SALUS UNIVERSITY
The Eye Institute

Pennsylvania College of Optometry
The Focal Point
February 2024 Edition

Sonya Aminov
Traditional Class of 2025

Hometown: NYC, New York
Undergrad: Queens College
Major: Psychology
Favorite Animal: Dog
Favorite instrument: Piano
Hobby: Cooking and Baking
Last Show I binged: The Vampire Diaries

Stephanie Leburg
Class of 2016, Pennsylvania College of Optometry

Hometown: Jamestown, New York
Undergrad: SUNY Binghamton
Major: Biology
Hates: Banana flavored candy
Hobby: Video games with toddler
Favorite TV Genre: Post-apocalyptic/Sci-Fi

Surprise! Staphyloma Struggles and the Importance of Specialty Referral

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Demographics 70 year old Black male
Chief complaint: Blurry Vision OU

- Patient presents with his brother who assisted in detailed history
- Patient reports his vision has been significantly decreasing over the past 10 years
- LEE was Fall 2020, patient was referred to Neuro-ophthalmology at that time
- Blood work and MRI were ordered by Neuro OMD, both unremarkable

History of present illness

Character/signs/symptoms: blurred vision at distance and near with current spec RX
Location: OU
Severity: severe, has been worsening over the past 10 years
Nature of onset: ~10 years
Duration: Frequency: constant
Exacerbations/remissions: none
Relationship to activity or function: worsened difficulty reading
Accompanying signs/symptoms: none

Patient ocular history (-) Glaucoma (-) Ocular Surgery (-) Ocular Trauma (+) Presbyopia
Family ocular history

Mother: No reported history of Glaucoma or blindness
Father: (+) Glaucoma

Patient medical history (-) DM (+) HTN (-) Hyperlipidemia (+) Prostate cancer 2017 s/p radiation therapy 3 years ago (+) Depression(+) Parkinsons
(+) Dementia
Medications taken by patient: Amlodipine 10 mg- valsartan 320 mg tablet, Acetaminophen ER 650 mg tablet, Carbidopa 25 mg tablet - levodopa 100 mg tablet, Clopidogrel 75 mg tablet, Cyanocobalamin (Vit B-12) 1,000 mcg tablet, Diclofenac 1% topical gel, Folic acid 1 mg tablet, Gabapentin 100 mg capsule, Losartan 100 mg tablet, Methotrexate sodium 2.5 mg tablet, Mirtazapine 30 mg tablet, Quinapril 5mg tablet, Rosuvastatin 20 mg tablet, Sertraline 100 mg tablet, Tizanidine 2 mg tablet, Vitamin B-1 100 mg tablet
Patient allergy history No known drug allergies
Family medical history

Mother: (+) HTN
Father: (+) HTN

Review of systems

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

Mental status

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

Clinical findings

BVA:

	Distance	Near
OD:	4/400	0.4/ 5.0M
OS:	20/60+2	0.4/ 2.0M

Pupils: PERRL (+) APD OD

EOMs: Restricted adduction and abduction OD and OS

Confrontation fields: Constricition 360 degrees OD and OS, poor reliability

Hirschberg: symmetric

Ishihara Color Plates: OD: 1/14 OS:2/14

Subjective refraction:

		VA Distance	VA Near
OD: -5.50 -3.25 x 150	ADD +2.50	20/200	0.4/1.6M
OS: +1.25 sph	ADD +2.50	20/50 +2	0.4/1.6M

Slit lamp:

Lids/Lashes/Adnexa: adnexa normal OU, superior capped glands OU

Conjunctiva: white and quiet bulb conj, pink & quiet palpebral conj OU, diffuse melanosis OU

Cornea: normal epithelium, stroma, endothelium and tear film OU, 360 arcus OU

Anterior Chamber: deep and quiet OU

Iris: Scattered diffuse iris atrophy OU

Lens: 1+ Nuclear Sclerosis OU, 1+ Cortical Spoking OU

Vitreous: Posterior vitreous detachment OU

IOPs/method: 12/13mmHg via GAT

Blood pressure: 160/90 mmHg RAS

Fundus OD:

C/D: 0.3/0.3, question of temporal pallor, NRR intact, no edema, no notching

Macula: flat, no hemorrhages, exudates, pigmentary changes, no macular edema
(+) Type VII staphyloma, (-) lacquer cracks , (-)CNVM

Periphery: flat x 360 degrees, no RD, no breaks

Fundus OS:

C/D: 0.35/0.35; question of temporal pallor, margins distinct, no elevation, NRR intact, no edema, no notching

Macula: flat, no hemorrhages, exudates, pigmentary changes, no macular edema

Periphery: flat x 360 degrees, no RD, no breaks

Case Images:

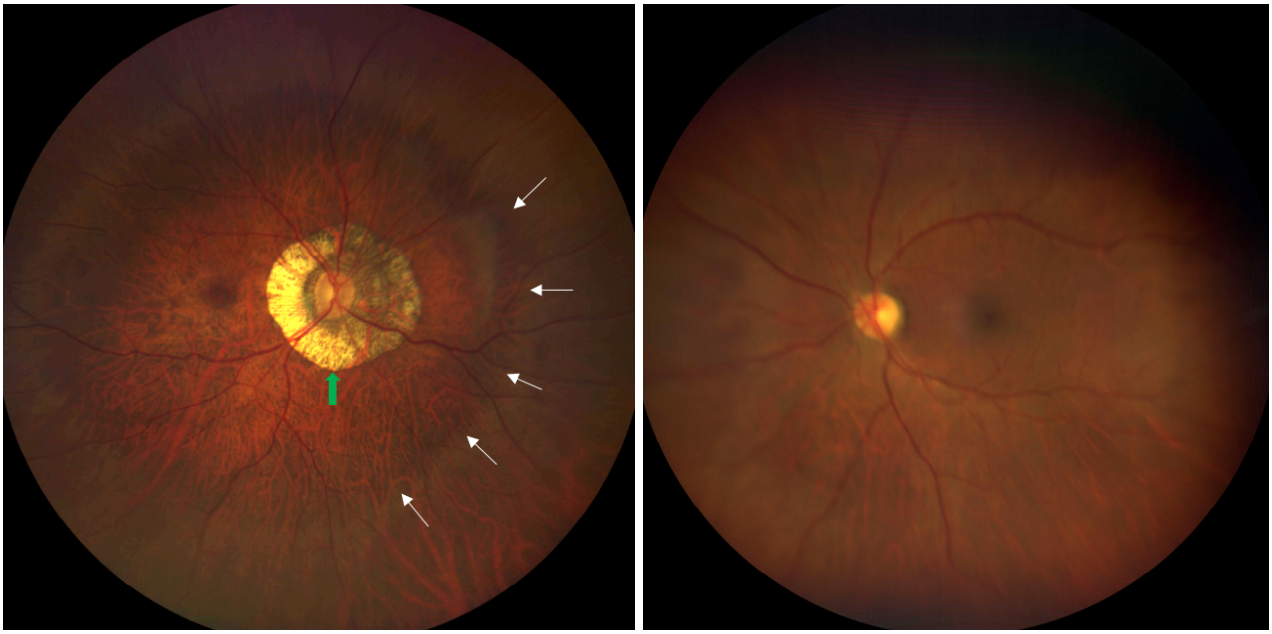

Figure 1: Clarus colored fundus photographs of posterior pole OD and OS, respectively. OD fundus photo reveals Type VII Staphyloma. Note the green arrow indicating the inner peripapillary staphyloma surrounding the optic nerve head and the white arrows delineating the larger posterior pole staphyloma. OS fundus is unremarkable, though the view is hazy due to poor quality image.

