group, or perhaps from having a mentor who has SCD, who is closer to their age and to whom they can relate more easily than older adults. This may be especially apt in adolescents, who expressed in Study 2 that they only want to speak about SCD with people who can relate to their experiences.

An issue that may undermine communication and support is secrecy surrounding SCD which was prevalent in all children in Study 1 and generally encouraged by mothers and also reported by healthy siblings in Study 2. This is an important issue that needs to be explored further by interviewing mothers and understanding their views and considering how communities/society can begin to alleviate the perceived health-related stigma and better support families affected by SCD. However, secrecy regarding SCD may have led to children developing their own coping mechanisms, such as the use of religion, especially by healthy siblings, as found in the qualitative studies. Moreover, it may be that because most children and adolescents with SCD seem to have adapted to their condition over the years (as suggested in the qualitative studies) they generally do not feel the need for much additional support. Therefore, further research could explore whether

children/adolescents may benefit from additional support or some sort of peer support, or perhaps more open discussions about SCD with their immediate/extended families or other health professionals like counsellors.

This support should also include healthy siblings as the findings of Study 1 suggested that, as children, they are affected by their sibling's condition. Further research should explore the psychosocial effects of SCD in healthy siblings in a larger, more ethnically diverse sample and what the determinants are, and different avenues of support that could be provided to them. The findings of Study 2 suggested that adolescent healthy siblings may be less affected by their sibling's condition and some were more involved, such as setting reminders for them to take their medication. It may be helpful to encourage this involvement, as a form of problem-focused coping.

6.6.1.2 Health behaviours and coping in SCD

The relationship between health behaviours and HRQL was a key contribution of this thesis. In Study 1, there was some discrepancy between children's current and ideal selves which was influenced by their desire to participate in more physical activities, and Study 3 showed that increased exercise predicted better HRQL, although parents discouraged participation in exercise (Studies 1 and 2). Exercise is important to children and adolescents, both for their HRQL and general health, so a balance needs to be struck. Providing specific advice to parents is key at an early stage (i.e. to parents when their child is a toddler). Also, focusing on promoting suitable exercise in this population and highlighting the importance of increased moderate exercise (e.g. walking, cycling, skateboarding, playground activities) for 60 minutes every day, perhaps through educational programs with parents and schools/colleges is imperative. Moreover, in Study 2, adolescents reported moderating their participation in exercise as a means of coping with

the condition. Thus, the role of exercise in coping with SCD could be further promoted as a problem-focused strategy. This is especially important because in Study 3 greater levels of disease severity (i.e. crises and days missed from school in the past year) predicted lower HRQL reports, so strategies that may help to reduce the number of crises and days missed from school are important. This information should be disseminated to parents, schools and healthcare professionals. Although other behaviours in Study 3 were not linked to HRQL, they may still serve as useful coping strategies so it may be beneficial for future research to look in more detail at the use of health behaviours as a source of coping.

6.6.2 Promoting health behaviours

The findings of the qualitative studies suggest that whilst parents, children/adolescents and schools are aware (to varying degrees) of healthy behaviours for children with SCD, these are not routinely followed. There has been some progression since Dyson et al's., (2010a; 2010b) pioneering research into the school experience relating to health behaviours, and some momentum with the publication of more recent work (e.g. Karlson et al., 2017; Melo et al., 2018; Omwanghe et al., 2017). In practice, it is still necessary to highlight the importance of good health behaviours at home and school. In fact, there could be clear guidance advising schools/colleges on the type (e.g. dancing, badminton, easy swimming), intensity (i.e. moderate) and frequency (60 minutes in healthy children/adolescents; NHS, 2018; World Health Organization, 2019) of daily exercise in SCD, as well as on the importance of regular water consumption. Additionally, it seems that current healthcare plans in schools may not be fully followed, so this needs further investigation.

Education for children, adolescents and parents (who did not always share the same health beliefs as their child/adolescent) is also important. While Study 4 did not test whether the TPB predicts intentions and behaviour in children/adolescents with SCD, so

future research still needs to do this, drawing on the TPB in Study 4 suggested that certain beliefs may be important. Child and parent SNs were the key predictors of increased exercise, in adolescents, a more positive parental attitude predicted increased exercise, and in children/adolescents their own attitude and their parents' attitude predicted water consumption. Thus, education should focus on changing unfavourable perceptions of these behaviours, including managing perceptions that exercise may trigger a vaso-occlusive crisis, as discussed in section 6.3.2. For example, leaflets could be distributed during hospital appointments to parents that promote the benefits of hydrating for SCD and children's/adolescents' general physical and mental health (including aspects that may appeal to children/adolescents such as improved complexion and hair). Technology could also be useful. Focus group discussions in Study 2 revealed that adolescents with SCD often learn about SCD online, and that some adolescents use smartphone apps to help adherence to medicine regimens. Earlier studies have also reported that adolescents with SCD would use smartphone apps or technology to learn about SCD (Badawy et al., 2016). Therefore, further research should explore the effectiveness of smartphone apps for improving SCD knowledge of and participation in health behaviours. However, since being male was a predictor of increased exercise in adolescents, there also needs to be further explorative research into female adolescents' reduced participation in exercise which may include broader issues such as body image as well as parental influence and SCD. It may be the case that more targeted intervention is needed to promote exercise in female adolescents.

Education and the use of technology may also be useful for targeting alcohol consumption, as a more positive attitude to drinking and unexpectedly, lower PBC predicted alcohol use in Study 4. Study 2 also revealed a dearth of knowledge about the implications that alcohol use may have on SCD, as well as the influence of peer pressure,

and that adolescents with SCD drink more alcohol than healthy siblings. Thus hospital workshops may be useful here. In particular, these could be facilitated by young adults with SCD or young adults who have overcome alcohol problems, who adolescents can relate to more than authority figures in their lives, who they may wish to rebel against.

6.7 Conclusions

The present research shed light on the HRQL and health behaviours of children and adolescents with SCD. Motivated by the lack of qualitative research and the absence of research that has taken a theoretical approach, a mixed methodological approach was adopted drawing on two theories. Drawing on a GT approach, it was found that children with SCD appeared to have more discrepancy in physical HRQL whereas healthy siblings seemed to have more in psychosocial HRQL. However, even at a young age children had begun to normalise SCD and reassuringly, adolescents appeared to cope with the condition even better and accept it as part of their identity. Consistent with previous research, greater disease severity (in particular fewer days missed from school) predicted increased HRQL but increased moderate exercise also predicted better HRQL in children. Examining the link between health behaviours and HRQL is novel and future research should investigate this further using longitudinal methods and perhaps using healthy siblings or peers as a comparison group in order to understand whether the findings are unique to SCD.

This thesis also contributed to our understanding of health/risky behaviours in children and adolescents with SCD, since there is a dearth of research in this area. Drawing on the TPB constructs, it was found that the most important TPB predictors of children's exercise were child and parent SNs. However, attitude appeared especially important. It was the key predictor of child and adolescent water consumption and of adolescent alcohol use, and parental attitude was also a predictor of child and adolescent water consumption

and adolescent exercise. Therefore, promoting the importance of health behaviours for SCD to parents and children and in schools from the very start of their education is imperative. Additionally, while alcohol use in adolescents was not excessive (and drug use was not prevalent), this may exacerbate SCD and lead to poor disease management in later life. Therefore, intervening at an early stage (i.e. preadolescence) and addressing adolescents' attitude to alcohol is key.

SCD is a chronic life-long condition that has some devastating clinical manifestations like vaso-occlusive crisis but some aspects of it can be managed at home and at school. The present research has shown that children and adolescents have adapted to, and have normalised SCD, developing their coping strategies, especially in adolescence. This thesis has also demonstrated that engaging in healthy behaviours can help manage SCD and that there is the potential for behaviours such as exercise to improve HRQL, if barriers to these behaviours caused by SCD can be overcome. Thus raising awareness of how behaviours can affect, and be affected by SCD is important among all people and organisations that are involved in the care of children/adolescents with SCD.

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Appendix A

Schedules (Studies 1 and 2), Example of Male Focus Group Transcript and Final Coding

Semi-Structured Interview Schedule for Children (5-12 year olds)

Key

Questions for children with Sickle Cell Disease (SCD) only

Questions for healthy siblings only

Italics – for chief investigator’s/researcher reference (applicable to stage 4 and onwards)

Stage 1 - Gaining Informed Consent

- Before gaining any type of consent describe the research.
- Be clear that their participation is voluntary and that this will have no affect on their hospital care and that they can withdraw any time.
- First written informed consent is provided by the parent or carer and then written consent is provided by the child.
- Parent or carer is asked to complete a demographic information sheet.

Stage 2 - Establishing a Rapport

- Establish a rapport with the child, aim to make them feel comfortable, for example would they like the door opened or closed or let them explore the room.
- Make it clear that they can have a drink or snack at any time and use the toilet at any time.

Stage 3 - Introduce the Research

- Briefly introduce the research and remind the child that everything discussed is private.
- Child is told that in special situations if I’m a bit worried about them then I will talk to a doctor, but I will tell them first.
- At the beginning of the interview the child is made aware that “If they don’t want to draw or talk about something I have asked, please just tell me, it is up to you”.

Stage 4 - Beginning the Interview

- Child is asked, just to start off with, can you tell me something about yourself? For example, your age, school year, how many brothers and sisters you have?
- Child is given the chance to ask me questions if they wish. For example, where I am studying?

Question 1 - Here is a piece of paper and some coloured pencils and crayons. Please can you draw a picture of you doing something you normally do? This may be with your family, friends or other people? You can use any colours that you like!

Ask them what they are going to draw. Ensure it is a daily activity they would normally do. Comment on the picture, saying something positive.

Question 2 - Here is another piece of paper. There may be some things that you can't normally do, maybe things that your friends can do but that you can't. So this time please can you draw a picture of you doing something you wish you could do? This might be with family, friends or maybe some other people? Remember this should be something that you normally wouldn't do but would like to do? You can use any colours that you like!

Ask them what they are going to draw. Ensure it is a daily activity that they would like to do.

Comment on the picture, saying something positive.

- What are you doing (*point to each picture*)? *Explore any differences in what they're doing in each picture and why they can't normally do what they have drawn in their second picture?*
- What kinds of things do you normally do outside of school?
- Are there things you would like to do that you can't normally do e.g. maybe things that your friends do but you can't? How does that make you feel?
- How are you feeling (*point to each picture*)? Why?
- Do you feel like this a lot? (*Explore any differences in feelings between the 2 pictures*).
- What kinds of things make you feel happy? What kinds of things make you feel sad or worried? Do you often worry or feel sad? Who do you speak too?
- Who are in your pictures? Why have you chosen to draw them? (*Explore any differences between the 2 pictures*).
- Can you tell me about your family and friends?
- Would you like to have more family or friends? Why is that?
- How do you feel about school? (Prompts: What are the people like; how do you get along with them? Can you tell me about your schoolwork and homework?)
- Do you ever get behind or have to miss school? Why?
- What's it like having Sickle Cell Disease (SCD)? (*If they talk about feeling ill, pain, going to the Doctor explore how often, how they feel, what happens*).
- Have you told your friends that you have SCD? Why/why not? How did they react? How did you feel about that?
What do your friends know about SCD?
Have they asked you questions about SCD? What?
- How do you feel about having a brother or sister who has SCD?
- How does it affect you?

Question 3 - Can you tell me about any types of exercise and sport you do every day?

This might include walking and riding a bike, anything where you are moving about?

- What do/don't you like doing?
- Are there things you can't do (things that maybe your friends can do)? Why can't you do these things? Who tells you that you shouldn't do them? If you feel ill or tired or have pain what things can't you do then?
- Are there things you would like to do?
- If you do a lot of exercise or sport do you get tired/feel ill/pain?

(Within these questions explore influence of parents, friends, teachers, school, likes/dislikes and having SCD).

Question 4 - Can you tell me about what you eat and drink?

- What do/don't you like eating and drinking? Why?
- What do you eat and drink at home/school/hospital?
- Are there things you can't eat and drink that maybe your friends are allowed to? Why can't you eat these things? Who says you can't eat or drink these things? When you're ill or tired or have pain what can't you eat and drink?
- *If you don't eat and drink does that ever make you feel ill or in more pain?*
- Are there things you would like to eat and drink?

(Within these questions explore influence of parents, friends, teachers, school, likes/dislikes and having SCD).

Stage 5 - Ending the Interview

- The child is asked, "Is there anything else you would like to say or ask me before the interview ends?"
- Thank you for taking part today. It was very interesting to hear your likes and dislikes. For example,.....
- Is it ok if I still keep/photocopy your pictures?

Stage 6 - After the Interview

- *Debriefing after the interview with the child.*
- *Make sure the child feels 'ok' after the interview has ended and there is nothing else they would like to talk about.*
- *Debriefing with their parent or carer.*

Focus Group Schedule for Adolescents (13 to 18 year olds)

Stage 1 - Gaining Informed Consent

- Before gaining any type of consent describe the research.
- Be clear that their participation is voluntary and this will have no affect on their hospital care and that they can withdraw any time.
- First written informed consent is provided by the parent or carer and then written consent is provided by the young person. If they are 16 years old and older then consent is only needed from the young person.
- Parent or carer or young person aged 16 years and older is asked to complete a demographic information sheet.

Stage 2 - Establishing a Rapport

- Door will be closed, to maintain confidentiality.
- Establish a rapport with the young people, aim to make them feel comfortable. For example, ask them how they would like to arrange the chairs i.e. in a circle, rows.
- Make it clear that they can have a drink or snack at any time and use the toilet at any time.

Stage 3 - Introduce the Research

- Briefly introduce myself and the research and remind the young people that everything discussed is confidential.
- Young people are informed that confidence is only broken in exceptional circumstances, if I'm very concerned about them, but I will talk to them before speaking with a consultant.
- At the beginning of the focus group the young people are informed "If I ask you something that you don't want to discuss in the group, please just tell me that you'd rather not go into that. It is your choice".

Stage 4 - Beginning the Focus Group

- Young people are asked, just to start off with, to introduce themselves? For example, your age, favourite band/singer, something interesting about yourself?
- Icebreakers to ensure the young people are comfortable.
- The chief investigator/researcher will then lead a discussion referring to the following questions:

(Nb: The chief investigator/researcher notes that the group discussion may naturally develop to cover some of these areas, however, these questions have been drafted as a point of reference to prompt and direct discussion if necessary).

Part One: Health-related quality of life

Let's consider your daily lives....

- 1) What kinds of things do you normally do in your free time like evenings and weekends?
 - a. Who or what influences what you choose to do and why them?

- 2) Are there things you would like to do in your free time that you can't normally do e.g. maybe things that your friends do but you can't?
- Who or what stops you from doing these things now, and why?
 - How does that make you feel?

Important people in your lives...

- 3) Can you tell me about the important people in your lives?
- In what ways are they important to you?
 - How do they help you?
 - How do they make you feel?

4) Is there anything you'd like to change about the people in your lives or the relationships you have with people? *Example provided for prompting if necessary: for example, would you like to know more people or see people more or get on better with people?*

- In what ways would this make things better for you?

Now let's consider school/college/training/work...

- 5) How do you feel about your school/college/training/work?
- What are the people like; how do you get along with them?
 - What are the positives and negatives of going to school, college, training or work?
 - Are there any things you would like to do at school, college, training or work but can't?
 - Do you ever miss school, college, training or struggle to get work done? If so, why? How does this make you feel?

6) Are there things that you would like to change about school/college/training/work, and if so what? *Example provided for prompting if necessary: for example, you wish you had more friends or found the work easier?*

I would like to discuss your emotional well-being and physical health...

7) How do you feel emotionally in your day-to-day lives? *Example provided for prompting if necessary: for example, what kinds of things make you feel happy or what kinds of things make you feel sad or worried?*

- Who would you speak to and why them? Can we think of any examples?

8) How do you feel physically in your day-to-day lives? *Example provided for prompting if necessary: for example, you end up falling behind on your school work or you can't go out with friends?*

- Does being unwell or in pain interfere with any part of your life? Can we think of examples?

Having Sickle Cell Disease (SCD) or a sibling with SCD...

- 9) We have already spoken about this a bit e.g. you said that..... but how does having SCD (*having a brother or sister with SCD**) affect your day-to-day lives? *Example provided for prompting if necessary: for example, you have to regularly attend the hospital or you can't do things when you're (your brother or sister is*) in pain?*
- 10) Who knows that you (*your brother or sister**) have SCD?
- 11) What do your friends know about SCD? Can you please give examples?
- a. Are things about SCD and how it affects your lives (*your brother or sister* lives*) that you think your friends don't understand?
 - b. Do they treat you the same as their other friends? In what ways?

Part Two: Health Behaviours

Now let's consider your daily exercise...

- 12) Can you please tell me about any types of exercise you participate in? *Example provided for prompting if necessary: for example, walking to school/college/training, going to the gym or other types of exercise, being part of a sports team or individual sport etc.*
- a. Who or what influences the types of exercise you choose to participate in? (*Explore influence of their attitudes i.e. likes and dislikes, friends and family attitudes and behaviours, school and teachers, media, SCD*).
- 13) Are there things you'd like to do but can't do, e.g. maybe things your friends can do? Why can't you do them? (*Explore influence of parents and teachers i.e. what they're told to do/not to do, and daily influence of SCD as well as crises and hospitalisation*).
- a. How does this make you feel?
- 14) How do you feel after participating in different types of exercise?

How about your diet...

- 15) Can you tell me about what you normally eat and drink?
- a. What or who influences what you decide to eat and drink? (*Explore influence of their attitudes i.e. likes and dislikes, friends and family attitudes and behaviours, school and teachers, media, SCD*).
- 16) Are there things that you can't eat and drink and would like to? Why? (*Explore influence of parents and teachers i.e. what they're told to do/not to do, and daily influence of SCD as well as crises and hospitalisation*).
- a. Does anything you eat or drink make you feel ill?

Part Three: Risky Behaviours

Ok, lastly I would like to ask you about risky behaviours...

- 17) What do you know about alcohol, smoking and other substances? *Example provided for prompting if necessary: are these behaviours good/bad for your health, do they have short/long-term effects, do they lead to addiction etc.*
- 18) Has anyone ever tried alcohol, smoking or other substances?
- Can you tell me about the first time you tried it and describe what it was like? *Only applicable to young people who have participated in risky behaviours.*
 - Have you drunk alcohol, smoked or taken other substances more than once, and if you have, why? How often do you do this now? *Only applicable to young people who have participated in risky behaviours.*
 - What types of alcohol or other substances including smoking would you be tempted to try, and why? *Only applicable to young people who have not participated in risky behaviours.*
- 19) Who or what influenced your decision to try/not try alcohol, smoking or other substances? *(Explore influence of their attitudes, friends and family attitudes and behaviours, media, SCD).*
- 20) Why do you think it's important to be careful about how much you engage in the health behaviours we have spoken about today like exercise, what you eat and drink, and drinking alcohol and smoking?
- Do you think they have any affect on your SCD? *Only applicable to young people with SCD.*
- 21) Are there any areas, not previously discussed today that you think would be relevant, or that you think we should consider related to your lives, health behaviours and risky behaviours?

