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Title: A Qualitative Exploration of Health-Related Quality of Life and Health Behaviours in Children with Sickle Cell Disease and Healthy Siblings

Short title: *Exploring paediatric sickle cell disease*

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Abstract

Objectives*:** This study explored the health-related quality of life (HRQL) and health behaviours of children with sickle cell disease (SCD) and healthy siblings, drawing on Gap theory, which suggests HRQL is the discrepancy between current and ideal selves. ***Design: Thirty-two interviews, facilitated by children's drawings of their current and ideal selves were thematically analysed. ***Results*:** Two themes were identified. First, limitations of SCD and adjusted expectations. Children with SCD report some discrepancy in HRQL as they would like to participate in more physical activity, but overall, they appear to have normalised their condition and adjusted their expectations in the context of the limits of their condition. Healthy siblings worry about their sibling and have greater expectations about engaging in adventurous activities and for their future. Second, coping with SCD. Children have limited social support, although children with SCD seek support from their mothers. They also modify health behaviours, like reducing exercise to help prevent and cope with sickle-related pain. ***Conclusion*:** Children have some discrepancies in their HRQL but adjusted expectations among children with SCD may reduce discrepancy. Adapting health behaviours may help to cope with SCD but it is important that reductions in physical activity do not impair HRQL.

Keywords:

children; chronic illness; health-related quality of life; qualitative methods; health behaviour.

Introduction

Sickle Cell Disease (SCD) is one of the most prevalent inherited haemoglobinopathies (blood disorders) (Bennett, 2011). Sickle cell anaemia (SCA) is the most common and severest form (Ilesanmi, 2013). Globally, more than 300,000 babies are born each year with SCD (Ware et al., 2017). In the United Kingdom (UK), SCD is reported in one in every 4,600 people, which means approximately 14,000 are living with SCD (Dormandy et al., 2018). The condition affects males and females equally, with over a third of children coming from single-parent or divorced families who are typically of lower socio-economic status (SES) (Ilesanmi, 2013), and it is most prevalent among individuals of Black African and Black Caribbean descent (Dormandy et al., 2018; Streetly et al., 2010). In children with SCD the most prevalent complication is vaso-occlusive crises which occur when blood vessels become blocked by sickle-shaped red blood cells. These may be triggered by, for example, physical or mental stress, infection, dehydration, excessive strenuous exercise or cold weather (Ballas et al., 2010; Brown, 2012; Kanter & Kruse-Jarres, 2013). Vaso-occlusive crises are characterised by the sudden onset of severe, acute painful episodes, with continuous and throbbing pain which may occur throughout the body, including in bones, muscles and organs (Kanter & Kruse-

Jarres, 2013). These episodes commonly last for hours to days, and in rarer instances for weeks, and can be extremely painful and debilitating (Ballas et al., 2010). Other physical complications of SCD include anaemia, infection, acute chest syndrome and stroke (Kanter & Kruse-Jarres, 2013).

As well as the physical consequences, children with SCD may suffer from psychosocial problems such as increased depression, anxiety, fatigue and more social problems compared to healthy children, as well as hospitalisations and frequent absences from school (Ünal et al., 2011). Social-cultural factors including the shared values and circumstances of families with SCD, may also exacerbate psychosocial issues and lead to poorer disease adjustment and coping. For example, financial hardships and a reluctance to discuss illness among their community, family or friends, especially in African cultures may be problematic (Barbarin & Christian, 1999), although they often draw on their religious beliefs such as prayer and spirituality (alongside medication) to help cope with SCD (Clayton-Jones & Haglund, 2015; Ilesanmi, 2013).

These physical and psychosocial consequences may contribute to lower health-related quality of life (HRQL) (Panepinto & Bonner, 2012). HRQL is a multidimensional concept that assesses individuals physical, social, emotional and school/work functioning (Panepinto et al., 2005). Questionnaires are the most prevalent method of measuring HRQL in children with SCD (Panepinto & Bonner, 2012), where child and parent-proxy reports are generally found to be lower than in healthy children (Panepinto et al., 2005). However, this research assesses current HRQL, often using the Pediatric Quality of Life Inventory, which is not based on a theoretical approach. In contrast, using measures based on a Gap theory (GT; Calman, 1984) approach, where HRQL is considered the gap or discrepancy between children's current and ideal selves, children with chronic conditions generally do not seem to have a lower quality of life (QoL) compared to healthy children in research on SCA (Constantinou et al., 2015), chronic kidney disease (Heath et al., 2011) and asthma (Eiser et al., 2000). For example, the Generic Children's Quality of Life Measure, which assesses self-reported discrepancy QoL, was administered to children with SCA and their QoL was not lower compared to a matched healthy sample (Constantinou et al., 2015). Parent proxy-reported QoL discrepancy scores were also not lower compared to child self-reports (Constantinou et al., 2015). Moreover, GT (Calman, 1984) aligns with the World Health Organization (WHO) definition of QoL; *"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).

