An integrative review of factors that influence reproductive decisions in women with sickle cell disease

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Abstract

Sickle cell disease is one of the most prevalent genetic diseases in the world. Improvements in care have enabled those affected to experience an improved quality of life, with many affected persons now reaching reproductive age. However, pregnancy poses significant risks for women with the disease and this may affect their reproductive decisions. We conducted an integrative review of the published relevant scientific evidence to ascertain the factors that may influence these women with their reproductive decisions. The electronic databases MEDLINE, CINAHL, and PsychInfo and relevant journals were searched for peer-reviewed papers published between 2005 and 2015. Of the 440 papers identified, six papers satisfied the inclusion criteria and were assessed for quality. Results were analysed and synthesised using a thematic approach to produce a narrative report of the findings. Two main themes were identified: (1) factors influencing reproductive decision-making and (2) experiences during pregnancy. Education regarding disease knowledge and genetic implications may improve participation into screening programmes, facilitate the effective treatment needed to create trust in healthcare services, and promote better self-management. Educating professionals involved in family planning will facilitate women with this condition to make informed decisions regarding pregnancy. However, further research is required to explore the understanding of the risks involved with sickle cell disease, the best methods to educate people with sickle cell disease, and the influence that partners and families may have on women's reproductive decisions.

Keywords Integrative review · Women · Pregnancy · Decision-making · Sickle cell disease · Reproductive decisions

Introduction

With improved life expectancy in the recent years, women with sickle cell disease (SCD) are now faced with numerous considerations regarding their reproductive lives and quality of life (QOL) (Naik and Lanzkron 2012). The term SCD encompasses a variety of different genotypes, where the abnormal sickle cell (HbS) allele can be inherited, or co-inherited with another abnormal haemoglobin allele in an autosomal recessive pattern (Rees et al. 2010). The most severe form is

HbSS, where a child inherits an abnormal sickle cell allele (HbS) from both parents. A generally milder form is HbSC, where a child inherits an abnormal sickle cell allele (HbS) from one parent and another abnormal haemoglobin allele (C) from the other parent, although there are considerable similarities in the symptoms experienced. Another of the most common types of SCD is the combination of an abnormal sickle cell allele and an abnormal allele for beta thalassaemia, a disorder that causes a reduction in haemoglobin production. HbS beta thalassaemia can produce a severe or milder form of the disease, depending on the alleles inherited (United States: Centers for Disease Control and Prevention 2015; Dalibalta et al. 2010; Creary et al. 2007). Inheritance of these pathogenic alleles causes red blood cells to develop a characteristic sickle shape and potentially cause blockages within blood vessels. These blockages cause crises, which are recurrent and often extremely painful events requiring immediate medical attention, as the circulatory impairment can result in irreversible organ and tissue damage (Panepinto and Bonner 2012). SCD is most prevalent in people of African, Hispanic

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and Mediterranean descent, but as a result of international migration, it is now one of the most common inherited diseases worldwide (James 2014) and is considered an important public health issue in regions such as sub-Saharan Africa, the Caribbean, and Brazil (Aygün and Odame 2012).

QOL is a term that encompasses factors that can have an impact on a person's life, such as employment, education, relationships, and health (Mann-Jiles and Morris 2009). Authors of several studies (Thomas and Lipps 2011; Van Tuijn et al. 2010; Mann-Jiles and Morris 2009) found that the overall QOL scores of adults with SCD were lower than those of the general population and comparable to the QOL scores of people affected by other chronic conditions, such as rheumatoid arthritis, chronic obstructive pulmonary disease, or psoriasis. Employment and education influenced how adults rated their QOL, as many were unemployed or in unskilled, low paid employment (Pereira et al. 2013; Mann-Jiles and Morris 2009). Psychological wellbeing and social relationships were associated with improved QOL scores in studies by Pereira et al. (2013), Jenerette et al. (2011), and Jenerette and Lauderdale (2008), whose participants reported that their greatest satisfaction in life was related to their children, despite many being advised against childbearing and having experienced difficult pregnancies and births.

