

Adult-Onset Neurological Vision Impairment

Dr. Erin Draper



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Dr. Draper has no relevant financial relationships to disclose



Overview

- Discuss optometry's role in the NVI Team
- Useful exam elements outside of "routine" eye exam
- Case studies of impaired visual function
- Discussion of how to work with an optometrist to maximize impact

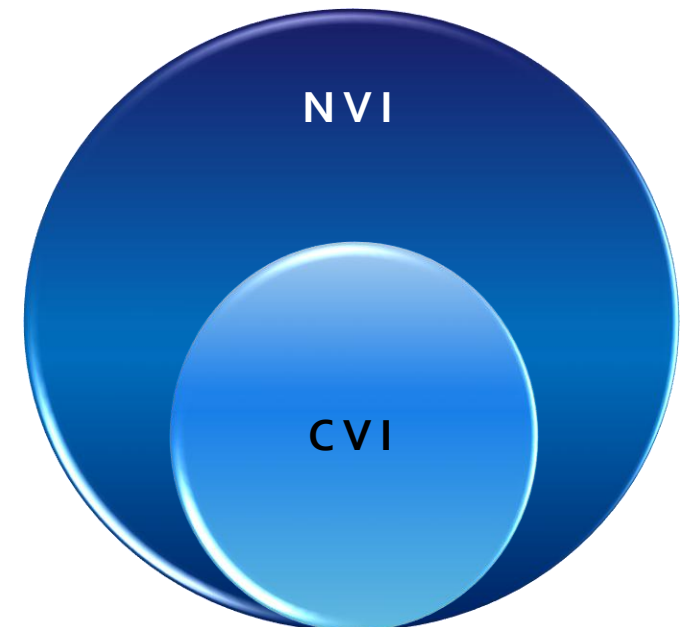
Neurological visual impairment (NVI) results from any condition within the brain that interrupts the typical flow of vision through the brain across the lifespan.

Includes adult acquired brain-based VI

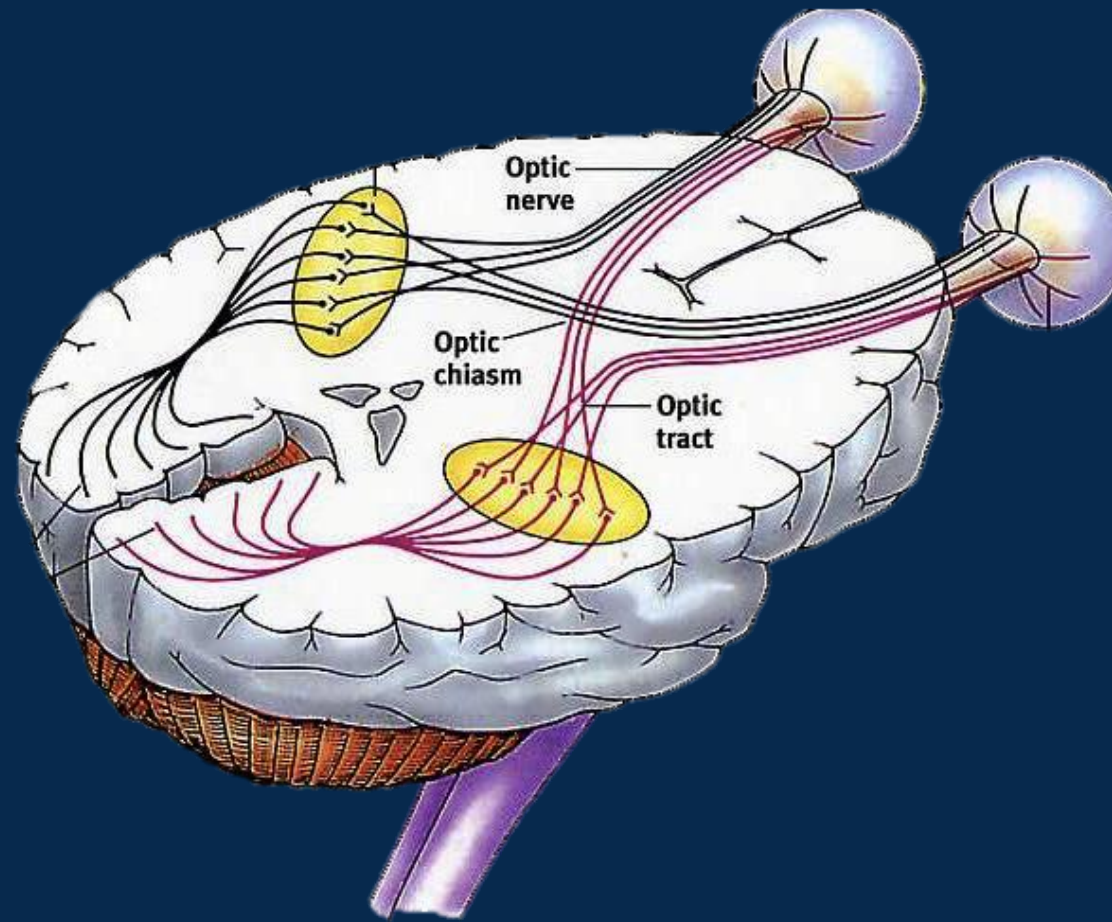
Includes pediatric CVI Spectrum

Location and the extent =

1000's of different manifestations of NVI



Anatomy & Physiology



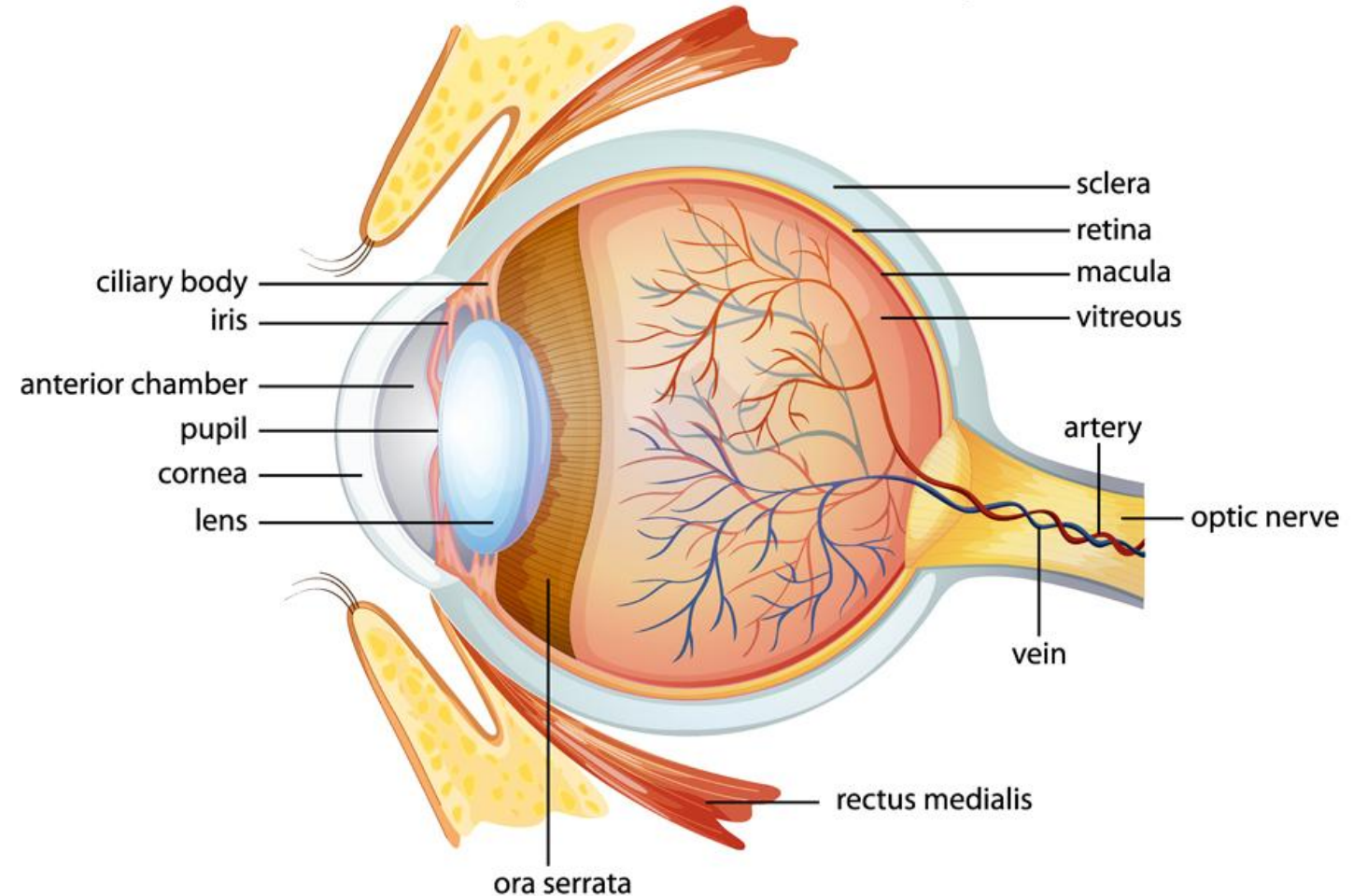
DREXEL UNIVERSITY
Elkins Park
Campus

Eye's job = Send a Clear Message

The purpose of the eye is to send a focused visual message to the brain.

- **Captures light**
- **Converts it into electrical signals**
- **Transmits this visual information to the brain to create images**

Beyond the globe is where the fun begins...

