Sickle cell disease

Appropriateness in access to automated red blood cell exchange:

The patient perspective

Health Innovation Yorkshire & Humber

SICKLE CELL SOCIETY

Health Innovation Network

Contents

Acknowledgements

This report will allow commissioners and providers of services to have more of a holistic view of the issues faced by patients, and the capacity issues within the healthcare system. These findings will be shared with NHS England and other relevant stakeholders to help improve policy, and hopefully improve access to automated red blood cell exchange across England.

We feel privileged to have had the valuable contributions of numerous individuals living with sickle cell in the development of this report. To have had those with lived experience involved has led to a rich and meaningful learning opportunity.

Thank you to the Sickle Cell Society and the Clinical Reference Group for Haemoglobinopathy for helping with the delivery of this report. Thank you to the National Spectra Optia Working Group for co-ordinating this work and providing such valuable input.

Introduction

A national delivery strategy and plan has been developed to increase patient access to automated red cell exchange for sickle cell disease patients as part of NHS England's (NHSE) MedTech Funding Mandate policy.

NICE Guidance MTG 28[1] describes the use of the Spectra Optia machine to enable automated red cell exchange for sickle cell disease patients and was added to the MedTech Funding Mandate (MTFM) policy[2] from 1 April 2022. Initial research, led by the Health Innovation Network and NHS England's Accelerated Access Collaborative (AAC) policy team, showed a relatively high prevalence of machines across England's NHS as the machine has multiple uses. However, intelligence also suggested that the machine capacity was often taken up for alternative use, so there was a significant opportunity to increase sickle cell patient access to automated red cell exchange.

Therefore, to ensure the NHS in England is providing more equitable access to treatment for those patients who require both routine, elective care for their sickle cell, and for those in crisis, who arrive at night in significant pain and need, the key national organisations and stakeholders from across England came together to agree a strategy and plan.

The AAC MedTech Funding Mandate (MTFM) Policy team and Health Innovation teams worked with NHS Commissioning Specialised Services team, Haemoglobinopathy Coordinating Centres (HCCs) and NHS Blood & Transfusion (NHSBT) to develop a coordinated and collaborative approach to understanding and improving sickle cell patient access to automated red cell exchange, and improvement to sickle cell pathways. This formed the basis of a National Spectra Optia/MTG28 Working Group. Preliminary working group discussions about patient access and the All-Party Parliamentary Group (APPG) report indicated that patients may travel long distances to access trusted providers for sickle cell care. As a result, understanding patient perspectives on care accessibility was prioritised.

It has been essential to understand where the MTFM fits in the wider sickle cell clinical pathway improvement programme. There is also a need to ensure that the MTFM plays its part in fulfilling the recommendations within the APPG-driven roundtable parliamentary party meetings to look at the 'No One is Listening' recommendations (link here).

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1 Spectra Optia for automatic red blood cell exchange in people with sickle cell disease Medical technologies guidance [MTG28]Published: 02 March 2016 www.nice.org.uk/guidance/mtg28

2 MedTech Funding Mandate policy: guidance for NHS commissioners and providers of NHS-funded care 10 March 2022 www.england.nhs.uk/publication/medtech-funding-mandate-policy-guidance-for-nhs-commissioners-and-providers-of-nhs-funded-care

Patient Voice

To ensure that the spread and adoption of Spectra Optia is as effective as possible when improving access to treatment across England, it was important to ask sickle cell patients themselves how they viewed the appropriateness of their access to automated red cell exchange treatment. To do this, NHS England's Patient and Public Involvement and Engagement Team in the Innovation, Research and Life Sciences Group, the Sickle Cell Society, and Health Innovation Yorkshire & Humber (formerly known as Yorkshire & Humber AHSN), designed a patient survey which was distributed to patients via the Haemoglobinopathy Co-ordination Centres and the Sickle Cell Society members.

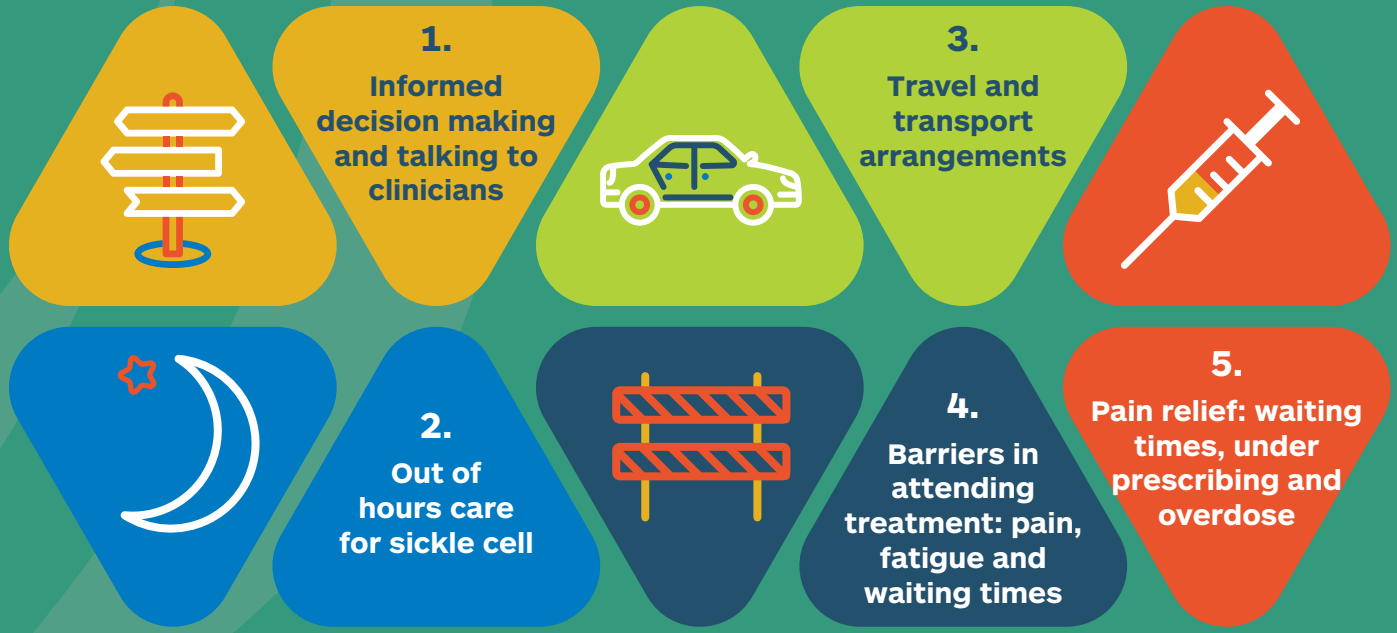
A jointly presented webinar with the Sickle Cell Society and Health Innovation Yorkshire & Humber was also held on 20 April 2023, which enabled the team to present the results of the survey to the Sickle Cell Society members and other interested stakeholders. Inputs from this webinar have been used in this report.

Executive Summary

157 responses were received following dissemination of the survey, 153 of these were completed fully enough to be analysed. 46 respondents were Sickle Cell Society members, and 107 were submitted by individuals via the Haemoglobinopathy Coordination Centre networks.

80% of respondents were from people living with sickle cell disease themselves, with the remainder being from a carer, relative or friend of someone living with sickle cell (on behalf of a patient with sickle cell). This meant that 100% of the information received and analysed within this report came directly from those with lived experience of the condition.

Analysis of the results indicated a need for change across five priority areas within the sickle cell treatment pathway.



Patient Engagement

The table below illustrates the characteristics of the respondent population from 153 valid responses.