Advances in the care and management of people with SCD have enabled those affected by the condition to have an increased life expectancy (Andemariam and Browning 2013; Naik and Lanzkron 2012), with many now reaching reproductive age; defined by the World Health Organization (2017) as 15–49 years. This means that increasing numbers of affected women have the opportunity to have children. However, whilst pregnancy outcomes for women with SCD have improved significantly over several decades, systematic reviews evaluating pregnancy outcomes (Boafer et al. 2015; Oteng-Ntim et al. 2015) found that women with SCD were at an increased risk of maternal mortality, pre-eclampsia, and caesarean section, when compared to women without SCD. Babies born to mothers with SCD were more likely to be small for their gestational age and at increased risk of stillbirth and neonatal death. Boafer et al. (2015) also found that adverse maternal outcomes for women with SCD were found in both high and low income countries, which may demonstrate a lack of knowledge and provisions in managing pregnant women with SCD. Naik and Lanzkron (2012) argue that, with the correct knowledge and care, almost all women with SCD are now able to achieve a successful pregnancy. However, James (2014) states that these women may have suffered severe complications by the time they are of reproductive age, which could create significant maternal problems during pregnancy such as chronic renal disease or pulmonary hypertension. All of these factors could influence reproductive decision-making in women affected by SCD.

In addition to considering the risks associated with pregnancy, these women also have to consider the implications of their decisions on any future children, taking into account their partner's SCD status and the options that pre-implantation or prenatal diagnosis (PND) can offer (Boafer et al. 2015; James 2014). One of the aims of genetic counselling is to inform those affected by a genetic condition, such as SCD, of the options available to them to improve their own health and potentially the health of their children and support them to make healthcare and/or reproductive decisions (Harper 2010). Within this context, patients should be supported to make the reproductive choices that best suit them, without any element of coercion, to maximise their psychological wellbeing (Skirton et al. 2014).

Material and methods

The aim of this study was to systematically explore the factors that influence women with SCD in their decisions regarding pregnancy. An integrative review, conducted in a systematic manner, allows for the evidence from the varied studies to be synthesised into a meaningful summary of the available literature (Offredy and Vickers 2010). This research was conducted according to the guidelines published by the University of York's Centre for Reviews and Dissemination (2009). Ethical approval was not required for this study.

Search strategy

The following databases were used to search for literature to include in the review: Medline, CINAHL, and Psycinfo. In addition, relevant journals and reference lists were hand-searched to ensure the inclusion of all possible research. Search terms related to SCD ("SICKLE CELL" OR MeSH terms for sickle cell that included: "ANAEMIA" OR "ANEMIA" OR "SICKLE CELL" OR "SICKLE CELL TRAIT") AND terms related to pregnancy ("PREGNAN*" OR "REPRODUCT*" OR "CHILDBEAR*") AND terms related to and decision-making ("INFORM*" OR "SUPPORT*" OR "NEED*" OR "COUNSEL*" OR "PSYCHOLOG*" OR "DECISION*" OR "RISK*" OR "PSYCHOSOCIAL*") were combined and searched for within any part of the text.

Inclusion and exclusion criteria

Papers were included if they were as follows: (i) reports of research using quantitative, qualitative, or mixed methods design, (ii) published in English between 2005 and 2015, (iii) focused on the reproductive experiences of women with SCD, and (iv) focused on factors that influenced women with SCD in their decision-making regarding pregnancy. Papers were excluded if they were as follows: (i) about the care and

management of people with SCD that was unrelated to reproductive issues, (ii) about the reproductive decisions of women who were carriers and not physically affected by SCD, (iii) where the data from women with SCD could not be differentiated from the overall data, (iv) where the data regarding reproductive issues was not underpinned by the opinions/ attitudes of the women with SCD themselves, or (v) if they did not include primary empirical data.

Search outcome

The Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) process was followed, with details of the search outcome presented in the PRISMA flow diagram (Supplemental Fig. 1) (Moher et al. 2009). The initial search identified 440 potential papers for inclusion. Of these, 87 were duplicates, leaving 353 papers for examination. The titles and abstracts of each paper were assessed for eligibility against the inclusion and exclusion criteria, by two independent researchers. Twenty-five papers were included for assessment of the full text. Of these, 19 were excluded (see Supplemental Fig. 1). The six remaining papers were subjected to quality assessment.

Quality assessment

The six papers that met the selection criteria were assessed using the appraisal tool developed by Kmet et al. (2004). This tool provides a systematic method of evaluating both quantitative and qualitative research. The appraisal tool comprises a series of quality criteria questions: a judgement was made as to whether the criterion is “fully met,” “partially met,” or “not met.” The author and a second appraiser independently assessed each paper, with the scoring discussed until a consensus was reached, to reduce subjective judgement (Gerrish and Lacey 2010). All studies were included in the review and notes on the quality assessment can be found in the [Supplemental table](#).