HRQL has rarely been explored using qualitative methods in children with SCD, although the experiences of these children have been considered more generally, often focusing on pain rather than the entire experience of living with SCD (Panepinto et al., 2012; Stegenga et al., 2004). Although children appear to have a good understanding of SCD, including pain triggers, pain is linked to

psychological distress, fear of dying and social isolation from their peers (Stegenga et al., 2004). Children discussed that pain was unpredictable and recurrent and that it interfered with their daily lives like planning or participating in social and physical activities (Panepinto et al., 2012; Stegenga et al., 2004). As play involves engaging in an activity for enjoyment and recreation, if SCD interferes with social and physical activities, it may also reduce opportunities for play.

Play is an important part of childhood because it contributes to key areas of child development including cognitive (e.g. creativity, imagination, problem solving), physical (e.g. motor skills and control of physical actions), emotional well-being (e.g. positive attitude and independence) and social skills (e.g. developing interaction with peers) (Ginsburg, 2007). Through play children also create and explore their own worlds, often practicing adult roles, for example, as a mother or in a career like a teacher or doctor (Ginsburg, 2007). Play is often restricted, especially by parents, to promote safety, but excessive safety concerns may paradoxically threaten development, as well as health, wellbeing and life opportunities (Brussoni et al., 2012; Wyver et al., 2010). For children with SCD this is likely to be a particular issue, and indeed some adolescents and adults with SCD recall having limited or disrupted physical activity and play, during their childhoods, which is partly because of their parents being overprotective (Forrester et al., 2015; Thomas & Taylor, 2002). Therefore, there is potential for SCD to impact important features of childhood.

Play activities may also be used to facilitate research with children, and drawing is one such activity. Evidence supports the use of drawings as a communication tool alongside interviews (Driessnack & Furukawa, 2012). Children who discussed their problems through 'drawing and telling' were found to provide twice as much clinically relevant information compared to 'telling' alone (Woolford et al., 2013). This methodological approach has been used in previous research with children with chronic conditions, including SCD. For example, Cotton et al (2012) asked children with SCD to draw things that help them cope with being sick. Stefanatou and Bowler (1997) asked children with SCA to draw two pictures; one of themselves and one of themselves in pain, to help understand their feelings about pain. They concluded that drawings and discussion was a useful approach in engaging with children as young as five years old, about their condition and how it affects their lives (Stefanatou & Bowler, 1997).

Qualitative research on paediatric SCD, including research using drawings, tends to focus on children with the condition. However, healthy siblings, who are comparable in terms of, for example, ethnicity and socioeconomic status, and may themselves experience some psychosocial problems (Hijmans et al., 2009; Lee et al., 1997), may form a representative comparison group. Their inclusion could help identify issues that are unique to children with SCD and also how healthy siblings' HRQL is affected by having a chronically ill sibling. This has not been explored using qualitative methods

and has only been examined in a quantitative study by Hijmans et al. (2010) who found that there was no significant difference between the HRQL of children with SCD and healthy siblings. This is in contrast to the abundance of quantitative research reporting that children with SCD have an impaired HRQL compared to healthy children more generally (e.g. Panepinto & Bonner, 2012).

Good management of SCD may support HRQL (Ahmed et al., 2015; Fisak et al., 2012). SCD is managed through regular hospitals visits, immunisations, blood transfusions but also at home, where adherence to medication is crucial. For example, to ease pain experienced during a vaso-occlusive crisis, painkillers are taken, and where children have recurrent vaso-occlusive crises they are prescribed daily hydroxyurea medication to help prevent further episodes and hospitalisations, and to improve HRQL (Dampier et al., 2002; Kanter & Kruse-Jarres, 2013). Additionally, some health behaviours may help manage symptoms. The behaviours that have been found to be most helpful are drinking water and avoiding strenuous or prolonged exercise, without precautions such as taking regular rest breaks, drinking extra fluids and staying warm by dressing in layers of clothing (Brown, 2012; Knight-Madden et al., 2011). Overexertion, dehydration and temperature extremes may trigger a vaso-occlusive crisis (Brown, 2012; NHS Choices, 2019) and increased fluids can help treat vaso-occlusive crises (Beyer et al., 1999). Research on health behaviours in children with SCD has shown that fewer children with SCD participated in daily vigorous to moderate/moderate physical activity for 60 minutes compared to healthy children and that they drank on average four glasses of water at home each day (Karlson et al., 2017). In a school environment, Omwanghe et al., (2017) reported that 90% of participants with SCD attended physical education classes, whereas Melo et al., (2018) found that they participated in significantly less physical education classes and sports compared to healthy children. Moreover, Dyson et al. (2010a) found that 46% of participants with SCD reported they were prevented from drinking water in class. Ievers-Landis et al. (2001) found that half of parents reported they frequently experienced difficulty getting their child to hydrate and take adequate rest to help prevent a vaso-occlusive crisis, although the authors did not explore an explanation for this finding. SCD itself may have an influence by potentially undermining health behaviours, but it is important that it does not hinder participation in suitable exercise, as the NHS recommends that children with SCD should exercise regularly to maintain good health (NHS Choices, 2019). However, the impact of SCD on engaging in health behaviours and the impact of health behaviours on managing SCD has rarely been examined. If children are able to have better control over their symptoms, for example, by keeping hydrated and avoiding excessive exercise, this may help reduce hospitalisations, school absences and psychosocial problems and improve HRQL.