1.	Greatest number of respondents in age bracket 35-49. 59% between ages 35 and 64
2.	Around 2/3rds of respondents were female
3.	90% of respondent identified as black
4.	47% of respondents are disabled
5.	No respondents said they are currently pregnant
6.	88% of respondents list their main language as English
7.	79% of respondents have been educated to either degree or A level*
8.	32% of respondents are currently unemployed**

* It should be noted that nearly 80% of respondents were educated to either degree or A-level standard. An assumption could therefore be made that these people may be in higher income brackets, which will influence whether they are able to afford a car, or pay for taxis. We should therefore be conscious that this survey may not represent those who are not digitally literate, or those population groups where English is not the first language. Those patients who are not engaged with their HCCs or Sickle Cell Society members may also have not had an opportunity to complete this survey.

** Risk of emergency readmission and inpatient mortality was found to be higher for patients living in more deprived areas - this is important context.

3 Research and analysis: Labour market value of higher and further education qualifications: a summary report. Updated 9 February 2023 www.gov.uk/government/publications/labour-market-value-of-higher-and-further-education-qualifications-a-summary-report

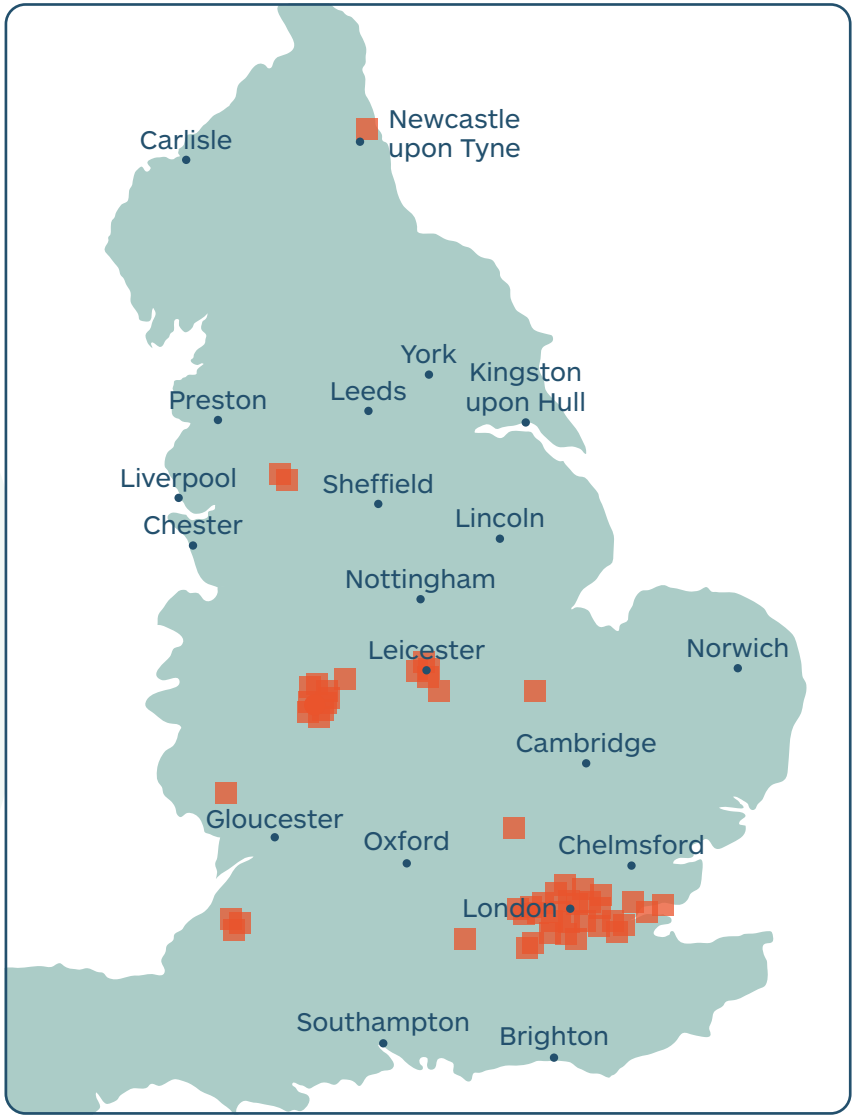
4 Aljuburi Get al. Socio-economic deprivation and risk of emergency readmission and inpatient mortality in people with sickle cell disease in England: observational study. J Public Health (Oxf). 2013 Dec;35(4):510-7. doi: 10.1093/pubmed/fdt100. Epub 2013 Oct 29. PMID: 24169414. https://pubmed.ncbi.nlm.nih.gov/24169414

Respondents were asked to share some information on their own experiences of sickle cell treatments.

1. 80% of responses are from people living with sickle cell.
2. 90% of respondents have received treatment for sickle cell with 58% currently having or have previously received Hydroxycarbamide.
3. 74% of respondents regularly visit their hospital outpatient clinics for sickle cell treatment.
4. 93 respondents have discussed automated red cell exchange during a conversation with a healthcare professional of which 60 (64%) felt they made an informed decision to undertake treatment and had equal input into the discussions.
5. 30 respondents answered they currently receive automated red blood cell exchanges with nearly all receiving treatment at a frequency of four weeks or longer.
6. 80% of respondents have needed treatment for sickle cell as an emergency out of normal working hours with over 4 out of 5 patients receiving medication as treatment.
7. 65% of respondents that have regularly received automated red blood cell exchanges felt that it had a positive effect on their day to day lives, with 47% saying they felt it had helped greatly.

Of the survey responses, 153 of them contained enough information to analyse. In order to get an insight into the accessibility of automated red blood cell exchange across England the locations of the patients and the trusts where they were receiving treatment was important to determine. 75% (114) respondents were receiving automated exchange and a partial postcode so that length of travel could be determined.

