

NO ONE'S LISTENING:

AN INQUIRY INTO THE AVOIDABLE DEATHS
AND FAILURES OF CARE FOR SICKLE CELL
PATIENTS IN SECONDARY CARE



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ABOUT THE SCTAPPG

The All-Party Parliamentary Group on Sickle Cell and Thalassaemia (SCTAPPG) was formed in December 2008 with the aim of keeping sickle cell and thalassaemia on the political agenda and facilitating a two-way dialogue between policymakers and those affected by sickle cell and thalassaemia.

The SCTAPPG holds regular meetings and has published a number of policy reports on issues such as the institutional failures of Personal Independence Payment for those living with sickle cell and thalassaemia, the lack of representation of sickle cell and thalassaemia in the education of pre-registration nurses and midwives, the impact of prescription charges for those living with sickle cell and thalassaemia and the effect of the Covid-19 pandemic on the sickle cell community.



FOREWORD

The All-Party Parliamentary Group on Sickle Cell and Thalassaemia (SCTAPPG) exists to argue the case for more understanding of sickle cell and better treatment for those who live with the condition.

In the past we have produced reports on the treatment of people living with sickle cell in the fields of employment, NHS care and the benefits system.

This report was triggered by the Coroner's report into the death of Evan Nathan Smith in North Middlesex hospital. Evan was a young man with his whole life in front of him. The mistakes made in his treatment leading to his early and avoidable death brought into sharp focus the lack of understanding of sickle cell, the battles patients have to go through to get proper treatment and the terrible consequences which can come about as a result.

Following the publication of the Coroner's report earlier this year, the APPG held three evidence sessions, hearing from patients, clinicians and politicians. We took evidence from a wide range of witnesses and received over a hundred written submissions. We are profoundly grateful to all who contributed. This report is a result of that evidence.

The findings in this report reveal a pattern of many years of sub-standard care, stigmatisation and lack of prioritisation which have resulted in sickle cell patients losing trust in the healthcare system that is there to help them, feeling scared to access hospitals, expecting poor treatment from some of those who are supposed to care for them and fearing that it is only a matter of time until they encounter serious care failings.

Underneath the individual recommendations in the report are two more fundamental points. The first is a deep sense of anger and frustration that many of these failings have been pointed out in different

ways before but have not been properly acted upon, leaving people with sickle cell to go through the same enormously distressing experiences over and over again.

The second is the question of race. Sickle cell is a condition that predominantly affects black people. People of every race have a right to equality in health treatment. Yet the experience of people living with sickle cell is that the failings in treatment and the lack of understanding outlined in this report show deep inequality in the healthcare system. This is a serious and longstanding issue which must be addressed.

The publication of this report must lead to major change in the care sickle cell patients receive. We have made a number of recommendations based on the evidence we received and call on the Secretary of State for Health and Social Care, NHS England & NHS Improvement and the numerous other stakeholders we have directed recommendations at to prioritise taking action.

In the APPG we hope that the greater awareness of health inequalities following the pandemic results in urgent action to ensure sickle cell patients finally receive care at a standard to which they are entitled, and for which they have waited far too long.

Rt Hon Pat McFadden MP
Chair, All-Party Parliamentary Group
on Sickle Cell and Thalassaemia

EXECUTIVE SUMMARY

Our inquiry sought to examine the level of care sickle cell patients receive when accessing secondary care and to determine the action that is required to improve care for sickle cell patients.

While many patients we heard from were keen to highlight their gratitude to those healthcare professionals who go above and beyond in the care they provide, we heard that this level of care is sadly not the norm.

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. While care in specialist haemoglobinopathy services is generally felt to be of a good standard, this is far from the case on general wards or when accessing Accident & Emergency (A&E) departments. Care failings have led to patient deaths over decades and 'near misses' are not uncommon. There is routine failure to comply with national care standards or NICE standards around pain relief when patients attend A&E. Shockingly, this sub-standard care has led many patients to fear accessing secondary care, or even outright avoid attending hospitals.

A significant factor in the sub-standard care sickle cell patients often receive is a lack of effective joined-up care. The evidence we received highlighted that communication failings between different departments within the same hospital often impact sickle cell care. Patient care plans that have been specifically developed to ensure routine care are often ignored.

Community care for sickle cell patients is generally inadequate or non-existent which leads to unnecessary admissions to hospitals.

We were told that awareness of sickle cell among healthcare professionals is low, with sickle cell patients regularly having to educate healthcare professionals about the basics of their condition at times of significant pain and distress. We heard from patients and clinicians alike that this low awareness arises from inadequate training in the condition for trainee nurses and medics.

Partially as a result of the low levels of awareness and insufficient training in sickle cell, patients are regularly treated with disrespect, not believed or listened to, and not treated as a priority by healthcare professionals. Many of those we received evidence from highlighted the role of racism in the negative attitudes towards sickle cell patients, which overwhelmingly affects people with African or Caribbean heritage.

We also heard that there is inadequate investment in sickle cell care. Services are under-resourced and under-staffed and there has been a distinct lack of investment in sickle cell research and treatments over decades, right up to the present day.

Based on the evidence we received, the SCTAPPG makes the following recommendations, separated below by the section of our report in which they appear:

Sub-standard care on general wards and in A&E

- The North London Integrated Care System to develop a plan for improving sickle cell services, in partnership with relevant stakeholders, and share learnings with other ICSs across the country.
- Department of Health and Social Care to commission an evidence review by the Getting It Right First Time programme examining the case for and against implementing dedicated sickle cell wards at all specialist centres.
- North Middlesex University Hospital NHS Trust to engage with Betty & Charles Smith regarding an appropriate memorial tribute to their son Evan, such as the naming of a ward after Evan, in line with their wishes.
- NHS Trusts to share findings of all internal reviews into incidents involving serious sickle cell care failings with the National Haemoglobinopathy Panel so that learnings can be communicated to haemoglobinopathy teams across the country.
- Health Education England to develop an e-learning module based on the national standards of care developed by the Sickle Cell Society in partnership with clinical experts and the UK Forum on Haemoglobin Disorders, which should be mandatory for all healthcare professionals providing sickle cell care in high-prevalence areas.
- All NHS Trusts to develop an action plan setting out how they will ensure compliance with the NICE clinical guideline around the delivery of pain relief within 30 minutes for sickle cell patients, with appropriate advice from the NHS England Clinical Reference Group for Haemoglobinopathies pain sub-group.
- Care Quality Commission to adopt compliance with the NICE clinical guideline for delivery of pain relief within 30 minutes for sickle cell patients as essential criteria when assessing NHS Trusts.
- NICE to revise clinical guideline around pain relief for sickle cell patients to set out standards relating to pain management in the entirety of a sickle cell crisis, not just delivery of the first dose.
- Royal College of Emergency Medicine and Royal College of Physicians to develop guidance for staff working in A&E and on general wards making clear that sickle cell patients should be prioritised for treatment as a medical emergency due to the high risk of fast medical deterioration, to be distributed by NHS Trusts.
- Care Quality Commission to undertake a thematic review of sickle cell care in secondary care, involving direct input from patients and the Haemoglobin Disorders Peer Review Programme Clinical Leads, providing guidance around what good care should look like.
- National Haemoglobinopathy Panel to work with Haemoglobinopathy Coordinating Centres to plan equitable access to psychological support services for sickle cell patients who require such support.

Failings in providing joined-up sickle cell care

- All NHS Trusts to require that haematology teams are informed whenever a sickle cell patient accesses or is admitted to the hospital to ensure the patient's clinical history is known and advice can be passed on regarding their care, with compliance reported via the NHS England and NHS Improvement Specialised Services Quality Dashboards.
- NHS Trusts to develop individualised care plans for, and in partnership with, each sickle cell patient, with the patient and any relevant carers provided with a copy of the plan.
- National Haemoglobinopathy Register to develop capability to host sickle cell patient care plans that are accessible across the NHS.
- The Secretary of State for Health and Social Care to instruct all Integrated Care Systems to develop plans to provide community care for sickle cell patients in their area, including integration with third sector providers and community care organisations.

Low awareness of sickle cell among healthcare professionals and inadequate training

- All universities to include comprehensive training in sickle cell as part of curriculums for trainee healthcare professionals, covering diagnosis, presentations, management, acute complications (such as pain, acute chest syndrome, stroke) and ongoing care and featuring direct contributions from sickle cell patients.
- The Nursing and Midwifery Council and the General Medical Council to urgently commission a review of their approach to sickle cell training, in collaboration with the sickle cell community.
- The NMC and GMC to strengthen requirements around the level of sickle cell training required for university curriculums to be approved.
- Royal College of Pathologists to include as part of haematology speciality training a compulsory rotation to a large regional haemoglobinopathy centre for trainees in low incidence regions who would not otherwise have as much opportunity to gain direct experience of managing sickle cell patients.
- Health Education England to provide additional funding for sickle cell training programmes for healthcare professionals, including for training in the delivery of blood transfusions for non-specialist doctors.

Negative attitudes towards sickle cell patients

- Secretary of State for Health and Social Care to implement charge-free prescriptions for sickle cell patients.
- Health Education England, the Nursing and Midwifery Council, the General Medical Council, universities and other medical training providers to ensure training programmes address diversity and racial bias awareness.
- NHS Race and Health Observatory, working closely with Haemoglobinopathy Coordinating Centres, specialist haemoglobinopathy teams, community sickle cell teams, other professionals involved in care provision and the sickle cell community, to undertake a study into sickle cell care in relation to race and ethnicity, examining the impact of racist attitudes and the extent of inequalities in funding and prioritisation for sickle cell compared with other conditions.
- NHS England & NHS Improvement to require NHS Trusts to conduct and report regular audits of patient involvement in decisions about their care, utilising patient feedback, in line with NICE clinical guideline stating that sickle cell patients (and their carers) should be regarded as experts in their condition.
- NHS England & NHS Improvement to establish formal sickle cell patient advisory groups, based on consultation with the Patient and Public Voice Assurance Group, to work in partnership with and conduct oversight of NHS sickle cell services.

Inadequate investment in sickle cell care

- NHS England & NHS Improvement to provide increased funding for sickle cell services in recognition of the consistent underfunding of sickle cell services when compared with services for other conditions. This should include dedicated funding for NHS Trusts to improve apheresis capacity across the country.
- Clinical Commissioning Groups and local authorities to provide additional funding for third sector providers and community care organisations for social prescription in relation to sickle cell to reduce pressure on NHS services.
- Department of Health and Social Care to convene organisations including Health Education England, the General Medical Council, the Nursing and Midwifery Council, the medical royal colleges and medical and nursing schools to come together with senior sickle cell service representatives to engage in effective workforce planning for sickle cell services, including the allocation of specialist training opportunities.
- All NHS Trusts to ensure that specialised service funding is invested in meeting recommended sickle cell service staffing numbers.
- UK Research and Innovation and the National Institute for Health Research to launch dedicated sickle cell research opportunities, including supporting and funding research into genetic therapies to cure sickle cell disorder.
- NHS England & NHS Improvement to report results of Managed Access Programme for Crizanlizumab to support roll-out following the drug's approval.

SICKLE CELL IN SECONDARY CARE: NOT A PRIORITY?

In May 2021, the SCTAPPG launched an inquiry into the care sickle cell patients receive when accessing secondary care services in the UK. The inquiry followed a number of high-profile examples of failings in care for people with sickle cell disorder which contributed to growing awareness of the challenges sickle cell patients still often face in receiving appropriate care.

Among the most notable of these was the tragic death of sickle cell patient Evan Nathan Smith in North Middlesex University Hospital in April 2019, which received renewed focus following the publication of the coroner's inquest into Evan's death in April 2021. The inquest found that Evan's death would not have happened were it not for failures in the care he received. With healthcare professionals, sickle cell patients and their families having repeatedly highlighted similar incidents, including avoidable deaths and 'near misses', over many years, the SCTAPPG was determined to highlight the issues sickle cell patients face when accessing secondary care.

The inquiry, chaired by SCTAPPG Chair Rt Hon Pat McFadden MP, featured three oral evidence sessions held in June 2021, with SCTAPPG members receiving testimony from expert witnesses including sickle cell patients, patients' carers and family members, clinicians and representatives from relevant healthcare bodies. In addition, the SCTAPPG issued a call for written evidence which resulted in the receipt of over 100 submissions from key stakeholders.

Below, we explore the main themes that emerged from the evidence we received.

SUB-STANDARD CARE ON GENERAL WARDS AND IN A&E

One of the most consistent themes of the evidence we received was related to sickle cell patients receiving sub-standard care when admitted to general wards or attending Accident & Emergency (A&E) departments.

Variations in care: “It really is like a lottery”

Many patients felt that the quality of care they received was dependent upon factors such as which staff happened to be on duty. One told us that “if it is the staff who are familiar with me then the care is great, if the staff do not know me then it can be problematic”¹. Another stated that some staff “are exceptional – dedicated, committed and loved by patients”, but that “unfortunately, this is the exception rather than the norm.”²

The latter also noted that care varies “from hospital to hospital” and the geographical differences in sickle cell care was another strong theme of the evidence we received. Liz Blankson-Hemans, a sickle cell patient, told us: “The standard of care for sickle cell disease in the UK ranges from very good to extremely patchy depending on where you live in the UK.”³

Often, this was thought to be attributable to some areas having less ethnically diverse populations, and thus fewer sickle cell patients. One patient told us that: “Hospitals in areas without a significant ethnic minority population tend to know very little about [sickle cell] and treat you like some alien life form”⁴

A haematologist based in an area with few sickle cell patients said that “in consequence staff do not build up an experience base in management of sickle cell disease, in particular with acute complications requiring urgent review or admission.”⁵

However, others felt that geographical differences in care standards were apparent regardless of the patient population in the area. Liz Blankson-Hemans wrote that “even in ‘good’ areas... it can vary depending on which pockets you live in, such as for example, London compared to Hertfordshire, although they share boundaries and populations of African, Caribbean or South Asian and Mediterranean people”.⁶

Another patient told us: “I live just outside of the M25 and considering the prevalence of the disease in London ... I would have expected the care to be equally as good in my area both in primary and secondary care and it never fails to surprise me the lack of knowledge and help that the local healthcare staff have.”

