SALUS UNIVERSITY
The Eye Institute

Pennsylvania College of Optometry

The Focal Point

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Traditional Class of 2023

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Class of 2021, SUNY College of Optometry

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Undergrad: The George Washington University
Major: Biological Sciences; **Minor:** History/PoliSci
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Splaying, Sunbursts, and Salmon Patches, Oh My! A Pediatric Case of Sickle Cell Retinopathy

Some details of the case have been changed to protect patient confidentiality. Though the NBEO PAM-Style format is utilized for the case write-up, there is no affiliation with NBEO.

Demographics: 16 year old Black male
Chief complaint: Floaters, area of missing vision OU
History of present illness

Character/signs/symptoms: floaters which are stationary and block his vision (nasal, inferior central)
Location: OU
Severity: 7/10
Nature of onset: suddenly 2 weeks ago, after becoming light-headed and losing consciousness for ~30 seconds
Frequency: constant
Exacerbations/remissions: none
Relationship to activity or function: none
Accompanying signs/symptoms: MRI and blood work performed, unremarkable

Patient ocular history: Simple hyperopia, longstanding photophobia, longstanding scleral icterus secondary to sickle cell, Sickle cell retinopathy (resolved salmon patch OU) 4 years prior; Denies any injury/surgery
Family ocular history:

Mother: unremarkable
Father: unremarkable

Patient medical history

(+) Sickle cell thalassemia, with last crisis and blood transfusion 4 months prior
(+) Low hemoglobin (7.5-8 gm/dL), longstanding history and "his normal"

Medications taken by patient: None
Patient allergy history: No known allergy
Family medical history

Diabetes (maternal grandfather)

Review of systems

Constitutional/general health: denies
Ear/nose/throat: Cardiovascular: denies
Pulmonary: Endocrine: denies
Dermatological: denies
Gastrointestinal: denies
Genitourinary: denies
Musculoskeletal: denies
Neurologic: denies
Psychiatric: denies
Immunologic: denies
Hematologic: denies

Mental status

Orientation: oriented to person, place, and time
Mood/Affect: normal

Clinical findings

BVA:

	Distance	Near
OD:	20/20	.4/.4M
OS:	20/20	.4/.4 M

Pupils: PERRL (-) APD
EOMs: Full and Smooth
Confrontation fields: FTFC OU
Hirschberg: symmetric
Slit lamp:

lids/lashes/adnexa: unremarkable OU
conjunctiva/sclera: icterus OU; (-) vasculature changes OU
Cornea: clear epithelium, stroma, and endothelium OU
anterior chamber: deep and quiet OU; (-) cells/pigment/hyphema OU
Iris: flat and intact OU; (-) NVI OU
lens: clear OU
Vitreous: clear OU; (-) cells OU

IOPs/method: 16/14 mmHg OD/OS with Goldmann Applanation Tonometry
Fundus OD:

C/D: 0.4/0.4; (-) NVD
Macula: splaying (saucerization of the foveal pit) with inferior macular pigmentation changes
Posterior pole: salmon patch inferior mid periphery
Periphery: flat x360; superior nasal sunburst lesion, (-) NVE

Fundus OS:

C/D: 0.35/0.35; (-) NVD
Macula: splaying (saucerization of the foveal pit) with inferior macular pigmentation changes
Posterior pole: ½ DD size salmon patch, 5DD nasal to ONH
Periphery: flat x360; temporal pigment changes and nasal pigmentation; questionable old seafan retinopathy inferior, (-) NVE

Case Images:

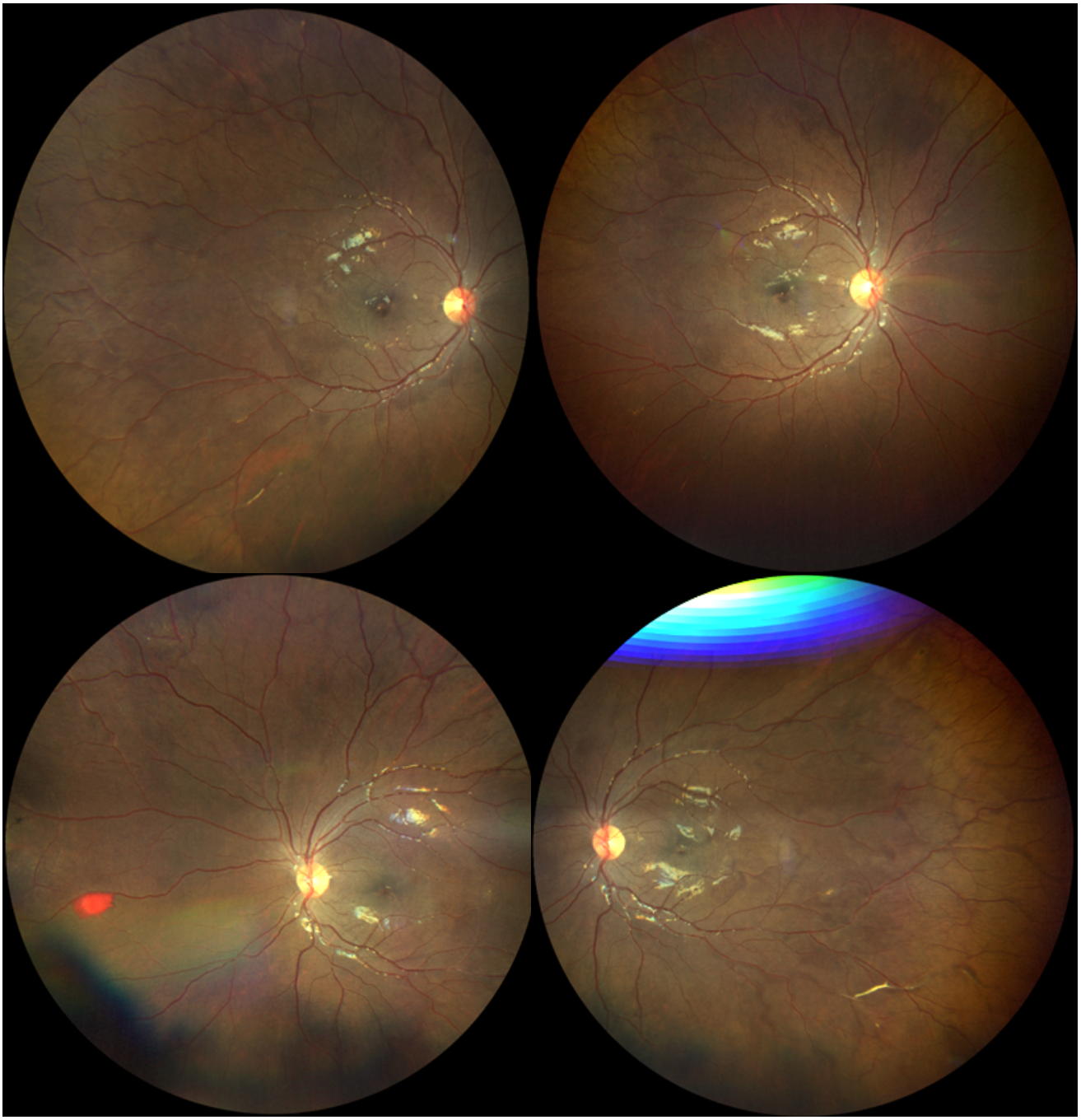

Image 1: Colored fundus photographs of the right eye (top) and left eye (bottom). Attention is drawn to the salmon patch hemorrhage in the nasal retina OS and patchy pigmentation of the retina/choroid OU in addition to pigment mottling in the foveal region OU.

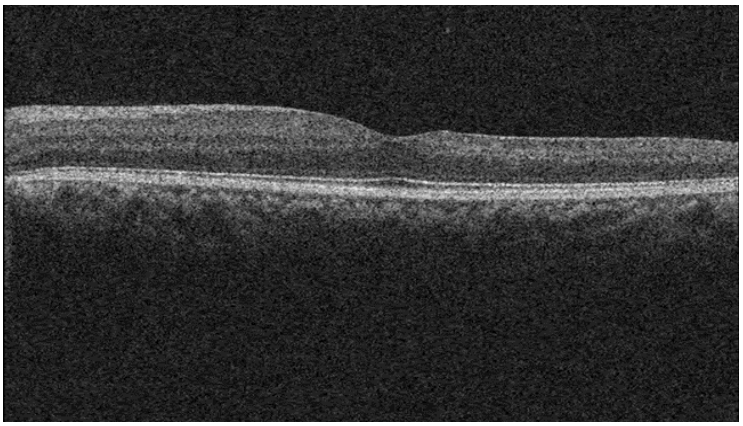

Image 2: OCT of the left macula demonstrating loss of foveal contour and shallowing of the foveal umbo along with thinning of the temporal macula as compared to the nasal tissue, indicating retinal ischemia. A mild epi-retinal membrane can be seen on the nasal macula.

