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‘A qualitative literature review on the experiences of young people living with sickle cell anaemia’

Abstract

This qualitative literature review examines the experiences of young people with Sickle Cell Anaemia (SCA). Sickle cell anaemia (SCA) is a condition acknowledged for its unpredictability, painful episodes and life threatening nature. The aim was to explore the impact SCA has on young people and the experiences they face. After reviewing potential articles found from the EBSCOhost platform, an inclusion and exclusion criteria was used and six appropriate studies were found with the majority of the participants in the 10-25 age range. However, one study had some participants in the early childhood and another had some of the sample in the young adult age range. These articles concentrated on the UK and North America instead of a broader international experience. The critical appraisal skills programme (CASP) was then used to evaluate the articles found. Thematic analysis identified three themes: acceptance, support and unpredictability, with subthemes of spirituality and discrimination. It was clear that SCA affected multiple areas of young peoples' lives, and health professionals need to consider young people's views.

The terms Sickle Cell Anaemia and Sickle Cell Disease are used interchangeably throughout this review.

Key words:

Sickle cell anaemia, experiences, views, adolescents, young people and qualitative.

The World Health Organisation (2011) describes SCA as a haemoglobin disorder that affects how oxygen is delivered around the body due to haemolysis (accelerated destruction of abnormal red blood cells) and increase in red cell production (Revell, 2005; Kato et al, 2017) which leads to types of crisis (vaso-occlusive, spleen sequestration, chest crisis and aplastic crisis). It is an inherited condition from both parents, and leads to the modification in the shape of red blood cells from biconcave discs to sickle shapes (WHO, 2011). The distorted cells lack manipulability and can block small blood vessels, reducing the amount of blood flow and oxygen being transported around the body (Anglin, 2015). The life span of the red blood cells are shortened to between 10 and 20 days leaving the blood chronically short of these cells, a state referred to as anaemia. Development of the condition can lead to chronic acute pain, severe bacterial infections and necrosis (WHO, 2016). Young people and their families have to live with the chronic condition and have acute exacerbations or crisis leading to admission to hospital.

The terms Sickle cell disease and sickle cell anaemia are often used interchangeably in the literature. However sickle cell disease is an umbrella term which encompasses three main genotypes, HbSS, HbSC and HbS/thal. (Ware et al, 2017). Sickle cell anaemia (HbSS) is the most common genotype and the most severe form the genotype as people with HbSS are generally very anaemic, which is not necessarily the same for the other two. If someone has HbSC they are not referred to as having sickle cell anaemia. They also have a more moderate course of the disease. Although used interchangeably depending on the context could sometimes mean different prognosis for individuals.

Today millions of people throughout the world are known to be affected by this disease (Global Burden of Disease Collaborators Study, 2013; Collaborators, 2015). The WHO (2016) however notes that SCA is most common among people with particular ancestry, with the highest prevalence occurring in people of African or African–Caribbean origin (Tewari & Rees, 2013). In these populations, 1 in every 200 people have sickle cell anaemia (SCA) (NICE, 2010). However ethnicity is becoming increasingly unreliable as a predictor of sickle cell disorders, as evidence from the UK national screening programme demonstrates. India is also known to have a high prevalence which can vary across different tribal groups as outlined by Colah et al (2015) and Serjeant et al (2016).

It is now the most common genetic condition at birth, with 1 in every 2500 live births in England being screened with sickle cell anaemia and thalassemia (Public Health England, 2016). Furthermore, it is estimated that there are more than 12,500 people with SCA in England (NICE, 2010). Such high figures supports the need for better understanding of this chronic condition and how it affects patients. For this reason, analysing research on young people's experiences of SCA will be useful to current and future nursing approaches giving insight into how young people cope with long-term conditions within a hospital setting and wider community.

Aim

The aim of this qualitative narrative literature review focusses on the experiences SCA has on young people and recognising the impact. The review addresses the following research question “What are the experiences of young people living with SCA”? The research objective was to critically review selected primary research on this topic area and make recommendations for practice, education and research.

The terms “Adolescent” and “Young People” were used based on the World Health Organisation (2011) definition. Children are defined as under 19, Adolescent, age 10-19 and young people/ adult- over 19 years.

Methods

The systematic research selection process initially involved the use of ‘the university library’ and ‘Google Scholar’ to establish whether there was an adequate amount of studies focused on young people. Thereafter, EBSCOhost was used to conduct the search. The search was carried out by using combined databases including ‘Academic search complete’, ‘CINAHL’, ‘Medline’, ‘Psycarticles’ and ‘PsycINFO’. Boolean operators were used to merge key words ensuring that articles with the same key words were being found. Initially the search used the terms sickle cell AND adolescen*OR Child* AND experience OR views but numerous irrelevant articles were being retrieved. The concluding terms used were:

1. "Sickle cell" AND adolescen* AND Experience.
2. "Sickle Cell” AND experience.

A hand search was also conducted from the references lists within the chosen articles to find additional articles. A total of 153 articles were generated from the first search, and 235 from the second. The inclusion and exclusion criteria below were introduced to retrieve all eligible papers (see Appendix A)

Inclusion criteria	Rationale
Scholarly (peer reviewed) articles	This ensured that articles are written and reviewed by experts and professionals.
Academic journals	To ensure the research being retrieved was credible.
Date 2000-2016	The search was initially restricted to the last decade, but this gave few results. The search bracket was then broadened to the past 16 years. It is likely that older studies are still relevant as the experiences that young people face will be very similar to those noted in older studies.
English Language	In order to understand the research.
Title and Abstract	To prevent inappropriate studies being retrieved.