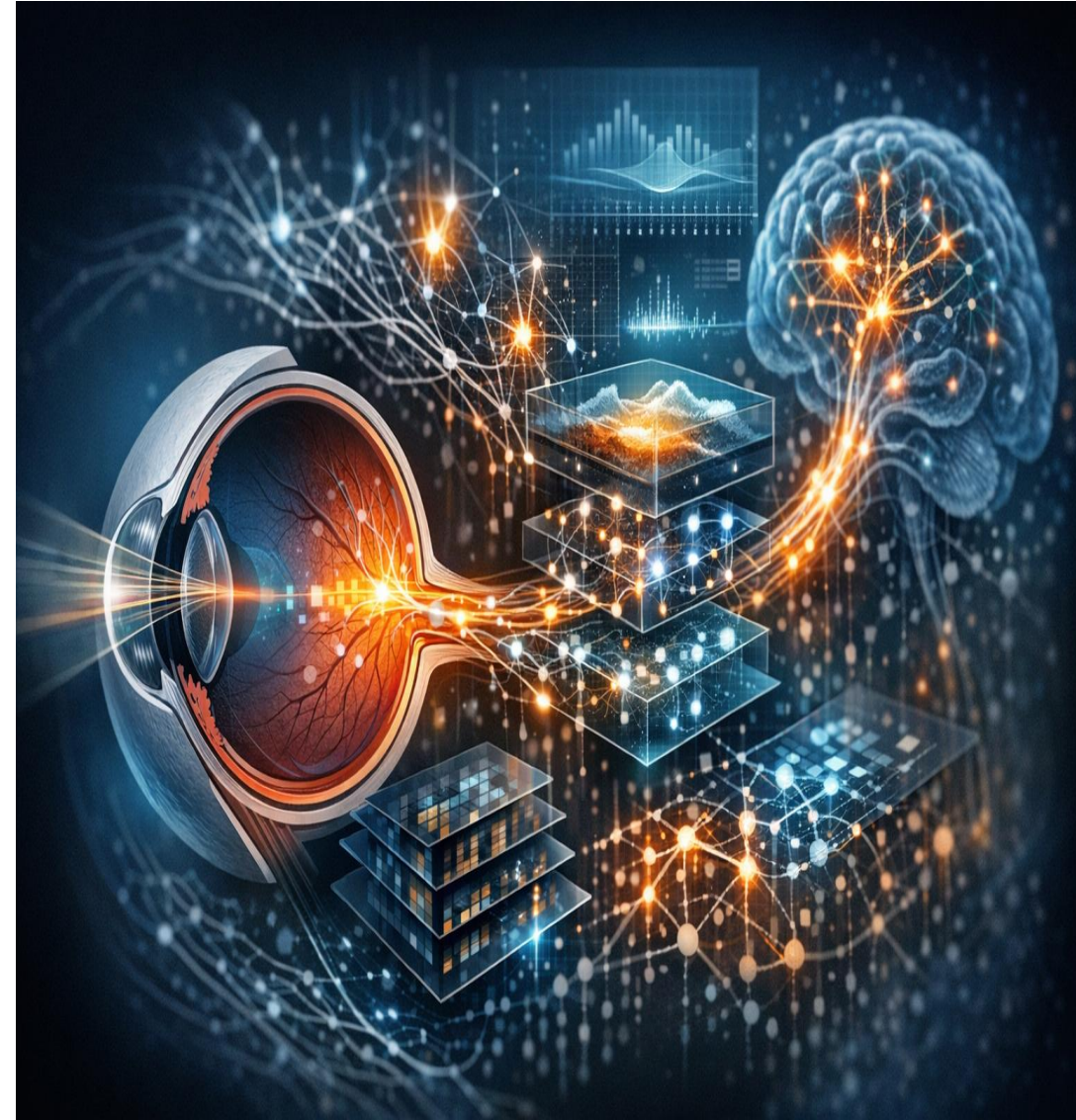


Brain's job = Process the message

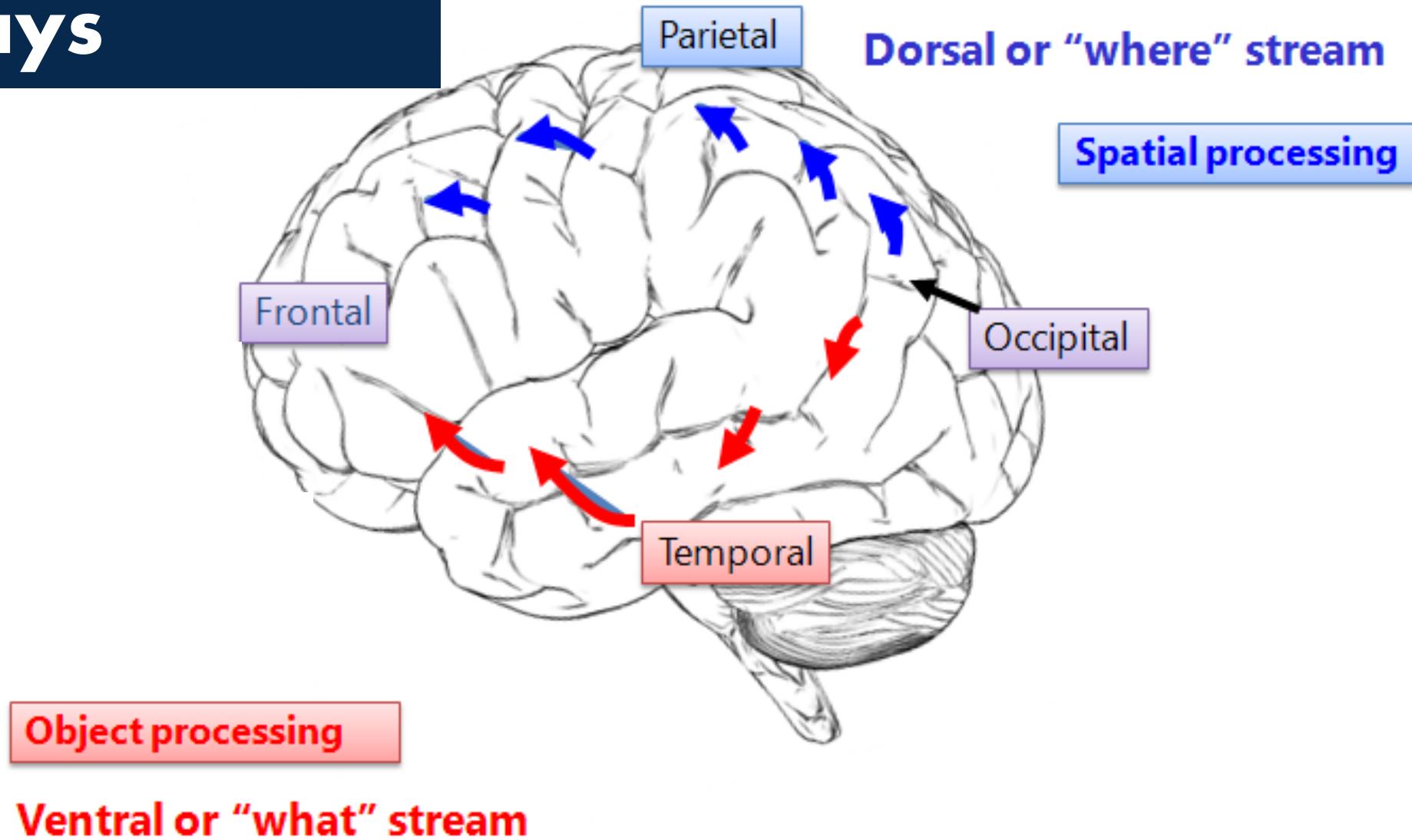
Copilot generated image

Visual Perception

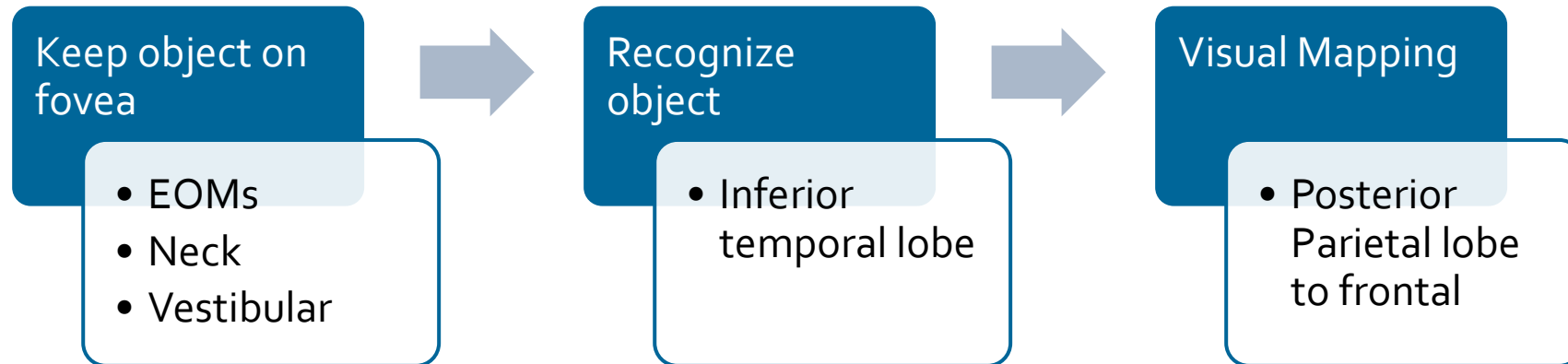
- Keep foveas pointed at objects of interest
- Coordinate eye movements
- Locating, fixating, tracking, shifting
- Visual Recognition
- Perception of motion and space
- Coordinate body movements with vision



