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Exploring health-related quality of life, exercise and alcohol use in adolescents with sickle cell disease and healthy siblings

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ABSTRACT

Objective: This study explored the health-related quality of life (HRQL) and health behaviours of adolescents with sickle cell disease (SCD) and healthy siblings, drawing on Gap theory which suggests HRQL is the discrepancy between current and ideal selves.

Design: Twenty-three adolescents with SCD and 21 healthy siblings aged 13 to 17 years participated in eight focus groups.

Results: Thematic analysis identified three themes: learning to accept SCD, coping with SCD and influences on health behaviours. Adolescents appear to have normalised and adapted to SCD. Adolescents with SCD have learnt effective coping strategies, such as moderating engagement in exercise. Unlike heathy siblings, they were not encouraged to exercise by parents but were content with their level of participation. Both groups were influenced to exercise by role models or wanting to socialise, and influenced to drink alcohol by peers, but there was limited understanding of the potential negative impacts of alcohol on SCD.

Conclusion: There does not appear to be a discrepancy between adolescents' current and ideal selves, providing optimism about their HRQL. Further consideration of engaging in healthy behaviours is needed, but it is important to strike a balance so that modifications to lifestyle do not impair HRQL.

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Adolescents; chronic illness; health-related quality of life; qualitative methods; exercise; alcohol use

Introduction

Sickle cell disease (SCD) is a lifelong genetic red blood cell disorder (Ware et al., 2017). It is common, with approximately 14,000 people living with SCD in the UK (Dormandy et al., 2018) and is prevalent in people of Black African or Caribbean descent (Streetly et al., 2010). In adolescents, SCD has some debilitating consequences such as increased vaso-occlusive crises (severe acute painful episodes), growth failure, delayed puberty and dermatological manifestations (Bennett, 2011; Knight-Madden

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et al., 2011). A review of research on adolescents' experiences of living with SCD suggests that the condition affects their school experience due to absenteeism, treatment and fatigue; their emotional well-being and coping, including greater levels of depression and anxiety; and their social relationships with their family members, who are their primary means of support (Poku et al., 2018). This suggests SCD impacts health-related quality of life (HRQL).

HRQL or quality of life (QoL) are often used interchangeably (Taylor et al., 2008). This is not recommended in paediatric research (Taylor et al., 2008), because QoL is rated higher (better) than HRQL and different variables predict each concept (Feldman et al., 2000). QoL is a broad, multidimensional concept of well-being. HRQL is also multidimensional but has a focus on the impact of an individual's health condition, so includes physical functioning, and also how a condition may affect psychosocial functioning including social, emotional and school functioning (King & Hinds, 2012; Panepinto et al., 2005). In medicine and associated peer-reviewed research, the term HRQL is preferred to QoL (Rapley, 2003), and a review found that the term HRQL is generally adopted in paediatric SCD research (e.g. Panepinto & Bonner, 2012). However, where previous research has used the term QoL, this has been maintained in the discussion of the literature below.

Quantitative research frequently uses the Pediatric Quality of Life Inventory Generic Core (PedsQL™) and the Pediatric Quality of Life-Sickle Cell Disease Module (PedsQL™ SCD Module) to assess HRQL in SCD (Stokoe et al., 2022). This research has consistently demonstrated that children and adolescents with SCD have a significantly lower (worse) HRQL than healthy peers (Panepinto & Bonner, 2012; Stokoe et al., 2022), and also that HRQL is lower if the child is older, female, has greater disease severity, greater pain, and more frequent hospitalisations, fatigue and school absences (Stokoe et al., 2022). Self-management interventions have shown minimal effectiveness in improving HRQL/QoL in children and young people with SCD, but where there were improvements, this was only in the social domain (Poku et al., 2022).

While most research focuses on current HRQL, some definitions of QoL or HRQL suggest that the gap between current and ideal selves is central. For example, the World Health Organization (WHO) refers to QoL 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). Calman's (1984) Gap Theory (GT) suggests 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.

Similarly to QoL, some researchers have argued that HRQL should examine the discrepancy between an individual's perceived current and ideal selves (Panepinto & Bonner, 2012), but research on QoL or HRQL has rarely taken a GT approach. However, quantitative research applying GT has explored adult mental health using a measure called the QoL-GAP to assess discrepancy between actual and ideal selves (Welham et al., 2001). In children and adolescents, GT has been applied in asthma (Eiser et al., 2000), SCD (Constantinou et al., 2015) and chronic kidney disease (Heath et al., 2011) using a measure called the Generic Children's Quality of Life Measure