Exclusion criteria	Rationale
Research not conducted in the UK or USA	Research not conducted in the UK or USA. The search was originally confined to UK studies; however, it was widened after few hits. The US studies were included due to its similar economic and healthcare provisions.
No relevance towards adolescent's viewpoint	The focus of the question was adolescents and young peoples' experiences, therefore studies centred on parent's views; siblings and adults were excluded.
Research done in less economically developed countries.	Although experiences may be similar to the UK elsewhere, living in a developing country may impact the experiences adolescents and young people have.
Studies that focused on a specific treatment	The question seeks to explore experiences of SCA as a whole.
Studies on transitioning	Many of the studies retrieved were based on experiences of transition to adult services, as opposed to experiences as an adolescent and young people.
Systematic Reviews	The aim was to look at primary research in order to develop a review.
Wrong age group	The focus group were either adults or young children instead of adolescents and young people.

The number of articles were reduced to 14 after applying the above criteria. Once their suitability was assessed a further eight were excluded after reviewing the full texts as they did not meet the inclusion criteria and therefore 6 articles met the inclusion criteria. (see Appendix A).

The Critical appraisal skills programme (CASP, 2014) was used within this review as a quality assessment tool to identify the strengths and limitations of each study. The structured approach outlined by Clarke and Braun (2013) was used through becoming familiar with the data; reviewing, defining and synthesising the themes to create a wider understanding. In order to interpret the findings of each article, a process of thematic analysis was used based on (Braun and Clarke, 2006; Clarke and Braun, 2013) recommendations. The process included reading each article several times to identify core findings, which were later developed into key themes and subthemes. Themes were then compared between the studies to identify common topic areas discussed by the participants. Further refinement and review resulted in 3 core themes and 2 sub-themes.

Results

The findings of the articles were compared and contrasted (See Appendix B for article summaries). Investigating how trustworthy an article is crucial to appraisal. Lincoln and Guba (1985) developed and recommended a criterion for analysing transferability, dependability, confirmability and credibility (Morse, 2015) which were used. Each of the final 6 articles were analysed in relation to the following:

- Aim
- Methods
- Date
- Country
- Sample size and gender
- Ethics and Consent
- Key Findings
- How does the study answer the research question?

The six studies came from the UK and USA and were published between 2001 and 2013. All used qualitative methods to capture the views and experiences of the participants. Erskine (2011), Valenzuela et al (2013), Dyson et al (2012) and Stegenga & Burks (2013) all took place in the last five years. Conversely Atkin & Ahmad (2001) and Thomas & Taylor (2002) studies occurred in the last 15 years. However, these studies all had similar results, thus could be considered confirmable as well as having greater dependability. Dyson et al (2012), Erskine (2011), Atkin & Ahmad (2001), Thomas & Taylor (2002) were all UK studies which enhances the dependability of these results, but indicates a research gap as the other two studies were from the USA.

Sample size and gender:

All the studies, bar Erskine (2011), gave perspectives from both genders increasing the credibility. Erskine (2011) recruited male adolescents only. Polit & Beck (2013) found that gender bias was generally observed when females alone were sampled and in the Erskine (2011) study it was good to have the male perspective as it can be challenging to recruit males to such studies. The sample size of a study dictates the quantity of information the study will generate (Cleary et al, 2014), particularly in relation to qualitative research which aims not to generalise but to encourage the production of rich data (WHO, 2004). The majority of the studies involved 25 or less participants (See Appendix B) which is common in qualitative research.

Method:

All of the studies examined used a qualitative approach, although differed in how they collected the data. Stegenga & Burks (2013) & Valenzuela et al (2013) used photovoice which "is often valued for its ability to uncover rich descriptive information" (Catalani & Minkler, 2010; pp.441). Erskine (2011) used Interpretative phenomenological analysis (IPA) which uses a psychological focus to help explore participant's experiences, understandings, perceptions and views (Brocki & Wearden, 2006). Atkin & Ahmad (2001) and Thomas & Taylor (2002) were the only studies conducted over a longer period of time. Atkin & Ahmad (2001) directed two interviews within a six-month period and Thomas & Taylor (2002) were over eight weeks. Thomas & Taylor (2002) was also the only study that used focus groups as a means of collecting data, which increases the depth, stimulates data from a range of perspectives and

in turn trustworthiness and reliability (Rauf et al, 2014). Dyson (2012) was the only study that used mixed methods of: questionnaires, recorded interviews and then case studies conducted with ten of the young people which enabled triangulation (Wilson, 2014). Atkin & Ahmad (2001) was the only study which clearly mentioned about participants having the choice of an interviewer. The researchers took into consideration the ethnicity and culture of partakers as they were likely to touch sensitive issues. Stegenga & Burks (2013), Atkin & Ahmad (2001), Dyson et al (2012) all stated that all interviews were fully transcribed.

Ethics:

Erskine (2011), Dyson (2012), Valenzuela et al (2013), Stegenga & Burks (2013) & Thomas &Taylor (2002) received either approval from an institutional review board or hospital research ethics committee. Although Atkin & Ahmad (2001) do not explicitly state that they sought ethical approval, they chose individuals from the case loads of health professionals therefore it may be assumed that permission was sought beforehand. Atkin & Ahmad (2001) appeared to take greater care by sharing how pseudonyms were used to maintain confidentiality. The acknowledgement of rights and the appropriate use of consent and assent were found amongst the majority of the studies.