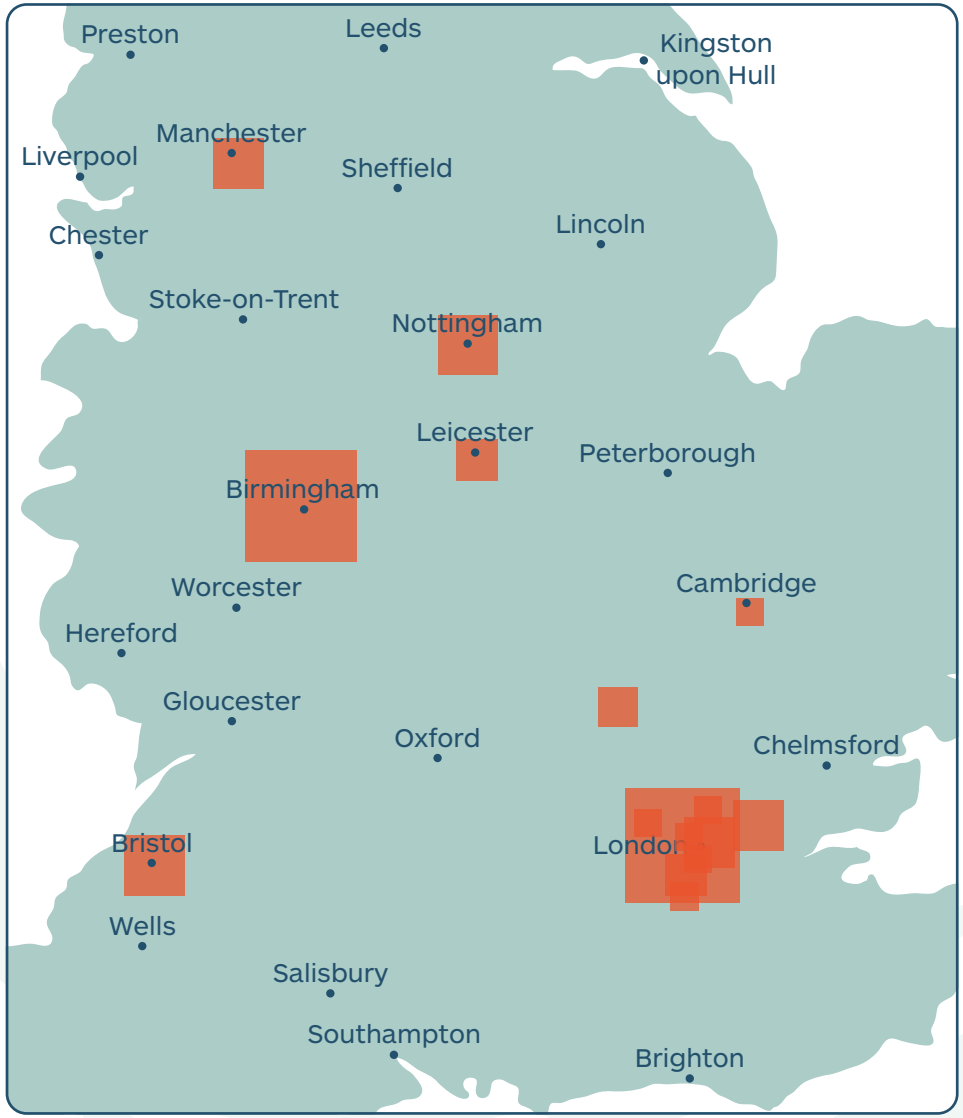
Health Innovation name	Responses	Commissioner Name	Respondents
East Midlands	10	NHS East Leicestershire and Rutland CCG NHS Leicester City CCG	5 5
Eastern	4	NHS Bedfordshire, Luton and Milton Keynes CCG NHS Cambridgeshire and Peterborough CCG NHS West Essex CCG	2 1 1
Health Innovation Manchester	3	NHS Manchester CCG	3
Health Innovation Network	7	NHS South East London CCG NHS South West London CCG	5 2
Imperial College Health Partners	8	NHS North West London CCG	8
Kent, Surrey & Sussex	3	NHS Kent and Medway CCG NHS Surrey Heartlands CCG	2 1
North East & North Cumbria	1	NHS North Tyneside CCG	1
Oxford	1	NHS Berkshire West CCG	1
UCL Partners	52	NHS North East London CCG NHS Thurrock CCG NHS North Central London CCG NHS Basildon and Brentwood CCG NHS Castle Point and Rochford CCG NHS Southend CCG	40 5 3 2 1 1
West Midlands	14	NHS Black Country and West Birmingham CCG NHS Birmingham and Solihull CCG NHS Herefordshire and Worcestershire CCG NHS South East Staffordshire and Seisdon Peninsula CCG	7 5 1 1
West of England	3	NHS Bristol, North Somerset and South Gloucestershire CCG	3



As the survey was distributed by the Sickle Cell Society to its members and via the Clinical Reference Group to the Haemoglobinopathy Coordination Centres across England, the survey received a good number of responses. However, it is worth noting that while the geographical spread of respondents was somewhat varied, they were largely concentrated in London and the West Midlands. With 16 responses from Imperial College Healthcare NHS Trust and 15 from Sandwell and West Birmingham Hospitals NHS Trust. As both these regions have local access to automated red blood cell exchange, this needs to be taken into account when considering patient travel times from more rural areas.

Which hospital(s) (name and location) do you visit to receive your automated red blood cell exchange? Response rate = 56% (85) 21 unusable responses

Trust and Health Innovation volumes in table, map shows geographical spread of trusts with density of square bigger for higher volumes



Health Innovation name	Responses	Trust Name	Respondents
East Midlands	6	Nottingham University Hospitals NHS Trust University Hospitals of Leicester NHS Trust	4 2
Eastern	1	Cambridge University Hospitals NHS Foundation Trust	1
Health Innovation Manchester	3	Manchester University NHS Foundation Trust	3
Health Innovation Network	7	Lewisham and Greenwich NHS Trust St George's University Hospitals NHS Foundation Trust Epsom and St Helier University Hospitals NHS Trust King's College Hospital NHS Foundation Trust Guy's and St Thomas' NHS Foundation Trust	2 2 1 1 1
Imperial College Health Partners	17	Imperial College Healthcare NHS Trust London North West University Healthcare NHS Trust	16 1
UCL Partners	11	Barts Health NHS Trust Barking, Havering and Redbridge University Hospitals NHS Trust Bedfordshire Hospitals NHS Foundation Trust University College London Hospitals NHS Foundation Trust Homerton Healthcare NHS Foundation Trust North Middlesex University Hospital NHS Trust	3 3 2 1 1 1
West Midlands	15	Sandwell and West Birmingham Hospitals NHS Trust	15
West of England	4	University Hospitals Bristol and Weston NHS Foundation Trust	4

Due to the lack of geographical spread of respondents the survey did not produce a comprehensive visual regarding the length of travel for patients receiving automated red blood cell exchange, and further analysis will need to be completed.

Patient Choice

Interestingly, during the webinar, some sickle cell patients, or those caring for others, gave an alternative view to the issues around the distance travelled. Rather than seeking care and treatment closer to home, patients were prepared to travel further in order to receive care from those trusts, and NHS professionals that they knew and trusted. When commissioning these new or expanded services, it is important to consider that patient perspectives on service accessibility directly impact their care experience. Therefore, initiatives aimed at enhancing red cell exchange accessibility should be accompanied by efforts to enhance the overall patient experience.

‘I live in Luton and my local hospital is the Luton & Dunstable Hospital. This hospital works in collaboration with the UCLH in Euston to allow some sickle cell patients to have the Red Blood Cell Exchange’

‘My local hospital doesn’t understand whereas my main one does’

‘My main hospital looks after me well. Nothing is too much. My local hospital has very little knowledge and no empathy’

‘Here in * pretty bad, the doctors didn’t have much information about sickle cell disease. in Leicester I am having a good experience’**

Informed decision-making and talking to clinicians

18 people directly affected by sickle cell disease specifically chose to write comments based on their experiences of initially accessing treatment. One of those comments was positive, with ten being negative in nature, and seven being neutral.

	Responses
Yes	60 (64%)
No	24 (26%)
Not sure	9 (10%)

26% **of those affected by sickle cell felt that they had not made an informed decision to undertake automated red cell exchange during a conversation with a healthcare professional, nor had equal input into the discussions.**

	Responses
Yes	93 (61%)
No	51 (33%)
Not sure	9 (6%)

33% **of those affected by sickle cell described themselves as not having a conversation about receiving blood transfusions with a healthcare professional.**

Views obtained via the survey illustrate a varying picture of the information patients receive and the discussions they have with healthcare professionals. It was not possible to clearly determine from the results, where in the treatment pathway, help and support was being offered (whether it was primary or secondary care), however, there was evidence that patients have to persevere to access treatment.

‘My GP doesn’t attend to me; I do complain but I don’t have the time to see a doctor each time I call’ and my GP I don’t really have access to talk to a doctor anytime I have pains’

‘Being listened to after a lot of pressure’

‘It’s a bit hit and miss but, I was even refused a new treatment protocol’

‘In the last 3 years my care has been non-existent I have complained to the trust concerning the treatment and non-communication concerning my sickle cell care plan. I found it very different to get a response for my concerns’

‘Now I get transfusions which I had to fight for’

‘I would like a blood transfusion but in my adult years it’s never been offered to me’

Furthermore, numerous positive experiences were reported, predominantly occurring after patients had sought care from specialised units within secondary healthcare.

