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Keeping Warm with Sickle Cell Disease Research Project



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Report for the Cheshire Lehmann Fund

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and Social Care
Research**

Executive summary

Sickle cell disease (SCD) can lead to frequent hospitalisation for extremely painful sickle crisis and to several severely disabling secondary conditions or even death. 12,000-15,000 people in the UK have SCD and it affects people of all ages. One of the main triggers for a crisis is getting cold. Therefore staying warm, as well as eating well, drinking plenty and avoiding stress, is recommended by healthcare professionals as a preventative strategy.

As SCD is most common among Black British, Black African and Black Caribbean groups, who are already at higher risk to fuel poverty than the general population, combined with increasing fuel costs, maintaining a home warm enough to avoid a crisis may be extremely challenging. This research looks at challenges people with SCD have in staying warm enough to prevent a painful sickle crisis.

15 participants were interviewed, using a semi-structured questionnaire with adults with SCD or parents of a child with SCD. Interviews were done across West and South Yorkshire; Manchester and Birmingham. Room temperature and humidity readings were taken for a maximum of 2 weeks for 6 of the participants.

The results revealed five major themes. These were: 1. How SCD crisis was linked to cold; 2. Heating behaviours of people with SCD; 3. Housing and energy efficiency; 4. Why people with SCD are at particularly high risk of fuel poverty; and 5. How heating impacts on other triggers for SCD.

Participants cited different triggers for a crisis, including getting cold, not eating well, dehydration and stress. Participants described different examples of where they had got cold, which led to their being hospitalised with a crisis and was seen by many, but not all, as a major trigger. Because pain was experienced rapidly with exposure to cold, participants believed there was a causal effect between getting cold and feeling pain. Whilst the trigger of cold could be a minor event the consequences could be severe, resulting in days in hospital and in some cases leading to a critical condition. Using the temperature monitors, a drop in room temperature at night in the bedroom of one participant was shown to coincide with the onset of a crisis that led to 2 days in hospital.

The room temperatures that participants reported being comfortable at were between 20 °C and 30 °C and all said they needed to be warmer than people without SCD. The temperature monitoring revealed lower room temperatures than what was said to be comfortable. This was partly explained by people being out of the house or having no money to put on the meter. 'Spatial shrink' was practised by some participants so they could save money on heating by staying in one heated room, but this could result in pain when moving to a colder area in the home. Some participants had trouble getting landlords to improve the energy efficiency of their homes and others had trouble finding the time to seek grants or discounts for heating or energy efficiency due to illness or caring burdens. In addition to heating, wrapping up warm was seen as important for staying warm both indoors and outdoors. Social isolation was experienced where participants could not leave their homes due to cold or rainy weather or where the place they were visiting was not expected to be warm enough.

In addition to being at high risk of harm from cold, the condition can also put people at risk of periods with no income whatsoever. This was a particularly critical situation for those on gas and electricity meters and who rely on sufficient cash availability to top up the meters. Having no income or a rapid drop in income, for some participants, was the result of SCD being a fluctuating condition. Because of its fluctuating nature and many people with SCD being relatively young and appearing to be well, participants did not always qualify for disability benefits. However when they were put on Jobseekers Allowance (JSA) they were sometimes ill or in hospital on the day of their signing on appointment and were immediately sanctioned, even if they rang up to explain. Three of the fifteen participants had no money at all at the time of the interview due to sanctions. Whilst the JSA was usually reinstated it could take weeks to come through due to delays in obtaining medical evidence and letters getting lost in welfare offices. Maintaining employment was also difficult without an understanding employer or full employment rights to allow for periods of sickness or caring for a sick child. The difficulty in maintaining a steady income, even a low one, combined with the higher costs of heating had a knock-on effect for other triggers – not eating well, dehydration (warm drinks are sometimes essential) and stress.

It would be hard to find another group that is at such high risk of physical harm from cold homes but at the same time at such high risk of fuel poverty. On the other hand there is much potential to reduce cold related harm in this group through a range of fuel poverty interventions such as improving energy efficiency, income and finding the best payment method and supplier. Hospital admissions due to SCD crisis cost thousands of pounds per admission and risk leaving the person with SCD with lifelong consequences such as organ failure. With an annual heating bill of around £1,000, it may be more cost effective to part or fully fund heating bills for people with SCD, so providing increased capacity to stay warm, eat well, drink plenty and avoid stress and repeated crisis. Helping people with SCD could be made even more cost effective by taking into account the positive effect on health for other members of the household with other conditions affected by cold.

In addition, working out how to address heating among the SCD population could help find solutions for people with other conditions, especially fluctuating conditions such as MS, arthritis and mental illness where people are at similar risk of not being able to work, qualify for disability benefits or avoid JSA sanctions. The challenge will be to match investment in addressing fuel poverty in groups with cold-vulnerable conditions to savings in health and social care and to see 'heat as medicine'.

Background

A sickle cell disease (SCD) crisis can lead to severe pain, stroke, anaemia, osteomyelitis, acute chest syndrome, kidney damage, cardiac failure, multi-organ failure and complications from treatment¹. Child SCD sufferers lose weeks of schooling per year² and adults may find it difficult to maintain regular employment if they experience regular attacks. The average lifetime hospital costs of a person with SCD at 35 years of age can be as much as £185,000³. One percent of all children born in the UK and 9% of ethnic minorities carry the trait for a haemoglobin disorder and there are between 12,000 and 15,000 people with SCD in the UK⁴. People with SCD are often told by doctors that cold can trigger a painful sickle cell crisis and that they must keep themselves warm⁵. They also experience hypersensitivity to cold⁶ which may greatly influence their heating choices. A recent study shows how intensely painful cold can be for people with SCD, whereas those without the condition are not affected; for example, when a child is sent out to the playground without a coat: "It was, they were just like, 'Oh it's not that cold'. (...) I got really numb and I got really still and then I just started to cry. I was in one spot crying real hard and I just felt this pain and a half." Participant G7 p 472.⁷ Almost all SCD crisis prevention and care guidance recommend that people with SCD avoid getting cold and that they wrap up warm⁸.

There is a growing evidence base on the association between cold temperatures and sickle cell crisis with subsequent complications^{9,10,11,12,13,14}. People with SCD have specific heating

¹ N.C.E.P.O.D (2008) *A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death*. http://www.ncepod.org.uk/2008report1/Downloads/Sickle_report.pdf

² Dyson op cit

³ Karnon J, Zeuner D, Ades AE, Efimba W, Brown J, Yardumian A (2000). The effects of neonatal screening for sickle cell disorders on lifetime treatment costs and early deaths avoided: a modelling approach. *Journal of Public Health Medicine*. 22(4):500-11.

⁴ N.I.C.E. (2012). *Sickle cell acute painful episode: management of an acute painful sickle cell episode in hospital* Available at: <http://www.nice.org.uk/guidance/cg143>

⁵ Brousse V (2014) Management of sickle cell disease in the community *British Medical Journal* 2014;348:g1765

⁶ Brandow AM, Stucky CL, Hillery CA, Hoffmann RG, Panepinto JA. (2013) Patients with sickle cell disease have increased sensitivity to cold and heat. *American Journal of Hematology*. 88(1):37-43.

⁷ Dyson S, Atkin K, Culley L, Dyson S, Evans H, (2011). Sickle cell, habitual dyspositions and fragile dispositions: young people with sickle cell at school *Sociology of Health and Illness* 33 (3):465-483

⁸ Oni L, Dick M, Smalling B, Walters J. (2013) *Care and management of your child with Sickle Cell Disease - A parents' guide*. The NHS Sickle Cell & Thalassaemia Screening Programme. Available at <http://sct.screening.nhs.uk/parentsguide>

⁹ Jones S, Duncan ER, Thomas N, Walters J, Dick MC, Height SE, Stephens AD, Thein SL, Rees DC. (2005). Windy weather and low humidity are associated with an increased number of hospital admissions for acute pain and sickle cell disease in an urban environment with a maritime temperate climate. *British Journal of Haematology*. 131(4):530-3.

¹⁰ Mohan J, Marshall JM, Reid HL, Thomas PW, Hambleton I, Serjeant GR. (1998) Peripheral vascular response to mild indirect cooling in patients with homozygous sickle cell (SS) disease and the frequency of painful crisis. *Clinical Science*. Feb;94(2):111-20.

¹¹ Molokie RE, Wang ZJ, Wilkie DJ. (2011). Presence of neuropathic pain as an underlying mechanism for pain associated with cold weather in patients with sickle cell disease. *Medical Hypotheses*. 77(4):491-3.

¹² Paul R, Castro O, O'Neal P (2011) Acute Chest Syndrome - Sickle cell disease *European Journal of Haematology* 87(3):191-207

¹³ Rogovik AL, Persaud J, Friedman JN, Kirby MA, Goldman RD (2011). Pediatric vasoocclusive crisis and weather conditions. *Journal of Emergency Medicine*. 41(5):559-65

requirements and need to be warm at all times to avoid painful exacerbations and often lengthy stays in hospital. How people with SCD manage to stay warm at home, especially where there are problems with housing conditions and difficulties paying for fuel bills, has not been fully explored.