(GCQ; Collier et al., 2000). The GCQ refers to a hypothetical group of girls/boys with two sets of questions: the first set asks the child to select the girl/boy who is most like them (their current self), and the second set is the same questions that ask the child to select the girl/boy who they would most like to be like (their ideal-self) (Collier et al., 2000). The discrepancy between the two sets of questions assesses the 'gap' in QoL (Collier et al., 2000). These studies applying GT found that the QoL scores of children and adolescents with chronic conditions and healthy peers were not significantly different, so the gap between current and ideal selves was similar across groups in all studies. However, they did not use measures of HRQL and were all quantitative studies. A qualitative study applying GT to HRQL, children with SCD aged 5 to 12 years old raised issues about their body image (being small and weak) and participating in less exercise than they would like, but overall they appeared to have adjusted their expectations, thus reducing discrepancy between their current and ideal selves (Constantinou et al., 2021). However, this study did not include adolescents. It is important to also study adolescents because their SCD symptoms, for example, vaso-occlusive crises, and health care utilisation tend to increase (Theodore et al., 2015), which may reduce HRQL (Stokoe et al., 2022). Adolescence is also a period of other age-related changes and difficulties; from biological (i.e. puberty) and cognitive to psychosocial and emotional (Swanson et al., 2010).

Healthy siblings may also suffer from psychosocial problems (Hijmans et al., 2009). They have a comparable HRQL to children and adolescents with SCD (Hijmans et al., 2010). They experience some anxiety about their sibling with SCD and some limits to their recreational activities but also appear to have higher expectations than children with SCD (Constantinou et al., 2021). Again, this study did not include adolescents, but the inclusion of healthy siblings may help identify issues that are unique to adolescents with SCD. Stokoe et al. (2022) postulate that the inclusion of comparison groups, such as healthy siblings, in paediatric SCD research would be a strength.

During adolescence teenagers generally gain more independence and take more responsibility for their lives, including their health (Poku et al., 2018). Good management of SCD is crucial and this would include engaging in or moderating health behaviours (Ahmed et al., 2015; Crosby et al., 2022; Fisak et al., 2012). For example, limiting alcohol consumption may be important as it can cause dehydration and increase the risk of a vaso-occlusive crisis (Brown, 2012). Similarly, as overexertion, temperature extremes and dehydration may trigger a vaso-occlusive crisis (Brown, 2012), care must be taken with strenuous and prolonged exercise (Brown, 2012; Knight-Madden et al., 2011). Regular hydration is also important to prevent a vaso-occlusive crisis (Brown, 2012). A review found that adolescents with SCD are aware that regular hydration and avoiding strenuous exercise and risky behaviours can benefit their condition (Poku et al., 2018). This could help adolescents have some control over SCD, avoid exacerbating symptoms, and thus may improve HRQL. Indeed, Mahdy Fouda et al. (2021) found that a self-management intervention was associated with improvements in disease knowledge and self-care practices, such as nutrition and pain management, and improved QoL in adolescents with SCD. However, in a review of self-management interventions in children and young people, improvements in HRQL or health behaviours were not consistently demonstrated (Poku et al., 2022).

One possible reason for the inconsistent link between self-management interventions and HRQL in the Poku et al., (2022) review is that self-management that involves moderating certain behaviours may also have unintended consequences. For example, exercise is important for good health (World Health Organization, 2020) but children and adolescents with SCD participate in less daily vigorous and moderate exercise for 60 min compared to healthy children and adolescents (Melo et al., 2018; Omwanghe et al., 2017). Furthermore, adolescents reported that SCD disrupts aspects of their lives such as participating in physical and social activities (Atoui et al., 2015; Panepinto et al., 2012), and they strive for normality (Poku et al., 2018). Similarly, children with SCD participate in less exercise than they would like compared to healthy siblings, and their parents are overprotective (Constantinou et al., 2021). Therefore, an unintended consequence of moderating certain behaviours may be that it creates a discrepancy between perceived current and ideal selves, as suggested by a GT approach to HRQL, and therefore HRQL may be undermined.

There is an abundance of research examining HRQL and SCD, which is largely quantitative (top–down), however the present study sought to explore adolescents' experiences from their perspective. This is especially important as there is limited qualitative research on children and adolescents with SCD that has a specified HRQL underpinning to explore the experience of living with SCD (e.g. Panepinto et al., 2012; Stegenga et al, 2004) and no research that focuses on both HRQL and health behaviours in both adolescents and healthy siblings. There is also no research examining the HRQL of adolescents with SCD through a GT lens. This is a novel approach and thus adopting a qualitative (bottom-up) approach is appropriate. Furthermore, with some exceptions (e.g. Porter et al., 2014), research on adolescents with SCD has used interviews. Using focus groups may help adolescents feel more comfortable and able to share their views and personal stories about living with SCD, since speaking with peers is a more natural situation than a one-to-one interview with an adult (Heary & Hennessy, 2002; Peterson-Sweeney, 2005). In addition, group discussions allow for different forms of communication that occur in day-to-day life such as joking and anecdotes, producing richer data (Wong, 2008). Therefore, the aim of the current study was to use focus groups to explore the HRQL and health behaviours of adolescents with SCD compared to healthy siblings, including any discrepancies between their current and ideal selves.