1 Anonymous, written evidence

2 Anonymous, written evidence

3 Liz Blankson-Hemans, written evidence

4 Anonymous, written evidence

5 Anonymous, written evidence

6 Liz Blankson-Hemans, written evidence

The patient went on to state that he will soon be going to university and worries about the standard of care he will receive if he attends a university in an area where sickle cell is less prevalent, noting that it should be his right to receive “high quality care regardless of where I am and where I access the care”.⁷

Reflecting on his experience of living in many different areas of the country as a sickle cell patient, Shubby Osoba concluded: “It really is like a lottery with regards to the kind of care that you receive. Sometimes the care can be really good, and that normally is if you’re being seen by a team who knows you, who have an understanding of what sickle cell is, and in particular an understanding of you. I think one of the issues is that whilst you can get lucky and find someone that does know what sickle cell is, if you’re in the right part of the country, if you go into hospital at the right time of day, on a weekday, all of that can help, but if not, then the chances of meeting someone who even knows what sickle cell is can be slim.”⁸

Global Blood Therapeutics’ submission noted that geographical variation in care “is particularly important as the geography of [sickle cell] is starting to change with patients increasingly moving out of London – home to 25 out of 53 of the listed Sickle Cell Centres in the UK – to the wider South East and other urban areas ... All patients, regardless of where they live, must have equal access to the most effective care and support available.”⁹

Cedi Frederick, Chair of North Middlesex University Hospital NHS Trust, told us that the development of Integrated Care Systems present an opportunity for hospitals and other providers to work together to improve services for sickle cell patients and ensure a more consistent standard of service. While we welcome Mr Frederick’s assurance that sickle cell will be a focus of the North London Integrated Care System, it was disappointing that our invitation for a representative from the North London ICS to provide evidence to our inquiry was turned down and that there has been no subsequent contact from ICS representatives with the SCTAPPG or the Sickle Cell Society. Such lack of engagement does little to dispel the perception that sickle cell is not a priority for healthcare leaders.

The recent commissioning of Haemoglobinopathy Coordinating Centres and designation of specialist haemoglobinopathy services by NHS England & NHS Improvement are welcome steps towards addressing the levels of variation in sickle cell care but there remains much work to be done to achieve uniformly high-standard services. With all 42 Integrated Care Systems expected to be fully operational in England by April 2022, ICS leaders must ensure that progress continues to be made in the effective commissioning of sickle cell services.

Recommendation: The North London Integrated Care System to develop a plan for improving sickle cell services, in partnership with relevant stakeholders, and share learnings with other ICSs across the country.

7 Anonymous, written evidence

8 Shubby Osoba, oral evidence session, 9 June 2021

9 Global Blood Therapeutics, written evidence

Specialist v non-specialist variation: “Clinicians had not got a clue or the care was so poor it was negligent”

Patients and their relatives often emphasised the contrast between their positive experiences of care in haematology departments with the care received on general wards or in A&E. One relative of a sickle cell patient, for example, told us that the care provided to her husband on the haematology ward is “consistently of a high standard”, with staff who are “caring and are experts in their field. They understand the physical and emotional strains of the illness and are highly skilled, knowledgeable, and compassionate.” By contrast, the submission described repeated issues in A&E, including delays in receiving treatment, lack of awareness of sickle cell among staff and, “almost every time my husband presents at A&E”, having to “battle” for effective pain relief.¹⁰

Richard Patching described the care his wife Carol receives at her regular outpatient appointments at the haematology unit as “very good” but noted that the “problems arise” when she attends A&E or is admitted to a general ward.¹¹

June Okochi told us her experience of specialist services is “really positive”, noting that she has had “great relationships” with the haematology teams at every hospital she has been admitted to. However, June added: “Where I have found the quality of care to be very poor is Accident & Emergency. In those specific situations, there’s been a couple of near misses where my outcomes could have been more dire than they were. I would say the general care on

the wards as well can be quite poor, depending on what time of the day, what time of the week, your relationship with the nurses, etc.”¹²

Similarly, Zainab Garba-Sani said that her care on haematology wards tends to be “much better” than on a general ward “because at least the nurses know what they’re doing ... so you’re not having to educate people whilst you’re already in quite a vulnerable position”.¹³ Kye Gbangbola told us: “Some of my haematologists have said, ‘call me if you need me to speak to the hospital doctors’; those calls, I have no doubt, have saved my life several times over, both when clinicians had not got a clue or the care was so poor it was negligent.”¹⁴

The general consensus among patients and their carers that care is of a lower quality outside of haematology departments was supported by healthcare providers. One haematologist told us that “many hospitals have insufficient beds for patients with sickle disorders and as such they may be placed in non-haematology wards where, at best, their care needs are not fully met and, at worst, their condition may deteriorate.”¹⁵ Another healthcare professional noted that “patients have described difficult experiences of care when they present at hospital outside of the working hours of the [sickle cell] specialist care team.”¹⁶ Whittington Health NHS Trust acknowledged inpatient care as an area in need of improvement, following “significant feedback from patients that care has deteriorated since being transferred to a different ward”.¹⁷

¹⁰ Anonymous, written evidence

¹¹ Richard Patching, written evidence

¹² June Okochi, oral evidence session, 9 June 2021

¹³ Zainab Garba-Sani, oral evidence session, 30 June 2021

¹⁴ Kye Gbangbola, written evidence

¹⁵ Anonymous, written evidence

¹⁶ Anonymous, written evidence

¹⁷ Whittington Health NHS Trust, written evidence

Some felt the solution to the often-sub-standard care on general wards is to ensure sickle cell patients are always placed on haematology wards. A haematologist told us she felt she was “lucky to work in an environment where my colleagues understand and support the need for appropriate sickle cell inpatient management.

This is especially in the case of inpatient care where sickle cell patients are seen as haematology patients and prioritised for care on the haematology wards. I know that this does not always occur in other centres but I feel this helps to maintain good knowledge of the staff caring for patients when they are admitted.”¹⁸

Others argued that there should be dedicated wards for sickle cell patients. The UK Forum on Haemoglobin Disorders wrote: “Similar to other specialist conditions e.g. cardiac, renal etc., care of sickle patients in a dedicated ward where nursing staff are specially trained and acquire the knowledge, skills and competence to care for this patient group is essential. Specialist teams can provide good effective pain relief in a holistic, supportive setting allowing for the rapid reduction of stress and pain ... Conversely, being nursed on a general ward, without specialist knowledge, with a low patient to nurse ratio, often results in delayed pain relief, more pressure on the nursing teams and a more antagonistic environment.”¹⁹

Betty Smith, Evan Nathan Smith’s mother, told us that “sickle cell patients, particularly those with underlying conditions should not be moved around the hospital or placed in unsuitable wards with no access to oxygen or a nurse call bell. It would be ideal to have a dedicated sickle cell ward in hospitals”.²⁰

Specialist wards were also advocated on the basis that it would help to mitigate the risk of immunocompromised sickle cell patients picking up infections on general wards.²¹

Clinicians giving evidence to the inquiry also felt that dedicated sickle cell wards could be useful, albeit caveated with reservations about the potential implications of their introduction. Dr Arne de Kreuk, Consultant Haematologist at North Middlesex Hospital, told us that “a dedicated ward is something that all healthcare professionals have on their wish list, where you have your own team of nurses and doctors surrounding you. I think a dedicated ward would make a difference. However, it could also backfire, because sickle cell patients can have other problems, for example post-surgical problems”.²²

Similarly, Dr Emma Drasar, Consultant Haematologist at The Whittington Hospital and University College London Hospital, said: “We’d like to have, potentially, our own unit staffed by haematology specialists. The problem is that can lead to the level of knowledge within the rest of the Trust falling even further down and potentially prejudicial attitudes becoming more entrenched. So it’s very difficult. I think it’s a balance. I think having our own unit where we can give significant high-quality care, that’s the aim of all haematologists and nursing staff, people involved in looking after these people.”²³

The general consensus of the evidence we received is that sickle cell patients should either be treated on dedicated sickle cell wards or on specialist haematology wards. We believe it would be beneficial for the Department of Health and Social Care to commission an evidence review looking further into the case for and against implementing dedicated sickle cell wards.

18 Anonymous, written evidence

19 UK Forum on Haemoglobin Disorders, written evidence

20 Betty Smith, oral evidence session, 30 June 2021

21 Anonymous, written evidence; Anonymous, written evidence

22 Dr Arne de Kreuk, oral evidence session, 16 June 2021

23 Dr Emma Drasar, oral evidence session, 16 June 2021

Recommendation: Department of Health and Social Care to commission an evidence review by the Getting It Right First Time programme examining the case for and against implementing dedicated sickle cell wards at all specialist centres.



Deaths and ‘near misses’: “How many other people have ended up dying in the way that she did?”

We were told of a number of incidents in which failures in care resulted in patient deaths. Bell Ribeiro-Addy MP, a member of the SCTAPPG, told the inquiry of the death of her close friend Adjuah Annan, who died after being given an overdose of a morphine-based painkiller during a sickle cell crisis. Compounding the tragedy, this followed the deaths of “a few of her cousins [with] sickle cell [who] had all died before the age of 25”. There was no inquest into Adjuah’s death, leading Bell to ask: “how many other people have been through what [she] went through and ended up dying in the way that she did and were not... investigated?”²⁴

Betty and Charles Smith outlined a catalogue of failures that led to the avoidable death of their son Evan. These failings began with Evan’s treatment for gallstones, a condition more common among sickle cell patients, which involved Evan having a stent placed in his biliary duct and his gall bladder removed. Evan faced repeated delays in receiving appropriate treatment and there were numerous errors by medical staff, including failing to develop and share care plans and missing crucial medical developments that should have been identified.

Evan contracted sepsis and klebsiella during the procedure to have his stent removed and was admitted to North Middlesex University Hospital the following day. Here, there were again repeated failings, with the haematology team initially not being informed of Evan’s admission, despite the A&E medical staff having been informed that he had an underlying sickle cell condition.

Once informed of his admission, the haematology team declined to take lead responsibility for Evan,

meaning his care was led by the gastroenterology team, who were not specialists in his haematology condition. There were further treatment delays and oversights, which included the nurse responsible for Evan’s care failing to recognise that he was experiencing a sickle cell crisis, which “resulted in Evan having to call the ambulance from his bed to plead for oxygen, but it was refused because he was already in a hospital bed”. Doctors failed to escalate findings that confirmed low oxygen saturation levels and possible onset of a crisis and opportunities were missed to provide Evan with a blood transfusion that the coroner’s inquest found would have saved his life.²⁵

“Sometimes it feels like you’re living on borrowed time because you’ve been in those situations and you’re just lucky that you’re still alive to be able to tell the story”

– Zainab Garba-Sani, sickle cell patient

It is clear that what happened to Evan was an example of experiences that are far too common for sickle cell patients. In a stark illustration of the scale of the problem, results of a Coroner’s inquest into yet another avoidable death of a sickle cell patient arrived shortly before publication of this report. The inquest found that Tyrone Airey’s death from a morphine overdose during a sickle cell crisis in Northwick Park Hospital in March 2021

²⁴ Bell Ribeiro-Addy MP, oral evidence session, 30 June 2021

²⁵ Charles Smith, oral evidence session, 30 June 2021

was avoidable, with nursing staff having insufficient training to provide the care that would have prevented Tyrone's death.²⁶

Rather than the failings in Evan and Tyrone's care being isolated incidents, we were told of numerous 'near misses' experienced by sickle cell patients in which they could have had a worse outcome. Sadeh Graham told us that delayed treatment on a general ward and the absence of a haematologist led to his admission to an intensive care unit, remarking: "That admission, and others have been similar in terms of the neglect and inadequate care Evan Nathan Smith received ... [the] difference is that I escaped with my life."²⁷

As in Evan's case, Alex Luke described an incident in which the care he received on a general ward was so poor that he felt compelled to call an ambulance from his ward bed "because the doctors were very delayed to come to my rescue – for a few days, actually – and the pain was intensifying, and my mental health was going down the hill at that point. I was asking myself, what's the whole point of staying here, really?"²⁸

Most of the clinicians we heard from had experienced 'near misses' involving sickle cell patients, often involving failures during blood transfusions. These often arise due to poor communication or low awareness of sickle cell, we were told. A Paediatric Clinical Lead based in a haemoglobinopathy team said she had encountered "several" near misses, "usually relating to failure to identify potential seriousness of the situation or propensity to deteriorate rapidly".²⁹ Dr Emma Drasar told us she had seen "a significant number of near misses with my sickle cell patients during my career, the majority of which have been caused by single point of failure systems [where one failing part of a system causes the entire system to collapse, such as having an

overreliance on one consultant with a specialist interest in red cell conditions, whose absence causes problems] and poor education and understanding of sickle cell disorders in combination with a lack of resource".³⁰

Professor Jo Howard, Consultant Haematologist at Guy's and St Thomas' Hospital and Chair of the NHS England Haemoglobinopathies Clinical Reference Group, said that she has encountered "three or four" such incidents and that they "have always been used as a learning experience and led to review of services and service improvements". However, she added: "It is important that the same happens with the tragic case of Evan Nathan Smith. Unfortunately, the case and the lessons learnt have not yet been shared with the national haemoglobinopathy community and it is vital this is done with some speed."³¹

While findings from North Middlesex University Hospital NHS Trust's review have since been shared with the haemoglobinopathy community, it is concerning that it took so long for this to occur. The very least that should happen after serious incidents of the type outlined above is that lessons are learned and shared to avoid repetition.