Acceptance

Acceptance was one of the common themes established, although two different sub-themes emerge. One sub-theme was how individuals understand and make sense of their condition in relation to who they are. The second is about how they makes sense and accept their condition in relation to other people. As SCA is a disease which can affect the whole body, it was noted that participants struggled to accept or avoided the reality of their condition. Atkin & Ahmad (2001) expressed that individuals would view themselves as normal to maintain a positive self-identity. This sense of normality and acceptance was also raised by Valenzuela et al (2013, page 102) who defines normal as “the importance of normal everyday activities and interests” and similarly Erskine (2011) discussed this as a comparison between the participants perception of themselves and their peers and their own past condition physically. This is reflective of the adolescent stage of development as engagement with peers is critical. In contrast, Dyson et al (2012) interviewed a 25-year-old who described SCA as a burden and showed resentment towards it. One male partaker from Thomas & Taylor’s (2002) study

expressed that people were preventing him from living normally, however this study was carried out 11 years ago so perhaps perceptions and normalisation of SCA were different. Erskine (2011: pp.24) focuses around comparisons to the past as one participant explains "Well I am normal. I'm getting more normal as I get older". This shows how the nature of sickle cell crises can change and in this study explored how the frequencies of crises and admissions to hospital improved as the participants got older.

Erskine's (2011) study also developed discussion around denial/non acceptance of the diagnosis as one 17-year-old participant challenged the validity of the diagnosis given by his doctor and parents. He expressed anger and resentment towards his parents for his inheritance of SCA, which was interesting as others in the same study did not understand the pattern of inheritance (Erskine, 2011, page 26). This was the only study which explored hatred towards SCA, particularly in relation to pain and inability to do things their peers could do. Equally this was the only study that used IPA method, which focuses on the psychological aspects and lived experiences of individuals. However, it was important to note the statements used by Erskine (2011) were all from male only perspectives which could have influenced their perceptions as stereotypes in society about men may influence their perception of being strong and able to cope with the pain.

Within acceptance, the sub theme of discrimination/stigma within school and the work place was apparent. Individuals believed racism formed more of an obstacle than their actual illness. It was noted by Atkin &Ahmad (2001) that the low expectations from teachers were not just down to their SCA but some felt it was also their African-Caribbean descent that contributed. Dyson et al (2012) whose participants were of an average age of 16 years; was the only study that focused specifically on school and ethos. They stated that individuals were being physically and verbally abused in relation to their condition. Their participants stated they had comments made about being jaundiced, their size, height and missing time from school. Furthermore, Erskine's (2011) study expressed they felt belittled and ridiculed due to their physical immaturity and obvious differences.

Supportive network

Support was another eminent factor discovered within the studies as support from family, friends and health professionals was mentioned by each of the articles. However, the level of

support needed and achieved does vary according to the age, stage of development and the wider support network. Valenzuela et al (2013) emphasised the importance of support and relationships, 18 of the 80 photographs taken were a representative of support of others. This study also identified pets as a supportive source. This possibly attributed to the method of photovoice where participants were able to express and capture their lives through pictures. Although this method was also used by Stegenga & Burks (2013) participants here were restricted within a campsite but the importance of friends was also mentioned. It seemed that participants felt a sense of security as this camp only consisted of individuals with SCA and smaller proportion with thalassaemia. Atkin & Ahmad (2001) found that the use of emotional/social support from family and friends was more popular amongst girls and younger children, while many older boys saw it as futile. The study revealed that older boys regarded emotional responses, like crying as a threat to masculinity, yet this was the only study that acknowledged family background. Out of the participants, 12 lived at home with only their mothers indicating that they may be imitating a strong father-like figure and displaying emotionally stability.

It was noted that family support varied amongst studies as Atkin & Ahmad (2001) mentions that solitude was valued as they did not want to burden others, however it was recorded that parents remained their best allies. Erskine (2011; pp.25) supports this statement as one participant in adolescence stated "Hospitals can't help. If I have a crisis we (my family) know how to deal with it". Yet there is still a reliance on professional care due the nature of the condition.

Thomas & Taylor's (2002; pp.350) study observed the contrasting issue of parents unable to come to terms with the child having SCA, making relationships difficult. This study stated one young person's perspectives that their parent would often cope through denial. One individual stated "Like when I'm in hospital, she never comes to see me". Conversely, this was the only study that had a criteria of recruiting participants who had at least three hospital admissions, which may indicate that participants had a greater health impact from their SCA compared to the other studies. Some studies noted that the support from health professionals impacted their experiences. One individual from Erskine's (2011) work expressed their appreciation towards their doctor. Appreciation was mentioned in Thomas & Taylor (2002) findings, participants found that staff made efforts to deal with their pain

promptly, but a lack of empathy amongst some hospital staff concerning their excruciating levels of pain. The lack of understanding, empathy and feelings of being ignored can lead to distress and anger. These pessimistic experiences were noted in Valenzuela et al (2013; pp.102) study where a 14-year-old boy stated "my school nurse is mean and I try not to go there". However, there is little elaboration on why this boy held this opinion. In contrast Atkin & Ahmad (2001) revealed young people valued positive contact with health professionals who helped them to accept their condition and maintain normalisation. Again, it is critical to acknowledge that development, the context either within the hospital setting, community or family can impact the perceptions or experiences of the individuals.

Spirituality was found to be a source of support for some participants particularly during painful crises which are evident with SCA. Perhaps this is due to the fact that the majority are from an African-Caribbean background where religion is commonly practiced. Atkin & Ahmad (2001) & Erskine (2011) recognised that individuals considered prayer as a coping strategy, but others believed their condition was either a test from God or punishment. The use of religion was often more evident in the younger participants ages 11-13 as identified by Atkins and Ahmad (2001).