Methods

Participants

A total of 270 adolescents with SCD aged 13 to 18 were invited to participate in the study. The healthy siblings of these adolescents were also invited. Adolescents with SCD had to be attending the haematology outpatient clinic at one of two London hospitals. Having co-morbidities could have an impact on HRQL, so, with the support of medical staff, their medical records were checked to confirm that they did not have any physical health conditions, such as cancer, diabetes, HIV or cystic fibrosis, other than those associated with SCD, and also that they did not have any diagnosed psychological conditions. Parents of healthy siblings (and adolescents with SCD) were also asked to confirm that their child had no diagnosed physical or psychological health conditions.

In total, 26 adolescents with SCD came forward and 23 participated, and 23 healthy siblings came forward and 21 participated. Three participants were not able to attend the focus group (due to a vaso-occlusive crisis, a bereavement and a job interview), and two did not participate because no further groups were being organised. Participants who were the first to respond to the invite and provide consent were included. Recruitment was closed after the number of participants noted above had come forward, as it was felt that data saturation had been reached during the focus groups that were underway. Participants were not all related; some healthy siblings participated and their sibling with SCD did not and vice versa. Demographics of adolescents and their parent who gave consent are shown in Table 1. The vast majority of parents were female, around half were married, three-quarters were employed and two-fifths had a degree. Half the adolescents who participated were female, the adolescents' ages ranged from 13 to 17 years old with one-third aged 16 to 17 years old, and more than three-quarters had the most severe form of SCD.

Procedure

Once ethical approval was granted by the authors' department's ethics committee and participating NHS hospitals, adolescents and their parents were posted information sheets. They were then approached by the researcher (the first author) during their next routine outpatient appointment where the researcher read through the information sheet. If a parent said a healthy sibling who did not attend the appointment may be interested in participating, the researcher discussed the research with the healthy sibling by phone. Parents provided written consent for adolescents aged 13 to 15 years old and adolescents aged 16 years or older provided their own written consent. Parents (or adolescents aged 16 years or older) completed a demographic information sheet.

Table 1. Demographic characteristics of adolescents with SCD and healthy siblings.

	Adolescents with SCD M(SD) or % n	Healthy siblings M(SD) or % n
Adolescent		
Age	14.87(1.39)	14.81(1.44)
Gender	48% Female	52% Female
Ethnicity	83% Black African, 17% Black Caribbean	81% Black African, 19% Black Caribbean
Education	78% School/Full-time, 22% College/Full-time	71% School/Full-time, 24% College/Full-time, 5% Apprenticeship
SCD phenotype	78% HbSS, 22% HbSC	100% HbAA
Parent		
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
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

Note: HbSS refers to sickle cell anaemia which is the most prevalent, and severe form of SCD; HbSC refers to haemoglobin SCD which is a milder form of the condition.

Focus groups took place in a private outpatient clinic room. There were between five and six adolescents in each group. The groups were same sex and four groups comprised adolescents with SCD and four comprised healthy siblings. Adolescents who were related, friends or acquaintances did not participate in the same focus group. Recruitment and data collection took place over a 3-month period before the COVID-19 pandemic. The researcher (primary author) who is experienced in conducting focus groups with adolescents, facilitated discussions which ranged from 73 to 96 min and were audio recorded and transcribed verbatim.

Table 2 shows focus group questions. This was used as a guide but adolescents were encouraged to explore issues or areas that resonated with them and thus additional and follow-up questions varied by group (Willig, 2013). GT (i.e. current vs. ideal selves) was used to inform the way some of the questions around aspects of HRQL were asked, including what they do in their free time, their feelings, and the social and school aspects of their lives. In these areas, participants were asked if there were things they would like to do but cannot and things they would like to change. Participants received 20 GB pounds to cover costs of travel and as an acknowledgement of their time.

Data analysis

Focus group transcripts were thematically analysed with the aid of QSR NVivo 10. Thematic analysis (Braun & Clarke, 2006) is a flexible technique, is not bound to any pre-existing theoretical framework, and is appropriate for larger datasets. It provides a detailed description of the dataset, allows for the identification of patterns and interpreting the meaning of these patterns, and can highlight similarities and differences across the dataset (Braun & Clarke, 2006). The researcher took a critical realist epistemological position, with an inductive approach to identify

Table 2. Example focus group questions.