Data abstraction and synthesis

The majority of the papers were qualitative in design, which made a meta-analysis of the findings inappropriate (Polit and Beck 2010). With both quantitative and qualitative studies included into the review, a narrative approach was the most appropriate method for identifying themes and analysing the findings of studies that are diverse in design (The University of York: Centre for Reviews and Dissemination 2009; Gough et al. 2012; Polit and Beck 2010). A table of the individual characteristics of the studies was prepared (Table 1), then the thematic analysis approach, described by Braun and Clarke (2006), was used to produce a narrative report with both authors involved in the analysis and synthesis of the data.

Results

The six papers represented research conducted in four different countries; Brazil, Cameroon, Jamaica, and the USA. The majority of the researchers used a qualitative approach with the use of interviews as their data collection tool, whilst Wonkam et al. (2014) utilised a quantitative approach and a structured questionnaire. Table 1 provides original data from the included studies. It should be noted that in three of the studies the same sample was used, but we have included all three studies as they focused on different aspects of the research question and the data in all three studies were relevant to this review. Two main themes emerged from synthesis of the six studies and the results are presented under those themes.

Theme 1. Factors influencing reproductive decision-making

In the context of reproductive decision-making, knowledge of the health and social implications and cause of the condition could be deemed to be relevant. Information or impressions gained from healthcare professionals were found to have had an impact on women’s reproductive knowledge and decisions. In the first instance, this related to potential fertility. Da Cruz Santos et al. (2012) and Silva Gomes Xavier et al. (2014) found that participants received varied advice from doctors regarding fertility and contraceptive use. Some were told they should not have any problems getting pregnant, whilst others were told they were unable or unlikely to get pregnant because they had SCD, or reported receiving the general impression that a pregnancy would be unlikely. Having received the impression or advice that they were unable to become pregnant, many women chose not to use contraceptives (Da Cruz Santos et al. 2012; Silva Gomes Xavier et al. 2014), which effectively reduced their ability to make their own reproductive decisions.

Although knowledge of the condition has a potential impact on reproductive decision-making, authors of only one study (Anderson and Asnani, 2015) explored their participants’ general knowledge about SCD. Almost two-thirds (63.3%; $n = 19$) of their participants, who lived in Jamaica, believed that their white blood cells were attacking their red blood cells. Of these, 84.2% ($n = 16$) reported that they had been given this information by a healthcare professional or had read it in the literature they had obtained from their local sickle cell clinic. Whilst almost all participants (83.3%) in the same study correctly knew that SCD was inherited from both parents, however the knowledge regarding the probability of transmission varied and was often incorrect.

Decision-making regarding pregnancy was also shown to be influenced by other factors, including the participant’s attitude to the condition, their beliefs about their ability to manage their child’s condition and the actual risks of inheritance.

