Retinopathy Severity and Presentations in Teenagers with Sickle Haemoglobinopathies

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Introduction

Sickle cell disease can cause sudden visual loss from severe proliferative sickle retinopathy, vitreous haemorrhage and retinal detachment.
Sickle retinopathy in children was known but thought of uncommon and treatment unnecessary.

Purpose

To investigate disease prevalence, retinopathy severity, genotypes in teenagers with known sickle cell disease.

Design

>Retrospective notes review/analysis.
>Single eye centre study.
>Notes identifcations from sickle eye clinics in 2022-23.
>Inclusive sickle patients age ≤19 years at first eye clinic presentation.

Non-proliferative retinopathy signs	Salmon patch, sunburst scar, iridescent spots, (hue change, white- without-pressure)
PSR Stage I	Peripheral arteriolar occlusion (peripheral ghost vessels, angiographic evidence)
PSR Stage II	Peripheral arteriovenous anastomosis (vascular remodelling)
PSR Stage III	Seafan retinal neovascularization
PSR Stage IV	Vitreous haemorrhage
PSR Stage V	Retinal detachment (Tractional/ rhegmatogenous)

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

Results

- 35 teenagers(70 eyes) with confirmed sickle cell retinopathy.
- Genotypes categories: 23/35(65.7%) HbSC, 9/35(25.7%) HbSS, 3/35(8.6%)HbSThalassaemia.
- 54% females.
- Mean presenting age:16.9yrs (SD:2.07,range10-19yrs).

Source of referrals

- 28/35(80%) Screening from hematologists/ pediatricians.
- 7/35(20%) from opticians and general ophthalmologists.

- 58/70 (82.9%) eyes asymptomatic.

Follow Up (FU):

- Mean follow-up period: 7.6yrs (SD:5.9, range 0.5-19.2yrs).
- 5 patients (=60 eyes) had no FU visit or discharged after one visit.

(4 SC patients, 1 SS patient, 1 SThal patient)

- 25/35 (71.4%) patients DNA more than once of scheduled visits.

Laser treatment (sector / barrier laser)

- performed in 29/70 eyes (41.4%) for PSR III–V disease
- repeatable if new seafans developed, traction risk at each clinic review visit.

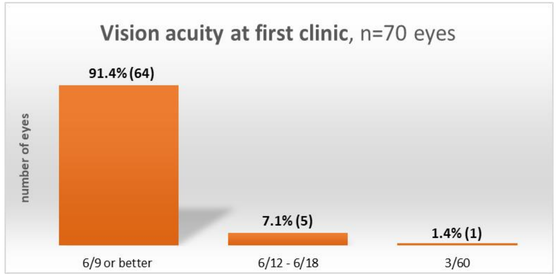


Figure 1. Majority of patients had good VA at presentation (91.4%).

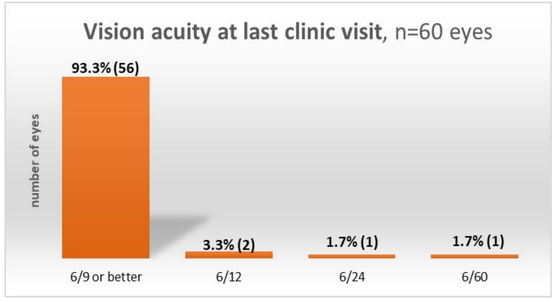


Figure 2. Majority of patients still achieved good VA at last review (93.3%).

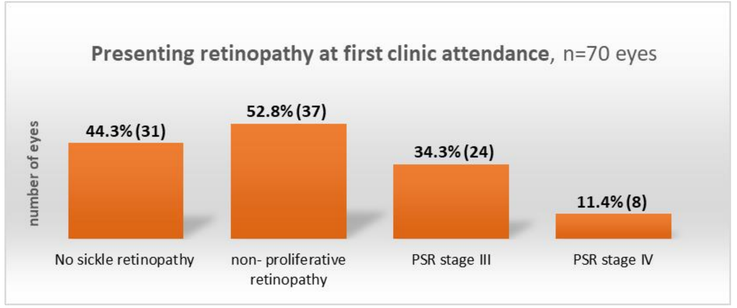


Figure 3. At first eye clinic attendance: 44.3% eyes had no sickle retinopathy, non-proliferative signs were found in 52.8% eyes (salmon patch was seen in 8.6% eyes), 34.3% showed sickle stage III disease, 11.4% eyes already vitreous haemorrhage.

Presenting retinopathy	**HbSC,** n=46eyes	**HbSS,** n=18eyes	**HbSThal,** n=6eyes
PSR Stage III	20 (43.5%)	1 (5.6%)	3 (50%)
PSR Stage IV	6 (13%)	0%	2 (33.3%)
PSR Stage V	0%	0%	0%
Progressed retinopathy	**HbSC,** n=38eyes	**HbSS,** n=16eyes	**HbSThal,** n=6eyes
PSR Stage III	8 (21%)	4 (25%)	0%
PSR Stage IV	10 (26.3%)	0%	0%
PSR Stage V	2 (5.3%)	0%	0%

Table 2. Severe retinopathy PSR stage 3 and 4 were higher in sickle SC patients at presentation and follow-up; more sickle SThalassaemia patients with severe retinopathy than sickle SS patients at presentation. Two (5.3%) eyes (were SC patients) developed PSR stage V with localised retinal-detachment.



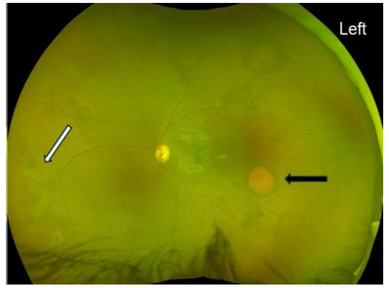


Figure 4: Example of fundus finding at first eye clinic presentation of a school boy with known sickle SC, aged 12. Vision 6/5 each eye, both eyes had auto-infarcted seafans scars on traction (white arrows). Right eye had vitreous haemorrhage (red arrows), flat retina. Left eye lesion a salmon patch in mid-temporal retina (black arrow).

Conclusion

Our cohort showed teenagers (age 10-19 years old) with sickle haemoglobinopathies can present with severe retinopathy, higher in HbSC genotype. Visual loss however is uncommon. Retinopathy screening could be beneficial from age 10 to detect early treatable sickle retinopathy to prevent progression to traction and detachment.