Unpredictability

The unpredictability and controlling of symptoms is a primary element to living with SCA. Being watchful and managing symptoms to prevent decline of the condition especially with regard to pain was an apparent theme revealed in four of the articles reviewed. Stegenga & Burks (2013), Valenzuela et al (2013) mentioned that young people with SCA were sensitive to environmental influences, and the constant necessity to be alert to prevent any pain or crisis. Individuals were more inclined to photograph things that related directly to their disease and its management. Initially this theme was only apparent in these two studies as the use of photovoice enables children to express and think, however this topic was evident in Erskine's (2011) study that used IPA. This study acknowledged the unpredictability of SCA and the managing of pain. Atkin & Ahmad (2001) also explained that the respondents persistently juggled the uncertainties of their illness which created a tension in their lives. To which one respondent commented that he was overwhelmed by his illness and was concerned about the effects on his future. This study clearly outlines the unpredictability for

those with SCA due to reoccurring crises. Thomas & Taylor's (2002) study also showed evidence that the unremitting nature of SCA has an immense impact; the unpredictability can lead to depression, suicidal thoughts and hopelessness. This was the only study which involved focus groups, but it is interesting to note how one participant was open in this sense, although they may have found reassurance knowing that everyone within the group all had the condition. The severity of the condition can vary and therefore this also has an impact on the predictability of SCA.

Discussion

The three main themes identified: acceptance, support and unpredictability, as well as two subthemes; discrimination and spirituality will be explored further.

Acceptance

For most participants' acceptance was something either reached or worked towards. For many it was difficult to acknowledge they had SCA, however this may vary according to developmental stage and the wider supportive networks. The thought of being different at an age where comparisons and being accepted by peers is classed as significant, resulted in some not expressing their condition with peers or work. Cotton et al (2009) found that many individuals just accepted their SCA and projected a positive outlook. One participant stated "even though I have sickle cell I wake up and know I have my day on Earth... but I just thank God he woke me up this morning" Cotton et al (2009, pp.319).

Forrester et al (2015) found that respondents predominately felt good about themselves and accepted their condition, but negative feelings were apparent especially during periods of painful crises. In a Nigerian study, Adeyemo et al (2015) noticed the attachment of stigmatisation on individuals with SCA, and reported low self-esteem and feelings of hopelessness. Similarly, Musumadi et al (2012) researching in the UK stated that adolescents had challenges adjusting to SCA and developing an optimistic outlook. From these studies it is apparent that there can be similarities in individuals' experiences in different countries. Burnes et al (2008) study of how mothers of children with SCA accepted the condition supported the views of young people in this paper. They also found experiences of loneliness and separation anxiety. From this, it is clear that the acceptance of SCA was not just a difficult

task for young people but also mothers and siblings. This is an aspect, although beyond the remit of this paper, which could be further explored to triangulate with young people's views. It is also key to recognise that acceptance for young people can be influenced by parent perspectives and their acceptance also.

Support

Individuals noted that having a supportive network had a positive impact on their experiences. Yet there was a divide regarding the support individuals received from parents, schools and also health professionals. Some studies mentioned that parents were a source of strength, but others found that where parents coped through denial, it altered relationships and created difficulty. Mitchell et al (2007) expressed that parents relied on children to monitor their symptoms. This approach could be seen as unsupportive, however parents who listened to recommendations regarding treatment, promoted knowledge and independence within young people. Although appreciation was given towards health professionals, results also found a lack of empathy and understanding amongst them. A study which focused purely on priapism showed individuals avoided hospitals except as a last resort. Some participants also found health professionals to have a lack of knowledge (Addis et al, 2007). This essentially is a concern as patients need confidence in professionals. The role of the sickle cell anaemia clinical nurse specialist (Anionwu and Leary, 2012) is critical to enhancing the young people's views of support and enabling them along their personal journeys, advocating on their behalf with others and facilitating them to navigate through denial and crises. This vital role can help young people to build their trust in health care professionals.

Amongst the studies, schools were labelled as unsupportive, and staff were seen as lacking awareness about SCA and inconsiderate towards students (Dyson et al, 2012; Thomas and Taylor, 2002; Atkin &Ahmad, 2001). Knight-Madden et al (2011) emphasised the impact schools and staff have on young people with SCA by playing their part to prevent triggers of painful crises, and also being a source of encouragement. In the literature more broadly primary support also came from mothers, intimate partners, close friends, primary care nurses and counsellors (Derlega et al, 2014).

It was apparent in the Atkin & Ahmad (2001) & Erskine (2011) studies that religion and spirituality was found to be a source of support for some individuals. Prayer was seen as a

basis of strength for SCA for some. Cotton et al (2009) supported this view; by demonstrating adolescents saw God as a source of emotional support who aided them in times of need.

Unpredictability

The unpredictability of SCA made it difficult for individuals to cope and live their lives without the constant fear of a crisis. This persistent fear often created a boundary to living 'normally'. Individuals often knew their limits, however for some crises would occur with a trigger. Coleman et al (2016) also reinforced this theme in their study with adults' age 24-57 years. The irregularity and changeable nature of SCA was problematic for individuals, and for many this was the element that caused the most issues.

The variability and unpredictability of sickle cell pain highlights the importance of individualised pain management (Fosdal, 2015). Although the prevention of sickle cell related pain is a difficult factor to control, studies have observed ways in which this unpredictable pain can be minimised. Psychological and behavioural approaches have been found to be beneficial using methods such as distraction and entertainment. Furthermore, non-pharmacological approaches like massages and heat pads may provide relief (Rees et al, 2003) but the effectiveness of these approaches has to be person-centred for young people with SCA.