1. What kinds of things do you do in your free time? Who or what influences what you choose to do and why? 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? Who or what stops you doing these things? How does that make you feel?
2. Tell me about the important people in your lives? How do they help you? How do they make you feel? Is there anything you'd like to change about the people in your lives or the relationships you have with people? In what ways would this make things better for you?
3. How do you feel about your school? What are the people like? What are the positives and negatives of school? Are there things you would like to do at school but can't? Do you ever miss school or struggle to get work done? Why? How does this make you feel? Are there things you would like to change about school. What?
4. How do you feel emotionally in your day-to-day lives, e.g. what kind of things make you feel happy or sad or worried? Who would you speak to? How do you feel physically in your day-to-day lives? Does being unwell or pain interfere with any part of your life?
5. How does having SCD/*having a sibling with SCD* affect your day-to-day lives? Who knows that you/*your sibling* have SCD? What do your friends know about SCD? Are there things you think they don't understand? Do they treat you the same as their other friends?
6. What types of physical activity do you participate in? Who or what influence the types of physical activities you do? Are there things you'd like to do but can't do, e.g. maybe things your friends can do? Why? How does this make you feel? How do you feel after participating in physical activity?
7. Can you tell me about what you normally eat and drink? Who or what influences what you eat or drink? Are there things that you can't eat and drink and would like to? Does anything you eat or drink make you feel ill?
8. What do you know about alcohol, smoking and other substances? Can you tell me about any experiences of trying these? Who or what influenced your decision to try these? 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 physical activity and drinking alcohol? Do you think they have an affect on your SCD?

semantic themes, since this form of coding focuses on the explicit and surface meanings within the data (Braun & Clarke, 2006). This position assumes that through their language participants can convey their meanings and experiences (Braun & Clarke, 2006). As discussed earlier, GT was used to inform the way some of the questions were asked, but an inductive approach to analysis was applied. As part of the thematic analysis, codes relating to discrepancy between adolescents' current and ideal selves were sought, but it was felt that it was important to allow adolescents to speak for themselves and ensure aspects of their experience of living with SCD were not missed, so a deductive approach, imposing a pre-existing theoretical framework was not taken. Data were analysed across all participants, although any differences in views and experiences of adolescents with SCD and healthy siblings were drawn out.

The thematic analysis was based on Braun and Clarke's (2006) six stages. The researcher became familiar with the data by transcribing, and then reading and re-reading the transcripts. Initial codes were generated and the researcher began to arrange these into potential themes. The second and third authors reviewed the codes and potential themes alongside the transcripts. Disagreements between authors were resolved through discussion until agreement was reached. The researcher reviewed the agreed themes in relation to the coded extracts and the whole dataset. The researcher ensured there were clear definitions and names for the themes and subthemes before writing up the themes supported by compelling data extracts. Participants were given pseudonyms.

Results

Three themes were identified; learning to accept SCD (including two subthemes: daily life with SCD and normalising SCD), coping with SCD (including two subthemes: emotional support and moderating health behaviours) and influences on health behaviours (including two subthemes: exercise and alcohol use).

Learning to accept SCD

Adolescents discussed aspects of their HRQL, from engaging in sport and exercise and socialising, to their emotions and future aspirations, including the extent to which SCD affects these in their daily lives (subtheme 1). They also discussed normalising SCD (subtheme 2).

Daily life with SCD

Adolescents with SCD and healthy siblings reported similar concerns unrelated to SCD that might impact their participation in recreational activities. For example, insufficient finances, a lack of facilities in their local area, their school or college work, or other time constraints. However, some adolescents with SCD explained that they had a lack of energy or were not able to participate in exercise or sports to the same level as peers, although this does not deter them from participating.

> Yeah, I get more tired than my friends after doing sports, it's annoying but it's the situation. You just deal with it. (Akono, SCD, male, aged 15, group 5)

> Yeah it's not the worst thing in the world – getting a bit tired - my friends still get tired ((Agreement)). (Amber, SCD, female, aged 13, group 6)

Similarly, to the previous quote, they also made other social comparisons to their peers, suggesting little evidence of discrepancy between current and ideal selves.

> Actually, I can do things my friends can't do … I'd say pretty even. They might be able to do things you can't do, but you might be able to do things they can't do. (Ayo, SCD, male, aged 14, group 1)

Healthy siblings discussed experiencing some disruptions to life, but only when their sibling with SCD is unwell.

> Every day it's not that bad, just sometimes staying with them, keeping them occupied, I can deal with that, mostly it's when they're sick it goes crazy! (Bisola, sibling, female, aged 13, group 4)

Overall adolescents seemed to be emotionally well adjusted and satisfied with their lives, with similar reports from adolescents with SCD and healthy siblings. When discussing if there were things they would like to do but cannot and things they would like to change, there was little evidence of adolescents being dissatisfied with their current lives and wanting a different ideal. Within GT the lack of discrepancy supports good HRQL. For example, adolescents with SCD and healthy siblings suggested that they were happy (including with their social lives) and that there is nothing that they would want to change.

> Yeah, I feel up for it, alright, no problems I take life as it is. I have no day to day worries. Pretty relaxed, happy. (Bello, sibling, male, aged 14, group 7)

> I'm generally just a happy person and wouldn't want to change or anything more ((agreement)). (Adewale, SCD, male, aged 14, group 5)

Finally, adolescents with SCD and healthy siblings discussed similar ambitions, in terms of careers or future lifestyles, so those with SCD did not appear to feel that their futures were restricted.