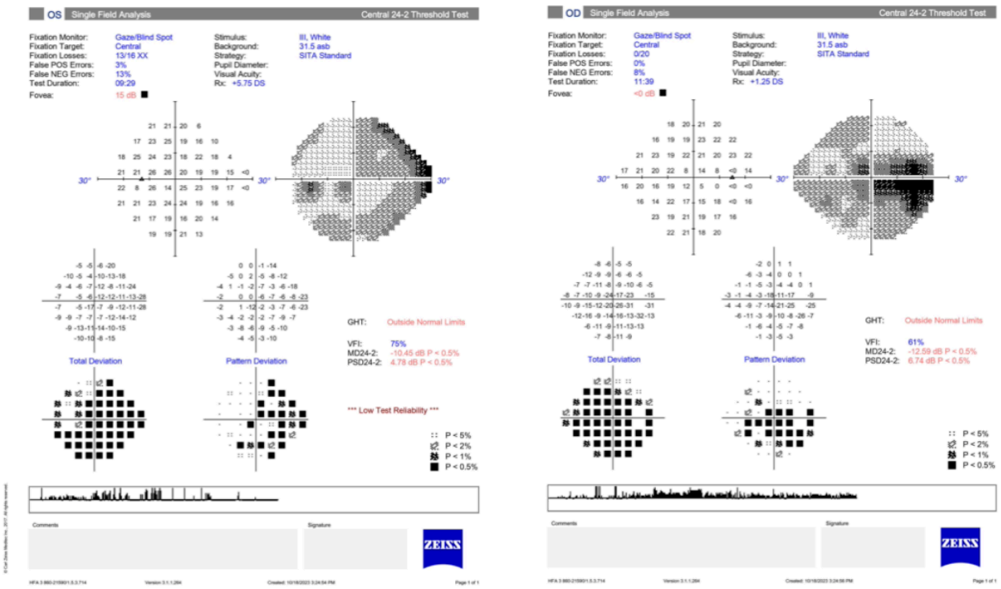

Figure 2: HVF 24-2, SITA Standard left eye and right eye, respectively. Test reliability questionable OU. Stimulus III, white used OU. Right eye shows repeatable enlarged blindspot defect, extending to the center of fixation, stable compared to past exams. Left eye shows low test reliability due to high fixation losses with superior and inferior nasal defects, difficult to compare to past exams.