Recommendation: North Middlesex University Hospital NHS Trust to engage with Betty & Charles Smith regarding an appropriate memorial tribute to their son Evan, such as the naming of a ward after Evan, in line with their wishes.

Recommendation: NHS Trusts to share findings of all internal reviews into incidents involving serious sickle cell care failings with the National Haemoglobinopathy Panel so that learnings can be communicated to haemoglobinopathy teams across the country.

26 MyLondon, Sickle cell sufferer, 46, left screaming in agony died after hospital neglect, <https://www.mylondon.news/news/sickle-cell-sufferer-singer-songwriter-21730986>. Accessed 8 October 2021.

27 Sadeh Graham, written evidence

28 Alex Luke, oral evidence session, 9 June 2021

29 Anonymous, written evidence

30 Dr Emma Drasar, written evidence

31 Professor Jo Howard, written evidence

Lack of compliance with national care standards: “It is life threatening!”

A significant factor in the sub-standard care sickle cell patients often receive in secondary care is the lack of adherence to national care standards, a source of frustration to patients and clinicians alike.

Sickle cell patient Liz Blankson-Heman asked: “Why is the standard of care so abysmal in some pockets despite [there being] a fully authoritative document written by experts in the field and applicable to the whole of the UK?” She added that, from her experience, it seemed that national care standards “may not be routinely used”.³²

Professor Jo Howard appeared at the same oral evidence session as Betty and Charles Smith and noted that a number of the failings in the care their son Evan received would not have occurred had national care standards been adhered to. For example, she said: “One of the national standards of care that we’ve produced in the document with the Sickle Cell Society a few years ago was that each specialist unit should have specialist guidance on looking after patients with sickle cell disease and having preoperative guidance is one of those things, so either there was guidance in place at the hospital and it wasn’t followed, or it wasn’t there.”³³

Likewise, Professor Howard added, “the national standards for sickle cell say that [for] any patient with sickle cell admitted to hospital, the haematology team should be informed, so the 48-hour delay initially in them even being informed [in Evan’s case] is pretty shocking and that’s something that would be outside standards of care. [Informing the haematology team] should have happened.”³⁴

“There are many examples of excellent guidelines about how to look after people with sickle cell disorders around the country. However, these are of no use if no one looks at them.”

Dr Emma Drasar, Consultant Haematologist,
The Whittington Hospital and University
College London Hospital and Chair,
Haemoglobinopathy Coordinating Centres

Written evidence from NICE referred to its clinical guidelines which state that “an acute painful sickle cell episode should be treated as an acute medical emergency ... and that analgesia should be offered within 30 minutes of presentation at hospital”.³⁵ In practice, standards around delivering pain relief are regularly not met.

Jaspreet Kaur told us that “overdue pain relief was the norm” during her friend’s admissions as an inpatient³⁶ and Stephanie George wrote that “90% of the time, I will receive pain relief between 45 minutes to over 60 minutes [after] attending A&E”.³⁷ Angela Thomas described waiting “in A&E for two to three hours while my pain got steadily worse until I was screaming out in pain”³⁸, while another patient referred to an incident in which they were left in “paralysing pain” for almost 24 hours, only to discover

32 Liz Blankson-Hemans, written evidence

33 Professor Jo Howard, oral evidence session, 30 June 2021

34 Ibid.

35 NICE, written evidence

36 Jaspreet Kaur, written evidence

37 Stephanie George, written evidence

38 Angela Thomas, written evidence

when a new doctor came on shift that the medication which was part of their care plan, and which they had been informed was not available, had been available all along.³⁹

Another patient wrote: “I have also seen some sickle cell patients wait so long for nurses to come with their pain relief, to the point where the patient was crying so much they could not breath properly because of the pain.”⁴⁰ Kye Gbangbola told us: “Every time I have been in hospital, I have constantly suffered more pain than necessary due to ward staff not responding to my medical needs”, including pain relief.⁴¹

Evidence from patients highlighted that lack of compliance with pain relief standards has been a persistent issue for years, rather than being a development related to recent pressures on the health service. The consensus of the evidence we received was that the failure to deliver pain relief within the time limits set out by NICE is often a result of low awareness of sickle cell among healthcare professionals and stigmatising attitudes that mean patients are not listened to or taken seriously. Both of these issues are explored further later in this report.

Clinicians also highlighted the lack of compliance with standards around delivering pain relief. University College London Hospital’s written evidence referred to an audit conducted in 2021 of compliance with NICE sickle cell pain management recommendations in A&E, which showed “very suboptimal adherence (30%)”.⁴² Dr Shivan Pancham, a Consultant Haematologist in the West Midlands area, told us that compliance with the NICE clinical guideline on pain relief in A&E in her NHS Trust is around 20%,

compared with over 90% in the haemoglobinopathy unit.⁴³ We were referred to a 2016 survey looking at experiences of pain relief among sickle cell patients, which found that only 30% of adults, 48% of children and 42% of parents felt that pain relief was provided to them in a timely manner in their most recent emergency healthcare episode”.⁴⁴

Dr Emma Drasar noted the frequent failure to deliver pain relief within 30 minutes and added that there are also “often delays with subsequent doses which again leads to poorly managed pain”. Dr Drasar suggested that this might require a change to the NICE clinical guidelines to focus not just on the timing of the first dose but on “overall pain control within the episode and requisite observations being performed”.⁴⁵

Patients and clinicians told us that sickle cell patients must be prioritised for treatment, in line with national care standards. One patient said that “once it is identified that the patient has sickle cell it should be escalated as their medical condition can deteriorate quickly into a life-threatening situation”.⁴⁶ Similarly, Betty Smith told us: “Patients with sickle cell condition should be prioritised as a matter of urgency particularly where deadlines and timescales for procedures are specified in patients’ records.”⁴⁷ Dr Shivan Pancham also noted that sickle cell patients in A&E “should automatically move into our priority line. The guidelines are there.”⁴⁸

Whittington Health NHS Trust told us that they will be developing and implementing a plan to increase compliance with the NICE guideline for patients to receive pain relief within 30 minutes⁴⁹

39 Anonymous, written evidence

40 Anonymous, written evidence

41 Kye Ggbangbola, written evidence

42 University College London Hospital, written evidence

43 Dr Shivan Pancham, oral evidence session, 9 June 2021

44 Dr Subarna Chakravorty, written evidence

45 Dr Emma Drasar, written evidence

46 Elizabeth Aiyedofe, written evidence

47 Betty Smith, oral evidence session, 30 June 2021

48 Dr Shivan Pancham, oral evidence session, 9 June 2021

49 Whittington Health NHS Trust, written evidence

and NHS England & NHS Improvement's submission stated: "Further work is underway to improve the management of acute pain (percentage of patients being given pain relief within half an hour of presentation, usually in A&E settings). In February the [NHS England Clinical Reference Group for Haemoglobinopathies] formed a multi-stakeholder pain subgroup to look at new ways of treating acute and chronic pain in [sickle cell disorder] and to improve education and research in this area."⁵⁰

It is evident that such work needs to be an absolute priority for the NHS, given the current widespread failures to comply with NICE guidelines or to meet the national standards of care developed by the Sickle Cell Society, in partnership with clinical experts and the UK Forum on Haemoglobin Disorders. As one patient put it to us: "It is life threatening! Delays to managing the pains leads to ... organ damage and death ... We are presenting at a hospital because we need help to make us better."⁵¹ Sickle cell patients are currently being failed by the system that should be providing them with this help and the consequences of these failings can be extremely serious.

Recommendation: Health Education England to develop an e-learning module based on the national standards of care developed by the Sickle Cell Society in partnership with clinical experts and the UK Forum on Haemoglobin Disorders, which should be mandatory for all healthcare professionals providing sickle cell care in high-prevalence areas.

Recommendation: All NHS Trusts to develop an action plan setting out how they will ensure compliance with the NICE clinical guideline around the delivery of pain relief within 30 minutes for sickle cell patients, with appropriate advice from the NHS England Clinical Reference Group for Haemoglobinopathies pain sub-group.

Recommendation: Care Quality Commission to adopt compliance with the NICE clinical guideline for delivery of pain relief within 30 minutes for sickle cell patients as essential criteria when assessing NHS Trusts.

Recommendation: NICE to revise clinical guideline around pain relief for sickle cell patients to set out standards relating to pain management in the entirety of a sickle cell crisis, not just delivery of the first dose.

Recommendation: Royal College of Emergency Medicine and Royal College of Physicians to develop guidance for staff working in A&E and on general wards making clear that sickle cell patients should be prioritised for treatment as a medical emergency due to the high risk of fast medical deterioration, to be distributed by NHS Trusts.

Fear and avoidance of hospitals: 'I do not trust the people who have sworn to protect us, because many times they have failed'

A large number of patients told us that their experiences of sub-standard care meant that they feared accessing secondary care, while others told us that they feel compelled to avoid attending hospital altogether.

50 NHS England & NHS Improvement, written evidence

51 Anonymous, written evidence

Patients described a “real reluctance” to attend hospital⁵², “apprehension and avoidance of hospital”⁵³ and feeling “traumatised and afraid to go into hospital”⁵⁴.

“Why should any one of us have to prepare ourselves mentally before we go into hospital?”

– Ifunanya Obi, sickle cell patient

Kye Gbangbola said that his experiences have left him with a sense that “it’s better to suffer at home, at least I will have some level of pain relief”.⁵⁵ Stephanie George wrote: “I have anxiety when I have to attend the hospital because I’m scared of the care I am about to receive. I do not trust the people who have sworn to protect us, because many times they have failed.”⁵⁶

Jaspreet Kaur told us her friend who has sickle cell “does everything she can to avoid a hospital admission, to avoid the mental strain of another battle with the doctors and nurses when she does not have the energy to advocate for herself” and that “delaying admission to hospital sometimes means that her clinical condition deteriorates rapidly as a consequence.”⁵⁷

Shubby Osoba said that his experiences of secondary care have been so poor that he saved up £3,000 for an oxygen machine and “would much rather try and care for myself ... as opposed to taking the gamble of going into hospital, potentially being sat in A&E for hours whilst someone tells you, ‘Have some paracetamol, have some ibuprofen.’”⁵⁸

Dr Emma Drasar noted that the difference between sickle cell and many other conditions is that sickle cell patients will continually have to access healthcare throughout their lives. Therefore, when patients have poor experiences: “They’re going to be afraid of going into a healthcare environment then, and they might stay at home longer when, perhaps, from a health perspective, that’s not what they should do, and not what I’d advise them to do as their consultant.”⁵⁹

The fact that so many sickle cell patients have had such poor experiences of secondary care that they avoid hospital altogether is an outrage. Such evidence demonstrates a deep failing in the care sickle cell patients receive.

Recommendation: Care Quality Commission to undertake a thematic review of sickle cell care in secondary care, involving direct input from patients and the Haemoglobin Disorders Peer Review Programme Clinical Leads, providing guidance around what good care should look like.

Recommendation: National Haemoglobinopathy Panel to work with Haemoglobinopathy Coordinating Centres to plan equitable access to psychological support services for sickle cell patients who require such support.

52 Claire T, written evidence

53 Anonymous, written evidence

54 Anonymous, written evidence

55 Kye Gbangbola, written evidence

56 Stephanie George, written evidence

57 Jaspreet Kaur, written evidence

58 Shubby Osoba, oral evidence session, 9 June 2021

59 Dr Emma Drasar, oral evidence session, 16 June 2021

FAILINGS IN PROVIDING JOINED-UP SICKLE CELL CARE

A significant factor in the sub-standard care sickle cell patients often receive is a lack of effective joined-up care. Such failings include poor communication between healthcare professionals within the same hospital, non-adherence to patient care plans and the lack of an appropriate level of community care for sickle cell patients.

Poor coordination within hospitals: “They blamed each other for what had happened to me when it was an obvious lack of communication”

The coordination of sickle cell care within hospitals was highlighted as a particular issue, with a consistent theme being the failure to alert haematology teams to the arrival of a sickle cell patient to another part of the hospital.

Araba Mensah noted that there is often no coordination with the haematology team when her daughter accesses other departments such as orthopaedics.⁶⁰ Others highlighted failures to alert the haematology team when accessing A&E, with a patient referring to “a lack of willingness to make contact with the relevant specialists to seek advice which resulted in severe prolonged pain and trauma”.⁶¹

Sickle Cell Suffolk wrote: “Once we are admitted to a ward we have to ask the ward [whether they] have advised the haematology department we have been admitted. The response is always ‘not yet’. It is our experience that the haematologist visits the patient on day three. This is not adequate [and] is purely because they have not been made aware.”⁶²

“Our haematologist should be informed immediately of our admissions. Not hours or days after but immediately!”