Implications for Future Practice

Themes from the review	Practice & Health Care Delivery	Education	Research
Acceptance of own diagnosis and condition, acceptance by peers, teachers and employers.	The opportunity for the young person to explore their emotions and acceptance of their condition through counselling and psychosocial interventions and discussions with the CNS.	SCA education and learning programs could be introduced, particularly to the school curriculum for peers and teachers.	Further research to explore peers, teachers, employers' perspectives of SCA through qualitative approaches.
Support needed from parents, peers, schools and health professionals	Peer mentors for young people with sickle cell. Create a forum for young people with SCA to meet with health professionals and peers and create opportunities to discuss positive and areas for development which impact on their health care. This forum could gain feedback from young people to health	A support service within schools could be created to help students with SCA which should involve teachers, parents, students and school nurses. Students would find a sense of security within this group, and could make any concerns	Explore supportive networks young people find useful in interviews and make recommendations.

	professionals, providing an opportunity for the young person to be heard.	known or academic difficulties.	
Unpredictability of the condition with crises	Encourage staff to be reflective practitioners. Johns (2010) expresses that reflection creates fresh insights and recognition for improvement. The CNS manage the young people's treatment during crises.	Educating student nurses throughout their degree about SCA would prepare them before becoming a registered nurse. This would allow a greater understanding and knowledge on how to deal with SCA patients but will minimise the stigma of health professionals having a lack of awareness and empathy as outlined in the review.	
	Develop guidelines with the assistance of young people with SCA and parents as part of a	Implement an educational programme for young people, peers, teachers and parents about the	

	health promotion strategy.	nature of sickle cell anaemia and how this may differ with individuals.	

Limitations

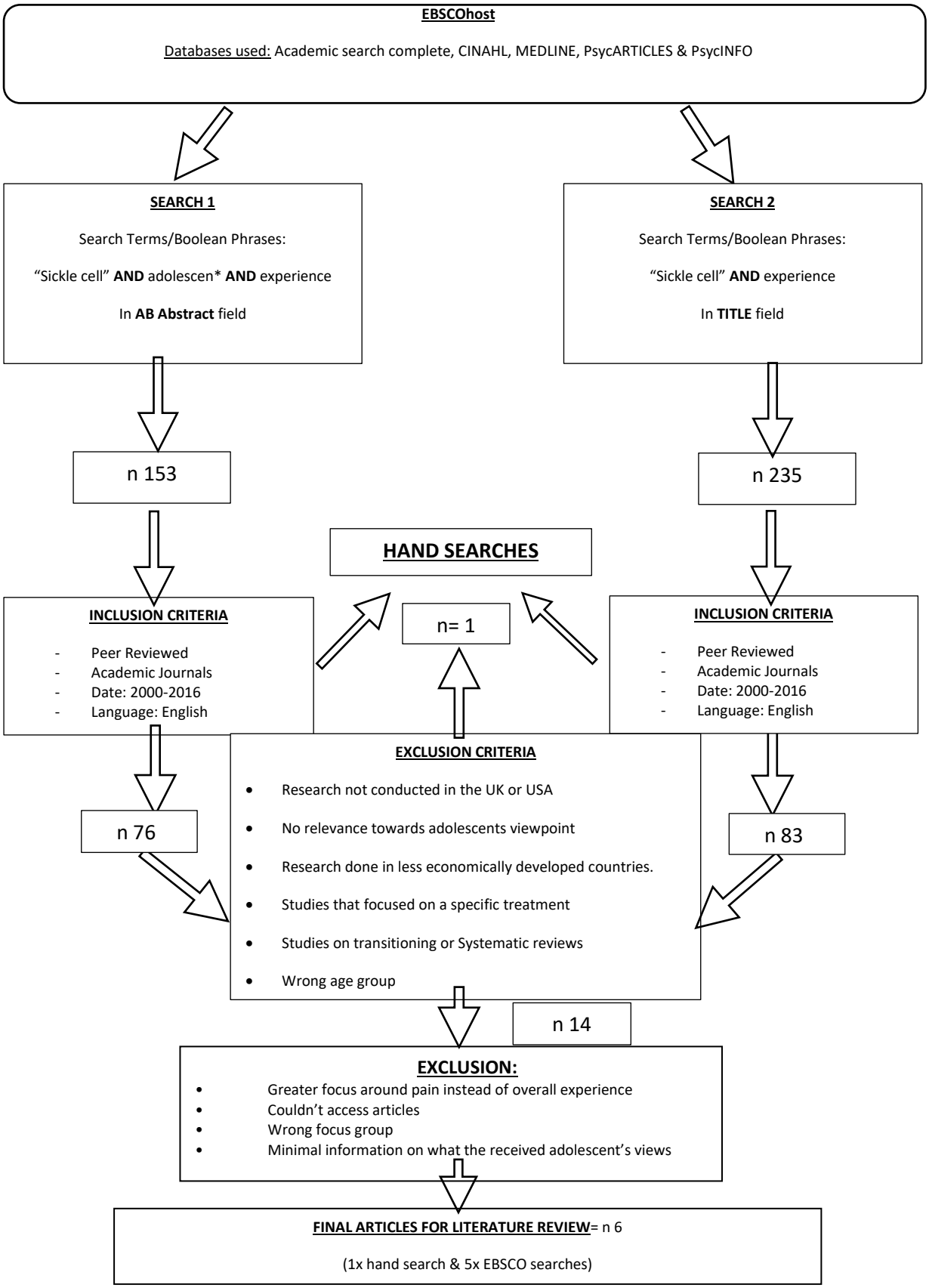
Possible limitations and strengths that could have altered results are as follows:

Limitations	**Strengths**
Articles from only two geographical areas	Articles from two geographical areas- Strengthens transferability
The research was conducted in a variety of different settings, including hospitals and a campsite	Articles used different types of methods to involve the young people
Age of articles- more contemporary research needed	Articles included different genders
Limited research specifically found in UK and USA	Similar themes found amongst studies
In ability to triangulate the young people's views with health professional and families views	Each of the articles were all from a small sample providing rich data
The research papers do not all differentiate the samples who have sickle cell anaemia rather than sickle cell disease and therefore the severity for the individual may vary.	This does emphasise the complexity of the condition and how the impact on individuals may vary.
This was an unfunded review and therefore the scope of the assessment of risk of bias at the level of the individual studies was limited due to time constraints	The small number of studies found led to a depth of analysis using the critical appraisal tool

Conclusion

In conclusion, it is evident that young people generally found it difficult to comprehend living with SCA. The impact it had on their daily lives was quite severe. This review has answered the research question as it has explored individuals' views through thematic analysis; revealing the challenges faced and attitudes toward their SCA. Nevertheless, it is clear that additional research within the UK for this age group is needed, so that potential strategies can be developed such as: 1) education within schools for students and teachers, 2) more qualitative research conducted within the UK, 3) development of mechanisms so healthcare professionals can hear the young people's voices influencing the care they provide, 4) formulate a health promotion strategy in the hope to improve the overall experiences for young people with SCA.