An Example of a Male Focus Group Transcript

- Res Ok so we've talked about religion a lot. And that most of you choose to attend Church or worship God in your own way. So who or what influences the things other things you choose to do? So hanging out with your friends, playing sports like football or basketball, is that right? ((agreement))
- Res So why do you choose to do those things?
- Ayo Mainly because you just get to get some time off school without always having to see your friends in school. You can meet them outside of school and ((pauses)) do everything like you want. Makes own choices regarding free time
- Res Oh, Ok. What about anyone else? So why do you choose to do those things?
- Ayo ((hesitation)) Just to have fun. Because you can't really speak as much talk as much, do as much when you're in school rather than at weekends.
- Res And ██████, you were going to say something? Enjoys socialising with friends outside of school. This provides fun.
- Abidemi It's usually different from what everyone else is doing, so, and if you enjoy it, it's better than doing nothing. Makes own choices in relation to sports
- Res You're talking about playing basketball...
- Abidemi Yeah.
- Res When you say that. So you just enjoy playing it, so you choose to do it.
- Abidemi Well, yeah, I'm tall, well, for my age, kind of. Has the physique/height for basketball

- Res For your age, yeah. So, you all seem to hang out with your friends. I'm wondering why you don't hang out with your family in your free time or maybe you do...? Spends some time with his family in free time Family live in Nigeria / outside of London
- Adom Well, truthfully, I do hang out with my family, ((pauses)) but the majority of the time, usually my brothers, they're always out because they've got either trials for teams, ((hesitation)) or they usually go to their friends' houses. And some of my family ((hesitation)) live out of London and abroad like in Nigeria, so I can't really hang out with them, and... Siblings are busy
- Res Are they older, then, your brothers?
- Adom Yeah.
- Res Uh huh. So they do their own thing, and other people are nodding. So is that the same for other people? Spends less time with older siblings
- Ayo Brothers and sisters are older, so, I do see them a lot, at home, but they're older so they do their own things and have their own lives with their friends.
- Res When you say older, a few years older, or ten, fifteen years older?
- Ayo Yeah, about, for me, ten, fifteen. Large age gap between siblings
- Res Yeah, so it's quite a big age gap, yeah.
- Adom About four. Four, five.
- Res Ok. But still, obviously quite a bit older so they want to do their own thing. What about other people? So how come you don't hang out with your family as much as your friends? Is there any particular reason, or..?
- Ayo My brothers are usually out, busy.
- Res Sorry, your parents or your brothers?

Ayo Brothers.

Abidemi I find it boring, staying at home. Being at home is boring

Res You don't like to stay home, it's boring. Fair enough. Do you have brothers or sisters?

Abidemi Yeah, younger.

Res Oh, Ok, they're all younger. And ████?

Azi Yeah, I do socialise with my family every now and then. Spends less time with his family socially

Res But not regularly. Oh, how come? Is it an age thing as well?

Azi No, it's just...

Res Alright, you just don't. Do you live with your parents ████?

Azi Yeah.

Res Ok. Do you have brothers and sisters?

Azi Yeah. I'm just always out. Chooses to spend time outside of the family home

Res Oh, alright, you're just always out.

Azi Yeah.

Res Ok. So next, is there anything that you would like to do that you normally can't do in your free time? Friends moved away from London

Adom Well, ((pauses)) I would sometimes like, say for instance that my friends, they've moved out of London. And they're not allowed to come back. ((hesitation)) Because, I don't know why, but, I would like... Smaller support network (friends)

Res Do you mean they've moved school, or they moved out, or..?

Adom They moved out of London.

- Res Oh, they just moved out completely?
- Adom Yeah. So I would like to go visit them, but sometimes I won't be able to do that because I either don't have the money for it, or sometimes some issues with my friends. Money issues / how low income impacts seeing friends
- Res Ah ok, so some issues with friends and because of money. Does anyone else experience the same thing, so money or friend issues means you can't do things you'd like to do?
- Ayo No. Money is not an issue.
- Abidemi No. Money is generally not an issue
- Azi Nah, there's nothing I can think of that I want to do now that I can't do.
- Adegoke No, nope, money doesn't hold me back, there's free stuff in London too. Parks are free, like, for example. Low income / finances are not a barrier Free recreational activities
- Res Ok, so we can move on, then. So next we're going to talk about important people in your lives, so, can you name me some important people in your lives? Who comes to mind?
- Ayo Mum.
- Abidemi Yeah.
- Adegoke Mum, yeah mum.
- Adom Mum, Dad, broth... Mum, brothers...
- Abidemi My coach is... His coach is an important person
- Res Your coach is?
- Abidemi Yeah.

- Ayo Mum, nephews, brothers, sisters, friends. Family especially parents are the most important people in their lives
- Adom And teachers. Teachers are important people
- Ayo Some teachers.
- Res Teachers, really? Ok.
- Azi Yeah, just family and friends.
- Res Family and friends. ██████, who would you say, like..?
- Atu Family, friends. Generally family and friends are important people
- Res Family and friends. And why are these people important to you? How are they important?
- Adom Well, ((pauses)) my mum is important because at the end of the day, if for her, I wouldn't've been born ((pauses)) and I wouldn't be here right now.
- Yeah, and... Appreciates his mum. Acknowledges her importance
- Res That's a very good point!
- Adom For my teachers, they're important as well because they would help you in life, they teach you, ((hesitation)) to get qualifications. And they can help you to get sponsors and stuff. Teachers help to obtain qualifications / sponsors Ambitious
- Res What do you mean, sponsors?
- Adom Help you to get... Say for instance you were playing football, they'd help you to get into a team and then get sponsored by something like Nike or.
- Res Oh, Ok. Do they encourage that, then?
- Adom Yeah.
- Res Oh, Ok. Is that the same with anyone else?

Ayo Yeah, our school gets sponsored by Nike, so we can do things like sports and stuff. School is sponsored by Nike

Res Is it an academy, that you go to or normal school?

Ayo Yeah, academy.

Res Uh huh. So why are the people that you guys have named like mum, coaches, so important to you? Important people in their lives provide support

Ayo Mostly because of support. The main thing they do is support you to get you to where you want to be. Adolescents consider their goals

Res Mm-hm. Is that your coach, teachers or your mum, dad or other people...?

Ayo Teachers and your parents, because without them, at a young age, you wouldn't be able to get where you are now. They're the ones who help you, physically, mentally, financially and all of that. Main role models are adolescents' parents and teachers, supporting different aspects of their lives

Res What do you mean physically and mentally?

Ayo Just if you feel like you can't get somewhere, they'll always say 'Oh yeah, you can do it, come on get the strength and try.' Important people are motivating

Res Mm-hm, so they're encouraging, motivate you to do things. Ok, and [REDACTED] and [REDACTED], why have you named those people. So you said family and friends, is that right?

Atu Yeah, because my family and close friends, they're always there for me.

Res They're consistent throughout your life. Family / close friends provide stability

Atu Yeah.

- Res And do they help you in any particular way? Can you think of any ways they might help you?
- Atu For example ((hesitation)) if I'm not feeling right, ((hesitation)) they'll try and make me laugh, or make me feel happy. Generally, important people in their lives makes them feel better / tries to help them
- Res And [REDACTED], did you say your teachers or your coaches, as well?
- Abidemi My coach, yeah.
- Res Why are they so important to you? Sports participation requires them to be disciplined
- Abidemi I see a lot of other clubs, doing different sports, some of them aren't that disciplined. But then, I just thought, because he seems different from others, like, can I see the difference between them. And our coaches give us discipline, and you're able to do work ((pauses)) and like at school, outside, they give you options to try to do coaching jobs and stuff. Coaches provide them with opportunities (like work experience)
- Res Uh huh. So they give you opportunities...Is that the same with your parents as well, or...?
- Abidemi Yeah, my parents are in touch with the coaches but it's the coaches that push me more and teachers, not my parents really. Coaches are more encouraging than parents (sports related)
- Res Uh huh [REDACTED]. sorry, were you going to say something?
- Adom Oh yeah, and my brothers as well.
- Res Your brothers. How come your brothers? Brothers are role models / supportive
- Adom Because, say for instance I didn't have my parents, I could rely on my brothers, because they've always got my back, no matter what.
- Res Mm-hm, Anything else you want to add? [REDACTED]? No other people are important?

- Azi Parents and friends and family.
- Res Ok. How do your friends support you?
- Azi They're just there, really.
- Res Just there. No particular way.
- Azi No, I just know that they're there which is basically more important than anything else they can do. Just that they're there.
- Res Mm-hm. So is there anything that you would like to change about the people in your lives, or the relationships you have with them? Ah, people are nodding no?
- Adom Well ((hesitation)) see my brothers more obviously would be the only thing.
- Res See your brothers more, yeah. That's it, just see your brothers more.
- Adom Yeah.
- Res Would anyone else like to see their friends or family more? No?
- Azi I see them enough.
- Res You see them enough.
- Adom Yeah.
- Res What about other people? Would you like to have any more people in your lives?
- Adom Yeah.
- Res Yeah? How come [REDACTED]?
- Adom Because ((hesitation)) I think that I'm old enough to not just look after myself, to look after other people as well.

Would like to spend more time with his brothers

Independent?
Self-sufficient

Would like to look after others

- Res Ah. So you're saying you want younger brothers and sisters?
- Adom No, I've got younger brothers. A nephew or niece would be good.
- Res Uh huh. So you want to have the responsibility of looking after them, to help out. Would anyone else like more family or friends? Or to get on with your family or friends better? Everyone's nodding no. Does everyone live with Mum and Dad?
- Ayo Only my mum.
- Adom Just Mum.
- Res Just your mum, just your mum. Mum and Dad?
- Atu Mum.
- Adegoke Mum.
- Res Yeah, so, two Mum, just Dad. Do you see your dad, if you don't mind me asking...?
- Ayo He visits sometimes, but I've talked to my dad once, yeah.
- Res Aw.
- Ayo Yeah it's mum and dad's choice.
- Res Both of their choice.
- Ayo He visits ((hesitation)) quite a few times. But I've seen dad once.
- Res Mm-hm
- Ayo I don't think that I will ever really hang out with him that much. Because he's usually busy with work and stuff.

Most adolescents live with their mum only

Distant relationship with his dad

Met his dad once

His dad is busy with work

- Res Uh huh. He's busy with work. [REDACTED], do you know why...? No. Ok. Can I just ask, are other parents really busy with work, or..?
- Azi Yeah but that's standard that parents work for everyone.
- Ayo Normal, like [REDACTED] said they need to work, like all adults need to work to pay rent, bills, food and yeah all that. Mature / realistic attitude regarding parents busy lives / work commitments
- Res Just normal, just like your friends' parents. Ok, so next I just want to talk about school or college. Can you tell me how you feel about your school, academy or college?
- Azi It's good. Likes school / positive attitude about school
- Res Good? Why?
- Azi College provides education.
- Res Mm education. Anything else you can say?
- Azi Socially.
- Res Socially good. So...
- Azi Yeah, I get on with everybody, I'm easy going, even with people who aren't my people, we alright and yeah, I'm the class clown but I get good grades!
- Res Uh huh What about your teachers [REDACTED]?
- Azi Yeah, the teachers, they're brilliant.
- Res Why? Teachers are role models Teachers are encouraging regarding their future
- Azi The teachers are fair, encouraging and help us improve and want us to make something of ourselves, want us to go somewhere, be someone. I go after-school clubs, sometimes, it depends if it applies to me and teachers run them.

Res Mm-hm so they're very supportive [REDACTED]. What about everyone else? How do you feel about school?

Ayo I'm going to do GCSEs in a couple of months. Some teachers are really good. Yeah, it's a great way to meet and learn more about the world ((hesitation)) because we have things like citizenship day, where we talk more about, go off the topic of school stuff but talk more about the world around us, and how we can live better lives.

School provide different opportunities i.e. citizenship day

Discusses improving his life

Res Oh, Ok. What about everyone else, how do you feel about school?

Adegoke It's Ok.

Res It's Ok.

Abidemi Some people don't talk to you, some teachers don't talk too much, you have to get over it.

Res What do you mean some teachers don't talk to you? If they don't teach you, or..?

Abidemi You don't talk to those kind of teachers, if you don't like their kind of teaching you just have to get over it. Because you need them for your future.

Res And how do you get along with the other people in your class?

Mature attitude. Focussed on future

Ayo Fine.

Res No problems with people?

Ayo Except some people who do it on purpose just to try and disrupt the class and be funny.

Res Like attention-seekers...?

Ayo Yeah.

Res Mm-hm..but no bullying or anything like that?

Ayo No.

Res Do you all go to mixed schools?

Ayo Yeah.

Adom Yeah.

Figure A. An Example of Final Coding for Study 2 (Focus Groups)

Initial Codes	Final Codes	Final Subtheme	Final Theme
<ul style="list-style-type: none"> • Spends some time with his family in free time 	<ul style="list-style-type: none"> • Condition does not appear to impact their social lives 	<p>Normalising and acceptance</p>	<p>Coping with SCD</p>
<ul style="list-style-type: none"> • Makes own choices regarding free time • Enjoys socialising with friends outside of school • Socialising with friends provides fun 			

Initial Codes

- Most adolescents live with their mum only
- Distant relationship with his dad
- Met his dad once
- His dad is busy with work
- Siblings are busy
- Family live in Nigeria / outside of London
- Smaller support network (family)

- Generally family and friends are important people
- Appreciates his mum. Acknowledges her importance
- Family / close friends provide stability
- Generally, important people in their lives makes them feel better / tries to help them

- Friends moved away from London
- Smaller support network (friends)

Final Codes

- Relationships with family
- Importance of support of mum / important people to adolescents
- Smaller support networks

Final Subtheme

Emotion-focused coping

Final Theme

Coping with SCD

Initial Codes	Final Codes	Final Subtheme	Final Theme
<ul style="list-style-type: none">• Independent? Self-sufficient• Would like to look after others	<ul style="list-style-type: none">• Becoming more independent	Self-management of SCD	Coping with SCD
<ul style="list-style-type: none">• Makes own choices to do with sports• Has the physique/height for basketball	<ul style="list-style-type: none">• Own choice to participate in sports• Has the physique/ability to participate in particular sports		Influences on health behaviours

Initial Codes

- Money is not an issue
- Low income / finances are not a barrier
- Free recreational activities

- Adolescents consider their goals
- Future
- Ambitious / motivated
- Discusses improving his life
- Sports participation requires them to be disciplined

- Likes school / positive about school
- Important people are motivating
- Teachers are encouraging regarding their future
- Teachers / school help to obtain qualifications / sponsors e.g. Nike
- Coaches/school provide opportunities (like work experience or citizenship day)
- Coaches are more encouraging than parents (sports related)

- Parents / brothers are role models
- Mature / realistic attitude regarding parents busy lives / work commitments

Final Codes

- Self-motivated and ambitious
- Aspirations for a better lifestyle
- Understanding of families financial circumstances

- Role models for education and future
- Differences between individuals at school and family members (mainly parents)

Final Theme

Education
and beyond

Appendix B

Information sheets, assent/consent forms, demographic information sheet and debriefing forms (Studies 1 and 2)



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Principle Investigator: [REDACTED]

Consultant Paediatrician

Telephone: [REDACTED]

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Participant Information Sheet for Young People (5-12 year olds)

Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

You are being asked to take part in our research study. I will explain to you why we are doing this. Then you can make up your mind if you would like to help me with it. Please talk to your mum, dad or carer if you have any questions. You can also ask me.

I am happy to read through this information sheet with you or you may like your mum, dad or carer to read it with you.

You will be asked to write your name on a sheet to say that you are happy to take part in the research study.

What is research?

Research is a way we try to find out the answers to questions and how you see things.

Why is this project being done?

We want to learn more about how happy you are with different parts of your life, like school, family and friends. We also want to learn about things like what foods you eat and what kinds of activities you do when you are at school and at home.

If you have sickle cell disease we also want to learn about how your diet and activities change when you are in hospital or when you are in pain.

This research is being carried out as part of my University studies.

Why have I been asked to take part?

All children with sickle cell disease at the [REDACTED] and their brother or sister have been asked to take part in the research.

Did anyone else check the research is OK to do?

Before any research is allowed to happen, it has to be checked by a group of people called a Research Ethics Committee. They make sure that the research is fair. This research has been checked by the NRES Committee London – Hampstead. It has also been checked by Middlesex University where I am studying.

Do I have to take part?

You don't have to take part if you don't want to. It is your choice. I will describe what you have to do. You can change your mind at anytime and stop taking part.

What will happen to me if I take part in the research?

We want you to draw some pictures and answer some questions. It will take about 45 minutes. You can have a break, snack or drink whenever you like.

I'll ask you to draw and answer the questions, on the ground floor at the [REDACTED]. Or if you, or your mum, dad or carer prefers I can visit your home and you can take part in the research then.

Might anything about the research upset me?

If you feel upset or worried please tell me and we will stop straightaway.

Will joining in help me?

It will give you something to do while you wait for your appointment. It is a chance to draw and talk about how you feel about family, friends, school and your health.

What if there is a problem?

If you have any problems during the interview, I will stop the interview straightaway. Then, I will suggest you talk to Dr [REDACTED], one of the sickle cell nurses or a member of the psychology team. I can also get your mum, dad or carer if you would like me to. It is your choice who you decide to talk to.

If you have any worries after the interview you can again talk to Dr [REDACTED], one of the sickle cell nurses or a member of the psychology team at any time. If you are not sure who to talk to you can always contact me first.

We can support you for as long as you need, so please just talk to us about any concerns you have. Contact details for Dr [REDACTED] and me are at the [top of page 1](#) and details for the [psychology team](#) are [at the end](#) of this page. Sickle cell nurses work at the clinics you attend, or if you want I can ask them to meet you at any time.

We will also not use any information you do not want us to use like your drawings or what you said to me.

Will my taking part in the study be kept confidential?

Anything you say will be private. I will only talk to doctor if I was very worried about you. But I would make sure you were ok with this first.

Support (see page 1 for contact details)

- For advice about if you should join in the research please talk to Dr [REDACTED].
- If you feel upset, worried or have any other problems you can talk to a member of the psychology team on [REDACTED].
- You can also phone ChildLine on 0808 11 11 where you can talk to a counsellor straightaway about anything that is upsetting or bothering you.

Thank you for taking the time to read the information sheet!



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Institutional Contact Details:

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Date: 18/05/2015

Principle Investigator: [REDACTED]

Consultant Paediatrician

Telephone: [REDACTED]

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Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

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Participant Information Sheet for Young People (13-18 year olds)

Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

You and one or more of your healthy brothers or sisters are being invited to take part in our research. Before you decide to take part, it is important for you to understand why the research is being done and what it will involve. Please take your time to read the information below carefully and discuss it with others if you want to. Please ask if there is anything that is not clear or if you would like more information. If you would like I, (Christina Constantinou) can read through this information sheet with you?

You will be given a copy of the information sheet and asked to sign a consent form before taking part in the research.

What is the purpose of the research?

Health-Related Quality of Life (HRQL) is important especially for young people with Sickle Cell Disease (SCD) and their brothers and sisters. We are keen to learn more about how young people with SCD and also some siblings of young people with SCD who do not have the condition see their HRQL. In other words, to find out how happy you feel with different parts of your life such as school, friendships and health.

