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The Sickle Eye Project: Prevalence of visual impairment due to sickle cell retinopathy and maculopathy in the United Kingdom

Submission date 07/02/2024	**Recruitment status** No longer recruiting	[_] Prospectively registered [X] Protocol
Registration date 19/03/2024	**Overall study status** Completed	[_] Statistical analysis plan [_] Results
Last Edited 24/04/2025	**Condition category** Eye Diseases	[_] Individual participant data [X] Record updated in last year

Plain English summary of protocol

Background and study aims
Sickle cell disease (SCD) is one of the most common genetic disorders in the United Kingdom, affecting over 14,000 people. It can cause reduced vision, most commonly by affecting the film that coats the inner part of the eye – known as the retina. It causes unwanted blood vessels to grow on the retina, which can either bleed or pull the retina away from the wall of the eye causing significant sight loss. However, these unwanted blood vessels can shrink away on their own. We are not currently able to predict in which eyes the blood vessels will bleed or pull the retina and in which eyes the blood vessels will shrink on their own. We also do not know how many people with sickle cell disease in the UK have suffered loss of vision due to the effects of sickle cell disease on the retina.
The aim of this study is to find out how common it is for people with sickle cell disease to have reduced vision due to the effects of sickle cell disease on their retina. The researchers also want to know how the severity of sickle cell disease affects the chance of having reduced vision due to sickle cell disease. They also want to know how any reduction in vision affects quality of life when you have sickle cell disease. The researchers will do this by measuring the vision and pictures of the retina taken from 600 people with sickle cell disease from different parts of the UK. They will also complete a questionnaire assessing how their vision affects daily activities and whether they find attending the eye clinic for an eye check acceptable. Participation in the study will only require one visit and tests that are quick, painless and commonly performed in the NHS.

Who can participate?
Anyone over the age of 16 years with a diagnosis of sickle cell disease

What does the study involve?
Participants will be asked to attend an eye clinic appointment where their pupils will be dilated and eye tests and scans will be performed. Participants will also be asked to complete two questionnaires, one to asses vision-related quality of life and one to assess the acceptability of the tests carried out.

What are the possible benefits and risks of participating?
There will be no direct benefits to participants. However, it is hoped that the findings from this study will help to improve our understanding of how many people with sickle cell disease are affected by sight loss and how we can design better healthcare services for them. It is also hoped the information gathered will help us to understand how the treatment of sickle cell disease affects the complications that may develop in the eye and support the design of treatments that prevent sight loss.

Where is the study run from?
London North West University Healthcare NHS Trust (UK)

When is the study starting and how long is it expected to run for?
September 2021 to March 2025

Who is funding the study?
National Institute for Health and Care Research (NIHR) (UK)

Who is the main contact?
For more information please contact the study project team at lnwh-tr.sickleiproject@nhs.net

Contact information

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Public

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Additional identifiers

Clinical Trials Information System (CTIS)
Nil known

Integrated Research Application System (IRAS)
333849

ClinicalTrials.gov (NCT)
Nil known

Protocol serial number
IRAS 333849, CPMS 58836

Study information

Scientific Title
The Sickle Eye Project: Prevalence of visual impairment due to sickle cell retinopathy and maculopathy in the United Kingdom

Acronym
Sickle Eye Project

Study objectives
Sickle cell disease (SCD) is one of the most common genetic disorders in the United Kingdom, affecting over 14000 people. It can cause reduced vision, most commonly by affecting the film that coats the inner part of the eye – known as the retina. It causes unwanted blood vessels to grow on the retina, which can either bleed or pull the retina away from the wall of the eye causing significant sight loss. However, these unwanted blood vessels can shrink away on their own. We are not currently able to predict in which eyes the blood vessels will bleed or pull the retina and in which eyes the blood vessels will shrink on their own. We also do not know how many people with sickle cell disease in the UK have suffered loss of vision due to the effects of sickle cell disease on the retina.

In this research study, we want to find out how common it is for people with sickle cell disease to have reduced vision due to the effects of sickle cell disease on their retina. We also want to know how the severity of sickle cell disease affects the chance of having reduced vision due to sickle cell disease. We also want to know how any reduction in vision affects quality of life when you have sickle cell disease. We will do this by measuring the vision and pictures of the retina taken from 600 people with sickle cell disease from different parts of the UK. We will also ask them to complete a questionnaire assessing how their vision affects daily activities and whether they find attending the eye clinic for an eye check acceptable. Participation in the study will only require one visit and tests that are quick, painless and commonly performed in the NHS.

Ethics approval required
Ethics approval required

Ethics approval(s)
approved 06/10/2023, South Central - Oxford A Research Ethics Committee (Ground Floor, Temple Quay House, 2 The Square, Bristol, BS1 6PN, United Kingdom; +44 (0)2071048171; oxforda.rec@hra.nhs.uk), ref: 23/SC/0363

Study design
Observational cross sectional study

Primary study design
Observational

Study type(s)
Quality of life, Screening

Health condition(s) or problem(s) studied
Visual impairment, retinopathy and maculopathy in patients with sickle cell disease

Interventions
People with sickle cell disease attending their haematology clinic will be informed about the study by their clinical team. They will be provided with the participant information sheet detailing the study and have the opportunity to ask questions about the study. If they wish to participate, they will attend the eye clinic, where their understanding of the study will be confirmed and they will be consented to participate in the study. They will proceed to have their visual acuity measured in the eye clinic. Next, they will have their pupils dilated which will allow the clinical team to examine their eyes using a slit lamp biomicroscope. Next, they will have optical coherence tomography (OCT), optical coherence tomography angiography (OCT-A) (if available) and ultra-wide field fundus photography performed. Lastly, they will complete the vision-related quality of life questionnaire and the acceptability questionnaire. Their clinical team will manage their eyes as usual and participating in the study will not influence their care in any way.