Appendix A flow chart



Appendix B– CASP

CASP/author	Atkin & Ahmad 2001	Erskine 2011	Valenzuela et al 2013	Dyson et al 2012	Stegenga & Burks 2013	Thomas et al 2002
1) Was there clear statement of the aims?	Yes- to provide a detailed understanding of young people's experiences of living with SCD and thalassemia	Not extremely clear but explains the study is to examine young men's experiences with this condition. Overall objective was to consider the health care needs of research participants. -evidence of poor engagement with health service in afro Caribbean yp with chronic illnesses	Yes-pilot study to understand how children and adolescents with SCD perceive their lives and disease using photo voice. The study is important as it's the first of its kind to engaging youth with SCD in a participatory method to better understand their disease within a community of their peers.	Yes - To attempt to make sense of variable experiences unrelated to disease severity or to teacher/peer awareness of sickle cell disorder To provide an understanding of the experiences of young people with sickle cell at school. To provide plausible explanation of variation in school experiences.	Yes-Examine the perspectives of children and adolescents with SCD using photovoice Limited research to date has used qualitative design to give voice to the perspectives of yp with SCD	No-minimal information regarding the set aim of study only brief stated in abstract
2) Is the qualitative method appropriate?	Yes-in depth interviews with young people (YP) aged (10-19) Interviewed x2 in 6months to reflect variability of condition and explore dynamic in greater detail.	Yes- qualitative method-suited to exploring personal accounts of illness. Interpretive phenomenological analysis. Eight boys aged between 13- 17 years with sickle cell anaemia	Yes- photo voice and reviews/group discussions were completed after each project	Yes- A mixed methods approach was selected First questionnaires to ascertain the experiences of young people Secondly tape- recorded 1 hour interviews in the form of a guided conversation Thirdly, case studies were conducted which encompassed: 1 hour taped interview with young person, main carer And a 2-week dairy of their school experiences	Yes- photovoice- and Completed qualitative interview afterwards	Yes- 32 hours of focus groups sessions to understand impact of sickle cell
3) Was the research design app to address the aim of research?	Yes-qualitative methods- (such methods allow an examination of complex and contingent situations, behaviours, beliefs and interactions).	Yes- IPA is a qualitative method that has been used extensively to investigate how people give meaning to bodily states and health care decisions	Yes- photovoice is a method whereby people are given cameras to document their lives. They share and engage a critical dialogue about the photos in order to capture info about lives/issues.	Yes- Mixed methods approach- facilitates collection of a range of data from a variety of sources.	Yes- studies have demonstrated photovoice to be a useful tool to examine psychosocial aspects of life, by allow participants to express their perspectives through a tool they can control,	Yes- increase the depth of an inquiry and stimulate data from a range of perspectives

					researcher gain more specific info tailored to the individual without limitations of structured measures. Previous success using photovoice-researchers felt it would provide a unique and beneficial approach to qualitatively explore important themes when this method is used.	
4) Was the recruitment strategy app to aims of research?	Yes- sample drawn from records of health professionals- sample of 26-12 males and 14 females- 18 had SCD aged between10-19.	Yes- medical records scrutinized to identify male participants aged between 13-17 yrs with a diagnosis of HbSS. 17 males who met criteria were sent info on study and parents of those under 16. 8 were recruited (13-17 yrs), participation was voluntary. No info on why some individuals did not participate	Yes- children recruited as part of an annual SC research and education day. Youth who met criteria were identified at event. Study was verbally explained and consent forms given that outlined the study and risks and benefits of participating-	Yes- participants recruited from three London outpatient clinics and primary care settings and 12 local sickle cell support groups Recruitment age 4-25 but majority were within compulsory schooling age.	YES- recruited at a sickle cell clinic in large Midwest hospital prior to attending a summer camp for children with SCD and their siblings Potential participants approached at camp drop off site no participants or parents who were approach declined study or withdrew after consenting screening done prior to camp start	Yes - twenty-five participants with sickle cell disease were recruited from four study centres Recruitment at the hospitals was conducted with the assistance of the consultant haematologists, sickle cell nurse specialists, and sickle cell and thalassaemia counsellors Participants needed to meet criteria
5) Was the data collected in a way that address the research issue?	Yes- All interviews conducted in English and were tape recorded. A topic guide identified a number of key themes they opted to explore during interviews-guide developed from review of relevant lit.	Yes-interviews schedule guide- included self-concept and identity, the experience of illness, family and peer relationships and sources of support. Open ended questions. Interviews tape recorded. At the end participants able to debrief and any outstanding issues were addressed.	Yes - all youth received digital cameras- they were given 3 assignments related and youth spent time photographing their lives according to the assignment- reviews/group discussions were completed after each project using the adapted version of SHOWeD questioning technique One additional session was conducted for both parents and youth to complete satisfaction surveys to measure acceptability of this method. Brief demographics on participants	Yes- mixed methods approach: Questionnaire, 1-hr tape recorded interview and case studies conducted.	Yes-Participants took photographs during camp and completed qualitative audio taped interview afterwards for participants to explain the message each photo conveyed about their life with SCD Interviews were subsequently content analysed and coded for salient themes related to the experiences At the end therapeutic benefit was examined through open ended	Yes- focus groups were tape-recorded and transcribed verbatim

