Birmingham Adults Sickle Cell Retinopathy: Prevalence, Presentations and Treatment Outcomes

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BMEC Birmingham & Midland Eye Centre Established 1823

NHS Sandwell and West Birmingham Hospitals NHS Trust

Introduction

- Sickle cell disease affects African/Afro-Caribbeans.
- In Birmingham population, 13.5% are of African/Afro-Carribean ethnicity (vs. 5.5% in England).
- 600 patients (4%) were registered in our single-hospital haematology sickle clinic (vs 0.4% sickle in England).
- Haematologist has routine referral to BMEC for retinopathy screening.

Purpose

To report adults' sickle retinopathy prevalence, disease severities and treatment outcomes from Birmingham sickle eye clinic.

Design

- Single eye centre study: from an established one-stop sickle eye clinic with same day screening-diagnosis-treatment.
- Retrospective data collection from 2021-2023.
- Adult sickle patients (age ≥20 years) at first eye clinic presentation.

Non-proliferative retinopathy signs	Salmon patch, sunburst scar, iridescent spots, hue change, white-without-pressure
PSR stage 1	Peripheral arteriolar occlusion (peripheral ghost vessels, angiographic evidence)
PSR stage 2	Peripheral arteriovenous anastomosis (vascular remodelling)
PSR stage 3	Seafan retinal neovascularisation
PSR stage 4	Vitreous haemorrhage
PSR stage 5	Retinal detachment (tractional/rhegmatogenous)

Table 1. Goldberg Proliferative Sickle Retinopathy Staging (PSR) AJO, 1971

Results

Initial Presentation

- 160 patients (319 eyes)
- Genotypes: 61.25% HbSC, 31.2% HbSS, 4.4% HbSThalassaemia, and 3.1% other.
- Gender: 53.1% female.
- Mean presenting age: 35.45 years (SD 11.64, range 20-75 years).
- 78.1% routine referral from haematologists
- 35.6% asymptomatic at presentation.
- 91.8% eyes best-corrected visual acuity (BCVA) ≥6/12 (64.2% ≥6/6, 3.7% ≤6/60)
- Initial proliferative sickle retinopathy staging
  - Stage 0: 34.8%
  - Stage 1-2: 15.6%
  - Stage 3: 49.5% (=severe)
  - Stage 4: 11.3% (=severe)
  - Stage 5: 6.3% (=severe)
- 22.2% advanced peripheral elevated fibrotic scar
- 40% bilateral severe PSR (stage ≥3)

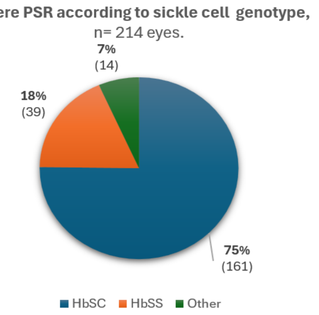


Figure 1. Correlation of sickle genotype to severe PSR (stage 3-5); sickle HbSC group has a high prevalence (75%) of severe PSR.

Follow Up (FU)

- Mean follow-up period: 11.6 years (SD 6.6, range 0.4-21.8 years).
- 13.1% had no FU visit (discharged or non-attendance).
- 88.4% BCVA ≥6/12, 3.97% ≤6/60
- OCT done in 207 eyes
  - 36.2% temporal macular thinning
  - 71.5% epiretinal membrane(ERM)/pseudohole
  - 10.1% significant macular preretinal fibrosis/traction
- Complications
  - 28.5% vitreous haemorrhage (at least one episode)
  - 5% retinal hole/tear/operculum
  - 5% tractional retinal detachment

Treatment

This sickle eye clinic has applied only retinal laser but not intravitreal anti-VEGF injection for sickle retinopathy treatment, in view of risk of retinal detachment (RD) in fibrovascular traction (common in advanced sickle retinopathy).

- Sector/barrier laser performed (at least once) in 53.4% of PSR stage 3 active disease or as barrier laser for established fibrovascular traction with risk of RD.
- Vitrectomy in 6.1% (RD 4.7%, ERM 1.4%)

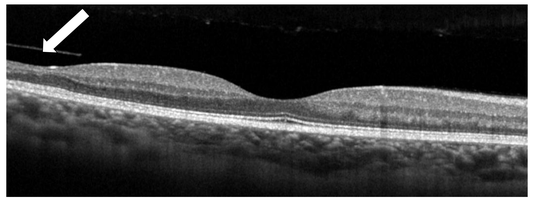


Figure 2. Despite OCT showing good normal foveal contour in sickle patients, asymptomatic macular temporal thinning (white arrow) is a feature noted in 36.2% of our cohort.

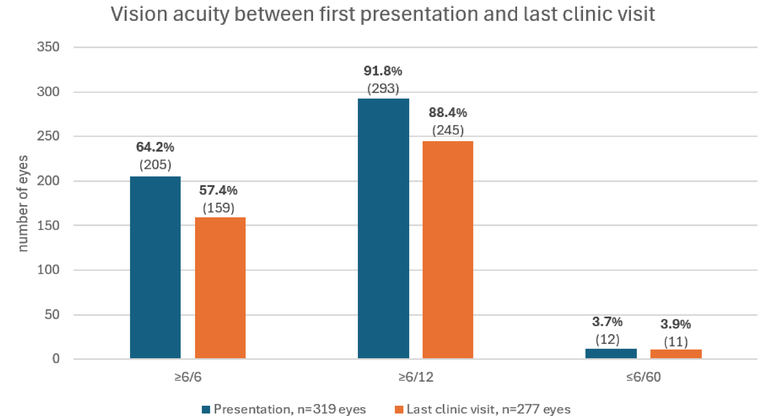


Figure 3. Sickle patients in our cohort had good vision better than 6/12 at presentation (91.8%) and at follow-up (88.4%).

Conclusion

- Most adult sickle patients attended eye clinic via retinopathy screening program through haematology (78.1%).
- Initial presentation with severe PSR (stage 3-5) is high (49.5%); notable high prevalence is in sickle HbSC genotype group.
- Good visual acuity is common (91.8% were 6/12 or better) in our cohort and is maintained after treatments despite severe PSR staging (67.5%).
- Laser treatment is effective in preventing disease progression /complications. Vitrectomy rate applied to initial severe presentation need only.
- One-stop sickle retinopathy screening-diagnosis-treatment programme allows early treatment intervention and prevents disease progression to blinding complications.