The present study

The present study is the first to adopt a GT approach using qualitative methods to explore HRQL in children with SCD. Thus, unlike past research, which tends to be quantitative and assesses perceived current HRQL, this study examines children's perceived current and ideal selves. Children can potentially exert some control over SCD by engaging in health behaviours, and yet there is a lack of research in this area, so children's water consumption and exercise levels are also explored in the current study. Finally, a comparison group may be useful in identifying and understanding issues that are unique to children with SCD. Healthy siblings of children with SCD have rarely been studied. They are included in the present study as they provide a representative comparison group that may themselves be affected by their sibling's condition. Therefore, the present study explored the HRQL and health behaviours of children with SCD compared to health siblings, including any discrepancies between their current and ideal selves.

Materials and Methods

Design

Qualitative methodology using drawing tasks with semi-structured interviews was adopted. Thematic analysis was conducted and children with SCD and healthy siblings were compared.

Participants

A total of 150 children with SCD and their parents currently attending the haematology outpatient clinic at a London hospital were contacted by post and children and healthy siblings were invited to participate. Inclusion criteria required children with SCD to be aged 5-12 and have no co-morbidities that were not related to their condition as this may have affected their HRQL. Healthy siblings were required to be aged 5-12, have a sibling who was diagnosed with SCD and to 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 (78% with HbSS, which is generally the most severe form) and 14 healthy siblings was recruited. The mean age of children was 8 (*SD* = 2.31), 50% were male, all were black African or Caribbean, 57% of parents were married, 69% were employed and 44% had a degree. There were no differences between the two groups. These participants were the first to volunteer and the remaining children were not asked to take part as data saturation was considered to have been reached.

Procedure

After ethical approval was obtained for this study from the authors' Department's Ethics Committee and the participating NHS site, children and their parent were posted age-appropriate information

sheets to their home address one week prior to the start of data collection. Children and their parent were then approached during their outpatient clinical appointment in an NHS hospital. The researcher (the first author) read through the information sheets with all participants, including in person with healthy siblings, if they had attended their sibling's outpatient appointment, or over the phone, after gaining the parent's verbal consent to do so. The same process could not be followed for all children because healthy siblings were not patients and therefore did not always accompany their parent and sibling with SCD to the outpatient clinical appointment.

Before the semi-structured interview began, the parent was asked to provide written informed consent for their child's participation and complete a demographic information sheet and the child also gave written assent for their participation. Half of the interviews took place in a private room in the outpatient clinic and the rest in their home.

Only the researcher and child were present during the interview which took approximately 45 minutes to complete and all were digitally recorded. To aid communication, children were asked to draw a picture of themselves doing something they normally do, and then to draw a picture of themselves doing something they normally cannot do but wish they could. In both cases they were told this could be with family, friends or other people. This was followed by the interview (see Table 1 for example questions). Children were encouraged to speak openly, discuss their pictures and expand on issues that were important to them. After the interview, the researcher verbally debriefed all participants.

INSERT TABLE 1 HERE

Data analysis

Digital recordings were transcribed verbatim by the researcher and thematically analysed with the assistance of QSR NVivo 10. Thematic analysis (Braun & Clarke, 2006) is a flexible technique which is not set to any pre-existing theoretical framework, it is well-suited to large datasets and can help to highlight similarities and differences across the dataset, which helped in the present study 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 researcher took a realist perspective and an inductive approach, identifying semantic themes across the dataset (Braun & Clarke, 2006).

The six stages suggested by Braun and Clarke (2006) were used for analysis. Stage 1 involved familiarisation with the data where the researcher transcribed and then actively read, and re-read the data and repeatedly viewed the drawings to become familiar with issues arising from the children.

Stage 2 involved the researcher generating initial codes i.e. coding interesting features of the data and then identifying data extracts to support these codes. Stage 3 involved the researcher beginning to arrange the initial codes into potential themes and collating the supporting data extracts. During stage 4, 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. The second and third authors also reviewed the codes and themes to ensure that they represented the data. The penultimate stage involved defining and naming themes, and ensuring that the themes were representative of the data extracts and children's drawings. The final stage involved assigning all participants pseudonyms, ensuring that there was a coherent story across each theme and subtheme and selecting data extracts and drawings to reflect the story.