					questions assessing positive and negative aspects of methodology	
6) Has the relationship between researcher and participant been adequately considered?	Partially- The researcher has not critically examined their own role but they have examined their participants- as they were able to choose the sex of interviewer and were also offered one of the same ethnic background (which was more important for parents for both language and cultural reasons)	Can't tell- Nil info on bias or influence A provisional interview schedule guided by sickle cell and health psychology literature Questions were developed in consultation with a young man with SCD and a paediatrician and clinical psychologist consultant Open ended questions guided by interview schedule. One participant requested further help-referral made to CAHMS	No- no info on relationship between participant and interviewer	No- no info on relationship between participant and interviewer	Partially - interviews by a study team member in a quiet private room in the clinic The study team member was a paediatric haematology/ oncology nurse with almost 20 yrs. experience working with children and adolescents and a researcher with 10 yrs. experience. further insight into challenges and opportunities related	No-
7) Have ethical issues been taken into consideration?	Partially- No mention about ethical approval Mention about confidentiality-use of pseudonyms to protect respondent's identities.	Yes- Ethical approval obtained from hospital research ethics committee. Medical records scrutinized. Info sheets sent either at outpatient's appointment or sent via post Parents for children under 16 given info sheet as well. Participants gave written consent. It was emphasized that participants could withdraw at any time.	Yes- Approval was received from the institutional review board before any study procedures were initiated. Considered consent and parents asked to sign a consent form giving permission and children given asset form	Yes- Approval from De Montfort university Human research ethic committee A multi-centre health services ethics committee	Yes - Institutional review board obtained before commencement of study parents' permission and child assent	Yes- Ethical approval was obtained from the local ethics committee of each of the hospitals and written consent was obtained from each participant.
8) Was the data analysis sufficiently rigorous?	Yes- topic guide used then interviews transcribed and organised according to analytical headings.	Yes- verbatim transcripts obtained and reviewed to identify recurrent themes. Themes elaborated and revised.	No	No	Partially- information regarding qualitative content analysis used	Partially – mention about thematic analysis used A phenomenological methodology was used to guide analyses

9) Is there a clear statement of findings?	Yes- And in conclusion No mention regarding credibility of their findings. Yes, they relate findings back to question. Consider both sides of themes.	Yes- table of 3 key themes then detailed sections of results- discussions from both pro and con	Yes- findings clear- sectioned into 4 themes No info regarding credibility	Yes- brief introduction to findings and use of subheadings	Yes- 3 themes emerged Very minimal detailed info around themes- but small sample so very limited results Looked at both sides etc.	Yes- findings clear and detailed. 6 themes revealed.
10) How valuable is the research?	Can't tell- No mentions on policies does relate back to previous research Nil info on identifying new research areas. Does relate info found to other research on chronic illnesses. Explain that yp struggles can only be understood in relation to wider context	Comparisons to what precious literature has mentioned. Young people drawn to health care initiatives conducted outside traditional settings Quality of communication is important- for preparing yp to manage own health and with health professionals Initiatives such as psychological educational groups Assessment and interventions for those who have suffered or who are at risk of stroke. Problems in failure to provide adequate pain relief -weaken trust in health professionals The study's validity is supported by the internal coherence of the themes and its coherence with the background literature.	Can't tell- nil mentions around current policies or practice Explain that photovoice is an effective way for yp to express feelings n SCD, share work and develop a sense of accomplishment to provide opportunity for youth to develop leadership and critical thinking skills.	Information expressed about school policies and further research should be conducted to assess transferability of ideas mentioned to other long term conditions.	A limitation was the small sample size but appropriate for a pilot study of this method It limits generalisability of results but yields key strategies for future research Future research conducted within home setting exploring SCD on daily lives would allow a larger picture on the impact of SCD on the individual and family. Photovoice and children with SCD can have a powerful impact on policy makers on the community, local and state levels Data collection during camp may not have allowed important themes to emerge Recommended future research on technology, flexibility and location.	Can't tell- nil information about implications to practice etc. but expressed that caution should be applied in generalising the present findings to all people with sickle cell disease

Appendix C- article table

Article	Date	Country	Purpose of study	Type of study	Method	Sample	Ethics	Key findings	Question answered
Living a normal life: young people coping with thalassaemia major or sickle cell disorder. **Atkin & Ahmad**	2001	UK	➢ Provide a detailed understanding of young people experiences of living with these conditions ➢ Explore how young people live and cope with conditions ➢ Address the worries and difficulties young people face ➢ The coping resources and strategies they employ to live a 'normal' life	Qualitative study	➢ In depth interviews x2 within a 6-month period	26 young people with SCD and 25 young people with thalassaemia major (aged 10-19) largely of south Asian and African Caribbean origin	➢ No mention regarding consent ➢ Consideration of confidentiality with use of pseudonyms	➢ Difficult to cope- compared themselves with other individuals who were worse- help develop a sense of normalisation ➢ Parental support was seen as protection ➢ Questions about why SCD re occurs during stressful events/changes in status life transitions	Yes – different areas of issues covered
The psychological experience of people with sickle disease and its impact on quality of life **Thomas & Taylor**	2002	UK	➢ To gain understanding of the psychological impact of sickle cell disease. ➢ To determine whether theses experience could be conceptualised in terms of quality of life defined by the	Qualitative study	➢ 32 hours of focus group transcripts. ➢ Each session was tape-recorded and transcribed verbatim ➢ Data were analysed using a	Twenty-five participants with sickle cell were recruited from four study centres. Participants need to have haemoglobin HbSS and be aged	➢ Ethical approval was obtained from the local ethics committee of each of the hospitals ➢ written consent was obtained	➢ Denial and lack of understanding from parents made it difficult to adjust - caused relationship issues with parents ➢ Education-absence from school- lack of support from teachers- strain on academic achievements	Yes- detailed responses given