Fuel poverty research in the UK has demonstrated that a significant proportion of the population in the UK find it hard to maintain a sufficiently warm home¹⁵. Fuel poverty grants and schemes providing financial help, energy efficiency measures and improved heating systems have not been able to reduce the number in fuel poverty, partly because of increasing energy prices and reductions in household incomes. People from ethnic minority communities in the UK are not only more likely to suffer from SCD but are also much more likely to be on a low income¹⁶ and therefore at higher risk of fuel poverty. Furthermore, there is a great awareness among people with SCD and their carers of the need to keep warm¹⁷ and, with their higher sensitivity to cold, they are likely to have particular concerns about maintaining a warm home and the associated cost. There is therefore an urgent need to initiate research into fuel poverty, heating needs and behaviour and the prevention of temperature-related sickle cell crisis.

There is little or no research on SCD and fuel poverty, and that on heating use appears to be almost non-existent. Alongside this there is a similar general paucity of research on the heating needs of people with chronic conditions exacerbated by cold. This research would be a starting point, aimed at developing more appropriate guidelines for maintaining homes that are not so cold that they are detrimental to health and where energy services are fair and accessible for all groups in society.

AIMS OF THE STUDY

To explore the heating and energy requirements and behaviours of people with sickle cell disease and to understand what opportunities there are to increase effectiveness and efficiency of delivery and take-up of energy use and services.

¹⁴ Smith W, Bauserman R, Ballas S, McCarthy W, Steinburg M, Swerdlow M, Waclawiw M, Barton B (2009). Climatic and geographic temporal patterns of pain in the Multicenter Study of Hydroxurea *Pain* 146 (1-2):91–98

¹⁵ Friends of the Earth and Marmot Review Team (2011). *The Health Impacts of Cold Homes and Fuel Poverty*.

¹⁶ Majeed A, Banarsee R, Molokhia M. Health disparities and community participation in England. (2009). *Journal of Ambulatory Care Management*. 32(4):280-4.

¹⁷ Molokie op cit

METHODS

A semi-structured questionnaire was used and room temperature and humidity readings taken where possible using TinyTag2 recorders. An advisory group was set up including a fuel poverty advisor, a representative of a sickle cell disease charity who also had the condition and a Warm Homes worker for a metropolitan council. Ethics approval was obtained from Sheffield Hallam University's Health and Wellbeing Faculty Ethics Committee.

i. Selection criteria

Adults with SCD or carers of a child (<18 years old) with SCD

ii. Recruitment

Recruitment was facilitated through email, telephone or face to face contact with people with SCD by ASYABI (Association of Sickle Cell Sufferers of Yorkshire and Africa and Basic Information); Oscar Birmingham; Oscar Sandwell; information leaflets left at a sickle cell clinic and the contacts of Maria Berghs (University of York), a posting on social media from The Sickle Cell Society and through snowballing with participants.

iii. Research process

After explaining the research, consent was taken and interviews conducted for a total of 15 adults with SCD or parents of a child with SCD. Nine of the participants were interviewed face to face, 3 participated in a small group interview and another 3 were interviewed by telephone. All of the participants had the full blown SCD, apart from one who had sickle cell trait but had experienced a crisis. The location of interview and option to record room temperatures depended on what was most convenient for the participants. Interviews lasted between 25 minutes and 70 minutes. The interviews were recorded using a digital recorder and transcribed. These methods were used to suit the participants. It was originally planned that the participants would be just from the Yorkshire area to reduce costs but when recruitment was found to be slow the geographic area was spread to include, Manchester and Birmingham as well. This was ultimately an advantage because it increased the level of anonymity, which can be difficult when undertaking research with people with a rare disease, especially where they might be expressing concerns about local services. Participants were interviewed where it was most convenient for them – in their own home, in a room of a voluntary sector organisation or by telephone. The participants were offered a £20 high street gift voucher as a gesture of gratitude for their participation, as well as travel expenses where incurred.

Six of the participants had room temperatures and humidity recorded in their own homes. This involved two visits, one to drop off the temperature monitors and approximately two weeks later conduct the interview. The TinyTag2 recorders automatically recorded room temperature and humidity every hour for a maximum of two weeks.

The characteristics of the participants are given in the table below, which shows the participants had a mixture of ages, tenure, employment status and payment type. All but one participant had gas central heating and all participants were either Black British, Black Caribbean or Black African. There were no criteria for selecting by ethnicity in the recruitment

process. The ethnicity of the participants reflects the higher incidence of SCD in groups of African origin. Unless stated, the information below refers to the participant – i.e. the adult with SCD or the parent of a child with SCD.

Table 1 Participant characteristics

Characteristic	Number in category	
Age of person with SCD (n=15)	<18 years	5
	19-35	4
	36-45	3
	41-55	1
	56-65	2
Gender of person with SCD	Male	6
	Female	9
Household tenure (n=15)	Housing association	3
	Local Authority landlord	3
	Private rented	3
	Home owner	2
	Living with parents	1
	Living with grandparent	1
	Homeless	1
Employment/income status of participant (n=15)	Working	5
	Claiming benefits	6
	No work or income	3
Highest qualification of participant (equivalent to UK) (n=12)	University level	6
	A Level	2
	GNVQ	4
Ethnicity of participant (n=15)	Black British	10
	Black African	2
	Black Caribbean	3
Fuel bill payment type (n=12)	Meter	4
	Direct debit/bill	7
	Taken out of benefits	1
Heating system (n=15)	Gas central heating	14
	Storage heating	1
	Electric radiators used as well as heating system (n= 11)	9

RESULTS

The themes were developed inductively by manually extracting all emerging themes and sub-themes and mapping them out using mind maps. There were no pre-conceived frameworks applied to the themes that emerged; these came entirely from the data. (Charmaz 2014, Strauss & Corbin 1998). The main themes that emerged are presented below.

1. How sickle cell crisis and pain are linked to being cold

Current evidence around the link between cold temperatures and SCD crisis focuses on climates and seasons. This evidence looks at associations of temperature and SCD pain and causal effects or explanations are rarely addressed. Whilst there is no sure way to determine what has triggered a crisis, and there may be many factors, cold was reported by the participants in this study to be a direct cause of pain. This is evidenced by multiple explanations of how they became cold, how quickly the pain developed and the belief that cold can trigger a crisis often came from years of lifestyle choices involved in preventing pain and a crisis. The below results discuss the triggers and experience of a crisis, where cold is identified as a trigger for a crisis. There is also a case study of a crisis that was attributed directly to cold by the participant, including room temperature data at the time of the onset of the crisis. How cold affects people with sickle cell trait is also discussed. Whilst not all the examples are about heating in the home, they show how extremely vulnerable people with SCD are and how easily a crisis can be triggered by simple effects of cold.

i. Signs, symptoms and experience of a crisis

Symptoms of sickle cell disease can be mild, but are said to constitute a crisis when the pain or symptoms can no longer be managed at home through increasing heating and taking over the counter pain killers and need admission to hospital, in particular for pain management. The main triggers for crisis reported by the participants were getting cold, dehydration, stress, infection and not eating well. Other triggers were strenuous activity and fizzy drinks. The main signs of a crisis starting were pain in the joints or other areas of the body, tingly lips, yellowing of the eyes and feeling tired.

I know it gets serious when my lips start to tingle. If my lips don't tingle then I know I don't, I'll be all right kind of thing. But my wrists will go, my wrists and my knees are the main things, when I can feel it about to start, they'll just start to sickle so I know I need to go, get some paracetamol, turn the heating up, put my dressing gown on, put my thermal socks on, cover my knees, cover my wrists. So I'll put leg warmers on my wrists, stuff like that, just to try and keep warm. And then jump in bed and just wait for the paracetamol to kick in. And most of the time it works. Sometimes everywhere starts to hurt, so I need to go to hospital or stuff. P1

Other signs reported were hardening of the spleen (a life-threatening emergency) and being unable to walk or speak. This was problematic for trying to prevent the crisis escalating by turning the heating on. Because of the pain and inability to walk one participant feared falling down the stairs whilst going to turn the heating on when she woke up cold and in pain at night and at times had to crawl on her hands and knees to get to the heating switch:

because I physically don't have the energy to get up, because like my oxygen levels get really low and I struggle to breathe. I struggle to talk sometimes when my oxygen levels drop. And yeah, like when, I've had to crawl around before to like get my tablets and stuff, because like it affects my shins sometimes. And they just keep, if I stand, my legs just give way, so I have to keel, have to crawl. It can get really, really bad. P1

Participants sometimes found it hard to describe the level of pain experienced in a crisis because of its severity:

my back and my stomach was in agony. And I was trying to put off going to hospital on that Sunday, but my mum could just hear me like screaming and just like in pain. She came and massaged my back for me, my back and my stomach, but it didn't really have an effect. So she was like right, you're going hospital, so I went to hospital. P1

Parents were also very aware of the pain their children suffered in a crisis:

You can say the ambulance, sometimes it's just delayed before they come, or she will be screaming and screaming, it's really painful. P3

Signs and symptoms were reported to be difficult to discern in very young children because of their inability to explain what they felt. Preventing hospitalisation for a crisis through avoiding the triggers was a high priority among carers of and people with SCD, as it also was once symptoms had started.

