SICKLE CELL SOCIETY

NHS RACE & HEALTH OBSERVATORY

Sickle cell comparative review to inform policy report

Providing evidence-based recommendations to tackle inequalities

June 2025

IMPERIAL

Contents

Foreword

Sickle cell is the most common genetic disorder in the UK, yet it has endured years of inadequate attention and investment, resulting in stark and persistent inequalities in healthcare for those living with the condition. This detailed research is invaluable in pinpointing where discrimination and bias continue to occur—whether in resource allocation, access to specialist care, or research funding. Crucially, the findings reveal that this neglect is not due to a lack of need: hospital admissions for sickle cell rose by 42% between 2013 and 2022.

The report goes beyond identifying disparities. It uncovers the root causes of these inequalities and offers an evidence-based foundation for meaningful change. It highlights a long-standing pattern of underinvestment and de-prioritisation that has eroded trust among many people with sickle cell in the healthcare system. Too often, they encounter poor knowledge of their condition or substandard treatment from the very services designed to support them.

In this context, the report is more than just a call to action – it is a roadmap for change. Its six key recommendations for policy, practice, and research provide actionable, practical solutions aimed at addressing the systemic failures exposed. If adopted, these measures could improve outcomes, rebuild trust, and ensure that people with sickle cell across the country receive the care that they need and deserve.

Moreover, this comparative research holds lessons that extend beyond sickle cell care. By examining disparities in funding and resource allocation compared to conditions such as cystic fibrosis and haemophilia, the report identifies both areas of progress and opportunities for improvement across multiple conditions. It demonstrates the impact of sustained investment in specialist care. The goal is to learn from gains made in other conditions in order to raise the standard of care for people with sickle cell to the same level – ensuring equity.

We extend our sincere gratitude to the research team, the advisory group, and the many experts and key partners who contributed to this vital work. Special thanks go to the people with sickle cell, their families, nurses, and haematologists across the NHS, whose lived experiences and insights have enriched the report's technical rigour and practical relevance. Their voices remind us why change is not only necessary but urgent.

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Executive summary

Sickle cell is often described as a neglected disease, both globally and in the UK. Comparative research can provide valuable insights into inequalities to guide public health policies. In this report, we analysed data across a wide range of indicators, by i) reviewing the literature and previous reports; ii) looking at research funding, financial resources of charities, scientific publications, clinical trials, approved drugs, dedicated disease registries and online awareness for sickle cell, cystic fibrosis and haemophilia; iii) analysing hospital admission data for these three conditions in relation to waiting times, length of hospital stays and estimated costs to the NHS; and iv) collecting evidence from patients about their lived experiences to complement the other parts of our analyses.

Some of our data reveals striking inequalities, which were often reflected in the experiences of patients living with sickle cell across the country. Nevertheless, evidence of these inequalities is not new, and this report adds further to the urgent need to address the underlying problems affecting the quality of care for patients with sickle cell. Importantly, this comparative research shows that improvements are also needed for other severe chronic conditions, such as cystic fibrosis and haemophilia, and that lessons can be learned from successes achieved in other specialties. This report includes a set of recommendations that aim to help tackle inequalities associated with sickle cell in the UK.

Graphic summary of findings

20%
of babies with **sickle cell** were not seen by a specialist by three months of age

42%
increase in hospital admissions for **sickle cell** between 2013 and 2022

Total estimated annual hospitalisation costs
2X HIGHER
for **sickle cell** than **cystic fibrosis**

The median age of death for
sickle cell 47
cystic fibrosis 46
general population 84

~2
specialist nurses per 100 patients for **cystic fibrosis** vs
~0.5
for **sickle cell**

>2.5X
more research funding for **cystic fibrosis** than for **sickle cell** or **haemophilia**

On average, patients with **sickle cell** managed
7 PAIN CRISES
without medical help

In a multi-country study, people with **sickle cell** in the UK had the **highest** annual mean hospitalisations for pain crises

Introduction

Rationale for the project

Recent evidence has highlighted striking inequalities in the health and care of individuals with sickle cell. These inequalities are not new, but this evidence has provided momentum for the long-term transformational change needed in the management and care of individuals with sickle cell, as illustrated by the funding commitment of the NHS Race & Health Observatory (RHO). To support this change, comprehensive evidence, from the literature and available data, is needed to further document these inequalities, including through comparisons with other comparable conditions. A better understanding of the current context underlying these inequalities would inform efforts to reduce and eliminate them in the future. Such a change cannot be achieved without the active involvement of key stakeholders from patients and their families to nurses, haematologists and the wider NHS. In this report, we try to bring together these different pieces of evidence to provide a compelling case for recommendations and the transformational change that patients with sickle cell deserve.

Why this work is important

Sickle cell is the most common genetic disease in the UK.[1] It predominantly affects people of African or Caribbean heritage. The "No One's Listening" report of the Sickle cell Society and All-Party Parliamentary Group on Sickle cell and Thalassaemia (SCTAPPG) highlighted inequalities in healthcare experience and variability in treatment for people living with sickle cell. This report also exposed a worrying shortfall in adequate care and treatment for sickle cell patients. Health inequalities result in part from structural biases in the healthcare system. Comparative research can play a valuable role in evidencing these biases and their consequences as a basis for evidence-based policy recommendations.

Conditions considered

We decided to consider three conditions for this comparative analysis: sickle cell, cystic fibrosis and haemophilia. We deliberately limited the number of conditions considered to enable a thorough multi-criteria comparison, rather than an eclectic superficial approach looking at a larger number of conditions.

Although "sickle cell" is not an official clinical terminology, we have opted to use it throughout this report instead of "sickle cell disease" or "sickle cell disorder" based on patient input. By "sickle cell", we mean different forms of sickle cell disease including sickle cell anaemia (HbSS), sickle cell–haemoglobin C (HbSC) and sickle cell-beta-thalassaemia (S-βthal).

Sickle cell is the primary focus of this work. Cystic fibrosis is often used in the UK and in other countries (e.g. US) as a typical comparator.[2, 3] Typical estimates of the numbers of individuals with these two conditions tend to suggest that the number of patients affected are quite similar: ~14,000 for sickle cell[1] and ~12,000 for cystic fibrosis.[4] These two conditions are also included in the universal newborn screening programme in place in the UK.

We believe that haemophilia, another blood disorder, was also worth considering in this comparison because it is also managed by haematologists and provides effective models and benchmarks for sickle cell. Although it affects less than 10,000 patients in the UK, it is often perceived as comparatively well-resourced and supported compared with sickle cell. It was also central to the Infected Blood Inquiry which published its final report towards the start of this work.

Throughout this report, we will consistently use the following colours for the three conditions considered to help with the reading and interpretation of our findings.

Sickle cell **Cystic fibrosis** **Haemophilia**

In line with the remit of the NHS to provide comprehensive, universal and free services at the point of delivery, specialist care and management should be available to all patients, independently of their demographics, place of residence, or condition by which they are affected. Although the primary focus of this report is to highlight inequalities affecting patients with sickle cell, the evidence presented suggests that improvements could also be made for other conditions, including cystic fibrosis and haemophilia.

Aim and objectives

The primary aim of this work is to provide evidence of the differences and inequalities in the perception and care that patients with sickle cell experience compared with other inherited disorders such as cystic fibrosis and haemophilia. It will help to better document and understand inequalities in the care of people with sickle cell across England, in order to ensure that the NHS can provide all patients with safe care and treat them with respect and dignity, independently of race, location or socio-economic status, and to guide recommendations about how these inequalities could potentially be reduced.

Our objectives have been to collect comparative evidence using three sets of data:

- **the literature**, including scientific publications, policy reports, etc (**Evidence Set 1**)
- **routine data**, including selected online indicators and hospital admission data (**Evidence Set 2**)
- **lived experiences** of patients through an online survey (**Evidence Set 3**)

We want to highlight again that more is needed to further improve the quality of life of all patients with sickle cell, cystic fibrosis, and haemophilia as well as of those suffering from any other severe chronic conditions, rare or common, across the UK. Throughout our analyses, we have objectively highlighted areas of improvement for any of the three conditions considered, based on data presented in this report.

The Research Team

We are a multidisciplinary team with a lot of expertise in quantitative data analysis of routine health data, literature reviews, patient engagement and involvement, and patient care.

- **Dr Frédéric Piel** is an epidemiologist within the School of Public Health at Imperial College London, with expertise of sickle cell and cystic fibrosis.
- **Dr Rutendo Muzambi** is an epidemiologist within the School of Public Health at Imperial College London, with a focus on health inequalities.
- **Professor Alex Bottle** is a statistician within the School of Public Health at Imperial College London, with expertise in measuring and monitoring the quality of healthcare in hospital and in the community for patients with chronic disease.
- **Dr Daniel Dexter** is a haematologist at Kings College London and looks after patients with sickle cell and haemophilia.

The Advisory Team

- **Ganesh Sathyamoorthy** is the Deputy Director of the Ethnicity & Health Unit at Imperial College London and a trustee of the Sickle cell Society.
- **Dr Cherelle Augustine** is an Engagement Coordinator in the Ethnicity & Health Unit at Imperial College London, living with sickle cell.
- **Professor Mark Layton** is a senior haematologist at Hammersmith Hospital, part of the Imperial NHS Trust, and looks after patients with sickle cell and haemophilia.
- **Professor Siobhan Carr** is a consultant in paediatric respiratory medicine based at the Royal Brompton Hospital, looking after children with cystic fibrosis.
- **John James** is the CEO of the Sickle cell Society.
- **Dr Carl Reynolds** is a Senior Clinical Advisor at the NHS RHO, with a leading role on sickle cell inequality.

The Patient Contributors

- **Jeannine Joseph** is a patient with sickle cell and provided input on the online survey and the report.
- **Funmi Dasaolu** is another patient with sickle cell, who also provided input on the online survey and the report.

Ethic approval and disclaimer

The use of NHS Hospital Episode Statistics (HES) data, through the Dr Foster Unit, has approval from the Health Research Authority to use HES data for research and measuring quality of delivery of healthcare, from the London - South East Ethics Committee (REC ref: 20/LO/0611).

The work presented in this report was funded by the NHS Race & Health Observatory.
Our aims were to be rigorous, systematic, and as comprehensive as possible in our data collection and reporting. Given the relatively short timeframe of this project, we relied mostly on open access evidence and routine health data. We acknowledge that there might be additional relevant evidence which could have been included in this report.
All the analyses were conducted independently by the research team at Imperial College London. The Advisory Team provided guidance regarding the methodology used, the data collection, the interpretation of the findings, and the wording of the report. The Patient Contributors used their lived experience to guide the questions of the online survey and the wording of the report.

The views expressed in this report are those of the authors and not those of their institutions.

List of abbreviations

A&E: Accident & Emergency

AMRC: Association of Medical Research Charities

APC: Admitted Patient Care

BNF: British National Formulary

CFR: Cystic Fibrosis Registry

CFTR: Cystic Fibrosis Transmembrane Conductance Regulator

DHSC: Department of Health & Social Care

EU: European Union

GP: General practitioner

HCC: Haemoglobin Coordinating Centre

HES: Hospital Episode Statistics

HMIC: Health Management Information Consortium

HRG: Hospital Resource Group

ICD: International Classification of Diseases

IMD: Index of Multiple Deprivation

IQR: Interquartile range

LSOA: Lower layer support output area

MHRA: Medicines and Healthcare Products Regulatory Agency

MRC: Medical Research Council

NHD: National Haemophilia Database

NHR: National Haemoglobinopathy Register

NHS: National Health Service

NICE: National Institute of Clinical Excellence

NIHR: National Institute for Health & Care Research

ONS: Office for National Statistics

PCA: Patient-controlled analgesia

RHO: Race & Health Observatory

SC: Sickle Cell

SCTAPPG: Sickle Cell & Thalassaemia All-Party Parliamentary Group

UK: United Kingdom

UKRI: UK Research & Innovation

US: United States

Methodology

This work was primarily centred around three sets of evidence: a review of the literature, a data analysis of key comparative indicators and of routine hospital data; and engagement with patients with sickle cell about their lived experiences. Below we provide further details about the methodology used to gather these three sets of evidence.