Results

Thematic analysis identified two themes; limitations of SCD and adjusted expectations (exercise and recreational activities, psychological well-being and education and ambitions), and coping with SCD (social support and health behaviours as prevention and coping).

Limitations of SCD and adjusted expectations

Children identified some limitations in their lives including less participation in exercise and recreational activities, as well as some affects of SCD on their psychological well-being, education and ambitions, which created some discrepancies between their current and ideal selves, but children with SCD also appeared to have adjusted expectations.

Exercise and recreational activities. Both groups of children reported that they participated in some types of exercise during school but healthy siblings were more likely to also participate in exercise in their leisure time.

> 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)

Many children with SCD described being unable to participate in exercise to the same level as their peers, which made them feel different from their friends and, at times, meant that they could not be part of school teams. This created some discrepancy between their current and ideal selves, for

example, in his current self drawing Jaheem was playing football on his PlayStation, while in his ideal self drawing he was outdoors playing football with his friends.

> 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)

Children with SCD were concerned that participating in exercise and sports that are particularly physically demanding, like running, football, rugby or basketball, especially in cold environments may trigger a vaso-occlusive crisis, and some children had experienced this. Children with SCD generally reported that their mothers discouraged them from participating in exercise for this reason. This exacerbated discrepancy between their current and ideal selves.

> 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)

However, even in their ideal self drawings some children could not imagine themselves without the limitations of SCD and expressed apprehension about doing new activities. For example, in Oria's current self drawing she is playing with her friends (Figure 1) whilst in her ideal self drawing she is trying to do a cartwheel by herself (Figure 2).

> 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! (Oria, SCD, female, aged 6)

INSERT FIGURES 1 AND 2 HERE

In contrast, healthy siblings reported that their parents encourage them to participate in exercise because it is beneficial to their physical and mental health.

> 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)

However, healthy siblings also reported limitations to participating in exercise as a result of their sibling's condition, such as not being able to play with their sibling when they are ill or in hospital or parents being too busy to take them to sports activities due to caring for the sibling with SCD.

> 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. (John, sibling, male, aged 12)

Nevertheless, healthy siblings were generally satisfied with their level of engagement in exercise, compared to children with SCD who talked of wanting to do cartwheels, or play football or basketball. Instead healthy siblings often spoke about wanting to do more exciting recreational activities. For example, healthy siblings' ideal self drawings included them and their family meeting Mickey Mouse in Disneyland (Rihana, sibling, female, aged 6), going to LEGOLAND (Oban, sibling, male, aged 9) and going on a rollercoaster at Alton Towers (Elizabeth, sibling, female, aged 12).

Psychological well-being. Children with SCD generally do not allow their condition to monopolise their lives and some even discuss some positive aspects of receiving treatments and 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)

In fact, with one exception, SCD children's current self drawings generally did not represent 'sickness', nor did they draw themselves without SCD in their ideal self drawings. They showed evidence of having normalised and accepted this life-long condition.

> 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)

In contrast, healthy siblings experienced anxiety about their sibling with SCD when they go to hospital or experience pain.

> 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. (Ibrahim, sibling, male, aged 10)

Although children with SCD seem to have normalised their condition, one area that may be problematic for them is body image. Most children, especially boys, with SCD spoke about how their physiques were affected, often describing themselves as small, short and weak compared to their friends. This is reflected in David's (SCD, male, aged 9) current self drawing (Figure 3) 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 (Figure 4) David was taller than his friend and was leaping over him.

> 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 and he don't…….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 strong in this one! (David, SCD, male, aged 9)

INSERT FIGURES 3 AND 4 HERE

In contrast, healthy siblings were more positive when they discuss their body image ideals such as expressing that their growing physique could lead to a career.

> I like doing running, hula hoops, football and basketball in school and press ups so I get bigger muscles and sometimes when I sleep my muscles grow bigger! Then I can be a policeman or or a fireman and save people! (Adebayo, sibling, male, aged 6)

Education and ambitions. While a few children with SCD described missing substantial periods of school due to feeling sick, vaso-occlusive crises and hospitalisations, most children reported that the condition had little impact on their education besides experiencing the occasional day of disruption. Nevertheless, relatively few children with SCD discussed current successes or future ambitions. Where they did, 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, healthy siblings do not have to contend with the limitations of SCD, so they were more ambitious and had already achieved some success which may help them in the future. They reported they were facilitated in their achievements by, for example, their parents providing them with private tutors (which was not reported by children with SCD). For example, Aisha's (sibling, female, aged 9) current self drawing showed her playing the trumpet, for which she won a scholarship from the Major of London, and her ideal self drawing showed her medical career aspirations. The lack of limitations of SCD may explain why healthy siblings reported having higher ambitions compared to children with SCD.

> 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)

Coping with SCD

Children discussed the different strategies used to cope with the condition including social support, religious beliefs and using health behaviours to aid the day-to-day management of their condition and to help them during a vaso-occlusive crisis.