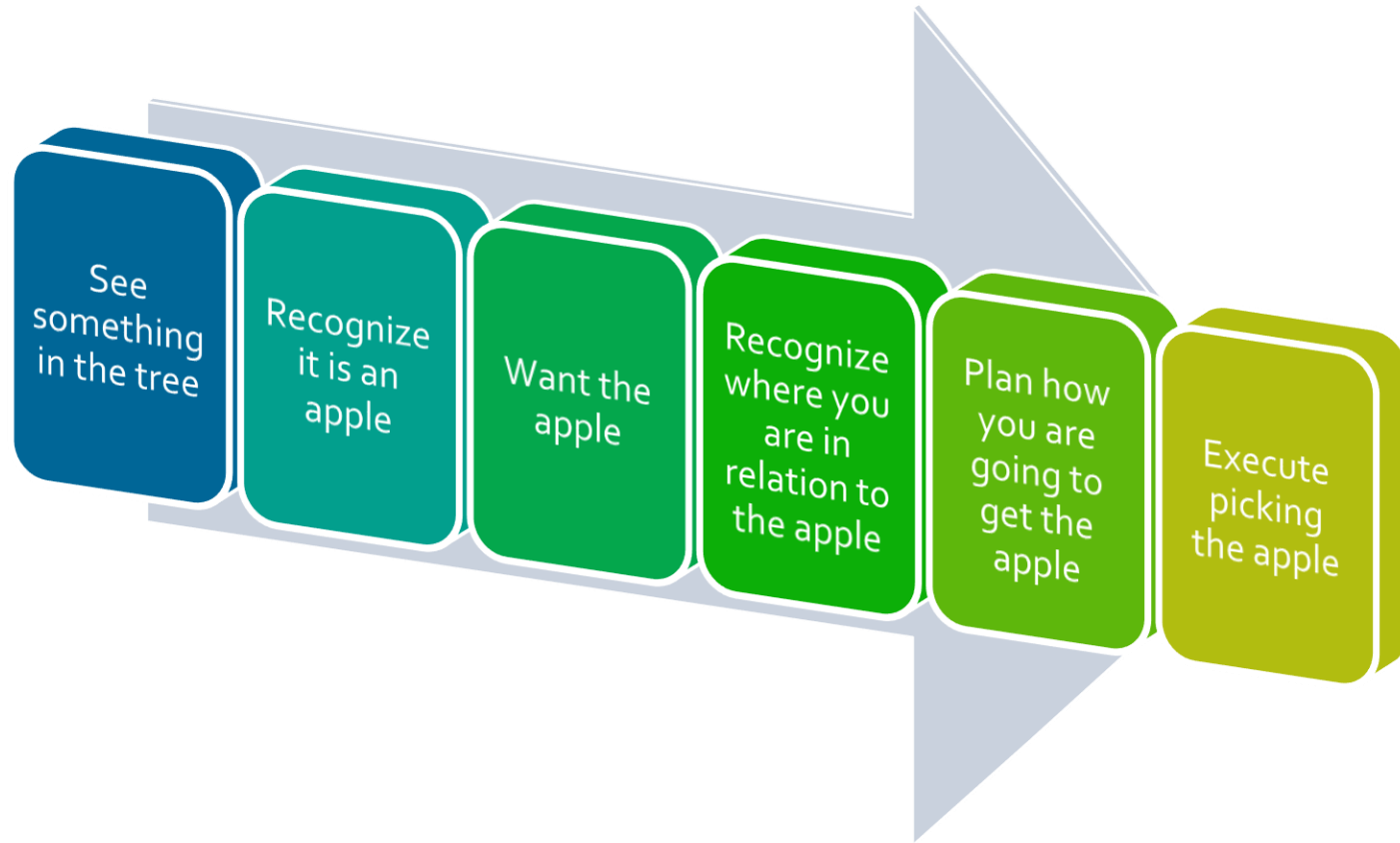
Processing Pathways

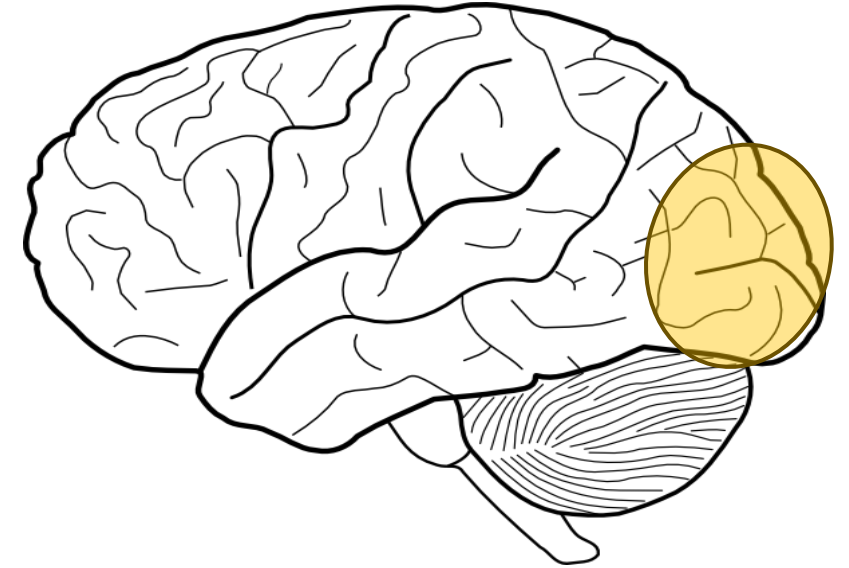
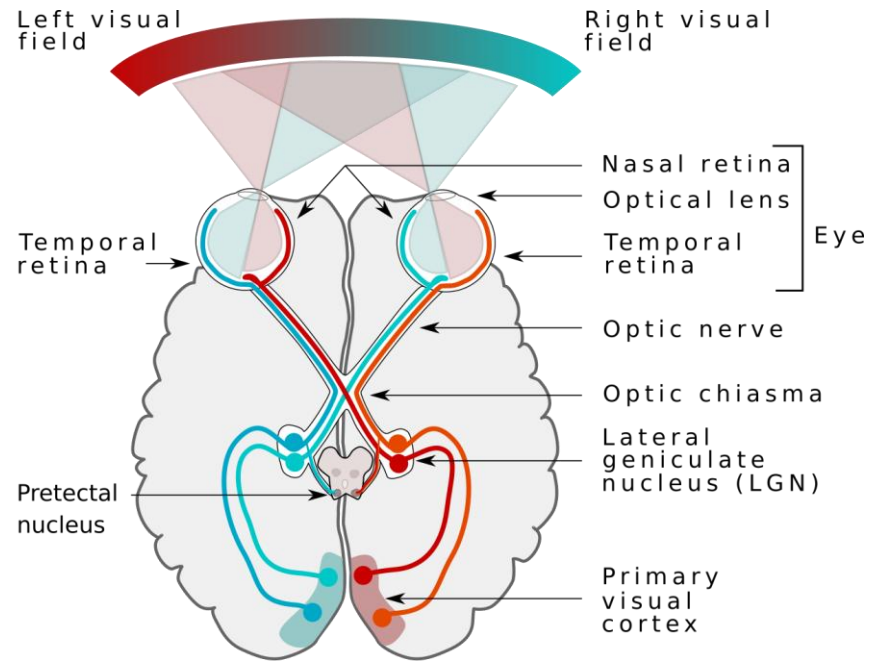


Visual Information Processing



Visual Function





See something
in the tree

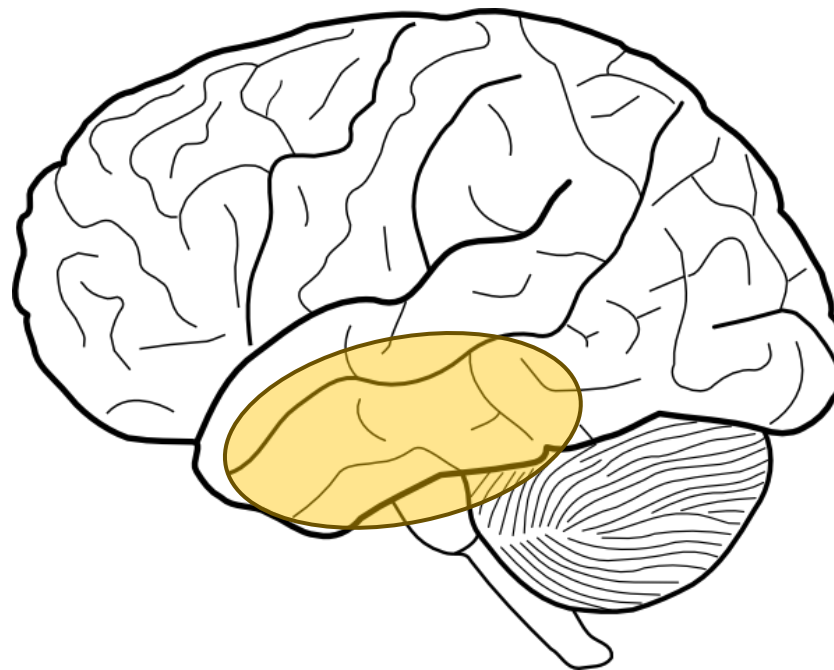
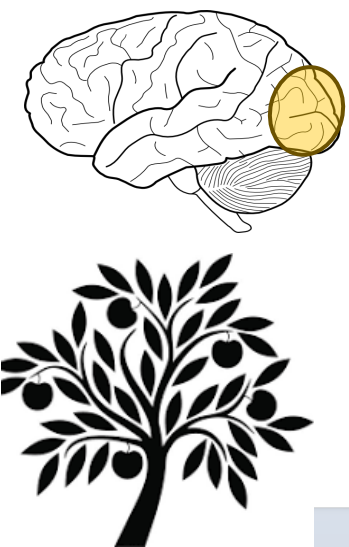
Recognize it
is an apple