Evidence Set 1: Literature review

The literature review included evidence from both the peer-reviewed (e.g. academic publications) and grey (e.g. policy reports) literature. We searched three electronic databases for relevant studies: the Health Management Information Consortium (HMIC) database, MEDLINE (Ovid interface) and PsycINFO. The study period covered 15 years, from 1st January 2010 to 31st December 2024. Subject headings and keywords relating to the three conditions considered and health inequalities were used, with a focus on the United Kingdom, through combination of Boolean logical operators. Further details of our full search strategy translated across the electronic databases can be provided upon request. Additional searches were conducted through Google searches, and targeted websites and organisations such as the Sickle Cell Society, Cystic Fibrosis Trust and the Haemophilia Society. We also identified additional studies through citations or webpage links included in reports or articles identified through our searches. All records identified were screened based on their title and abstract. Full text screening of selected records was then performed based on our inclusion and exclusion criteria. Information was extracted on the author, year of publication, study design, setting, population and main findings, and categorised under recurring themes.

Evidence Set 2: Data analysis

2.1 Prevalence of the three conditions considered

To put the evidence presented in context, it is necessary first to define the number of people affected by sickle cell, cystic fibrosis and haemophilia in the UK. We extracted data from the National Haemoglobinopathy Register (NHR), Cystic fibrosis Registry (CFR) and National Haemophilia Database (NHD), focusing on reports for 2021-2022.[5-7] The NHR and NHD reports used were published in the financial year April 2021 to March 2022 while the 2021

CFR report spans from January 2021 to December 2021.[5-7] In Section 3.1, we also compare these data with the numbers of patients with sickle cell, cystic fibrosis and haemophilia in the NHS Hospital Episode Statistics (HES). The birth prevalence of sickle cell and cystic fibrosis was obtained from the 2018-2019 newborn screening programme and prevalence at birth of haemophilia was estimated using 2017 data from the NHD.[8, 9] In addition, the most recent estimates of median survival in 2022 were obtained from the CFR and NHD.[10, 11] However, nationally representative estimates of median survival for sickle cell are not available. We therefore used data from a 2016 single site study at King's College Hospital (London, UK).[12]

2.2 Selected comparative indicators

We collected data on seven comparative indicators, the choice of which was driven by previous research and the availability of publicly available data:

2.2.1 Number and value of successful grants from the three main UK public health research funders

We assessed research funding allocated to sickle cell, cystic fibrosis and haemophilia from the National Institute for Health & Care Research (NIHR), UK Research and Innovation (UKRI), and Wellcome. UKRI brings together multiple research councils including the Medical Research Council (MRC). We searched databases obtained from the research funders webpages for the period 1st January 2010 to 31st December 2023.

2.2.2 Financial resources available to dedicated national charities.

The charity commissions for England and Wales publishes financial annual reports for all registered charities. We searched their database (https://register-of-charities.charitycommission.gov.uk/) on 10th September 2024 using the search terms "sickle cell", "cystic fibrosis" and "haemophilia". We conducted a more in-depth analysis for the three main national charities: the UK Sickle Cell Society, the Cystic fibrosis Trust and the Haemophilia Society.[13-15] Annual financial reports are only available for the last five years, so we compiled data from the reporting years 2019 to 2023 and report the 5-year average and 95% CI.

2.2.3 Quality and completeness of disease registries

Descriptions of the NHR, CFR and NHD were searched to compare key characteristics of these national registries. We used a combination of information from annual reports, scientific publications and additional resources listed on the respective disease registry webpages.

2.2.4 Number of registered clinical trials

We searched the National Library of Medicine Clinical Trials database (from 1st January 2010 to 31st December 2023) and the EU Clinical Trials Register (1st January 2010 to 31st December 2020) to obtain information on all clinical trials for sickle cell, cystic fibrosis and haemophilia. Additionally, we restricted our search to focus on trials conducted in the UK. The status and protocol of UK Trials in the EU Clinical Trials Register have not updated since January 2021.

2.2.5 Number of drug approvals

We used the Medicines and Healthcare products Regulatory Agency (MHRA) and the National Institute of Clinical Excellence (NICE) specifically the NHS Indicative price from the British National Formulary (BNF) to search the number of drug approvals for sickle cell, cystic fibrosis and haemophilia. The NHS indicative price refers to the price the NHS pays for drugs and medicines and provides an indication of the relative cost. This price does not take into account VAT, professional fees and other overheads. We also used NICE guidance reports to determine the list price per treatment for gene therapy.

2.2.6 Number of scientific publications

We searched Pubmed, a free database of published scientific articles in the field of biomedical and life sciences, managed by the US National Library of Medicine. We used the following terms "sickle cell", "cystic fibrosis" and "haemophilia" as generic search terms. Although Pubmed includes publications from the 19th century, we restricted our study period to between 1st January 2010 and 31st December 2023.

2.2.7 Online disease awareness

We used data from Google Trends and Glimpse as a proxy of disease awareness of sickle cell, cystic fibrosis and haemophilia amongst the general public. To retrieve relevant information for all indicators, we used the following broad search terms for both databases: sickle cell, cystic fibrosis and haemophilia in the United Kingdom during the period of 1st January 2010 to 31st December 2023. The google trends database reports the relative search interest over time on a scale of 0 to 100 with 100 representing the peak interest in that search term for a given period and region. The Glimpse database reports the absolute search volume.

2.3 Routine hospital data analysis

We conducted a retrospective study of routine hospital data using the NHS Hospital Episode Statistics Admitted Patient Care (HES APC) database.[16] HES APC contains information on all hospital admissions to English NHS hospitals or to independent hospitals funded by the NHS. The dataset includes any episode requiring a hospital bed covering a period from 1989-90 onwards. Diagnoses are recorded using the International Classification of Diseases version 10 (ICD-10). To increase the specificity of our analyses, we included only individuals with a primary diagnosis for sickle cell, cystic fibrosis or haemophilia. We extracted data from all individuals of any age with the following ICD codes recorded between 1st January 2013 and 31st December 2022:

- **D57.0** for sickle cell with crisis;
- **D57.1** for sickle cell without crisis;
- **E84** for cystic fibrosis;
- **E84.0** for cystic fibrosis with pulmonary manifestations;
- **D66** for hereditary factor VIII deficiency;
- **D67** for hereditary factor IX deficiency.

We accounted for age (0-9, 10-19, 20-29, 30-39, 40-49, 50-59 and 60+); gender (men and women); ethnicity (White, South Asian, Black, Mixed, and other); region (Channel Islands, East Midlands, East of England, Greater London, Isle of Man, North East, North West, Northern Ireland, Scotland, South East, South West, Wales and West Midlands), and socio-economic status, through linkage of the lower layer super output areas (LSOAs) from the 2011 Office for National Statistics (ONS) Census to quintiles of the 2019 English Indices of Multiple Deprivation (IMD) - one representing the most deprived areas and five representing the least deprived. LSOAs are official geographies used in the UK. There were 33,755 LSOAs in England in the 2021 Census. They comprise between 400 and 1,200 households and have a usually resident population between 1,000 and 3,000 persons.

We considered four main outcomes in this study:

2.3.1 The number and proportion of hospital admissions

We calculated the proportion of individuals with 1, 2, 3, 4 or 5+ hospital admissions in a year for each condition stratified by type of admission, specifically emergency or elective (planned) admissions. Our elective admissions included regular day and night cases as well as ordinary and day cases. We calculated the total number of bed days per year for each condition by summing the total number of days individuals admitted to hospital in 2022. For admissions in which individuals stayed in hospital for less than a day, we counted it as half a day.

2.3.2 30-day emergency hospital readmissions

Individuals were defined as having a readmission if they had an emergency hospitalisation within 30 days of a previous hospital discharge. We calculated the proportion of individuals with 4 or more 30-day emergency re-hospitalisations in a period of 12 months.

2.3.3 The length of stay in days for emergency admissions

We focused on non-disease specific health complications affecting the general population such as acute appendicitis, long bone fracture and sepsis. A short stay was defined as one day. One day was added to admissions with a zero length of stay and all other stays were defined as long stays.

2.3.4 The average cost per hospitalisation per patient per year

We used the National Cost Collection Index from the financial years 2019/20 to 2021/22 to calculate the costs of elective and emergency hospitalisations to the NHS for sickle cell, cystic fibrosis and haemophilia in each year for each Hospital Resource Group (HRG).

We used the following descriptions:

- paediatric sickle cell anaemia with crisis,
- sickle cell anaemia with crisis
- sickle cell anaemia without crisis
- cystic fibrosis

- paediatric cystic fibrosis
- coagulation defect (for haemophilia)
- paediatric coagulation disorders (for haemophilia)

Costs for paediatric sickle cell anaemia without crisis were not reported in the data. For simplicity and consistency across the three conditions, individuals under 18 years were classified as paediatric admissions.

Evidence Set 3: Patient engagement

Here, we focused exclusively on people with sickle cell. To assess whether the data from the other two Evidence Sets accurately reflected the lived experiences of people with sickle cell, we created an online survey (see Appendix 1) in collaboration with patient representatives and members of the Sickle Cell Society. Based on previous input from patients, we ensured that the survey was concise and included a prize draw (£50 voucher) for all the participants who completed the survey. The anonymous online survey, set up in Qualtrics, launched on 29th November 2024 and closed on 20th December 2024. Summary statistics were then generated for all the questions and key themes identified from the open text comments. The survey was disseminated via social media platforms (LinkedIn, X [formerly known as twitter], patient and public voice groups such as the Northwest London Haemoglobin Coordinating Centre (HCC) Patient and Public Voice group, Imperial Sickle Cell Warriors, the Ethnicity and Health Unit and the NHS Race and Health Observatory. Individuals were eligible to participate if they were:

- living with sickle cell or caring for someone with sickle cell;
- aged 18 years and older;
- living in the United Kingdom.

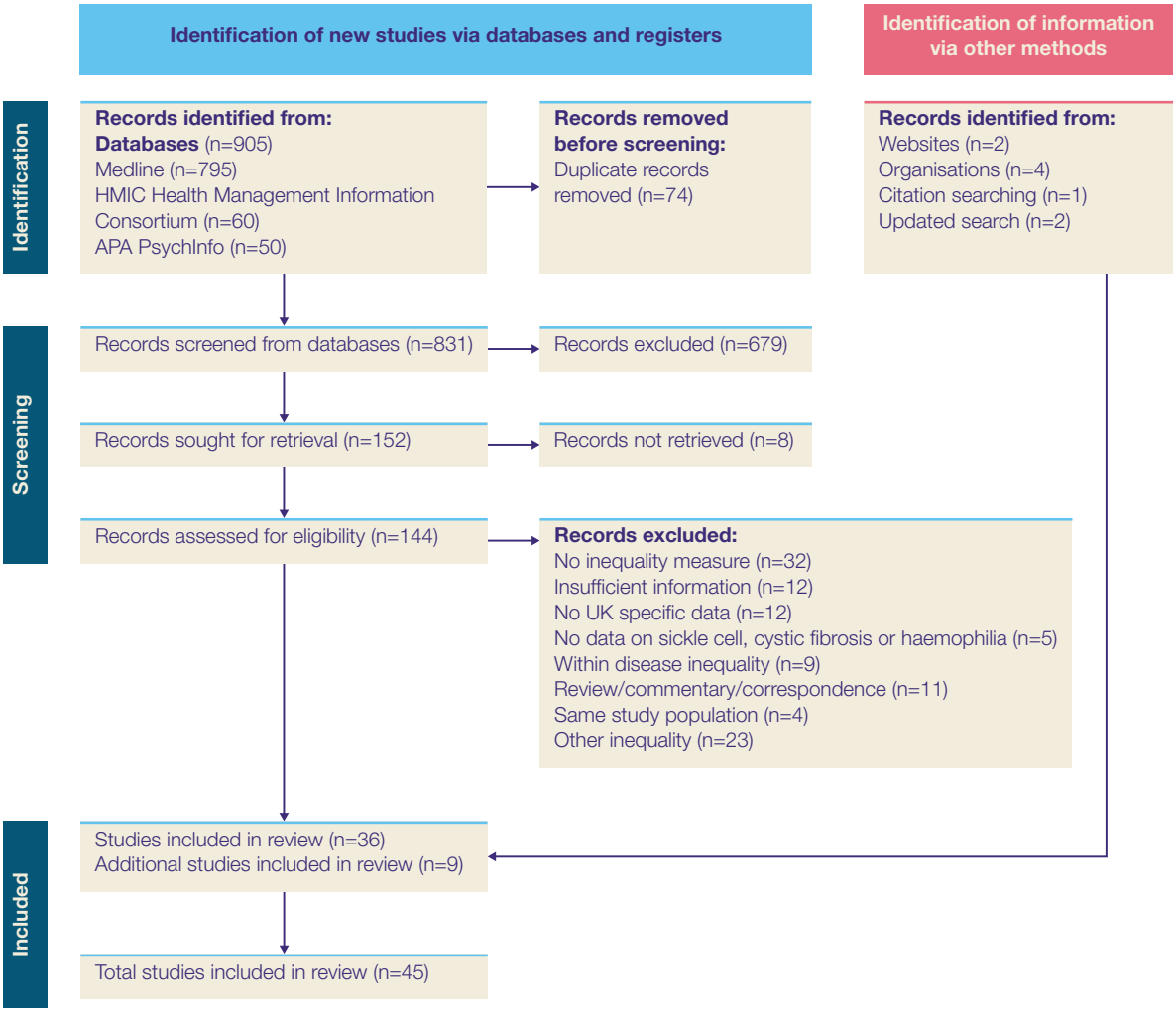
The results were analysed using a thematic analysis and grouped together responses that shared similar meanings.