Also, we want to ask you about your health behaviours such as the kinds of things you eat and drink and more risky health behaviours such as drinking alcohol or caffeinated drinks and smoking and substance use. Your health behaviours may or may not affect your HRQL and your HRQL may or may not affect your health behaviours. This is what we want to find out.

This study is being carried out as part of my PhD.

Why have I been invited?

All young people who are 13-18 years old with SCD and who attend the [REDACTED] [REDACTED] have been invited to take part. Also, all brothers and sister from the same family who are aged 13-18 years old have been invited to take part.

Do I have to take part?

It is up to you to decide whether or not you want to take part. I will describe the study and go through the information sheet with you, if you want. You can withdraw from the study at any time up to publication, without giving a reason. This will have no affect on the care you receive from the hospital.

Expenses.

Parents or carers who accompany their child to the [REDACTED] will receive up to a maximum of £10.00 for public transport costs or up to a maximum of £10.00 if they travel by car when their child's participation occurs on a day when the child does not have an outpatient clinic appointment. Young people who are 13-18 years old will receive £20.00 to cover their travel costs and to acknowledge the time demands of participating in a focus group.

What will happen to me if I take part?

You will participate in a digitally recorded focus group with up to another four people of the same sex. Nobody from the same family will be in the same focus groups. The discussion in the focus groups will cover your experiences of HRQL, health behaviours and risky health behaviours. The focus group should last approximately one hour and will be digitally recorded and later typed up. If you want to participate I will contact your parent by telephone or email to arrange a date and time that is convenient for both of you. You and your parent will be asked to provide written consent on the day of the focus group; either of you can change your mind with no affect on the care you receive at the hospital. If you are aged 16 years or older only you will need to provide written consent. The focus group will take place here in the outpatient clinic at the [REDACTED]. The focus group will not be during school time.

There is also a brief form containing questions about background information such as your age for your parent to complete, or for to you complete if you are aged 16 years or older.

What are the possible disadvantages of taking part?

There should be no risk for you at all. If you feel upset or anxious please tell me and we will stop the focus group straightaway. Participation in this research is entirely voluntary.

What are the possible benefits to taking part?

It is a chance to think about how you feel about your HRQL so things like your family, friends, health and school and your health behaviours. Also, it is an opportunity to meet with your peers who may be experiencing the same positive or negative feelings as you.

What if there is a problem?

If you have any problems please talk to me. Or you can talk to your parent or carer or a member of staff at the [REDACTED].

If you decide to take part you may withdraw at any time up to publication, without giving a reason. Any contribution you have made during the focus group will not be typed and the digital recording will be destroyed.

Will my taking part in the study be kept confidential?

Yes. The information you provide will remain completely confidential. The tapes of the focus groups will be destroyed once they are transcribed. Anonymous transcripts will be kept on a password protected computer.

Only in exceptional circumstances I may become concerned about your life or well-being is at risk would I speak to a consultant about what you have discussed with me. This is very unlikely and I would tell you before I speak to a consultant.

I, (Christina Constantinou) or Dr [REDACTED] may look through your medical records only where it is relevant. We will only do this if you have SCD.

Who has reviewed the study?

This study has been reviewed and approved by the NRES Committee London – Hampstead and the Middlesex University Psychology Department’s Ethics Committee.

Further information and contact details (see previous page)

- For general information regarding the research please contact Christina Constantinou.
- For specific information about this research project please contact Christina Constantinou or Dr Nicky Payne.
- For advice as to whether you should participate please contact Dr [REDACTED].
- For independent information about participating in research please contact Patient Advice and Liaison Service (PALS) on [REDACTED] or email pals@[REDACTED].
- To make a complaint, please contact Dr [REDACTED] or Dr Nicky Payne.

Thank you for taking the time to read the information sheet.



Chief Investigator/Researcher Name: Christina Constantinou

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Date: 18/05/2015

Principle Investigator: [REDACTED]

Consultant Paediatrician

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Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Participant Information Sheet for Parents/Carers

Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Your child/children with Sickle Cell Disease (SCD) and one or more of any healthy children you have are being invited to take part in our research. Before you decide we would like you to understand why the research is being done and what it will involve for your child/children. Please take your time to read the following information carefully, and discuss it with others if you wish. Please ask if there is anything that is not clear or if you would like more information. Take your time to decide whether or not you would like your child/children to take part. If you do decide that your child/children will participate in the research study then I, (Christina Constantinou) will read through the age-appropriate information sheet with them. You will be given another copy of the information sheet and asked to sign consent forms prior to your child/children's participation in the research.

What is the purpose of the research?

Health-Related Quality of Life (HRQL) is important for us to consider when thinking about treating chronic illnesses and also for the general well-being of all children. We are keen to learn more about how children with SCD and healthy children see their HRQL. In other words, to find out how happy they feel with different parts of their life such as school, friendships and their health.

Also, we want to find out about their health behaviours such as the kinds of things they eat and drink and the amount and kinds of activities they take part in. For children 13 years and older we are also interested in more risky behaviour such as whether they drink alcohol or caffeinated drinks and have ever tried smoking or other substances. We want to know whether health

behaviours may affect their HRQL which includes their physical health and psychological well-being.

This study is being carried out as part of my doctoral research (i.e. PhD).

Why have I been invited?

All children who are 5-18 years old with SCD attending the [REDACTED] have been invited to take part. Also, all healthy children from the same family who are aged 5-18 years old have been invited to take part.

Do my children have to take part?

It is up to you to decide whether or not you want your child/children to take part. All of your children with SCD and your healthy children can take part or just your child/children with SCD, it is your decision. I have described the study in the form and would be happy to describe it to you in person at the hospital when you come in for your child's appointment if you wish. Your child/children can withdraw from the study at any time up to publication, without giving a reason. This will have no affect on the care your child receives from the hospital.

Expenses.

Parents or carers who accompany their child to the [REDACTED] will receive up to a maximum of **£10.00** for public transport costs or up to a maximum of £10.00 if they travel by car when their child's participation occurs on a day when the child does not have an outpatient clinic appointment. Young people who are 13-18 years old will receive **£20.00** to cover their travel costs and to acknowledge the time demands of participating in a focus group.

What will happen to my child if they take part?

If your child/children is/are aged 5 - 12

Your child/children will be asked to take part in drawing tasks and an interview which will be digitally recorded and later transcribed (i.e. typed up). The drawing and questions during the interview will focus on their HRQL and health behaviours.

The drawing and interview should approximately 45 minutes to complete with each child, here in the outpatient clinic at the [REDACTED] or if you prefer the drawing and interview can take place at your home. Your child with SCD may participate when they attend their outpatient clinic appointment at the hospital as there will be sufficient time while they wait to be seen by the consultant and the nursing staff. If they have a brother or sister aged 5-12 who does not have SCD and will also participate in the research, he/she would participate at a later date; I will contact you by telephone or email to arrange a convenient day and time to visit your home. If their brother or sister decides not to participate when I visit your home this is completely their decision. You and your child will be asked to provide written consent on the day of the drawing and interview, either of you can change your mind with no affect on the care they receive at the hospital. You must provide written consent for your children's participation in the research. You may find it easier for me to visit your home if both of your children are going to participate.

If your child/children is/are aged 13 - 18

Your child/children will be asked to participate in a digitally recorded focus group with up to another four children of the same sex. Any children who are from the same family will not be in the same focus groups and therefore they will be arranged on different days. The discussion in the focus groups will cover your children's experiences of HRQL, their health behaviours and risky health behaviours. The focus group should last approximately one hour. It will be digitally recorded and later transcribed. It will take place here in the outpatient clinic at the [REDACTED] at a date and time that is convenient to you and your child. If your child or children wish to participate I will contact you by telephone or email to arrange a convenient day and time. You and your child will be asked to provide written consent on the day of the focus group; either of you can change your mind with no affect on the care they receive at the hospital. If your child is aged 16 years or older you will not need to provide written consent on their behalf, their consent is sufficient. The focus groups will not be during school time and will not interfere with your children's studies.

If you have children in each of these two age groups, one or more may participate in the drawings and interviews and one or more may participate in the focus group.

There is also a brief form containing questions about background information (such as "What is your occupation / job title?") for you to complete.

What are the possible disadvantages of taking part?

There are no risks involved if your children take part in this research. Whether they chose to do so or not will not influence your child's care at the [REDACTED]. If during participation your child becomes upset for any reason or wishes to stop we will stop immediately.

What are the possible benefits to taking part?

This may give your children a chance to discuss how they feel about different parts of their life. For children aged 13-18 it is also an opportunity for them to meet with their peers who may be experiencing the same positive or negative feelings as them.

What if there is a problem?

If you have any problems please talk to me or a member of staff at the [REDACTED]. If your child has a problem they can immediately speak with me or a member of staff from the Sickle Cell and Thalassaemia Service or Paediatric Psychology Department. If you decide your children may take part you may still withdraw at any time up to publication, without giving a reason. Any digital recordings, interview transcripts or drawings related to your children will be destroyed.

Will my child's taking part in the study be kept confidential?

Yes. The information your children provide will remain completely confidential. The tapes of the interview or focus groups will be destroyed once they are transcribed. Anonymous transcripts will be kept on a password protected computer. Anonymous drawings and consent forms will also be kept securely.

Only in exceptional circumstances where I may become concerned that your child's life or well-being is at risk would I speak to a consultant about what your child discussed with me. This is very unlikely and I would tell your child before I speak to a consultant.

I, (Christina Constantinou) or Dr [REDACTED] may look through your child's medical records only where it is relevant. This will only be for children or adolescents with SCD.

Who has reviewed the study?

This study has been reviewed and approved by the NRES Committee London – Hampstead and the Middlesex University Psychology Department's Ethics Committee.

Further information and contact details (see previous page)

- For general information regarding the research please contact Christina Constantinou.
- For specific information about this research project please contact Christina Constantinou or Dr Nicky Payne.
- For advice as to whether you should participate please contact Dr [REDACTED].
- For independent information about participating in research please contact Patient Advice and Liaison Service (PALS) on [REDACTED] or email [REDACTED].
- To make a complaint, please contact Dr [REDACTED] or Dr Nicky Payne.

Thank you for taking the time to read the participant information sheet.



Written Assent form for Young People (5-12 year olds)

(to be completed by the child and their parent/guardian)

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Chief Investigator/Researcher: Miss Christina Constantinou

Participant Identification Number:

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

- I have read the information sheet with the researcher or the researcher has read the information sheet to me. This is dated **23/09/2014** (Version IF 1B) for the above study. Yes/No
- The researcher has talked to me about the study. I have been able to ask the researcher questions. The researcher has answered my questions. Yes/No
- I understand and am happy to take part in drawing tasks and a recorded interview. Yes/No
- I am happy to let the researcher keep my two drawings or make colour photocopies of my two drawings. Yes/No
- Do you understand it's OK to stop taking part at any time? 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 can write your name below...

Your name

Date

Name of person taking consent
Thank you for your help.

Signature

Date



Written Informed Consent for Parents or Carer's

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Participant Identification Number:

Please initial box

- I have read and understood the information sheet dated **18/05/2015** (**Version IF 2**) for the above study.
- I understand that data collected during the research will not be identifiable, and that my child has the right to withdraw from the study at any time, up to publication without any obligation to explain their reasons for doing so.
- I consent that some relevant sections of my child's medical notes may be looked at by responsible individuals from the [REDACTED], from regulatory authorities or from the NHS Trust, where it is relevant to taking part in the study.
- I have been given contact details for the researcher in the participant information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- I consent that the researcher can ask my child about their health-related quality of life and health behaviours during drawing tasks and a recorded interview and that I will complete a demographic information sheet.
- I consent that the researcher can keep my child's original two drawings, or colour photocopy them and return the two original drawings to my child.
- I further understand that the anonymous data they provide may be used for analysis and subsequent publication, and provide my consent that this might occur. I also understand that all recordings will be destroyed once transcribed.

Name of participant

Date

Signature

Name of person taking consent

Date

Signature

To the participants: Data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures. Although this would happen in strict confidentiality, please tick here if you do not wish your data to be included in audits: _____



Written Informed Consent for Young People (13-18 year olds)

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator/Advisor (*only for students*): Dr [REDACTED]

Participant Identification Number:

Please initial box

- I have read and understood the information sheet dated **18/05/2015 (Version IF 3)** for the above study.
- I understand that my participation is entirely voluntary, the data collected during the research will not be identifiable, and I have the right to withdraw from the study at any time without any obligation to explain my reasons for doing so.
- I consent that some relevant sections of any of my medical notes may be looked at by responsible individuals from the [REDACTED], from regulatory authorities or from the NHS Trust, where it is relevant to my taking part in the study.
- I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- I consent that the researcher can ask me about my health-related quality of life, Health behaviours and risky behaviours during a recorded focus group with my peers. I will complete a demographic information sheet (*only applicable if aged 16-18 years*).
- I further understand that the anonymous data I provide may be used for analysis and subsequent publication, and provide my consent that this might occur. I also understand that recordings will be destroyed once transcribed.

Name of participant

Date

Signature

Name of person taking consent

Date

Signature

To the participants: Data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures. Although this would happen in strict confidentiality, please tick here if you do not wish your data to be included in audits: _____



Written Informed Consent for Parents or Carer's

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator/Advisor (*only for students*): Dr [REDACTED]

Participant Identification Number:

Please initial box

• I have read and understood the information sheet dated **18/05/2015** (**Version IF 2B**) for the above study.

• I understand that my child's participation is entirely voluntary, the data collected during the research will not be identifiable, and that they have the right to withdraw from the study at any time, up to publication without any obligation to explain their reasons for doing so.

• I consent that some relevant sections of any of my child's medical notes may be looked at by responsible individuals from the [REDACTED], from regulatory authorities or from the NHS Trust, where it is relevant to their taking part in the study.

• I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.

• I consent that the researcher can ask my child about their health-related quality of life, health behaviours and risky behaviours during a recorded focus group which will also involve their peers and that I will complete a demographic information sheet.

• I further understand that the anonymous data they provide may be used for Analysis and subsequent publication, and provide my consent that this might occur. I also understand that recordings will be destroyed once transcribed.

Name of participant

Date

Signature

Name of person taking consent

Date

Signature

To the participants: Data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures. Although this would happen in strict confidentiality, please tick here if you do not wish your data to be included in audits: _____



Written Informed Consent for Young People (13-18 year olds)

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator/Advisor (*only for students*): Dr [REDACTED]

Participant Identification Number:

Please initial box

- I have read and understood the information sheet dated **18/05/2015 (Version IF 3)** for the above study.
- I understand that my participation is entirely voluntary, the data collected during the research will not be identifiable, and I have the right to withdraw from the study at any time without any obligation to explain my reasons for doing so.
- I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- I consent that the researcher can ask me about my health-related quality of life, health behaviours and risky behaviours during a recorded focus group with my peers. I will complete a demographic information sheet (*only applicable if aged 16-18 years*).
- I further understand that the anonymous data I provide may be used for analysis and subsequent publication, and provide my consent that this might occur. I also understand that recordings will be destroyed once transcribed.

Name of participant

Date

Signature

Name of person taking consent

Date

Signature

To the participants: Data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures. Although this would happen in strict confidentiality, please tick here if you do not wish your data to be included in audits: _____



Written Informed Consent for Parents or Carer's

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator/Advisor (*only for students*): Dr [REDACTED]

Participant Identification Number:

Please initial box

• I have read and understood the information sheet dated **18/05/2015** (Version IF 2B) for the above study.

• I understand that my child's participation is entirely voluntary, the data collected during the research will not be identifiable, and that they have the right to withdraw from the study at any time, up to publication without any obligation to explain their reasons for doing so.

• I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.

• I consent that the researcher can ask my child about their health-related quality of life, health behaviours and risky behaviours during a recorded focus group which will also involve their peers and that I will complete a demographic information sheet.

• I further understand that the anonymous data they provide may be used for analysis and subsequent publication, and provide my consent that this might occur. I also understand that all recordings will be destroyed once transcribed.

Name of participant

Date

Signature

Name of person taking consent

Date

Signature

To the participants: Data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures. Although this would happen in strict confidentiality, please tick here if you do not wish your data to be included in audits: _____

6. What is your highest level of educational attainment? (Please circle as appropriate):

Higher Degree	Degree	Foundation Degree/Diploma
<i>A Levels or equivalent</i>	<i>GCSEs or equivalent</i>	Not Applicable

Information Regarding the Child:

7. Is your child (please circle as appropriate): Male Female

8. Age _____ years old.

9. What is their ethnic group (please circle as appropriate):

White	Mixed <i>(please specify)</i>	Asian <i>(please specify)</i>	Black Caribbean
_____	_____	_____	_____
Black African <i>(please specify)</i>	Black Other <i>(please specify)</i>		Other Ethnic Group
_____	_____		_____

10. Is your child currently in (please circle as appropriate):

School / Full-time Education	College / Full-time	College / Part-time
Training <i>(please specify)</i>	Employment <i>(please specify)</i>	Unemployed
_____	_____	

11. What type of sickle cell does your child have (please circle as appropriate):

Sickle Cell Anaemia / Hb SS	Hb SC	Hb Sbeta+
Hb Sbeta ⁰	Other <i>(please specify)</i>	Not Applicable

Thank you for taking the time to fill in the demographic information sheet.



Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept.,
Middlesex University,
Town Hall, The Burroughs, Hendon,
London , NW4 4BT
Email: c.constantinou@mdx.ac.uk

Date: 02/07/2014

Principle Investigator: [REDACTED]

Consultant Paediatrician

Telephone: [REDACTED]

Email: [REDACTED]

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Debriefing Form for Young People (5-12 year olds)

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Points to discuss with children once they have completed their drawings and semi-structured interview.

- a) Your help in my research has finished.
- b) Thank you very much for helping with my research.
- c) You may have some worries or questions for me about the research or anything you have not understood.
- d) You may want to ask your parent or carer some questions. Or you may prefer to ask a member of staff here at the [REDACTED] some questions.
- e) Would you like to ask me anything else? Or talk to a member of staff here at the [REDACTED] about anything else?
- f) Thank you again!



Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept.,
Middlesex University,
Town Hall, The Burroughs, Hendon,
London , NW4 4BT
Email: c.constantinou@mdx.ac.uk

Date: 02/07/2014

Principle Investigator: [REDACTED]

Consultant Paediatrician

Telephone: [REDACTED]

Email: [REDACTED]

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Debriefing Form for Young People (13-18 year olds)

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Your participation in the research has finished. You may have some questions for either me or another member of staff.

About the research....

- a) Past research shows that adolescents with Sickle Cell Disease (SCD) report a lower HRQL compared to their healthy peers.
- b) Questionnaire based research was carried out with adolescents with SCD at the [REDACTED]. It was found that adolescents with SCD rate their quality of life as no different from their healthy peers. Therefore, we wanted to look at this in more detail by speaking to adolescents, and also some of their healthy brothers and sisters.
- c) There has been limited research that has looked at adolescents views of their health behaviours including risky health behaviours in the United Kingdom and how this may influence their HRQL. This applies to adolescents with SCD and their brothers and sisters who do not have the condition.

Here are some points to consider.....