> I desperately want to have a proper career not just a job, like I said, a theoretical physicist or a games developer…. That will lead onto a good life, smokin house, holidays, watches. I'm determined to get there. I have as much chance as anyone else – there's no colour, no sickle cell. (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 and live in a nice house, have a nice car, stability…. We're all capable of success in our future ((general agreement)). (Basma, sibling, female, aged 16, group 8)

Normalising SCD

Adolescents accept that SCD is part of their or their sibling's identity. In a GT approach to HRQL, normalising plays a role in reducing discrepancy, thus supporting good HRQL.

> 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)

> My siblings 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)

They even suggested some protective elements of SCD, indicating they try 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)

Both adolescents with SCD and healthy siblings made social comparisons to peers with other conditions such as diabetes, dyslexia and allergies to highlight that others also have problems. They also made comparisons to people with more serious or life-threatening diseases to provide some perspective.

> Sickle's just a small part of me and my life. It 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, 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. (Bobo, sibling, male, aged 17, group 7)

Coping with SCD

Adolescents discussed strategies that may help them cope with the condition, including emotional support, which was quite limited (subtheme 1), and moderating health behaviours (subtheme 2) as part of adapting their lifestyle.

Emotional support

While adolescents reported seeking some emotional support, this was limited for a number of reasons. Due to absent fathers and family members living abroad, they often sought emotional support from only one or two family members (often their mother), but this also 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, they're in Africa. My mum brought me up by herself and sorted sickle stuff, that's why I don't like putting anything else on her. (Adegoke, SCD, male, aged 13, group 1)

Adolescents reported feeling more comfortable discussing their problems with people who have SCD, because they are more likely to relate to their experiences. For example, Babukar (sibling, male, aged 16, group 7) said 'there's always my cousins to go to, they get sickle … so it's easier to tell them'. However, they did not know many young people with SCD.

> 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'm not close to anyone with sickle cell, let alone anyone my age, so I don't have that many options for conversations. (Awa, SCD, female, aged 17, group 2)

Opportunities for support were also limited by some adolescents not wanting to disclose their/their sibling's condition. Healthy siblings felt it was a private matter and worried about gossip. Adolescents with SCD felt it was not necessary, that it would not change anything, and they did not want to feel different or be treated differently.

> I hate special treatment … 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 … it looked like something I wouldn't want to miss. (Amber, SCD, female, aged 13, group 6)

Linking to the previous subtheme on normalising SCD, many adolescents felt that it was not necessary to seek emotional support because SCD is simply part of their life.

> Sickle has always been part of our family. There's no need for big conversations about it. (Amara, SCD, female, aged 17, group 6)

Religious beliefs, however, were reported to provide strength, comfort and hope in coping with SCD, to healthy siblings and adolescents with SCD, especially during a vaso-occlusive crisis or hospital admission.

> Praying gives strength with it gives you the energy to get on with life and it helps overcome the negatives like when she has a crisis, tiredness, goes hospital, all of it. Religion is the source of my and my families hope but, it's this is important, I pray every day! Easy, good and tough times. Rely on Him and give praise to Him every day! (Biko, sibling, male, aged 13, group 7)

Moderating health behaviours

The current research focused on health behaviours that all adolescents may engage in, and thus did not cover medication adherence. Nevertheless, discussions about the use of technology to manage medication adherence arose. Adolescents reported that they were attempting to exert some control and had learnt strategies to help cope with SCD, such as using their smart devices to improve medication adherence. Healthy siblings also helped.

> 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. (Anna, SCD, female, aged 13, group 2)

> 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)

Many adolescents made small changes to their routine or modified the type of sport or exercise they participated in to avoid more strenuous, contact sports that

were played outdoors, especially in cold weather. This helped their symptoms and they discussed these modifications as not being a big deal, suggesting little evidence of a gap between their current and ideal selves.

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

> 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).

Adolescents with SCD also felt they had learnt how to cope with pain more effectively during a vaso-occlusive crisis, for example, by not engaging in physical activities and instead engaging in relaxing activities.

> 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. (Abena, SCD, female, aged 16, group 6)

Adolescents understood the importance of eating a balanced and healthy diet for their general health, although they did not link it to SCD. They understood that drinking water can help prevent a vaso-occlusive crisis, but most adolescents with SCD and healthy siblings reported drinking water on a daily basis anyway.