Many of the participants were in hospital at least two or three times a year for stays of approximately 3 days to a month. Staying in hospital was not easy, some children were afraid of needles and some people with SCD reported receiving poor care due to a lack of understanding of the immediate, high level of pain relief required.

ii. Cold as a trigger for crisis

Hospitals were also the source of some infections which could trigger another crisis. In addition to the premature loss of life to SCD in their own families or communities some people experienced life and death situations themselves. One participant had been told he was about to die and he needed to call his parents in to say their goodbyes. He survived but suffered kidney failure as a result of the crisis. One of the mothers participating had lost her 11 year old daughter to SCD after fleeing a war in an African country. Whilst sheltered by a family in a neighbouring country her daughter had been contributing to the host household by doing the washing up in cold water. This triggered a crisis, exacerbated by stress and dehydration and the mother was unable to pay the hospital fees in time for life saving treatment. This mother was now looking after her surviving daughter, also suffering from SCD, in the UK. She is now extra vigilant and fearful of triggers:

even when she says I'm feeling my head, I'm not going anywhere. I know, I've experienced it; I saw my daughter die and you can see a big woman like me, I was just.. it's shock, I can't believe. It's so quick! It's so quick! P3

Cold was identified as a major trigger for sickle cell crisis leading directly to hospitalisation. Participants identified 12 different instances when they believed cold temperatures were the trigger for their crisis. Therefore being able to heat the home adequately was of high importance:

It's [heating] very, very, very, [important] because any time, if [son] says I'm cold I start panicking, you know, because he's not supposed to be cold at any time. P7

The examples below are useful to illustrate the different ways people with SCD get cold. Also being admitted to hospital with a crisis can be triggered by cold that would be more easily tolerated by people without SCD:

Fig 1. Examples of where cold triggered a sickle crisis and hospitalisation

1. Being unprepared for very cold weather when the radiators were turned off at night. (crisis and hospitalised)
2. Went swimming and didn't get dry quick enough afterwards (crisis and hospitalised for 4 weeks)
3. Bedroom temperature dropped to lowest point overnight (crisis and hospitalised for 2 days)
4. Was given a job selling door to door in cold weather (crisis and hospitalised for 4 weeks for crisis and a chest infection)
5. Child ate an ice-cream (crisis and hospitalised)
6. Child washed up dishes in cold water (child died)
7. Went out in snow (crisis, pneumonia and hospitalised)
8. Flat got flooded so participant was moved into temporary accommodation in a hotel, hotel was cold (crisis and hospitalised)
9. Came back from holiday in a hotter country, brother brought him coat to stay warm but wasn't enough (crisis and hospitalised)
10. Got caught in downpour and couldn't get dry whilst travelling home (crisis and hospitalised)
11. Studying late at night and felt cold draught on back (crisis and hospitalised)
12. Got cold which lead to a 'bony crisis', felt more pain than a normal crisis (crisis and hospitalised)

iii. Case study of a hospitalisation attributed to cold during the study

Over the two week period room temperatures were recorded one participant said she had woken up feeling cold and with pain in the early hours of the morning. This was during a cold snap when it had been snowing some days earlier. She was a young adult living with her parents in a well insulated, new build house. She had been getting up at night to put the heating on and take painkillers when she felt pain quite frequently and this would allow the pain to go away. On this occasion the pain had not gone by the morning and her mother tried to massage her to no effect so they decided to go to hospital where she was admitted:

Well I woke up with the pain at ten to five [am]. That's when I woke up on the Sunday with the pain. And that's when I went downstairs to, did I turn the heating on, I turned the heating on, I think, got my tablets, got my water, went back to bed, so all that happened between then and nine o'clock. P1

Below are the temperature readings for her bedroom. Her bedroom temperature was 19.1 °C when she woke with pain. The average room temperature over the 2 week period was 20.5°C so pain was felt at below this temperature. The room temperature continued to drop to 18.7°C during the period she said her pain had not gone away. Whilst these temperatures may be tolerable for people without SCD this may be cold enough to bring on symptoms in people with SCD. On return from hospital her mother bought her a 13 tog duvet to replace her 10.5 tog duvet. She stopped waking up as much with cold in the night with the extra insulation.

Table 2 Bedroom temperature readings for participant 1 when crisis was triggered

Time and date	Room temperature (bedroom)	Reported effect of temperature
31 Jan 2015 21:00	20.9 °C	None
31 Jan 2015 22:00	21.5 °C	
31 Jan 2015 23:00	21.8 °C	
1 Feb 2015 00:00	21.5 °C	
1 Feb 2015 01:00	21.0 °C	
1 Feb 2015 02:00	20.5 °C	
1 Feb 2015 03:00	19.9 °C	
1 Feb 2015 04:00	19.4 °C	
1 Feb 2015 05:00	19.1 °C	Woke up feeling cold, turned heating on, took painkillers and drink and went back to bed
1 Feb 2015 06:00	18.8 °C	Couldn't get rid of pain
1 Feb 2015 07:00	18.7 °C	
1 Feb 2015 08:00	20.3 °C	
1 Feb 2015 09:00	21.0 °C	Admitted to hospital for 2 days with crisis
1 Feb 2015 10:00	21.7 °C	

iv. Effect of cold on SCD carriers

There is a misconception that SCD carriers (trait) are merely carrying a silent gene. Medical advice however does state that carriers may sometimes feel milder symptoms, or experience a crisis if under extreme physiological stress¹⁸. Being a hereditary disease people with SCD would almost invariably have family members and carers who had the SCD trait. Family members with SCD trait reported feeling mild pain in their joints when they felt cold. Two participants with the trait reported being in intensive care as a result of a crisis causing complications combined with other medical problems. The health of carriers is not as fragile as those with the full blown disease but there are clearly some health risks involved to a much larger population of 240,000 with SCD trait in the UK¹⁹.

¹⁸ Key N, Connes P, Derebail (2015) Negative health implications of sickle cell trait in high income countries: from the football field to the laboratory *British Journal of Haematology* (published online)

¹⁹ NHS Sickle Cell and Thalassaemia Screening Programme (2015) *Facts and Figures/Statistics* <http://sct.screening.nhs.uk/statistics>

2. Heating behaviours of people with SCD

People with SCD use multiple strategies simultaneously to keep warm at all times. They exercise a 'warm chain' practice where they try to minimise getting cold at all times. For example a teenage girl with the condition would walk home from school with several layers on and her mum would pre-warm the house ready for her arrival. When she arrived she still felt a bit cold so would go to bed under a thick duvet for 15 minutes to re-warm herself and then get up and help her mum with housework. Because pain can be felt almost instantly on exposure to cold this may aid the understanding of an effective cause and effect association that allows people to learn how to stay warm. In addition health professionals recommend they stay warm, and children with SCD will often be taught throughout their childhood that they must wear extra layers or observe the consequences. Whilst not all of these examples are of heating use, they show just how concerned and sensitive people with SCD are to effects of cold.

i. Preferred temperatures

All the participants reported they felt they needed warmer room temperatures than family members without the condition. Out of 7 participants who specified a temperature the temperatures were 20°C; 20-30°C; 25°C, 25°C, 25C, 30°C and 30°C. This is above the Department of Health's recommended temperature of 18°C for health²⁰. How warm their homes were will be looked into in more detail with the temperature and humidity readings.