- a) Please note that all recordings, focus group transcripts and demographic information will be confidential to the researcher and her supervisors at Middlesex University. All results will be published anonymously.
- b) If you have any concerns, you can talk to me or a member of staff at the [REDACTED]. This will all be in confidence.
- c) If you would like to receive a brief leaflet outlining the results of the study once it is completed, please provide me with an email or postal address I can forward them too. Please understand that this may take some time.

Thank you for participating in the study.



Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept.,
Middlesex University,
Town Hall, The Burroughs, Hendon,
London , NW4 4BT

Email: c.constantinou@mdx.ac.uk

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Date: 02/07/2014

Principle Investigator: [REDACTED]

Consultant Paediatrician

[REDACTED]

[REDACTED]

Telephone: [REDACTED]

Email: [REDACTED]

Debriefing Form for Parents/Carer's

Title of Study: Health-Related Quality of Life (HRQL) and Health Behaviours in Paediatric Sickle Cell Disease (SCD) Populations

Your child's or children's participation in the research has finished. You may have some questions for either me or another member of staff.

About the research.....

a) Past research shows that children and adolescents with Sickle Cell Disease (SCD) report a lower HRQL compared to their healthy peers.

b) Questionnaire based research was carried out with children and adolescents with SCD at the [REDACTED]. It was found that children and adolescents with SCD rate their quality of life as no different from their healthy peers. Therefore, we wanted to look at this in more detail by speaking to children and adolescents, and also some of their healthy siblings.

c) There has been limited research that has looked at children's or adolescents' views of their health behaviours and how this may influence their HRQL. This applies to children and adolescents with SCD and siblings who do not have the condition. Also, no research has been conducted in the United Kingdom exploring risky health behaviours in adolescents' with SCD or their healthy siblings.

Here are some points to consider.....

a) Please note that all drawings, interview and focus group transcripts and demographic information will be confidential to the researcher and her supervisors at Middlesex University. All results will be published anonymously.

b) If you or your children have any concerns, you can talk to me or a member of staff at the [REDACTED]. This will all be in confidence.

c) If you or your children would like to receive a brief leaflet outlining the results of the study once it is completed, please provide me with an email or postal address I can forward them too. Please understand that this may take some time.

Thank you for participating in the study

Appendix C

Favourable letters from the research ethics committee reference number 14/LO/1548
(Studies 1 and 2)



Health Research Authority

National Research Ethics Service

NRES Committee London - Hampstead

Barlow House
3rd Floor 4 Minshull Street
Manchester
M1 3DZ

Telephone: 0161 625 7832

Fax: 0161 625 7299

15 September 2014

Miss Christina Constantinou



Dear Miss Constantinou

Study title:	An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)
REC reference:	14/LO/1548
Protocol number:	N/A
IRAS project ID:	157322

The Research Ethics Committee reviewed the above application at the meeting held on 10 September 2014. Thank you for attending to discuss the application.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details, unless you expressly withhold permission to do so. Publication will be no earlier than three months from the date of this favourable opinion letter. Should you wish to provide a substitute contact point, require further information, or wish to withhold permission to publish, please contact the REC Manager Dr Ashley Totenhofer, nrescommittee.london-hampstead@nhs.net.

Ethical opinion

The members of the Committee present gave a favourable ethical opinion of the above research on the basis described in the application form, protocol and supporting documentation, subject to the conditions specified below.

Conditions of the favourable opinion

The favourable opinion is subject to the following conditions being met prior to the start of the study.

- Please send a copy of the Lone Worker Policy that would be followed when making home visits.
- On page 2 of the Participant Information Sheet for Young People aged 5 -12, please add further information about what would happen if a participant was upset. Please explain exactly what would be done to make sure a participant was okay and what sources of support would be available.
- Please revise the address for the REC given on page 21 of the protocol. The correct address is:

HRA Centre
 Manchester 3rd Floor,
 Barlow House 4
 Minshull Street
 Manchester
 M1 3DZ

You should notify the REC in writing once all conditions have been met (except for site approvals from host organisations) and provide copies of any revised documentation with updated version numbers. The REC will acknowledge receipt and provide a final list of the approved documentation for the study, which can be made available to host organisations to facilitate their permission for the study. Failure to provide the final versions to the REC may cause delay in obtaining permissions.

Management permission or approval must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission (“R&D approval”) should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements.

Guidance on applying for NHS permission for research is available in the Integrated Research Application System or at <http://www.rdforum.nhs.uk>.

Where a NHS organisation’s role in the study is limited to identifying and referring potential participants to research sites (“participant identification centre”), guidance should be sought from the R&D office on the information it requires to give permission for this activity.

For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.

Sponsors are not required to notify the Committee of approvals from host organisations.

Registration of Clinical Trials

All clinical trials (defined as the first four categories on question 2 of the IRAS filter page) must be registered on a publically accessible database within 6 weeks of recruitment of the first participant (for medical device studies, within the timeline determined by the current registration and publication trees).

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to contest the need for registration they should contact Catherine Blewett (catherineblewett@nhs.net), the HRA does not, however, expect exceptions to be made. Guidance on where to register is provided within IRAS.

It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).

Ethical review of research sites

NHS Sites

The favourable opinion applies to all NHS sites taking part in the study taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see “Conditions of the favourable opinion” below).

Summary of discussion at the meeting

Social or scientific value; scientific design and conduct of the study

The Committee discussed that SCD can affect all organs in the body and can have a devastating effect. Patients can end up addicted to narcotics; consequently research in this field is very important and potentially valuable.

The Committee had noted that if a participant chose to withdraw from the research their data would also be withdrawn. It was thought that this was a waste of data but it was agreed that it is the researcher’s choice.

The Committee queried how the study would work if the patient did not have a healthy sibling.

The Committee noted that smoking and drinking alcohol are not the only risky behaviours that young people engage in.

The Committee were aware that there is a lot of existing literature relating to children with SCD and quality of life. The Committee asked what could be learnt by carrying out this study. You explained that this study would incorporate gap theory and utilise the WHO definition of quality of life; something which has not been done in previous research.

The Committee were of the opinion that this was an interesting idea but had agreed that there were a lot of variables and asked what new information could be gained from the study. You advised that this would be the first study to use a comparison group of healthy siblings who would have their own set of issues. Looking at risky behaviours would also be novel.

The Committee advised that it may be that these areas had already been researched so they would appreciate confirmation of this. You advised that work had not been done which utilised the multi-dimensional health related quality of life concept.

Favourable risk benefit ratio; anticipated benefit/risks for research participants (present and future)

The Committee agreed that this was a low risk study. It may be beneficial for patients to give more thought to their behaviour.

The Committee asked for more information about visiting families in their homes. You explained that it would be more convenient for siblings to be visited at home. It is common to do such visits and there have been no incidences in the past. A time would be arranged and it would be clear to participants that they could withdraw from the study.

The Committee asked you whether you would adhere to the Trust’s lone worker policy.

You advised that you have carried out home visits in the past and would follow the policy.

Care and protection of research participants; respect for potential and enrolled participants' welfare and dignity

The Committee noted that a psychologist would be available to help a distressed patient and there was a de-briefing sheet.

The Committee discussed the importance of there being a pleasant, quiet space available to carry out the research.

The Committee asked you for further information about how you would manage the focus groups. You advised that these patients attend clinic every 6 months. It is in clinic that you would approach the patients and introduce the research. If they were willing to participate you would obtain their contact details. The focus groups would be run on weekends or during half-term so that children did not have to miss school and they would be reimbursed for their travel expenses.

The Committee advised that focus groups are quite a sophisticated concept, especially for young people who may not be at ease or understand the boundaries. The Committee queried how you would help the participants to understand the ground rules. You explained that you had run focus groups in the past involving young people with chronic conditions. The young people would be partnered up and would tell each other their name, school year and something interesting about themselves. Their partner would then feed this back to the group. You described how you would set the boundaries, including that the discussion in the group would remain confidential.

The Committee asked about those young people who might be nervous, anxious or upset. You explained that there would be a mental health worker and paediatric psychologist in the hospital. You would also report this to your line manager who would take the necessary steps.

Informed consent process and the adequacy and completeness of participant information

The Committee asked whether the suitability of the Participant Information Sheet for 5-12 year olds had been tested. You advised that you had tested the suitability of all Information Sheets, the interview schedule and focus group schedule. So far they had been tested by healthy individuals but they would also be tested by patients with SCD. The Committee advised that they had some further points to make and these would be sent in writing.

Other ethical issues were raised and resolved in preliminary discussion before your attendance at the meeting.

Approved documents

The documents reviewed and approved at the meeting were:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Indemnity insurance cover letter from Middlesex University]		12 September 2013
Interview schedules or topic guides for participants [Semi-Structured Interview Schedule for Young People (5-12 year olds)]	IS 1	02 July 2014
Interview schedules or topic guides for participants [Focus Group Schedule for Young People (13 to 18 year olds)]	FGS 2	02 July 2014
Letter from sponsor [Middlesex University]		08 August 2014

Other [CV. Olga van den Akker]	CV. Ovd A	25 July 2014
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Other [Debriefing Form for Young People (5-12 year olds)]	DF 1	02 July 2014
Other [CV. ██████████ (principle investigator)]	CV.BI	22 July 2014
Other [Debriefing Form for Parents/Carer's]	DF 2	02 July 2014
Other [Debriefing Form for Young People (13-18 year olds)]	DF 3	02 July 2014
Other [Demographic Information Sheet]	DIM 1	02 July 2014
Participant consent form [Young People (13-18 year olds)]	C4	02 July 2014
Participant consent form [Young People (5-12 year olds)]	C1	02 July 2014
Participant consent form [Parents or Carer's]	C7	02 July 2014
Participant consent form [Young People (13-18 year olds)]	C5	02 July 2014
Participant consent form [Parents or Carer's]	C2	02 July 2014
Participant consent form [Parents or Carer's]	C3	02 July 2014
Participant consent form [Parents or Carer's]	C6	02 July 2014
Participant information sheet (PIS) [Young People (13-18 year olds)]	IF 3	02 July 2014
Participant information sheet (PIS) [Young People (5-12 year olds)]	IF 1	02 July 2014
Participant information sheet (PIS) [Parents/Carers]	IF 2	02 July 2014
REC Application Form [REC_Form_11082014]		11 August 2014
Research protocol or project proposal [Protocol]	1A	02 July 2014
Summary CV for Chief Investigator (CI) [Christina Constantinou]	CV.CC	11 August 2014
Summary CV for student [Christina Constantinou]	CV.CC	11 August 2014
Summary CV for supervisor (student research) [Nicola Payne]	CV.NP	22 July 2014

Membership of the Committee

The members of the Ethics Committee who were present at the meeting are listed on the attached sheet.

After ethical review

Reporting requirements

The attached document "After ethical review – guidance for researchers" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The NRES website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website:

<http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/>

We are pleased to welcome researchers and R&D staff at our training days – see details at <http://www.hra.nhs.uk/hra-training/>

14/LO/1548 Please quote this number on all

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




**On behalf of
Miss Stephanie Ellis
Chair**

E-mail: nrescommittee.london-hampstead@nhs.net

Enclosures: List of names and professions of members who were present at the meeting and those who submitted written comments

“After ethical review – guidance for researchers”

Copy to: Dr Gordon Weller

Karen Ignatian,




Health Research Authority
National Research Ethics Service

NRES Committee London - Hampstead

Barlow House
3rd Floor 4 Minshull Street Manchester
M1 3DZ

Telephone: 0161 625 7815
Fax: 0161 625 7299

25 September 2014

Miss Christina Constantinou Department of



Dear Miss Constantinou

Study title: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

REC reference: 14/LO/1548

IRAS project ID: 157322

Thank you for your email of 23 September 2014. I can confirm the REC has received the documents listed below and that these comply with the approval conditions detailed in our letter dated 15 September 2014.

Documents received

The documents received were as follows:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Other [redacted] NHS Foundation Trust Lone Worker Policy]		
Participant information sheet (PIS)	IF 1B	23 September 2014
Research protocol or project proposal	1B	23 September 2014

Approved documents

The final list of approved documentation for the study is therefore as follows:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Evidence of Sponsor insurance or indemnity (non NHS Sponsors)		12 September 2013

only) [Indemnity insurance cover letter from Middlesex University]		
Interview schedules or topic guides for participants [Semi-Structured Interview Schedule for Young People (5-12 year olds)]	IS 1	02 July 2014
Interview schedules or topic guides for participants [Focus Group Schedule for Young People (13 to 18 year olds)]	FGS 2	02 July 2014
Letter from sponsor [Middlesex University]		08 August 2014
Other [CV. ██████████ (principle investigator)]		22 July 2014
Other [Demographic Information Sheet]	DIM 1	02 July 2014
Other [CV. Olga van den Akker]		25 July 2014
Other [Debriefing Form for Parents/Carer's]	DF 2	02 July 2014
Other [Debriefing Form for Young People (13-18 year olds)]	DF 3	02 July 2014
Other [Debriefing Form for Young People (5-12 year olds)]	DF 1	02 July 2014
Other [Guy's & St Thomas' NHS Foundation Trust Lone Worker Policy]		
Participant consent form [Young People (13-18 year olds)]	C5	02 July 2014
Participant consent form [Parents or Carer's]	C6	02 July 2014
Participant consent form [Young People (13-18 year olds)]	C4	02 July 2014
Participant consent form [Parents or Carer's]	C2	02 July 2014
Participant consent form [Parents or Carer's]	C3	02 July 2014
Participant consent form [Young People (5-12 year olds)]	C1	02 July 2014
Participant consent form [Parents or Carer's]	C7	02 July 2014
Participant information sheet (PIS) [Young People (13-18 year olds)]	IF 3	02 July 2014
Participant information sheet (PIS)	IF 1B	23 September 2014
Participant information sheet (PIS) [Parents/Carers]	IF 2	02 July 2014
REC Application Form		11 August 2014
Research protocol or project proposal	1B	23 September 2014
Summary CV for Chief Investigator (CI) [Christina Constantinou]		11 August 2014
Summary CV for supervisor (student research) [Nicola Payne]		22 July 2014

You should ensure that the sponsor has a copy of the final documentation for the study. It is the sponsor's responsibility to ensure that the documentation is made available to R&D offices at all participating sites.

14/LO/1548

Please quote this number on all

Yours sincerely



Dr Ashley Totenhofer REC Manager

E-mail: nrescommittee.london-hampstead@nhs.net

Copy to: Dr Gordon Weller – Middlesex University
 Karen Ignatian - ██████████ Foundation NHS Trust Professor Olga van den Akker – Middlesex University
 Dr Nicola Payne - Middlesex University



Health Research Authority

National Research Ethics Service

NRES Committee London - Hampstead

Barlow House
3rd Floor 4 Minshull Street Manchester
M1 3DZ

Tel: 0161 625 7849

11 June 2015

Miss Christina Constantinou



Dear Miss Constantinou

Study title:	An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)
REC reference:	14/LO/1548
Amendment number:	Version 1C
Amendment date:	18 May 2015
IRAS project ID:	157322

- The amendment consists of an increase in the number of research participants from 60 to 70.
- In addition research participants aged 13-18 years will receive £20 instead of £10 for travel costs and time given to the study.
- A study end date extension has also been proposed.

The above amendment was reviewed at the meeting of the Sub-Committee held on 11 June 2015 by the Sub-Committee in correspondence.

Ethical opinion

The members of the Committee taking part in the review gave a favourable ethical opinion of the amendment on the basis described in the notice of amendment form and supporting documentation.

There were no ethical issues raised.

Approved documents

The documents reviewed and approved at the meeting were:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Notice of Substantial Amendment (non-CTIMP)	Version 1C	18 May 2015
Participant consent form [13-18 Year old]	C4 B	18 May 2015
Participant consent form [13-18 Year old (Healthy Sibling Volunteer)]	C5 B	18 May 2015
Participant consent form [Parent or Carers]	C6 B	18 May 2015
Participant consent form [Parent or Carers (Healthy Sibling Volunteer)]	C7 B	18 May 2015
Participant information sheet (PIS) [Parent or Carers]	IF 2B	18 May 2015
Participant information sheet (PIS) [13-18 Years old (Healthy Sibling Volunteer)]	IF 3B	18 May 2015
Research protocol or project proposal	1C	18 May 2015

Membership of the Committee

The members of the Committee who took part in the review are listed on the attached sheet.

R&D approval

All investigators and research collaborators in the NHS should notify the R&D office for the relevant NHS care organisation of this amendment and check whether it affects R&D approval of the research.

Statement of compliance

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

We are pleased to welcome researchers and R & D staff at our NRES committee members' training days – see details at <http://www.hra.nhs.uk/hra-training/>

14/LO/1548:	Please quote this number on all correspondence
--------------------	---

Yours sincerely



Signed on behalf of Miss Stephanie Ellis Chair

E-mail: nrescommittee.london-hampstead@nhs.net

Enclosures: *List of names and professions of members who took part in the Review*

Copy to: *Karen Ignatian [REDACTED] Foundation NHS Trust*

Dr Gordon Weller, Middlesex University

Appendix D

Information sheets, assent/consent forms, debriefing forms and demographic information sheet (Studies 3 and 4)



IRAS project ID: 211257

Date: 22/03/2017

Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept.,
Middlesex University,
Town Hall, The Burroughs, Hendon,
London, NW4 4BT

Email: c.constantinou@mdx.ac.uk / **Tel:** 0208 411 6076

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Tel: 0208 411 5467 / 0208 411 6953

Participant Information Sheet for Young People (5-12 year olds)

An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

My name is Christina Constantinou and I am a researcher at Middlesex University. You are being asked to take part in my research study. I will explain to you why I am doing this. Then you can make up your mind if you would like to help me with it. Please talk to your mum, dad or carer if you have any questions. You can also ask me.

I am happy to read through this information sheet with you or you may like your mum, dad or carer to read it with you.

You will be asked to write your name on a sheet to say if you are happy to take part in the research study.

What is research?

Research is a way to find out the answers to questions and how you see things.

Why is this project being done?

I want to learn more about how happy you are with different parts of your life, like school and friends. I also want to know how you cope with sickle cell disease, so do you ask someone for help when you get a crisis?

I also want to learn about things like what kinds of exercise and sports you do and when you drink water.

This research is being carried out as part of my University studies.

Why have I been asked to take part?

All children with sickle cell disease at the hospital have been asked to take part in the research.

Did anyone else check the research is OK to do?

Before any research is allowed to happen, it has to be checked by a group of people called a Research Ethics Committee. They make sure that the research is fair. This research has been checked by the NRES London – City and East Committee. It has also been checked by Middlesex University where I am studying.

Do I have to take part?

You don't have to take part if you don't want to. It is up to you. I will describe what you have to do. You can change your mind at any time and stop taking part.

What will happen to me if I take part in the research?

I want you to answer some questions. Some of the questions are about how you feel (like do you feel sad) and how you feel about your school. There are other questions about what sports you do and about drinking water. It will take about 25 minutes. I'll help you do this, in a private room near the waiting room at the hospital.

Might anything about the research upset me?

If you feel upset or worried please tell me and I will stop asking you questions straightaway.

Will joining in help me?

It will give you something to do while you wait for your appointment. It is a chance to talk about how you feel about friends, school, your feelings and your health.