> To stay hydrated for Sickle Cell. To help the blood flow smoothly. To slow down, to stop a crisis. It's just like our energy tablet. Water. It's basically like energy to keep your body working like a car needs oil. (Azi, SCD, male, aged 17, group 1)

Influences on health behaviours

Under the previous subtheme some adolescents with SCD reported moderating their participation in exercise, but under this theme both they, and healthy siblings discussed influences on their exercise (subtheme 1), as well as influences on their alcohol use (subtheme 2). There was limited discussion of influences on diet or drinking water, other than that discussed in the previous subtheme. Additionally, adolescents had limited experience with risky behaviours, for example, only one healthy sibling had tried smoking tobacco cigarettes. Therefore, this theme focuses on exercise and alcohol.

Exercise

Adolescents explained that participation in exercise was influenced by their parents. Those with SCD were not encouraged to exercise, as parents were concerned about the negative consequences for SCD, but healthy siblings were encouraged.

> 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)

Nevertheless, adolescents with SCD did not appear troubled by this lack of encouragement and while, some moderated health behaviours (as discussed under the

previous subtheme), others felt SCD had little impact or appeared content with their participation in exercise, showing little evidence of discrepancy between their current and ideal selves.

> I do and try whatever sport I want. It's not like when you're a kid and you need permission. (Asir, SCD, male, aged 15, group 5)

> I don't experience problems, doing sport and I'm in the swimming team and cross-country team…. it's all about belief and getting on with life, not being held back by some label, some 'Sickle Cell' label given at birth and living your life. (Azi, SCD, male, aged 17, group 1)

Adolescents with SCD and healthy siblings reported similar influences on exercise. For example, female adolescents reported participating in physical activity for enjoyment and to socialise with friends, while the media had some positive influence on male adolescents, as they described watching sports on television and trying to achieve the physiques or sporting abilities of role models.

> I watch every Arsenal match to reproduce the tricks, movement and skill of Alexis Sánchez. I do specific exercises that I read he does to get his physique (Akono, SCD, male, aged 15, group 5)

> It's probably more athletics that I watch. And yeah, just athletics to be honest. I'd like to be like them, I try but obviously I can't get to their level because they train every day for 8 hours or something! (Babatu, sibling, male, aged 14, group 3)

Male adolescents with SCD, in particular, were motivated to improve their physiques to show strength and maintain some control over their physical appearance and health when they feel it is harder to control SCD. The discipline and maturity developed through having SCD were reported to facilitate this. Once again, they made social comparisons to peers.

> I can control what my body looks like, my basic health, not crisis, jaundice, ulcers, and still be on par with my friends. (Abdel, SCD, male, aged 17, group 5)

Alcohol use

More adolescents with SCD reported drinking alcohol compared to healthy siblings, but they did not report drinking excessively. Alcohol use appeared to be considered acceptable to both adolescents with SCD and healthy siblings because some parents drink and allowed them to have a drink on a special occasion.

> Drugs and probably smoking, nah, my parents wouldn't allow but with alcohol, yeah, 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)

However, peers mostly influenced adolescents to drink alcohol when they were socialising at parties or in the park.

> I've always had that temptation to try drinking, 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. (Ada, SCD, female, aged 15, group 6)

> I think it was at a party, it was just there, and people there were drinking it and like saying, 'come on, try', so I thought ok. Like the first time it was just I'll try it, and found out it was alright. (Bina, sibling, female, aged 17, group 4)

Thus, overall, both adolescents with SCD and healthy siblings reported similar influences on their alcohol use. Having SCD appeared to have little influence, with the exception that some female adolescents with SCD suggested that having SCD may actually provide strength and maturity to resist peer pressure to partake in risky behaviours.

> I think most people with sickle cell tend to have a lot of self-conf… 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)

Healthy siblings spoke of less serious health consequences related to alcohol use compared to smoking and drug use which they view as risky behaviours.

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

Similarly, adolescents with SCD generally believe smoking and drug use are more likely to trigger a vaso-occlusive crisis. These opinions tended to be based on observations of other adolescents with SCD drinking alcohol.

> 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 but taking alcohol wouldn't. Around me people with sickle cell take alcohol regularly and medication. (Afram, SCD, male, aged 16, group 5)

Discussion

The findings of this study highlight few discrepancies between the current and ideal selves of adolescents with SCD or healthy siblings and thus provide little evidence of poor HRQL or differences between the two groups in HRQL. Both groups (as evidenced by the mirroring of many quotes in the findings) reported similar feelings, experiences and influences on their lives. For example, they reported general issues such as finances that got in the way of recreational activities, being influenced to engage in exercise by role models or wanting to socialise, and being influenced to drink alcohol by peers. They appeared well adjusted and reported being happy and had similar aspirations for the future. Both groups reported some disruptions to life caused by SCD, for example, those with SCD get tired and lack energy, are not encouraged to exercise by parents, and have moderated some aspects of their lifestyle. However, they are not deterred from engaging in activities, do not perceive tiredness or adjustments as a big deal, and feel that there is nothing they cannot do that they would like to do or that they would like to change. Thus, there is little evidence of discrepancy between their current and ideal selves. Within GT a lack of discrepancy means good HRQL, and this supports previous research that has taken a GT approach to HRQL in children (Constantinou et al., 2015).