Social support. Children with SCD reported that their mothers provided both instrumental support by managing their day-to-day treatment and emotional support when they were 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)

Nonetheless, children with SCD and healthy siblings generally did not report discussing the condition or their feelings about it with anyone, even though they discussed other issues such as arguments with friends. There were relatively few circumstances where children with SCD reported

requiring support for their condition from their friends, but 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)

One reason for not seeking social support may be that they have a limited social network from which to seek support, as many of their friends and family were not aware of their condition. 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)) [my 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 and family may gossip about them, be mean or react badly. Children were often encouraged by their mothers to keep their condition a secret, and this appeared to result from concerns about stigma.

> 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 operating around stigma and reluctance to disclosure. For example, in certain cultures it is less acceptable to discuss 'disease', even amongst family.

> 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)

Healthy siblings did not speak about social support but they did find their religious beliefs to be a comfort in coping with their sibling's illness which was often influenced by their parents and grandparents. This was not discussed by children with SCD.

> No she [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)

Health behaviours as prevention and coping. Children with SCD demonstrated a good understanding of what they should be doing (e.g. drinking water, taking medication, avoiding overexertion without precautions and the cold) and how their actions could trigger a vaso-occlusive crisis. Healthy siblings also had a good understanding of what their sibling should be doing.

> 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)

> [He] 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)

As discussed under the exercise and recreational activities subtheme, children with SCD reported that mothers discouraged them from participating in exercise to help prevent a vaso-occlusive crisis, but this was also seen by children as key to helping to cope with a crisis.

> 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)

Some children with SCD reported that their school was also aware that overexertion or playing outdoors when it is cold may adversely affect their condition, however, other children with SCD reported that their school lacked knowledge about this, or at least did not enforce it.

> 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)

Furthermore, children reported that at school everyone was encouraged to have a water bottle in class, but some children with SCD reported that their school made additional allowances for their condition, for example, some teachers reminded them to drink water 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)

This was not always considered to be positive as there were also some children who explained that they did not want to draw attention to themselves by being seen to receive special treatment from the teacher.

> 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)

In contrast, some children reported that their school restricted when they were allowed to drink water and use the toilet, even when they were aware of their condition, so 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)

At home, children reported that water consumption was not notably encouraged. However, children with SCD were aware that drinking water may help them to cope with a vaso-occlusive crisis, but most instead had drinks that they enjoyed and found comforting. Some children, such as Emmanuel reported avoiding water in favour of a more comforting drink.

> 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. (Emmanuel, SCD, male, aged 11)

Discussion

The present study provides a unique contribution to the literature by taking a GT approach (Calman, 1984) to explore HRQL and health behaviours, including any discrepancies between the current and ideal selves, of children with SCD compared to healthy siblings. There were some discrepancies between the current and ideal selves of children with SCD and healthy siblings but these differed between the two groups. Additionally, children with SCD drew on social support from their mothers and on health behaviours to cope with SCD, while healthy siblings used their religious beliefs.

Children with SCD appeared to have little discrepancy between their current and ideal selves, except in the physical functioning aspect of their HRQL, where they described participating in exercise during school time but wished they were able to do more in their leisure time. Melo et al., (2018) found that children with SCD were significantly less likely to participate in leisure time physical activity than healthy peers. In the current study, some children with SCD were apprehensive about participating in exercise and sports, especially outside when it was cold. 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 realistic regarding their physical abilities and the limitations of their condition, and in this study some were also concerned about participating in new activities even in their ideal self scenario. This suggests that children may be realistic that these limitations will always be part of their lives and have adjusted expectations accordingly, which may reduce the gap between their current and ideal selves.

Healthy siblings generally did not have a discrepancy between their current and ideal selves in the physical functioning domain of their HRQL but their expectations appeared to be higher. They reported that both they and their parents had high expectations for their futures and they wanted to participate in more exciting recreational activities. Siblings of children with chronic conditions sometimes described feeling 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). They also experienced some anxiety about their siblings' condition (supporting Plumridge et al., 2011). Altogether, this suggests some discrepancy in the psychosocial domain of healthy siblings' HRQL. Past research has found a higher prevalence of depression among healthy siblings than children with SCD (Lee et al., 1997).

Some children with SCD expressed feeling dissatisfied with their body image as a consequence of growth failure (supporting Bennett, 2011). However, other than this, in the psychosocial functioning domain of HRQL, they did not seem to be greatly affected by their condition. This is consistent with earlier studies that have taken a gap approach, where children with SCD did not have a lower discrepancy QoL compared to their healthy peers (Constantinou et al., 2015). For example, in this study, compared to healthy siblings, they did not have the same ambitions for the future. They and their parents may approach life on a day-to-day basis rather than focusing on the future. They may also have adjusted, and possibly more realistic ideals and expectations, as well as different priorities (Eiser et al., 2000), thus reducing discrepancy between their current and ideal selves. There is evidence from the present study that children with SCD have normalised their condition, do not see it as the focal point of their lives and do not see themselves without it even in their ideal self scenario.