What if there is a problem?

If you have any problems when you answer the questions, I will stop straightaway. Then, I will ask if you want to talk to a doctor, one of the sickle cell nurses or someone from the psychology team. I can also get your mum, dad or carer if you would like me to. It is up to you who you want to talk to.

If you have any worries after finishing the questions you can again talk to a doctor, one of the sickle cell nurses or a member of the psychology team at any time. If you are not sure who to talk to you can always contact me first.

Will my taking part in the study be kept confidential?

Yes. Anything you say will be private. Anything you fill in, like the questions you answered, won't have your name on.

During the research I will keep your papers in a locked cupboard at the university where I am studying. Only I can get into the cupboard.

At the end of the research, a doctor will make sure that all of the papers you filled in are put in locked cupboards at the lead hospital, [REDACTED]. These papers will not have your name on. Only people like the doctor and his team can open these cupboards. After 10 years the doctor will make sure that all of the papers are ripped up. He will also make sure that anything saved on a computer is deleted.

I will only talk to doctor if I was very worried about you. But I would make sure you were ok with this first.

What will happen next?

After the research has finished, I will write it up for my University studies. I will also write about the research in academic magazines and talk about it in front of other people like doctors. Your name will not be included on anything I write or talk about.

Support (please see page 1 for contact details)

- For advice about if you should join in the research please talk to the consultant.
- If you feel upset, worried or have any other problems you can talk to a member of the psychology team, I'll give you their telephone number.
- You can also phone ChildLine on 0808 11 11 where you can talk to a counsellor straightaway about anything that is upsetting or bothering you.

Thank you for taking the time to read the information sheet!

IRAS project ID: 211257

Date: 22/03/2017

Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept., Middlesex University,
Town Hall, The Burroughs, Hendon,
London , NW4 4BT.

Email: c.constantinou@mdx.ac.uk / **Tel:** 0208 411 6076

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / **Tel:** 0208 411 5467 / o.vandenakker@mdx.ac.uk

Participant Information Sheet for Young People (13-18 year olds)

An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

My name is Christina Constantinou and I am a psychology PhD student at Middlesex University. You are being invited to take part in my research study. Before you decide to take part, it is important for you to understand why the research is being done and what it will involve. Please take your time to read the information below carefully and discuss it with others if you want to. Please ask if there is anything that is not clear or if you would like more information. If you would like I can read through this information sheet with you.

You will be given a copy of the information sheet and asked to sign a consent/assent form before taking part in the research.

What is the purpose of the research?

Health-related quality of life is important especially for adolescents with sickle cell disease. I am keen to learn more about how adolescents with sickle cell disease see their health-related quality of life. In other words, to find out how happy you feel with different parts of your life such as school, friendships and health. I am also interested to see how adolescents cope with sickle cell disease e.g. do you ask someone for help when you have a crisis?

Also, I want to ask you about your health behaviours such as the kinds of exercise and sports you do and more risky health behaviours such as drinking alcohol, smoking and substance use. Your health behaviours may or may not affect your health-related quality of life and your health-related quality of life may or may not affect your health behaviours. This is what I want to find out.

This study is being carried out as part of my PhD.

Why have I been invited?

All young people who are 13 to 18 years old with sickle cell disease who attend this hospital have been invited to take part.

Do I have to take part?

It is up to you to decide whether or not you want to take part. I will describe the study and go through the information sheet with you, if you want. You can withdraw from the study at any time up to data analysis, without giving a reason. This will have no affect on the care you receive from the hospital.

What will happen to me if I take part?

You will be asked to complete four questionnaires which measure your perception of your health-related quality of life, health behaviours and also risky behaviours including daily exercise and sport participation, drinking water and alcohol use and lastly a coping questionnaire. The coping questionnaire explores how adolescents cope with sickle cell disease e.g. do you ask someone for help when they're in pain, have a crisis or are tired. All of these questionnaires were developed for adolescents to complete. They should take about 25 minutes. I can help you to complete them if you like, here in a private room near the waiting room at the hospital.

If you are aged 16 years or older only you need to provide written consent for taking part in the research. If you are younger your parents/carers will also be asked to sign a consent form for you.

Your parents/carers will also be asked to complete two similar questionnaires which measure their perception of your health quality of life and your health behaviours including daily exercise and sport participation and also drinking water. They will also complete a form containing questions about background information (such as your age or gender).

What are the possible disadvantages of taking part?

There should be no risk for you at all. If you feel upset or anxious please tell me and you can immediately stop completing the questionnaires. Participation in this research is entirely voluntary.

What are the possible benefits to taking part?

The questionnaires may help some time pass by while you wait for your appointment. Also, it is a chance to think about how you feel about your health-related quality of life so things like your friends, health and school and your health behaviours.

What if there is a problem?

If you have any problems please talk to me. Or you can talk to your parent or carer or a member of staff from the paediatric sickle cell team or paediatric psychology department. If you decide to take part you may withdraw at any time up to data analysis, without giving a reason. All of the questionnaires that you completed will be destroyed.

Will my taking part in the study be kept confidential?

Yes. The information you provide will remain completely confidential.

All of the materials including demographic information sheets, questionnaires and the database where this information will be entered will be anonymous i.e. have a reference number and will not contain your name. During data collection, questionnaires and demographic information sheets will

be stored at Middlesex University in a secured cupboard that can only be accessed by Christina Constantinou, the researcher. After the study is completed Dr [REDACTED] (a Consultant at [REDACTED]) will be responsible for securely storing all of the anonymised materials in locked cupboards at the lead hospital, [REDACTED]. Consent/assent forms will be stored separately from the questionnaires so that no individual can be identified. Only Dr [REDACTED] and members of his team can access these cupboards. Also, the database will be stored on NHS and Middlesex University computers which are password protected and that can only be accessed by Christina Constantinou, Dr [REDACTED] and members of his team. After 10 years Dr [REDACTED] will be responsible for destroying i.e. shredding all materials and deleting the database.

Only in exceptional circumstances where I become concerned that your life or well-being is at risk would I speak to a consultant about the questionnaires you completed. This is very unlikely and I would tell you before I speak to a consultant.

What will happen next?

Once the research has been completed, I will analyse the anonymised group data which will be written up and form part of my PhD thesis. I will also use the anonymised group data for journal publications and conference presentations.

Who has reviewed the study?

This study has been reviewed and approved by the NRES London – City and East Committee and the Middlesex University Psychology Department’s Ethics Committee.

Further information and contact details ([please see previous page](#))

- For general information regarding the research please contact Christina Constantinou.
- For specific information about this research project please contact Christina Constantinou or Dr Nicky Payne.
- For advice as to whether you should participate please contact the sickle cell consultant.
- For independent information about participating in research please contact Patient Advice and Liaison Service (PALS) on [REDACTED] or email [REDACTED].
- To make a complaint, please contact the sickle cell consultant or Dr Nicky Payne.

Thank you for taking the time to read the information sheet.

IRAS project ID: 211257

Date: 22/03/2017

Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept., Middlesex University, Town Hall,
The Burroughs, Hendon, London, NW4 4BT

Email: c.constantinou@mdx.ac.uk / **Tel:** 0208 411 6076

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Tel: 0208 411 5467 / 0208 411 6953

Participant Information Sheet for Parents/Carers

An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

My name is Christina Constantinou and I am a psychology PhD student at Middlesex University. You and your child are being invited to take part in my research study. Before you decide to participate, it is important for you to understand why the research is being done and what it will involve. Please take your time to read the following information carefully, and discuss it with others if you wish. Please ask if there is anything that is not clear or if you would like more information. Take your time to decide whether or not you wish to take part. You will be given another copy of the information sheet and asked to sign consent forms prior to your child's participation in the research. If you do decide that you and your child will participate in the research study then I will read through the age-appropriate information sheet with them and they will also be asked to write their name on a simple assent form if they are happy to participate.

What is the purpose of the research?

Health-related quality of life is important for us to consider when thinking about treating chronic illnesses. I am keen to learn more about how children with sickle cell disease see their health-related quality of life. In other words, to find out how happy they feel with different parts of their life such as school, friendships and their health. Also, I want to find out about their health behaviours such as the kinds of exercise and sports they do and also, for children over 12 years old, more risky behaviour such as whether they drink alcohol and have ever tried smoking or other substances. I want to know whether health behaviours may affect their health-related quality of life which includes their physical health and psychological well-being. This study is being carried out as part of my doctoral research (i.e. PhD).

Why have I been invited?

All children who are 5 to 18 years old with sickle cell disease and their parent/carer attending this hospital have been invited to take part.

Do my children have to take part?

It is up to you to decide whether or not you want you and your child to take part. I have described the study in the form and would be happy to describe it to you in person at the hospital when you come in for your child's appointment if you wish. You and your child can withdraw from the study at any time up to data analysis, without giving a reason. This will have no affect on the care your child receives from the hospital.

What will happen to me and my child if we take part?

There are two questionnaires for parents/carers to complete which measure your perception of your child's health quality of life and their health behaviours including daily exercise and sport participation and also drinking water. There is also a form containing questions about background information (such as "are you currently employed?") to complete. The questionnaires should take about 25 minutes to complete, in a private room near the waiting room at the hospital.

Your child will also be asked to complete some very similar questionnaires; if they are aged 5 to 12 years old they will be asked to complete age appropriate versions of a health-related quality of life questionnaire, a health behaviour questionnaire and a kids coping questionnaire. The kids coping questionnaire explores how children cope with sickle cell disease e.g. do they ask someone for help when they're in pain, have a crisis or are tired. Also, adolescents aged 13 to 18 years old will also be asked to complete a risky behaviour questionnaire. The questionnaires were developed for children to complete. They should take about 25 minutes. I will help children to complete them, here in a private room near the waiting room at the hospital. If your child is aged 16 years or older you will not need to provide written consent on their behalf, their consent is sufficient.

What are the possible disadvantages of taking part?

There are no risks involved if your children take part in this research. Whether they chose to do so or not will not influence your child's care at the hospital. If during participation your child becomes upset for any reason or wishes to stop I will stop immediately.

What are the possible benefits to taking part?

This may give your children a chance to discuss how they feel about their health-related quality of life and health behaviours.

What if there is a problem?

If you have any problems please talk to me or a member of staff at the hospital. If your child has a problem they can immediately speak with me or a member of staff from the paediatric sickle cell team or paediatric psychology department. If you decide that your child may take part you may still withdraw at any time up to data analysis, without giving a reason.

Will my child taking part in the study be kept confidential?

Yes. The information you and your child provides will remain completely confidential. All of the materials including demographic information sheets, questionnaires and the database where this information will be entered will be anonymous i.e. have a reference number and will not contain

you or your child's name. During data collection, questionnaires and demographic information sheets will be stored at Middlesex University in a secured cupboard that can only be accessed by Christina Constantinou, the researcher. After the study is completed Dr [REDACTED] (a Consultant [REDACTED]) will be responsible for securely storing all of the anonymised materials in locked cupboards at the lead hospital, [REDACTED]. Consent/assent forms will be stored separately from the questionnaires so that no individual can be identified. Only Dr [REDACTED] and members of his team can access these cupboards. Also, the database will be stored on NHS and Middlesex University computers which are password protected and that can only be accessed by Christina Constantinou, Dr [REDACTED] and members of his team. After 10 years Dr [REDACTED] will be responsible for destroying i.e. shredding all materials and deleting the database.

Only in exceptional circumstances where I may become concerned that your child's life or well-being is at risk would I speak to a consultant about the questionnaires your child has completed. This is very unlikely and I would tell your child before I speak to a consultant.

What will happen next?

Once the research has been completed, I will analyse the anonymised group data which will be written up and form part of my PhD thesis. I will also use the anonymised group data for journal publications and conference presentations.

Who has reviewed the study?

This study has been reviewed and approved by the NRES London – City and East Committee and the Middlesex University Psychology Department's Ethics Committee.

Further information and contact details (please see previous page)

- For general information regarding the research please contact Christina Constantinou.
- For specific information about this research project please contact Christina Constantinou or Dr Nicky Payne.
- For advice as to whether you should participate please contact the sickle cell consultant.
- For independent information about participating in research please contact Patient Advice and Liaison Service (PALS) on [REDACTED] or email [REDACTED].
- To make a complaint, please contact the sickle cell consultant or Dr Nicky Payne.

Thank you for taking the time to read the participant information sheet.

IRAS project ID: 211257

Written Assent form for Young People (5-12 year olds)

(to be completed by the child and their parent/guardian)

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

Chief Investigator/Researcher: Miss Christina Constantinou

Participant Identification Number:

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

- I have read the information sheet with the researcher. Yes/No
This is dated **22/03/2017 (Version IF 1B)** for the above study.

- The researcher has talked to me about the study. I have been able to ask the researcher questions. The researcher has answered my questions. Yes/No

- I understand and am happy to take part. Yes/No

- Do you understand it's OK to stop taking part at any time? Yes/No

If any answers are 'no' or you don't want to take part, don't write your name!

If you do want to take part, please can write your name below.

Your name

Date

Name of person taking consent

Signature

Date

Thank you for your help.



IRAS project ID: 211257

Written Assent form for Young People (13-15 year olds)

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors *(only for students):* Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator *(only for students):* Dr [REDACTED]

Participant Identification Number:

Please initial box

- I have read and understood the information sheet dated **22/03/2017** **(Version IF 2B)** for the above study.
- I understand that my participation is entirely voluntary, the data collected during the research will not be identifiable, and I have the right to withdraw from the study at any time up to data analysis without any obligation to explain my reasons for doing so.
- I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- I further understand that the anonymous data I provide may be used for analysis and subsequent publication, and provide my consent that this might occur.
- I understand that data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures.
- I agree to take part in the above study.

Name of participant	Date	Signature

Name of person taking consent	Date	Signature

IRAS project ID: 211257

Written Informed Consent for Young People (16-18 year olds)

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator (*only for students*): Dr [REDACTED]

Participant Identification Number:

Please initial box

- I have read and understood the information sheet dated **22/03/2017** (Version IF 2B) for the above study.
- I understand that my participation is entirely voluntary, the data collected during the research will not be identifiable, and I have the right to withdraw from the study at any time up to data analysis without any obligation to explain my reasons for doing so.
- I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- I further understand that the anonymous data I provide may be used for analysis and subsequent publication, and provide my consent that this might occur.
- I understand that data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures.
- I agree to take part in the above study.

Name of participant

Date

Signature

IRAS project ID: 211257

Written Informed Consent for Parents or Carer's

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

Chief Investigator/Researcher: Miss Christina Constantinou

Academic Supervisors (*only for students*): Dr Nicky Payne and Professor Olga van den Akker

Principle Investigator (*only for students*): Dr [REDACTED]

Participant Identification Number:

Please initial box

- I have read and understood the information sheet dated **22/03/2017** (**Version IF 3B**) for the above study.
- I understand that my child's participation is entirely voluntary, the data collected during the research will not be identifiable, and that they have the right to withdraw from the study at any time, up to data analysis without any obligation to explain their reasons for doing so.
- I have been given contact details for the researcher in the information sheet and had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- I further understand that the anonymous data they provide may be used for analysis and subsequent publication, and provide my consent that this might occur.
- I understand that data may be inspected by the Chair of the Psychology Ethics panel and the Chair of the School of Social Sciences Ethics committee of Middlesex University, if required by institutional audits about the correctness of procedures.
- I agree that my child and I will take part in the above study.

Name of participant

Date

Signature

Name of person taking consent

Date

Signature

7. What type of sickle cell do you have (please circle as appropriate):

Sickle Cell Anaemia / HbSS

HbSC

Hb β +Hb β ⁰

Sickle cell trait

Other (*please specify*)

I do not have sickle cell disease

Information Regarding the Child:**8. Is your child (please circle as appropriate):** Male Female**9. Age** _____ **years old.****10. What is their ethnic group (please circle as appropriate):**Asian (*please specify*)

Black African

Black Caribbean

Black Other (*please specify*)Mixed (*please specify*)

Other Ethnic Group (*please specify*)

White

11. What religious faith does your child follow (please circle as appropriate):

Christianity

Hinduism

Islam

Traditional African religion

Other Religious faith (*please specify*)

They are not religious

Does not wish to disclose

12. Is your child currently in (please circle as appropriate):

School / Full-time Education

College / Full-time

College / Part-time

Training *(please specify)*Employment *(please specify)*

Unemployed

13. What type of sickle cell does your child have (please circle as appropriate):

Sickle Cell Anaemia / HbSS

HbSC

Hb β +Hb β ⁰Other *(please specify)*

Not Applicable

14. How many crisis has your child encountered in the last 12 months i.e. 1, 2 etc?

_____ crisis

15. What treatment did they receive (please circle as appropriate indicating the amount of times they received this treatment i.e. 1, 2 etc.)?

Treated at home _____

GP _____

Medication required

A&E attendance _____

Hospital attendance _____

Other _____

16. How many days has your child missed from school due to illness related to sickle cell disease in the last 12 months?

_____ Days

Thank you for taking the time to fill in the demographic information sheet



Date: 02/01/2017

Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept., Middlesex University,
Town Hall, The Burroughs, Hendon,
London , NW4 4BT

Email: c.constantinou@mdx.ac.uk /**Tel:** 0208 411 6076

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Tel: 0208 411 5467 / 0208 411 6953

Debriefing Form for Young People (5-12 year olds)

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents

Points to discuss with children once they have completed their questionnaires.

- a) Thank you very much for helping with my research which is now finished.

- b) Do you have some worries or questions for me about the research or anything you have not understood?

- c) Would you like to ask me, your parent or carer or the hospital staff here at the hospital some questions?

- d) If you feel sad or worried at any time after leaving the hospital:
 - You can talk to your parent or carer;
 - ask your parent or carer to call a doctor or nurse from the sickle team and you can talk to them;
 - or you can talk to a counsellor from Childline for free on 0800 1111. Childline is private.

- e) **Thank you again!**

Date: 02/01/2017

Chief Investigator/Researcher Name: Christina Constantinou

Institutional Contact Details:

Psychology Dept., Middlesex University,
Town Hall, The Burroughs, Hendon,
London, NW4 4BT

Email: c.constantinou@mdx.ac.uk / **Tel:** 0208 411 6076

Academic Supervisor's Names: Dr Nicky Payne and Professor Olga van den Akker

For Contact Details: see institutional contact details

Email: n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk

Tel: 0208 411 5467 / 0208 411 6953

Debriefing Form for Young People (13-18 year olds)

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

Your participation in the research has finished. You may have some questions for either me or another member of staff.

About the research.....

- a) Past research shows that children and adolescents with Sickle Cell Disease (SCD) report a lower Health-Related Quality of Life (HRQL) compared to their healthy peers. Also, parents' reports of children's HRQL are often lower than the child's.

- b) Good health behaviours, for example regularly drinking water, should be a part of children's daily lives and may help to improve different aspects of their HRQL including psychological well-being and physical health, although there has been little research in this area.

- c) Previous research about HRQL and health behaviours was carried out in London hospitals involving a small number of children with SCD and their healthy siblings. The research found that children with SCD would like to exercise more often and also are less likely to drink water at home compared to at school. Also, some adolescents with SCD drink alcohol and to a much lesser extent, some have tried smoking and substances like shisha.

d) It is important to see whether these findings are the same in a larger sample of children and adolescents with SCD. Also, we wanted to learn why children choose to engage in some health behaviours and not others because, for example, drinking water may help their condition. We also wish to see whether this might affect their HRQL.