– Sickle cell patient

As referred to above, failures to coordinate care had a particularly tragic outcome for Evan Nathan Smith. Betty Smith told us there was no evidence from Evan's records that advice was sought from the sickle cell team prior to his stent removal procedure, despite the procedure placing Evan at an increased risk of sepsis.⁶³

Furthermore, Evan's father Charles set out the failure of A&E medical staff to alert the haematology team to Evan's presentation the following day,

60 Araba Mensah, written evidence

61 Claire T, written evidence

62 Sickle Cell Suffolk, written evidence

63 Betty Smith, oral evidence session, 30 June 2021

despite Evan having informed them that he had an underlying sickle cell condition: “It was later revealed that the haematology team were not informed until two days later of Evan’s admission and a series of missed opportunities and delays transpired over the following five days before his rapid deterioration and death”. Even once the haematology team had been made aware of Evan’s presence in the hospital, a failure to coordinate resulted in there being a lack of clarity as to which department had overall responsibility for Evan’s care, leading, Charles told us, to “substandard care” that led to Evan’s death.⁶⁴

As Betty Smith told us, Evan’s death was the result of “a lack of integrated and joined-up working within the medical teams caring for Evan. Medical teams should not work in silos when caring for sickle cell patients, rather in collaboration ... to optimise outcomes for patients.”⁶⁵

Another patient described experiencing severe pain while under the care of the rheumatology department and being refused a request to be seen by a haematologist “because I was under the rheumatologist’s care”. A CT scan revealed that the patient had had a stroke, at which point the haematologist took responsibility for their care. The patient concluded: “In my opinion this could have been prevented if they had just communicated with the rheumatologists about my sickle cell. By this time, it was far too late for the haematologists to act ... When both consultants came to talk to me, they blamed each other for what had happened to me when it was an obvious lack of communication.”⁶⁶

The transition from paediatric care to care as an adult for sickle cell was also highlighted repeatedly as an area of concern. One patient carer told us that the transition for her niece took place “without adequate preparation of what to expect or how different adult

care is. One minute the family is involved and can talk to doctors and the next minute [it’s], ‘Sorry, we can only talk to your niece and, whatever your concerns, we are sorry, but she is now an adult’, which is very unhelpful in an already complex situation.”⁶⁷

Araba Mensah described her and her daughter’s carers being halfway through singing happy birthday on her daughter’s 17th birthday when a porter arrived to take her to the adult ward: “We were not given any warning that she was going to be transferred to the adult ward and there was no preparation whatsoever. It was so abrupt and totally brutal.” Once on the adult ward, her daughter was regularly moved around and received very little interaction other than to be given her medicine: “To go directly, without any preparation, from the children’s ward where there are teachers, play specialists and one’s parent, to complete isolation on the adult ward was devastating. The situation was so horrendous that she felt abandoned, unwanted and uncared for to such an extent that she became severely depressed.”⁶⁸

“Better communication is needed between staff. You communicate with one staff member and they do not tell others or write it down, therefore we are always explaining things to different staff”

– Sickle Cell Suffolk patient group

64 Charles Smith, oral evidence session, 30 June 2021

65 Betty Smith, oral evidence session, 30 June 2021

66 Anonymous, written evidence

67 Anonymous, written evidence

68 Araba Mensah, written evidence

The transition from paediatric to adult care is a known problem. University College London Hospital noted that progress has been made in recent years but that a 2020 peer review of sickle cell services found that “many services still lack the robust processes needed to ensure a safe transfer of care to adult services”.⁶⁹ Dr Fatima Kagalwala, a Paediatric Haematology Lead, called for better national guidance on making the transition from paediatric to adult care, which suggests those working on the ground feel that they lack appropriate support to improve the situation.⁷⁰

Professor Jo Howard told us of concerted efforts within her Trust to improve the coordination of care, including providing joint clinics with renal physicians, orthopaedic doctors, neurologists, respiratory physicians, urologists, the pain-management team, obstetricians and cardiologists. A policy for

peri-operative management was developed with the anaesthetic team and the haematology team is informed of every patient with sickle disease who is having surgery, which results in a daily visit by the haematology team.⁷¹ The development of such multidisciplinary teams and procedures should be adopted by all NHS Trusts, with guidance from NHS England & NHS Improvement

Recommendation: All NHS Trusts to require that haematology teams are informed whenever a sickle cell patient accesses or is admitted to the hospital to ensure the patient’s clinical history is known and advice can be passed on regarding their care, with compliance reported via the NHS England and NHS Improvement Specialised Services Quality Dashboards.



69 University College London Hospital, written evidence

70 Dr Fatima Kagalwala, written evidence

71 Professor Jo Howard, written evidence

Failure to comply with patient care plans: “They said, ‘That care plan is not for this hospital.’ I was very shocked”

In theory, patient care plans exist to prevent the type of failings in joined-up care outlined above. However, we were told that sickle cell patients often have their care plans ignored or disregarded when accessing secondary care.

One patient told us that they had worked with consultants to agree a care plan but that other healthcare professionals, such as junior doctors, “decide to do something else”.⁷² Another wrote: “I have seen far too many human errors and mistakes that could have been well avoided if the nurses or doctors just took the time to read their patient’s notes or even talk to the patient and listen to them in order to get an understanding of the patient’s care plan.”⁷³

Following repeated incidents of poor care in A&E, one woman told us, she and her husband made a complaint to the hospital which led to the agreement of a protocol between the Consultant Haematologist and A&E Consultant. However, she told us, “it is rarely followed correctly and consistently ... One of the medical staff actually told my husband if he wouldn’t accept the alternative pain relief offered ... then he could not have anything – all in spite of his having an agreed protocol written by Consultants at the very same hospital.”⁷⁴

Richard Patching wrote of his wife Carol’s experiences: “From A&E, Carol is always transferred to the acute medical unit (AMU). Pre-Covid times, I would be with her for this transfer and I would have to tell the staff on AMU all about Carol’s care requirements. The hope then is that the staff will consult Carol’s care plan and that they will in any case have the basic knowledge of how to care for a sickle cell patient. In practice, Carol is always moved

onto another general medical ward and always in the middle of the night. Her care plan and all the advice I gave never go with her.”⁷⁵

Similar experiences of care plans being ignored were recounted in the oral evidence we heard. Alex Luke told us about being refused the pain relief medication he requested and asking the doctors to look at his care plan which outlined that he should be given it if in severe pain: “They said, ‘That care plan is not for this hospital.’ I was very shocked.”⁷⁶

Kye Gbangbola referred to having been given a letter by a doctor, “very much like [a] care plan”, to give to healthcare professionals if refused appropriate care. Nevertheless, “I’ve had healthcare workers ignore that letter. When they do this, the reason for it is, ‘You have to wait your turn.’”⁷⁷

It seems perverse that patient care plans specifically designed to ensure patients receive appropriate and consistent care are then ignored by healthcare professionals, often working in the same NHS Trust that developed the care plan. It is clear that a crucial part of improving care for sickle cell patients is greater adherence to patient care plans.

Recommendation: NHS Trusts to develop individualised care plans for, and in partnership with, each sickle cell patient, with the patient and any relevant carers provided with a copy of the plan.

Recommendation: National Haemoglobinopathy Register to develop capability to host sickle cell patient care plans that are accessible across the NHS.

72 Anonymous, written evidence

73 Anonymous, written evidence

74 Anonymous, written evidence

75 Richard Patching, written evidence

76 Alex Luke, oral evidence session, 9 June 2021

77 Kye Gbangbola, oral evidence session, 9 June 2021

Lack of community care: “Community care is deficient”

The lack of an appropriate level of community care for sickle cell patients is another example of failing to provide joined-up care, which adds to pressure on hospitals and fails sickle cell patients.

One haematologist told us that “community care is deficient”, with the lack of integration across health and social care systems contributing to the sub-standard care sickle cell patients receive. Where there are successful projects, they struggle to secure funding, they added, citing as an example “an excellent Sickle Cell Society pilot scheme providing practical domestic support to patients suffering pain and wishing to remain at home ... despite demonstrating the clear benefits of such an approach, no further funding was made available, with the project falling somewhere between health and social care.”⁷⁸

Professor Jo Howard described community nursing support for sickle cell as “very patchy”, with some areas having “excellent” support and others having none available.⁷⁹ Liz Blankson-Hemans wrote that community care for sickle cell is “completely randomised and not comprehensive”, even in areas of high-prevalence.⁸⁰

As noted by the Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee, by assisting with social needs, community services can “keep patients well and out of hospital”.⁸¹ Whittington Health NHS Trust told us it has been “very successful at reducing hospital admissions through our community offering” and plans to look into how this can be expanded “to help patients better manage their condition at home and therefore reduce A&E attendances and hospital admissions”.⁸²

This clearly also benefits patients, as noted by Dr Rachel Kesse-Adu: “Sickle patients do not want to have their lives interrupted by hospital admission so bolstering our community services and listening to patient and clinician groups to focus on what keeps our patients well at school, home and work, and what supports in the community will allow this, is fundamental.”⁸³

NHS Blood and Transplant suggested that the development of Integrated Care Systems “should help NHS providers and other key stakeholders to work across organisational boundaries and deliver improved access to treatment. The ICSs should focus on reducing the existing bureaucracy around contracting and funding between organisations that currently acts as a major barrier to access for patients across the NHS.”⁸⁴

Given ICSs have been explicitly designed to bring services together and ensure better joined-up care, we agree that the development of ICSs offers an excellent opportunity for a renewed approach to the delivery of community care for sickle cell patients which will ensure joined-up care, reduce pressure on hospitals and improve patient experience.

Recommendation: The Secretary of State for Health and Social Care to instruct all Integrated Care Systems to develop plans to provide community care for sickle cell patients in their area.

78 Anonymous, written evidence

79 Professor Jo Howard, written evidence

80 Liz Blankson-Hemans, written evidence

81 Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee, written evidence

82 Whittington Health NHS Trust, written evidence

83 Dr Rachel Kesse-Adu, written evidence

84 NHS Blood and Transplant, written evidence



LOW AWARENESS OF SICKLE CELL AMONG HEALTHCARE PROFESSIONALS AND INADEQUATE TRAINING

Low levels of awareness of sickle cell among healthcare professionals is another significant factor in the sub-standard care sickle cell patients receive in secondary care. These low levels of awareness are a result of inadequate training around sickle cell for healthcare professionals and trainee nurses and medics.

Lack of awareness of sickle cell: “I am teaching them more than they are doing the job at hand”

Almost all of the evidence we received during the inquiry referred to low levels of awareness of sickle cell among healthcare workers on general wards and in A&E.

A representative assessment of the situation came from Denise Owusu-Ansah, who wrote: “In my experience, the poorer quality care I have received has primarily been due to a lack of knowledge and/or experience of my condition on the part of the healthcare professional. There appears to be a very superficial level of knowledge of the condition and little if any understanding of the degree of pain that can be caused by a sickle cell crisis, the range of symptoms a sickle cell patient can experience and the very basic first steps that should be taken in the event of a sickle cell crisis.”⁸⁵ Patients contrasted

the low awareness of sickle cell among healthcare professionals with other similar conditions such as cystic fibrosis.⁸⁶

One patient told us: “I have been hospitalised in wards where the doctors have asked me, “what do we do for you, I have no idea at all?”.⁸⁷ Another referred to experiences of presenting to A&E and lack of awareness of sickle cell among the healthcare workers leading to “a lot of delay and Googling/ discussing with [a] colleague”⁸⁸, an understandable cause for alarm.

We heard from a patient who referred to seeing approximately five different members of staff in A&E and “it became obvious none of them knew what I was talking about and didn’t know what to do, which they admitted”⁸⁹. A number of different patients testified to having been asked how long they had

85 Denise Owusu-Ansah, written evidence

86 Carol Burt, written evidence; Anonymous, written evidence; Kye Gbangbola, written evidence

87 Anonymous, written evidence

88 Anonymous, written evidence

89 Anonymous, written evidence

had sickle cell or when they ‘caught it’ by healthcare professionals who evidently did not understand that the condition is present from birth.⁹⁰

Araba Mensah highlighted the consequences for her daughter’s care of the low levels of awareness of sickle cell among healthcare professionals, which included not being administered oxygen or blood transfusions at the correct time, failure to deliver pain relief and missing associated conditions because of a lack of understanding that they could be linked to her sickle cell disorder.⁹¹

“It should be as shocking for a senior trained medical staff [member] to say they have never heard of sickle cell disorder as it would be for them to say they had never heard of cystic fibrosis or diabetes.”

– Liz Bankson-Hemans, sickle cell patient

Patients reported that, due to the low levels of awareness among the healthcare workers they encounter, they feel that they have to educate staff themselves. The mother of a sickle cell patient told us that “sickle cell patients and relatives are forced to be their health advocates as knowledge of the condition is sparse”⁹², while one patient wrote that due to the lack of specialist nurses on the ward he accesses, “I find that I am teaching them more than they are doing the job at hand”⁹³. While many patients value being able to advocate on their own behalf and

rightly consider themselves to be the expert on their own condition, there is a world of difference between a patient having the opportunity to contribute to decisions around their care with an informed expert and feeling forced to explain basic information about their condition during a time of significant pain and distress.