In order to deal with the physical consequences of SCD, children with SCD sought support from their mothers during a crisis, but overall they did not report seeking emotional support to discuss their feelings about their condition. If they have normalised their condition they may not feel the need to do this because it is not something out of the ordinary that requires discussion. However, Patel and Pathan (2005) revealed that children with SCD were concerned that their condition was a burden for their immediate family members and therefore may intentionally not seek this kind of support. The present study suggested that social support options available to children may also be limited because of a reluctance to disclose. Mothers' reluctance may be related to their own difficulty accepting SCD or feelings of guilt for passing the condition to their child (Thomas & Taylor, 2002). It may also be because culturally or socially there is more responsibility placed on mothers than fathers for SCD (Marsh et al., 2011), or due to general concerns about stigma, especially in African communities (e.g. Burnes et al., 2008). There may also be a reluctance to disclose chronic diseases in these communities because of reprisals or cultural beliefs (Barbarin & Christian, 1999).

Healthy siblings do not have to focus on dealing with the physical symptoms of the condition and may have similar difficulties in seeking social support, so religion, which is an important aspect of many African people's lives and identity (Clayton-Jones & Haglund, 2015), provided them with comfort in the current study. Previous research has shown that children with SCD use religion as a coping mechanism but also that some believed their condition was a punishment from God (Cotton et al., 2012). However, religion was not raised by children with SCD in the present study.

Additionally, children with SCD reduced any type of exercise during a vaso-occlusive crisis, but they did not drink more water, despite understanding the importance of hydration. This is consistent with past evidence which found that children with SCD do not hydrate during a vaso-

occlusive crisis but instead were more likely to use pain medication (which is a recommended treatment for vaso-occlusive crises) or diversional activities (Maikler et al., 2001). The same has been found among adults (Okomo & Meremikwu, 2012), suggesting that childhood behaviours may be transferred into adulthood, although research in older children suggests they may develop more effective coping strategies with age (Panepinto et al., 2012).

Health behaviours were also preventative. Within a school setting there were a diverse range of experiences. Some children with SCD disclosed that their schools were aware that some types of exercise may affect their condition, that teachers reminded them to drink water, and that drinking water is generally encouraged. However, some children also reported that they did not like receiving special treatment (supporting Panepinto et al., 2012). Other children revealed that they were unable to drink water and use the toilet when needed (supporting Dyson et al., 2010a; 2010b) and their schools did not limit physical exertion. Similarly, Dyson et al., (2010a) reported that approximately a third of participants with SCD 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 and an individual healthcare plan is then devised (Department for Education, 2015). It may be that some parents in the current study did not disclose their child's condition, or for those who did, Dyson et al., (2010b) postulate this may not always be communicated to all staff or perhaps staff forget or do not have the time or sufficient knowledge to take action.

Parents discouraged children with SCD from participating in exercise. This is in line with retrospective studies of adults with SCD where parent overprotection during childhood limited their involvement in physical activities (Forrester et al., 2015; Thomas & Taylor, 2002). Parental protectiveness is understandable given the potential risks. However, the condition should not undermine their participation in moderate exercise, which is good for general health (NHS Choices, 2019). Retrospective studies in adults with SCD revealed that parental protective behaviours during childhood negatively affected their self-care behaviours, health outcomes and QoL (Jenerette & Murdaugh, 2008; Jenerette & Valrie, 2010). Furthermore, in this study, leisure-time physical activity was considered "having fun and play time" by children. Thus, to the extent that SCD hinders physical activity, it may also hinder play. Participating in different types of play in different settings is a crucial part of children's cognitive, physical, emotional and social development (Ginsburg, 2007) and excessive restrictions due to safety concerns may threaten this (Wyver et al., 2010). It has been suggested children in general should be kept *as safe as necessary* rather than *as safe as possible* (Brussoni et al., 2012), which has implications for the management of SCD.

Implications

In the present study there was some discrepancy between the current and ideal selves of children with SCD which was influenced by their desire to participate in more physical activities. Exercise is important to children both for their HRQL and general health, so a balance needs to be struck. The findings also suggest that whilst parents, children and schools were aware (to varying degrees) of healthy behaviours for children with SCD, these were not routinely followed. Therefore, it is necessary to highlight the importance of the role of health behaviours at home and school, and that healthcare plans in schools are implemented and followed. Communication and support are also important issues. Consideration needs to be given to how communities can reduce stigma and enable greater support for families, which, in particular, should also include healthy siblings.