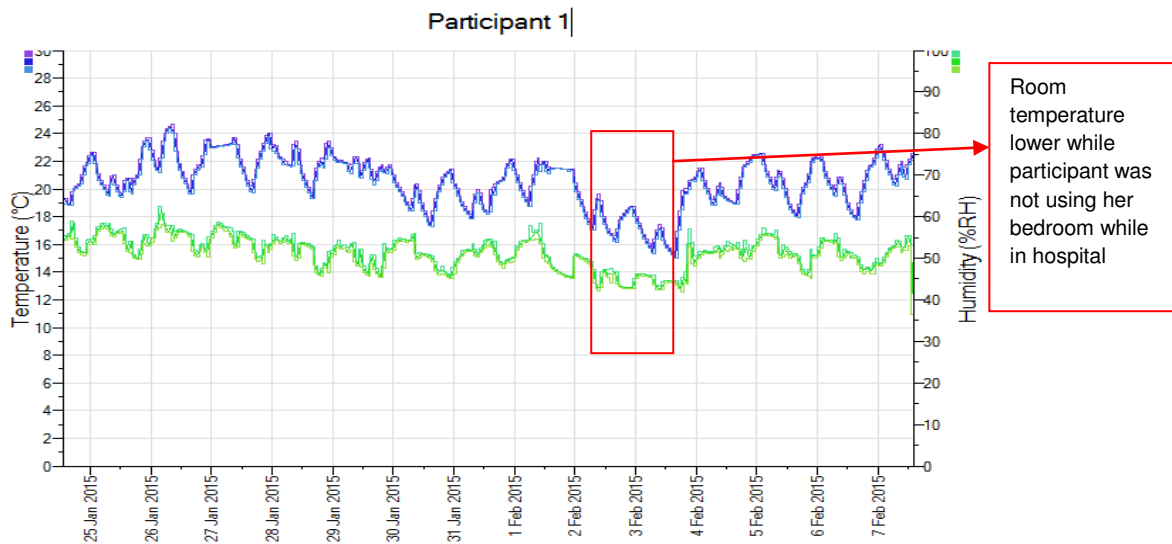
ii. Room temperature readings from Tiny Tag Temperature measuring devices

There were 6 participants who were able to have their room temperatures recorded over a maximum of 2 weeks.

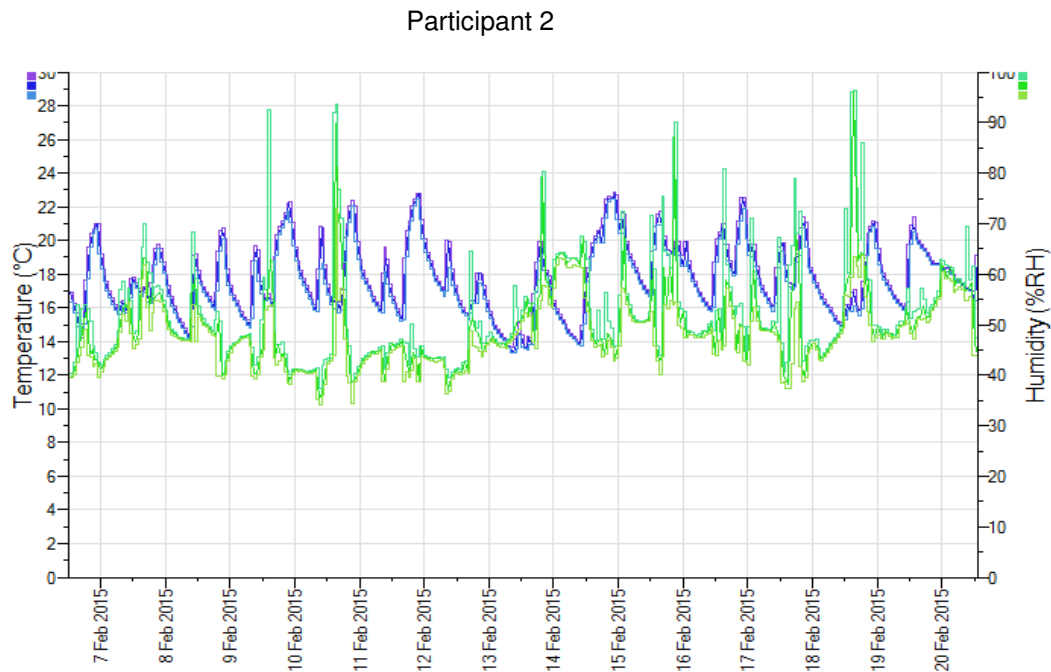
Participant number	Maximum Temperature (°C)	Minimum temperature(°C)	Average temperature (°C)	Room measured
P1	24.7	15.0	20.5	Bedroom
P2	22.8	13.4	17.7	Various
P8	29.1	17.1	20.9	Living room
P9	20	12.7	15	Living room
P10	25.4	12.6	14.4	Living room
P12	25.3	18.7	21.2	Living room

The charts below illustrate the temperature (in blue) and relative humidity (in green) recorded over a maximum of 2 weeks.

²⁰ R. Wookey, A. Bone, C. Carmichael, A. Crossley (2014) 'Minimum home temperature thresholds for health in winter – A systematic Review' Public Health England https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/365755/Min_temp_threshold_for_homes_in_winter.pdf Accessed 18.12.14

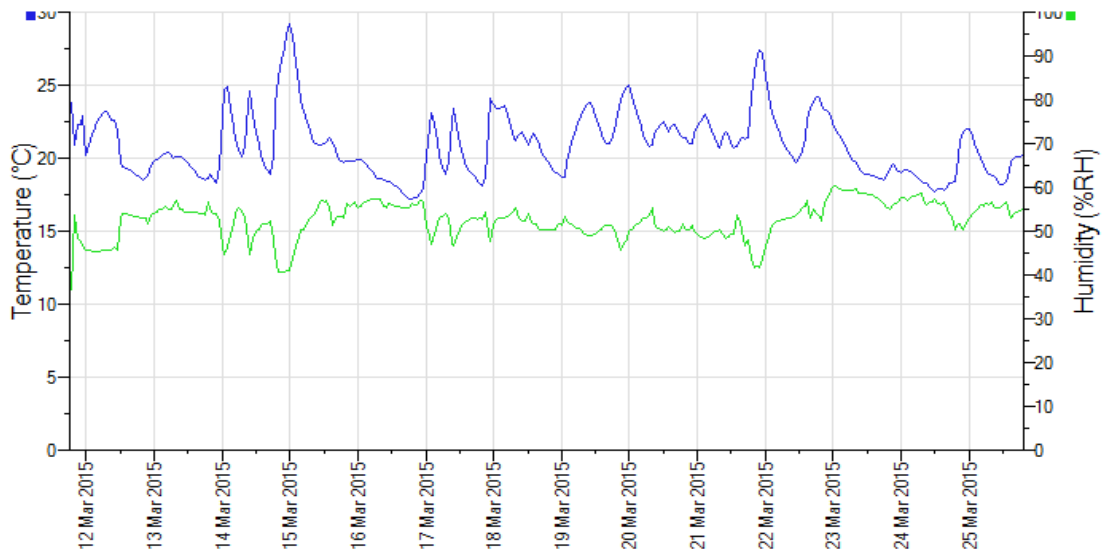


Participant 1 was a young adult with SCD living with her parents in a new build, well insulated property owned by her parents. The heating was turned off at night unless she felt pain during the night and got up to turn the heating on. A crisis was experienced (see above) in the early hours of the 1st February. While she was in hospital (1-3rd February 2015) a noticeable drop in room temperature can be seen.



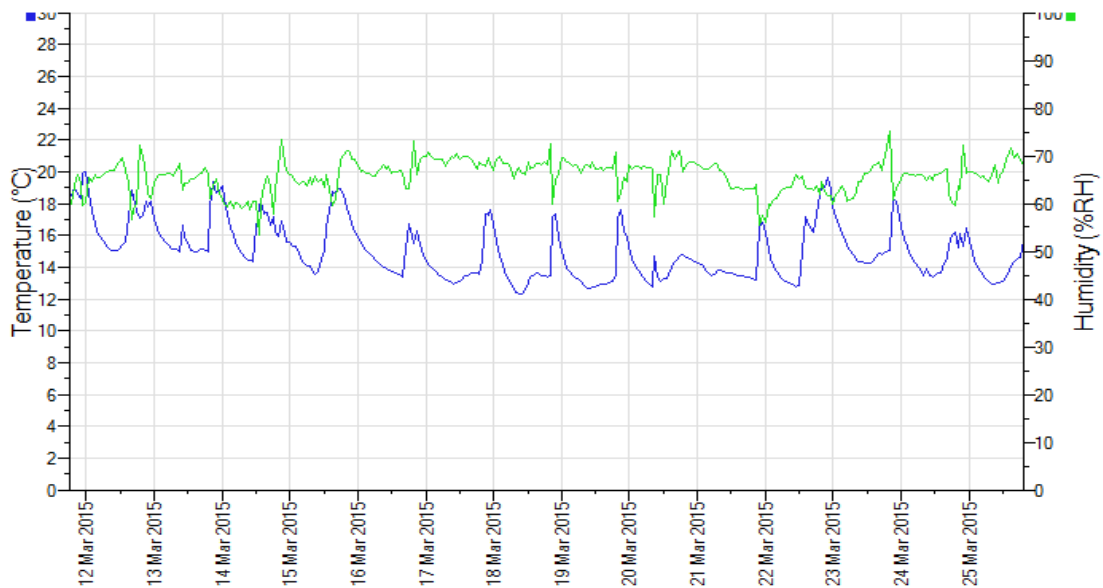
Participant 2 was the mother of an 8 year old boy with SCD living in a mid-terrace post war council house. The participant moved the Tiny Tag monitor around the house during the measurement period. During the second week the children were at home for half term holidays but were not in the house all the time. Heating was used mostly in the evenings when they were home.