Findings

Evidence Set 1: Literature Review

In total, we identified 905 records from our electronic database searches (Figure1). After removing 74 duplicates, we were left with 831 studies for title and abstract screening. Based on our inclusion and exclusion criteria, 152 studies were then selected for full text screening. Of these, 36 were eligible for inclusion into the final review. An additional nine records were identified from other sources. Therefore, in total 45 records were included into our final review.

Figure 1: Flowchart of the different inclusion and exclusion steps used in our literature searches.



After reviewing the information included in the 45 data sources identified, five key themes emerged: i) the sub-standard care often received by patients; ii) relatively poor disease outcomes; iii) a generally low disease awareness amongst healthcare professionals, partly due to inadequate training; iv) frequent negative attitudes towards patients, often driven by racism and stigmatisation; and v) a systemic lack of investment for the benefit of patients. We provide further details below of the evidence found through the literature on each of these themes. Most of the evidence presented relates to sickle cell in the UK, but, where possible, comparisons are made with cystic fibrosis and haemophilia, or with data from other countries.

1.1 Sub-standard care

People with sickle cell experience a range of barriers in accessing primary care services.

Access to primary care

- In a cross-sectional study in London (n=40), most sickle cell patients reported that they were not satisfied with the quality of care that they received from their general practitioner (GP).17

- Almost half of them (47%) reported that they did not use their GP to manage a painful sickle cell crisis and instead preferred to attend the emergency department.[17]

- Similarly, in a small study of parents of a child living with sickle cell, parents also reported preferring to go to the hospital instead of the GP due to the GP's lack of knowledge of sickle cell.[18]

- People with sickle cell report feeling ignored by GPs who lack comprehensive knowledge of sickle cell, particularly in terms of pain management. They felt that GPs were not actively interested or engaged in the progress and treatment of their condition.[19, 20]

- Sub-standard care also affects infants with sickle cell. In a cohort study of all babies born with sickle cell in England between 2010 and 2015, 20% of babies were not seen by a specialist by three months of age. The acceptable standard according to the NHS Sickle Cell and Thalassaemia screening programme is that 90% of babies should be seen by a specialist by three months of age. Regional differences were present as infants living in London were less likely to be seen by the GP within the 90-day target.[21]

- In comparison, GPs mentioned employing a holistic care approach for conditions with a large input from secondary care such as cystic fibrosis and believe that patients with cystic fibrosis who do not tend to see their GP often should be proactively followed up with.[22]

Disease management

- Pain management in sickle cell has been consistently reported as poorly managed in NHS settings.[23]

- Qualitative studies involving patients with sickle cell have shown that patients fail to receive pain relief in a timely manner. In a focus group and online survey, only 30% of adults and 42% of parents felt that pain relief was provided in a timely manner in their most recent emergency healthcare episode.[24]

- In a recent evaluation of haemoglobinopathy review programmes, long waiting times for pain relief for sickle cell patients were observed in emergency departments.

- Despite the 2012 NICE guidelines indicating that pain relief should be offered within 30 minutes of presentation with an acute painful sickle cell episode in hospital, nearly all centres reviewed did not meet this target.[25 26]

- In another qualitative study in England, participants reported experiencing a mean number of painful episodes in the last year of approximately seven in which they did not see a doctor. Patients who were admitted to A&E and those aged 18 years and older were less satisfied in terms of pain control.[27] Geographical differences have been observed in the UK and beyond. In a multi-country study, people with sickle cell in the UK had the highest annual mean hospitalisations for pain crises (mean of 2.98 hospitalisations per year) at nearly twice the rate for the United States.[28]

- Pain is also common in haemophilia. In a recent study, 59% of people with haemophilia reported experiencing frequent pain. In contrast with sickle cell, 70% felt well supported and listened to when speaking to clinicians about pain and 63% reported that discussing pain resulted in a referral to a physiotherapist.[29]

- Besides pain relief, evidence also suggests that people with sickle cell experience delays in other treatments. While the NHS Sickle Cell and Thalassaemia Screening Programme Standards 2011 recommend that 90% of babies are offered and prescribed Penicillin V or alternative by three months of age as an acceptable standard (with 99% as the achievable standard), only 80% of babies with sickle cell were prescribed penicillin by three months of age, so that 20% missed out on treatment.[21]

- Sickle cell patients also experience delays in surgical treatment. In a prospective clinical audit from five urgent and community clinics in the UK, 70% of patients who underwent hip arthroplasty for femoral head osteonecrosis experienced a delayed surgery of beyond 18 weeks.[30] In comparison, only 21% of sickle cell patients in Nigeria experienced delay in surgical management.[31] This delay in surgery impacts on the quality of life of patients and

these findings also suggest a lack of knowledge and experience of sickle cell among UK health care professionals in managing hip arthropathy or osteonecrosis in sickle cell.

1.2 Poor disease outcomes

Hospital admissions

- Sub-standard care in primary and secondary care services leads to poor health outcomes. Hospital admissions for sickle cell have risen by more than 50% over a 10-year period between 2001/02 and 2009/10 increasing from 21.2 per 100,000 to 33.5 per 100,000 with 74.9% of all sickle cell admissions in England occurring in London. [32]

- Over a 20-year period (1999-2019), hospitalisation rate for sickle cell increased by 1.95-fold.[33]

- The number of cystic fibrosis hospital admissions in Scotland also increased steadily over time between 1989 and 2009 with high 12-month re-admission rates of over 80%.[34]

- High hospital admissions in sickle cell have been associated with mortality. In a London study of sickle cell patients, a high hospital admission rate was associated with more than three-fold risk of death.[12]

- Another London study found that 74% of the total number of admissions were multiple admissions.[35]

- In a paediatric sickle cell population, rates of hospital admission had fallen from 111.3 admissions per 100 patient years to 41 admissions per 100 patient years over a 50-year period (1960-2010).[36]

Mortality

- In a study from King's College London Hospital, the median survival for sickle cell (HbSS/HbSβ0) was 67 years and the median survival for patients with high hospital admission rates was 60 years old which was significantly lower than that in patients with low admission rates.[12]

- The median age of death for sickle cell was reported as 47 years (interquartile range: 33-58) in a study covering a 10-year period between 2009 and 2018. The crude 10-year mortality rate was 5.3% in the entire cohort and a higher mortality rate of 25.3% was observed among those aged 50 years and older.[37]

- Survival in cystic fibrosis in England and Wales has improved substantially over time, with the median age of death increasing from age band 0-4 years to 25-29 years between 1959 and 2008.[38] This is supported by a paediatric London study which reported a small number of deaths in children with cystic fibrosis (11 out of 1,022) over a period of between 2000 and 2009.[39]

- Another study using the UK Cystic Fibrosis Registry found an overall trend of increasing age of death for all people with cystic fibrosis with the median age at death increasing from 25.0 years in 2007 to 29.0 years in 2010.[40] A more than 2-fold improvement in the age at death for cystic fibrosis was found between 1968 and 2009.[41] In the 2023 annual report of the UK Cystic Fibrosis Registry, the median age at death reported was 46.[42]

- Mortality rates for cystic fibrosis decreased annually by 2% between 2006 and 2015.[43]

- The median survival time for people with moderate haemophilia A and B between 1996 and 2023 was 78 and 77, respectively. [11]

1.3 Low awareness among healthcare professionals and inadequate training

- Patients consistently reported a lack of knowledge of sickle cell from healthcare professionals.[44]

- Sickle cell patients report that their GP has little or no knowledge of sickle cell,[20] with 55% of patients reporting that they did not visit their GP for general advice about their condition. 23% reported that they rarely visited their GP and 27% did not use their GP to manage a painful crisis and preferred to attend the emergency department instead.[17]

- Sickle cell patients have reported GPs being unreliable during a crisis and feeling ignored. In addition, they perceived that GPs were not interested in their progress or treatment and felt that GPs lacked comprehensive knowledge of sickle cell.[19]

- In a study of mothers of children with sickle cell, they reported preferring to go to the hospital instead of the GP due to GPs having poor knowledge of sickle cell.[18] They reported immediate access to care in a haematology department in a positive light.[18]

- Low awareness of sickle cell in A&E has often been reported, resulting in delays in receiving treatment and sub-standard care.[45] In a survey of 722 patients of sickle cell and their carers, respondents reported a lack of awareness from A&E staff which resulted in a poor experience of sickle cell care.

- Only 45% of respondents indicated that healthcare professionals in emergency care 'definitely knew enough about sickle cell, while 76% thought so in a planned healthcare setting.[24]

1.4 Negative attitudes towards patients

- Racial inequality has been identified as a key factor in the poor and sub-standard treatment reported by sickle cell patients. In the No-One's Listening report, patients reported multiple accounts of racism which included patients being called profanities by health care staff to staff assuming patients are "drug seekers".[45]

- In the NHS RHO Sickle Cell Digital Discovery report, patients also reported experiencing mistrust, lack of compassion and being perceived as 'drug seekers' by health care professionals.[46]

- In a qualitative study of pain management in sickle cell, 59% of patients in London reported at least one in-hospital concern-raising behaviour. More specifically, 39% reported disputes with staff, 20% were suspected or accused of analgesic misuse and 14% self-discharged from hospital.[47]

- 49% of sickle cell patients reported that healthcare professionals were not completely sympathetic and understanding in emergency care settings, while 32% felt the same in planned settings.[24]

- Participants in a qualitative study in the midlands regions of England reported experiencing racism when receiving healthcare. They reported unethical care and fears of receiving 'second class' treatment and being labelled as aggressive.[44]

- In other studies patients report not being satisfied in terms of respect, dignity and staff attitudes and behaviour.[27, 48]

- Institutional racism and stereotypes of ethnic minority patients having a lower pain threshold has been identified as a key issue that patients with sickle cell face which leads to inadequate care. [23]

- The Infected Blood Inquiry report included evidence of stigmatisation and indignities for patients with haemophilia infected with HIV and hepatitis viruses throughout the 1980s and 1990s.[49]

1.5 Inadequate investment

We found evidence of inadequate investment in staffing, research and novel treatments for sickle cell.

