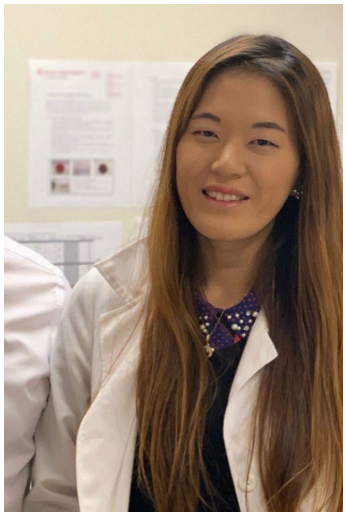
SALUS UNIVERSITY
The Eye Institute

Pennsylvania College of Optometry
The Focal Point
October 2022 Edition

Tanner Johnson
Traditional Class of 2024

Hometown: New Bern, North Carolina
Undergrad: Campbell University
Major: Biology
Favorite Subject: Posterior Segment Disease
Optometry Goal: Pass boards on the first try
Favorite food: Cheeseburger
Hobby: Playing with my dog
Last Show I binged: Game of Thrones



Alice Lim
Class of 2018, Pennsylvania College of Optometry

Hometown: North Wales, Pennsylvania
Undergrad: Arcadia University
Major: 3+4 accelerated program for Optometry, Interdisciplinary Sciences
Favorite Diagnostic Instrument: B-scan
Hates: Mushroom and cilantro
Hobby: painting things

The SLOW but FUN Parts of Congenital Nystagmus: How to Recognize and Manage

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.

The Case

Demographics

42 yo Black male, concession stand owner

Chief complaint:

Blurry vision at near

History of present illness

Character/signs/symptoms: eye strain with near work
Location: OU
Severity: mild
Nature of onset:
Duration: long standing
Frequency: everyday
Exacerbations/remissions: none
Relationship to activity or function: none
Accompanying signs/symptoms: none

Patient ocular history

h/o congenital nystagmus s/p “muscle” surgery, age 3 @ Wills Eye
Poor vision since childhood, “never saw 20/20”

Family ocular history

Non-contributatory

Patient medical history

Hypertension

Medications taken by patient

Carvedilol, Losartan, Nifedipine

Patient allergy history

NKDA

Family medical history

Father: Hypertension, DM 2

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 oscillopsia, vertigo, dizziness
Psychiatric: denies
Immunologic: denies
Hematologic: denies

Mental status

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

Clinical findings

Entering VA:

	Distance (sc)	Near (sc)
OD:	20/50-2	0.4/0.6M
OS:	20/60	0.4/0.5M
OU:	20/40	

Pupils: PERRL (-) APD OU
EOMs: Full and no restriction with nystagmus present in all gazes
Other Exam findings: Right beat nystagmus OD/OS in primary gaze, (+) latent component, worse with occlusion (+) Null zone with convergence (+) Jerk form (+) right beat on upgaze
Confrontation fields: constriction 360 OU
Hirschberg: Symmetric

Subjective refraction:	VA Distance	VA Near
OD: +0.50-1.75x167	20/30	0.4/0.4M
OS: +1.50-2.75x180	20/50	0.4/0.4M
ADD: +1.00		

Slit lamp:

Lids/lashes/adnexa: Normal
Conjunctiva: white and quiet bulb conj, pink and quiet palp conj with limbal melanosis OU
Cornea: normal endothelium, stroma, epithelium, and tear film OU
Anterior chamber: deep and quiet OU
Iris: flat and intact OU (-) TID OU
Lens: clear lens capsule, cortex and nucleus OU, Mittendorf dot OD
Vitreous: syneresis and synchysis OU

IOPs/method:

The Case

iCare: 15/13mmHg

Fundus OD:

C/D: 0.5/0.5 anomalous appearance/segmental hypoplasia, oblique insertion

Macula: flat and intact

Posterior pole: abnormal nasal vasculature branching

Periphery: flat x 360, no RD, no breaks; I/N CHRPE

Fundus OS:

C/D: 0.5/0.5 anomalous/segmentally hypoplastic disc, extreme oblique insertion

Macula: flat and intact

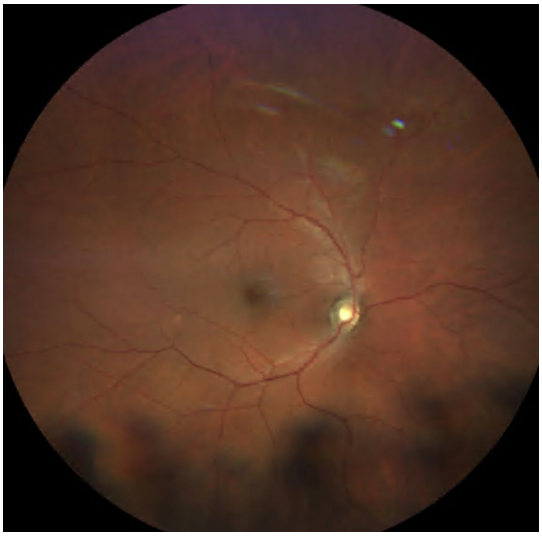
Posterior pole: abnormal nasal vasculature branching

