## BEACON

**Full Title: O**utcomes of **r**etinal detachment **i**n **s**ickle cell retinopathy **Acronym: ORIS** study

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What you are studying: The surgical methods used and the corresponding outcomes in patients with sickle cell retinopathy (proliferative or non proliferative) developing retinal detachment (tractional, rhegmatogenous or combined tractional/ rhegmatogenous)

What is your primary research question: What are the surgical methods used by surgeons in these eyes (PPV, scleral buckle, combined treatment) and what are the postoperative outcomes?

What are the characteristics of retinal detachment developing in sickle cell retinopathy patients?

What are the risk factors for primary surgery failure?

What are the functional, anatomical outcomes and surgical complications in these eyes?

## Background and importance:

Sickle cell disease is one of the commonest genetic disorders in the UK and sickle cell retinopathy is a significant cause of blindness in several parts of the world. At the same time, there is concern regarding the lack of guidance on sickle cell retinopathy treatment in the UK. There are several unique characteristics in sickle cell retinopathy related retinal detachment (e.g. the fibrovascular proliferation in proliferative sickle cell retinopathy develops in the retinal periphery or many of these patients are quite young). At the same time, there are only limited data in the literature coming from small case series about the outcomes of different surgical options for the management of the disease.

## **Case definition:**

A sickle retinopathy (proliferative or non proliferative) patient developing retinal detachment (tractional, rhegmatogenous or combined) and undergoing surgery for the condition.

## Inclusion criteria:

- Eyes with tractional, rhegmatogenous or combined tractional/rhegmatogenous retinal detachment and sickle cell retinopathy
- Any age
- Any macula status (on/ off)
- Minimum of 3 months follow up post primary repair

## Exclusion criteria:

- Tractional or combined retinal detachment developing secondary to other conditions (e.g. diabetes mellitus)
- Typical rhegmatogenous retinal detachment developing in patients with sickle cell disease but no sickle cell retinopathy
- Less than 3months of follow up

## Likely incidence:

Sickle cell related retinal detachment is a major cause of retinal detachment in several parts of the world and a global problem nowadays. Despite that, there is a paucity of data in the literature and widespread concern regarding the lack of evidence and treatment guidance on the condition. Analysing preoperative characteristics and surgical outcomes will provide input about prognosis, optimal management strategies and outcomes in these patients.

## Combined prospective/retrospective data collection

## For retrospective

- Eligible study period: September 2018 September 2023 (5 years)
- Target completion for end of data collection: 1st May 2023

## **For Prospective**

Case collection period: August 2023 – August 2024 Follow up duration: 3 months minimum

## Primary outcome measure:

Primary surgery success rate (flat retina after 1 surgery – no tamponade in situ)

## Secondary Outcome measures:

Multiple surgery success rate (flat retina after multiple surgeries – no tamponade in situ) Best corrected visual acuity (BCVA) at final follow-up BCVA improvement Surgical intervention Adverse events

**Data collection form(s):** Formatted excel chart available from study leads via email. All data must be anonymised

Images required: nil

#### References

1. Amissah-Arthur KN, Mensah E. The past, present and future management of sickle cell retinopathy within an African context. Eye. 2018;32(8):1304-14.

2. Chen RW, Flynn Jr HW, Lee W-H, Parke III DW, Isom RF, Davis JL, et al. Vitreoretinal management and surgical outcomes in proliferative sickle retinopathy: a case series. American journal of ophthalmology. 2014;157(4):870-5. e1.

3. Williamson T, Rajput R, Laidlaw D, Mokete B. Vitreoretinal management of the complications of sickle cell retinopathy by observation or pars plana vitrectomy. Eye. 2009;23(6):1314-20.

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5. Dinah C, Greystoke B, Mueller I, Talks J. Action on sickle cell retinopathy: the time is now. Eye. 2022;36(6):1138-9.