Participant 8



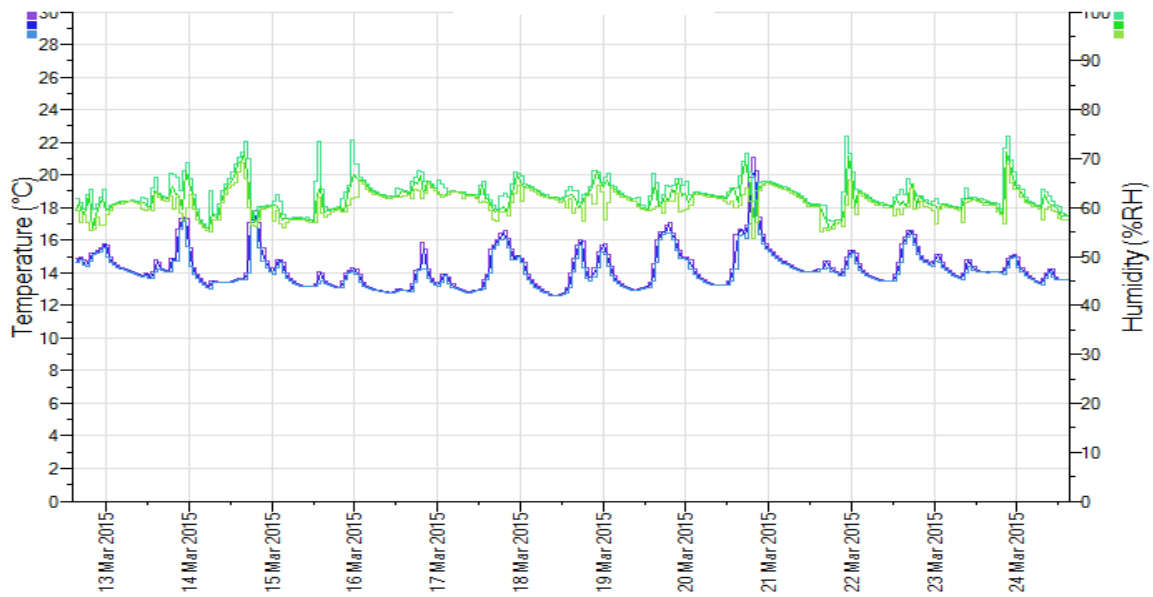
Participant 8 was the mother of a 16 year old girl with SCD. The girl lived across the road in her grandmother's house because it was warmer there. The house is a post-war council house. The measurements were taken in the grandmother's house.

Participant 9



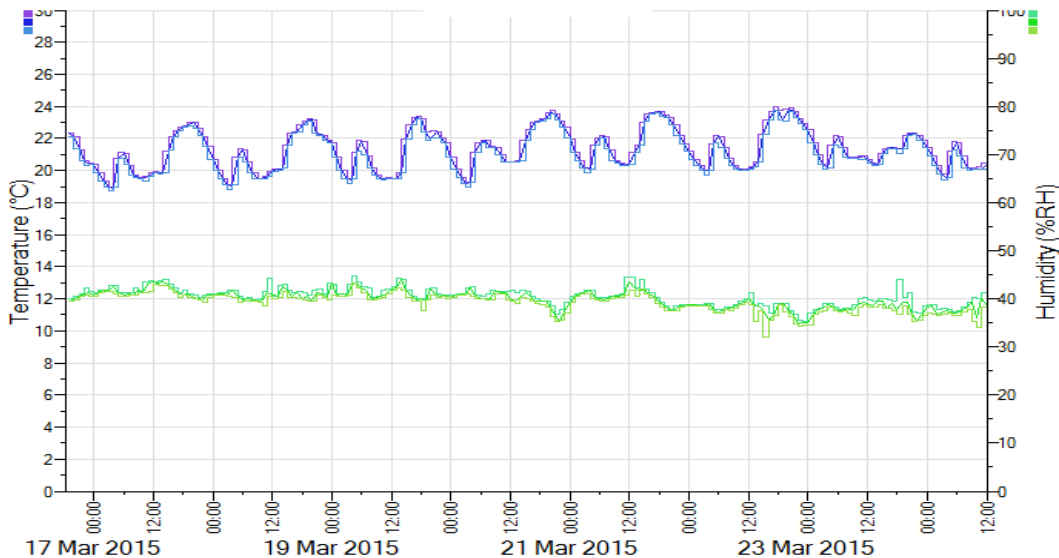
Participant 9 was a mother of a 13 year old girl with SCD. The house was a semi-detached post-war council house. The heating was only turned on when the girl was back from school. It looks possible that the heating was not put on in the mornings when the children were getting ready for school.

Participant 10



Participant 10 was a young adult with SCD living on his own in a ground floor flat. He had received no benefits during the measurement period because he was suddenly taken off Employment and Income Support after failing a work capacity assessment. With £30 debt on the gas meter he did not see the point of accepting £30 from a family member to top up his meter card because that would just be taken up by the debt. So he was not using his heating while he had no income. He spent most of the day at home.

Participant 12



Participant 12 was a retired lady with SCD living in a mid-terrace new build. She spent most of the day at home.

It can be seen that the average temperatures were not quite as high as some of the participants said they felt comfortable at. However participants may not have had the heating on all the time if they were not in the house. The two lowest averages were from participant 9 who only had the heating on when her daughter was back from school and from participant 10 who had no money to heat the house.

iii. Heating systems and 'spatial shrink'

Most people had gas central heating apart from one with electric storage heating. The storage heating was reported to be very inadequate as the heat ran out by mid-afternoon. Almost all participants relied frequently on additional portable electric radiators to top up their heating when it was too cold. 'Spatial shrink' was practised, i.e. where only parts of the home are used and heated. This featured one family who would retreat to the warmer bedrooms to eat and one father, who would only heat the living room with the electric heater while his children were at school, and turn the central heating on when they were back. Heating would also be restricted to certain rooms to cut costs:

Well actually she likes to spend more time in the living room watching telly. But I just encourage her to go inside, and we normally take dinner inside our rooms now. P3

This was mainly to cut heating costs but seen as problematic as it meant going from a warm room to a cold corridor, kitchen or bathroom brought on pain.

Some participants used less heating than they would have liked to save costs:

Oh yeah, in an ideal world I would love to have the heating on. But because it's on at a level that we can manage, whereas if we had it on, we've tried heating the house constantly all day and all night, but when we got the bill it was a shocker. So we thought we won't get ourselves into that kind of debt again, so we have to just try and manage it on timed without it being on constantly. P6

iv. People with SCD not in control of the heating in their home

Living with different ages, genders and health conditions could cause conflict in setting heating at a comfortable temperature. Those without the condition thought those with SCD had a room 'like a furnace':

Yeah, like my mum will walk in my bedroom and be like, and she'll just be like oh, I can't breathe kind of thing. But I'm comfortable in there. I do like it really hot. So she'll come in, turn it down, and it's just like a fight between us, so I'll keep turning it back up, she'll turn it back down. But yeah, she keeps saying like it's not healthy, like walking out from the passage where it's nice and cool, and then going into my bedroom where it's like a furnace kind of thing. So yeah there's that. Yeah. I like it hot, they don't. P1 (SCD)

Those in a dependant position, i.e. children or a homeless young lady taken in by a couple, didn't always feel able to ask for the heating to be turned up because they were conscious of other people's costs. However most did not have their heating on during the night and used bedding and hot water bottles to keep warm as overnight heating was felt to lead to dehydration (another trigger), cause headaches and nose bleeds in an unaffected sibling.