The researchers will use the NEI-VFQ 25 to determine vision-related quality of life, as it is the most widely used instrument in eye disease. However, it takes, on average, 15-20 minutes to complete, which can occasionally be too long for the participant or the clinical service. As such, the researchers will assess participation in the study over the initial 4 months and discuss with sites whether the NEI-VFQ 25 is preventing participation. If this is the case, the researchers have arranged to substitute the NEI-VFQ 25 with the EQ-5D with vision bolt-on, which takes approximately 5 minutes to complete and appears to be almost as accurate as the NEI-VFQ 25 but is a newer questionnaire for this purpose.

The OCT, OCT-A and ultrawide field fundus photography images will be pseudo-anonymised and sent securely to the Moorfields Reading Centre with the participants' permission. In addition, relevant parts of their medical and eye history will be entered by the research team into the electronic case report form which is also sent to the Moorfields Reading Centre.

The images and study data will be assessed and graded by the reading centre experts and analysed by the research team to determine the proportion of eyes with visual acuity of LogMAR ≥0.3 (approximately driving vision). Data analysis will also be conducted to meet the secondary study objectives.

Intervention Type

Other

Primary outcome(s)
The prevalence of visual impairment due to sickle cell retinopathy or maculopathy: best corrected visual acuity LogMAR ≥0.3 in at least one eye measured through best corrected visual acuity test at baseline visit

Key secondary outcome(s))
1. The prevalence of each stage of sickle cell retinopathy and maculopathy and correlation with:
1.1. Number of hospital admissions with acute sickle cell episodes in the past 12 months
1.2. Serum HbF level
1.3. Serum Hb level
Collection into source document of the number of hospital admissions in the past 12 months and serum HbF and Hb level results within 3 months of study entry at baseline visit
2. The impact of sickle cell retinopathy and maculopathy on vision-related quality of life measured using the NEI-VFQ 25 questionnaire at baseline visit
3. The acceptability to patients of retinal imaging and routine screening for sickle cell retinopathy and maculopathy measured using acceptability questionnaire at baseline visit

Completion date
31/03/2025

Eligibility

Key inclusion criteria
1. Willingness to participate
2. Ability to provide informed consent
3. Age 16 years or older
4. Diagnosis of SCD of any genotype

Participant type(s)
Patient

Healthy volunteers allowed
No

Age group
Other

Lower age limit
16 years

Upper age limit
100 years

Sex
All

Key exclusion criteria

1. Inability to consent
2. Poor image quality
3. Age <16 years
4. Sickle cell trait only

Date of first enrolment
21/11/2023

Date of final enrolment
31/10/2025

Locations

Countries of recruitment
United Kingdom

England

Study participating centre
London North West University Healthcare NHS Trust - Central Middlesex Hospital
Acton Lane
Park Royal
London
United Kingdom
NW10 7NS

Study participating centre
Whittington Health NHS Trust
The Whittington Hospital
Magdala Avenue
London
United Kingdom
N19 5NF

Sponsor information

Organisation
London North West Healthcare NHS Trust

ROR
https://ror.org/04cntmc13

Funder(s)

Funder type
Government

Funder Name
National Institute for Health and Care Research

Alternative Name(s)
National Institute for Health Research, NIHR Research, NIHRresearch, NIHR - National Institute for Health Research, NIHR (The National Institute for Health and Care Research), NIHR

Funding Body Type
Government organisation

Funding Body Subtype
National government

Location
United Kingdom

Funder Name
ROCHE PRODUCTS LIMITED

Alternative Name(s)
National Institute for Health Research, NIHR Research, NIHRresearch, NIHR - National Institute for Health Research, NIHR (The National Institute for Health and Care Research), NIHR

Funding Body Type
Government organisation

Funding Body Subtype
National government

Location
United Kingdom

Results and Publications

Individual participant data (IPD) sharing plan
The datasets generated during and/or analysed during the current study will be stored in a non-publicly available repository (Moorfields Grading Portal, https://grading.readingcentre.org/sign_in).
The type of data stored: data (basic demographics, sickle cell disease details and eye test

results) and images.
The process for requesting access (if non-publicly available): individual portal access is granted by the study project team to delegated staff in participating sites.
Dates of availability: portal access available for the duration of the recruitment period.
Informed consent is obtained from all participants prior to data collection.
No participant identifiers are collected into the case report form therefore only anonymised data are entered into the data collection system, likewise for images which are exported in an anonymised manner and uploaded into the portal.

IPD sharing plan summary

Stored in non-publicly available repository

Study outputs

Output type	Details	Date created	Date added	Peer reviewed?	Patient-facing?
Protocol file	version 1.5	18/12/2023	18/03/2024	No	No