Table 1 Summary of papers included in the review

Authors and reference (country of study)	Aim	Methodology	Sample and size	Method of analysis	Main results or findings	Quality assessment comments
Anderson and Asnani (2015) (Jamaica)	“To explore lay understandings of sickle cell disease (SCD) among Jamaicans living with the illness” (pg. 1).	Qualitative, exploratory, descriptive study.	Purposive, convenience sample of 30 participants (male & female) 18+ years, at routine SCD visits, not in acute pain.	Atlas.ti qualitative data analysis software for thematic analysis, constant comparison and case analysis.	Inaccurate knowledge of how SCD works. 25/30: inherited from both parents but probability of inheritance was varied. 19/30: for partner testing to avoid SCD. 16/30 willing to have child with SCD.	Sample obtained from one setting. No mention of reflexivity.
Thompson Ross (2015) (USA)	“To explore the motivations of women with SCD for identifying their partners’ sickle cell status—either trait or disease—via genetic testing” (pg. 37).	Qualitative, descriptive, phenomenological study.	28 African-American women with SCD aged 18–52 years. Recruited from 4 different places.	Colaizzi’s phenomenological method. Described in detail.	79% women had asked their partners to undergo testing. Motivations included: concerns about physical suffering, feelings of guilt, determining whether to enter into or remain in an intimate relationship.	Design not clear but not inappropriate. Reflexivity mentioned but not discussed. No mention of verification procedures.
Wonkam et al. (2014) (Cameroon)	“To study the attitudes of adult patients suffering with SCD on prenatal genetic diagnosis (PND) and possible TAP” (termination of an affected pregnancy) (pg. 615).	Quantitative, descriptive, cross-sectional study.	Volunteer, purposeful and convenience sample of 89 participants, aged 18+ with a confirmed diagnosis of SCD.	SPSS, V.17.0 for descriptive analysis. H test of Kruskal-Wallis or Z test Kolmogorov-Smirnov for variable relationships.	Had severe forms of SCD. 88.2% accepted PND for SCD. 40.9% accepted TAP for SCD. For: negative personal experiences, fear of more affected children with poor quality of life, ethical or economic reasons. Against: ethical and religious reasons	Qualitative approach may be more suitable. Possible answers could be better defined for clarification.
Silva Gomes Xavier et al. (2014) (Brazil)	“To meet some aspects related to the use of contraception by women with sickle cell anaemia” (pg. 28).	Qualitative, descriptive study.	Purposive, convenience sample of 25 women with reproductive experience and a diagnosis of sickle cell anaemia.	Barden’s content analysis—categorised analysis thematic mode.	All women became pregnant. Contraceptive usage increased after a negative pregnancy experience. Doctors influenced choices by informing women they were unlikely/could not get pregnant.	Theoretical framework not well described. Reflexivity or verification procedures were not discussed.
Silva Gomes Xavier et al. (2013) (Brazil)	“To analyse the perception of women suffering from sickle cell anaemia regarding pregnancy” (pg. 834).	Qualitative, exploratory, descriptive design.	A purposive, convenience sample of 25 women with reproductive experience, diagnosed with sickle cell anaemia.	Collective subject discourse	56% had 1+ pregnancy. 72% had complications during pregnancy. Main themes: Fear and anxiety, dilemmas regarding the abortion decision, desire to be a mother frustrated by miscarriage.	Theoretical framework and data collection methods not well described. No mention of verification procedures.
Da Cruz Santos et al. (2012) (Brazil)	“To describe the feelings experienced by women with sickle cell anaemia with regard to their reproductive experiences” (pg. 2934).	Qualitative, exploratory, descriptive design.	A purposive, convenience sample of 25 women with a confirmed diagnosis of sickle cell anaemia and previous reproductive experience.	Thematic content analysis. Not described.	Many believed that they were infertile. Pregnancy created: fear and guilt, happiness and victory, sadness and trauma, frustration, loneliness, regret due to consequences of complications.	Theoretical framework, sample, data collection and analysis not well described. No mention of verification.

Sixteen of the 30 (53.3%) participants in one study (Anderson and Asnani 2015) stated that they would want a child regardless of whether the child had SCD or not. Of these, 50% ($n = 8$) of the participants gained confidence through their own experiences of the condition, whilst 31.2% ($n = 5$) viewed the situation in a positive light, choosing to focus on the possibility that their unborn child might not inherit an abnormal sickle cell allele from both parents. Strategies to reduce the risks to any children included choosing to have only one child, paying close attention to the child's health and diet or planning to have children with a partner who was not a carrier. Participants in other studies were more concerned about having an affected child and used knowledge of their partner's sickle cell status to help them to plan for an unaffected pregnancy. For example, in a study exploring contraceptive use, Thompson Ross (2015) found the motivating factors for women requesting their partners be tested included a desire to protect future children from suffering physical pain. Women in that study had been advised by their families to avoid a partner carrying the sickle cell trait. As many as 79% ($n = 22$) asked their partners to undergo testing, some before becoming intimate with them and subsequently ended the relationship if their partner was found to be a carrier (Thompson Ross 2015). These findings are supported by those of Wonkam et al. (2014), who explored the views of people living with SCD on PND and the termination of an affected pregnancy, finding that 100% supported premarital screening to determine the genetic status of both partners.

However, obtaining information about the partner's carrier status was not always straightforward and the onus seemed often to fall mainly on the female partners. Whilst Anderson and Asnani (2015) reported that two of the three men who did not ask their partners to be tested stated that they were ashamed of their sickle cell status, half of the women with untested partners expressed frustration at their partner's refusal to engage in prenatal testing. Some of the women who participated in Thompson Ross's study (2015) reported witnessing the guilt that their parents experienced, whilst others expressed anger towards their parents, claiming that their behavior was irresponsible. However, some did acknowledge that there was a lack of information available to their parents at the time and that it was possible that they were unaware of their sickle cell status. Participants whose mothers knew their sickle cell status felt that it was their mother's fault for not getting their fathers tested, as they felt that mothering begins before conception and that taking partners to be tested is a part of a woman's responsible sexual behaviour.