Again, some felt that geographical differences were apparent in the levels of awareness of sickle cell. One patient told us that “... outside of London, in my experience medical staff do not have an understanding of what sickle cell is or how to manage it”. Their submission went on to explain that they had been admitted to hospital while away at university but the seriousness of their condition was only understood when they switched their care to a hospital in London, where they were admitted to intensive care and informed by their doctor that if they had stayed at the original hospital, “I would have died, as my crisis was very life threatening and was not being taken seriously”.⁹⁴ NHS Blood and Transplant’s submission noted that, among those providing care, there is “less expertise where hospitals see fewer patients”.⁹⁵

Another patient told us that staff turnover was a factor: “We are only seen by our main consultants occasionally and treated by junior doctors with minimal knowledge about sickle cell. The high turnover rate of these junior doctors has an impact on our care.”⁹⁶

Evidence from the Haemoglobin Disorders Peer Review Programme Clinical Leads also highlighted their findings around low levels of awareness of sickle cell, leading to poor care: “Urgent care of patients in non-specialised settings were fraught with poor experience of care. Most patients pointed to the knowledge deficit among emergency department

90 Sadeh Graham, written evidence; Vanessa Williams, written evidence

91 Araba Mensah, written evidence

92 Anonymous, written evidence

93 Anonymous, written evidence

94 Amanda, written evidence

95 NHS Blood and Transplant, written evidence

96 Anonymous, written evidence

(ED) and general practice staff in management of [sickle cell] and frequently expressed stigmatisation and allegations of drug-seeking behaviour.”⁹⁷

Referring to the same peer review, University College London Hospital’s submission noted: “There was suboptimal awareness and expertise amongst nursing staff in relation to this disease especially in the non-specialist centres ... This was indeed reflected in the feedback from some patients during the recent peer review, emphasising the lack of knowledge of some ward staff about sickle cell disease when they were admitted in an emergency to the ED and to general wards.”⁹⁸

The Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee cited low staff awareness as a factor behind adverse events related to inappropriate transfusion: “Due to lack of staff awareness, patients with sickle cell may be transfused inappropriately or with blood not

meeting specific requirements.” Their submission referred us to data reported to the Serious Hazards of Transfusion (SHOT) UK haemovigilance scheme between 2010 and 2019, which showed that 2.8% of all Specific Requirement Not Met errors occurred in patients with sickle cell disorder.⁹⁹

Generally, in contrast to on general wards and in A&E, patients felt satisfied that sickle cell is well understood by haematology teams. However, this was not uniform. Sickle Cell Suffolk told us that in their experience of a local hospital, “the haematology staff do not have enough knowledge on sickle cell and are not able to advise the medical staff adequately”, citing an incident where one of their members was on a general ward and “the haematologist asked the nursing staff why she was on a fluid drip ... Given this is a basic need for a sickle cell patient, it did not fill the patient with confidence about her care.”¹⁰⁰

Inadequate training: “The negative impact of this on patients’ care cannot be overstated”

The clear consensus from those who provided evidence to our inquiry was that the low levels of awareness of sickle cell among healthcare professionals is a result of inadequate training in the condition.

Carol Burt told us that during her training as a nurse in the 1980s, she recalled receiving training material with “less than six lines” on sickle cell “compared with pages and reams of literature for cystic fibrosis. In reality, I have nursed three people with cystic

fibrosis in the whole of my career and can’t mention how many individuals with sickle cell disease”.¹⁰¹ Similarly, Stephanie George said that she had a single one-hour session on sickle cell during her midwifery training, and concluded: “how are staff going to know about [sickle cell] when the teaching itself is substandard?”¹⁰²

97 Haemoglobin Disorders Peer Review Programme Clinical Leads, written evidence

98 University College London Hospital, written evidence

99 Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee, written evidence

100 Sickle Cell Suffolk, written evidence

101 Carol Burt, written evidence

102 Stephanie George, written evidence

“We are concerned why patients with sickle cell conditions should be nursed on wards where nurses are not fully trained to understand the complexities of this condition and respond appropriately. This, to us, is evidence of substandard care.”

– Betty Smith, mother of Evan Nathan Smith

One Consultant Haematologist felt that medical undergraduate and postgraduate training does a good job of including training around sickle cell alongside other aspects of haematology but that “[t]his is not the case across all areas of the medical profession ... there is a lack of training within nursing studies, especially [on] recognising the long-term health implications of the condition”.¹⁰³

However, other haematologists felt even this was too positive an assessment of the level of sickle cell training. One told us that most A&E departments “are staffed by clinicians (doctors and nurses) who have little training or awareness of [sickle cell]”.¹⁰⁴ Dr Emma Drasar wrote: “... education about sickle cell disorders is extremely patchy ... Even when it is included it is given comparatively little time on the curriculum ... Outside of haematology e.g. in general or speciality medicine the situation is significantly worse and people can become consultants having never been taught about sickle cell disorder or

having had very limited education and clinical experience. Similar issues occur in other allied healthcare professional groups including nursing.”¹⁰⁵

Professor Jo Howard told us: “In the nursing training, there is no set educational information about sickle cell disease so you can complete your nursing training with very little information about sickle cell disease, and every nurse should have that. Likewise, the training for medics on sickle cell disease is very, very poor so both those things should be improved. Anyone who’s likely to look after patients with sickle cell disease, so any general medical staff, should all have additional education. My personal thought is that should be mandatory and it’s not ... It wouldn’t be that difficult, it would need some money and some time to develop some national training that everyone had to undergo so at least they had some kind of basic understanding of sickle cell and when it was important.”¹⁰⁶

University College London Hospital said that the “welcome” recent restructuring of haemoglobinopathy provision needs to be accompanied by “major investment in staff and training ... healthcare providers in other interconnecting specialties such as A&E and intensive care need targeted and funded retraining, so that prejudicial assumptions that often exist about the genuine needs of patients and therapeutic options available to sickle patients do not harm patients either physically or psychologically.”¹⁰⁷

Responding to the widespread concern around the level of training around sickle cell for nurses, Dr Geraldine Walters from the Nursing and Midwifery Council (NMC) explained to us that the NMC assesses nurses against “high-level, outcome-focused standards” rather than listing specific conditions in the regulatory standards. The NMC is responsible for assessing and approving university curriculums, however, and Dr Walters told us that “the

103 Anonymous, written evidence

104 Dr Subarna Chakravorty, written evidence

105 Dr Emma Drasar, written evidence

106 Professor Jo Howard, oral evidence session, 30 June 2021

107 University College London Hospital, written evidence

curriculum-level can focus on specific diseases within [those high-level standards]”, with NMC’s “visitors check[ing] with the university whether the right components have been included.¹⁰⁸

It is clear to us from the evidence we received, including from the NMC, that change is needed in the way nurses are trained to ensure sickle cell patients receive the care they deserve. While we understand that the NMC’s approach to regulatory standards is based on high-level standards, rather than requirements around specific conditions, it is apparent from the evidence that too often these high-level standards are not being met.

Dr Walters told us the high-level standards include “things like patient assessment, cultural competence, understanding of tests and investigations, person-centred care, diversity”, as well as pain management and that “you need to listen to the patient. You need to have respect for them. You need to be aware of any issues which might impact the way you treat them, and you need to know the boundaries of your own competence.”¹⁰⁹ However, as will be outlined further in the next section of the report, sickle cell patients often do not feel they receive care in a “person-centred” manner, nor with respect to diversity or appreciation of cultural differences. Sickle cell patients often do not receive assessments in an appropriate manner when, for example, attending A&E and, as explored above, far too frequently do not receive appropriate pain management.

“Every single person in healthcare knows that if your face droops, you have to call an ambulance because of a stroke. Everyone knows that if you’ve got pain on your chest that radiates into the left arm, every second matters. I think we need to get out there that sickle cell presents with X, Y and Z; it’s a similar medical emergency.”

– Dr Arne de Kreuk – Consultant Haematologist, North Middlesex Hospital and Deputy Lead, North London Haemoglobinopathy Centre

Furthermore, even if accepting that the appropriate level for ensuring nurse training focuses on specific conditions is at university curriculum-level, it seems apparent that the current curriculums are not sufficient to ensure nurses have an appropriate level of knowledge of sickle cell, given the overwhelming consensus of the evidence we received was that nurses still too often have low awareness of sickle cell. It was welcome that Dr Walters acknowledged that “there might be other ways that we can strengthen our quality assurance around what goes into the curriculum”¹¹⁰, and we recommend that the NMC prioritises reassessing its requirements around the level of training in sickle cell required to ensure university curriculums are passed as meeting the NMC’s standards.

108 Dr Geraldine Walters, oral evidence, 16 June 2021

109 Ibid.

110 Ibid.

This is particularly important given the concerns we heard about the regional variations in ‘on the job’ training nurses and other healthcare professionals are exposed to around sickle cell. Dr Walters told us that: “Half of the hours of training are spent in clinical practice; half are in the university. So we know that there are some people who qualify who might have had quite a lot of exposure to sickle cell and thalassaemia. Others will have had relatively little.”¹¹¹

Haematologists also expressed concern at the regional variations in gaining experience around sickle cell during training. Professor Jo Howard told us: “It is very easy for nurses and doctors (particularly in low prevalence areas) to complete their training without learning about [sickle cell] and without ever seeing a patient with [sickle cell disorder]”. Noting that haematology trainees outside London “may not receive adequate hands-on training”, Professor Howard advocated “a short period in a sickle centre” as part of training activity.¹¹²

A second haematologist agreed, writing: “I trained in Haematology in the East of England, which has historically always had only a small number of patients with sickle cell disease. No formal training opportunity existed to go on secondment to a larger city centre (e.g. in London) to gain experience in management in a regional centre. Most of the training offered is via courses ... rather than with actual patient care. Training is not adequate in the low frequency regions, and specialist training for haematology speciality training should include a compulsory rotation to a large regional haemoglobinopathy centre for trainees in low incidence regions who would not otherwise gain much experience.”¹¹³

Another haematologist felt that there is good training in haemoglobinopathy in London and the south-east but that “for trainees outside large urban centres with smaller population this can be a bit patchy”, adding:

“For nursing and medical student training, very little time is spent in haematology as a whole, and even less so in haemoglobinopathy, and hence sickle patient management ... The negative impact of this on patients’ care cannot be overstated”.¹¹⁴

A number of healthcare professionals and providers referred us to existing or planned training around sickle cell. Dr Arne de Kreuk told us that he would like to see more use of “drills and simulations of what can happen”, which he uses in his own teaching, telling us: “I always start with two or three cases that start similarly and end very differently. I challenge the students and doctors and nurses, ‘Okay, what would you do? What would your management plan be here?’ I think a very practical, hands-on, maybe with modern technology, simulation module where you can actually see what happens, would be very vital.”¹¹⁵ University Hospitals of Leicester NHS Trust also highlighted the use of simulation training for junior doctors in its hospitals.¹¹⁶

The National Haemoglobinopathy Panel (NHP)’s submission referred to its monthly multidisciplinary team (MDT) meeting for clinical specialists as a key educational opportunity for those involved in patient care, including non-members such as senior consultants, nurses, psychologists and trainee doctors attending as observers. In addition to holding webinars and seminars on specific areas of sickle cell care, the NHP’s future plans include establishing a repository of complex cases that could be accessed by clinicians for learning, as well as analysing and sharing lessons from the first twelve months of NHP MDTs.¹¹⁷

We also received examples of good practice in the delivery of training around sickle cell from individual hospitals. This included regular training in sickle cell for non-specialist staff at Evelina London Children’s Hospital and Guy’s and St. Thomas’ NHS Foundation

111 Ibid.

112 Professor Jo Howard, written evidence

113 Anonymous, written evidence

114 Dr Rachel Kesse-Adu, written evidence

115 Dr Arne de Kreuk, oral evidence session, 16 June 2021

116 University Hospitals of Leicester NHS Trust, written evidence

117 National Haemoglobinopathy Panel, written evidence; Professor Baba Inusa, oral evidence session, 16 June 2021

Trust.¹¹⁸ The West London Haemoglobinopathy Coordinating Centre noted that a pilot initiative embedding appropriate learning on sickle cell in the nursing and medical curriculum at Imperial College London “has received positive feedback from students”.¹¹⁹

The UK Forum on Haemoglobin Disorders and the Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee referred us to educational opportunities they provide and efforts they have made to expand such training to non-specialist healthcare professionals.¹²⁰ However, the latter told us “we could do more with more targeted training days for specific groups of healthcare professionals.”¹²¹

Similarly, NHS Blood and Transplant informed us that it could, “if so commissioned and funded appropriately, provide nationwide teaching on transfusion in haemoglobin disorders to all staff groups ... We want to increase the audience of our courses to healthcare professionals in training, not just haematology trainees to transfusion requirements and haemoglobinopathies, as it is usually not specialist doctors that initially see haemoglobinopathy patients when they are acutely unwell, and they may have little awareness of appropriate management.”¹²²

We welcome the examples cited to us of existing training around sickle cell and planned or potential future training. We hope to see continued development and sharing of best practice in training provision around sickle cell from individual hospitals and healthcare bodies. Nevertheless, despite these specific examples of good practice, it is clear from our inquiry that nothing less than a fundamental step change is needed in relation to training for healthcare professionals around sickle cell. Much existing training, while certainly useful and welcome, does

not reach those who are most in need of it because it relies on healthcare professionals choosing to undertake it or having the time in which to do so. Comprehensive pre-qualification training in sickle cell for all healthcare professionals, alongside retraining for existing healthcare professionals is, therefore, essential.

Recommendation: All universities to include comprehensive training in sickle cell as part of curriculums for trainee healthcare professionals, covering diagnosis, presentations, management, acute complications (such as pain, acute chest syndrome, stroke) and ongoing care and featuring direct contributions from sickle cell patients.

Recommendation: The Nursing and Midwifery Council and the General Medical Council to urgently commission a review of their approach to sickle cell training, in collaboration with the sickle cell community.

Recommendation: The NMC and GMC to strengthen requirements around the level of sickle cell training required for university curriculums to be approved.

Recommendation: Royal College of Pathologists to include as part of haematology speciality training a compulsory rotation to a large regional haemoglobinopathy centre for trainees in low incidence regions who would not otherwise have as much opportunity to gain direct experience of managing sickle cell patients.

Recommendation: Health Education England to provide additional funding for sickle cell training programmes for healthcare professionals, including for training in the delivery of blood transfusions for non-specialist doctors.

118 Evelina London Children’s Hospital, written evidence and Guy’s and St. Thomas’ NHS Foundation Trust adult haematology service, written evidence

119 West London Haemoglobinopathy Coordinating Centre

120 UK Forum on Haemoglobin Disorders, written evidence and Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee, written evidence

121 Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee, written evidence

122 NHS Blood and Transplant, written evidence



NEGATIVE ATTITUDES TOWARDS SICKLE CELL PATIENTS

Partially as a result of the low levels of awareness and insufficient training in sickle cell, patients are frequently subject to prejudicial attitudes, treated with a lack of respect or prioritisation and undermined or disbelieved when accessing secondary care. The weight of the evidence suggests that such negative attitudes towards sickle cell patients are also often underpinned by racism.