Understaffing

- Healthcare professionals have reported disproportionate understaffing in sickle cell services with considerable regional differences. Only 2 out of 10 regional sickle cell care networks were above minimum threshold for good standard of care with most being poorly staffed having one full-time specialist nurse for every 199.7 patients.[50]

- For cystic fibrosis, there was a median of 1.8 and 1.3 whole time equivalent (WTE) nursing staff per 75 patients in paediatric and adult care for cystic fibrosis, respectively.[51]

- Regional disparities have been reported in terms of staffing and funding with 6 out of 10 regional sickle cell care networks having a suboptimal nurse/patient ratio and other areas of the country not benefitted from funding.[50]

- In addition, insufficient training has been reported from numerous sources. Funding for specialist sickle cell nursing posts is perceived as harder to secure than other nursing positions.[50]

Research

- In a report of rare disease research across the UK from 2016 to 2021, cystic fibrosis received the second greatest number of awards (140 awards) funded by the Association of Medical Research Charities (AMRC). Sickle cell and haemophilia did not feature in the top 30 rare diseases awarded funding by AMRC members.[52] Cystic fibrosis was the top condition being researched by industry in the UK between 2016 and 2021 after being awarded over 25 projects during this period. Sickle cell ranked at number 9 and was awarded less than ten projects. Haemophilia was listed as one of the top conditions being researched in individual years but did not feature in the top 30 conditions.[52]

Lack of treatment options

- A lack of research into sickle cell can impact the availability of treatment options. hydroxyurea was the only medication approved for sickle cell until 2021. In October 2021, Crizanlizumab, indicated for preventing sickle cell crises, was recommended by

NICE as the first new therapy for sickle cell in 20 years.[53] However, in January 2024, the MHRA withdrew the marketing authorisation of Crizanlizumab.[54] A similar story happened shortly after with Voxelotor. Voxelotor, a sickle haemoglobin polymerisation inhibitor, was approved by the MHRA in 2022 and received NICE funding in May 2024.[55, 56] However, in September 2024, Voxelotor was suddenly withdrawn from the market.[56] Although such withdrawals are driven by poor efficacy or safety issues from clinical trials, these instances have further highlighted the lack of treatment options for sickle cell patients.

- Casgevy, which aims to cure sickle cell, was the first gene therapy approved by the MHRA in 2023.[57] The decision to make this groundbreaking treatment available to patients on the NHS has been announced in January 2025.[58] It is expected that about 50 people with sickle cell per year might receive this treatment on the NHS. Issues related to access of treatment and affordability are important to consider.[59]

- Breakthroughs in cystic fibrosis and haemophilia drug development have led to multiple treatment options including the recombinant factor VIII and factor IX concentrates, human DNAase, dornase alfa, and the novel cystic fibrosis transmembrane conductance regulator (CFTR) modulator, Ivacaftor, which was available on the NHS in 2016.[60, 61]

- In more recent years, the first gene therapy (Etranacogene Dezaparvovec) and monoclonal antibody therapy for haemophilia (Emicizumab) were approved by the MHRA in 2021 and 2023, respectively, and other CFTR modulators such as Kaftrio, Symkevi and Orkambi recommended for use on the NHS in 2024.[62, 63]

Evidence Set 2: Data analysis

2.1 Prevalence of sickle cell, cystic fibrosis & haemophilia

Prevalence

As previously documented, there is no reliable estimate of the total number of people living with sickle cell in the UK, to support the commissioning and provision of services for people with sickle cell.[1] The following number of patients were reported as registered in the NHR, CFR and NHD between 2021 and 2022 respectively: 15,841, 10,908, and 7,774. These numbers depend on the completeness of the databases. To our knowledge, there is currently no data available on the representativeness of the NHR. Claims that the CFR covers 99% of people with cystic fibrosis are often cited, but it is difficult to know how close this figure is to the reality.

Birth prevalence

Sickle cell and cystic fibrosis are both included in the universal newborn screening in the UK. As a result, there are comprehensive data on the birth prevalence of these conditions. The screen positive rate in 2019 was comparable for sickle cell and cystic fibrosis (4.08 and 4.17 per 10,000 births, respectively). The prevalence of haemophilia at birth estimated from the NHD in 2017 was 2.6 per 10,000 male births for haemophilia A, and 0.5 per 10,000 male births for haemophilia B.

Survival

Nationally representative estimates on survival showed that haemophilia had the greatest median survival at 77 and 79 years of age for severe haemophilia A and B, respectively, while the median survival for cystic fibrosis was lower at 56 years. In a cohort study from a single hospital with excellent specialist services in London, the median survival for sickle cell was 67 years.

2.2 Selected comparative indicators

2.2.1 Number and value of successful grants from the three main UK public health research funders

In total, there were 387 projects awarded by NIHR, UKRI and Wellcome for the three conditions considered between 2010 and 2023 (Figure 1). Out of these, 73.6% were awarded on cystic fibrosis, 19.4% to sickle cell and 7.0% to haemophilia. Total research funding to all three conditions amounted to £182,448,399 over our 14-year study period, of which £107,323,136 were devoted to cystic fibrosis (58.8%), £40,798,305 (22.4%) to sickle cell and £34,326,958 (18.8%) to haemophilia.

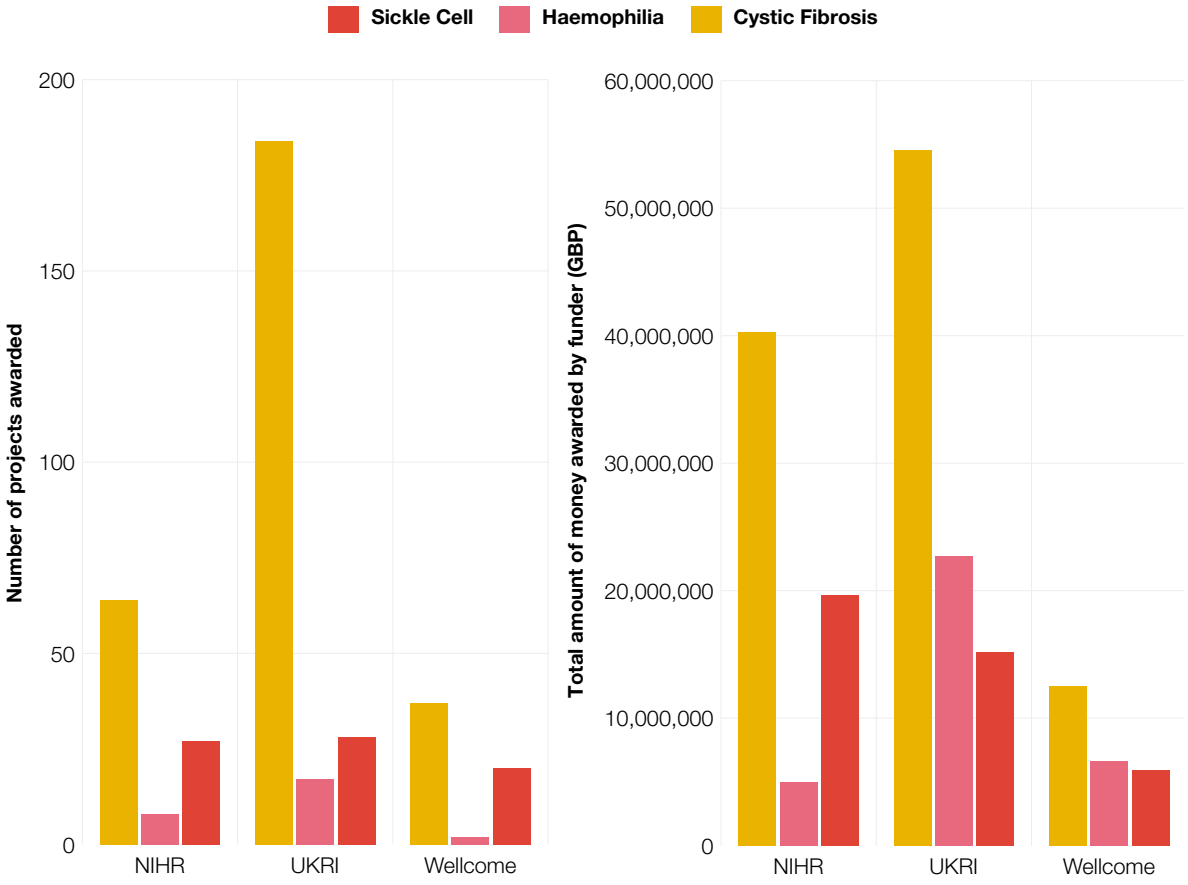


Figure 1. A. The number of projects awarded on cystic fibrosis, haemophilia and sickle cell by the National Institute for Health and Care Research (NIHR), UK Research and Innovation (UKRI) and Wellcome between 2010-2024. B. Total amount of money awarded by NIHR, UKRI and Wellcome to projects on these three conditions between 2010-2024.

Based on the number of patients reported above, we calculated the average annual research funding per person for individuals with one of the three conditions considered. This amount

was lowest for sickle cell at £184 (95% CI: £172 - £196), compared with £315 (95% CI: £226 - £404) per person with haemophilia and £703 (95% CI: £697 - £709) per person with cystic fibrosis.

2.2.2 Financial resources available to dedicated national charities

In total, there were 53, 27 and 8 registered charities in September 2024 dedicated to sickle cell, cystic fibrosis and haemophilia, respectively. The average funding per person from the last recorded funds of the charities in total was £137 for sickle cell, £1,422 for cystic fibrosis, and £132 for haemophilia. Over a five-year period, the average annual funding for the Sickle Cell Society, the Cystic Fibrosis Trust and the Haemophilia Society were: £761,042 (95% CI: £680,038 - £842,046), £15,805,800 (95% CI: £14,589,270 - £17,022,330) and £909,820 (95% CI: £726,975 - £1,092,666), respectively. Accounting for the number of people with these conditions, the funding per person was: £1,449 (95% CI: £1,337 - £1,561) for cystic fibrosis, £48 (95% CI, £43 - £53) for sickle cell, and £117 (95% CI: £94 - £141) for haemophilia.

2.2.3 Quality and completeness of disease registries

Table 2 shows a comparison of the NHR, CFR and NHD. There are similarities in the activities of these three registries in terms of service commissioning and disease monitoring. However, there are key differences in the funding, data collected and uptake. The NHR is estimated to only include 70% of people with sickle cell while the CFR reports over 99% of people with cystic fibrosis consenting to their data being submitted to the registry.[1, 4] Although the NHD reports that it is designed to be as inclusive as possible, we could not find any information about the proportion of people with haemophilia included in their database.

Table 2. Comparison of disease registries for sickle cell, cystic fibrosis and haemophilia

	UK National Haemoglobinopathy Registry (NHR)[64]	UK Cystic Fibrosis Registry (CFR) [65]	National Haemophilia Database (NHD)[66]
Conditions Monitored	Sickle cell disease, thalassaemia syndromes and rare inherited anaemias [5]	Cystic Fibrosis [4]	Haemophilia A/B, Von Willebrand Disease and other bleeding disorders [7]
Inception	Commissioned by the Department of Health and Social Care in 2008 [1, 67]	Established in 1995 at the University of Dundee, Scotland [4]	Established in 1968 by the Department of Health in the UK [68]
Uptake	Register estimated to contain 70% of patients with sickle cell [1]	Over 99% people consent to their data being submitted to Registry [65]	
Sources of funding	Public Health England; specialist commission – since 2013	Sponsored and hosted by Cystic Fibrosis Trust [6]	Mainly funded by NHS and receives additional funding from pharmaceutical industry [69]
Activities	Support NHS England in service commissioning. Supports national sickle cell care networks by holding patient information, guidelines and protocols, education materials and national sickle cell disease information	Commissioning care. Pharmacovigilance, drug safety and efficacy, support pandemic response [4]	Disease monitoring, healthcare planning, funding, research, pharmacovigilance, drug safety and efficacy.[69] Commissioning of services. Epidemiology and clinical care. Surveillance and Safety. Quality of care
Steering committee	Steering committee with patient representatives and stakeholders (commissioners, clinicians, patient societies, HCC data manager) [5, 70]	Includes patient representative, clinicians, England, Wales and Scotland Commissioner, Centre and Registry data managers [10]	Yes
Sources of Data	Haemoglobinopathy coordinating centres, Newborn screening program and Newborn outcomes project. NHS Blood and Transplant.	Newborn screening program centres and outcomes project [10]	Local Haemophilia centres. Haemtrack to monitor therapy and bleeding symptoms. Direct at home patient entry via electronic system.
Data Input	NHS system encrypted database	NHS employees at specialist centres via online encrypted web system	Data input into encrypted NHS network by NHS employees at Specialist haemophilia centres. Patient-held on-line or mobile system to collect individual data from patients
Data collected	Demographic, diagnosis, hospital admissions, annual Reviews, complications, therapies, iron chelation, transfusion and Vaccination status	Demographic, annual reviews, incidence and new registrations, therapies, complication rates and respiratory function [6]	Demographic, natural history of disease, pharmacovigilance data about inhibitors - Prospective data collection for inhibitor risk factors.
International Network/Registry coordination?	No	European CF registry	Yes

2.2.4 Number of registered clinical trials

In total, there were 2,625 clinical trials registered in the National Library of Medicine, including for 29.1% for sickle cell, 45.9% for cystic fibrosis, and 25.0% for haemophilia, globally. Of the clinical trials conducted in the UK, sickle cell accounted for the lowest proportion of trials, comprising 14.3% (n=58) and 15.3% (n=41) of trials in the National Library of Medicine Clinical trials and EU clinical trials database, respectively, compared to 50.9% and 44.4% for cystic fibrosis (n=206 and 119) and 34.8% and 40.3% for haemophilia (n=141 and n=108).