Want the
apple

Recognize
where you
are in relation
to the apple

Plan how you
are going to
get the apple

Execute
picking the
apple



See something
in the tree

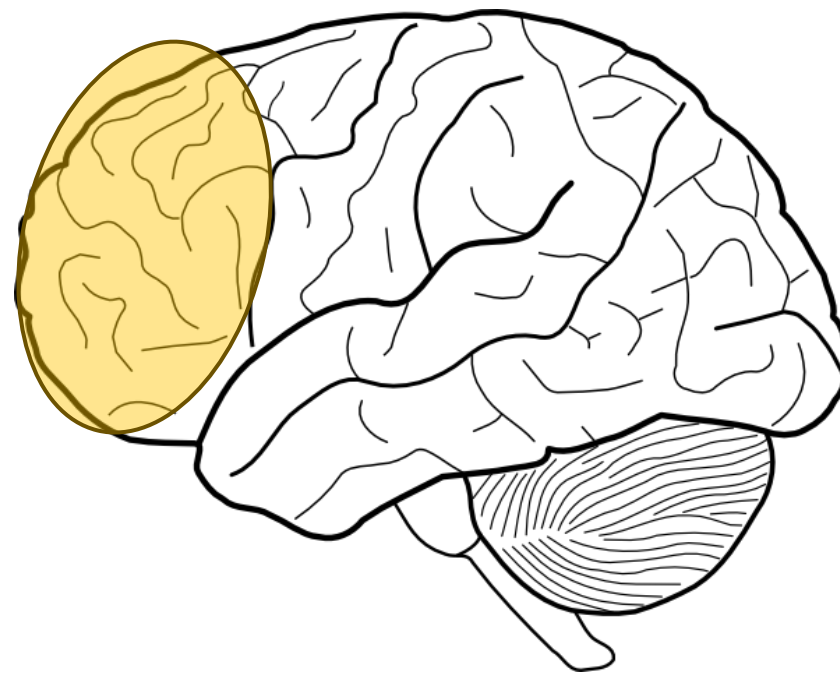
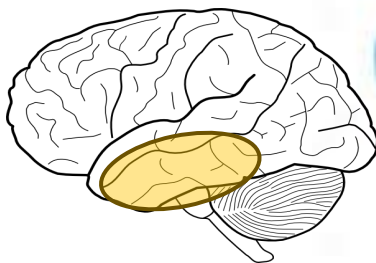
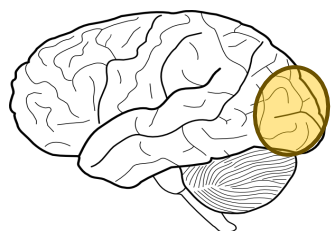
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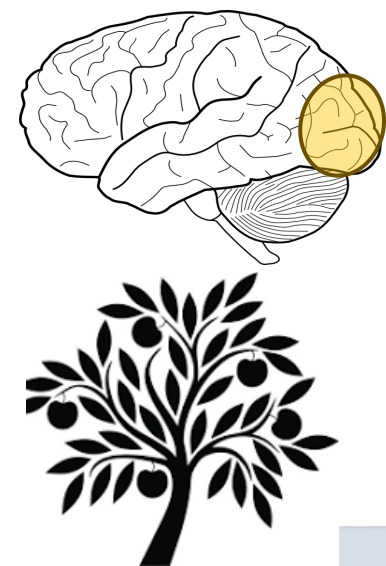
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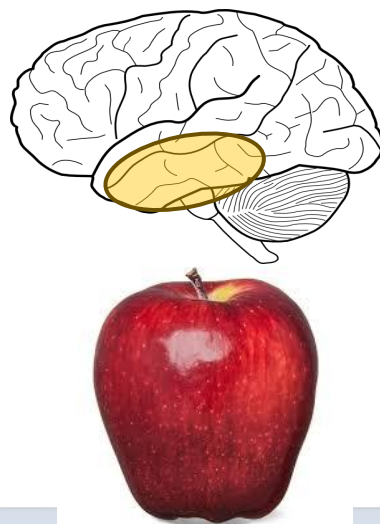
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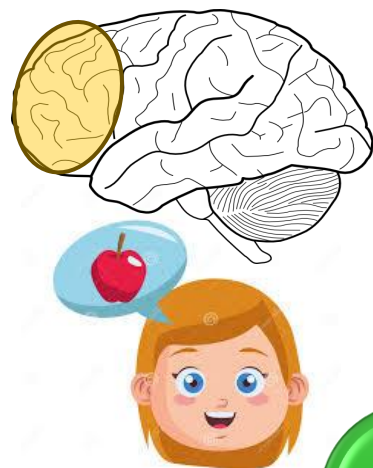
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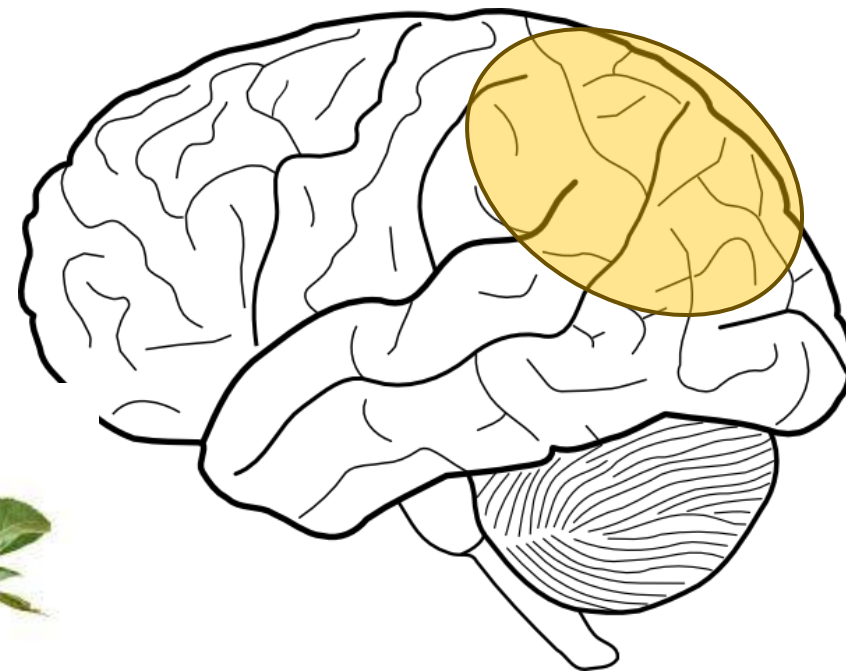
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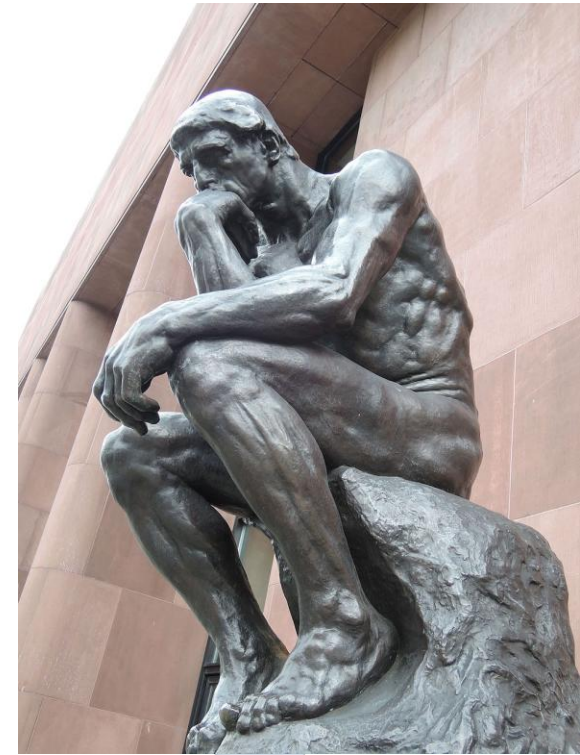
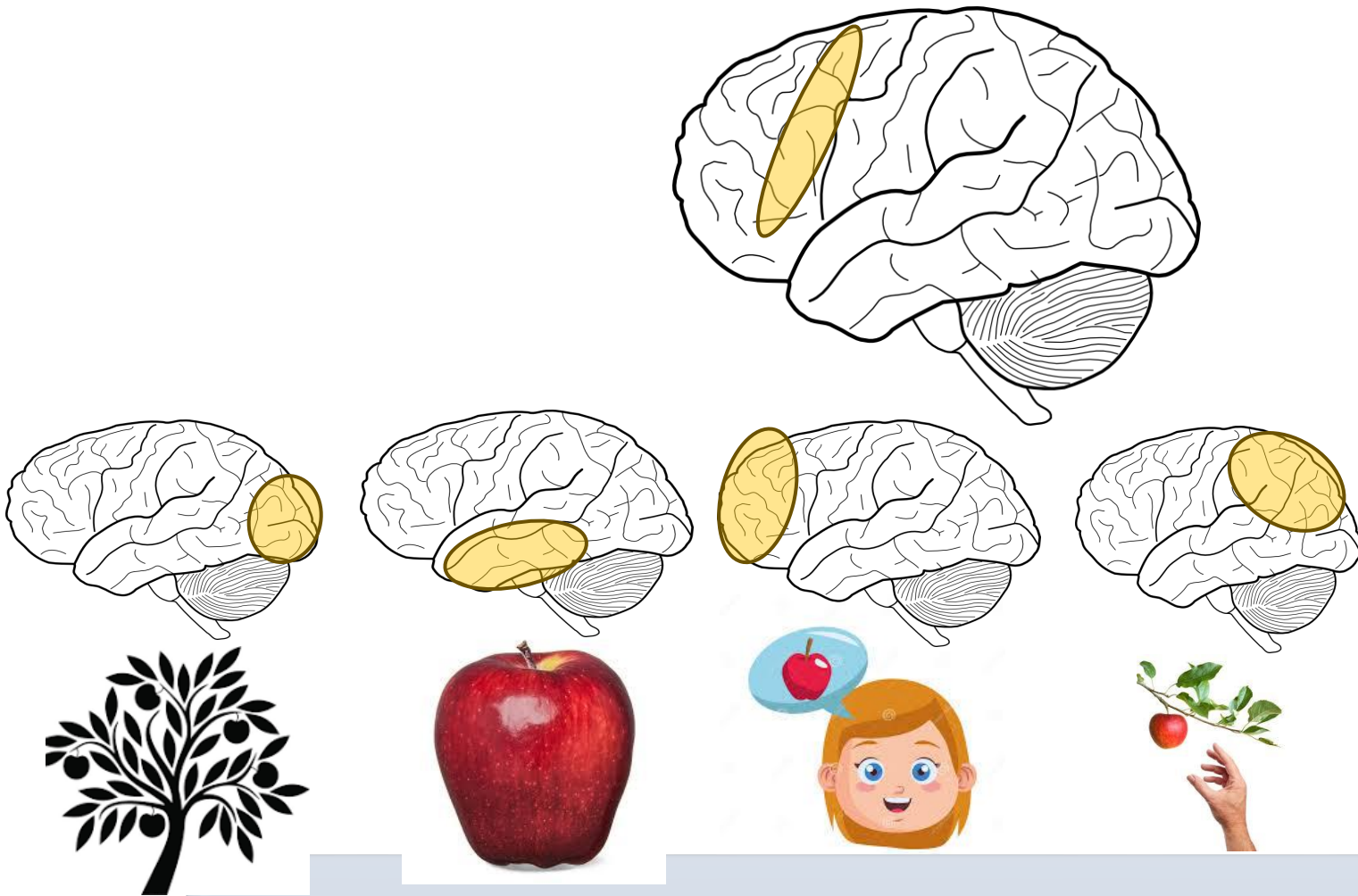


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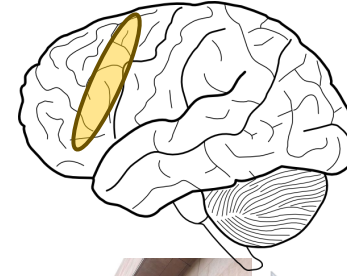
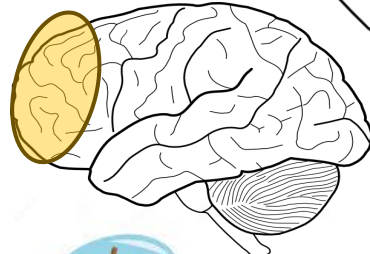
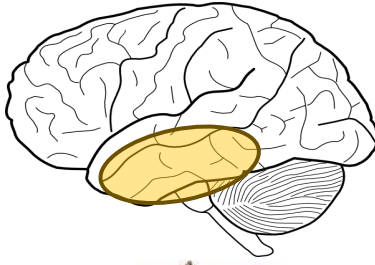
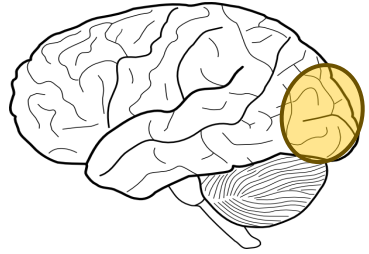
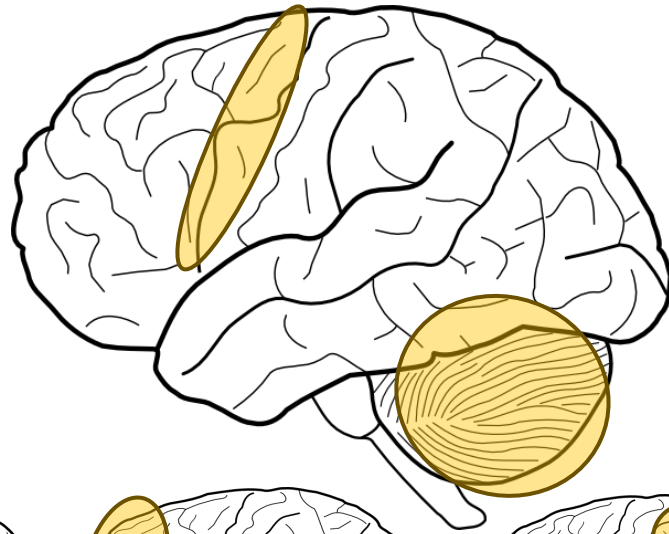
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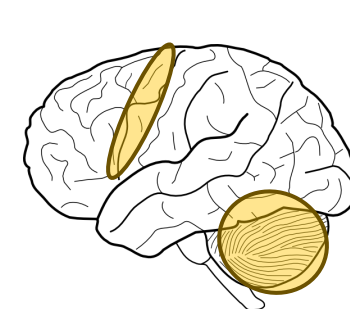
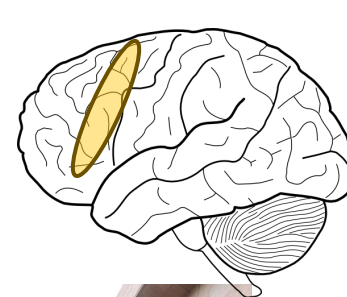
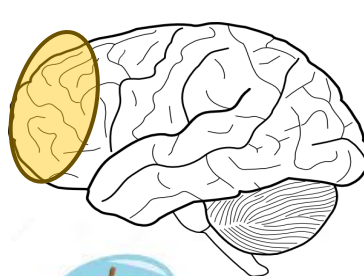
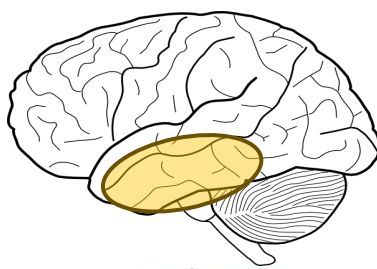
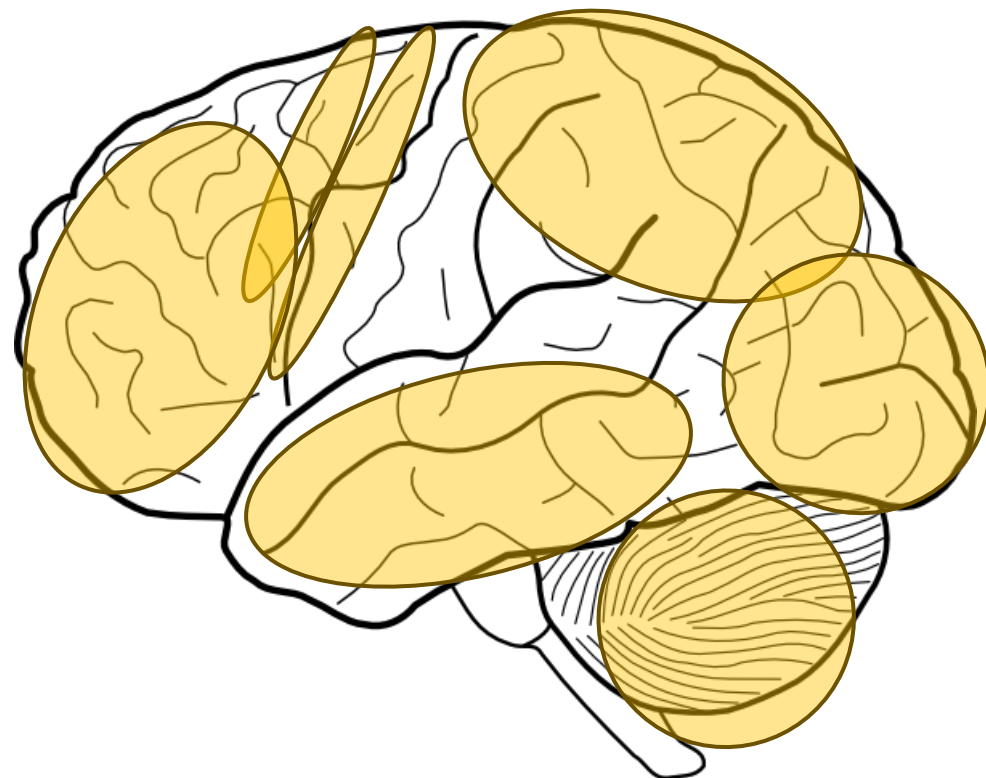
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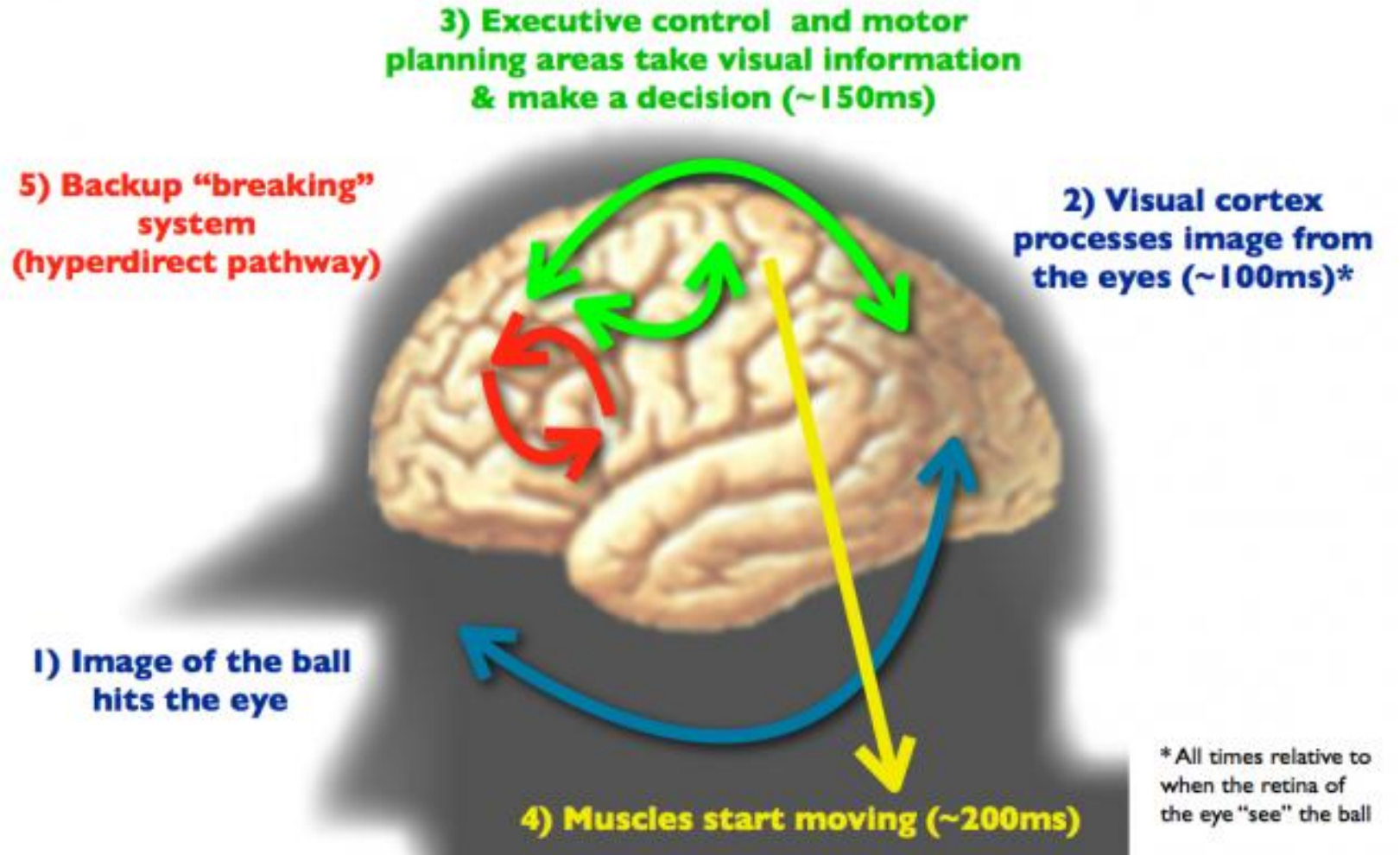
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Neuroanatomy of hitting a baseball