Discussions around the amount, and quality of information that patients receive was the first issue discussed at the 'Sickle Cell Disease: Appropriateness in Access to Automated Red Blood Cell Exchange' webinar. Evidence suggests that automated red blood cell exchange is far preferable than having manual exchange, it is quicker, uses less blood, causes less iron build up in the patient's body, and is more cost effective to the NHS[5]. This treatment option is not the only, or even the best course of treatment for some sickle cell patients. However, it is important that patients are aware of the procedure and can make informed decisions about their options. So, that 33% of the respondents completing the survey have never discussed the option with their healthcare professional is concerning.

'Sickle teams work with patients to ensure we are informed about choices and treatments. Specialist sickle cell nurses are doing a great job in aftercare support and with providing guidance'

'The specialist nurses I have encountered live and breathe the service. They aim to give everyone a positive experience and it really makes a difference'

'I have been informed all the way through of what has affected my crisis, things I can do to better my day-to-day life and about the new treatment I am on by my doctors'

'The specialist outpatient team are wonderful and frequently jump through hoops of fire to ensure my needs as a patient are met'

5 Spectra Optia for automatic red blood cell exchange in people with sickle cell disease Medical technologies guidance [MTG28]Published: 02 March 2016 www.nice.org.uk/guidance/mtg28

In general patients felt that they had a good relationship with their haematologists, and gave some examples of robust personalised care plans, with some patients trying automated exchange and then not tolerating the procedure. Examples of patients working with their clinicians to find the right treatment for them through trial and error were presented. People affected by sickle cell disease felt that the information provided on the Sickle Cell Society website and via the National Hemoglobinopathy Panel were good resources.

However, this being the case, there is an absolute necessity for the provision of more information, individuals who do not have access to specialist advice need to be able to access accurate information, either via websites, support groups or their GPs. Delegates on the webinar mentioned, *'patients not going to hospital'*, and people *'not knowing what the treatments are'*. A mother of children with sickle cell presented that she felt that *'you don't hear about sickle cell treatments until the time is right'* or *'at a time dictated by either your consultant or your illness'*, *'when things get really bad'*. She had a perception that automated red blood cell exchange was an emergency, not an elective procedure.

Another noteworthy concern regarding patients missing their appointments stems from two main factors: some feel well and consequently lose touch with specialized services, while others refrain from attending treatment due to a lack of understanding. Raising the question: can more be done to follow up these patients? Could more information be available for those who are not engaging regularly with healthcare services?

Clinicians taking part in the webinar were also concerned about this issue as they felt that a good quality management system is needed to ensure patients were adequately followed up where necessary. This improvement could be driven by a change in the coding of patients as they travel through the system. For example, a sickle cell patient who is experiencing another illness may be recorded as presenting with that illness, rather than sickle cell. This can be problematic, as like any other chronic conditions such as diabetes or high blood pressure, even if there is no pain, there can be silent organ damage, therefore it is important all patients can be identified and managed.

Information should also include the wider needs of the sickle cell patient, for example, travel and transport arrangements, counselling, the psychological effects of treatment, information around cultural and religious issues for example, regarding the taking of blood.

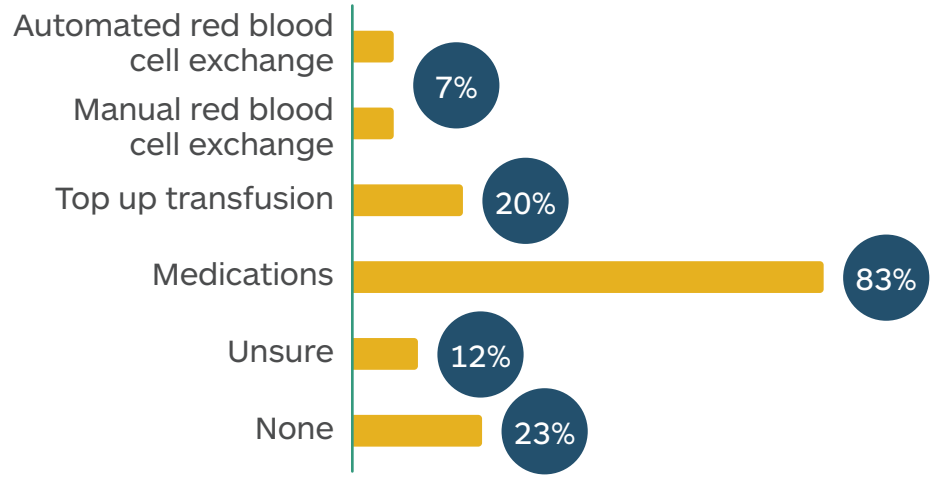
'The machine, if you like, we used to call it 'our best place'. And because my daughter came in and she was really unwell, and she had an emergency transfusion. So, I knew that there was such a thing. I just never knew that it was a programme, and it was available where you could come in and regular, you know, a regular interval to keep yourself well. So, I didn't actually get to know that until my son was really poorly and again my daughter was experiencing some really bad back pain after that. And we then made the decision to do the exchange programme'

Out of hours care for sickle cell

121 (80%) of the 151 respondents affected by sickle cell disease have needed treatment for their sickle cell as an emergency out of normal working hours (i.e. outside of 9 am to 5 pm Monday to Friday). This is very revealing, as within the 'No One's Listening' report[6] if it is A&E the patients attend, it is universally a poor service.

	Responses
Yes	121 (80%)
No	25 (17%)
Unsure	5 (3%)

If 'Yes'. did you receive any of the following treatments?

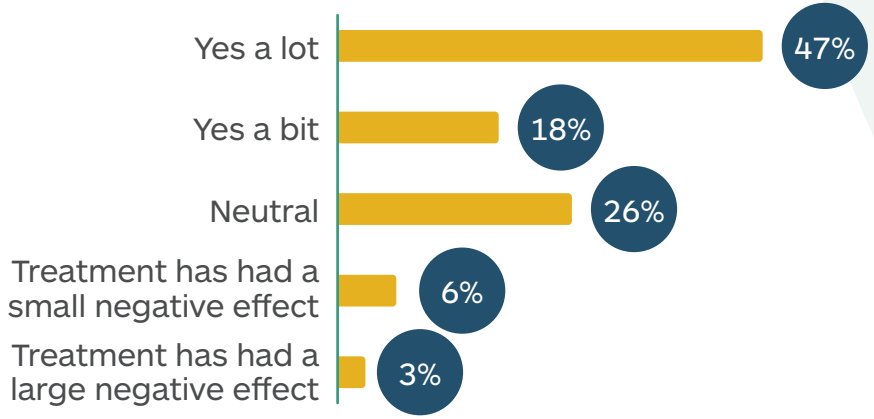


Receiving treatment Out of Hours is not unusual, as sickle cell is an unpredictable condition and patients could experience crisis at any time. It was noted that of those 121 (80%) patients attending out of hours, 34% needed a blood transfusion and top up, yet only 7% received an automated exchange. Clinicians on the webinar cited lack of available resources and capacity as major issues, including access to Spectra Optia machines, the availability of staff and even the physical space required to undertake the treatment which were all deemed to be barriers to providing optimal treatment. It is hoped that the MedTech Funding Mandate can help provide organisations with the necessary resources to expand their apheresis services.