			World Health Organisation with the intention of informing psychosocial research.		phenomenological approach	between 15 to 35 years with three or more hospital admission. Only 17 patients (7 males, 10 female) attended the sessions.	from each participant	➢ Impact of the unremitting nature- unpredictability of disease- hard to cope/manage ➢ Employment- disclosure of condition, jeopardise job opportunity ➢ Strain on relationships, - difficult to relate with others ➢ Hospitalisation-prevent going to hospital- adds more stress.	
Adolescent boys with sickle cell disease: A qualitative study **Erskine, R.**	2011	UK	➢ To investigate the experiences of adolescent boys with Sickle cell disease ➢ To consider the health care needs of research participants, taking account of social, cultural and personal beliefs about their illness and their attitudes towards medical intervention	Qualitative study	Qualitative methods to examine young men's experiences- By using Interpretive phenomenological analysis	Eight boys aged between 13- 17 years with sickle cell anaemia. All described themselves as either black African or black Afro- Caribbean All practicing Christians	➢ Ethical approval was obtained from the hospital Research Ethics Committee ➢ Participants met criteria provided with an information sheet describing the. ➢ Parents of those aged under 16 were given an information sheet prepared for parents and carers.	➢ Keeping watchful-being mindful to prevent any painful/crisis episodes ➢ Holding onto normality- bullied due to physical immaturity ➢ Comparisons to others ➢ Connecting and disengaging- good support from health care professionals makes a difference to experience ➢ Others prefer family support ➢ Religion has a big impact on managing and coping.	Yes-aims covered

School ethos and variation in health experiences of young people with sickle cell disease at school **Dyson et al**	2012	UK	➢ To attempt to make sense of variable experiences unrelated to disease severity or to teacher/peer awareness of sickle cell disorder ➢ To provide an understanding of the experiences of young people with sickle cell at school. To provide plausible explanation of variation in school experiences.	Qualitative study	➢ A mixed methods approach was selected ➢ First questionnaires to ascertain the experiences of young people ➢ Secondly tape-recorded 1 hour interviews in the form of a guided conversation ➢ Thirdly, case studies were conducted which encompassed: 1 hour taped interview with young person, main carer ➢ A 2-week dairy of their school experiences	Young people with sickle cell aged 4 -25 years old- 21 females and 19 male participants for recorded interviews and 10 participants for the case study	➢ Approval from De Montfort University Human Research Ethics Committee, NHS and six health services research and development offices in English localities ➢ Written consent/ assent	➢ Young people with SCD faced challenges to their physical health at school. ➢ Some teachers are understanding ➢ Being labelled as lazy by teachers. ➢ Low expectations from teachers create negative ethos. - Racism ➢ Students are physically abused in relation to SCD- individual explained better off dead than living and suffering with SCD young ➢ Positive ethos-bullying and discrimination was not tolerated in school-head spoke to child and parents of the bully ➢ Teachers and Students are uneducated about SCD ➢ Parents and children prefer not to inform the school that child has SCD- they see it as embarrassing and they are worried how they will be treated. ➢ Students with SCD don't want to be treated differently but their needs should be taken into consideration ➢ Students experienced social isolation	Yes- variation was shown within participant's answers regarding their sickle cell disease
Understanding the experience of youth living worth sickle cell disease: a photo voice pilot	2013	USA	➢ To further understand young people's lived experience with SCD and examine the acceptability,	Qualitative study	➢ Photovoice- all youth received digital cameras- they were given 3 assignments related and youth	Youth who met the study inclusion (SCD diagnosis, age range within 8-17) during a	➢ Consent and assent obtained	➢ The impact of SCD and its management ➢ Coping with SCD- just trying to fit it and be normal ➢ importance of everyday activities/interest- keeps	Yes- children took pictures and explained how each picture represented

Valenzuela et al			feasibility and utility of a photovoice pilot method in that population		spent time photographing their lives according to the assignment- ➢ reviews/group discussions were completed after each project using the adapted version of SHOWeD questioning technique ➢ One additional session was conducted for both parents and youth to complete satisfaction surveys to measure acceptability of this method.	comprehensive sickle cell centre event- Enrolled participants were either placed in One group for children age 8-12 or adolescents age 13-16. A total of 16 youth participated as photographers		individuals busy, don't focus life around SCD-participate in 'normal' activities ➢ individuals don't allow SCD to prevent future/education etc.-positive thinking just go along with it ➢ Importance of family and support- acknowledge the support especially from parents and grandparents ➢ Pets were also identified as a source of support. ➢ Friends and peer relationships- few individuals tell friends or only closest know. Many have been teased but one expresses how supportive her best friends are.	their sickle cell experience
Using photovoice to explore the unique life perspectives of youth with sickle cell disease: a pilot study **Stegenga & Burks**	2013	USA	➢ Examine the perspectives of children/adolescents with SCD using photovoice both to gain understanding of the perspectives of these patients and the utility of the research method with this population.	Qualitative study	➢ Participants took photographs during camp ➢ Completed qualitative interview afterwards	12 participants with SCD between 6-14	➢ Institutional review board approval ➢ Parent permission ➢ child assent was obtained	➢ Importance of friends ➢ Controlling symptoms ➢ Importance of camp and the importance of health professionals and counsellors at camp	To some extent however, Information from children's perspective very minimal

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