2.2.5 Number of drug approvals

Sickle cell has the lowest number of approved drugs (n=2) while the number of approved drugs with NHS funding was higher for both cystic fibrosis (n=7) and haemophilia (n=7).

In terms of costs, the maximum NHS indicative price listed in the BNF was highest for haemophilia at £12,076 (Emicizumab, Hemilibra) compared to £8,346 for cystic fibrosis (Ivacaftor with Tezacaftor and Elexacaftor, Kaftrio) and £500 for sickle cell (Hydroxycarbamide, Siklos).

There is currently no gene therapy approved for cystic fibrosis, however, gene therapies for haemophilia and sickle cell are currently available for NHS funding with an official list price of £2,600,000 per treatment and £1,651,000 per course of treatment, respectively.

2.2.6 Number of scientific publications

Over the 14-year study period, there was almost double the number of research articles in PubMed on cystic fibrosis (n=34,594), compared with sickle cell (n=18,757) and haemophilia (n=16,560) (Figure 2). The number of publications on cystic fibrosis was roughly three-fold that of those on sickle cell and haemophilia in 2010, which reduced to a two-fold difference by 2023. The increase in publications on haemophilia during our study period appeared lower than for the other two diseases.

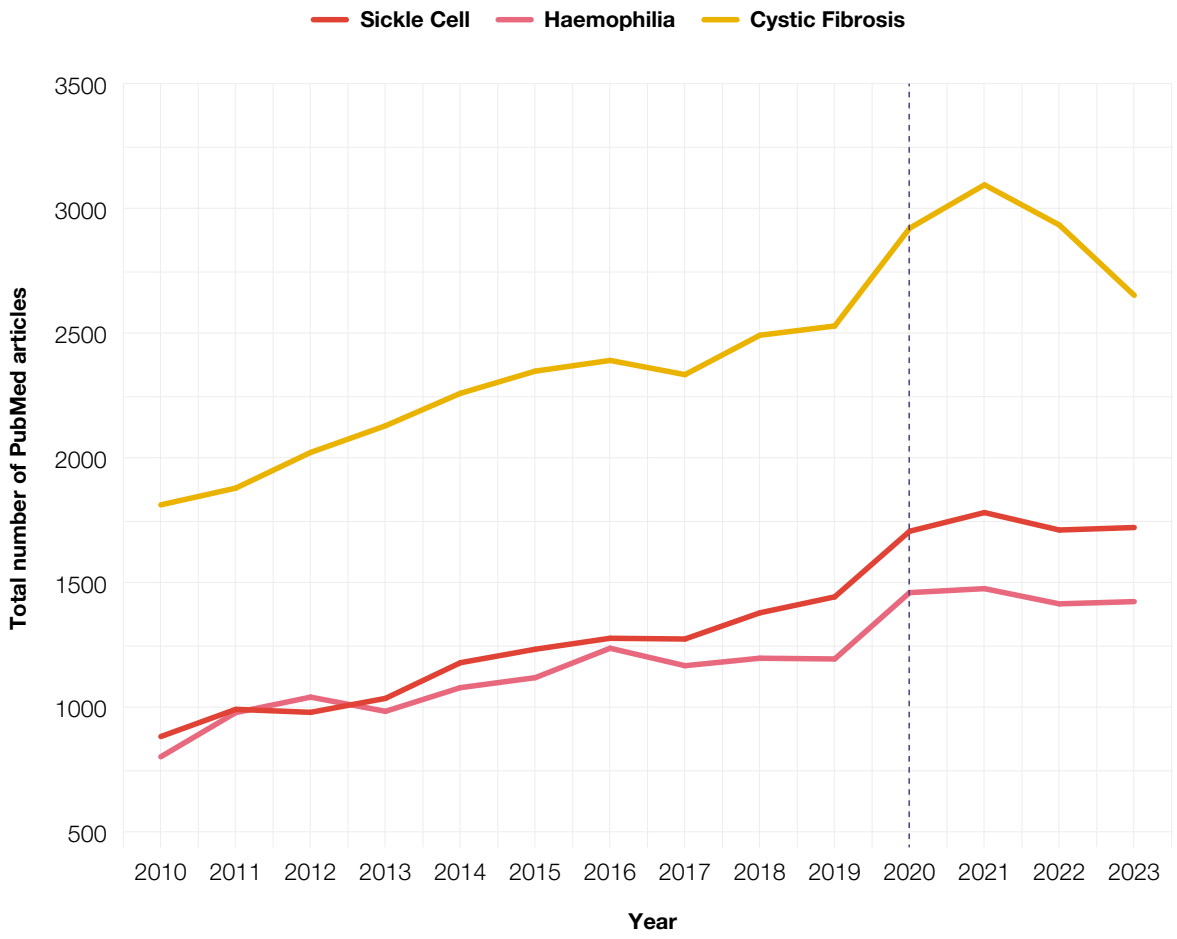


Figure 2. Total number of PubMed articles retrieved from a search of the following search terms, "sickle cell", "cystic fibrosis" and "haemophilia or hemophilia". Articles search between 1st January 2010 and 31 December 2023. The dashed line represents January 2020 indicating the start of the Covid-19 pandemic.

2.2.7 Online disease awareness

The absolute volume of Google searches over our study period was greater for sickle cell (n=23,962,462) compared to cystic fibrosis (n=15,961,489) and haemophilia (n=2,764,486). Figure 3 illustrates the relative interest in Google searches for sickle cell, cystic fibrosis and haemophilia over the same period. The relative interest in Google searches was highest for cystic fibrosis over time and lowest for haemophilia. Relative interest for sickle cell increased from the COVID-19 pandemic period. The biggest spikes in searches were seen in April 2021 and April 2022 for sickle cell and cystic fibrosis, respectively, likely coinciding with the high-profile deaths of individuals living with these conditions.[71,72]

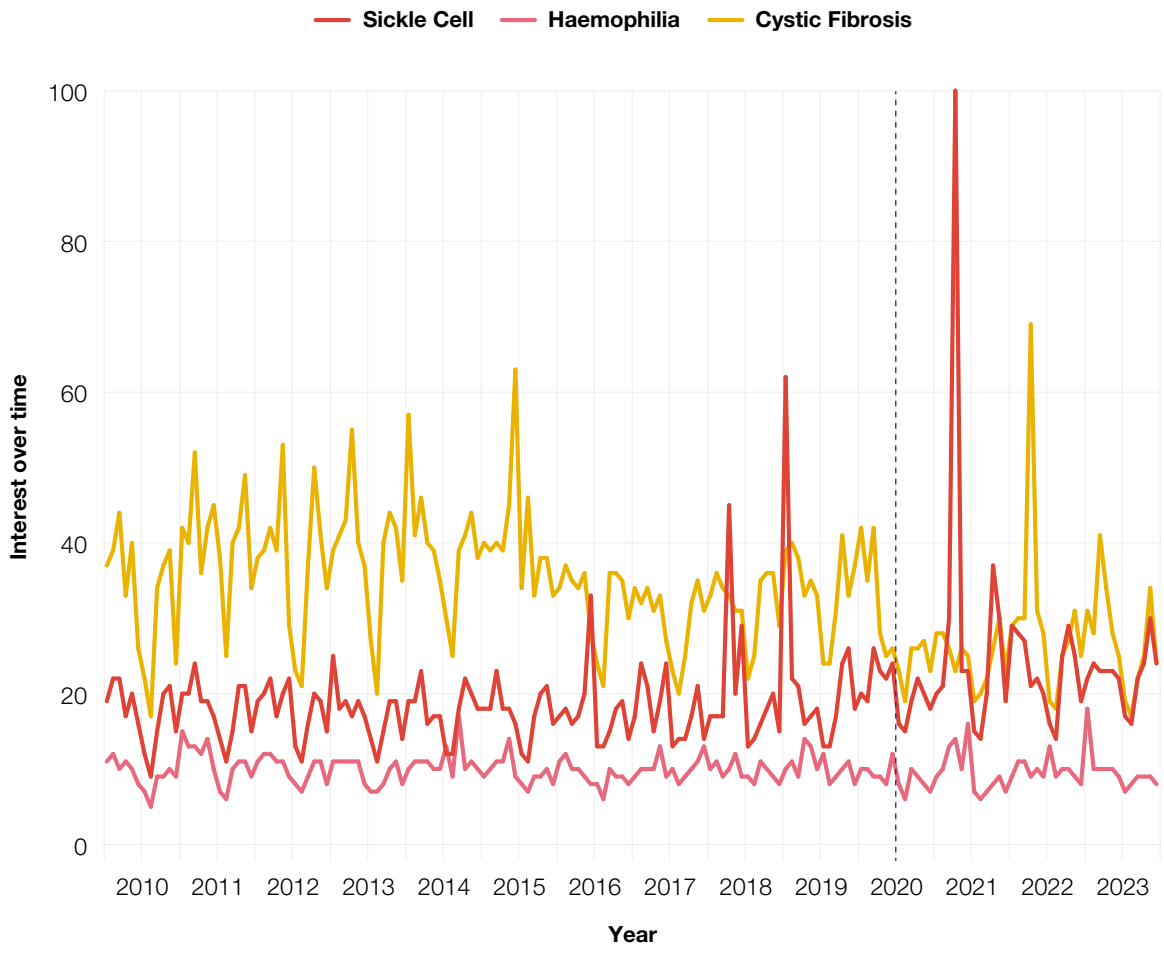


Figure 3. Interest over time in Google searches in the UK between 1st January 2010 and 31st December 2023. Google search interest for sickle cell, cystic fibrosis and haemophilia relative to the highest point on the chart in the UK between January 2010 and December 2023. Interest over time represents search interest relative to the highest point on the chart for the given region and time. A value of 100 is the peak "popularity" for the term. The main peaks in this figure are depicted by letters and correspond to the following: A) a value of 64 for search interest for cystic fibrosis in June 2015, B) a value of 59 for search interest for sickle cell in January 2019 C) 100 representing peak "popularity" for sickle cell in April 2021 and D) a value of 71 for cystic fibrosis search interest in April 2022. The dashed line represents January 2020 indicating the start of the Covid-19 pandemic.

2.3 Routine hospital data analysis

In total, between January 2013 and December 2022, there were 19,506, 9,569 and 7,289 individuals with a primary diagnosis of sickle cell, cystic fibrosis, and haemophilia respectively in the NHS Hospital Episode Statistics dataset (Table 3).

Key characteristics of people with these three conditions are:

- Age: Individuals with cystic fibrosis were the youngest.
- Gender: Individuals with haemophilia included in our study were male. We excluded females to avoid including carriers.
- Ethnicity: Most people with sickle cell were of Black ethnicity while those with cystic fibrosis and haemophilia were predominantly in the White ethnic groups.
- Socio-economics status: A greater proportion of individuals with sickle cell were in the most deprived socio-economic group.
- Geographical distribution: Individuals with sickle cell and haemophilia largely resided in Greater London while a greater proportion of individuals with cystic fibrosis lived in the Northwest of England.

Compared with individuals with a primary diagnosis for the conditions considered, individuals who had a secondary diagnosis code for sickle cell, cystic fibrosis or haemophilia, a greater proportion of individuals aged 60 years and older were represented for cystic fibrosis (28.2% vs 2.1%) and a lower proportion of people with sickle cell from Black ethnicity (57.4% vs 77.1%).