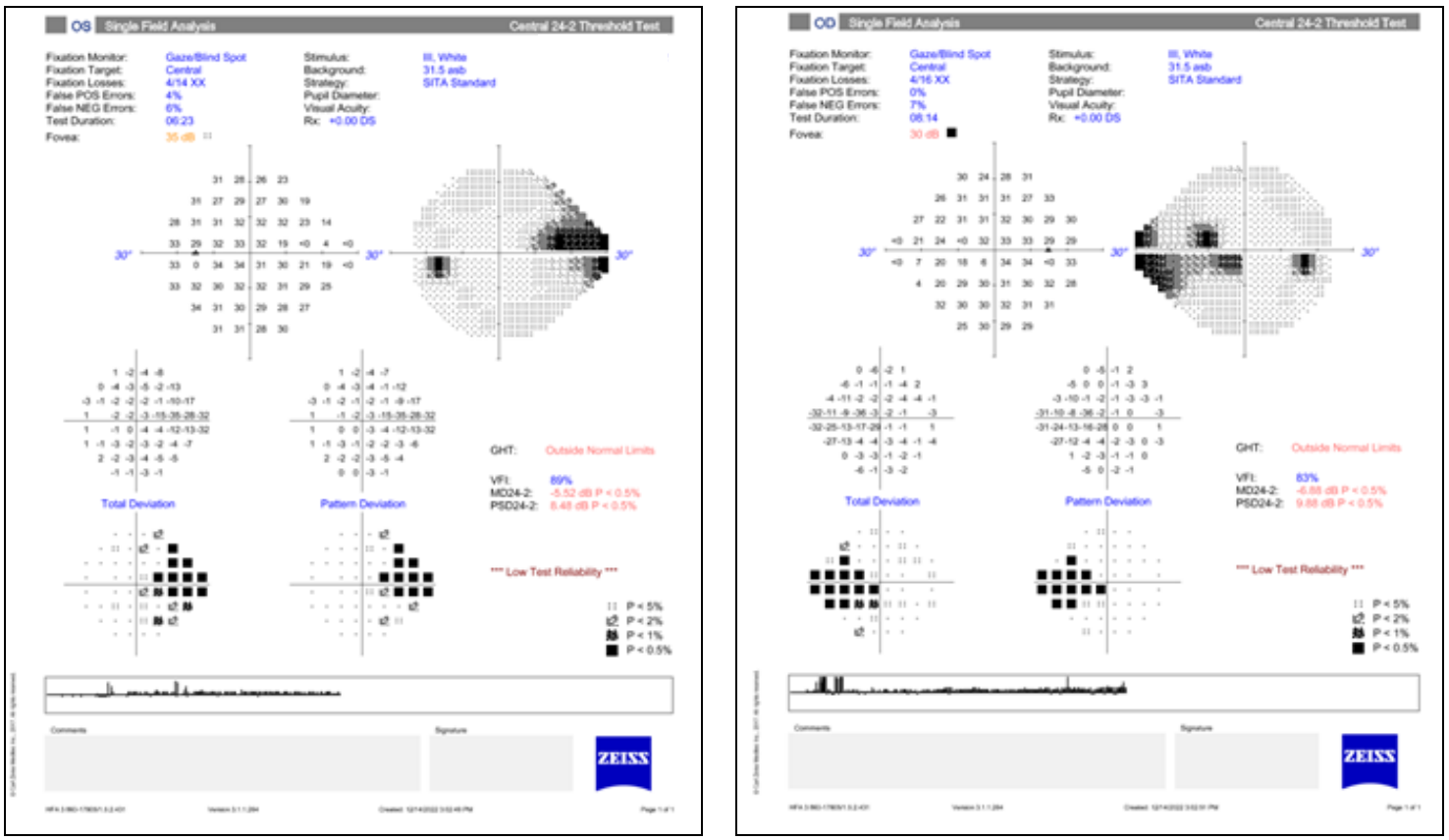

Image 3: Humphrey Visual Field 24-2 testing of the left and right eye, respectively. The left HVF demonstrates nasal visual field loss outside of the macular region indicating temporal mid-peripheral or peripheral retinal ischemia.The right HVF demonstrates visual field damage into the central 4 'macular' points indicating temporal retinal and parafoveal ischemia of the retina. HVF 10-2 would be needed to confirm visual field damage to the central 10 degrees. OCT GCC showed superior temporal thinning, which may correlate with a future HVF 10-2.

Case Management Summary

- Peripheral retinal exam remarkable for sunburst lesion OD, questionable old seafan OS, and salmon patch lesions OU
- OCT → shows foveal splaying OU (saucerization of the foveal pit) with temporal and superior temporal paramacular thinning
- HVF 24-2 → binasal defects, likely corresponding to the chief complaint of “stationary floaters” (10-2 and FA would be needed to correlate with macular structure and highlight areas of retinal ischemia)
- Fundus photos taken to document
- Referred to hematology for consultation
- Referred for Retina consultation
    - Results of Retina findings → No evidence of active proliferative retinopathy or hemorrhage. OCT shows temporal macular thinning consistent with focal microvascular ischemia. Peripheral retinal exam is remarkable for sunburst lesion OD, questionable old seafan OS, and salmon patch lesions OU. Monitor yearly or as needed.

Case Pearls

- Sickle cell retinopathy is mostly known for causing peripheral retinal issues however, clinicians should be aware of potential posterior pole complications. Macular infarctions associated with sickle cell can cause changes in the thickness of retinal tissue and cause reduced BCVA or visual field defects, for which a patient may be more symptomatic.
- Although fundus findings in proliferative disease are often considered to be most prominent in peripheral tissue, patient symptoms and close posterior pole evaluation may warrant further structural and functional testing in order to uncover anomalous findings.
- Foveal splaying, or loss of the foveal contour seen as saucerization of the foveal pit, can be a sign of macular ischemia. Structural testing in the form of OCT may yield thinning in the parafoveal area, indicating retinal atrophy in these cases.
- It is important to closely consult and co-manage with hematology and ophthalmology when monitoring patients with sickle cell retinopathy.