What to do if you feel upset after taking part.....

- a) If you have any concerns, you can contact me ([see contact details at the top of this form](#)) and I will contact a member of staff at the hospital or you can contact someone from the sickle cell team directly using the telephone number you already have for them. This will all be in confidence.
- b) Or you may prefer to talk about your concerns to a counsellor from Childline just call them for free on 0800 1111. Childline is confidential.

Here are some points to consider.....

- a) Please note that all questionnaires will be confidential to the researcher and her supervisors at Middlesex University. All data will be published anonymously.
- b) If you would like any information about the results of the study once it is completed, please contact me or ask your parent or carer to contact me and provide an email address or postal address ([see contact details at the top of this form](#)). Please be aware that there will be a prolonged period of time before you receive the results of the study.

Thank you for participating in the study.

Date: 02/01/2017

Chief Investigator/Researcher Name: Christina Constantinou**Institutional Contact Details:**Psychology Dept, Middlesex University,
Town Hall, The Burroughs, Hendon,
London, NW4 4BT**Email:** c.constantinou@mdx.ac.uk / **Tel:** 0208 411 6076**Academic Supervisor's Names:** Dr Nicky Payne and Professor Olga van den Akker**Email:** n.payne@mdx.ac.uk / o.vandenakker@mdx.ac.uk**Tel:** 0208 411 5467 / 0208 411 6953

Debriefing Form for Parents/Carers

Title of Study: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

Both your and your child's participation in the research has finished. You may have some questions for either me or another member of staff.

About the research.....

- a) Past research shows that children and adolescents with Sickle Cell Disease (SCD) report a lower Health-Related Quality of Life (HRQL) compared to their healthy peers. Also, parents' reports of children's HRQL are often lower than the child's.
- b) Good health behaviours, for example regularly drinking water, should be a part of children's daily lives and may help to improve different aspects of their HRQL including psychological well-being and physical health, although there has been little research in this area.
- c) Previous research about HRQL and health behaviours was carried out in London hospitals involving a small number of children with SCD and their healthy siblings. The research found that children with SCD would like to exercise more often and also that they are less likely to drink water at home compared to at school. Also, some adolescents with SCD drink alcohol and to a much lesser extent, some have tried smoking and substances like shisha.
- d) It is important to see whether these findings are the same in a larger sample of children and adolescents with SCD. Also, we wanted to learn why children choose to engage in some health behaviours and not others because, for example, drinking water may help their condition. We also wish to see whether this might affect their HRQL.

What to do if you/your child feel upset after taking part.....

- a) If you/your child have any concerns, you can contact me ([see contact details at the top of this form](#)) and I will contact a member of staff at the hospital or you can contact someone from the sickle cell team directly using the telephone number you already have for them. This will all be in confidence.
- b) Or you may prefer to talk about your concerns to the Samaritans just call them for free on 116 123 or you can email them; jo@samaritans.org. Your child may also prefer to talk about their concerns to a counsellor from Childline, they can call them for free on 0800 1111. The Samaritans and Childline are confidential.

Here are some points to consider.....

- a) Please note that all questionnaires and demographic information will be confidential to the researcher and her supervisors at Middlesex University. All data will be published anonymously.
- b) If you or your children would like any information about the results of the study once it is completed, please contact me and provide an email address or postal address ([see contact details at the top of this form](#)). Please be aware that there will be a prolonged period of time before you receive the results of the study.

Thank you for participating in the study

Appendix E

Questionnaires (Studies 3 and 4)

ID#	_____
Date:	_____

PedsQLTM

Paediatric Quality of Life Inventory

Version 4.0 - English (United Kingdom)

YOUNG CHILD REPORT (ages 5-7)

Instructions for interviewer:

I am going to ask you some questions about things that might be a problem for some children. I want to know how much of a problem any of these things might be for you.




Show the child the template and point to the responses as you read.

*If it is **not at all** a problem for you, point to the smiling face.*

*If it is **sometimes** a problem for you, point to the middle face.*

*If it is a problem for you **a lot**, point to the frowning face.*

I will read each question. Point to the pictures to show me how much of a problem it is for you. Let's try a practice one first.

	Not at all	Sometimes	A lot
Is it hard for you to click your fingers?			

Ask the child to demonstrate by clicking his or her fingers to determine whether or not the question was answered correctly. Repeat the question if the child demonstrates a response that is different from his or her action.

Think about how you have been doing over the last few weeks. Please listen carefully to each sentence and tell me how much of a problem this is for you.

After reading the item, gesture to the template. If the child hesitates or does not seem to understand how to answer, read the response options while pointing at the faces.

PHYSICAL FUNCTIONING (problems with...)	Not at all	Some times	A lot
1. Is it hard for YOU to walk?	0	2	4
2. Is it hard for you to run?	0	2	4
3. Is it hard for you to play sports or exercise?	0	2	4
4. Is it hard for you to lift bio things?	0	2	4
5. Is it hard for you to have a bath or shower?	0	2	4
6. Is it hard for you to help in the home (like picking up your toys)?	0	2	4
7. Do you have aches and pains <i>Where?</i>	0	2	4
8. Do you ever feel too tired to play?	0	2	4

MOTIONAL FUNCTIONING (problems with...)	Not at all	Some times	A lot
1. Do you feel scared?	0	2	4
2. Do you feel sad?	0	2	4
3. Do you feel angry?	0	2	4
4. Do you have trouble sleeping?	0	2	4
5. Do you worry about what will happen to you?	0	2	4

SOCIAL FUNCTIONING (problems with...)	Not at all	Some times	A lot
1. Is it hard for you to get on with other children?	0	2	4
2. Do other children say they do not want to play with you?	0	2	4
3. Do other children tease you?	0	2	4
4. Can other children do things you cannot do?	0	2	4
5. Is it hard for you to keep up when you play with other children?	0	2	4

SCHOOL FUNCTIONING (problems with...)	Not at all	Some times	A lot
1. Is it hard for you to pay attention in school?	0	2	4
2. Do you forget thinos?	0	2	4
3. Is it hard to keep up with schoolwork?	0	2	4
4. Do you miss school because of not feelino well?	0	2	4
5. Do you miss school because you have to go to the doctor or hospital?	0	2	4

ID# _____
Date: _____

PedsQLTM
Paediatric Quality of Life
Inventory

Version 4.0 - English (United Kingdom)

PARENT REPORT for YOUNG CHILDREN (ages 5-7)

INSTRUCTIONS

On the following page is a list of things that might be a problem for **your child**. Please tell us **how much of a problem** each one has been for **your child** during the **past ONE month** by circling:

- 0** if it is **never** a problem
- 1** if it is **almost never** a problem
- 2** if it is **sometimes** a problem
- 3** if it is **often** a problem
- 4** if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has your child had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking 100 metres	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activities or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores, like picking up his or her toys	0	1	2	3	4
7. Having aches or pains	0	1	2	3	4
8. Feeling tired	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Getting on with other children	0	1	2	3	4
2. Other children not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not being able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

SCHOOL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with school activities	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

ID# _____

Date: _____

PedsQLTM
Paediatric Quality of Life
Inventory

Version 4.0 - English (United Kingdom)

CHILD REPORT (ages 8-12)**INSTRUCTIONS**

On the following page is a list of things that might be a problem for you. Please tell us **how much of a problem** each one has been for you over the **PAST MONTH** by circling:

- 0** if it is **never** a problem
- 1** if it is **almost never** a problem
- 2** if it is **sometimes** a problem
- 3** if it is **often** a problem
- 4** if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

Over the **PAST MONTH**, how much of a **problem** has this been for you...

ABOUT MY HEALTH AND ACTIVITIES (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard for me to walk more than a couple of streets (about 100 metres)	0	1	2	3	4
2. It is hard for me to run	0	1	2	3	4
3. It is hard for me to do sports activities or exercise	0	1	2	3	4
4. It is hard for me to lift heavy things	0	1	2	3	4
5. It is hard for me to have a bath or shower by myself	0	1	2	3	4
6. It is hard for me to do chores around the house	0	1	2	3	4
7. I have aches and pains	0	1	2	3	4
8. I feel tired	0	1	2	3	4

ABOUT MY FEELINGS (problems with.)	Never	Almost Never	Some-times	Often	Almost Always
1. I feel afraid or scared	0	1	2	3	4
2. I feel sad	0	1	2	3	4
3. I feel angry	0	1	2	3	4
4. I have trouble sleeping	0	1	2	3	4
5. I worry about what will happen to me	0	1	2	3	4

How I GET ON WITH OTHERS (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. I have trouble getting on with other children	0	1	2	3	4
2. Other children do not want to be my friend	0	1	2	3	4
3. Other children tease me	0	1	2	3	4
4. I cannot do things that other children my age can do	0	1	2	3	4
5. It is hard to keep up when I play with other children	0	1	2	3	4

ABOUT SCHOOL (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard to pay attention in class	0	1	2	3	4
2. I forget things	0	1	2	3	4
3. I have trouble keeping up with my school work	0	1	2	3	4
4. I miss school because of not feeling well	0	1	2	3	4
5. I miss school to go to the doctor or hospital	0	1	2	3	4

ID# _____

Date: _____

PedsQLTM
Paediatric Quality of Life
Inventory

Version 4.0 - English (United Kingdom)

PARENT REPORT for CHILDREN (ages 8-12)**INSTRUCTIONS**

On the following page is a list of things that might be a problem for **your child**. Please tell us **how much of a problem** each one has been for **your child** during the **past ONE month** by circling:

- 0** if it is **never** a problem
- 1** if it is **almost never** a problem
- 2** if it is **sometimes** a problem
- 3** if it is **often** a problem
- 4** if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has your child had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking 100 metres	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activities or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores around the house	0	1	2	3	4
7. Having aches or pains	0	1	2	3	4
8. Feeling tired	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Getting on with other children	0	1	2	3	4
2. Other children not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not being able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

SCHOOL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with schoolwork	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

ID# _____
Date: _____

PedsQLTM
Paediatric Quality of Life
Inventory

Version 4.0 - English (United Kingdom)

TEENAGER REPORT (ages 13-18)

INSTRUCTIONS

On the following page is a list of things that might be a problem for you. Please tell us **how much of a problem** each one has been for you over the **PAST MONTH** by circling:

- 0** if it is **never** a problem
- 1** if it is **almost never** a problem
- 2** if it is **sometimes** a problem
- 3** if it is **often** a problem
- 4** if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

Over the **PAST MONTH**, how much of a **problem** has this been for you...

ABOUT MY HEALTH AND ACTIVITIES (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard for me to walk more than a couple of streets (about 100 metres)	0	1	2	3	4
2. It is hard for me to run	0	1	2	3	4
3. It is hard for me to do sports activities or exercise	0	1	2	3	4
4. It is hard for me to lift heavy things	0	1	2	3	4
5. It is hard for me to have a bath or shower by myself	0	1	2	3	4
6. It is hard for me to do chores around the house	0	1	2	3	4
7. I have aches and pains	0	1	2	3	4
8. I feel tired	0	1	2	3	4

ABOUT MY FEELINGS (problems with)	Never	Almost Never	Some-times	Often	Almost Always
1. I feel afraid or scared	0	1	2	3	4
2. I feel sad	0	1	2	3	4
3. I feel angry	0	1	2	3	4
4. I have trouble sleeping	0	1	2	3	4
5. I worry about what will happen to me	0	1	2	3	4

How I GET ON WITH OTHERS (problems with.)	Never	Almost Never	Some-times	Often	Almost Always
1. I have trouble getting on with other teenagers	0	1	2	3	4
2. Other teenagers do not want to be my friend	0	1	2	3	4
3. Other teenagers tease me	0	1	2	3	4
4. I cannot do things that other teenagers my age can do	0	1	2	3	4
5. It is hard to keep up with other teenagers my age	0	1	2	3	4

ABOUT SCHOOL/ COLLEGE (problems with..)	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard to pay attention in class	0	1	2	3	4
2. I forget things	0	1	2	3	4
3. I have trouble keeping up with my school / college work	0	1	2	3	4
4. I miss school /college because of not feeling well	0	1	2	3	4
5. I miss school / college to go to the doctor or hospital	0	1	2	3	4

ID# _____

Date: _____

PedsQLTM

Paediatric Quality of Life Inventory

Version 4.0 English (United Kingdom)

PARENT REPORT for TEENAGERS (ages 13-18)

INSTRUCTIONS

On the following page is a list of things that might be a problem for **your teenager**.

Please tell us **how much of a problem** each one has been for **your teenager** during the **past ONE month** by circling:

- 0** if it is **never** a problem
- 1** if it is **almost never** a problem
- 2** if it is **sometimes** a problem
- 3** if it is **often** a problem
- 4** if it is **almost always** a problem

There are no right or wrong answers.

If you do not understand a question, please ask for help.

In the past **ONE month**, how much of a **problem** has your teenager had with ...

PHYSICAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Walking 100 metres	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activities or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores around the house	0	1	2	3	4
7. Having aches or pains	0	1	2	3	4
8. Feeling tired	0	1	2	3	4

EMOTIONAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

SOCIAL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Getting on with other teenagers	0	1	2	3	4
2. Other teenagers not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other teenagers	0	1	2	3	4
4. Not being able to do things that other teenagers his or her age can do	0	1	2	3	4
5. Keeping up with other teenagers	0	1	2	3	4

SCHOOL FUNCTIONING (problems with...)	Never	Almost Never	Some-times	Often	Almost Always
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with schoolwork	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

The Kids Coping Scale

When your **sickle cell disease is bad**, like when you're in pain, have a crisis or tired, what do you normally do?

	Never	Some- times	A lot
1. You tried to think of different ways to solve the problem.			
2. You did not want to think about it.			
3. You thought about what others might do.			
4. You tried your best to make things better.			
5. You avoided the problem.			
6. You asked someone to help.			
7. You tried hard to fix the problem.			
8. You did things to stop thinking about it.			

Health Behaviour Questionnaire - Child Version

Section 1: Daily Exercise and Sport

1. During a normal week, so Monday to Sunday, how many times on average do you do the following kinds of exercise for more than 15 minutes during your free time? This can be at break times, lunch times, after school, evenings and weekends...

1a. STRENUOUS EXERCISE (heart beats very fast) _____ times per week (e.g., playing chase, running, football, rugby, basketball, tennis, gymnastics, dancing that uses lots of energy, swimming fast, riding your bike fast for a long time or up a hill)

1b. MODERATE EXERCISE (not very tired) _____ times per week (e.g., walking fast, dancing, badminton, easy swimming, riding your bike or scooter, skateboarding, roller skating)

1c. MILD EXERCISE (not a lot of effort) _____ times per week (e.g., easy walking, golf)

2. I like doing exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

3. I think that doing exercise helps me be healthy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

4. I think that doing exercise helps my sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. I think that doing exercise might make me get a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my life (e.g. family, friends, teachers and doctors) exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

7. Important people in my life (e.g. family, friends, teachers and doctors) encourage me to exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

8. Important people in my life (e.g. family, friends, teachers and doctors) think that doing exercise might be bad for my sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

9. Who influences whether you do, or don't exercise the most? **Please circle one**

Mum/dad

Grandparent(s)

Brother or sister

Best friend(s)

Friend(s)

Classmates

Teacher(s)

Doctor

Nurses

Media or sports people

10. It's difficult to do exercise because we don't have much money:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

11. It's difficult to do exercise when my parents are busy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

12. Because of my sickle cell disease it's difficult to do exercise when its cold outside:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

13. It's difficult to do exercise because sickle cell disease makes me tired:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

14. It's difficult to do exercise because I worry that I might not be good at it:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Section 2: Drinking water

1. How many glasses of water do you drink in one day at...

...school:

0 1 2 3 4 5 6 or more

...afterschool:

0 1 2 3 4 5 6 or more

... the weekend:

0 1 2 3 4 5 6 or more

2. I like drinking water:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

3. I think that drinking water helps me be healthy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

4. I think that drinking water helps my sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. Important people in my life (e.g. family, friends, teachers and doctors) drink water:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my life (e.g. family, friends, teachers and doctors) encourage me to drink water:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

7. Important people in my life (e.g. family, friends, teachers and doctors) think that drinking water will help my sickle cell disease e.g. help stop me from getting a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

8. Who influences whether you do, or don't drink water the most? **Please circle one**

Mum/dad

Grandparent(s)

Brother or sister

Best friend(s)

Friend(s)

Classmates

Teacher(s)

Doctor

Nurses

Media or sports people

9. It's difficult to drink water during lessons at school like maths class:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

10. It's difficult to drink water at school as I don't like getting special treatment because I have sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

11. It's difficult drinking water when I'm busy doing other things:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Health Behaviour Questionnaire - Adolescent Version

Section 1: Daily Exercise and Sport

1. During a typical 7-day period (a week), how many times on average do you do the following kinds of exercise for more than 15 minutes during your free time e.g. break times, lunch times, after school/college/work, evenings and weekends? Please write on each line the appropriate number.

1a. STRENUOUS EXERCISE (heart beats rapidly) _____ times per week
(e.g., running, football, rugby, basketball, tennis, gymnastics, energetic dancing, vigorous swimming, vigorous long distance bicycling or cycling on hilly ground)

1b. MODERATE EXERCISE (not exhausted) _____ times per week (e.g., walking fast, dancing, badminton, easy swimming, cycling on level ground, skateboarding, rollerblading)

1c. MILD EXERCISE (minimal effort) _____ times per week (e.g., easy walking, yoga, archery, golf)

2. I like doing exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

3. I think that doing exercise helps me be healthy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

4. I think that doing exercise helps my sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. I think that doing exercise might make me get a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my life (e.g. family, friends, teachers and doctors) exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

7. Important people in my life (e.g. family, friends, teachers and doctors) encourage me to exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

8. Important people in my life (e.g. family, friends, teachers and doctors) think that doing exercise might be bad for my sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

9. Who influences whether you do, or don't exercise the most? **Please circle one**

Mum/dad	Grandparent(s)	Brother or sister
Best friend(s)	Friend(s)	Classmates
Teacher(s)	Doctor	Nurses
		Media or sports people

10. It's difficult to do exercise because we don't have much money:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

11. It's difficult to do exercise when my parents are busy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

12. Because of my sickle cell disease it's difficult to do exercise when its cold outside:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

13. It's difficult to do exercise because sickle cell disease makes me tired:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

14. It's difficult to do exercise because I worry that I might not be good at it:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Section 2: Drinking water

1. How many glasses of water do you drink in one day at...

...school:

0 1 2 3 4 5 6 or more

...afterschool:

0 1 2 3 4 5 6 or more

... the weekend:

0 1 2 3 4 5 6 or more

2. I like drinking water:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

3. I think that drinking water helps me be healthy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

4. I think that drinking water helps my sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. Important people in my life (e.g. family, friends, teachers and doctors) drink water:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my life (e.g. family, friends, teachers and doctors) encourage me to drink water:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

7. Important people in my life (e.g. family, friends, teachers and doctors) think that drinking water will help my sickle cell disease e.g. help stop me from getting a crisis:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

8. Who influences whether you do, or don't drink water the most? **Please circle one**

Mum/dad	Grandparent(s)	Brother or sister
Best friend(s)	Friend(s)	Classmates
Doctor	Nurses	Media or sports people
Teacher(s)		

9. It's difficult to drink water during lessons at school like maths class:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

10. It's difficult to drink water at school as I don't like getting special treatment because I have sickle cell disease:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

11. It's difficult drinking water when I'm busy doing other things:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Section 2: Alcohol

1. I like drinking alcohol:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

2. I think that drinking alcohol helps me be healthy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

3. I think that drinking alcohol might make me get a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

4. Important people in my life (e.g. family, friends, teachers and doctors) approve of me drinking alcohol:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. Important people in my life (e.g. family, friends, teachers and doctors) think that drinking alcohol might make me get a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my life (e.g. family, friends, teachers and doctors) drink alcohol:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree7. Who influences whether you do, or don't drink alcohol the most? **Please circle one**

Mum/dad

Grandparent(s)

Brother or sister

Best friend(s)

Friend(s)

Classmates

Teacher(s)

Doctor

Nurses

Media or sports people

8. It's difficult for me to drink alcohol because it's expensive:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

9. It's difficult for me to drink alcohol because it's hard to get hold of:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

10. When I'm with my friends it's hard for me to say 'no' to drinking alcohol.

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Health Behaviour Questionnaire - Parent Version

Section 1: Daily Exercise and Sport

1. During a typical 7-day period (a week), how many times on average does your child do the following kinds of exercise for more than 15 minutes during their free time e.g. break times, lunch times, after school/college/work, evenings and weekends? Please write on each line the appropriate number.