For those who wish to have a child, PND of an affected pregnancy is a possibility, which may influence the decision to have a pregnancy. Exploring the opinions of those living with SCD on PND and termination of affected pregnancies, Wonkam et al. (2014) found that nearly all participants (96.5%) were in favour of PND in general, yet less (88.2%)

supported PND specifically for SCD. A similar result was found when participants reported how they felt about the termination of an affected pregnancy, with 53% supporting the termination of an affected pregnancy in general and 40.9% in favour of the termination of an affected pregnancy for SCD specifically. Of those who were against PND and the termination of an affected pregnancy for SCD, 45.5% stated religious considerations as the rationale and 78.1% stated ethical reasons, although these were not specified. Conversely, ethical reasons were also cited as the rationale of 61.5% of those in support of PND and the termination of an affected pregnancy for SCD. Personal negative experiences of the condition, economic stresses, and concerns about their child's QOL were also reasons for supporting PND and possible TOP.

Theme 2. Experiences during pregnancy

For those women who became pregnant, the experience of pregnancy had both positive and negative aspects: these experiences could influence decisions about subsequent pregnancies or could have an impact on the decisions of other women in their families or friendship groups. Many women had complicated pregnancies and births, necessitating significant medical intervention. Of the 25 women in the study by Silva Gomes Xavier et al. (2014), 72% ($n = 18$) experienced complications during pregnancy and/ or childbirth; deterioration in their anaemia, an increased need for transfusions with prolonged hospital stays, premature, and/ or low birth weights; and urinary tract infections. Twenty-eight percent ($n = 7$) of the women experienced spontaneous abortion, 20% ($n = 5$) experienced stillbirths, and 8% ($n = 2$) induced the abortion themselves.

For many women, there was an immense emotional and psychological impact connected with the pregnancy and this was sometimes connected with their perceived lack of healthcare support. Silva Gomes Xavier et al. (2013) explored women's perceptions of their pregnancy experiences in depth, finding that fear and anxiety were significant emotions felt by the women with SCD. Women shared their anxiety about their child's health and fears for their own death and that of their unborn child. They felt anxious throughout due to a lack of structured services, trained healthcare professionals who understood how to manage their condition, and a lack of beds for high-risk pregnancies in maternity hospitals, as they were often told to look elsewhere for care. In addition to this, these women often had a low economic status resulting in financial concerns and overall reported feeling no joy throughout their pregnancies, experiencing anger, crying, and hopelessness instead.

Da Cruz Santos et al. (2012) also explored women's feelings about their reproductive experiences, finding that they experienced guilt about the possibility that their child might be affected by SCD, and feared for their own death. Grief, sadness, shock, mourning, and trauma were described when discussing their experiences of abortions or stillbirths, along

with immense frustration at not being able to be a mother. Those whose pregnancies were successful experienced frustration at their inability to breastfeed their babies, whilst others spoke of feelings of regret, as they were faced with complications and consequences of the pregnancy, such as necrosis of the hip.

For some, the anxiety associated with the pregnancy influenced their wish to have an abortion. However, despite their fears for themselves and their babies, some of the women in the study by Silva Gomes Xavier et al. (2013) felt that they could not discuss the option of the termination of an affected pregnancy with anyone, as it was criminal offence (in their country), and expressed great concern over what their families might think of them. Others attempted, some successfully, to induce a termination themselves with teas and medicines as they felt they had no support, despite not seeking medical advice. They struggled with feelings of guilt as they felt what they were doing was a sin, praying for support and forgiveness.

However, having children was a very significant factor in the lives of some women. Some reported a loss of self-esteem whilst childless, as they did not feel like “normal” women, and some feared that they would lose their partners to other women if they could not bear a child (Silva Gomes Xavier 2013). These findings were further supported by reports of women whose pregnancies were successful, who experienced a sense of victory in overcoming their perceived limitations (Da Cruz Santos et al. 2012).