3. Housing, energy efficiency and other ways of keeping warm

1. Energy efficiency

Participants were aware that energy efficiency could reduce their fuel bills but some could not improve their situation. With limited time and energy they still found it hard to get through to schemes that could help:

Then by the time you reach there they say oh next time ...we've got enough people that have already applied, you can try again. Next year in December. You try in December, phone them in December, they say oh the deadline has been, everything, the deadline is in January. When you start to phone it's busy, busy, busy. When you phone in January it's all closing, you know what I mean, so you get fed up. Especially when she's [daughter] been ill, I have no time, time to speak on the phone, time to cook, time to clean, time to look after the child. It's all hard, it's very hard. P9

One participant had been asking her landlord to install double glazing for 10 years and had given up and decided to move house. Another lived in a high rise flat, meaning the whole tower block would need insulating and not just her property. This was the mother of the teenage girl with SCD who had already lost one daughter to a crisis in Africa when she could not pay the hospital bill in time. The property had no wall insulation and only electric storage heater, supplemented by portable electric radiators, which led to very high heating costs:

Because the lady said if they insulate it for you, you can spend £10 a week instead of spending how much. Whenever I say £50 people scream what, £50, are you sure of what you say? I said yes, and we're not wasting it. It's just too expensive. P3

ii. Moving away from family for a warmer house

Some participants also chose where to live according to warmth. One homeowner bought a mid-terrace house with the hope she would benefit from neighbours' warmth:

I chose this house because it was being built when I came around and I chose this one because it was the middle of two, because I thought it might be a bit warmer being in the middle. And I think it's worked out that way. P12

Another teenage girl moved in with her grandmother who was in her eighties and also needed a house that was warm at all times, and another participant moved hundreds of miles further south in the hope it would be warmer in that city. Moving away from family to be warmer isn't unheard of among people with sickle cell outside of this project. This is similar to the case of Imani Green, an 8-year-old girl with SCD from Tooting, London who was tragically caught in crossfire in Jamaica in January 2013. She was sent to stay with family there with the hope the warmer climate would ease her condition during the British winter. Her Head teacher talked about her condition: *"She dealt with her illness very bravely and coped well with the special arrangements we had to have in place to support her. She had been given special permission to travel to Jamaica so that she could benefit from the warmer*

climate and we had been in contact with the local primary school she was attending.'²¹ Choosing to move away from family to stay warm shows how much staying warm was prioritised.

iii. Clothing and bedding

Electric blankets may seem an obvious solution for overnight heating but some participants wouldn't use them for fear of fire risks:

I mean I used to hear the horror stories of people burning themselves to death with an electric blanket, that's the kind of vision I've got. But electric blankets are supposed to be fairly good now. When I used to go to one of these sickle cell centres to have my bloods done, they'd have an electric pad where they'll give you that and you can hold onto it and that will keep you warm during your transfusion. So that was a good idea, electric heat pads. P6

High tog (approx 13 tog) duvets were used for cold nights in bed and blankets were frequently used in living rooms in the day. One participant regularly used a high tog duvet on the sofa. How many layers of clothing were needed to prevent pain seemed to be learnt throughout childhood but young people felt under particular pressure to dress like their peers, wearing little clothing in cold weather. A male participant felt that peer pressure to wear few layers was harder for women than men. One young lady noticed a pattern of going out with her friends at the weekend without a coat and being in hospital with a crisis a couple of days later. She then started wearing a coat when she went for a night out, paying to keep it in a cloakroom and was supported by her family to do this:

My nan and granddad know that I need to keep warm, like the other night when I went out that night with my coat on, my cousin was like I hope you're dressing up warm and stuff, so I sent her a picture of what I was wearing and she was like she approved. So, yeah, like my extended family are good as well, really really good. P1

Hats were seen as essential items indoors and outdoors. Dressing gowns or housecoats were worn indoors and thermals were also used indoors and outdoors. The importance of good quality, warm clothing was recognised with one mother spending £75-£120 on outdoor coats for each of her children with SCD and 'getting away' with only spending £25 on coats for her other children.

iv. Avoiding cold environments

Another example of how strongly participants felt about keeping warm at all times was in restricting when they went out and whom they visited. Some stopped visiting family and friends in their houses because they did not think they would be warm enough there. In some cases their family and friends would turn the heating up, understanding how important heat was to them, and have blankets waiting for them. Avoiding going out, or stopping children with SCD going out in the rain or cold weather to play or for errands was a common practice. Cars and taxis were the preferred form of transport because warmth could be ensured, despite the extra cost of using taxis in cold weather. These avoidance behaviours contribute

²¹ BBC (2013) *Imani Green Jamaica killing: 'Happy girl', eight, shot dead* <http://www.bbc.co.uk/news/uk-england-21002359>

to social isolation as people with SCD do not feel free to go where they want when they chose to:

Bad. That's why I don't go anywhere; I don't go to people's houses because I need my heat. And you can't go in somebody's house and say can you turn on your heater? You can't do that. I just stay at home. P5

vi. Other methods of keeping warm

Consistent with the high importance participants placed on keeping warm, other methods were used in addition to the above. These included warm drinks, heat pads, hot water bottles and one family with younger children huddled together in the same bed to stay warm when they had no money for the gas meter.

4. Why people with SCD are at particularly high risk of experiencing fuel poverty

Evidence above shows that people with SCD are at very serious risk to harm from cold and invest a great deal of time and energy into staying warm at all times using multiple methods. However this research has revealed evidence that they may also be a group at extremely high risk of fuel poverty through multiple and complex factors affecting their access to education, employment and welfare support. For three of the participants the voucher for the research was the only source of money they had for the week they were interviewed.

i. Education

Frequent bouts of illness requiring hospitalisation and convalescence throughout childhood can interrupt education, resulting in reduced earning capacity as adults. Carers can find it hard to progress in adult education whilst taking on the responsibilities of caring for a sick child and investing time and energy in preventing future illness when their child is healthy. Entry to early years education was delayed with one child whilst the nursery awaited training from a specialist SCD nurse. School staff need to know how to spot an emergency:

I can't tell it all but there are so many things that you have to look for. Like for him for instance his spleen, you feel that area and it feels hard you have to rush with him because it can be fatal. And like him going to nursery and stuff, the sickle cell nurse has to go to the nursery or school and explain the illness. So anything staff know how to react. P7

Attending university, particularly away from home, is a financial challenge considering extra costs associated with the condition and financial challenges all students face. However, disability support was found to be available through university services. One participant gave up university after getting into arrears with his rent. War, language, several changes in country of residence, poverty and gender relations had an impact on educational opportunities for participants coming from some African and Caribbean countries.

One participant was studying and reported having no income whatsoever and was registered as homeless. She had been taken in by a couple she knew whilst her family were in a different city. They allowed her to contribute to the house whenever she could but she felt it was an imposition to ask them to turn their heating up. She had been homeless for 6 months and was on a list for re-housing.

ii. Employment

Some people with SCD found maintaining a job a challenge due to chronic pain, fatigue and frequent illness. Maintaining a job with SCD or caring for someone with SCD can depend on how understanding and flexible the employer is. One lady on a work placement year at University gave up on one employer because they didn't like her being off sick frequently and found a new job where her sick leave was supported:

So yeah, they have been really good, been really, really good. And work's been good as well. Like because before, when I first started placement I was at [employment 1] and I didn't like it, I didn't feel like.. I was in hospital two or three times, I think, and I just felt like they were like oh for god's sake, not again, kind of thing. I didn't think they understood. So, and I just didn't generally like it there anyway, so then I moved to the [employment 2]. I started there in November, and I've been in hospital twice because of my sickle cell since then, and they've been really really understanding. So it's just, I feel a lot better working there, like with my sickle cell, because they're just really nice about it. So I think that helps, definitely. I still feel bad when I don't go in, like when I'm in hospital. P1

Maintaining a job also depended on the kind of employment rights attached to the work contracts. Working on zero hours contracts or agency work meant no entitlement to sick pay or carers leave so when they were unable to work their income effectively ceased:

With my own situation, if my daughter is sick or I am sick, if I am sick, like I'm not like others, they are paid if they are sick, but for me nothing like that. So sometimes I just try if I see extra hours to just work and make up for the last days. Sometimes even when you're not sick but you just get tired, you want to stay, you know, on your own. P3

People with SCD and their carers can get caught in a cycle of low paid jobs, with probationary periods making it easy for employers to legally sack people with SCD or their carers for being off sick or caring for a sick child.

Because I have my nephew here as well, he's sickle cell, he's SS like [daughter], and he's 32. He's a very hard worker but sometimes you can see the agony. But he never claims. He always, he got disability allowance but always worked. But his work, but he missed so much job, every time, god knows how many agencies he has been. Because he'll be working, some agency he'll be working for three or four months, and then like he is off for a week, even though he explained that because I've been ill or because I've got this illness. But people don't understand, they keep sacking him. P9

One participant (a mother of a child with SCD) was in ICU after an obstetric emergency resulted in a crisis, even though she was a carrier and didn't have full blown SCD. Her husband had to take time off to look after her and their daughter with SCD. On returning to his work after 3 days he was sacked:

Off for maybe three or four days, because he was looking after me and after the kids. He went to work, honest to god, they sacked him. I told him even to, I said oh just go and sue them. He said, he turned round and said to me how many companies I should sue? Because this has always been our lives. Only because I'm lucky, I always work with one company but he never be lucky. Stick with one. But yeah they sack him. They didn't give him an explanation, they said to him you're not fit to work in this warehouse. I knew why,

because he's been off for three days. Even though he told him that my wife was in hospital, and then other time he had to stop with [daughter], he in the house because she wasn't well. P9

One participant solved working whilst being a carer by working night shifts while her husband was back from work and on her days off she worked on a second job during school hours. Another participant voluntarily gave up work because she was having too much time off work due to the condition and found work too stressful and tiring.