A key reason for this apparent lack of discrepancy appears to be that adolescents with SCD and healthy siblings have normalised and adapted to SCD. In a GT approach to HRQL, normalising plays a role in reducing discrepancy, and according to Calman (1984), reducing discrepancy does not mean lowering expectations. The present study showed that both groups of adolescents are ambitious to achieve future success. In contrast, research on younger children with SCD found they rarely spoke of their future ambitions in comparison to healthy siblings (Constantinou et al., 2021). The authors found evidence of normalising SCD in younger children, but this seems more pronounced among adolescents in the present study, who spoke of SCD being normal and just a small part of who they are/their sibling is. Thus, it may be that any discrepancy between current and ideal selves reduces with age. Adolescents report that they do not allow SCD to define them or dominate their lives and have learnt strategies to cope. Albrecht and Devlieger (1999) postulate that people with chronic illness are able to adapt socially and emotionally.

To aid normalisation, both groups of adolescents focused on positives. They made social comparisons to peers (e.g. 'on par with my friends', 'my friends still get tired', 'I can do things my friends can't do') and downward social comparisons to people with more life-threatening conditions or other problems. Making downward social comparisons is a defensive tendency that helps people to feel better about themselves or their situation (Wills, 1991) and may be used especially by people with chronic conditions in response to the threat of illness (Tennen et al., 2000). In adults with SCD, fewer depressive symptoms were related to downward social comparisons (Wilson et al., 1997).

In this study adolescents report sometimes seeking emotional support to help them cope but this is limited. The main person in their lives who provides support is their mother, which was also reported by younger children with SCD (Constantinou et al., 2021). However, similarly to Atoui et al. (2015), the present study found that adolescents with SCD do not want to burden their mothers. Adolescents also described feeling that their friends cannot relate to SCD and they would prefer to speak to someone with similar experiences, also supporting previous research (Atoui et al., 2015; Valenzuela et al., 2013). However, similarly to Constantinou et al. (2021), reluctance to disclose was an issue in the current study that undermined seeking emotional support. A lack of emotional support could be considered a concern, but due to their normalisation of SCD, adolescents in the present study also spoke of not needing to seek support. Nevertheless, adolescents did use their religious beliefs to help them cope which is in line with previous research (Clayton-Jones & Haglund, 2016) but in a study on younger children, this coping strategy was only reported by healthy siblings (Constantinou et al., 2021).

Findings of this study suggest that adolescents are trying to take greater responsibility for managing their SCD and that health behaviours play a key role. They appear to have adapted to SCD and learnt to cope effectively with it, a finding supported by Hijmans et al. (2010) and Panepinto et al. (2012). A sense of learning to cope and having some control supports the HRQL of adolescents with SCD (Goldstein-Leever et al., 2020). While the focus of the current research was not medication adherence, some use their smartphones to remind them or their sibling to

take medication. Past evidence has shown that daily use of a smartphone improved medication adherence in adolescents with SCD (Creary et al., 2014). An important health behaviour in preventing a vaso-occlusive crisis is regular hydration (Brown, 2012). Adolescents in the present study understood this, supporting Poku et al. (2018). Some adolescents spoke of moderating aspects of their daily routine that involve activities that could trigger a vaso-occlusive crisis, especially participation in strenuous exercise or exercise in cold weather (Brown, 2012; Poku et al., 2018). This explains previous evidence suggesting that adolescents with SCD participate in less exercise than their healthy peers (Melo et al., 2018), but this is also a possible concern as exercise is important for general health.

Unlike healthy siblings, adolescents with SCD were not encouraged to exercise by their parents. In previous research younger children with SCD were bothered by this and experienced discrepancy between their current and ideal selves because they wished to participate in more exercise (Constantinou et al., 2021). However, while adolescents with SCD in this study talked of acceptance of sometimes needing to be careful, they discussed not needing parental permission and making their own choices, which included participating in exercise, thus reducing discrepancy and suggesting this does not impact their HRQL. Overall, similar influences on exercise participation were reported by adolescents with SCD and healthy siblings, for example the social aspect was a motivator for female adolescents, and the media had a positive impact on male adolescents who were motivated to exercise by role models in sport. Male adolescents with SCD also felt exercise could help them exert some control over their general health and improve their physiques. Erskine (2011) established that male adolescents with SCD were self-conscious about their physiques and appearance in comparison to their peers. This may be because delayed growth in children with SCD is more pronounced in males (Bennett, 2011).