6 All-Party Parliamentary Group (APPG) inquiry report November 2021 www.sicklecellsociety.org/wp-content/uploads/2021/11/No-Ones-Listening-Final.pdf

I have had manual red cell exchanges which was extremely traumatic because the hospital said the automated machines wasn't available on the weekend. When attending A&E I am often left for hours waiting for IV paracetamol and morphine and ignored on the wards. I'm have also been put onto wards where nursing staff are unfamiliar with Sickle Cell, PCA is delayed for hours, and crisis become worse in hospital because of the delay. However, my consultant is excellent when I see him. It is just the care when being admitted into hospital that is very challenging.

Respondents were asked, if you regularly received automated red blood cell exchanges, do you feel that they have had a positive effect on your day-to-day life? (Ability to support everyday life and activities)?



This question confirmed that for those patients requiring treatment 65% of patients felt that it had a positive effect on their lives, with 47% stating that this positive effect was significant. It therefore makes sense, that where possible, this treatment option is offered.

'I cannot now live without it'.

'I find the frequency of my crisis is less'

'I have had one exchange. It was the best I ever felt'

'Receiving blood transfusion for over 30 years gave an amazing quality of life'

What is also interesting is that 35% of patients experienced a neutral or even negative effect, and more could be done to uncover why this was the case. Comments made within the survey noted that:

‘Can get violently sick during or just after exchange, the more I feel before exchange the longer it takes to recuperate which eats into my good period. Good period after exchange can kick in after anything from 3 days to about a week. Feel boost of energy for about 3-4 weeks then start to decline. Exertion, cold weather and infection speed up rate of decline post exchange’.

‘Could not continue due to difficulties to canulate’

Another key issue to consider is that in an acute situation it may be more clinically appropriate to give a top-up of blood rather than providing a full transfusion and having to find blood, venous access and putting in the lines. Top-ups could ease the patient’s suffering quicker.

Respondents often mentioned their preference to avoid A&E and self-manage their condition for as long as possible.

‘It’s too stressful to use A&E so I tried to avoid it’.

‘A&E staff to be trained on how to deal with sickle cell emergencies’.

‘Outpatient treatment at [the hospital] is very high quality. But the emergency inpatient care is still quite a negative experience’.

‘Improvements could be made within the emergency department where doctors need to listen to the patient more rather than make decisions without discussing it with the patient’.

‘There is a day unit that understands my condition, but they have limited beds…. most [of the] time I call because [I] am scared of going to A&E but they say no bed, no funding. They open Monday to Friday, 8-6, which makes it difficult to get the appropriate care you need’.

‘A&E is often delayed and not easy to access treatment’.

‘As an outpatient it is reasonable. I am scared to be admitted as an inpatient when I have a crisis now because I had such a bad time last time I was in hospital. I feel like there is still a lack of care and understanding from some carers and nurses on the wards when it comes to sickle cell’.

Out of hours visits for those affected by sickle cell are frequent, and there is a drive by NHSE to improve staff knowledge of the disease within Emergency Departments, which is echoed by the survey findings. One of the challenges with clinicians’ education and training is the disparity in prevalence across the country. Some regions such as London, Birmingham and Manchester have larger sickle cell prevalence, therefore staff within these areas are routinely treating patients. In other regions, such as the South-West and North-East, there are fewer patients, making it more challenging to maintain staff awareness and competencies.

Primary issues such as whether staff were giving support, providing compassion and presenting patients with kindness and empathy received a mixed response with 40% of respondents feeling this was a positive, and 60% having a more negative response.

Giving patients the confidence to attend emergency departments when they are in acute need is extremely important and will require a number of interventions across multiple stakeholders. Providing patients with the empathy and compassion they deserve and out of hours access to the most appropriate treatment and medications is crucial.

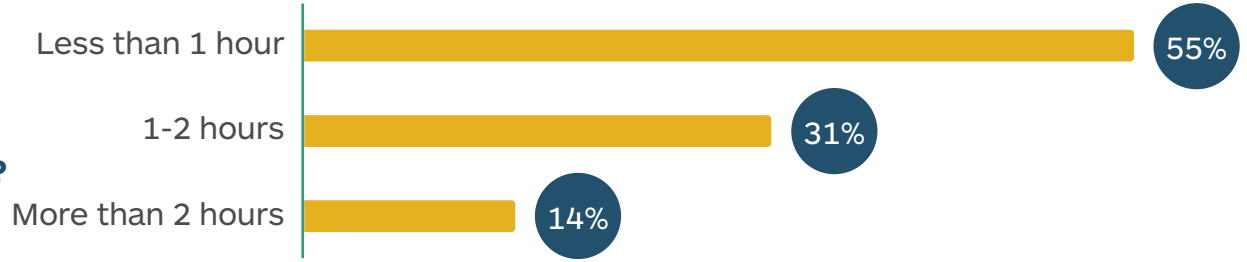
As part of the MedTech Funding Mandate, Haemoglobinopathy Coordinating Centres are being invited to submit business cases in order to expand their apheresis services, and in particular to increase access to automated red blood cell exchange. Commissions should be able to look at where any deficiencies in the system are so that they can be addressed.

Travel and Transport

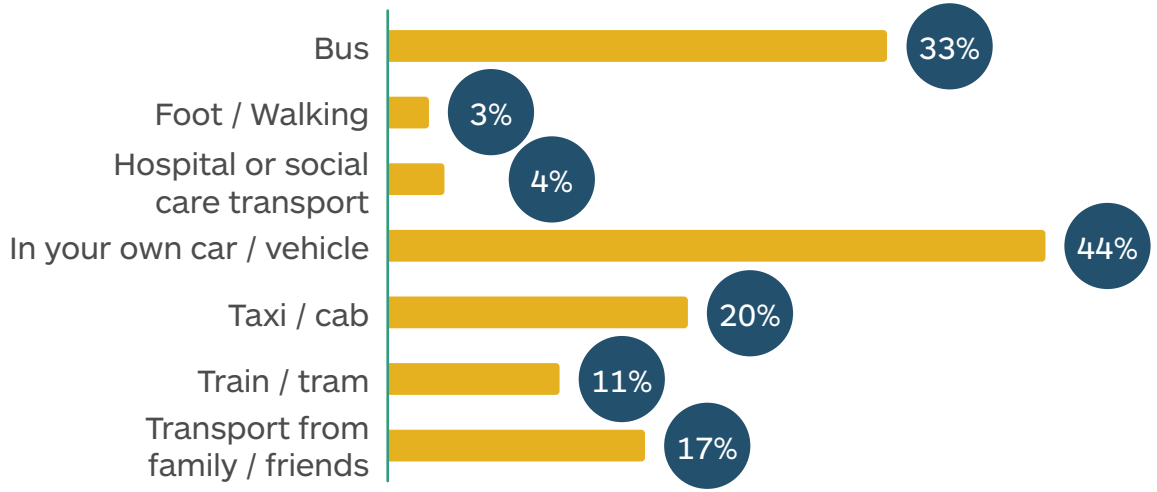
Anecdotal evidence shared at the MTFM National Working Group highlighted the inappropriate amount of travel that people were expected to make in order to receive their automated exchange treatment, and this survey confirmed it, with 55% of people travelling less than an hour; 31% between 1-2 hours and 14% over 2 hours.

The transport predominantly used was buses, cars and taxis. One of the respondents replying to this survey stated that they had 'opted not to try automated exchange because of the amount of travel involved'.

How long does it normally take you to travel to receive an Automated Red Blood Cell Exchange? (Response rate = 42% (64)



Which mode(s) of transport do you use to access your treatment?