*All times relative to when the retina of the eye "see" the ball

Source:

<https://www.psychologytoday.com/us/blog/white-matter-matters/201309/forget-chess-meet-the-ultimate-brain-challenge>

Medical Etiologies which can lead to NVI

Etiologies for Neurological Vision Impairment

Perinatal / Neonatal Brain Injury



- prematurity
- periventricular leukomalacia (PVL)
- intraventricular hemorrhage (IVH)
- hypoxic-ischemic encephalopathy (HIE)
- hypoxia/anoxia
- extreme temperature exposure
- excessive jaundice
- full-term birth associated with HIE



Vascular / Ischemic Injury

- stroke
- ischemia
- cerebral hypoxia/anoxia



Traumatic / Acquired Brain Injury

- traumatic brain injury (TBI)
- non-accidental trauma
- shaken baby syndrome



Infectious / Inflammatory Causes

- CMV
- congenital or acquired infections



Genetic / Metabolic / Structural Causes

- genetic conditions
- genetic syndromes
- metabolic disorders/diseases
- glyceimic disturbances
- hydrocephalus
- cerebral/brain tissue atrophy

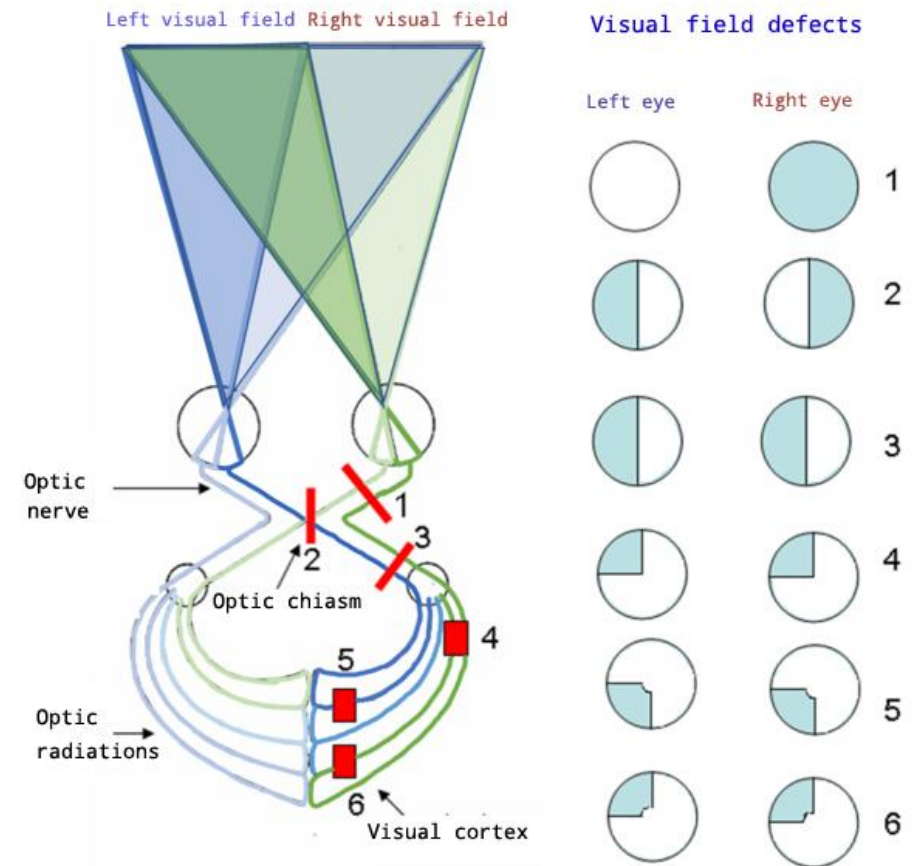


Neurological / Seizure-Related Causes

- infantile spasms
- epilepsy
- seizures
- cerebral palsy (CP)

Effects of disruptions to the Visual Process

**Lesion to the Visual
Pathway
Optometrists can
understand**



**These are
challenging to
detect on a routine
eye exam.**

Subtle Vision Impairments Not Easily Detected on Standard Eye Exams



1. Visual Attention

- Difficulty finding an object in a cluttered environment
- Losing place when reading
- Difficulty shifting attention between objects
- Visual overwhelm in busy environments



2. Visual Processing

- Difficulty recognizing familiar objects
- Difficulty interpreting complex scenes
- Trouble judging spatial relationships
- Difficulty identifying objects based on visual information alone



3. Visual-Motor Integration

- Poor visually guided reaching
- Difficulty copying shapes
- Problems with handwriting or drawing
- Difficulty navigating stairs or obstacles



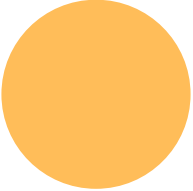
4. Visual Field/Hemifield Problems

- Visual neglect
- Subtle peripheral field loss

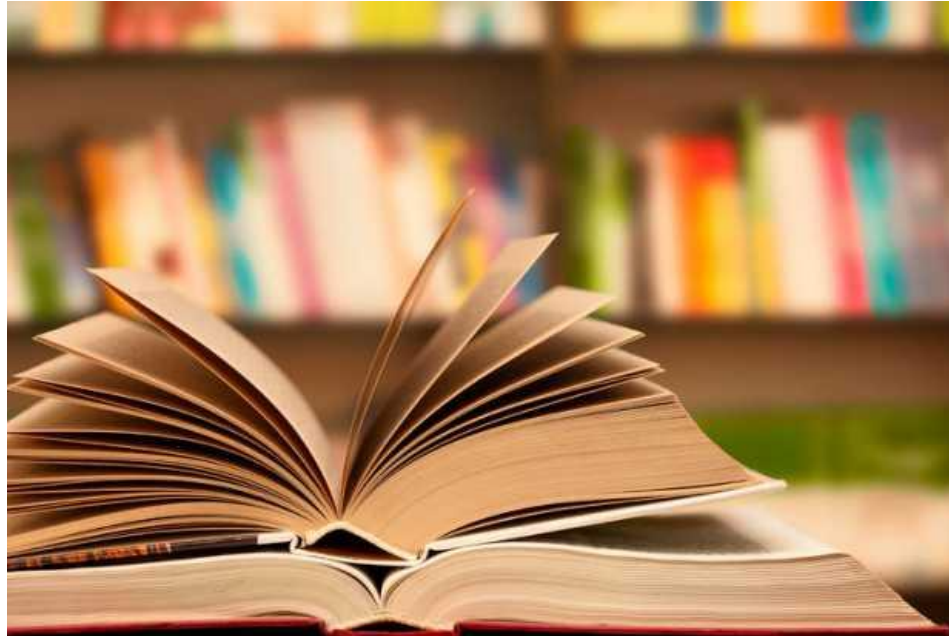


5. Motion and Spatial Processing

- Difficulty detecting or interpreting movement
- Difficulty navigating through crowds
- Problems with depth or spatial orientation