Racial inequality as a factor in sickle cell care: “Care is clouded by stereotypical perceptions of black people”

With sickle cell disorder primarily affecting people with African or Caribbean heritage, racism was regarded by many to be a key factor in the sub-standard care sickle cell patients often receive.

Some patients shared with us examples of particularly overt racism. Calvin Campbell told us he has “had to deal with doctors and nurses openly being racist towards me and others ... I’ve been called the ‘n’ word to my face and much worse”.¹²³

Another patient told us she had experienced “nurses who would witness you being racially abused and still treat you as the instigator or just assume before even getting the facts. I have witnessed patients being racially harassed by other patients and then the nurses would be rushing to placate the instigator rather than the victim”.¹²⁴

We were told of an occasion when a consultant told a patient that “the care I was receiving was much better than the care I would have received if it was in my parents’ country (in West Africa). She cannot compare the UK to Africa. I was born here so I should surely get the right treatment.”¹²⁵

“Don’t look at the colour of our skin first, look in our face and see the pain and help us.”

– Diane Crawford, sickle cell patient

Alex Luke described an incident in which he experienced a sickle cell crisis on the motorway and had to call an ambulance. When the ambulance arrived, he was asked to provide identification, an experience he ascribed to racist prejudice.¹²⁶

Patients told us that racist attitudes often affect healthcare professionals’ perceptions of sickle cell patients, for example in the frequent assumption that they are ‘drug-seekers’. Diane Crawford said that: “As sickle is mainly a black illness, they jump to the conclusion that we’re all ‘junkies’ and not in pain at all ... If we were cancer patients it would be totally different, they have high doses of morphine, no questions asked and extra if they need it because they are mainly white people.”¹²⁷

Similarly, June Okochi told us: “I definitely feel that race does play a significant role in how patients are treated, especially in A&E. I think there is the misconception that the drug-seeking patients are back here again”.¹²⁸ Dr Arne de Kreuk echoed this, telling us: “I do strongly feel that [racism] is a problem on the wards, in A&E and even among doctors. There are publications about this, that illustrate that the perception is that sickle cell patients are difficult, are after painkillers. That perception is still out there and is, I think, deeply rooted, possibly even in training programmes. That perception is something we come across a lot.”¹²⁹

Bell Ribeiro-Addy was among many to point to research “that [shows] people believed that black people experience less pain, and because they

123 Calvin Campbell, written evidence

124 Anonymous, written evidence

125 Anonymous, written evidence

126 Alex Luke, oral evidence session, 9 June 2021

127 Diane Crawford, written evidence

128 June Okochi, oral evidence session, 9 June 2021

129 Dr Arne de Kreuk, oral evidence session, 16 June 2021

believe they experience less pain, a lot of the time they're having to beg for pain killers and that creates a massive issue."¹³⁰

Kye Gbangbola referred to evidence from a study in the USA showing that doctors denied pain relief to black sickle cell patients based on their belief that black people have higher pain thresholds or are opioid addicts, yet research in the same journal showed people with sickle cell disorder display lower levels of addiction than the general population.¹³¹

Many cited the lack of prioritisation of sickle cell compared to other conditions, as outlined above, as being the result of racial inequalities. Stephanie George stated: "I do believe that if sickle cell predominately affected people who are not from African or Caribbean origins, then the care would be completely different ... If you compare [sickle cell] to cystic fibrosis, the difference of care and awareness is staggering. Cystic fibrosis affects fewer people in the UK than [sickle cell] but research has shown the level of awareness and funding for [cystic fibrosis] is much higher."¹³²

Araba Mensah told us that "care is clouded by stereotypical perceptions of black people", noting that, while it is sometimes said that sickle cell is not prioritised because it is an 'invisible condition', "there are other "non-visible" conditions that are treated positively. For example, there is a huge disparity between care for patients with sickle cell and care for other blood disorders like leukaemia. Unlike sickle cell, leukaemia patients are treated with dignity, empathy, compassion and sympathy."¹³³

Zainab Garba-Sani referred to the fact that hydroxyurea, until recently the only licensed treatment for sickle cell in the UK, is "free for cancer

patients and it's not free for sickle cell patients", which is "probably a chief indication of institutional racism".¹³⁴

A number of submissions argued that the very fact that there are few treatments and low levels of research into sickle cell is an example of racism. Araba Mensah wrote: "The illness has been marginalised and kept out of the mainstream and not seen as deserving or warranting research into treatments because it affects blacks and there is no money to be made in it."¹³⁵

Clinicians contrasted the level of funding and resourcing for sickle cell services with that available to conditions that primarily affect those of a Caucasian background. A haemoglobinopathy clinician wrote: "Compared to other inherited conditions, many of which tend to affect Caucasian populations e.g. cystic fibrosis and haemophilia, [sickle cell] is woefully under resourced in the UK."¹³⁶ Dr Emma Drasar made the same point: "... despite the recent changes by NHS England there is massive and chronic funding disparity and under-resourcing compared to similar genetic disorders e.g. cystic fibrosis and haemophilia which predominately impact Caucasians."¹³⁷

Professor Jo Howard told us that the UK Forum on Haemoglobin Disorders has run "very effective" anti-racism teaching.¹³⁸ We agree with the suggestion that this type of training needs to be expanded and incorporated as an essential element of training for all healthcare professionals.

Recommendation: Secretary of State for Health and Social Care to implement charge-free prescriptions for sickle cell patients.

130 Bell Ribeiro-Addy MP, oral evidence session, 30 June 2021

131 Kye Gbangbola, oral evidence session, 9 June 2021

132 Stephanie George, written evidence

133 Araba Mensah, written evidence

134 Zainab Garba-Sani, oral evidence, 30 June 2021

135 Araba Mensah, written evidence

136 Anonymous, written evidence

137 Dr Emma Drasar, written evidence

138 Professor Jo Howard, written evidence

Recommendation: Health Education England, the Nursing and Midwifery Council, the General Medical Council, universities and other medical training providers to ensure training programmes address diversity and racial bias awareness.

Recommendation: NHS Race and Health Observatory, working closely with

Haemoglobinopathy Coordinating Centres, specialist haemoglobinopathy teams, community sickle cell teams, other professionals involved in care provision and the sickle cell community, to undertake a study into sickle cell care in relation to race and ethnicity, examining the impact of racist attitudes and the extent of inequalities in funding and prioritisation for sickle cell compared with other conditions.

Disrespectful treatment: “No sympathy, no compassion, no empathy”

Patients and carers reported frequent disrespectful treatment from healthcare professionals. Araba Mensah, whose daughter has sickle cell disorder, provided a stark summary of her experience of the local hospital: “Staff are unfriendly, judgemental, prejudiced and have preconceived ideas about the patients. There is a definite air of hostility, suspicion and a “them and us” culture between the staff and patients which is really, really sad and distressing to see. Staff do not respect the patients. There is no sympathy, no compassion, no empathy and no appreciation of what the patients are going through.”¹³⁹

“Being ill with sickle cell vaso-occlusive crisis can feel tantamount to being invisible for the amount you feel heard or respected.”

– Kye Gbangbola, Chair of Trustees, Sickle Cell Society and patient representative

We were provided with countless examples of this disrespectful treatment. One patient told us: “I have experienced sneers and laughter with comments like ‘this is a movie in here’, commenting on my sickle cell pain crisis.”¹⁴⁰

A patient outlined an incident in which they were administered the wrong blood, resulting in severe side-effects. However, they told us: “The consultants blamed me and made me feel like I had done something wrong.”¹⁴¹

Another patient described being admitted to a general ward, and “upon arrival, staff felt it was appropriate to say ‘oh no, this one is going to be hard work’. When I questioned why this was said about me as they did not know me, the response was ‘well, sickle patients require a lot of work and can be difficult’.”¹⁴² This was echoed by a patient carer, who told us that healthcare professionals “label the patients as ‘demanding’ [or] ‘difficult’ just because the patients have to press and literally beg for pain medication or help.”¹⁴³

Patients reported feeling they had to consciously be aware of their tone during agonising pain to avoid being seen as too aggressive or demanding. One

139 Araba Mensah, written evidence

140 Anonymous, written evidence

141 Anonymous, written evidence

142 Claire T, written evidence

143 Anonymous, written evidence



wrote: “I am a patient and understanding person, so most healthcare professionals, do not view me as ‘demanding’; silence leads to better treatment.”¹⁴⁴ Another said: “I have had to take on certain roles so that the healthcare professional in charge will treat me well when I am brought into the A&E. For example, I will compliment them, be overly nice to them and explain I am a good person. I will explain the scenario that led to my crisis and beg them to help me.”¹⁴⁵

Angela Thomas told us: “... having a crisis is a scary thing when it happens, not just what physical pain your body goes through, but what treatment are you going to have ... although I am the one in excruciating pain, I still have to be aware of my tone speaking to staff as they have in the past ignored me or taken my pain for aggressive behaviour.”¹⁴⁶ Shubby Osoba said

that he feels he has to adopt his “professional voice” and even go to the lengths of changing into smarter clothes while experiencing a pain crisis before going to hospital so that he will be taken seriously.¹⁴⁷

It is unacceptable that sickle cell patients going through a highly distressing experience feel that they have to be act in a certain manner out of fear of receiving disrespectful treatment from healthcare professionals due to prejudicial attitudes.

144 Anonymous, written evidence

145 Anonymous, written evidence

146 Angela Thomas, written evidence

147 Shubby Osoba, oral evidence session, 9 June 2021

Failure to believe or listen to patients: “The first response is always one of disbelief”

As NICE highlighted in their written evidence submission, their clinical guideline around managing acute painful episodes in hospital for sickle cell disease states that “patients (and their carers) should be regarded as experts in their condition”.¹⁴⁸

This is evidently far too often not the case. While patients noted that there are many diligent, dedicated, kind healthcare professionals, sickle cell patients frequently encounter secondary care staff who do not believe them or fail to have regard for their expertise in their condition.

Patients often face scepticism that they are in as much pain as they say they are. Charlotte Mensah wrote: “Our pain is often downplayed, overlooked or straight up ignored. The doctors and nurses sometimes imply that we’re exaggerating, faking, or lying about our symptoms ... the NHS Constitution makes a point about how every patient should be treated with compassion and empathy, but in my experience, only 15-20% of doctors and nurses do this.”¹⁴⁹ Angela Thomas told us that “hospital staff can be unsympathetic and believe it is a cry for attention”.¹⁵⁰

Zainab Garba-Sani described being “not believed and undermined as a patient with sickle cell”, such as being told, “You could at least look a little bit more unwell, you look absolutely fine, what’s wrong with you, should you be here?”¹⁵¹ Araba Mensah told us: “... each time [my daughter] presents at the hospital with a crisis as well as any of these complications, the first response is always one of disbelief at the extent of her pain and suffering”.¹⁵² Likewise, another patient

felt that sickle cell patients often develop a high threshold for pain and so when they attend hospital in pain “we are looked upon as if we are lying about our pain, as most health professionals expect us to be rolling and crying out loud before they believe we are actually in pain”.¹⁵³

“Going into hospital as a sickle cell patient requires you to put on an armour because from the moment you reach A&E it becomes your job to convince everyone you are really in that much pain and are not simply there for medication”

– Sickle cell patient

This failure to believe how much pain patients are experiencing often leads to accusations of illicit drug-seeking behaviour by healthcare professionals who do not believe that they actually require the pain relief to which they are entitled. Among the many examples we heard of such incidents, Angela Thomas wrote that “because morphine is the medication that eases my pain”, she faces questioning while trying to deal with the pain “to fathom whether I am in pain or just want the pain medication because I am an addict”.¹⁵⁴

148 NICE, written evidence

149 Charlotte Mensah, written evidence

150 Angela Thomas, written evidence

151 Zainab Garba-Sani, oral evidence session, 30 June 2021

152 Araba Mensah, written evidence

153 Anonymous, written evidence

154 Angela Thomas, written evidence

Another patient recounted an occasion in which a doctor reduced the level of pain relief medication they had been administered by another doctor: “When I corrected them, they called me a liar and made comments with the nurse about me lying to get more pain medication. Later that night, when they checked my record, they realised they were wrong and mis-practised.”¹⁵⁵

Mikell Allison provided us with another example of such stigmatising attitudes leading to a serious outcome, after encountering nurses who have “preconceived ideas (prejudice) that sickle cell patients are ‘drug addicts’ only there for the morphine. On one such occasion [when] I was admitted in 2009 with a particularly bad crisis, a nurse refused to administer pain relief. Being at the peak of crisis I could only say, ‘You can’t do that. I have been prescribed the pain medicine’. He then gave me the medication but I ended up in intensive care as my condition worsened.”¹⁵⁶

Sadeh Graham, a sickle cell patient who works in the healthcare system, told us that he only received appropriate treatment when his professional status was known: “The handful of admissions that have been okay or the times I received the appropriate dose of opioids was only due to [healthcare professionals] knowing I was a clinical pharmacist. This is something I used to find heartbreaking because as a sickle cell patient alone I will never be believed”.¹⁵⁷

A number of patients felt that making formal complaints about poor care did not lead to improvements because they were not believed or ignored. One patient outlined a time in which a nurse, while trying to cannulate her vein “repeatedly hit my hand hard because the line didn’t go in, blaming

me. I tried to make a formal complaint but wasn’t taken seriously, and I had no witness, so I had to concede.”¹⁵⁸