Limitations and future research

There are some limitations of this study that highlight some avenues for future research. First, while the sample size was relatively small, this reflects a qualitative approach which facilitated the children expressing themselves openly through drawing and spoken narratives, which is a strength of this study. Nevertheless, the sample was from one hospital, and was not ethnically diverse. In England SCD is most prevalent in people of Black African or Caribbean origin (Streetly et al., 2010), as reflected in the present sample, but it can occur in other groups (Serjeant & Serjeant, 2001). Additionally, the sample did not include adolescents who have also been found to have an impaired HRQL compared to their healthy peers (Panepinto & Bonner, 2012). Second, a convenience sample was recruited during children's outpatient appointments. Individuals who attend regular hospital appointments may have a better understanding of SCD, greater levels of adherence and consequently adopt healthier behaviours and have a better HRQL. Third, the interview schedule delved into a number of areas of HRQL and health behaviours. As a result of this broad focus, but also due to interviewing young children, whose developmental stage means they tend to observe things about the world in quick succession, in-depth exploration of some matters were beyond the scope of this study. However, issues arising from the children's interviews and drawings, which have not been examined by the abundance of previous quantitative research, could be pursued with a sharper focus in further qualitative research. For example, children with SCD seemed to have adjusted expectations and ambitions. Further research could investigate whether this is a self-protective strategy as well as what causes this and how this affects them. Furthermore, the relationship between health behaviours (including those other than exercise and hydration) and HRQL could also be further investigated using quantitative methods. This would enable examination of whether relationships exist between

HRQL and objectively assessed health behaviours, as well as allowing data to be collected from a large, more diverse group of children, which would facilitate generalisability of findings.

Conclusion

Children's interviews and drawings revealed some discrepancy between the current and ideal selves of children with SCD in the physical functioning domain of HRQL, and in the psychosocial functioning domain for healthy siblings, providing some support for GT (Calman, 1984). While SCD may hinder some physical activity, and hence some opportunities for play, children with SCD appear to have normalised their condition and adjusted their expectations accordingly, thus reducing discrepancy. There is an assumption that children with chronic conditions live unhappy, restricted lives, and some previous, mainly quantitative research on paediatric SCD supports this. However, children with SCD in this study generally portray themselves as happy and well adjusted, with healthy siblings expressing more anxiety about their sibling with SCD. As the sample in this study was primarily of Black African or Caribbean descent, there may be some influence from the children's cultural backgrounds, such as a reluctance to disclose and discuss health issues, and thus less opportunities for social support and greater reliance on religious faith. However, modifying health behaviours, such as participating in reduced exercise compared to healthy siblings, was also used to help cope with SCD. However, it is important to strike a balance, so that reductions in physical activity do not impair HRQL.

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Table 1. Semi-Structured Interview Schedule

Example of Questions
1. 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?*
2. How are you feeling? 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?
3. 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?
4. How do you feel about school? 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?
5. What's it like having SCD? *If they talk about feeling ill, pain, going to the Doctor explore how often, how they feel, what happens.* OR How do you feel about having a brother or sister who has SCD? How does it affect you?
6. Have you told people like your friends that *you/your* sibling has SCD? Why/why not? How did they react? How did you feel about that? Have they asked you questions?
7. 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? 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?
8. Can you tell me about what you drink? What do/don't you like drinking? What do you drink at home/school/*hospital*? Are there things you can't drink (things that maybe your friends can do)? Who tells you that you shouldn't have this? If you feel ill or tired or have pain what things can/can't you drink? Are there things you would like to drink? If you don't drink does that ever make you tired/feel ill/pain?

Figure 1. Oria's current self drawing (SCD, female, aged 8).