Table 3. Characteristics of hospital admissions for patients with a primary diagnosis of sickle cell, cystic fibrosis or haemophilia between 2013-2022. IQR: Interquartile range. *For data protection, table cells containing fewer than 10 individuals were recorded with an asterisk.

	Sickle Cell	Cystic fibrosis	Haemophilia
Total number of patients	19,506	9,569	7,289
Median Age (years, IQR)	27.0 (12.0 to 40.0)	18.0 (7.0 to 29.0)	49.0 (25.0 to 59.0)
Mean Age (years)	27.4 (17.9)	19.9 (15.8)	42.9 (23.2)
Age group (years)			
0 to 9	4,335 (22.2)	2,982 (31.2)	1,019 (14.0)
10 to 19	3,302 (16.9)	2,376 (24.8)	548 (7.5)
20 to 29	3,662 (18.8)	2,099 (21.9)	618 (8.5)
30 to 39	3,404 (17.5)	1,164 (12.2)	554 (7.6)
40 to 49	2,224 (11.4)	545 (5.7)	1,136 (15.6)
50 to 59	2,122 (10.9)	199 (2.1)	1,902 (26.1)
60+	457 (2.3)	204 (2.1)	1,512 (20.7)
Gender			
Men	9,219 (47.3)	5,006 (52.3)	7,289 (100.0)
Women	10,287 (52.7)	4,563 (47.7)	NA
Ethnicity			
White	693 (3.6)	8,551 (89.4)	6,071 (83.3)
South Asian	205 (1.1)	355 (3.7)	335 (4.6)
Black	15,040 (77.1)	35 (0.4)	174 (2.4)
Mixed	710 (3.6)	111 (1.2)	124 (1.7)
Other	1,410 (7.2)	162 (1.7)	278 (3.8)
Missing	1,448 (7.4)	355 (3.7)	307 (4.2)

	Sickle Cell	Cystic fibrosis	Haemophilia
Deprivation			
1 - Most deprived	4,943 (25.3)	1,864 (19.5)	796 (10.9)
2	4,299 (22.0)	1,722 (18.0)	950 (13.0)
3	2,020 (10.4)	1,637 (17.1)	841 (11.5)
4	1,115 (5.7)	1,498 (15.7)	805 (11.0)
5 - least deprived	627 (3.2)	1,409 (14.7)	732 (10.0)
Missing	6,502 (33.3)	1,439 (15.0)	3,165 (43.4)
Region			
Channel Islands	*	*	12 (0.2)
East Midlands	419 (2.1)	727 (7.6)	270 (3.7)
East of England	533 (2.7)	784 (8.2)	414 (5.7)
Greater London	7,230 (37.1)	1,124 (11.7)	929 (12.7)
Isle of Man	*	*	*
North East	383 (2.0)	814 (8.5)	398 (5.5)
North West	1,086 (5.6)	1,388 (14.5)	653 (9.0)
Northern Ireland	*	15 (0.2)	*
Scotland	10 (0.1)	75 (0.8)	*
South East	995 (5.1)	1,244 (13.0)	526 (7.2)
South West	310 (1.6)	873 (9.1)	473 (6.5)
Wales	25 (0.1)	167 (1.7)	41 (0.6)
West Midlands	1,618 (8.3)	955 (10.0)	390 (5.4)
Missing	6,893 (35.3)	1,386 (14.5)	3,168 (43.5)

2.3.1 The number and proportion of hospital admissions

The total number of annual hospital admissions for sickle cell increased by 42% from 30,194 in 2013 to 42,934 in 2022. During the same period, annual admissions for haemophilia increased by 21%, while they decreased by 41% for cystic fibrosis. This decrease is mostly due to a sharp decline in 2020, which coincides with the start of the Covid-19 pandemic.

2.3.2 30-day emergency hospital readmissions

There was a higher proportion of patients with sickle cell (9.0%) and cystic fibrosis (7.5%) with four or more 30-day emergency readmissions in a 12-month period compared with haemophilia (3.2%). When we stratified by age group (Figure 4), we found that the 10-19 and 20-29 age groups had the highest proportion of individuals with emergency readmissions at 13.2% and 11.6% for sickle cell and 8.4% and 9.8% for cystic fibrosis, respectively. The highest proportion of readmissions for haemophilia (9.5%) was in the 0-9 age group (compared to 9.0% for sickle cell and 6.2% for cystic fibrosis).

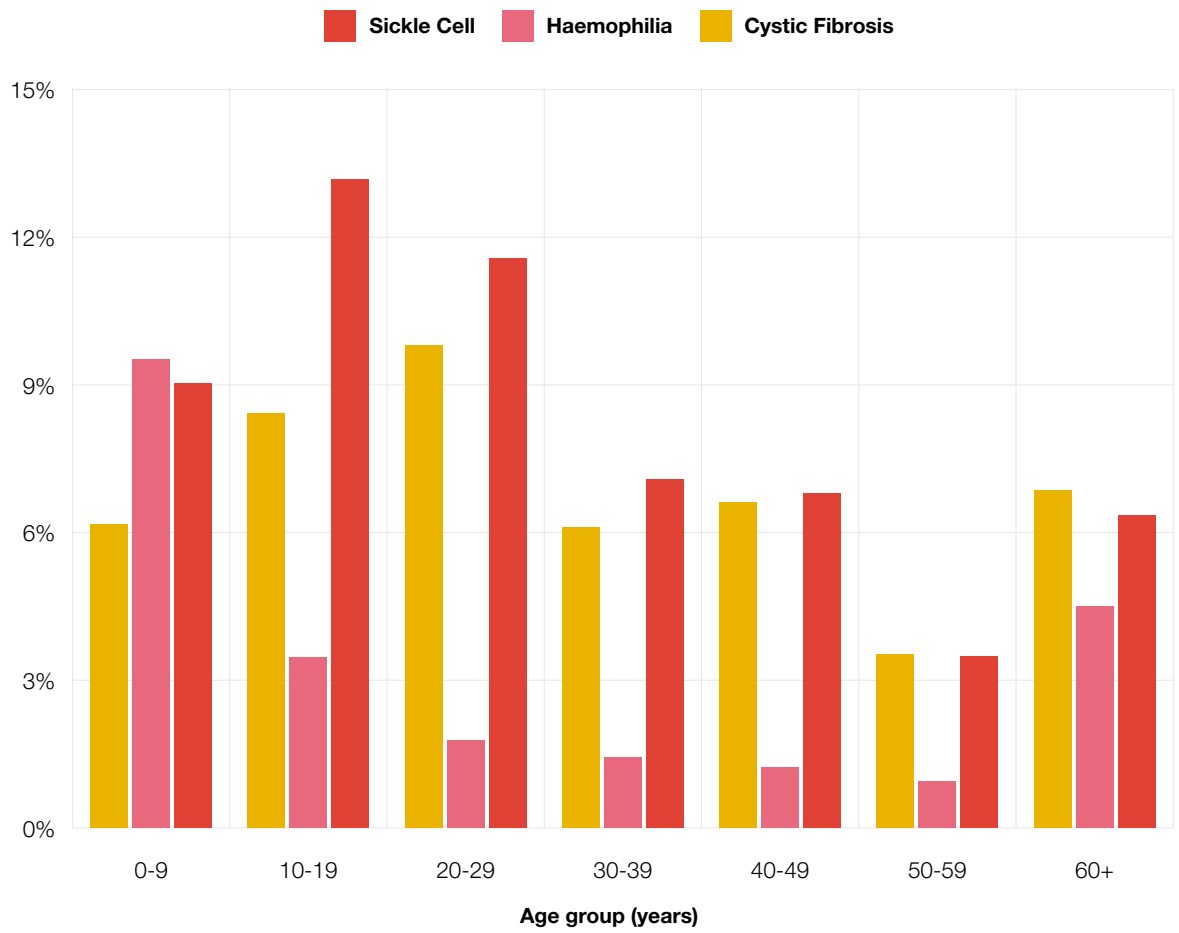


Figure 4. Proportion of individuals with four or more emergency readmissions in a 12-month period, stratified by age

2.3.3 The length of stay in days for emergency admissions

In our sub-cohort of 2,722 patients with emergency hospital admissions for a long bone fracture, sepsis or acute appendicitis, the median length of stay for appendicitis was similar over time for sickle cell, cystic fibrosis and haemophilia (Figure 5). There was greater variability in length of stay for fractures than appendicitis though the overall length of stay was similar for all three conditions. The overall length of stay for sepsis was shorter for people with haemophilia (median: 9.5 days) and sickle cell (median: 9.5 days) compared to cystic fibrosis (median: 13.0 days).

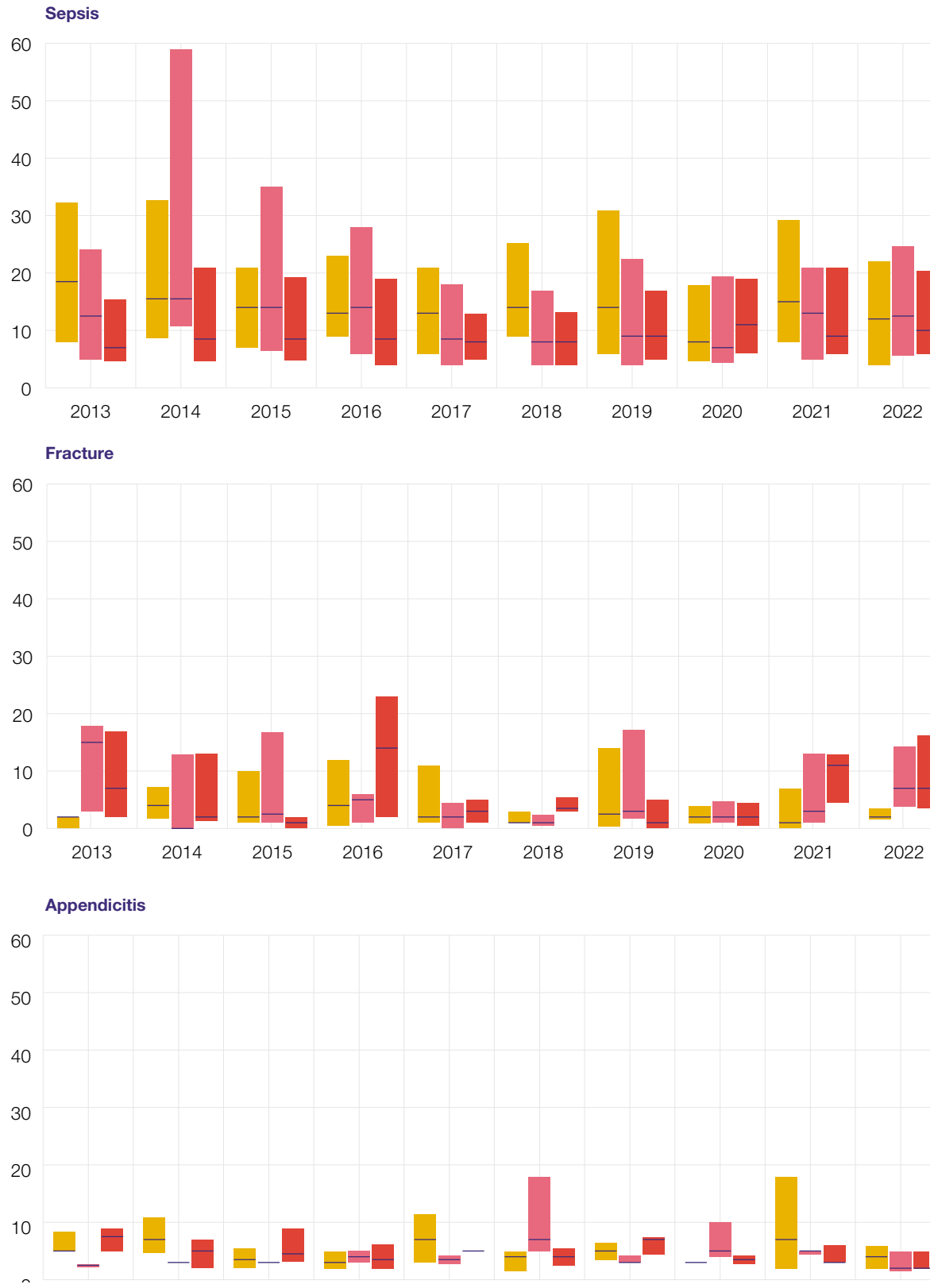


Figure 5. Length of hospital stay among those with sickle cell, cystic fibrosis or haemophilia with an emergency hospital admission for acute appendicitis, long bone fracture and sepsis. The total number of hospital admissions for appendicitis, fracture and sepsis was 53, 87 and

1,580, respectively, for people with sickle cell; 68, 106 and 463 hospitalisations had these reasons for admission respectively among people with cystic fibrosis; and there were 29, 100 and 275 hospital admissions among people with haemophilia for appendicitis, fracture and sepsis, respectively.