Adolescents with SCD also suggested that being mature for their age, and having to be strong and disciplined as a result of SCD helped them make healthier behavioural choices, including resisting peer pressure to drink alcohol. Nevertheless, overall, similar influences on alcohol use were reported by adolescents with SCD and healthy siblings, with peers and social situations being the main drivers, supporting a systematic review in healthy adolescents (Leung et al., 2014). Alcohol consumption appeared to be more prevalent among adolescents with SCD compared to healthy siblings. While this was only a small sample, this substantiated Asnani et al.'s (2014) findings. Poku et al., (2018) concluded that adolescents with SCD strive for a 'normal' adolescent life and assert their independency which may lead to more risk-taking behaviours. Alternatively, perhaps having a chronic condition is no longer a deterrent against engaging in risky behaviours (Sawyer et al., 2007). In this study, adolescents with SCD and healthy siblings believed that alcohol was less of a risky behaviour than smoking or drug use. Adolescents with SCD also erroneously believed alcohol was less likely to trigger a vaso-occlusive crisis, although in past research, older adolescents with SCD understood it was important to avoid alcohol (John-Olabode et al., 2015).

In summary, if health behaviours help manage symptoms or avoid vaso-occlusive crises (Brown, 2012), this is likely to positively influence HRQL, especially as quantitative research has found that disease severity/symptoms predict a lower (worse)

HRQL in children and adolescents with SCD (Stokoe et al., 2022). Although, in the present study, adolescents lacked an understanding of the potential impact of alcohol, they understood the importance of regular hydration and moderated their participation in strenuous exercise or sports. Of course, leading a normal adolescent life is important, and too much moderation may create a discrepancy between current and ideal selves, so it is important to strike a balance to support HRQL. This study has shown many similarities between adolescents with SCD and healthy siblings in terms of aspects of their lives, such as their social lives, emotions, future ambitions and influences on their recreational activities and health behaviours, and little evidence of discrepancy between current and ideal selves in these areas as a result of SCD.

Implications for practice

The findings of this study suggest there is a need for adolescents with SCD to have open and confidential discussions about risky behaviours with healthcare professionals (Surís et al., 2008). This is important because adolescents with chronic conditions who engage in risky behaviours are more likely to have poor disease management and treatment adherence (Scaramuzza et al., 2010). These discussions should aim to increase knowledge about alcohol use, and also healthy behaviours. This study found that adolescents endorsed healthy behaviours. They were aware of the benefits of engaging in behaviours, such as regular hydration, and of appropriate moderation, particularly of exercise. Therefore, there needs to be continued education surrounding healthy behaviours. This should include clear guidance about the importance of daily suitable, moderate exercise (e.g. walking, cycling), while also ensuring concerns about triggering a vaso-occlusive crisis do not undermine exercise participation. Thus, physical activity pacing should be promoted. Peer-led support groups or forums may be helpful in providing information about health behaviours and SCD. This would also provide a forum for sharing experiences and emotional support, as adolescents in the present study reported feeling more comfortable discussing their problems with people who have SCD. Healthy siblings could be included in such groups, and in supporting the management of their sibling's SCD to increase their sense of control. For example, in the present study, both adolescents with SCD and healthy siblings used smartphone apps to aid their/their sibling's adherence to medicine regimens. Future studies could explore the effectiveness of apps in supporting adolescents to moderate health behaviours to help cope with SCD, such as their use in supporting physical activity pacing.

Limitations

This study had some limitations which should be considered in interpreting the findings. First, a convenience sample was recruited during outpatient appointments at only two hospitals, so participants may be regularly monitored by hospital staff and consequently have a better HRQL and engage in healthier behaviours. Second, parents were asked to confirm the absence of any health conditions in healthy siblings. It is possible their reports may not have been reliable, but screening measures, such as psychological tests were deemed too demanding. Third, in-depth exploration of all aspects of HRQL and healthy/risky behaviours was not achievable in a single focus

group of an acceptable length, so further research is needed, for example, on alcohol use, where there is a dearth of research in this population. Fourth, each focus group included adolescents aged 13 to 17 years old which may mean they did not share similar experiences to facilitate discussion. Fifth, group discussions may have led some adolescents to be guarded about voicing insecurities, overemphasise alcohol use, or overstate how well they coped with SCD. However, group member influence is an important aspect of focus groups as this would happen in everyday conversations in natural settings (Peterson-Sweeney, 2005).

Conclusion

Despite these limitations, the findings provide optimistic results about the HRQL of adolescents with SCD and healthy siblings who appeared to differ little from each other and did not appear to have discrepancy between their current and ideal selves as suggested by GT. This suggests a reduced gap compared to that reported in younger children (Constantinou et al., 2021), where there was some evidence of discrepancies. With maturity, they have normalised and adapted further to SCD, and developed more effective coping strategies. Healthy behaviours may be part of this, but it is important to strike a balance so that modifications to lifestyle, such as moderating exercise, do not impair HRQL. Additionally, alcohol use amongst adolescents with SCD may be of concern as this can exacerbate their condition.

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Data availability statement

Data are not available due to ethical restrictions. Due to the nature of this research, participants of this study did not agree for their data to be shared publicly, so supporting data are not available.

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