Others reported making complaints that did not even receive a response or, worse, resulted in them receiving worse treatment. Charlotte Mensah said that, at her local hospital, “patients are often scared to stand up for themselves, call doctors out on their behaviour, or make a complaint, because it’s common knowledge amongst sickle patients ... that if you offend, upset or anger the doctors, the quality of your care (and by extension your health) will worsen.”¹⁵⁹ Likewise, another patient told us: “Patients feel afraid at times to make complaints ... or escalate a problem because of fear of being bullied or having their treatment impeded. I have personally experienced this myself in the past”.¹⁶⁰

Dr Emma Drasar told us there needed to be a change in behaviour among some healthcare professionals based on her experience of supporting sickle cell patients: “Patients often report that they feel stigmatised against, that people don’t listen to what they say. I’ve had patients contact me and other haematology colleagues directly to try and advocate on their behalf ... We’re all doing a lot of teaching, but if people don’t internalise that knowledge and change their behaviour based on it, then however good your teaching is, however good your guidelines are, people have to act on what they’re being taught.”¹⁶¹

A repeated theme of patients’ evidence was the importance of healthcare professionals understanding that patients are experts in their own condition and should be listened to and respected. One patient told us that too often “doctors and nurses

155 Anonymous, written evidence

156 Mikell Allison, written evidence

157 Sadeh Graham, written evidence

158 Anonymous, written evidence

159 Charlotte Mensah, written evidence

160 Anonymous, written evidence

161 Dr Emma Drasar, oral evidence session, 16 June 2021



have their ‘plan for me’ but fail to listen to what I’m saying about my history or what I’ve already used/ tried at home before presenting to hospital.”¹⁶²

“The lack of collaborative work between health professionals/healthcare workers and patient leads to poor and sometimes tragic outcomes”

– Daniel Gunn, sickle cell patient

Ifunanya Obi wrote: “I’ve heard too many times while being in hospital, ‘I didn’t know that, are you sure?’ Like they are implying I don’t know anything because I’m a patient ... I feel a lot of people giving care to us

think they know it all and can’t learn anymore which is really bad because it puts a bad name on those that really want to learn and help.”¹⁶³

Failing to listen to patients can have serious implications for the care they receive. A patient told us of an experience they had had where they requested the insertion of a femoral line into their groin to provide a blood transfusion, knowing that their veins were too damaged to be used: “I could see that he did not like being told and felt he knew better, I could feel his body language saying ‘I know what I am doing, I don’t need to be told, it will be fine.’ The pain that I felt from that needle trying to penetrate through hard scar tissue became evident by those screams echoing throughout the hospital theatre and corridors. I was quickly sedated before receiving an apology and a look of regret from a flustered anesthetist. He should have listened, I wasn’t telling him how to do his job I was just letting him know what my body needed because of my knowledge through my past experiences.”¹⁶⁴

162 Anonymous, written evidence

163 Ifunanya Obi, written evidence

164 Anonymous, written evidence

Such experiences highlight the importance of treating patients as experts in their condition, in line with NICE guidelines. As Bell Ribeiro-Addy put it: “Who is going to know better about their care and what needs to be done than the individual and their family members and the people that care for them?”¹⁶⁵

Recommendation: NHS England & NHS Improvement to require NHS Trusts to conduct and report regular audits of patient involvement in decisions about their care, utilising patient feedback,

in line with NICE clinical guideline stating that sickle cell patients (and their carers) should be regarded as experts in their condition.

Recommendation: NHS England & NHS Improvement to establish formal sickle cell patient advisory groups, based on consultation with the Patient and Public Voice Assurance Group, to work in partnership with and conduct oversight of NHS sickle cell services.

Lack of prioritisation: “People are treated as an added-on”

Sickle cell patients told us that they are often made to feel like they are not a priority for healthcare professionals. One patient told us that “we feel that the hospital here and nationwide actually puts sickle cell patients’ needs at the very bottom of healthcare. We feel and see we’re being undermined, undervalued, and not being listened to when we are trying to gain some semblance of peace and dignity while in hospital at our worst and weakest time in our already tumultuous lives.”¹⁶⁶

Calvin Campbell referred to experiences of having to wait too long for pain relief and then being told “you’re not the only sick person on the ward’ or ‘there are sick people I have to deal with’, as if someone with sickle cell in the middle of a crisis and in excruciating pain is not considered sick”.¹⁶⁷

Kye Gbangbola told us: “I have been taken to A&E and sat for many hours, waiting for doctors to attend and see me. I would repeatedly ask for pain relief, I’d repeatedly ask for doctors, including their resident sickle cell specialist, only to be told, ‘You have to wait.’ So you wait, and there is no one to tell that things are not going well ... Annoyed and angry healthcare workers, they make patients feel like a pest just for asking for pain relief.”¹⁶⁸

“Generally, it has been a constant battle to get adequate care for my child. I have to push for further investigation in all elements of my child’s healthcare and, speaking to other parents, they are experiencing the same.”

– Sickle cell patient carer

Another patient described being in hospital with an extremely high temperature and asking a nurse for some ice cubes and assistance with tepid sponging as “from personal experience I was concerned in case I started fitting”. After being refused the ice, “he continued to inform me that he has more important things to do than to stand beside me sponging me down”.¹⁶⁹

A member of the parent and child support group for Darent Valley Hospital, Kent highlighted a lack of prioritisation when taking her daughter for blood

165 Bell Ribeiro-Addy, oral evidence session, 30 June 2021

166 Anonymous, written evidence

167 Calvin Campbell, written evidence

168 Kye Gbangbola, oral evidence, 9 June 2021

169 Anonymous, written evidence

transfusions: “We are asked to come over by 10am and we will be sitting and waiting up till 1pm before the blood comes. We often leave around 6pm or 7pm and, at times, after the night staff have started their shift.”¹⁷⁰

Professor Jo Howard referred to similar findings of a national peer review, where “there was a lot of repeated examples about where people felt like second-class citizens, where they’re treated as an ‘added-on’, where they’re treated in cancer centres, because a lot of haematology is cancer so the sickle patients can just go along to the same centre.”¹⁷¹

There was a disturbing theme in the evidence we received of patients having their ability to call for aid while in hospital taken away or ignored. A friend of a patient recounted visiting her friend on a number of occasions and finding that the sound from her friend’s buzzer had been turned off, which she felt made it “easier for the staff to ignore the patients”. On another occasion, she witnessed a healthcare professional throw her friend’s buzzer out of reach.¹⁷² Another patient told us: “Some nurses will deliberately come and silence the call bell and walk off without notifying the appropriate staff members of the requests being made by the patient.”¹⁷³

Zainab Garba-Sani described being admitted onto a hospital ward “in quite a lot of pain and my pain medication was wearing off ... I buzzed the buzzer literally about every 30 minutes for about four hours, before then getting up myself and trying to find someone. I then found a nurse and the nurse said, ‘can you go and sit back down, we’ll come to you, just press the buzzer’. I was like, ‘well, that’s what I’ve been doing for the last how many hours’ ... It’s that

feeling of being completely ignored, not given the pain medications that you needed and that you’re requesting.”¹⁷⁴

Similarly, another patient told us: “I have met nurses who, in order nullify my cries of agony, pulled the bed curtains around me and ignored my cries for help ... At times I was in fear for my life. No one was listening to me but actively ignoring my cries for help, while attending other patients as they tiptoed around my bed.”¹⁷⁵

Patients and clinicians told us that sickle cell is often treated as less of a priority than other health conditions. According to Dr Arne de Kreuk: “If an A&E member of staff has to prioritise between a sickle cell patient in pain and someone who’s broken a leg, unfortunately, they’re not treated equally.

Often, sickle cell is regarded as something that can wait, despite the fact that the first line of the NICE guidance very clearly says, ‘Treat a sickle cell crisis as a medical emergency.’”¹⁷⁶

Madeleine Glover, a haematology nurse, told us that, in her experience, sickle cell patients often have their appointments for apheresis (automated exchange blood transfusion) procedures moved at short notice “to accommodate other patient groups”. She further outlined a number of ways in which access to specialist haematology services for sickle cell patients “is secondary to access allowed to other patients, principally those with cancer”, including capping the number of sickle cell patients who may attend day unit services, failing to consider current or likely demand for haemoglobinopathy patients when planning space in day care settings that also host cancer patients and giving priority for the use of side rooms and bed spaces to cancer patients.¹⁷⁷

170 Parent and child support group, Darent Valley Hospital, Kent, written evidence

171 Professor Jo Howard, oral evidence session, 30 June 2021

172 Anonymous, written evidence

173 Anonymous, written evidence

174 Zainab Garba-Sani, oral evidence session, 30 June 2021

175 Anonymous, written evidence

176 Dr Arne de Kreuk, oral evidence session, 16 June 2021

177 Madeleine Glover, written evidence

Patients also highlighted feeling that other conditions are prioritised more in secondary care. One said that, as their care was often provided alongside cancer patients, “we unconsciously are pitted against each other and cancer will almost always win...

For example, if a [sickle cell] patient requests pain relief before a cancer patient, though the medication is due to be given, the cancer patient will receive their medications before a sickle patient. We frequently hear the words, ‘I will be with you soon; I have other patients who need me’. In that moment, you are not one of their patients”.¹⁷⁸

Another felt that “there also seems to be some strange sort of competition or bias towards preferential treatment to those with white cell

conditions. This is unspoken, yet whenever [there are] any changes to ward structure or patient treatment, it is the sickle patients who always have to give ground.”¹⁷⁹

These examples all demonstrate the shocking extent to which sickle cell patients are treated as though they are not a priority when accessing secondary care and the frequency with which they are made to feel their condition is not as serious as others.

178 Anonymous, written evidence

179 Anonymous, written evidence

INADEQUATE INVESTMENT IN SICKLE CELL CARE

Sickle cell patients, carers and clinicians all noted the low levels of investment in sickle cell services and research into the condition, particularly when compared with other similar medical conditions. The recent move by NHS England & NHS Improvement to commission sickle cell as a specialised service, including the formation of Haemoglobinopathy Coordinating Centres, is welcome but was felt by many to have still not adequately addressed the problem of inadequate funding for sickle cell services.

Under-resourcing of sickle cell services: “It has the feeling of an underfunded and underinvested ‘Cinderella’ area of medicine”

We were told that under-resourcing of sickle cell services is a significant contributor to sub-standard care. One patient who attends a London hospital told us that there are only between four and eight beds allocated to sickle cell patients on the haematology ward they access and asked “how sickle cell patients are meant to feel safe when we can’t even get a bed on our own specialist ward”.¹⁸⁰

Sadeh Graham said that the haematology ward at the hospital in the West Midlands he attends has no access for sickle cell patients, with only nine beds in the haematology ward, which are reserved for patients with other conditions. The lack of available beds for sickle cell patients requiring pain management means patients are often “turned away to go home or sit in A&E for hours and be subject to poor care”, Sadeh told us.¹⁸¹

The under-resourcing of sickle cell services was also raised by many of the clinicians we received evidence from. Professor Jo Howard noted that “the majority of hospitals” are unable to provide apheresis out of hours, which results in “patients travelling halfway across the country”. Professor Howard described this as a “funding issue ... there hasn’t been enough investment in that”.¹⁸²

Evidence from the Haemoglobin Disorders Peer Review Programme Clinical Leads highlighted the lack of support sickle cell services receive from NHS Trust leaders to address areas of clinical care considered to be of ‘immediate risk’ or ‘concern’ during reviews. Whereas in a national renal care review in 2016, 63% of services stated that their Trusts had supported them to address such concerns in a post-hoc survey, “this was the case in a fraction of services in the [sickle cell] reviews. Specifically, 25% (in 2010-2011); 35% (in 2012-2013); 39% (in 2014-2016)

¹⁸⁰ Anonymous, written evidence

¹⁸¹ Sadeh Graham, written evidence

¹⁸² Professor Jo Howard, oral evidence session, 30 June 2021

and 54% (in 2018-2020) of haemoglobinopathy services received Trust support to address areas considered ‘immediate risk’ or ‘concern’ to patient care.” The Clinical Leads of the Haemoglobin Disorders Peer Review Programme concluded that “this reflects the lack of Trust executive-level interest in providing material and human resources required to improve care of people with haemoglobin disorders”.¹⁸³

Many clinicians contrasted the level of resource provided to sickle cell services with that provided to other similar conditions. One haematologist said that, in their opinion, sickle cell patients do not receive “anything like the level of care that other patient groups with chronic disease do, and it has the feeling of an underfunded and underinvested ‘Cinderella’ area of medicine”.¹⁸⁴

“While the recent restructuring of haemoglobinopathy provision is a welcome recognition of the existence of a problem, structural reorganisation without major investment in staff and training (especially of staff in other specialties) will not be enough.”