The distance patients need to travel in order to receive automated red blood cell exchange as a treatment is known to be a barrier for patients. What these survey results indicate is that there are inconsistencies that exist for patients, beyond travel being a disincentive. Commissioners and providers should consider producing a clearer policy about transport, looking into other conditions to see if there is a more inclusive model that can be adopted.

'I have never actually had the blood exchange, but I had to travel to the hospital to discuss it. One of the reasons I opted not to try it was because of the amount of travel involved'

Some patients are willing to travel longer distances to access specialized services, even if it means going beyond their local hospital, as they view this travel as essential to receive more tailored treatment for their condition.

'I am happy to travel to get what I need and to get that specialist input with those who are passionate about [sickle cell management]'

'I travel to * Hospital because *** staff have no clue. I am grateful to my consultants and nurses'**

Other issues regarding the wider impact of travel and transport were also made.

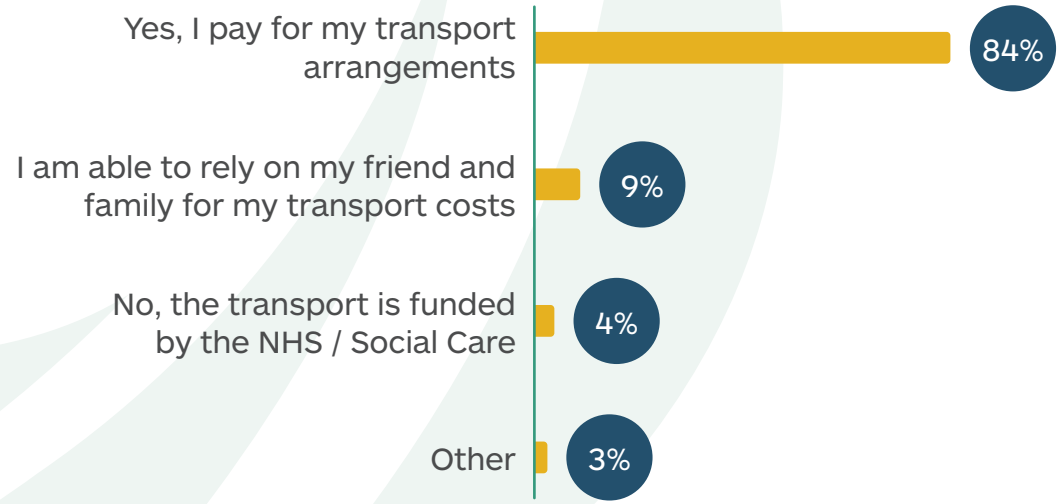
During the webinar, senior clinicians raised the issue of patients not attending their appointments. Delegates discussed whether the distance people have to travel, and the complexity of their journey contributed to the number of missed appointments. Patients who are delayed often miss their appointments as their slot has been given away because of capacity issues, meaning that providing treatment available closer to home could be more cost effective in the long run.

A key point made by a patient who received treatment in London was that although they had regular public transport, and three main hospital sites to choose from; following 4-5 hours of transfusion, navigating the underground, in rush hour was not the best experience, and especially not for children. Driving in London was also problematic with limited parking and parking costs. On more than one occasion patients would have to wait three hours, then go and move the car.

Return travel was also discussed by a patient who regularly travels between Milton Keynes and London on the train for treatment, and who found it so intolerable that they were going to give up treatment until they found out transport was available.

Perhaps the main issue highlighted was the variation in transport and travel across the county, and the inconsistency that patients experience. During the webinar, a delegate expressed the view that 66% of individuals with sickle cell disease reside in economically disadvantaged communities, which could pose a significant obstacle to patients experiencing deprivation due to travel expenses.

Do you normally cover the costs of your transport when accessing Automated Red Blood Cell Exchange? Response rate = 50% (76)



One sickle cell patient explained their experience of living in an area where there was no access to automated exchange in the local hospital. They required an ambulance to reach a larger hospital which was also the HCC. Only in this way, they were able to access the treatment that was right for them, but this took time as funding arrangements had to be agreed leading to a delay in the provision of treatment.

Barriers in attending treatment: pain, fatigue and waiting times

Respondents held generally positive views on their medical care, but negative views on how much they needed to wait for pain relief and access to treatment. As pain is a key feature of sickle cell crisis, NICE guidance recommends pain relief should be given to patients within 30 minutes of getting to the hospital.

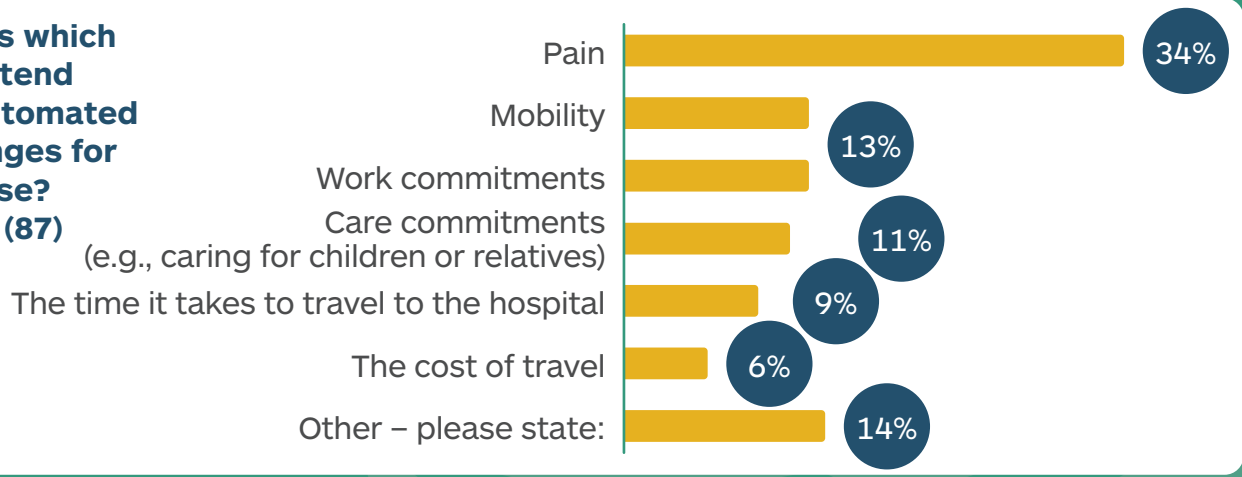
80% (4 out of 5) of respondents reported they received pain relief at once when attending hospital in a sickle cell crisis. It is worth noting that patients experiencing significant pain may not accurately gauge the duration of their wait, but their perception of when relief was provided remains invaluable.

Other comments regarding barriers to accessing care and treatment were:

- Apart from mobility, the fear of knowing that I'll have a lengthy wait at A&E
- Complications
- Fatigue
- Pain and the cost of travel
- Pain, mobility, travel time and cost
- The attitude of the nurses and some doctors
- Waiting time at hospital

Pain was identified as the biggest barrier to attending hospital or accessing automated red blood cell exchanges.

Are there any reasons which make it difficult to attend hospital or access automated red blood cell exchanges for your sickle cell disease? Response rate = 57% (87)



Delegates who attended the webinar were shown the results of the survey and were invited to discuss opportunities for improvement. One key suggestion was to ensure that medical staff within the NHS are well-informed about sickle cell disease. When a patient encounters a clinician upon arriving at the hospital who is either unaware of the condition or who asks how long the patient has had the condition for, it undermines the patient's confidence in the healthcare system and their belief that they will receive attentive and appropriate care. There were discussions about how a sickle cell patient presenting in crisis in the A&E could be seen quickly, perhaps directed to a more appropriate unit, which has capacity. Some responses suggested that capacity in these units need to be increased.