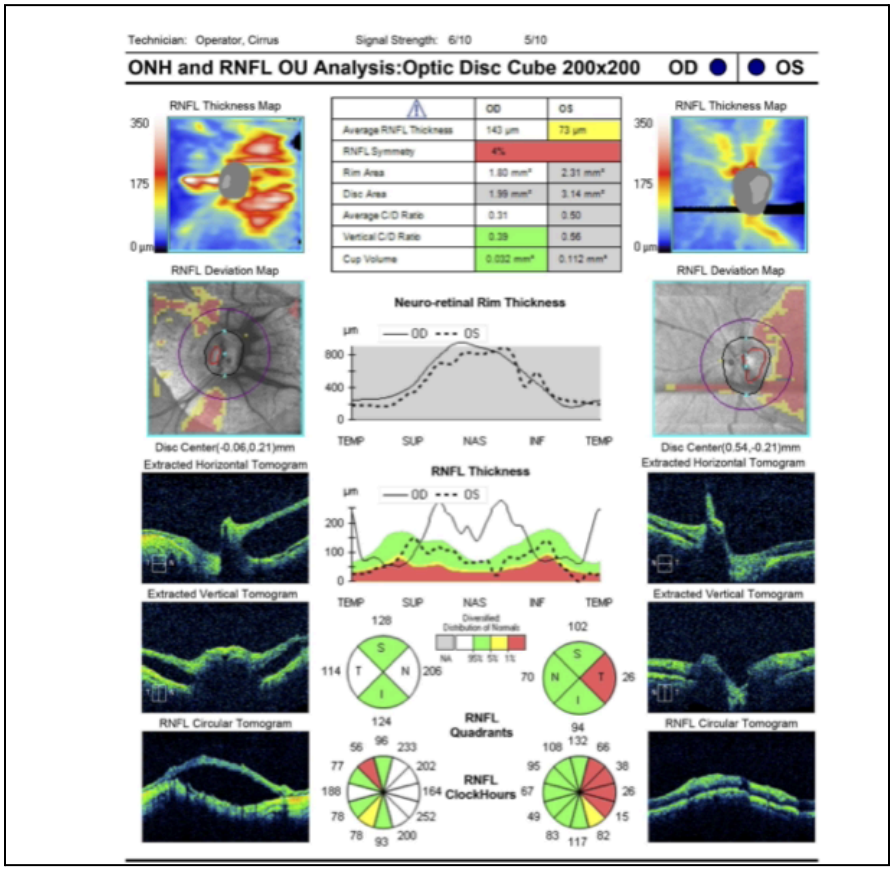

Figure 3: ONH and RNFL OU Analysis: Optic Disc Cube 200x200
Poor quality scan OS>OD. Myopic staphyloma and peripapillary atrophy seen in deviation map. Of note: horizontal, vertical, and circular tomograms visualize posterior herniation of peripapillary region and tissue schisis resulting from pathologic myopia changes. Average RNFL OD is falsely elevated due to peripapillary schisis. Poor reliability OS due to blink artifact on the inferior aspect of scan with temporal RNFL thinning which is questionably stable compared to past scans.

Case Management Summary

Assessment 1: Unspecified Optic Atrophy (H47.20)

- Unspecified optic atrophy OU. Reduced BCVA today in his good eye OS of 20/50. Patient was previously best corrected to 20/25 OS in 2020. This reduced vision is not definitively correlating with minor crystalline lens changes noted today
- Temporal pallor OD, OS with 360 constricted fields on confrontations
- Reduced color vision, however symmetric in both eyes
- ONH RNFL OCT: myopiconus with staphyloma and schisis OD and temporal RNFL thinning OS with questionable worsening from previous
- HVF 24-2: no definite worsening OD, unreliable OS
- Patient denies a history of trauma, alcohol toxicity, surgeries, hormonal imbalances, or nutritional deficiencies
- Patient does admit to a long history of Cocaine abuse from approximately age 20-60. He has been sober for at least the last 10 years.

Plan 1:

- Patient and brother educated on today's findings and the importance of follow up compliance to assess visual function. Records requested to obtain past records from Neuro-ophthalmology and for MRI/radiology reports.
- Patient referred to the internal Neuro-ophthalmic Disease service to consider if any additional work-up is indicated to rule out non-glaucomatous optic neuropathies.
- Patient educated on the option of returning to previous Neuro OMD and they preferred to establish care with the team at The Eye Institute.
- Patient educated on importance of keeping Neuro appointment here to determine if there is any treatable etiologies for vision loss OS (long history of reduced BCVA in OD presumed due to pathologic myopia and refractive amblyopia).

Assessment 2: Combined Forms of age Related Cataract, bilateral (H25.813)

- Exam revealed combined cataracts, not visually significant at this time. Patient denies problems with Activities of Daily Living (ADL's) at this time due to cataracts.

Plan 2:

- Patient educated on today's findings and clinical course of age related cataracts. Surgery not indicated at this time. Will continue to monitor in 1 year or sooner if visual changes are noted.

Assessment 3: Refractive Amblyopia, right eye (H53.021)

- Refractive amblyopia OD secondary to high myopia and anisometropia
- BCVA: 20/200 OD and 20/50 OS (previously OD 20/125 and OS 20/25)

Plan 3:

- Patient educated on our recommendation of a referral to the Low Vision service to further evaluate visual oriented goals and devices. Patient states he used to be an avid reader and does not feel like this near vision is good enough to read anymore. He would like to feel more independent with his vision as well. Patient also stresses difficulty with writing.

Addendum: LOW VISION EXAM: Patient was seen in William Feinbloom Vision Rehabilitation Center where they recommended apps for reading, as well as increasing ADD power. Patient was also referred to Bureau of Blindness and Visual Services for an Orientation & Mobility evaluation and training. He was also placed on the Vision Rehabilitation Therapy waitlist for assistance in increasing ADL independence in cooking and cleaning at home.

Case Pearls

Staphyloma… What is it?