1a. STRENUOUS EXERCISE (heart beats rapidly) _____ times per week
(e.g., playing chase, running, football, rugby, basketball, gymnastics, energetic dancing, vigorous swimming, vigorous long distance bicycling or cycling on hilly ground)

1b. MODERATE EXERCISE (not exhausted) _____ times per week
(e.g., walking fast, dancing, tennis, badminton, easy swimming, cycling on level ground, riding their scooter, skateboarding, rollerblading, roller skating)

1c. MILD EXERCISE (minimal effort) _____ times per week (e.g., easy walking, yoga, archery, golf)

2. I think that exercise is enjoyable for my child:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

3. I think that doing exercise helps my child be healthy:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

4. I think that doing exercise helps my child's sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. I think that doing exercise might make my child get a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my child's life (e.g. family, friends, teachers and doctors) exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

7. Important people in my child's life (e.g. family, friends, teachers and doctors) encourage them to exercise:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

8. Important people in my child's life (e.g. family, friends, teachers and doctors) think that doing exercise could be bad for their sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

9. Who influences whether your child does, or does not exercise the most? **Please circle one**

Mum/dad	Grandparent(s)	Brother or sister	
Best friend(s)	Friend(s)	Classmates	Teacher(s)
Doctor	Nurses	Media or sports people	

10. I think that it's difficult for my child to exercise because we don't have much money:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree
11. I think that it's difficult for my child to exercise when we are busy:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree
12. Because of my child's sickle cell disease I think that it's difficult for my child to exercise when its cold outside:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree
13. I think that it's difficult for my child to exercise because sickle cell disease makes them tired:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree
14. I think that it's difficult for my child to do exercise because they worry that they might not be good at it:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Section 2: Drinking water

1. How many glasses of water does your child drink in one day at...

...school:

0 1 2 3 4 5 6 or more

...afterschool:

0 1 2 3 4 5 6 or more

... the weekend:

0 1 2 3 4 5 6 or more

2. I think that my child likes drinking water:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree
3. I think that drinking water helps my child be healthy:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree
4. I think that drinking water helps my child's sickle cell disease:
Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

5. Important people in my child's life (e.g. family, friends, teachers and doctors) drink water:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

6. Important people in my child's life (e.g. family, friends, teachers and doctors) encourage them to drink water:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

7. Important people in my child's life (e.g. family, friends, teachers and doctors) think that drinking water will help their sickle cell disease e.g. help stop them from getting a crisis:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

8. Who influences whether your child does, or does not drink water the most? **Please circle one**

Mum/dad	Grandparent(s)	Brother or sister
Best friend(s)	Friend(s)	Classmates
Doctor	Nurses	Media or sports people
		Teacher(s)

9. I think that it could be difficult for my child to drink water during lessons at school:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

10. I think that it could be difficult for my child to drink water at school as they don't like getting special treatment because they have sickle cell disease:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

11. I think that it's difficult for my child to drink water when they are busy doing things:

Disagree: 1 : 2 : 3 : 4 : 5 : 6 : 7 : Agree

Appendix F

Favourable letters from the research ethics committee reference number 17/LO/0322
(Studies 3 and 4)



Health Research Authority

**London - City & East Research Ethics
Committee**

Bristol Research Ethics Committee Centre
Whitefriars Level 3, Block B Lewins Mead
Bristol BS1 2NT
Telephone: 02071048033/53

Please note: This is the favourable
opinion of the REC only and does not
allow

you to start your study at NHS sites in
England until you receive HRA Approval

16 March 2017

Miss Christina Constantinou


Dear Miss Constantinou

Study title:	An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)
REC reference:	17/LO/0322
Protocol number:	N/A
IRAS project ID:	211257

The Research Ethics Committee reviewed the above application at the meeting held on 02 March 2017. Thank you for attending to discuss the application.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this favourable opinion letter. The expectation is that this information will be published for all studies that receive an ethical opinion but should you wish to provide a substitute contact point, wish to make a request to defer, or require further information, please contact hra.studyregistration@nhs.net outlining the reasons for your request.

Under very limited circumstances (e.g. for student research which has received an unfavourable opinion), it may be possible to grant an exemption to the publication of the study.

Ethical opinion

The members of the Committee present gave a favourable ethical opinion of the above research on the basis described in the application form, protocol and supporting documentation, subject to the conditions specified below.

Conditions of the favourable opinion

The REC favourable opinion is subject to the following conditions being met prior to the start of the study.

1. *It should be clarified in the PIS that anonymised study data will be stored for 10 years at the lead NHS trust, after which it will be destroyed.*
2. *There are inconsistencies in the PIS as the terms “I”, “me” and “we” are used in a number of places. The PIS should be reviewed and revised accordingly.*
3. *The application and PIS mention that the participants can withdraw from the study at any point up to December 2017. This should be revised withdrawal from the study at any point up to the analyses of the data.*
4. *PIS does not clarify what will happen to the data once it has been analysed. The participants should be given some information on what will happen to the data and how it will be used.*
5. *The Committee agreed that the paragraph at the bottom of the consent forms with regards to inspection by ethics panel, should be included as a main consent statement.*
6. *The Committee noted a number of other errors and inconsistencies in the PIS detailed in the tracked changes PIS provided with the opinion letter as agreed by the Committee.*
7. *The Committee noted that in the demographic questionnaire there is a question about religious faith. There should be an option to say ‘does not wish to disclose’ or it should be clear that the person filling the form does not have to answer any question they would prefer to leave.*

You should notify the REC once all conditions have been met (except for site approvals from host organisations) and provide copies of any revised documentation with updated version numbers. Revised documents should be submitted to the REC electronically from IRAS. The REC will acknowledge receipt and provide a final list of the approved documentation for the study, which you can make available to host organisations to facilitate their permission for the study. Failure to provide the final versions to the REC may cause delay in obtaining permissions.

Management permission must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements. Each NHS organisation must confirm through the signing of agreements and/or other documents that it has given permission for the research to proceed (except where explicitly specified otherwise).

Guidance on applying for HRA Approval (England)/ NHS permission for research is available in the Integrated Research Application System, at www.hra.nhs.uk or at <http://www.rdforum.nhs.uk>.

Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity. For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation. Sponsors are not required to notify the Committee of management permissions from host organisations.

Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database. This should be before the first participant is recruited but no later than 6 weeks after recruitment of the first participant.

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to request a deferral for study registration within the required timeframe, they should contact hra.studyregistration@nhs.net. The expectation is that all clinical trials will be registered, however, in exceptional circumstances non registration may be permissible with prior agreement from the HRA. Guidance on where to register is provided on the HRA website.

It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).

Ethical review of research sites

NHS Sites

The favourable opinion applies to all NHS sites taking part in the study taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion" below).

Summary of discussion at the meeting

Favourable risk benefit ratio; anticipated benefit/risks for research participants (present and future)

The Committee requested you to clarify how long would the study data be stored for. You explained that the anonymised data will be stored for 10 years at the lead NHS trust, after which it will be destroyed. The Committee requested the same to be explained in the PIS. You agreed.

Please contact the REC Manager if you feel that the above summary is not an accurate reflection of the discussion at the meeting.

Approved documents

The documents reviewed and approved at the meeting were:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Middlesex University Indemnity]		09 September 2016
IRAS Application Form [IRAS_Form_10022017]		10 February 2017
Letter from sponsor [Letter from Sponsor for Christina Constantinou]		31 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Child Version]	HBQ1	02 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Adolescent Version]	HBQ2	02 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Parent Version]	HBQ3	02 January 2017
Other [Demographic Information Sheet]	DIS 1	02 January 2017
Other [Debriefing Form for 5-12 year olds]	DF 1	02 January 2017
Other [Debriefing Form for 13-18 year olds]	DF 2	02 January 2017
Other [Debriefing Form Parents or Carers]	DF 3	02 January 2017
Participant consent form [Child Assent form (5-12 year olds)]	C1 A	08 February 2017
Participant consent form [Adolescents Assent Form (13-15 year olds)]	C2 A	08 February 2017
Participant consent form [Consent Form for Older adolescents (16- 18 year olds)]	C4 A	08 February 2017
Participant consent form [Parent or Carer Consent Form]	C3 A	08 February 2017
Participant information sheet (PIS) [Participant information sheet for young people (5-12 year olds)]	IF 1	02 January 2017
Participant information sheet (PIS) [Participant information sheet for young people (13-18 year olds)]	IF 2	02 January 2017
Participant information sheet (PIS) [Participant information sheet for Parent or Carer]	IF 3	02 January 2017
Participant information sheet (PIS) [Participant information sheet for 5-12 year olds]	IF 1A	08 February 2017
Participant information sheet (PIS) [Participant information sheet for Parent (13-18 year olds)]	IF 2A	08 February 2017
Participant information sheet (PIS) [Participant information sheet for Parent or Carer]	IF 3A	08 February 2017
Research protocol or project proposal [Protocol]	1A	02 January 2017
Summary CV for Chief Investigator (CI) [CV]	CV.CC	27 January 2017
Summary CV for student [CV for Christina Constantinou]	CV.CC	27 January 2017
Summary CV for supervisor (student research) [CV for Nicola Payne]	CV.NP	27 January 2017
Summary CV for supervisor (student research) [CV for Olga van den Akker]	CV.OvdA	02 February 2017
Summary CV for supervisor (student research) [CV for ██████ (Principle Investigator)]	CV.BI	30 January 2017
Validated questionnaire [PedsQL]	4.0	
Validated questionnaire [The Kids Coping Scale]		

Membership of the Committee

The members of the Ethics Committee who were present at the meeting are listed on the attached sheet.

There were no declarations of interest

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

After ethical review

Reporting requirements

The attached document “After ethical review – guidance for researchers” gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: <http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/>

HRA Training

We are pleased to welcome researchers and R&D staff at our training days – see details at <http://www.hra.nhs.uk/hra-training/>

17/LO/0322

Please quote this number on all correspondence

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

Yours sincerely



pp Dr John Keen Chair

E-mail: nrescommittee.london-cityandeast@nhs.net

Enclosures: List of names and professions of members who were present at the meeting and those who submitted written comments

"After ethical review – guidance for researchers"

*Copy to: Dr Gordon Weller
Mays Jawad, [REDACTED]*



Health Research Authority

London - City & East Research Ethics Committee

Bristol Research Ethics Committee Centre

Whitefriars Level 3, Block B Lewins Mead

Bristol BS1 2NT

Telephone: 02071048033/53

Please note: This is an acknowledgement letter from the REC only and does not allow you to start your study at NHS sites in England until you receive HRA Approval

23 March 2017

Miss Christina Constantinou



Dear Miss Constantinou

Study title: An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)

REC reference: 17/LO/0322

Protocol number: N/A

IRAS project ID: 211257

Thank you for your response of 23 March 2017. I can confirm the REC has received the documents listed below and that these comply with the approval conditions detailed in our letter dated 16 March 2017

Documents received

The documents received were as follows:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Other [Demographic Information Sheet]	Version DIS 1A	22 March 2017
Participant consent form [Written Informed Consent for	Version C3 B	22 March 2017

Parents/Carers]		
Participant consent form [Assent form for Young People (5-12 year olds)]	Version C1 B	22 March 2017
Participant consent form [Assent form for Young People (13-15 year olds)]	Version C2 B	22 March 2017
Participant consent form [Written Informed Consent for Young People (16-18 year olds)]	Version C4 B	22 March 2017
Participant information sheet (PIS) [Participant Information Sheet for Young People (5-12 year olds)]	Version IF 1B	22 March 2017
Participant information sheet (PIS) [Participant Information Sheet for Young People (13-18 year olds)]	Version IF 2B	22 March 2017
Participant information sheet (PIS) [Participant Information Sheet for Parents/Carers]	Version IF 3B	22 March 2017

Approved documents

<i>Document</i>	<i>Version</i>	<i>Date</i>
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Middlesex University Indemnity]		09 September 2016
IRAS Application Form [IRAS_Form_10022017]		10 February 2017
IRAS Checklist XML [Checklist_23032017]		23 March 2017
Letter from sponsor [Letter from Sponsor for Christina Constantinou]		31 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Child Version]	HBQ1	02 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Adolescent Version]	HBQ2	02 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Parent Version]	HBQ3	02 January 2017
Other [Debriefing Form for 5-12 year olds]	DF 1	02 January 2017
Other [Debriefing Form for 13-18 year olds]	DF 2	02 January 2017
Other [Debriefing Form Parents or Carers]	DF 3	02 January 2017
Other [Demographic Information Sheet]	Version DIS 1A	22 March 2017
Participant consent form [Assent form for Young People (5-12 year olds)]	Version C1 B	22 March 2017
Participant consent form [Assent form for Young People (13-15 year olds)]	Version C2 B	22 March 2017
Participant consent form [Written Informed Consent for Young People (16-18 year olds)]	Version C4 B	22 March 2017
Participant consent form [Written Informed Consent for Parents/Carers]	Version C3 B	22 March 2017
Participant information sheet (PIS) [Participant information sheet for young people (5-12 year olds)]	IF 1	02 January 2017
Participant information sheet (PIS) [Participant information sheet for young people (13-18 year olds)]	IF 2	02 January 2017
Participant information sheet (PIS) [Participant information sheet for Parent or Carer]	IF 3	02 January 2017
Participant information sheet (PIS) [Participant Information Sheet for	Version IF	22 March 2017

Young People (5-12 year olds)]	1B	
Participant information sheet (PIS) [Participant Information Sheet for Young People (13-18 year olds)]	Version IF 2B	22 March 2017
Participant information sheet (PIS) [Participant Information Sheet for Parents/Carers]	Version IF 3B	22 March 2017
Research protocol or project proposal [Protocol]	1A	02 January 2017
Summary CV for Chief Investigator (CI) [CV]	CV.CC	27 January 2017
Summary CV for student [CV for Christina Constantinou]	CV.CC	27 January 2017
Summary CV for supervisor (student research) [CV for Nicola Payne]	CV.NP	27 January 2017
Summary CV for supervisor (student research) [CV for Olga van den Akker]	CV.OvdA	02 February 2017
Summary CV for supervisor (student research) [CV for ██████████ (Principle Investigator)]	CV.BI	30 January 2017
Validated questionnaire [PedsQL]	4.0	
Validated questionnaire [The Kids Coping Scale]		

You should ensure that the sponsor has a copy of the final documentation for the study. It is the sponsor's responsibility to ensure that the documentation is made available to R&D offices at all participating sites.

17/LO/0322

Please quote this number on all correspondence

Yours sincerely



Rajat Khullar REC Manager

E-mail: nrescommittee.london-cityandeast@nhs.net

Copy to: *Dr Gordon Weller*
Dr Mays Jawad, ██████████



Health Research Authority

Miss Christina Constantinou
 [REDACTED]
 [REDACTED]
 [REDACTED]

Email: hra.approval@nhs.net

05 April 2017

Dear Miss Constantinou

Letter of HRA Approval

Study title:	An Examination of Health-Related Quality of Life (HRQL) and Health Behaviours in Children and Adolescents with Sickle Cell Disease (SCD)
IRAS project ID:	211257
Protocol number:	N/A
REC reference:	17/LO/0322
Sponsor	Middlesex University

I am pleased to confirm that HRA Approval has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications noted in this letter.

Participation of NHS Organisations in England

The sponsor should now provide a copy of this letter to all participating NHS organisations in England.

Appendix B provides important information for sponsors and participating NHS organisations in England for arranging and confirming capacity and capability. **Please read *Appendix B* carefully**, in particular the following sections:

- *Participating NHS organisations in England* – this clarifies the types of participating organisations in the study and whether or not all organisations will be undertaking the same activities
- *Confirmation of capacity and capability* - this confirms whether or not each type of participating NHS organisation in England is expected to give formal confirmation of capacity and capability. Where formal confirmation is not expected, the section also provides details on the time limit given to participating organisations to opt out of the study, or request additional time, before their participation is assumed.
- *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* - this provides detail on the form of agreement to be used in the study to confirm

capacity and capability, where applicable.

Further information on funding, HR processes, and compliance with HRA criteria and standards is also provided.

It is critical that you involve both the research management function (e.g. R&D office) supporting each organisation and the local research team (where there is one) in setting up your study. Contact details and further information about working with the research management function for each organisation can be accessed from www.hra.nhs.uk/hra-approval.

Appendices

The HRA Approval letter contains the following appendices:

- A – List of documents reviewed during HRA assessment
- B – Summary of HRA assessment

After HRA Approval

The document “*After Ethical Review – guidance for sponsors and investigators*”, issued with your REC favourable opinion, gives detailed guidance on reporting expectations for studies, including:

- Registration of research
- Notifying amendments
- Notifying the end of the study

The HRA website also provides guidance on these topics, and is updated in the light of changes in reporting expectations or procedures.

In addition to the guidance in the above, please note the following:

- HRA Approval applies for the duration of your REC favourable opinion, unless otherwise notified in writing by the HRA.
- Substantial amendments should be submitted directly to the Research Ethics Committee, as detailed in the *After Ethical Review* document. Non-substantial amendments should be submitted for review by the HRA using the form provided on the [HRA website](#), and emailed to hra.amendments@nhs.net.
- The HRA will categorise amendments (substantial and non-substantial) and issue confirmation of continued HRA Approval. Further details can be found on the [HRA website](#).

Scope

HRA Approval provides an approval for research involving patients or staff in NHS organisations in England.

If your study involves NHS organisations in other countries in the UK, please contact the relevant national coordinating functions for support and advice. Further information can be found at <http://www.hra.nhs.uk/resources/applying-for-reviews/nhs-hsc-rd-review/>.