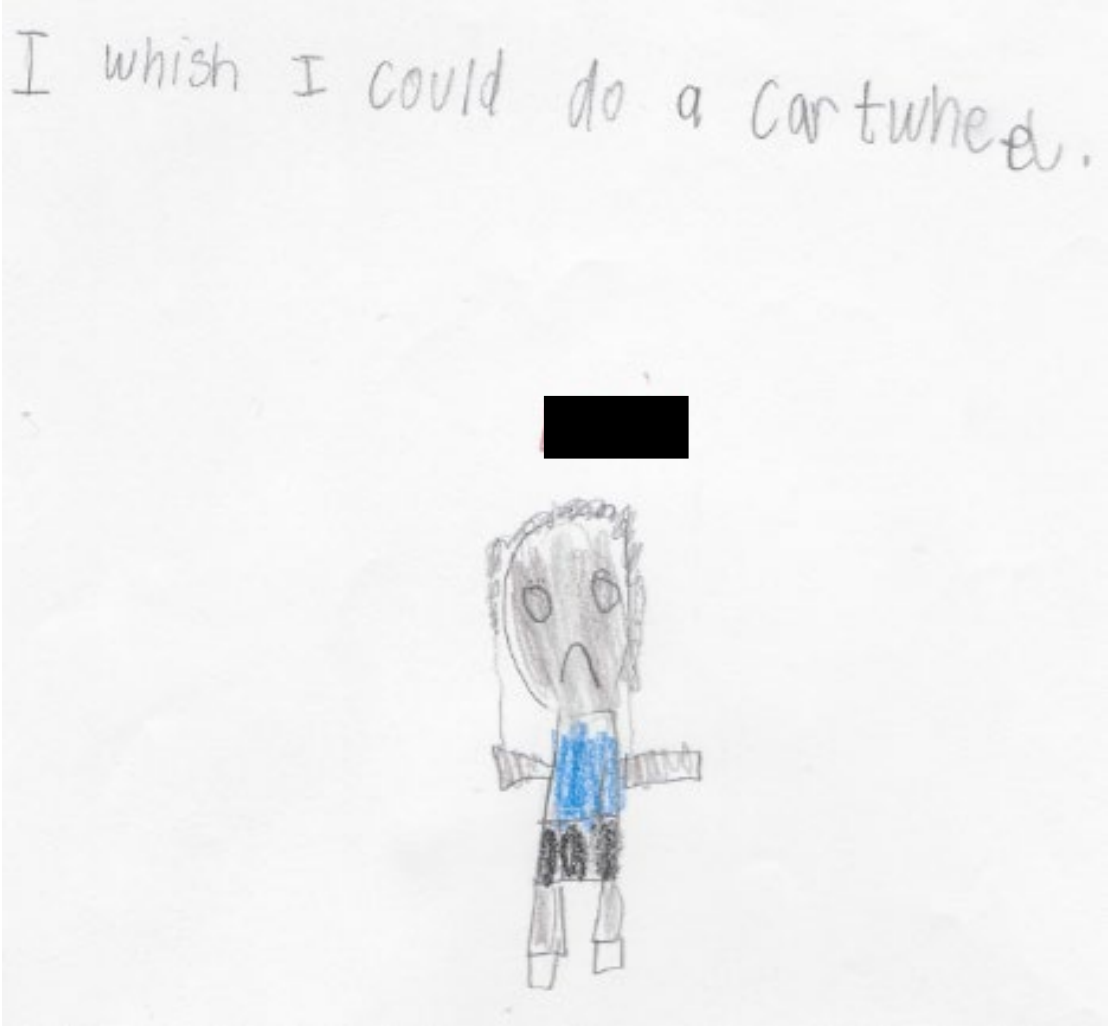

Figure 2. Oria's ideal self drawing (SCD, female, aged 8).

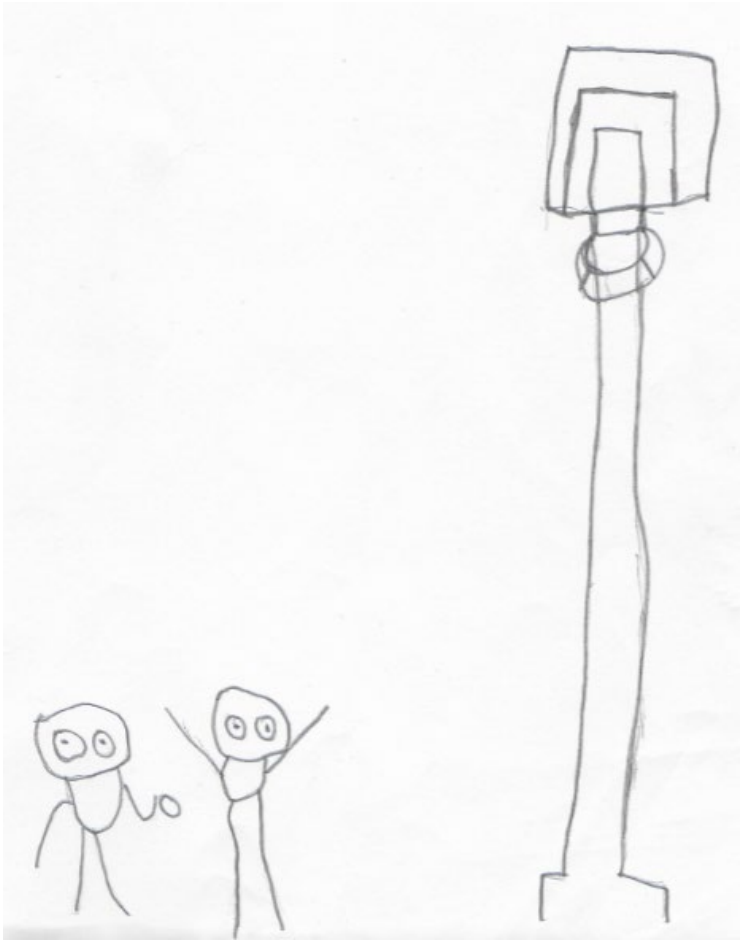

Figure 3. David's current self drawing (SCD, male, aged 9)

Figure 4. David's ideal self drawing (SCD, male, aged 9).