Parents with a child with SCD also found childcare a challenge because they found it unaffordable and did not always fit in with shift work. Not all participants had family nearby to help with childcare so some relied on help from friends or neighbours. Informal childcare arrangements may put children at risk. To be able to get to work before the nursery opened one mother was leaving her child with SCD with a neighbour who walked him up to nursery for her. When a new partner came to live with her neighbour she became wary of him and found the man had taken the child on his bike and the child had fallen off and cut himself. She stopped leaving her son with the neighbour the next day and came home to find the police arresting the man. In addition to finding normal childcare parents reported the difficulty of finding carers that understood how to prevent a crisis being triggered and spot the signs of an emergency if the child became ill.

iii. Welfare support

Participants claiming welfare benefits got stuck in a cycle of not qualifying for disability benefits but getting jobseekers allowance (JSA) payments sanctioned because they were unable to sign on if they were in hospital or caring for a child in hospital.

Disability benefits

Being a relatively rare condition of a fluctuating nature puts people with SCD at risk of failing to qualify for Disability Living Allowance (DLA), Personal Independence Payments (PIP) and Employment and Income Support Allowance (EIS). Participants had trouble qualifying for disability living allowance because the condition was fluctuating and they could appear well:

You can look at me and think oh he's an able bodied young man. But I just struggle walking here, hold onto my daughter, and then I've got other complications. I get short of breath because I've had lung problems... So it really is what you look on the surface, don't judge a book by its cover, you know, because what's going on underneath the bonnet, it's not really working too well P6

Participants had been asked what conditions they had 'on top' of SCD, and sometimes having another condition, caused by SCD, such as kidney failure, was a better understood kind of disability:

I don't think that enough people do know about sickle cell or what's involved with sickle cell, whereas you say renal failure or you say kidney failure, well everybody knows what the kidneys are and it's just because they don't know why it's failing or what causes it to fail, but just the word kidney failure, obviously it's not working, so obviously they'll probably grasp that a bit more than just saying, you've got sickle cell, but yeah. P13

One young man refused disability living allowance (DLA) as a teenager because he wanted to be normal like his peers. Unable to cope with employment, when he applied again for DLA he was refused. The reason given was that he had refused it before, and so therefore should not need it.

And then when he was 16 he said, we sat down and we spoke about it, he said I feel fit me, I want to go and get a job. So we decided not to re-apply for the carer's allowance, and we would try. Six months after that he had heart failure. He had had a couple of jobs up until then, but couldn't, it was about a year he got heart failure, then he had to be booked in to have open heart surgery for the second time. And when I went to re-apply for disability living allowance it had all changed and oh no, you said you didn't want it before so you're not having it now, you don't need it now. Did an appeal, that failed. P8

More positive experiences with claiming disability benefits were among participants with kidney failure and another one who was accepted as housebound. One participant was accepted for DLA because of his SCD and found the advisor who had done a course on SCD, very understanding.

No, they don't really know about sickle cell. But luckily the person that I had at the time had a course on sickle cell, so that's why they put me on employment support allowance for three years. So after that now as I said I'll have to be reassessed later on, sometime this year I think, to determine whether I can stay on the support allowance or I have to go on jobseekers.. Whereas some others have said that they've had one and they said so you've got sickle cell, what's that? Then he's explained what sickle cell, and they're saying well that doesn't determine that you can't work. Whereas I think it does, because sickle cell is a derivative and disabling disease. So for them to say what else have you got on top of sickle cell is ridiculous. P6

Jobseekers Allowance

Because of the fluctuating nature of the condition, being able to attend all JSA appointments was not always possible:

I wouldn't be able to make them regularly, because as I said there's days when I can hardly get out of bed, and there's days where the sickle cell takes over and I am literally left at home in a state where I can't even come out the house for a couple of days I'm in that much pain. So I can't really say oh I can make this appointment that day, whereas that's what they expect you to do, turn up on your appointment days... I can get from being happy and jolly one day to death's door the next day, so it really is up and down P6

For those on JSA the risk of being sanctioned for failure to sign on was high. Reasons for non-attendance given were being in hospital at the time or caring for a child in hospital. In all cases participants rang their Jobcentre to inform them, even in advance in the case of someone having an operation. Their benefits were stopped immediately when they did not attend their appointment and they were asked to provide medical evidence. One lady who was being sanctioned at the time of the interview was told by the hospital she would need to wait a couple of weeks to receive the letter, so in the meantime she stayed sanctioned. Another participant was ill at the time of his appointment and had to produce a note from his GP. He had to wait days to book an appointment and said that, in the past, he had sent doctors notes in and Jobcentre staff had lost them. In most cases the benefits were

reinstated with appeal and/or receiving medical evidence. However this meant they were without income for two or three weeks at a time and relying on family members to buy food for them and pay for their heating. When JSA was stopped so was housing benefit and this was not backdated so some of the participants were falling into arrears because of frequent periods of sanctioning. Of the 15 participants, 9 were claiming benefits. 3 of these had reported being sanctioned in the month previous to the interview, one had just come out of hospital and had been sanctioned for 2 weeks previous to the interview. Another participant was taken off Employment and Income Support Allowance after failing his disability assessment and had had no income for the previous 3 weeks because he was waiting for his JSA to come through and was appealing the disability assessor's decision. This participant had a signing on appointment on the day of the interview but had no money for the bus. When I asked how he was going to get to the appointment he said he planned to walk and take regular rests as he went despite his hip replacement and breathlessness caused by the SCD and a heart condition

Other factors affecting cost of living and income

Other reasons for people with SCD and their families being at more risk of fuel poverty were the costs associated with the condition such as higher costs of heating, taxis to get around, higher costs of more nutritious foods, higher quality clothing and bedding and supporting other family members in times of sanctions or low income. Participants reported finding it hard to save money on heating because they were either ill with the condition or too busy caring for someone or keeping up with paperwork for benefits and appeals to make frequent phone calls. One participant could not get through to a helpline for an energy efficiency discount and then was told that the scheme wasn't running. Switching energy companies was low down on their list of priorities although they recognised the value of it. One participant did not have internet and was referred to a council helpline for switching. The last time participants had switched energy companies was between 6 months and 15 years. The average time since the last switch was 6 years.

Carers

Carers were taking on considerable burdens for their families and worked hard to keep their relative with SCD healthy:

Which is why you have to be very protective and you have to, if you value your child's life you will do it anyway. But you work left right and centre, because you haven't got time to listen and be upset by ignorant comments. But remember these ignorant comments are coming from people who have the ability and your life in their hands, if not nurses then people who say whether or not you deserve £10 extra a week to be able to survive. So these people have got your life in their - so if he was on his own I don't think [son] would have survived. P8

Aside from working to support their own families and adult children who had left home and elderly relatives, they had other care roles on top of that. These included fostering, caring for their other children and advocating for better awareness and care for people with SCD on a local and national level. Some also worked as carers in home care, hospitals and day centres. The support provided by carers for someone with SCD is vital and very much based on preventative care. They needed to speak for the person with SCD when they were in a

crisis, ensure they received optimum care in hospital, aid recuperation at home, shop and cook, especially when the person they were caring for couldn't move or walk and advocate for them in benefits appeals. Care during well periods was just as vital to ensure their family member was warm, well fed and drinking plenty at all times and action taken promptly when there were signs of a crisis.

5. How problems with heating impact on other triggers of stress, dehydration, infection and not eating well

The top priorities for preventing a crisis were keeping warm, eating well, drinking plenty and avoiding stress. So how does heating impact on these? High heating costs make it harder to buy good quality food:

Someone like my daughter and myself and my son, because we're all trait and she's carrier. We have to have proper juice and proper meals; we can't keep buying like smart price food and everything. There is nothing wrong with it, but because you know that you've got medical condition you're scared. P9

People with SCD and their families can already experience an additional level of stress due to the experience of having the condition. They may experience a lack of understanding of the condition in employment, schools and with welfare agencies. Although many good examples of health care were mentioned there were also examples of poor care, particularly with children's services; the lack of understanding of the condition and inadequate pain relief, which can contribute to depression. This mother talked of inadequate pain relief for her son's hip replacement needed because of the effects of SCD:

He had crisis and had to endure the pain in a side ward with the door closed getting a couple of paracetamol and they said what do you mean he needs morphine? He's supposed to have a two year transition then in preparation for the operation, so he's been through some stuff that's put him in a dark place, very dark place... they didn't give him anything for hours, and so he felt the full impact of a crisis, and it ain't to be felt... He was in the actual hospital and they didn't know how to care for him. That's with two years preparation for the operation, so he really has lost all trust. P8

They may not be allowed to self-care, such as students being stopped from going to get water during class. Children may find injections stressful. They may also experience death of close family members or near death experiences themselves. They may also be subject to racist comments as the condition is sometimes incorrectly believed to exist only in people of African origin.