'When I went to A&E with my daughter with severe pain. My daughter was asked to wait outside, when the pain I had was so excruciating to do anything for myself. When I challenged the security staff at the door, I was told my daughter can queue for me. I do not think sickle cell patients should queue especially when going through crises'

'Everything is usually great apart from emergency visits where wait times are so long that have almost experienced death like experience'

'We often use A&E, but they are always overwhelmed or overcrowded. Can sometimes take a long time to get seen or treated. Our GP practices should be able to provide emergency support to manage a crisis, but we always have to go all the way to BCH'

'During crises I always have to wait in a queue at Accident and Emergency for hours to be seen'

'Better access for pain management, people don't always want to go to A&E which is why they attend scat and with only 4 beds available this is not always possible with the growing number of people now accessing this treatment'

'It's all the anxieties for me of going to my hospital. How I'm going to be treated. The fact that they don't really understand my condition makes your pain worse'

Pain relief: the wait, under prescribing and overdose

Although most sickle cell patients reported they tended to receive their pain medication ‘at once’ when attending the Emergency Department, when examining the responses, and after listening to patients’ experience throughout the webinar, it was apparent that timely and adequate pain relief was something which really needs to be addressed. The scope of this survey was to try and uncover patient experience on transport and the availability of access to automated red blood cell exchanges. However, some of the statements being made about pain should be addressed.

There was a consensus during the webinar that the pain caused by this condition was not always understood by clinicians leading patients to experience anxiety and vulnerability when seeking medical attention. This, in turn, acts as a significant barrier to their hospital attendance.

Treatment can be slow at times especially when the normal haematology team are not available. This usually means it can take longer to receive medication or treatment when presenting to A&E with a crises’

‘[Please improve the] urgency of which staff recognise and treat SC crises. Take our pain seriously, PLEASE. Trust me, I’d rather not be in A+E and I’m not hungry for drugs’.

‘I have found that the process of administering my pain relief also seems to be improving (e.g., my care plan is normally available, and I am given the right drugs without being asked what I am normally prescribed etc.) However, I do regularly still face long wait times before actually receiving that pain relief (far beyond the 30-minute NICE guidance)’

‘I find it really insulting to have to measure my pain score when I tell them it’s not actually on the scale, its off the scale’

‘I am often left waiting without pain relief in the general waiting area, which I find incredibly difficult. For example, if I have a pain crisis in my legs/hips the pain is made worse by sitting upright in the waiting room chairs’

‘General treatment once admitted to a ward is good, but being seen by a doctor in the initial stages from A&E and receiving pain relief can be a lengthy process where I’ve had to wait for hours in some cases’

‘No consistency with nursing staff and being treated like a drug addict. Have been overdosed on a few occasions due to misreading my notes and sensitivity to opioids’

‘During my last crisis, I called ahead so the team knew I was coming in by ambulance. It was late (approx. 2am). I was in agony and crying out for pain relief. It took hours for a Dr to attend and when he did, he gave me a very small dose of morphine. My pain level was 10 and this was not managed well at all’

‘My local NHS accident and emergency does not have a clue what sickle cell crisis is, the last time I was in crisis, I waited for more than 5 hours without pain relief when I tried to self-discharge, they gave me one’

During the webinar patients described that they knew their bodies well enough to say whether they are in crisis or whether they have an infection, and when they need pain relief.

‘It was more than three hours before I was given pain relief. They took me into A&E where I waited an hour for my family to come. I sat on a chair for a further two hours, because there were no beds available. Now, having my family watch that (I know they should be used to it by now) but it stresses me that they have to see that - I can’t do anything to help myself and the people that you’re talking to and not listening to you.

When I eventually did get pain relief. It wasn’t enough and I literally had to wait until I was put on Ward. I eventually spoke to a consultant, and I felt sorry for him because I’m crying. I’m putting my face in the bed because I don’t want to look at anybody. I don’t want anybody to see how much I’m struggling.’

It became obvious why the APPG report was entitled, ‘No one is listening’, as this was a message that came through strongly in the patient discussions.

‘I go into A&E with my care plan. There’s a digital one and I’ve got a soft copy on me, and I speak to the doctor in A&E will say to me, ‘that’s a lot of pain relief!’, looking at me in a patronisingly way, telling me, ‘ohh. You sure you should be on that?’, I have to explain that this document has been given to me by my consultant. So, I think my frustration really is the doctors not listening. They think they know better than my own consultant and try to make me feel like I’ve come there just to come over those on pain relief. So, what is the use of the consultant doing all that hard work only for another doctor?’

‘I’m not a patient, I’m a carer for my children, but I can understand the pain that they’re experiencing when they get into A&E. It really is. It’s horrible even for me as a parent. By the time I’m going to hospital, I’m beside myself. I’ve lost. I’ve lost control of the pain relief. There’s nothing more I can do. I need help like ASAP so I you know I really do understand that and to the point nowhere I don’t drive to the hospital when there’s an emergency anymore. I always phone the ambulance service.’

Inconsistencies in the administration of pain relief was mentioned on a few occasions. These inconsistencies started when the patient was onboarded by the ambulance teams, as the management of pain by patients taking part in the discussions ranged from receiving paracetamol to a morphine injection.

Sometimes you have to wait for a long time and sometimes you have to be giving the injection straight away. So, there's it depends on where, when, who you get, what time you get there, who you see and everything else.

Ultimately, the provision of pain relief that is adequate and timely is still inconsistent, and patients' needs must be listened to. There is also some learning for the NHS in terms of training and awareness across the system, including the ambulance trusts and A&E.

The positives

It is evident that experiences across the sickle cell treatment pathway is varied, and as such, a patient's experience is varied. Respondents to the survey made many extremely positive statements regarding their care and treatment.

Overall, how would you describe your care and treatment? (Response rate = 81% (124)

	Experience (#)			Experience (#)		
Total Responses	**Positive**	**Neutral**	**Negative**	**Positive**	**Neutral**	**Negative**
124	**67**	**39**	**18**	**54%**	**31%**	**15%**

'Hospitals, Doctors and Nurses are great. We just need improved and better treatment options to improve the quality of life'

‘Generally good, as the usual haematology team listen and tend to your needs efficiently. I have regular appointments and when I have a crisis and present to A&E am usually seen quite quickly.’

‘Staff attentiveness, proficiency of equipment, meaning I spend less time in the hospital. Friendliness of staff. Access to complimentary therapies’

‘I am the father of the patient of the sickle patient who is my daughter. She has never had an automated red blood exchange. But she is on medication. She is 4years old. She is treated very good by her doctor at Children’s and women hospital’

‘The blood transfusion has changed my son to a different person. He used to be in hospital every other month’

‘The Specialist Nurses I have encountered live and breathe the service. They aim to give everyone a positive experience and it really makes a difference.’

‘The specialist outpatient team are wonderful and frequently jump through hoops of fire to ensure my needs as a patient are met’

‘There are some really sweet caring people that help to ease the distress, pain & anxiety felt’

Opportunities for change and current developments

Opportunities for change, as determined by this report can be broken down into six areas. There is a huge amount of work being undertaken by NHS Race and Heath Observatory and the Health Innovation Network to improve the access, experiences and outcomes of healthcare services of sickle cell patients in England. It is important to take note of both the current actions being undertaken and the ongoing enhancements being implemented. These are highlighted in the table below.

