Adult Haemoglobinopathy Service

Eye complications in sickle cell disease: Clinical guideline

Acute complications

Acute ocular complications in patients with sickle cell disease (SCD) may occur secondary to trauma, infection and vaso-occlusive episodes leading to occlusion of the retinal vasculature, or progression of proliferative sickle retinopathy (PSR). All may have devastating consequences including permanent loss of vision.
Hyphema, central retinal artery occlusion (CRAO), vitreous haemorrhage, orbital and peri-orbital infections, orbital infarction, and orbital compression syndrome (OCS) **all require urgent assessment and therapy.** Late state changes associated with PSR such as non-clearing vitreous haemorrhage and retinal detachment may present with acute visual symptoms.

Indications for referral

The following should be referred **urgently to the local acute eye service** for assessment:

- Anyone with SCD presenting with eye trauma
- Any patient with SCD exhibiting signs and symptoms such as protrusion of the eye, changes in visual acuity (flashes or floaters), and unilateral or bilateral loss of vision including the appearance of a 'veil' or 'curtain'.

Chronic complications

Ocular complications due to SCD are uncommon in childhood; however retinal vessel occlusion may begin in adolescence, in particular with HbSC disease. Thus there is a requirement for ophthalmological assessment from puberty onwards.

Recommendations

All SCD patients:

- should be informed about the risk of ophthalmic complications and asked about visual symptoms at their annual review (reference the patient information leaflet).
- should have a baseline retinopathy screening and follow-up every 2-3years. Where ophthalmology is not available optician routine screening is adequate
- should be educated about acute symptoms (including trauma) and how to access help
- with visual symptoms should be referred for ophthalmic review
- with a history of retinopathy should have yearly ophthalmic review under the care of ophthalmology
- on regular transfusion regimens receiving Desferrioxamine or Deferasirox require annual ophthalmological assessment for the development of drug-induced retinopathy

Management

- Urgent referral to ophthalmology is required. Exchange is unlikely to be required for vitrectomy, but all cases should be discussed with a consultant haematologist pre-operatively

Tertiary Specialist Referral:

Oxford:

Prof Susan Downes

Consultant Ophthalmic Surgeon at the Oxford Eye Hospital, with interest in genetic retinopathies. Oxford Eye Hospital, Level LG1, John Radcliffe Hospital.

Southampton:

Ophthalmology Consultant or Spr on-call

Link document:

S20.1 Patient information: Sickle cell disease and sickle retinopathy

References:

Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK, 2nd edition © Sickle Cell Society 2018

NHLBI (2014) Evidence based management of sickle cell disease. Expert panel report https://www.nhlbi.nih.gov/sites/www.nhlbi.nih.gov/files/sickle-cell-disease-report.pdf

Downes SM, Hambleton IR, Chuang EL, Lois N, Serjeant GR, Bird AC. Incidence and natural history of proliferative sickle cell retinopathy: observations from a cohort study. Ophthalmology. 2005; 112 (11):1869-75.

Authors:

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Review

Name	Revision	Date	Version	Review date
Dr Wale Atoyebi	Pre-peer review	Jan 2013	1.1	Jan 2015
Dr Deborah Hay	Routine review	Aug 2015	1.2	Jan 2017
Dr Wale Atoyebi, Consultant haematologist. Sandy Hayes, ANP	Combining of acute and chronic conditions into one document.	Nov 2017	2.0	Nov 2019
Dr Noemi Roy, Dr Henna Wong, Prof. Downes	Review and update	Nov 2019	3.0	Nov 2021
Dr Noemi Roy	Update of the recommendation and management sections, logo update	Oct 2021	3.1	Oct 2023
Dr Wale Atoyebi	Full Review	May 2024	4.0	May 2026