If there are participating non-NHS organisations, local agreement should be obtained in accordance with the procedures of the local participating non-NHS organisation.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: <http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/>.

HRA Training

We are pleased to welcome researchers and research management staff at our training days – see details at <http://www.hra.nhs.uk/hra-training/>

Your IRAS project ID is **211257**. Please quote this on all correspondence. Yours sincerely

Catherine Adams

Senior Assessor

Email: hra.approval@nhs.net

Copy to: *Dr Gordon Weller, Sponsor Representative*
Dr Mays Jawad, [REDACTED]

<i>Document</i>	<i>Version</i>	<i>Date</i>
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Middlesex University Indemnity]		09 September 2016
IRAS Application Form [IRAS_Form_10022017]		10 February 2017
IRAS Checklist XML [Checklist_23032017]		23 March 2017
Letter from sponsor [Letter from Sponsor for Christina Constantinou]		31 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Child Version]	HBQ1	02 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Adolescent Version]	HBQ2	02 January 2017
Non-validated questionnaire [Health Behaviour Questionnaire - Parent Version]	HBQ3	02 January 2017
Other [Debriefing Form for 5-12 year olds]	DF 1	02 January 2017
Other [Debriefing Form for 13-18 year olds]	DF 2	02 January 2017
Other [Debriefing Form Parents or Carers]	DF 3	02 January 2017
Other [Demographic Information Sheet]	Version DIS 1A	22 March 2017
Participant consent form [Assent form for Young People (5-12 year olds)]	Version C1 B	22 March 2017
Participant consent form [Assent form for Young People (13-15 year olds)]	Version C2 B	22 March 2017
Participant consent form [Written Informed Consent for Young People (16-18 year olds)]	Version C4 B	22 March 2017
Participant consent form [Written Informed Consent for Parents/Carers]	Version C3 B	22 March 2017
Participant information sheet (PIS) [Participant information sheet for young people (5-12 year olds)]	IF 1	02 January 2017
Participant information sheet (PIS) [Participant information sheet for young people (13-18 year olds)]	IF 2	02 January 2017
Participant information sheet (PIS) [Participant information sheet for Parent or Carer]	IF 3	02 January 2017
Participant information sheet (PIS) [Participant Information Sheet for	Version IF	22 March 2017

Appendix A - List of Documents

The final document set assessed and approved by HRA Approval is listed below.

Young People (5-12 year olds)]	1B	
Participant information sheet (PIS) [Participant Information Sheet for Young People (13-18 year olds)]	Version IF 2B	22 March 2017
Participant information sheet (PIS) [Participant Information Sheet for Parents/Carers]	Version IF 3B	22 March 2017
Research protocol or project proposal [Protocol]	1A	02 January 2017
Summary CV for Chief Investigator (CI) [CV]	CV.CC	27 January 2017
Summary CV for student [CV for Christina Constantinou]	CV.CC	27 January 2017
Summary CV for supervisor (student research) [CV for Nicola Payne]	CV.NP	27 January 2017
Summary CV for supervisor (student research) [CV for Olga van den Akker]	CV.OvdA	02 February 2017
Summary CV for supervisor (student research) [CV for ██████████ (Principle Investigator)]	██████████	30 January 2017
Validated questionnaire [PedsQL]	4.0	
Validated questionnaire [The Kids Coping Scale]		

Other

Statement of Activities Schedule of Events

Appendix B - Summary of HRA Assessment

This appendix provides assurance to you, the sponsor and the NHS in England that the study, as reviewed for HRA Approval, is compliant with relevant standards. It also provides information and clarification, where appropriate, to participating NHS organisations in England to assist in assessing and arranging capacity and capability.

For information on how the sponsor should be working with participating NHS organisations in England, please refer to the *participating NHS organisations, capacity and capability* and *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* sections in this appendix.

The following person is the sponsor contact for the purpose of addressing participating organisation questions relating to the study:

Dr Gordon Weller
E-mail G.Weller@mdx.ac.uk Telephone 02084114509

HRA assessment criteria

Section	HRA Assessment Criteria	Compliant with Standards?	Comments
1.1	IRAS application completed correctly	Yes	A46 states that identifiable data will be stored for 10 years however supporting information indicates that this will be non identifiable data.
2.1	Participant information/consent documents and consent process	Pending	Changes have been made to information sheets to comply with HRA standards
3.1	Protocol assessment	Yes	No comments
4.1	Allocation of responsibilities and rights are agreed and documented	Yes	A statement of activities is in use. The sponsor is not requesting and does not expect any other site agreement.

Section	HRA Assessment Criteria	Compliant with Standards?	Comments
4.2	Insurance/indemnity arrangements assessed	Yes	Where applicable, independent contractors (e.g. General Practitioners) should ensure that the professional indemnity provided by their medical defence organisation covers the activities expected of them for this research study
4.3	Financial arrangements assessed	Yes	No funding is to be provided as indicated in the Statement of Activities
5.1	Compliance with the Data Protection Act and data security issues assessed	Yes	No comments
5.2	CTIMPS – Arrangements for compliance with the Clinical Trials Regulations assessed	Not Applicable	No comments
5.3	Compliance with any applicable laws or regulations	Yes	No comments
6.1	NHS Research Ethics Committee favourable opinion received for applicable studies	Yes	No comments
6.2	CTIMPS – Clinical Trials Authorisation (CTA) letter received	Not Applicable	No comments
6.3	Devices – MHRA notice of no objection received	Not Applicable	No comments
6.4	Other regulatory approvals and authorisations received	Not Applicable	No comments

Participating NHS Organisations in England

This provides detail on the types of participating NHS organisations in the study and a statement as to whether the activities at all organisations are the same or different.

All organisations will be undertaking the same activity (i.e. there is only one 'site-type') as detailed in the supporting documentation

The Chief Investigator or sponsor should share relevant study documents with participating NHS organisations in England in order to put arrangements in place to deliver the study. The documents should be sent to both the local study team, where applicable, and the office providing the research management function at the participating organisation. For NIHR CRN Portfolio studies, the Local LCRN contact should also be copied into this correspondence. For further guidance on working with participating NHS organisations please see the HRA website.

If Chief Investigators, sponsors or Principal Investigators are asked to complete site level forms for participating NHS organisations in England which are not provided in IRAS or on the HRA website, the Chief Investigator, sponsor or Principal Investigator should notify the HRA immediately at hra.approval@nhs.net. The HRA will work with these organisations to achieve a consistent approach to information provision.

Confirmation of Capacity and Capability

Principal Investigator Suitability

This confirms whether the sponsor's position on whether a PI, LC or neither should be in place is correct for each type of participating NHS organisation in England, and the minimum expectations for education, training and experience that PIs should meet (where applicable).

A Local Collaborator is required to facilitate study activity in each participating organisation.

GCP training is not a generic training expectation, in line with the [HRA statement on training expectations](#).

This describes whether formal confirmation of capacity and capability is expected from participating NHS organisations in England.

Participating NHS organisations in England **will be expected to formally confirm their capacity and capability** to host this research.

- The sponsor should ensure that participating NHS organisations are provided with a copy of this letter and all relevant study documentation, and work jointly with NHS organisations to arrange capacity and capability whilst the HRA assessment is ongoing.
- Further detail on how capacity and capability will be confirmed by participating NHS organisations, following issue of the Letter of HRA Approval, is provided in the *Participating NHS Organisations and Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* sections of this appendix.
- The [Assessing, Arranging, and Confirming](#) document on the HRA website provides further information for the sponsor and NHS organisations on assessing, arranging and confirming capacity and capability.

HR Good Practice Resource Pack Expectations

This confirms the HR Good Practice Resource Pack expectations for the study and the pre-engagement checks that should and should not be undertaken.

Where arrangements are not already in place, network staff (or similar) undertaking any of the research activities listed in A18 or A19 of the IRAS form (except for administration of questionnaires or surveys), would be expected to obtain an honorary research contract from one NHS organisation (if university employed), followed by Letters of Access for subsequent organisations.

This would be on the basis of a Research Passport (if university employed) or an NHS to NHS confirmation of pre-engagement checks letter (if NHS employed). These should confirm enhanced DBS checks, including appropriate barred list checks, and occupational health clearance. For research team members only administering questionnaires or surveys, a Letter of Access based on standard DBS checks and occupational health clearance would be appropriate.

Information to Aid Study Set-up

This details any other information that may be helpful to sponsors and participating NHS organisations in England in study set-up.

The applicant has indicated that they do not intend to apply for inclusion on the NIHR CRN Portfolio.

Appendix G

Tables of non-significant preliminary findings (Studies 3 and 4)

HRQL and coping

Table G1

Correlations between Mean HRQL Scores and the Kids Coping Scale for Children (n=106) and Adolescents (n=96)

	Child self-report			Parent proxy			Adolescent self-report			Parent proxy		
	Problem	Emotion	SS	Problem	Emotion	SS	Problem	Emotion	SS	Problem	Emotion	SS
<u>HRQL</u>												
HRQL total score	-.08	.04	.07	.04	-.07	.15	-.01	-.07	-.06	-.03	-.06	-.08
Physical health summary score	-.13	.14	.15	.02	-.07	.15	-.07	.03	-.12	-.06	.05	-.10
Psychosocial health summary score	-.04	-.03	.01	.06	-.05	.14	.02	-.12	-.02	-.02	-.13	-.05

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Problem = Problem-focused coping. Emotion = Emotion-focused coping. SS = Seeking social support.

HRQL / health behaviours and children's gender

Table G2

Gender Differences in Mean HRQL Scores for Different Dimensions of the PedsQL™ and Health Behaviour Scores for Children (n=106)

	Child self-report			Parent proxy		
	Male M(SD)	Female M(SD)	<i>t</i>	Male M(SD)	Female M(SD)	<i>t</i>
<u>HRQL</u>						
HRQL total score	71.42(17.94)	74.98(14.58)	-1.12	68.72(17.29)	73.54(16.83)	-1.45
Physical health summary score	69.44(20.15)	75.66(18.42)	-1.66	66.03(19.82)	72.00(21.18)	-1.50
Psychosocial health summary score	72.47(18.59)	74.62(14.68)	-.66	70.15(17.10)	74.36(16.17)	-1.30
<u>Health Behaviour</u>						
Weekly exercise	125.06(57.48)	144.04(59.33)	-1.67	129.26(61.00)	143.21(64.50)	-1.15
Strenuous exercise	62.83(47.64)	75.12(45.19)	1.36	62.33(48.54)	75.46(53.14)	-1.33
Moderate exercise	31.94(17.60)	34.42(19.01)	-.70	35.09(17.28)	34.81(18.12)	.08
Water consumption	29.37(12.70)	34.63(14.13)	2.02*	32.06(12.69)	37.13(12.69)	-2.06*

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

HRQL / health behaviours and adolescents gender

Table G3

Gender Differences in Mean HRQL Scores for Different Dimensions of the PedsQL™ and Health Behaviour Scores for Adolescents (n=96)

	Adolescent self-report			Parent proxy		
	Male M(SD)	Female M(SD)	<i>t</i>	Male M(SD)	Female M(SD)	<i>t</i>
<u>HRQL</u>						
HRQL total score	75.34(15.57)	70.77(18.17)	1.34	74.66(15.73)	68.70(18.15)	1.72
Physical health summary score	71.29(21.35)	65.10(21.27)	1.42	71.29(21.05)	64.13(20.14)	1.70
Psychosocial health summary score	77.50(14.37)	73.78(18.37)	1.10	76.46(14.57)	71.15(18.76)	1.55
<u>Health Behaviour</u>						
Weekly exercise	140.65(62.62)	75.79(58.38)	5.25***	133.92(63.27)	71.31(60.22)	4.97***
Strenuous exercise	70.50(49.13)	30.38(37.85)	4.48***	65.63(49.29)	29.63(40.84)	3.90***
Moderate exercise	33.65(18.44)	21.35(19.04)	3.21**	35.10(20.33)	19.06(16.62)	4.23***
Water consumption	42.27(15.23)	43.88(17.20)	-.48	42.46(16.22)	41.98(15.93)	.15

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

HRQL / health behaviours and age

Table G4

Correlations between Mean HRQL Scores for Different Dimensions of the PedsQL™ and Health Behaviour Scores and Children's (n=106) and Adolescents (n=96) Age

	Child self-report	Parent proxy	Adolescent self-report	Parent proxy
<u>HRQL</u>				
HRQL total score	-.14	-.20*	-.16	-.15
Physical health summary score	-.17	-.15	-.12	-.09
Psychosocial health summary score	-.10	-.21*	.16	-.17
<u>Health Behaviour</u>				
Weekly exercise	-.22*	-.20*	-.08	-.14
Strenuous exercise	-.14	-.16	-.08	-.08
Moderate exercise	-.23*	.11	-.22*	-.22*
Water consumption	.06	.07	.10	.13

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

HRQL / health behaviours and marital status (children)

Table G5

Parents Marital Status Differences in Mean HRQL Scores for Child Reports of Different Dimensions of the PedsQLTM and Health Behaviours (n=106)

	Child self-report			Parent proxy		
	Single M(SD)	Married M(SD)	<i>t</i>	Single M(SD)	Married M(SD)	<i>t</i>
<u>HRQL</u>						
HRQL total score	72.06(17.09)	74.36(15.69)	-.72	70.34(18.39)	71.89(15.87)	-.46
Physical health summary score	70.80(21.00)	74.33(17.33)	-.93	68.24(22.01)	69.73(19.19)	-.37
Psychosocial health summary score	72.73(17.14)	74.38(16.43)	-.51	71.45(17.97)	73.04(15.35)	-.49
<u>Health Behaviour</u>						
Weekly exercise	135.00(59.12)	133.69(59.23)	-.11	135.00(65.01)	137.29(61.02)	-.19
Strenuous exercise	67.25(45.72)	70.59(48.01)	-.37	67.91(53.72)	69.71(48.49)	-.18
Moderate exercise	33.27(19.15)	33.04(17.44)	.07	34.36(16.01)	35.59(19.33)	-.36
Water consumption	32.53(12.44)	31.33(14.88)	.45	35.13(11.67)	33.92(14.16)	.48

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

HRQL / health behaviours and marital status (adolescents)

Table G6

Parents Marital Status Differences in Mean HRQL Scores for Adolescent Reports of Different Dimensions of the PedsQL™ and Health Behaviours (n=96)

	Adolescent self-report			Parent proxy		
	Single M(SD)	Married M(SD)	<i>t</i>	Single M(SD)	Married M(SD)	<i>t</i>
<u>HRQL</u>						
HRQL total score	70.11(18.63) 76.25(14.52)		-1.79	69.02(17.80)	74.57(16.12)	-1.60
Physical health summary score	66.13(22.10) 70.45(13.55)		-.99	64.75(20.55)	70.92(20.83)	-1.46
Psychosocial health summary score	72.23(18.31)	79.35(13.55)	-2.15*	71.30(17.75)	76.52(15.70)	-1.52
<u>Health Behaviour</u>						
Weekly exercise	104.04(70.07) 112.76(67.18)		-.62	100.42(72.60) 105.00(65.64)		-.32
Strenuous exercise	49.14(47.98) 51.85(48.64)		-.27	48.06(49.55)	47.15(47.93)	.09
Moderate exercise	25.50(18.61) 29.67(20.69)		-1.04	25.00(19.25)	29.34(21.07)	-1.06
Water consumption	41.30(15.93) 45.00(16.40)		-1.21	41.02(16.28)	43.52(15.75)	-.76

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

HRQL / health behaviours and parents education (children)

Table G7

One-Way ANOVA of Different Dimensions of HRQL and Health Behaviours in Adolescents by Highest Parental Educational Attainment (n=96)

	Child self-report				Parent proxy			
	Group 1 M(SD)	Group 2 M(SD)	Group 3 M(SD)	<i>F</i>	Group 1 M(SD)	Group 2 M(SD)	Group 3 M(SD)	<i>F</i>
<u>HRQL</u>								
HRQL total score	72.40(17.12)	72.02(15.00)	74.40(16.73)	0.22	71.28(15.99)	67.34(16.05)	72.83(18.65)	0.78
Physical health summary score	69.65(22.70)	71.33(17.64)	75.49(17.32)	0.98	70.07(19.85)	63.72(18.48)	70.69(22.22)	0.96
Psychosocial health summary score	73.86(16.77)	72.39(15.44)	73.81(17.70)	0.07	71.93(16.03)	69.28(15.95)	73.96(17.74)	0.60
<u>Health Behaviour</u>								
Weekly exercise	133.61(59.99)	133.91(55.10)	135.24(61.08)	0.01	139.71(68.58)	135.78(56.83)	133.22(61.90)	0.11
Strenuous exercise	74.13(47.23)	61.43(41.66)	68.20(48.94)	0.53	76.50(55.59)	61.83(43.87)	65.80(50.69)	0.72
Moderate exercise	30.26(18.85)	35.43(14.37)	34.44(19.55)	0.76	33.29(18.02)	36.52(16.20)	35.56(18.22)	0.28
Water consumption	35.95(14.32)	32.22(11.24)	28.44(13.44)	3.27	39.58(13.57)	33.70(11.05)	30.73(11.95)	5.33**

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Group 1 = higher degree degree. Group 2 = foundation degree or diploma. Group 3 = A Levels or below.

HRQL and Health behaviours and parents education (adolescents)

Table G8

One-Way ANOVA of Different Dimensions of HRQL and Health Behaviours in Adolescents by Highest Parental Educational Attainment (n=96)

	Adolescent self-report				Parent proxy			
	Group 1 M(SD)	Group 2 M(SD)	Group 3 M(SD)	<i>F</i>	Group 1 M(SD)	Group 2 M(SD)	Group 3 M(SD)	<i>F</i>
<u>HRQL</u>								
HRQL total score	74.08(15.92)	70.98(17.69)	73.27(18.05)	0.24	71.40(17.36)	70.89(15.38)	72.54(18.47)	0.07
Physical health summary score	68.99(21.10)	64.81(21.10)	69.58(21.45)	0.38	67.71(20.58)	66.44(19.15)	68.57(22.63)	0.07
Psychosocial health summary score	76.79(15.56)	74.28(16.75)	75.25(17.77)	0.18	73.38(17.39)	73.26(14.73)	74.66(18.16)	0.07
<u>Health Behaviour</u>								
Weekly exercise	106.18(68.60)	85.96(55.57)	125.62(73.21)	2.67	94.92(65.19)	86.65(59.55)	122.24(76.25)	2.12
Strenuous exercise	51.69(47.52)	30.91(45.64)	62.21(47.44)	3.06	45.23(46.01)	30.52(42.54)	61.94(52.09)	3.09*
Moderate exercise	26.79(19.98)	26.09(18.64)	29.26(20.34)	0.22	25.38(19.85)	27.83(17.83)	28.83(22.31)	0.24
Water consumption	47.41(14.23)	39.65(15.02)	40.41(18.23)	2.45	44.15(15.52)	41.13(15.17)	40.74(17.26)	0.48

Note. * $p < .05$. ** $p < .01$. *** $p < .001$.

Group 1 = higher degree or degree. Group 2 = foundation degree or diploma. Group 3 = A Levels or below