	Challenge	Development
1	The clinical pathway is complex and in order to achieve change there is a need to work collaboratively.	The MTFM National Working Group for Spectra Optia meets regularly with a wide range of stakeholders across the NHS, all 15 organisations part of the Health Innovation Network, commissioners, providers and suppliers to work towards a shared goal: increasing access to services.
2	Increase patient facing information so people with sickle cell can make informed decisions on all eligible treatment options.	Partnership with the Sickle Cell Society, Terumo and NHSBT to create an AI Avatar for patients. NHS England working on programme to develop expert patients.
3	Guarantee out of hours care, the best possible care and access to automated red blood cell exchange. Sickle cell is not predictable, crises often occur outside of 9am-5pm Monday to Friday. Can we look at in-house solutions, or a collaboration with NHS Blood and Transplant?	MTFM has facilitated the production of business cases for the expansion of apheresis services where demand is exceeding capacity. In house expansion and contracting with NHSBT to provide more accessible hours are being realised.
4	Understand travel barriers. Where are patients going for their treatment, and can the Integrated Care Boards make this easier for them?	Analysis currently underway by NHSE to examine travel time via clinical datasets.

5	Work in collaboration to overcome barriers to accessing treatment such as poor experiences within A&E, staff understanding and awareness, fast tracking and ensuring care is given quicker, can we utilise innovation such as electronic patient records/apps.	In 2022, NHSE launched a dedicated programme to improve care for patients living with sickle cell disease. This includes roll out of an E- Learning module 'Addressing Inequalities in healthcare – Sickle Cell Disease'; pilot implementation of SCD hyperacute units and development of digital care plans. Thousands of people with sickle cell disease will benefit from quicker pain relief when experiencing a crisis thanks to new specialist NHS centres across the country. On World Sickle Cell Day, NHS England announced the creation of new expert clinics to provide specialist care when people need it most, allowing them to bypass A&E. Around 15,000 people in England have sickle cell disease. The condition can cause patients multiple periods of intense pain a year – known as a crisis – due to blood clotting in their arteries. The new 24/7 Hyper Acute Units will be set up in parts of the country with the highest number of sickle cell patients, initially London and Manchester, later this year. Around four in five people with sickle cell disease will receive specialist support from staff at the units, meaning they can avoid waiting in A&E, going straight to clinicians who understand their condition and can provide effective pain relief quicker. The new units are part of a raft of measures taken over the last year, including training for healthcare professionals on sickle cell symptoms, to address longstanding inequalities reported by patients in accessing the right care. NHS England and NHS Blood and Transplant announced a new partnership to provide genetic tests for patients with sickle cell and related conditions, enabling them to receive better-matched blood transfusions – decreasing the risk of side-effects. NHSE are working on a campaign to ensure those eligible for free prescriptions take up the offer to help them stay well at home, and steps towards a new digital care plan to ensure patients get the right care wherever they are.

	Challenge	Development
6	Tackle the complex issue of pain management, across the ambulance and healthcare settings; can we increase staff competence regarding sickle cell crisis pain?	NHSE ran a National Campaign – 'Can you tell it's sickle cell' for World Sickle Cell Day in 2022 and 2023. In June 2023 NHSE launched the Sickle Cell Patient Card for all patients living with sickle cell disorder. The card highlights a painful crisis as a medical emergency that should be managed according to NICE guidelines. On presentation at an ambulance or healthcare setting, the card will help a healthcare professional to swiftly pull up a patient's sickle cell crisis care plan and the patient will be given care according to their agreed protocol. Efforts are currently underway to digitize and make accessible the crisis care plans that every patient should have, particularly on interoperable digital platforms. The Haemoglobinopathy Coordinating Centres will continue to work with their Specialist Haemoglobinopathy Centres and Local Haemoglobinopathy Teams to improve staff competency in managing painful crisis. 'Can You Tell It's Sickle Cell resources' available here: https://campaignresources.dhsc.gov.uk/campaigns/can-you-tell-its-sickle-cell and the sickle cell e-learning module here: www.e-lfh.org.uk/programmes/health-inequalities
7	Combat the variation in experience; unpick where these challenges are and try and empower organisations to work across their boundaries to improve experience.	NHSE Specialised Commissioning team undertook a compliance exercise against the existing service specifications. This will inform a review of the national specifications to accurately reflect the requirements of effective, high-quality services for patients.

Experience

Trust

Pain

Information

Travel

Variation

Conclusions

Respondents were asked what were the positive elements of the care you received? (Response rate = 70% (107)

Key Theme	Total Responses	% of negative / neutral responses
Kindness and Empathy	31	28%
Medical care	30	27%
Staff knowledge of condition	15	14%
Communication	10	9%
General Condition	8	7%
Access to treatment	6	5%
Pain Management	6	5%
Wait for Pain Relief	3	3%
Transport	1	1%

When respondents were asked if anything could be improved, we received a response rate of 70% so 107 patients and/or their carers.

Key Theme	Total Responses	% of negative / neutral responses
General Condition	28	27%
Staff knowledge of condition	15	15%
Access to treatment	13	13%
Communication	13	13%
Kindness and Empathy	10	10%
Medical care	9	9%
Wait for Pain Relief	7	7%
Transport	4	4%
Availability of medical staff	2	2%
Pain Management	2	2%

These results clearly illustrate that there is a huge variation in experience from the patient's perspective. A view also presented within the No-Ones-Listening-Report, which, in its Executive Summary states that, 'Sickle cell patients too often receive sub-standard care, with significant variations in care depending on which staff happen to be on duty or which area of the country a patient is in'.

Just absorbing the information in the tables above shows that 28% of patients were shown kindness and empathy, whilst 10% were not. 27% of patients experienced good medical care whilst 9% did not. In order to ensure some more consistency in experience there is a need to identify those trusts that have more challenges. Perhaps in hospitals where sickle cell prevalence is lower, staff are not so well-versed in identifying and treating the condition quickly and effectively. Perhaps in some areas, clinical practice varies according to the time of the day and night that the patient arrives, whether haematology is open, and whether expert staff are on site and available to call.

These recommendations are being brought to the National Working Group on Spectra Optia to help inform efforts to improve patient access to automated red blood cell exchange. The working group will be reviewing patient travel time/distance as part of this work, as well as out of hours provision.

Role of the Haemoglobinopathy Coordinating Centres

Haemoglobinopathy Coordinating Centres (HCCs) are commissioned by NHS England to reduce levels of morbidity and mortality and to improve the experience of all haemoglobinopathy patients by reducing inequalities and improving timely access to high quality expert care. They are tasked with ensuring good governance which will improve both access to services and access to expertise. Also, to introduce good leadership to improve both the patient experience and outcomes. It is therefore essential that each HCC co-ordinates an in-depth governance review looking into their regional Specialist Haemoglobinopathy Teams and their Local Haemoglobinopathy Teams to determine where these shortfalls are and develop a plan for change. Reviews should consider patients' views, look at the care that they are receiving and indeed, whether they are travelling huge distances to access their treatment.

If possible, these reviews should consider the wider engagement within the Integrated Care Board networks and look at the interface with primary care, the ambulance services and social care, to try and create an opportunity to improve the entire clinical pathway from start to finish.

Where appropriate, HCCs can also engage with the regional Health Innovation Network to discover whether there are any opportunities within the MedTech Funding Mandate to apply for funding to expand existing services for automated red blood cell exchange.

Only by working with each centre, and by listening to the patients can we identify our challenges, and our opportunities for improvement. Only then, can we strive for a constant service for sickle cell patients in England.

Health Innovation
Yorkshire & Humber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