- Thinning of the lining of the globe which can cause an outpouching ocular structures and lead to vision loss
- Most commonly found in pathologic myopia since the elasticity of the ocular structures is compromised but can also be seen post trauma or infections
- Pathophysiology remains unclear; however current wisdom believes that staphyloma is due to decreased resistance of the sclera which then leads to

protruding of Bruch's membrane followed by structural changes to the retina
- Greatest symptom is decreased vision

Types of Staphyloma based on their location:

- Anterior Staphyloma: involves the cornea
- Intercalary Staphyloma: involved the area between the cornea and sclera
- Ciliary Staphyloma: involves the ciliary body
- Equatorial Staphyloma: occurs in the middle of the eye
- Posterior Staphyloma: *Most common type of staphyloma*. Occurs in the retina
- Peripapillary Staphyloma: RARE - often congenital. Outpouching around the optic nerve - differentials in this case would be morning glory syndrome or optic nerve coloboma

Types of Posterior Staphyloma:

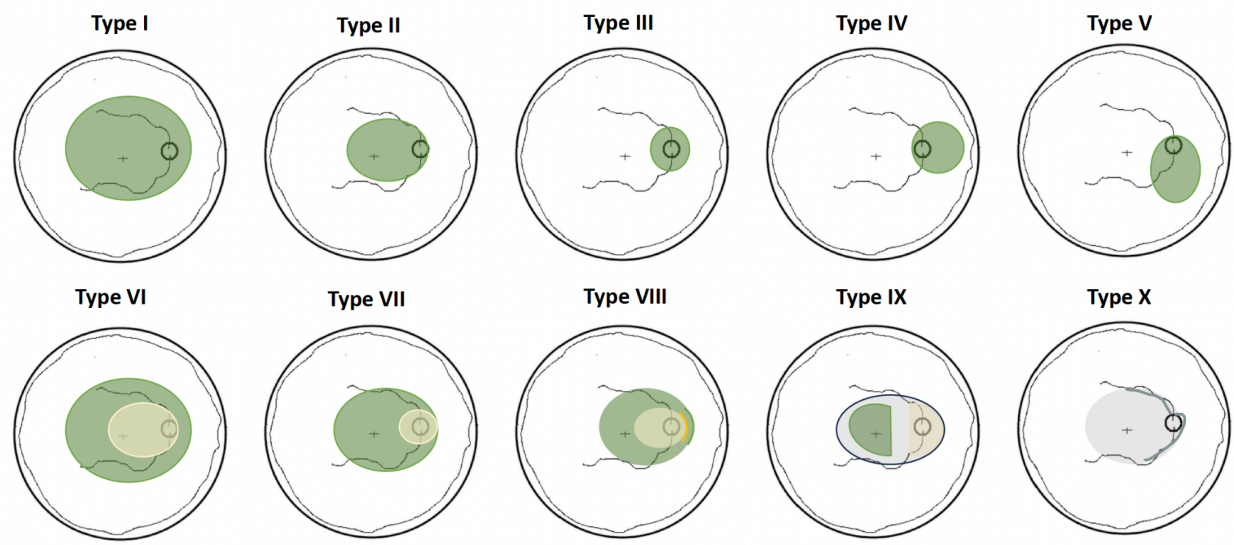

Figure 4: Types of Staphylomas based on Curtin[1].
Type I = posterior staphyloma
Type II = macular staphyloma
Type III = peripapillary staphyloma
Type IV = nasal staphyloma
Type V = inferior staphyloma
Type VI = combined staphyloma types I and II
Type VII = combined staphyloma types I and III - *this is what our patient had as visualized in Figure 1.*
Type VIII = tiered staphyloma
Type IX = septal staphyloma
Type X = plicated staphyloma

[1] Recreated from Curtin, B J. "The posterior staphyloma of pathologic myopia." *Transactions of the American Ophthalmological Society* vol. 75 (1977): 67-86.

Staphyloma Struggles and Co-Management

- Staphyloma most commonly occurs in pathologic myopia. Thinning of the retina may cause a significant decrease in vision, which can interfere with patients' ADL's.
- Furthermore, increase in axial length along with thinning of the ocular structures can result in schisis of retinal layers, breaks in the retina, or retinal detachments which further lead to loss of vision.
- Co-Management with Low Vision specialists, such as in this case, can help patients reach their daily goals.
- Co-Management with Retina specialists can help monitor for progression of and rule out additional retinal pathology such as breaks or detachments.