2.3.4 Annual hospitalisation costs

On average over the three financial years studied (2019-2020, 2020-2021 and 2021-2022), overall estimated hospitalisation costs were almost two-fold higher for sickle cell (£97,268,969) than for cystic fibrosis (£49,367,091) and nine-fold higher compared to haemophilia (£10,458,169).

Annual estimated costs decreased for cystic fibrosis, primarily driven by paediatric reductions (37% reduction), while they have substantially increased for sickle cell in adults (+42%) and haemophilia in both children (+111%) and adults (+63%).

Hospitalisation estimated costs for adults with sickle cell were more than 10-fold higher than for children, while they were roughly similar for paediatric and adult hospitalisation for cystic fibrosis (Table 4). Similarly, costs were substantially higher for adults with haemophilia compared to children.

Table 4. Costs of hospitalisation per patient per year for the three financial years. SCA: Sickle cell anaemia

		Total Costs			
Age group	**Condition**	**2019-20**	**2020-21**	**2021-22**	**Average**
Paediatric	**SCA with crisis**	£7,841,868	£7,474,950	£8,331,418	£7,882,745
	Cystic fibrosis	£27,838,367	£23,081,649	£17,624,635	£22,848,217
	Haemophilia	£1,764,897	£1,173,362	£3,721,262	£2,219,840
Adult	**SCA**	£71,523,304	£95,031,909	£101,603,459	£89,386,224
	with crisis	£35,080,019	£41,375,419	£45,882,963	£40,779,467
	without crisis	£36,443,285	£53,656,490	£55,720,496	£48,606,757
	Cystic fibrosis	£27,441,615	£25,534,542	£26,580,466	£26,518,874
	Haemophilia	£6,539,697	£7,484,288	£10,637,511	£8,220,499

Evidence Set 3: Patient engagement

In total, we gathered responses from 73 participants. Out of these, 62 (85%) had sickle cell while 11 (15%) were caregivers of people with sickle cell. 53 (73%) of respondents were female and 40 (55%) respondents lived in London.

The main themes covered in the responses mirrored some of the evidence presented in our literature review (Evidence Set 1). 59 (81%) people somewhat or strongly agreed that sickle cell is neglected. Four themes were identified from the subsequent question asking participants to explain why they thought sickle cell is or is not neglected: i) poor care; ii) negative attitudes; iii) lack of knowledge and awareness of sickle cell; and iv) inadequate investments. We include examples of citations to better reflect the lived experiences of patients and their caregivers. Some responses went beyond identifying shortcomings in care and suggested possible solutions.

3.1 Poor care

Amongst our participants, 42 (68%) reported experiencing unfair treatment within the last 12 months and 30% of carers felt dismissed within the last 12 months. Experiences of unfair treatment were centred around perceived inadequate care particularly in terms of pain management. The subthemes identified were: i) long waiting times to receive care; and ii) negative attitudes from healthcare professionals.

NICE guidance recommends that analgesia should be offered to people presenting with an acute sickle cell episode within 30 minutes of presentation in secondary care.[25] Respondents reported being often ignored in A&E when in pain and left for hours without care.

"I was crying in pain and the nurses were just laughing. They just shut the curtain."

"I had to plead to be allowed to have my regular blood transfusion at a specialist sickle cell centre and because of where I live I only get to see the sickle specialist once a year, the rest of the time I have to see a local haematologist who knows nothing about SCD, so can't make treatment decisions."

"Many areas of hospitals are not able to deal with sickle cell and this causes negative or at times devastating affects for patients and their quality of care."

"The care provided is not up to scratch and varied across the country. Sometimes it feels like a war zone."

"Being told I am not in pain, because I don't look like it."

"Up until about two years ago, I was never under a sickle cell team. The number of hospitals that have a haematology department in the UK is abysmal, even more so, the number of hospitals that do not encourage taking a sickle patient under their care."

3.2 Negative attitudes

The poor management of pain in sickle cell is related to negative attitudes from healthcare professionals towards people with sickle cell including ignoring patients, stereotyping patients as 'drug addicts' and laughing or mocking people with sickle cell. There were several mentions of racism and of staff refusing to provide adequate pain management or treatment, or of staff not believing reports of pain from people with sickle cell are experiencing and even mocking them.

"Sickle cell is neglected because it is suffered by the minority; hence the government and pharmaceutical companies are complacent about it, compared to cancer."

"Due to the colour of the patients skin."

"Racial disparity is enormous compared to other chronic health issues. Sickle cell is overlooked and under funded as a baseline."

"The NHS love to promote "patient centred care" but only applies to patients with "severe" conditions like cancer, diabetes or MS. Because sickle cell is mainly a "black" condition, our "centred care" only applies when it suits them."

"It's neglected possibly because it only affects minorities - Africans, Indians etc. it's not a "white" person's disease."

"There are also many nurses who believe that because we have a lot of pain and require high doses of painkillers, that we are addicts."

"I was really sick for almost a year always in hospital and he doesn't basically listen to me or what to hear my opinion he does what he likes no doctor patients rapport and he actually called me drug seeking."

"Less judgemental medical professionals, not assuming they just want drugs because they are addicted to pain meds. The pain is real!"

"I presented at A&E and was given Oramorph. Was discharged. Two days later when the medication wore off I went back and was told I was a drug addict and only coming in for morphine"

"Waiting 6hrs in A&E to be seen and doctors saying they can't do anything until they speak to a haematologist but I explained that they can at least prescribe some painkillers while waiting and it was ignored, whispering outside to other colleagues saying be careful it could be a drug seeking case."

"I have numerous situations. But I am been blatantly told by a medical professional that sickle cell patients are here for the painkillers. They use their image of us to then mistreat us by ignoring call bells when on the ward, or being demeaning and showing zero sympathy when in pain by simply telling us to calm down that there are other patients. This is clearly them assuming we just choose to put in a show by crying and screaming in pain."

3.3 Lack of knowledge

Participants reported a lack of knowledge and awareness of sickle cell from healthcare professionals and the public. Participants made references to the people having more knowledge of other conditions compared to sickle cell.

When asked what could be done to ensure people with sickle cell are treated in a fairer way when receiving care, key strategies suggested were centered around wider education and training for healthcare professionals including GPs and hospital staff to increase more knowledge and awareness of sickle cell nationally and for better recognition and management of a sickle cell crisis. Further suggestions on education included a patient led education programme mandatory for everyone associated with sickle cell including paramedics and A&E receptionists and regular training. Other suggestions included listening to patients and showing empathy, a better understanding of the pain experienced by patients, more funding into sickle cell services, greater support in the community and social care, a sub-ward for

sickle cell patients, greater mental health support, improve staffing, free prescriptions, all hospitals to have a unique and clear protocol/pathway for administering care to sickle cell patients, and to provide patients with a care plan and follow agreed care plan.

3.4 Inadequate investments

Participants reported a lack of funding as a reason why they thought that sickle cell is neglected. Specifically, participants mentioned the lack of new therapeutic options and lack of funding for existing novel therapies. The lack of free prescriptions was also cited as a reason for the perceived neglect of sickle cell.

Discussion

The evidence presented in this report is complex and only reflects some of the inequalities faced by patients on a day-to-day basis. Inevitably, some of this evidence mirror previous reports and publications on these inequalities, but we believe that most of the content of this report is novel. The complexity of such an analysis is reflected in what could first appear as contradictions in our findings. For example, while we found evidence that patients with sickle cell tend to manage many of their complications at home, the average number of hospitalisations for pain crisis was found to be the highest in England in a multi-country study.[73] Further work is required to fully understand these complexities.

Below, we link the different pieces of data presented in our three Evidence Sets to guide our recommendations. We identified 8 main themes, which align well with the global recommendations of the recent Lancet Haematology Commission on Sickle Cell Disease:[74]

1. There is a striking lack of reliable epidemiological information on the number and geographical distribution of patients, particularly those with sickle cell, in the UK. This is essential to plan and deliver the specialist care required by patients with such severe chronic conditions. The lack of nationally representative data on the survival of patients with sickle cell is an important gap which should be addressed. NHS data, complemented by information from the NHR and the Office for National Statistics (ONS), offer a unique resource in the world to support this, and have the potential to efficiently monitor changes in the number of patients, their characteristics and their needs. The various initiatives from the Cystic Fibrosis Trust to support researchers in using and accessing data from the CFR provide an excellent illustration of the benefits of having high quality data and ensuring that it is used for research and policies.

2. Our data suggests that there is a clear lack of dedicated research funding for sickle cell in the UK. It is likely that this is contributing to: the poor awareness of the disease amongst healthcare professionals and the general public; the limited number of clinical trials and registered drugs; and the fewer peer-reviewed publications. The NIHR has launched several dedicated calls for research proposals focusing on cystic fibrosis across different funding programmes. To the best of our knowledge, although this would align well with the NIHR's commitment towards improved equality, diversity and inclusion, no equivalent call has been announced for sickle cell yet. Such dedicated calls would likely have a far-

reaching impact and contribute to address some of the problems outlined above.

3. Sickle cell is a complex chronic disease with a wide range of clinical severity, and which can affect many different organs in the body. The is currently a lack of treatment options for sickle cell. Having a range of effective and affordable treatments that can be used individually or in combination, is essential to provide the best management programme to each individual patient. This is in line with the push towards personalised medicine. Although not accessible to all people with cystic fibrosis, the availability of different CFTR modulators has led to substantial improvements in the quality of life of people with cystic fibrosis and their carers.

4. Rapid developments towards curative treatments based on gene therapy offer new hope for patients, but further research is needed to ensure the long-term safety of these treatments. The substantial costs of such therapies also limit the number of patients per year who can be treated, raising important ethical questions about prioritisation. After the withdrawals of Crizanlizumab and Voxelotor, the approval of Casgevy on the NHS will certainly be very welcome by eligible patients.

5. Most patients face significant challenges related to lack of knowledge, stigmatisation and racism, in relation to their healthcare. Education of healthcare professionals, including GPs, A&E staff, and nurses, as well as of the public is really needed. A better awareness of sickle cell would likely substantially contribute: to a better understanding of the lived experiences of patients; to rebuild trust towards the healthcare services; to a better management of patients; and to important reductions in the stigmatisation and structural racism often faced by patients. The neglect of sickle cell and discrimination faced by patients discourages many of them to seek medical help and leads to them managing their complications at home and lose trust in the NHS. As reflected by the hospitalisation data, this could potentially lead to a suboptimal management of sickle cell crises, leading to more severe health outcomes and higher costs. It seems that the management of people with cystic fibrosis at home has substantially improved during the Covid-19 pandemic, resulting in sustained fewer hospital admissions. Lessons could therefore potentially be learned from their experience.

6. Patients experience different standards of care in different parts of the country, with some avoiding or fearing to travel. Some of these points were already highlighted in the NHS RHO Digital Discovery report on sickle cell produced by Public Digital.[46] There is various guidance in place in relation to the administration of penicillin in newborns or the maximum time expected in A&E for the management of a sickle cell crisis,[75] but targets tend to be poorly met, partly due to a lack of resources and staff. These shortfalls might be due to the often-underestimated number of patients affected with sickle cell. Additional financial resources and specialist staff, including nurses and haematologists,

are required to reduce these inequalities. This is likely to be a particular concern in areas where there are only a few patients affected by conditions like sickle cell or cystic fibrosis. Novel technologies (e.g. virtual consultations) might provide valuable tools to enable such patients to access the specialist care that they need.

7. Many of the sickle cell inequalities highlighted in this report are quite systemic and would require collective change to address them. Serious failings in the care of sickle cell patients were already evidenced in the No One's Listening report published in 2021.[45] Although this was not the purpose of this review, we found very few (if any) signs of improvements in the responses to our surveys. Key stakeholders can play a major role in triggering changes, but it will take a collective approach to address these inequalities.