Discussion

Factors influencing reproductive decision-making

The fertility of women with SCD can be affected by the wide array of complications that can arise from unpredictable episodes of crises and the medical therapies used to prevent or manage them (Smith-Whitley 2014). Therefore, it is possible that the varied medical opinions given to the women with SCD, regarding pregnancy and contraception, were accurate. However, Smith-Whitley (2014) also states that there are limited data available from well-designed, large clinical studies that could contribute towards the knowledge base regarding the fertility of women with SCD. Mütze et al. (2016) had a similar finding within their study to evaluate the services available to adults living with the genetic condition phenylketonuria (PKU), an inherited metabolic disorder. Increasing numbers of people affected by PKU are now reaching reproductive age, with most having had their conditions managed in paediatric centres and few specialised centres available to them once they reach adulthood. Most participants in the study reported not adhering to the recommended diet and treatment as strictly as they did during their childhood, meaning any potential children could have been subjected to the

severe embryopathy risks associated with mothers who have poorly-controlled phenylketone levels. Cammidge et al. (2016) also conducted a study to explore the motherhood experiences of women with another inherited condition, cystic fibrosis (CF) in which life expectancy has improved over the past few decades. The authors found that many women with CF perceived variations in the support they received from their CF teams regarding pregnancy, reporting little or inaccurate information and care.

Whilst some women in Thompson Ross (2015) study avoided relationships with partners found to have SCT, other women in Anderson and Asnani (2015) study experienced difficulty engaging their partners with testing. Atkin et al. (2015) conducted a study to try and understand the expectations and experiences of fathers engaging in a UK sickle cell screening programme, finding that whilst fathers rarely understood the implications of the tests, they were happy to undergo testing as they felt it was important to comply as a demonstration of commitment to the pregnancy. Fathers did not feel jointly involved and felt that their only contribution was as a source of genetic material, with a perceived pressure to produce a healthy child. This perceived pressure is reflected in findings from Anderson and Asnani's (2015) study, within this literature review, where some men stated that they were ashamed of their sickle cell status and women expressed frustration at their partner's refusal to engage in prenatal testing. To achieve a significant reduction in stigma, cultural and societal changes are needed, but education about the genetic implications of the disease with its recessive inheritance pattern may improve the involvement and understanding of genetic screening programmes.

A significant number of participants in this present review, both male and female, supported premarital screening and/ or expressed a preference to undergo testing to avoid having a child with SCD (Anderson and Asnani 2015; Wonkam et al. 2014). Memish and Saeedi (2011) exploring the outcome of a national premarital screening programme for SCD and β -thalassaemia demonstrated that despite the use and availability of screening for prevention, there remained a large number of people at risk of having children affected with SCD who chose to have children. This is reflected within the present review, as women explained that they chose not to encourage their partners to get tested because they were unwilling to limit their romantic and reproductive lives, and that they were grateful for the opportunity of life given to them (Thompson Ross 2015). This identifies a need to provide support and education for women regarding the variety of potential consequences of SCD, which can vary significantly from person to person (Andemariam and Browning 2013; Panepinto and Bonner 2012). Mütze et al. (2016) and Cammidge et al. (2016) had similar findings in their respective studies on PKU and CF. Mütze et al. (2016) reported that women with PKU were not discouraged from becoming mothers regardless of their prior

management of their condition, whilst Cammidge et al. (2016) found that some women with CF even reduced the contact they had with their CF teams or doctor, if they perceived a lack of support with their decision, which consequently had an impact on their own care. The risks some women are prepared to take to achieve their desire to become a mother is also highlighted in a case report by Godlewski and Castellanos's (2017); a woman living with spinal muscular atrophy (SMA) chose to become pregnant despite being counselled about the possible serious risks to herself and her child. At 27 weeks, the woman became unresponsive and an emergency caesarean was performed. She survived but her baby died after suffering hypoxia.

The majority of the participants with SCD enrolled into Wonkam et al.'s (2014) study stated that they were in favour of PND for SCD, whilst significantly fewer were in favour of the termination of an affected pregnancy. Negative experience and fear were the rationale for the termination of an affected pregnancy for other women, who expressed a fear of having a sick child with SCD, or a fear that their child or they themselves may die during pregnancy or childbirth (Silva Gomes et al. 2013). Ahmed et al. (2015) explored the decision-making related to PND and inherited retinal disease, finding that most participants were also in favour of PND but against TOP. However, participants did acknowledge that circumstance and experiences would have an impact on individual opinions of the subject. A majority of those participants did not feel that their QOL was impacted enough by their condition to justify a TOP, but felt that it may be more supported by others for severe diseases. This finding is supported by Hewison et al. (2007), who studied the attitudes that 420 women in the UK had towards PND and TOP for 30 different conditions. Similar results were found in respect of greater support for PND than TOP; however, exceptions were made for what were considered severe conditions, such as anencephaly, trisomy 13 or 18, quadriplegia, or Duchene muscular dystrophy.