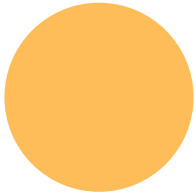


CASE Study



CASE HISTORY

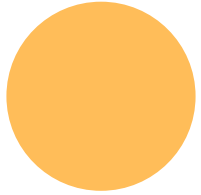
70-Year old woman



Chief Complaint	Difficulty with reading for past 2 years. Has tried magnifiers but do not help.
Systemic History	HTN, DM Type 2, hypercholesterolemia, hypothyroidism, (-) stroke
Ocular History	Cataract surgery 1.5 years ago, no improvement in vision Previous suspicious homonymous hemianopia – evaluated by neurology, MRI completed and normal
Medications	HCTZ, rosuvastatin, metformin, levothyroxine
Social History	(-) smoker, denies alcohol or drug use



AFFERENT VISUAL SYSTEM



VISUAL ACUITY

Snellen Chart

OD: 20/400

OS: 20/400

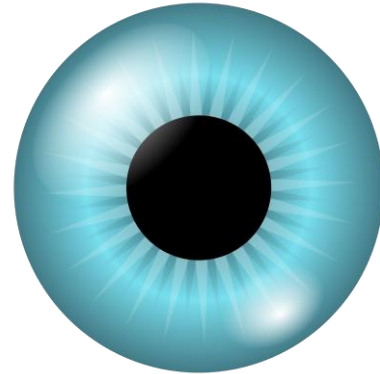
PH NI OU

Single Letter

OD: 20/40

OS: 20/40

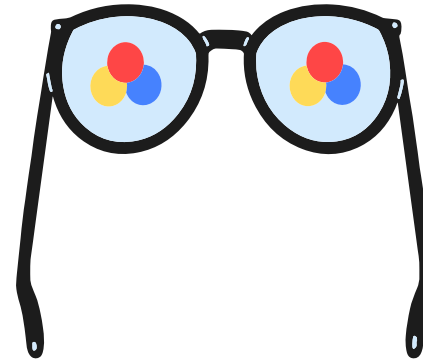
PH NI OU



PUPILS

OD: round
Bright: 2.5mm
Dim: 4mm

OS: round
Bright: 2.5mm
Dim: 4mm
– APD



COLOR VISION

OD: unable
OS: unable
Ishihara

Cannot ID test
plate but can ID
color when
asked

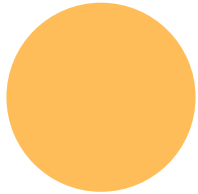


CONFRONTATIONAL FIELDS

Severe
Constriction OU



AFFERENT VISUAL SYSTEM



VISUAL ACUITY

Snellen Chart

OD: 20/400

OS: 20/400

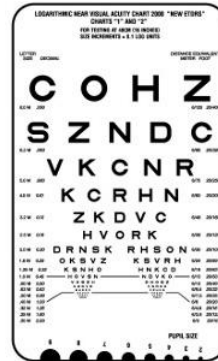
PH NI OU

Single Letter

OD: 20/40

OS: 20/40

PH NI OU



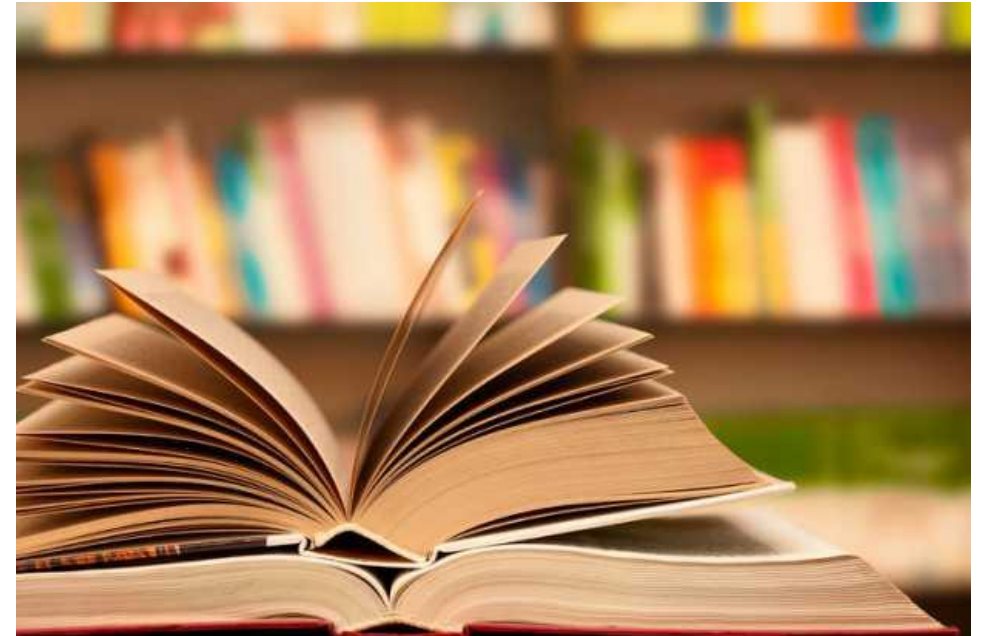
NEAR VA

OD: 20/50

OS: 20/50

(Snellen Equiv)

**Unable to read words longer than
2 letters regardless of amount of
enlargement. She could spell
words but not read them.**



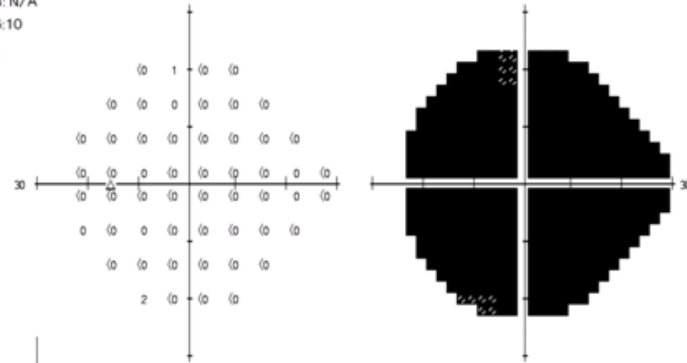
Fixation Target: Central
Fixation Losses: 2/13
False POS Errors: 0 %
False NEG Errors: N/A
Test Duration: 06:10

Background: 31.5 ASB
Strategy: SITA-Standard

Visual Acuity:
RX: DS DC X

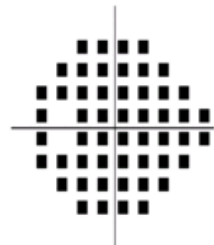
Time: 2:56 PM
Age: 70

Fovea: <0 dB



-27 -25 -26 -28
-29 -30 -28 -31 -30
-30 -31 -32 -32 -33 -32 -31 -30
-31 -31 -33 -34 -33 -32 -29 -28
-31 -33 -34 -34 -34 -33 -29 -28
-29 -32 -31 -33 -34 -33 -32 -30
-31 -32 -32 -33 -32 -31
-27 -31 -30

Total Deviation



∴ < 5%
∴ < 2%
∴ < 1%
■ < 0.5%

Pattern Deviation not
shown for severely
depressed fields. Refer
to Total Deviation.

Pattern Deviation

Pattern Deviation not
shown for severely
depressed fields. Refer
to Total Deviation.

GHT
Outside Normal Limits

VF1 0%

MD -31.49 dB P < 0.5%

PSD 2.03 dB P < 5%

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1200 West Godfrey Avenue
Philadelphia, PA 19141
Unit 4

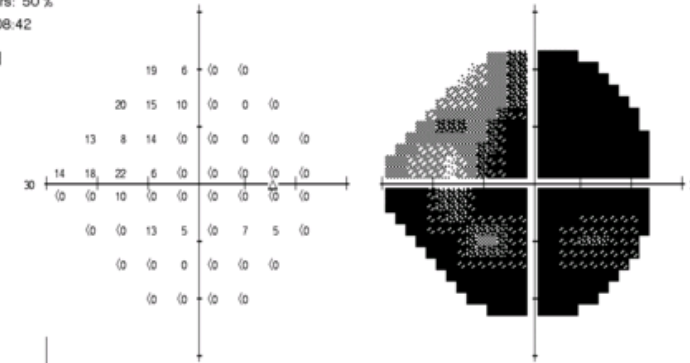
Fixation Target: Central
Fixation Losses: 5/15 xx
False POS Errors: 13 %
False NEG Errors: 50 %
Test Duration: 08:42

Background: 31.5 ASB
Strategy: SITA-Standard

Visual Acuity:
RX: DS DC X

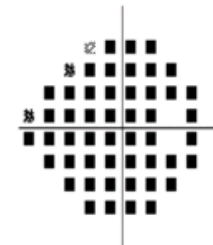
Time: 2:36 PM
Age: 70

Fovea: 5 dB



-7 -20 -28 -27
-8 -14 -19 -30 -26 -29
-15 -22 -16 -33 -32 -30 -31 -30
-12 -11 -8 -25 -34 -33 -33 -31
-28 -31 -21 -34 -34 -34 -33 -31
-30 -32 -18 -26 -33 -24 -26 -31
-31 -32 -31 -32 -32 -31
-30 -31 -31 -31

Total Deviation



∴ < 5%
∴ < 2%
∴ < 1%
■ < 0.5%

Pattern Deviation not
shown for severely
depressed fields. Refer
to Total Deviation.

Pattern Deviation

Pattern Deviation not
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*** Low Test Reliability ***

GHT
Outside Normal Limits

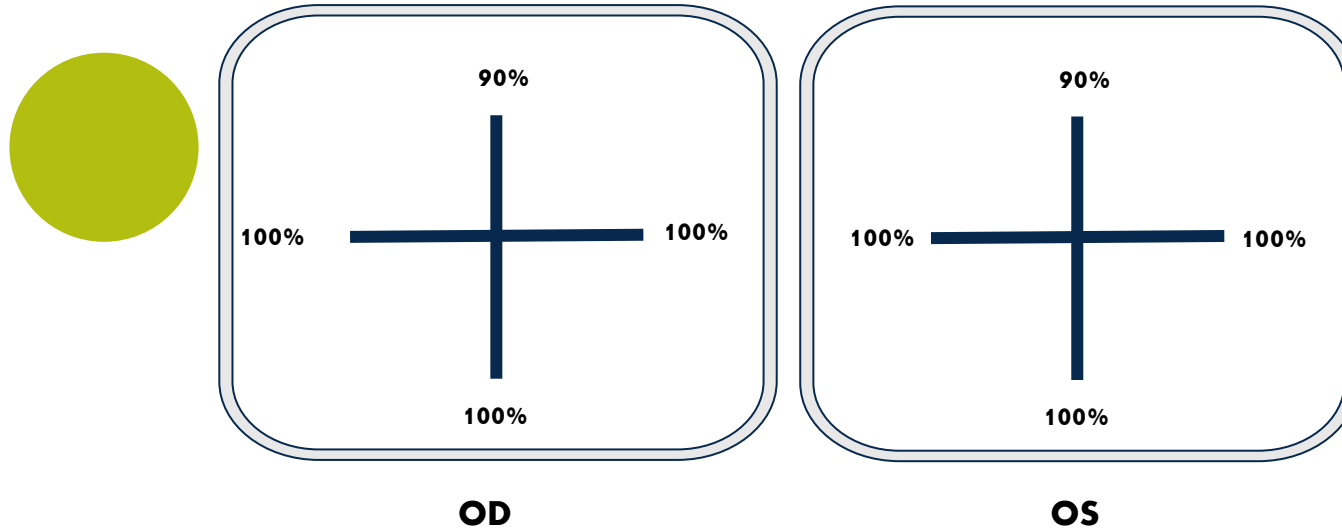
VF1 10%

MD -27.23 dB P < 0.5%

PSD 8.08 dB P < 0.5%

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1200 West Godfrey Avenue
Philadelphia, PA 19141
Unit 4