8. Finally, as for any medical condition, the sickle cell patients themselves and their carers have a unique knowledge based on their lived experience. Much can be learned from individuals and groups of patients about ways to improve the management of sickle cell and it is essential that patients are involved in decisions that affect their care, from how their data can be used to the design of clinical trials. Such involvement will contribute to (re)build trust towards the healthcare system and researchers. The UK Sickle Cell Society can play a key role in guiding and coordinating interactions between patients and other stakeholders.

This report is the result of one year of hard work to compile different pieces of comparative evidence to document inequalities. Given this timeframe, there are clearly limitations associated the work presented. First, apart from the HES APC data analysis and the patient survey, we have mostly relied on data publicly available. We have considered a wide range of themes and comparative indicators, but others could certainly also have been considered. Although we tried as much as possible to have consistency in the time periods considered for our different analyses, we were often limited by the availability of retrospective data or a lag in the release of evidence for the most recent years. Second, further work is required to better understand the relationships between the different pieces of evidence presented. Inequalities are driven by multiple interconnected factors, including geography, socio-economic status, education, policies and finances. They are documented through a mixture of quantitative and qualitative evidence and require a large multidisciplinary team to address them.

Recommendations

Our recommendations reflect the key findings of this review and are aligned with some of the global recommendations of the Lancet Haematology Commission on Sickle Cell Disease.[74]

Although implementation of these recommendations will require actions from a range of stakeholders and partners, key institutions which should lead on the recommendations are named.

Better Data

Improve Data Access and Use

Enable access to and use of routine epidemiological data on sickle cell, including mortality and life expectancy data, for monitoring and commissioning purposes (Department of Health and Social Care and NHS Race and Health Observatory).

Drive Accountability Through Data

Use data to ensure and track equitable provision of NHS sickle cell care, holding systems accountable and driving continuous improvement (Department of Health and Social Care).

Better Support

Fund Action-Focused Research

Prioritise dedicated funding calls for research aimed at improving patient access, experiences, and outcomes, in sickle cell care (National Institute for Health and Care Research, UK Research and Innovation, and Wellcome).

Strengthen Workforce Education

Expand education of healthcare professionals and the sickle cell workforce, including specialist nurses, to provide safe and timely care across the NHS for people with sickle cell (Department of Health and Social Care).

Better Treatment

Ensure Access to Curative Therapies

Guarantee affordability and accessibility of new curative treatments, such as Casgevy, for all eligible patients (Department of Health and Social Care).

Embed Equity in Clinical Guidelines

Ensure existing and future clinical guidelines for sickle cell proactively address and reduce racial bias and inequity (National Institute for Health and Care Excellence in collaboration with NHS Race and Health Observatory).

Graphic summary of recommendations

Appendices

Appendix 1. Monthly blog posts

Throughout this project, we shared updates with monthly posts on our blog: **https://blogs.imperial.ac.uk/uk-sickle/**. Below, we included a couple of examples of content.

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Sickle Cell Comparative Research to Inform Policy Project

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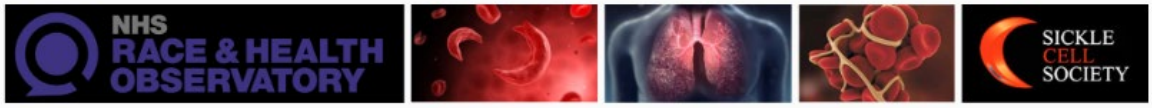

February & March – Bridging Patients & Research

Rutendo Muzambi
14 April 2025

A quick recap

In **January**, we shared updates on our NHS RHO comparative research project and a range of upcoming sickle cell events and projects.

What have we been up to?

In February and March, we focused on patient engagement events around sickle cell research and attended an in-person meeting on the latest sickle cell research in the UK and across the world.

1. Sickle Cell Research: A Day of Learning, Art, and Empowerment

Rutendo volunteered in planning and delivering a **Genomics England** funded sickle cell event which took place on Saturday 29th March at Chelsea Football Club. The event, dedicated to people living with sickle cell and their carers, focused on sickle cell research and how people with lived experience can get involved in research.

The day featured a rich variety of activities such as panel discussions, art exhibitions, music performances and workshops that brought together healthcare professionals, researchers, people living with sickle cell and artists.

ABOUT SICKLE CELL COMPARATIVE RESEARCH TO INFORM POLICY PROJECT
Sample Page

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RECENT POSTS
- February & March – Bridging Patients & Research
- January – New Year, New Milestones
- December – Thank you and season's greetings
- October & November – Time flies
- September – Sickle Cell Awareness Month

2. Patient-centred sickle cell disease management in sub-Saharan African (PACTS) annual meeting

Fred is leading a work package on Mapping and Epidemiology as part of a multi-country project between the UK, Ghana, Nigeria and Zambia. The project is funded by the National Institute of Health & Care Research (NIHR) and led by Professor Imelda Bates at the Liverpool School of Tropical Medicine. Reuben, who works with Fred, presented recent geo-mapped data collected from people with sickle cell attending 6 different health care facilities around Abuja, which will help us to better understand barriers and facilitators of access to healthcare in these settings.

3. Global Congress on Sickle Cell Disease

5th GLOBAL CONGRESS ON SICKLE CELL DISEASE

Scientists and healthcare professionals often meet up in conferences or events to present their latest findings, network and collaborate, and discuss particularly important issues. In early June, many of the international experts working on sickle cell disease will meet in Abuja, Nigeria for the 5th Global Congress on Sickle Cell Disease, organised jointly by the **Global Sickle Cell Disease Network** (GSCDN) and the **Sickle Cell Support Society of Nigeria** (SCSSN). Fred will be attending and presentting some of his work. If interested, you can find more information **here**.

Thank you for reading our blog. See you in May!

Rutendo & Fred

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Sickle Cell Comparative Research to Inform Policy Project

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December – Thank you and season's greetings

Rutendo Muzambi

19 December 2024

A quick recap

Last month, we launched our patient engagement survey on inequalities in sickle cell care, discussed our collaborative efforts and the recent **Marmot report** on racism and health inequalities.

1. Survey

We would like to express our deepest thanks to those who participated and shared our **survey**. Your incredible response and tremendous effort in disseminating the survey have been invaluable. Our NHS RHO funded project is coming to an end, so we are working hard to write it all up in a report and in scientific publications for wide dissemination. The winner of the virtual voucher will be contacted shortly. Findings from the survey and other parts of our work on inequalities in sickle cell will be published in spring 2025.

2. Imperial Sickle Cell Alliance Meeting

On Friday 6th December, Rutendo attended the **Imperial Sickle Cell Alliance** meeting were discussions with stakeholders across institutions, organisations and patient groups focused on health, advocacy, education, research and innovation projects. The Alliance is a fairly recent initiative, led by **Dr Steven Okoli** and **Professor Julie Makani**, with the aim to unite organizations and campaigns under a single umbrella to have a stronger, more unified voice in advocating for sickle cell patients.

3. The future of Sickle Cell Research survey

Genomics England, the **Sickle Cell Society** and the **James Lind Alliance** are conducting a **survey** to gather insights from people with lived experience of sickle cell, family members, carriers, caregivers, and healthcare professionals. The aim is to identify the most important research areas for sickle cell and how genomics can contribute to improving patient care and outcomes. Have your say for a chance to win a £150 Amazon voucher!

4. Thank you!

As we wrap up for the year, we thank you for following, supporting and engaging with our blog. We wish you a wonderful festive break and happy new year!

By Rutendo Muzambi

ABOUT SICKLE CELL COMPARATIVE RESEARCH TO INFORM POLICY PROJECT

Sample Page

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RECENT POSTS

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Appendix 2. Online Survey

Understanding inequalities in Sickle Cell

Understanding Inequalities in Sickle Cell Survey We would like to invite you to take part in this short anonymous survey looking at inequalities in the care of people with sickle cell. This study is part of an overall project at Imperial College London funded by the NHS Race and Health Observatory comparing inequalities in people living with sickle cell to those faced by people with cystic fibrosis and haemophilia. We are working in collaboration with the UK Sickle Cell Society. If you would like further information on this project, please visit our blog. Please click on the video below to watch a one-minute clip where Dr Piel and Dr Muzambi provide more details about this project and survey. This survey will only take 5-10 minutes to complete. If they wish, participants who complete this survey will be entered into a prize draw for a chance to win a £50 gift voucher. Please note that this survey will close on 20th December 2024.

Q1 Do you live with sickle cell or care for someone with sickle cell?

- o Yes, I have sickle cell
- o Yes, I care for someone with sickle cell
- o No, I do not have sickle cell and do not care for someone with sickle cell

Q2 Do you live in the United Kingdom?

- o Yes
- o No

Q3 How old are you?

- o 0 to 17 years old
- o 18-29 years old
- o 30 to 39 years old
- o 40 to 49 years old
- o 50 years and older

Q4a As a patient or carer, to what extent do you agree with the statement: 'Sickle cell is neglected'?

- o Strongly disagree
- o Somewhat disagree
- o Neither agree nor disagree
- o Somewhat agree
- o Strongly agree

Q4b Please explain why you think sickle cell is or is not neglected.

The following two questions were displayed for respondents who answered "Yes, I have sickle cell" to Q1.

Q5a Within the last 12 months, have you received unfair treatment when getting care for sickle cell?
(Unfair treatment can include being ignored or treated disrespectfully by healthcare professionals, receiving inadequate care, or experiencing discrimination.)

- o Yes, frequently
- o Yes, occasionally
- o Yes, once
- o No, never

Q5b Could you please provide at least one example of a situation where you experienced unfair treatment for sickle cell?

The following four questions were displayed for respondents who answered "Yes, I care for someone with sickle cell" to Q1.

Q5a Within the last 12 months, have you witnessed the person you care for receive unfair treatment when getting care for sickle cell?
(Unfair treatment can include being ignored or treated disrespectfully by healthcare professionals, receiving inadequate care, or experiencing discrimination.)

- o Yes, frequently
- o Yes, occasionally
- o Yes, once
- o No, never

Q5b Could you please provide at least one example of a situation where the person you care for received unfair treatment for sickle cell?

Q5c Within the last 12 months, have you been dismissed while trying to advocate for somebody you care for with sickle cell?

- o Yes, frequently
- o Yes, occasionally
- o Yes, once
- o No, never

Q5d Could you please provide at least one example of when you were dismissed while trying to advocate for the person you care for with sickle cell?

Q6 What do you think could be done to ensure people with sickle cell are treated in a fairer way when receiving care (e.g. GP, hospital or A&E visit)?

Q7a Please share any additional comments or experiences you would like to add.

Q7b We may use some of your responses in our report to the NHS Race and Health Observatory to better reflect lived experiences of people with sickle cell, alongside our data analysis. Would you agree for us to cite some of your anonymous answers in this report?

- o Yes, I consent to my comments being shared anonymously in the report.
- o No, I do not consent to my comments being shared in the report.

Q7c To enter the £50 prize draw, please provide your email address below so we can contact you if you win. Please note your email address will only be used to contact the winner. All email addresses will be deleted as soon as the prize is successfully awarded.

Thank you for your responses so far. We have three additional short questions regarding your gender, ethnicity, and the region in which you reside to help us better understand our survey results. Your answers will remain confidential.

Q8 What is your sex assigned at birth?

- o Male
- o Female
- o I prefer not to say

Q9 Please select the option that best describes your ethnic group or background

- o Black/ African/Caribbean/Black British (African, Caribbean, Any other Black/African/ Caribbean background)
- o Mixed/Multiple ethnic groups (White and Black Caribbean, White and Black African, White and Asian, Any other
- o Asian/Asian British (Indian, Pakistani, Bangladeshi, Chinese, Any other Asian background)
- o White (English/Welsh/Scottish/Northern Irish/British, Irish, any other White background)
- o Other ethnic group (Arab, Any other ethnic group)
- o Prefer not to say

Q10 Which city or country of the UK do you currently live in?

- o London
- o Birmingham
- o Manchester
- o Elsewhere in England
- o Wales
- o Scotland
- o Northern Ireland

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