Periphery: flat x360, no RD, no breaks

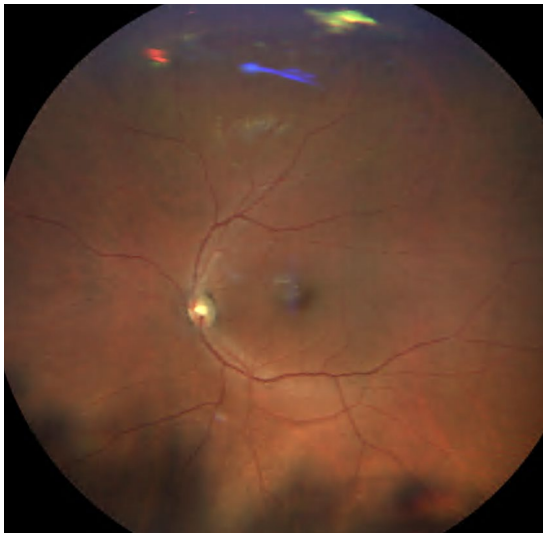
Blood pressure: 140/84mmHg RAS

Case Images:

OD Fundus



OS Fundus



Case Management Summary

A1: Compound hypermetropic astigmatism w/ Presbyopia OU

-Reduced vision 2' congenital nystagmus
-BCVA: 20/30 OD 20/50 OS and 0.4/0.4M OU at near

P1: Patient educated on finding. Updated finalized Rx given, discussed adaptation needed since patient is not used to wearing spectacle correction.

A2: Congenital nystagmus OU

-by history and confirmed with examination findings of SLOWFUN
-h/o EOM surgery (unsure type, could be Kestenbaum procedure)
-anomalous appearance of ONH OU, segmental hypoplasia could be contributory
-no apparent signs of ocular albinism, foveal hypoplasia, or fundus abnormality noted

P2: Patient educated on findings. No further work up needed at this time given long term stability and no development of acute symptoms or change. Records requested from Wills Eye. Soft CL not recommended at this time given patient has never consistently worn correction but consider in future. Monitor yearly.

A3: Optic nerve hypoplasia OU

-small, segmentally hypoplastic disc OU w/ oblique insertion
-constriction on confrontation noted, per patient, long standing peripheral vision deficit
-fundus photo documentation taken

P3: Patient educated on finding. Scheduled to return for HVF 24-2, and attempt OCT of ONH (expected to be difficult due to nystagmus, may have to utilize null zone of convergence).

Case Pearls

- Infantile nystagmus (previously known as congenital nystagmus) is a life-long, involuntary to-and-fro movement of the eyes that may affect quality of vision and function. It can occur in the first few months after birth, can be idiopathic, and inherited or associated with visually impairing conditions.
- In our case, the patient presented with prior knowledge of his nystagmus; however, we further confirmed that his nystagmus was most likely congenital and not acquired by utilizing SLOW FUN, an acronym for components of congenital nystagmus.
- **S: Symptomless** → There should be no reported acute double vision,

oscillopsia, or vertigo.
    - Our patient symptoms were related to emerging presbyopia, not his nystagmus.
- **L: Latent component** →Nystagmus intensity increases with occlusion with fast phase beating towards uncovered eye, and monocular VA tends to be worse.
    - Our patient's monocular VA was slightly worse than binocular VA, and movement worsened with occlusion.
- **O: Optokinetic Nystagmus (OKN) is abnormal** →OKN tends to be reversed.
    - This was not confirmed with our patient.
- **W: Waveform is mixed** →Both pendular and jerk form can be observed related to congenital nystagmus, but not rotary.
    - A right beating jerk waveform was observed on our patient.
- **F: Fixation** makes nystagmus worse.
    - Nystagmus worsened while performing VA's on our patient (could be also related to occlusion).
- **U: Upgaze keeps it horizontal** →Congenital nystagmus presents predominantly in the horizontal axis, vertical component is likely
    - We noticed the same jerk pattern, right beating nystagmus on our patient while performing Versions &pursuits.
- **N: Null zone** →The intensity of nystagmus is minimized at a certain gaze angle, which most often includes convergence.
    - We were able to observe decreased nystagmus intensity when our patient was converging on a near fixation point.
- Our patient also reported a history of "muscle" surgery at age 3. This potential procedure could have been Kestenbaum surgery which involves the recession and resection of the extraocular muscles to move the null zone into primary position of gaze. Although we could not confirm this with history or prior records.
- Perform a thorough ocular health assessment in order to rule out ocular conditions often associated with congenital nystagmus. Look for anterior segment opacities such as iris illumination defects to suggest albinism and congenital cataracts. Look for optic disc abnormalities such as optic nerve hypoplasia and fundus abnormalities such as foveal hypoplasia and abnormal pigmentation.
- Keep optical treatments in mind. Soft contact lenses can dampen nystagmus. If the null

zone is convergence, BO prisms can be used to induce positive fusional vergences and decrease nystagmus.