OTHER EXAM FINDINGS

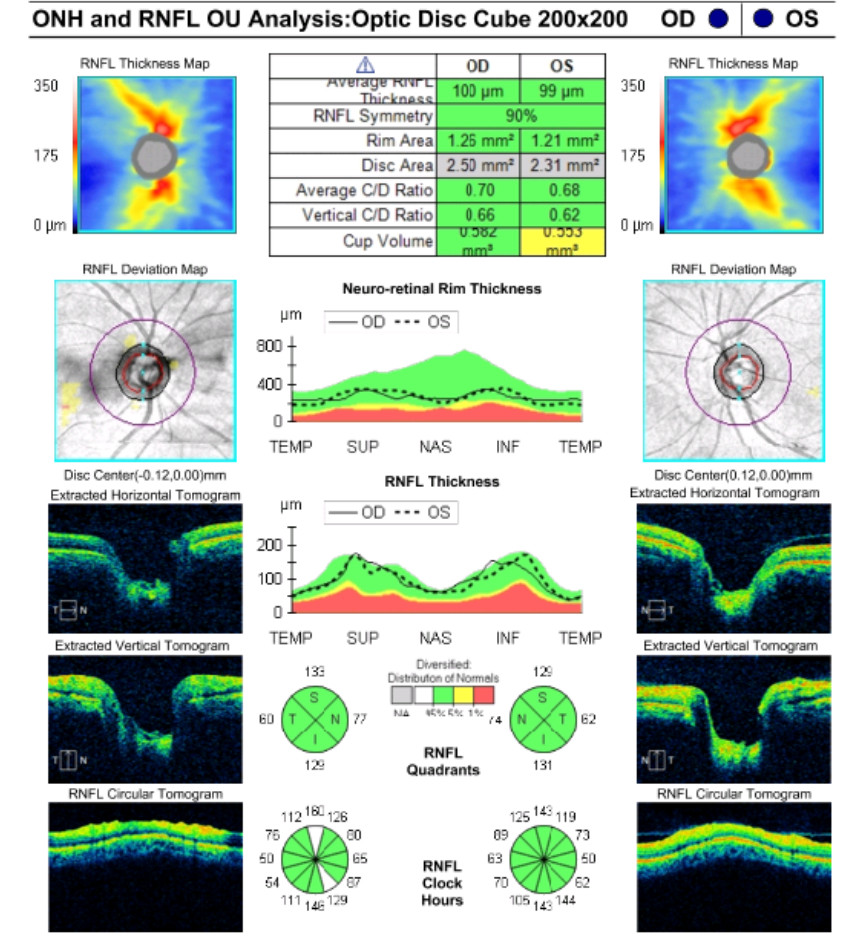


IOPs: OD 13 OS 13 mmHg

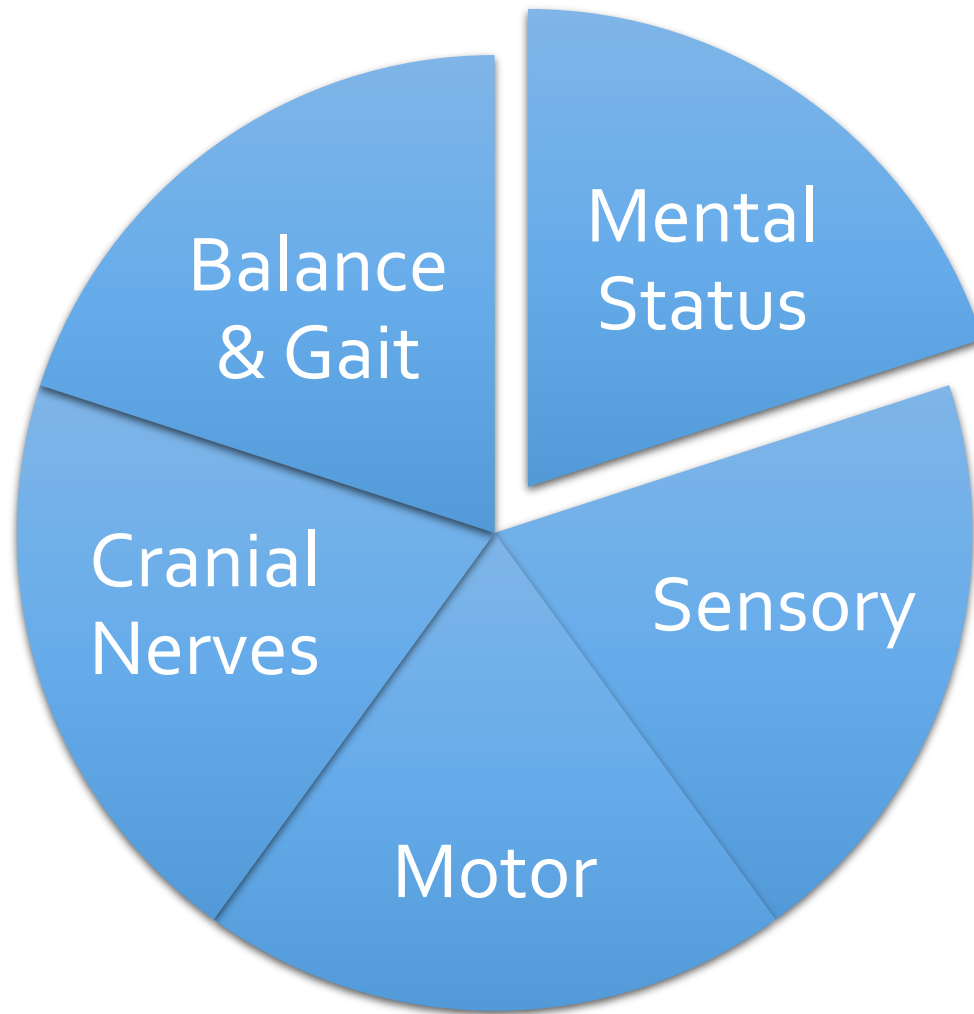
Fundus: OD: 0.65/0.65 no pallor

OS: 0.7/0.7 no pallor

**No anterior or posterior
segment pathology noted.**



NEUROLOGICAL EXAM



Neurological Exam Results

Generally within normal
Unsteady Gait

**Difficultly following instructions



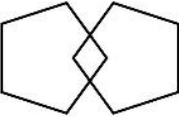
Mini-mental status exam showed
Severe Cognitive Impairment.

MINI MENTAL STATE
EXAMINATION
(MMSE)

Name:

DOB:

Hospital Number:

One point for each answer		DATE:		
ORIENTATION				
Year	Season	Month	Date	Time
Country	Town	District	Hospital	Ward/Floor
REGISTRATION				
Examiner names three objects (e.g. apple, table, penny) and asks the patient to repeat (1 point for each correct. THEN the patient learns the 3 names repeating until correct).				
ATTENTION AND CALCULATION				
Subtract 7 from 100, then repeat from result. Continue five times: 100, 93, 86, 79, 65. (Alternative: spell "WORLD" backwards: DLROW).				
RECALL				
Ask for the names of the three objects learned earlier.				
LANGUAGE				
Name two objects (e.g. pen, watch).				
Repeat "No ifs, ands, or buts".				
Give a three-stage command. Score 1 for each stage. (e.g. "Place index finger of right hand on your nose and then on your left ear").				
Ask the patient to read and obey a written command on a piece of paper. The written instruction is: "Close your eyes".				
Ask the patient to write a sentence. Score 1 if it is sensible and has a subject and a verb.				
COPYING: Ask the patient to copy a pair of intersecting pentagons				
				
TOTAL:				

MMSE scoring
24-30: no cognitive impairment
18-23: mild cognitive impairment
0-17: severe cognitive impairment

MONTREAL COGNITIVE ASSESSMENT (MOCA)				NAME : _____		Date of birth : _____																									
				Education : _____		Sex : _____																									
						DATE : _____																									
VISUOSPATIAL / EXECUTIVE End Begin Copy cube Draw CLOCK (Ten past eleven) (3 points)				POINTS		/5																									
NAMING 				POINTS		/3																									
MEMORY Read list of words, subject must repeat them. Do 2 trials. Do a recall after 5 minutes.				<table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th></th> <th>FACE</th> <th>VELVET</th> <th>CHURCH</th> <th>DAISY</th> <th>RED</th> </tr> </thead> <tbody> <tr> <td>1st trial</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td>2nd trial</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> </tbody> </table>			FACE	VELVET	CHURCH	DAISY	RED	1st trial						2nd trial						No points							
	FACE	VELVET	CHURCH	DAISY	RED																										
1st trial																															
2nd trial																															
ATTENTION Read list of digits (1 digit/ sec.). Subject has to repeat them in the forward order [] 2 1 8 5 4 Subject has to repeat them in the backward order [] 7 4 2				POINTS		/2																									
Read list of letters. The subject must tap with his hand at each letter A. No points if 2 or more errors [] F B A C M N A A J K L B A F A K D E A A A J A M O F A A B				POINTS		/1																									
Serial 7 subtraction starting at 100 [] 93 [] 86 [] 79 [] 72 [] 65 4 or 5 correct subtractions: 3 pts, 2 or 3 correct: 2 pts, 1 correct: 1 pt, 0 correct: 0 pt				POINTS		/3																									
LANGUAGE Repeat: [] I only know that John is the one to help today. [] The cat always hid under the couch when dogs were in the room. []				POINTS		/2																									
Fluency / Name maximum number of words in one minute that begin with the letters F [] _____ (N 2 or 11 words)				POINTS		/1																									
ABSTRACTION Similarity between e.g. banana - orange = fruit [] train - bicycle [] watch - ruler []				POINTS		/2																									
DELAYED RECALL <table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th></th> <th>FACE</th> <th>VELVET</th> <th>CHURCH</th> <th>DAISY</th> <th>RED</th> </tr> </thead> <tbody> <tr> <td>Has to recall words WITH NO CUE</td> <td>[]</td> <td>[]</td> <td>[]</td> <td>[]</td> <td>[]</td> </tr> <tr> <td>Category cue</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> <tr> <td>Optional Multiple choice cue</td> <td></td> <td></td> <td></td> <td></td> <td></td> </tr> </tbody> </table>					FACE	VELVET	CHURCH	DAISY	RED	Has to recall words WITH NO CUE	[]	[]	[]	[]	[]	Category cue						Optional Multiple choice cue						POINTS		/5	
	FACE	VELVET	CHURCH	DAISY	RED																										
Has to recall words WITH NO CUE	[]	[]	[]	[]	[]																										
Category cue																															
Optional Multiple choice cue																															
ORIENTATION [] Date [] Month [] Year [] Day [] Place [] City				POINTS		/6																									
© Z. Nasreddine MD Version November 7, 2004 Normal ≥ 26 / 30				TOTAL		/30																									
www.mocatest.org				Add 1 point if ≤ 12 yr edu																											



Mini-Cog®

Instructions for Administration & Scoring

ID: _____ Date: _____

Step 1: Three Word Registration

Look directly at person and say, "Please listen carefully. I am going to say three words that I want you to repeat back to me now and try to remember. The words are [select a list of words from the versions below]. Please say them for me now." If the person is unable to repeat the words after three attempts, move on to Step 2 (clock drawing).

The following and other word lists have been used in one or more clinical studies.^{1,3} For repeated administrations, use of an alternative word list is recommended.

Version 1	Version 2	Version 3	Version 4	Version 5	Version 6
Banana	Leader	Village	River	Captain	Daughter
Sunrise	Season	Kitchen	Nation	Garden	Heaven
Chair	Table	Baby	Finger	Picture	Mountain

Step 2: Clock Drawing

Say: "Next, I want you to draw a clock for me. First, put in all of the numbers where they go." When that is completed, say: "Now, set the hands to 10 past 11."