On top of this stress was direct stress regarding maintaining an adequate income to be able to afford fuel bills:

The heating can cause stress actually if you're worried about where the money's going to come from to paying each bill. Because it's like you don't switch the heating on and stuff, that can obviously cause a lot of stress, which then can participate in bringing on a crisis. P11

This ability was put in danger or made impossible with the difficulties experienced with sudden changes in income due to sanctions and sudden loss of DLA at a new assessment, adding to the other stresses:

But those people that they have on as frontline staff that make the decision between life and death are responsible for a lot of people hitting depression, hitting mental health, because they're so weak already, they're at the bottom of the pile for crying out loud, they don't have anything, and they're so weak already that they haven't got the strength to fight you. P8

Those with family nearby had more support in terms of being able to get out of their own house and go to where they knew it was understood that they needed warmth. Families acted as financial buffers, lending money or buying food and heating when the person with SCD had no money. Support was often offered by family members on low incomes themselves.

Maintaining a warm house, and one warmer than needed by the general population, was a strain and a worry for many participants:

Yeah, I think one of the biggest outgoings is paying your fuel bills really and a most necessary one that you have to keep up, because you can't live in the cold, simple as that, so you have to pay it. P12

Those on meters were at risk of 'self-disconnection', that is to say that energy companies have not cut them off, and they wouldn't be allowed to, but the lack of money meant no money in the meter cut their supply off. This supply could not be restored until they had cash again, which could be days later and debts on the meter may need to be covered before getting any heating:

because like I think it's been about three weeks now since I've had income, so with my gas, there was, what they do with my gas, I've got no debt on it, well I have now, but I had no debt on it, and because I haven't used it, there's like £30 worth of debt now. So when I do start getting something through, even if I was to pay the £30 to get rid of the debt, I'd have to find another £30 just so I could have some heating. So it just sets you back a lot. P10

Waiting until there was money to put on the meter again was not seen by some as an option as they believed cold could rapidly bring on pain or a crisis. One example is when a mother of a child with SCD realised late at night the meter needed more money. She didn't have any money at all on her and did not get paid the next day. She phoned her energy company to see if they could extend the emergency credit explaining her daughter was ill. Her energy company agreed but could not do it until the next day. With no family nearby she drove across the city to borrow £10 off a friend. However by the time she was back the shop selling the top up had already closed. The only way she felt she could keep her daughter warm and pain free was to huddle up in bed with her under the duvet. Not having money for the meter at all times was a worry for those who believed they needed to maintain a warm house to avoid a crisis:

With a direct debit as I say I've no idea of how much I pay a month, but yeah, I do definitely prefer, because actually if I've not got that money to top up a meter then I'm just sat in my house freezing which all then equates to be in hospital. P13

On the other hand, those on direct debits or paying bills monthly/quarterly were worried about large bills coming through and getting into debit. Therefore paying for heating could also act as a different trigger for SCD, that of stress.

Not having gas or electricity because of no money to top up the meter prepayment cards also had an impact on their diet and ability to drink plenty. Without gas or electricity they were unable to cook, keep food in the fridge fresh, and prepare warm drinks. Very cold drinks or foods were reported to risk a crisis. And when someone had got cold or started to feel pain one of the remedies was to prepare a hot drink:

Well to give him some painkiller, and I want to put him very good clothes, and I want to give him the warm drink. I give him the warm drink, the warm drink not very cold, drink warm drink and some painkillers and keep the house very warm. P2 (mother of 8 year old with SCD)

Other consequences of no credit in a meter were not being able to prepare a hot water bottle or wash up dishes or bathe in warm water. Some participants thought it was less stressful to pay by meter and others by direct debit/bills. Below is a table listing some of the reported advantages and disadvantages of the different payment methods for preventing stress:

Fig 2 Stresses of different payment methods

Payment method	Meter	Direct debit/bills
Advantages	<ul style="list-style-type: none"> • Avoids the debts and shocks of big bills • You can see what you are using 	<ul style="list-style-type: none"> • Can refuse to have a meter put in on health grounds • Can negotiate monthly payment • You can have heat even when you don't have money • Can't see what you're using • Can spread payments over year
Disadvantages (sources of stress)	<ul style="list-style-type: none"> • Gas or electricity can get cut off when emergency credit runs out • Shop may be shut to top up card • Have to go out in cold to get top up • Have to take young children out with you to get top up • If you have gone into debt even more is needed to get into credit + have some for heating • Standing charge for meter – even when no gas appliances connected • May not be able to crouch down for meter • Meter may be outside in cold • Paying up-front in winter 	<ul style="list-style-type: none"> • Frequent drops in income and sanctions can lead to fees for unpaid direct debit/unauthorised overdraft

One participant highly recommended a third way of paying. He had his fuel payments plus a small debt taken out of his benefit payment before he received his benefits. This meant he was happy that his heating was always paid for and he didn't have to worry about not being able to heat his house.

DISCUSSION

In 2010 there were 2,489 emergency admissions for 'sickle cell anaemia with crisis in children (cost per patient £4,198) and 7,714 emergency admissions for 'sickle cell anaemia with crisis or with complications and co-morbidities (CC)' in adults (cost per patient £11,367). In addition there were 906 emergency admissions for 'sickle cell anaemia without CC' in adults (cost per patient = £637). The approximate total cost to the NHS of these admissions was around £16.2 million.²²

Although the triggers for each sickle cell crisis can vary and are hard to determine there is a case for an intervention to support people with SCD with provision of heating. Hospital admissions for a SCD crisis cost between £637 and £11,367, whereas the average annual bill for gas and electricity is approximately £1,200.²³ It has been demonstrated above that keeping warm is a major factor in preventing a SCD crisis. In addition the cost of heating was also found to have an impact on other triggers of avoiding stress, eating well and drinking plenty. Therefore there is great potential for preventing crisis and savings for the NHS by helping people with SCD maximise their ability to heat their houses. This is particularly important for those most at risk of fluctuations in income such as benefits sanctions and DLA suddenly being withdrawn. One participant talked of heat as a medicine people with SCD need as much as drugs:

The part of their medicine is warm clothing, heating and fresh food and drink. If that's not endorsed by the health authorities it doesn't go into a care package. Now if you have arthritis and you need a certain drug for rheumatoid arthritis, that goes into your budget. So £10,000 per year for those drugs is what keeps that person healthy. But there's not that for a sickler. So there needs to be a health package, so much for heating, so much for clothing. P8

By helping one person with SCD per household stay warm there is also the potential to improve the health of other members in the household. Among the households in this study there were other conditions affected by cold homes. These were other people with SCD, people with SCD trait, young children with respiratory conditions, arthritis, mental health problems, Raynaud's Syndrome. People with SCD may have co-morbidities such as heart conditions and history of stroke that are affected by cold. A more in depth health economics study could be carried out to examine the potential savings to health and social care in households where there is a person with SCD.

However this is not always an easy group to conduct research with. This is partly because of the debilitating impact of the condition and sometimes because of difficulties communicating with participants when they cannot predict when they might become suddenly hospitalised with a crisis. There are other problems such as incomes so low that potential participants

²² National Institute for Health and Clinical Excellence (2012) Sickle cell acute painful episode. *Costing report Sickle cell acute painful episode Implementing NICE guidance*
<https://www.nice.org.uk/guidance/cg143/resources/cg143-sickle-cell-acute-painful-episode-costing-report2>

²³ Consumer Futures 2014. *Retail Price Information* <http://www.consumerfutures.org.uk/our-work/project4/retail-price-information#Bookmark4>

may not even have sufficient phone credit to contact a research project, or access to internet or email. However there are other problems caused by the industry of research itself. Participants can often find opportunities to be involved in research, particularly biomedical research trialling new medicines. Their past experience has not always been positive and can lead to reluctance to get involved in future research because they do not see any thing has come of their efforts in the past:

But you know what I find, there's a lot of people, I've been to a lot of meetings, research pertaining to sickle cell, and still I don't see anything being done, nothing at all. P5

To overcome these barriers to research participatory research methods may be suitable for this group because there is more ownership and control of the research and the use of the results.

In addition, working out how to address heating among the SCD population could help find solutions for people with other conditions, particularly fluctuating conditions such as M.S., Crohn's disease, M.E., arthritis and mental illness where people are at similar risk of not being able to work, qualify for disability benefits or avoid JSA sanctions. These conditions can also be affected by cold. Similar problems with sanctions have been reported with these conditions²⁴. The challenge will be to match investment in addressing fuel poverty in groups with cold vulnerable conditions to savings in health and social care and to see 'heat as medicine'.

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²⁴ MS Society, Arthritis Care, Parkinsons UK, Forward-ME Group, National AIDS Trust, Crohn's and Colitis UK (2011) *Employment and Support Allowance Work Capability Assessment review Making it work for fluctuating conditions* <http://www.nat.org.uk/media/Files/Publications/Apr-2011-Employment-and-support-allowance-WCA-review.pdf>