Ahmed et al. (2012) conducted a study to explore the attitudes of parents and relatives of children with recessive conditions. Their findings demonstrated that men were more in favour of PND and the termination of an affected pregnancy than women. Women made up the majority of Wonkam et al.'s (2014) sample, which may have influenced the lower percentage in favour of the termination of an affected pregnancy. The findings from these studies demonstrate that women may have differing opinions towards PND and the termination of pregnancy; however, they may be potentially influenced by their partners when making a final decision. Support and education for both partners are therefore required to facilitate informed reproductive decision-making.

Experiences during pregnancy

Grief, sadness, shock, mourning, and trauma were felt by women who experienced an abortion or stillbirth, in this

present study, along with the immense frustration at not being able to be a mother. These responses are similar to the findings of Sives (2016), who conducted research exploring the experiences of women who were unintentionally childless. Women reported frustration and anger and a sense of failure, especially when surrounded by other women with children. There was a perception that being a real woman involved being a mother and women felt that others judged them for not having children, leaving them feeling isolated in their grief and estranged from society.

Guilt was also a significant emotion experienced by women with SCD in this review. Women, who tried to induce a termination of their pregnancy, experienced guilt as they felt what they were doing was a sin (Silva Gomes Xavier et al. 2013). Robinson (2014) states that all pregnancy losses, whether unplanned or chosen, are associated with guilt and self-blame. Women with CF within Cammidge et al. (2016) study also reported experiencing similar emotions, despite having had a successful pregnancy. They acknowledged that their frequent hospital admissions, treatment, and shortened life expectancy led to upset, frustration, and feelings of guilt. However, no mother expressed any regrets over their decision to have children, with enjoyment, disbelief at becoming a parent, and feeling a great sense of achievement being reported from the findings. Women in this present study, whose pregnancies were successful, also reported experiencing a huge sense of victory and empowerment from overcoming their limitations and perceived capabilities. They experienced relief and happiness at having achieved a successful pregnancy (Da Cruz Santos et al. 2012). Participants with SCD, in studies by Jenerette et al. (2011) and Jenerette and Lauderdale (2008), expressed similar emotions when asked about their life satisfaction. Despite many being advised against pregnancy and having also experienced difficult pregnancies and births, the most frequent response regarding life satisfaction was related to their children. This highlights the important significance that having children has on the QOL of women with SCD, and efforts should be made to support women who would like to experience having their own family.

Strengths and limitations of the review

This integrative review was conducted systematically and in accordance with the protocol set out by the University of York's Centre for Reviews and Dissemination guidelines (2009). The choice of protocol was justified with the findings from a preliminary literature review, which identified the potential inclusion of both quantitative and qualitative studies. Relevant journals were hand-searched to improve study detection rate and the use of two independent assessors supported the rigour of the study. Restricting the criteria to peer-reviewed papers strengthens the ability to make recommendations for

improved evidence-based practice, but exclusion of grey literature may be viewed as a limitation. The exclusion of any papers not written in English, due to the authors only being literate in English, was also a limitation of the review. In addition, authors of four of the six studies recruited women only, and three of the studies were conducted using the same sample.

Conclusion and recommendations for practice

This review draws attention to the multiple factors that may influence women with SCD in their decisions regarding pregnancy. The findings demonstrate that there is a lack of knowledge regarding the reproductive capabilities of women with SCD, which if improved could lead to better experiences and reproductive outcomes for affected women. Women with SCD therefore need to have access to education about their condition and reproductive choices to enable them to make considered and informed decisions regarding what could be a potentially life changing event. The influence of partners in these situations should not be underestimated and efforts should be made to improve awareness and involve them if appropriate.

However, even with appropriate education and availability of screening and testing, there remains a large number of women who would be prepared to have a child with SCD and/ or are prepared to continue to try for a successful pregnancy. It is important that healthcare professionals are educated, skilled and have the resources to be able to offer support and provide safe care, to ensure equity of opportunity for these women.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethics approval and consent to participate This article does not contain any studies with human or animal subjects performed by any of the authors.

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