– National Haemoglobinopathy Panel

Dr Rachel Kesse-Adu told us that, despite feeling her NHS Trust has one of the best-resourced sickle cell

services in the country, “we do not even marginally compare when you hold us up to the resource and support both in the hospital and community that exists for other chronic conditions (such as cystic fibrosis) or other ailments such as cancer”.¹⁸⁵

Another haematologist told us that services for haemophilia and cystic fibrosis “provide a benchmark for holistic comprehensive care and sickle services generally fall below this standard” and that sickle cell is often the “poor relation” compared to cancer care in haematology departments. While this clinician welcomed the recent additional funding from NHS England, they told us that “the monies available did not match the requirements of the Specialised Haemoglobinopathy [Coordinating] Centre service specification, such that my employing Trust has accepted that it has to overspend on this budget”.¹⁸⁶ Similarly, Professor Jo Howard told us the funding of red cell exchange transfusion “is not adequate and the tariff received by centres is less than it costs”.¹⁸⁷

We also heard that the level of resource varies hugely across the country. Dr Thomas Lofaro, a Consultant Haematologist who previously trained and worked in London and is now based in Hertfordshire, told us that “it is very difficult to provide the same level of service because of the great difficulties in accessing funding and support for this condition outside of major centres ... patients may be fewer, but their needs are the same (or even more for lack of support) and the care we can provide is not the same”.¹⁸⁸

The under-resourcing of sickle cell services can have serious outcomes. For example, one patient told us that, aged seven, they required an exchange blood transfusion which could not be offered at their local hospital and were instead referred to a specialist paediatric intensive care unit in central London after ten days. This delay led to the patient being hospitalised for almost two months and in a

183 Haemoglobin Disorders Peer Review Programme Clinical Leads, written evidence

184 Anonymous, written evidence

185 Dr Rachel Kesse-Adu, written evidence

186 Anonymous, written evidence

187 Professor Jo Howard, written evidence

188 Dr Thomas Lofaro, written evidence

wheelchair for at least six months after that, followed by intensive physiotherapy, all of which impacted their education.¹⁸⁹

Recommendation: NHS England & NHS Improvement to provide increased funding for sickle cell services in recognition of the consistent underfunding of sickle cell services when compared with services for other conditions. This should include dedicated funding for NHS Trusts to improve apheresis capacity across the country.

Recommendation: Clinical Commissioning Groups and local authorities to provide additional funding for third sector providers and community care organisations for social prescription in relation to sickle cell to reduce pressure on NHS services.



Under-staffing of sickle cell services: “We are constantly facing a staffing crisis”

The lack of investment in sickle cell services is also apparent in the significant shortfall in appropriate numbers of healthcare professionals working in sickle cell care.

The British Society for Haematology told us that haemoglobinopathy “has a longstanding recruitment problem and an ageing staff demographic suggesting that shortages are likely to continue to be an issue”. They referred us to three recent workforce surveys, run by the Royal College of Physicians, the Royal College of Pathologists and the British Society for Haematology, which they said “demonstrate a marked shortfall in consultant numbers over the next few years across all areas of haematology”. In line with the evidence set out in the section above, the British Society for Haematology welcomed the recent changes to sickle cell service provision by NHS England but added, “the funding allocated to this service redesign was minimal, and for many centres did not cover the costs of establishing appropriately staffed core services”.¹⁹⁰

Haematologists we heard from echoed this concern around levels of staffing. Dr Emma Drasar told us that it is a struggle to attract enough staff to red cell haematology “which means we are constantly facing a staffing crisis ... I exist in a state of anxiety around sustaining my service, worried that my patients will not receive good care unless I am there and in fear that I am not doing the best for my patients due to external forces”.¹⁹¹ Professor Jo Howard noted that national recommendations for staffing levels per patient numbers “are universally not met” for sickle cell services, adding: “The workload

of [haemoglobinopathy] clinicians is huge and consistently exceeds contracted hours and ‘burnout’ is a major concern”.¹⁹²

“Chronic under-staffing, under-training and under-funding of clinical positions (doctors, nurses and psychologists) is likely to have contributed to the lack of appropriate standard of care for patients”

– Consultant Haematologist

Even more concerning, the situation is getting worse, according to the Haemoglobin Disorders Peer Review Programme. In the 2016 review, 35% of sickle cell services stated that they had problems with time available for senior clinicians to provide leadership of the service or availability of consultant medical staff. By the 2020 review, this had risen to an astonishing 84% of services.¹⁹³

The under-funding of services and inadequate levels of staffing can be a mutually reinforcing problem. The Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee told us that, as a result of the significant underfunding of sickle cell services, “there are significantly fewer numbers of specialised nurses, doctors, psychologists and support staff that

190 British Society for Haematology, written evidence

191 Dr Emma Drasar, written evidence

192 Professor Jo Howard, written evidence

193 Haemoglobin Disorders Peer Review Programme Clinical Leads, written evidence

have chosen to work within this service. They mainly move into oncology (white cells) and clotting with research and opportunities.”¹⁹⁴

Similarly, the UK Forum on Haemoglobin Disorders noted that “this under-resourced area has become increasingly challenging to recruit to ... Junior doctors struggle to find academic or research opportunities in haemoglobinopathies and will often take up research programmes in malignant or coagulation and hence will fall into that career path as Consultants”.¹⁹⁵ Professor Jo Howard said that “even when posts are funded it is difficult to fill specialist posts and many are vacant”.¹⁹⁶

In addition to concerns around the number of haematology doctors and nurses, many submissions also mentioned shortfalls in specialist psychologist staff and community nurses to support sickle cell patients. Professor Jo Howard cited the difficulties many services have faced in obtaining funding for psychologists and other specialist staff as evidence that “the funding of [sickle cell] care does not seem to be a priority”.¹⁹⁷

Under-staffing is a significant problem in sickle cell care and, with the consensus being that the problem is currently on course to get worse, it is imperative that NHS England & NHS Improvement take action to address the issue to improve the care sickle cell patients receive.

Recommendation: Department of Health and Social Care to convene organisations including Health Education England, the General Medical Council, the Nursing and Midwifery Council, the medical royal colleges and medical and nursing schools to come together with senior sickle cell service representatives to engage in effective workforce planning for sickle cell services, including the allocation of specialist training opportunities.

Recommendation: All NHS Trusts to ensure that specialised service funding is invested in meeting recommended sickle cell service staffing numbers.

Underinvestment in sickle cell research and treatment: “Research has been woefully inadequate”

The long-standing lack of investment in sickle cell research and new treatments was repeatedly highlighted in the evidence we received. There are currently a very limited range of treatments available for sickle cell patients in the UK, with the two most significant being the use of blood transfusions and the medicine hydroxyurea, which can reduce the frequency of sickle cell crises.

Shortly before publication of this report, NHS England & NHS Improvement approved Crizanlizumab, the first new treatment for sickle cell in over 20 years, a welcome development but one that is well overdue. The evidence we received suggested the lack of new treatments for over two decades is a reflection of the health inequalities associated with sickle cell disorder.

194 Royal College of Pathologists’ Transfusion Medicine Specialty Advisory Committee, written evidence

195 UK Forum on Haemoglobin Disorders, written evidence

196 Professor Jo Howard, written evidence

197 Ibid.

This lack of treatment availability is a result of low levels of research, we were told. Professor Jo Howard noted that there are very small numbers of sickle cell research-active clinicians in the UK and that it has been historically difficult to obtain funding, adding: “A lack of research into health outcomes hampers the introduction of new therapies as there is little data about the economic impact of sickle cell disorder.”¹⁹⁸

Araba Mensah was among those who highlighted the lack of research into sickle cell. She told us: “Considering the magnitude of the effect it has on sufferers’ lives, research has been woefully inadequate over the decades ... The majority of patients are left with no option but to take painkillers for their condition while they live in hope that one day a medical breakthrough will provide them some much-needed relief.”¹⁹⁹

Again, many noted the contrast between levels of research and treatment-availability for sickle cell with those for other, similar conditions. A Consultant Haematologist told us that, unlike for cystic fibrosis, no specific funding streams are available for sickle cell research, meaning sickle cell researchers have to apply to generic funding calls.²⁰⁰ Another said: “The lack of access to research is especially apparent when you compare the opportunities to patients with a cancer diagnosis to those in the sickle cell community, which is evident every day to those of us who work in environments where colleagues are involved in treating patients with cancer.”²⁰¹

While this lack of investment in research means that there are limited treatments available, frustration was also expressed that treatments available in other countries have not been approved in the UK. Professor Jo Howard told us there are “several new

drugs and therapies” available in other countries which have not been approved in the UK “and are unlikely to be available for many years”.²⁰²

University College London Hospital noted that NHS England has published clinical commissioning guidance for sibling Allogeneic Haematopoietic Stem Cell Transplantation for adults with sickle cell disease, which is potentially curative for those people with severe disease in whom other treatments have failed or have not been tolerated. Their submission called for additional funding from NHS England to ensure adequate investment in new clinical pathways to treat this cohort of patients.²⁰³

It is clear that decades of underinvestment in sickle cell research has led to a dearth of treatment options for sickle cell patients. Increasing the level of research and the availability of treatment options is key to improving sickle cell care outcomes.

Recommendation: UK Research and Innovation and the National Institute for Health Research to launch dedicated sickle cell research opportunities, including supporting and funding research into genetic therapies to cure sickle cell disorder.

Recommendation: NHS England & NHS Improvement to report results of Managed Access Programme for Crizanlizumab to support roll-out following the drug’s approval.

198 Ibid.

199 Araba Mensah, written evidence

200 Dr Subarna Chakravorty, written evidence

201 Anonymous, written evidence

202 Professor Jo Howard, written evidence

203 University College London Hospital, written evidence

CONCLUSION

In addition to the issues explored over the course of this report, a further common theme of the evidence we received from patients and specialist sickle cell clinicians was anger and frustration that the same issues have been highlighted time and again over many years without any action.

It is a damning indictment of the way sickle cell patients have been treated that so many told us they fear, or actively avoid, accessing secondary care services. The feeling that many sickle cell patients have been left with is that they are not a priority, that their suffering is not considered important and that treatment that would not be accepted for other patient groups is ignored when it relates to sickle cell. The only way this can be changed is by taking urgent steps to address the factors behind sub-standard care for sickle cell patients.

The shocking, tragic and avoidable death of Evan Nathan Smith was just the latest in a long line of deaths and near misses among sickle cell patients. Further avoidable deaths among sickle cell patients will be inevitable unless action is taken.

We urge all of those we have addressed recommendations to in this report to set out the steps they will be taking in response. More generally, we are calling for healthcare leaders, including the Secretary of State for Health and Social Care, the Chief Executive of NHS England & NHS Improvement and leaders of the new Integrated Care Systems to adopt improving sickle cell care as a key priority.

It is long past time that action is taken to improve sickle cell patients' experience of secondary care. The SCTAPPG looks forward to working with all relevant stakeholders to deliver the changes that are required.

ANNEX

EVIDENCE

The SCTAPPG would like to thank all those who provided evidence to the inquiry.

Oral evidence

The SCTAPPG conducted three oral evidence sessions with the following witnesses:

Wednesday 9th June 2021

- June Okochi (patient representative)
- Alex Luke (patient representative)
- Kye Gbangbola (Chair of Trustees, Sickle Cell Society and patient representative)
- Shubby Osoba (patient representative)
- Dr Shivan Pancham (Consultant Haematologist, Sandwell and West Birmingham NHS Trust)

Wednesday 16th June 2021

- Cedi Frederick (Chair, North Middlesex University Hospital NHS Trust)
- Dr Geraldine Walters CBE (Executive Director for Professional Practice, Nursing and Midwifery Council)
- Professor Baba Inusa (Consultant Paediatric Haematologist, Guy's and St Thomas' NHS Foundation Trust and Chair, National Haemoglobinopathy Panel)
- Dr Arne de Kreuk (Consultant Haematologist, North Middlesex Hospital and Deputy Lead, North London Haemoglobinopathy Centre)
- Dr Emma Drasar (Consultant Haematologist, The Whittington Hospital and University College London Hospital and Chair, Haemoglobinopathy Coordinating Centres)

Wednesday 30th June 2021

- Betty & Charles Smith (parents of Evan Nathan Smith)
- Professor Jo Howard (Consultant Haematologist, Guy's and St Thomas' NHS Foundation Trust and Chair, NHS England Haemoglobinopathies Clinical Reference Group)
- Bell Ribeiro-Addy MP (member, APPG on Sickle Cell and Thalassaemia and former care provider to sickle cell patient)
- Zainab Garba-Sani (patient representative)

Written evidence

The following individuals provided written evidence to the inquiry:

- Joana Allison Mikell Allison
- Liz Blankson-Hemans
- Carol Burt
- Calvin Campbell
- Dr Subarna Chakravorty
- Diane Crawford
- Dr Emma Drasar
- Kye Gbangbola
- Stephanie George
- Madeleine Glover
- Sadeh Graham
- Daniel Gunn
- Professor Jo Howard
- Dr Fatima Kagalwala
- Jaspreet Kaur
- Dr Rachel Kesse-Adu
- Dr Thomas Lofaro
- Araba Mensah
- Charlotte Mensah
- Ifunanya Obi
- Denise Owusu-Ansah
- Richard Patching
- Charles Phillip
- Mamme Prempeh
- Angela Thomas
- Dammy Shittu
- Dr Tullie Yeghen
- Amanda [surname withheld by request]
- Claire T [full surname withheld by request]
- We received a further 54 anonymous submissions.
- The following organisations provided written evidence to the inquiry:
 - British Society for Haematology
 - Crescent Kids
 - Darent Valley Hospital Paediatric Centre
 - Evelina London Children's Hospital
 - Global Blood Therapeutics
 - Guy's and St Thomas' NHS Foundation Trust adult haematology service

- Haemoglobin Disorders Peer Review Programme Clinical Leads
- National Haemoglobinopathy Panel
- National Institute for Health and Care Excellence
- NHS Blood and Transplant
- NHS England & NHS Improvement
- Royal College of Pathologists Transfusion Medicine Specialty Advisory Committee
- Serious Hazards of Transfusion
- Sickle Cell Suffolk
- Sickle Plus
- Sickle Cell Winning Ways
- South East Haemoglobinopathy Co-ordinating Centre
- UK Forum on Haemoglobin Disorders
- University College London Hospital
- West London Haemoglobinopathy Coordinating Centre
- Whittington Health NHS Trust

Parliamentarians who participated in the inquiry

Rt Hon Pat McFadden MP (Chair)

Janet Daby MP

Bell Ribeiro-Addy MP

Stella Creasy MP

Baroness Benjamin

Acknowledgement

This report was produced by Aidan Rylatt of Principle Consulting, working with the Sickle Cell Society as the SCTAPPG's secretariat.