Use preprinted circle (see next page) for this exercise. Repeat instructions as needed as this is not a memory test. Move to Step 3 if the clock is not complete within three minutes.

Step 3: Three Word Recall

Ask the person to recall the three words you stated in Step 1. Say: "What were the three words I asked you to remember?" Record the word list version number and the person's answers below.

Word List Version: _____ Person's Answers: _____

Scoring

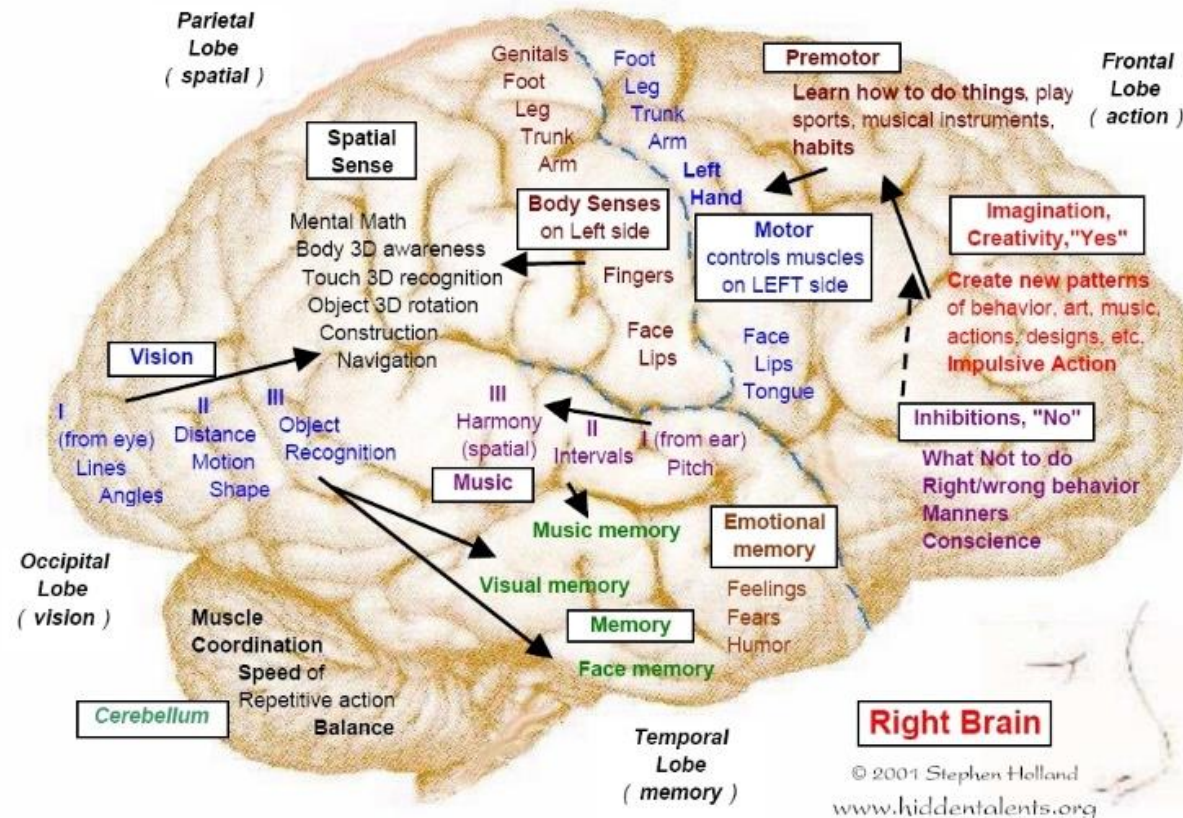
Word Recall: _____ (0-3 points)	1 point for each word spontaneously recalled without cueing.
Clock Draw: _____ (0 or 2 points)	Normal clock = 2 points. A normal clock has all numbers placed in the correct sequence and approximately correct position (e.g., 12, 3, 6 and 9 are in anchor positions) with no missing or duplicate numbers. Hands are pointing to the 11 and 2 (11:10). Hand length is not scored. Inability or refusal to draw a clock (abnormal) = 0 points. Total score = Word Recall score + Clock Draw score.
Total Score: _____ (0-5 points)	A cut point of <3 on the Mini-Cog™ has been validated for dementia screening, but many individuals with clinically meaningful cognitive impairment will score higher. When greater sensitivity is desired, a cut point of <4 is recommended as it may indicate a need for further evaluation of cognitive status.

Mini-Cog © S. Borson. All rights reserved. Reprinted with permission of the author solely for clinical and educational purposes. May not be modified or used for commercial, marketing, or research purposes without permission of the author (soob@uw.edu). v. 01.19.16



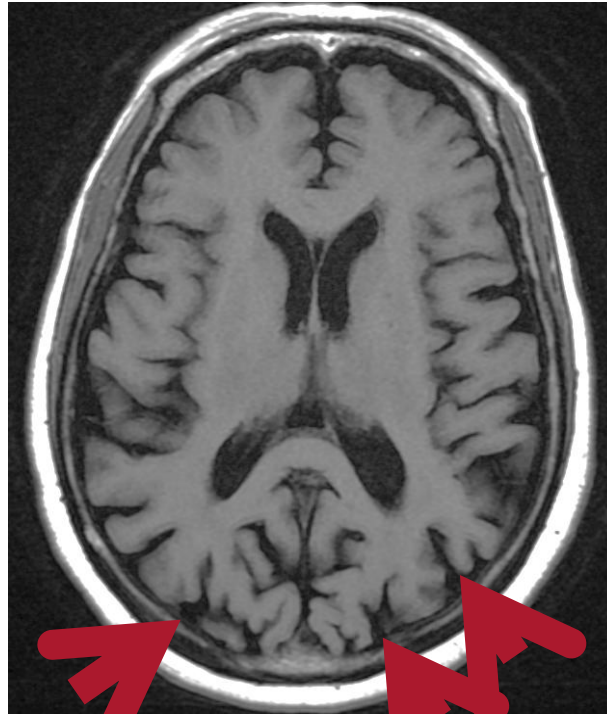
DIAGNOSIS

Vision loss likely secondary impaired visual information processing

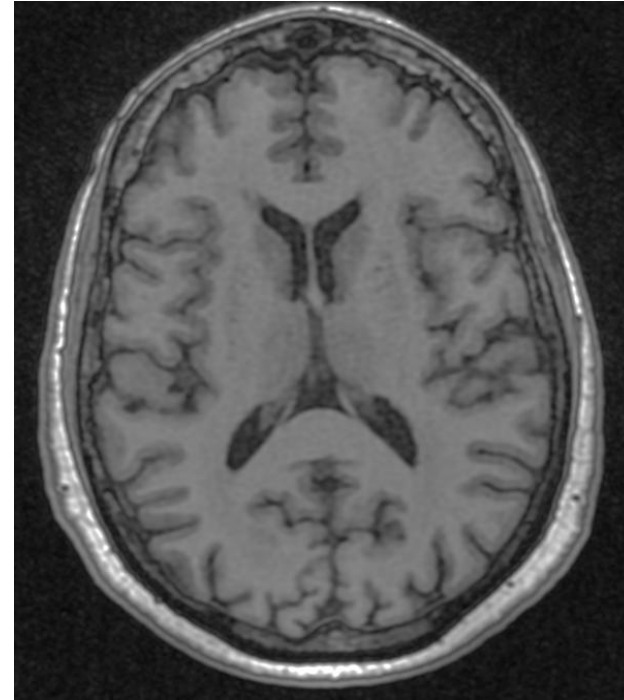


DIAGNOSIS

Patient



Age-Matched Normal



ARROWS indicate the areas of increased cortical atrophy in patient compared to the age-matched normal patient on the right.

Findings and history consistent with
Posterior Cortical Atrophy (PCA)

Features of PCA

First complaint is DIFFICULTY READING

Gradual onset and progression

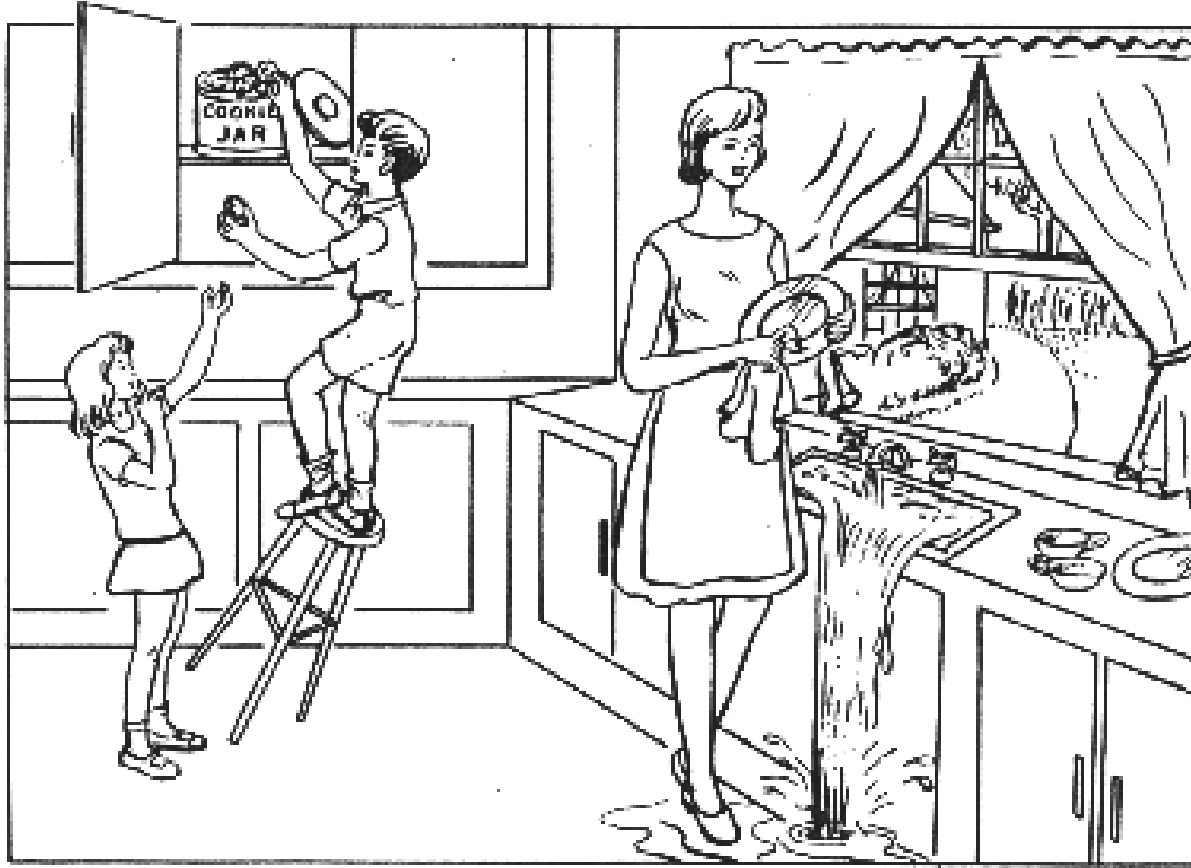
Relatively preserved episodic memory, verbal fluency and personal insight

Difficulty processing complex images

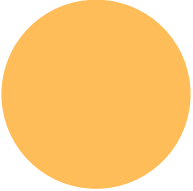
PCA

- Decreased Visuospatial
- Decreased Visuo-perceptual

Cookie Thief – another test to consider



Tests aphasia but also issues with
SIMULTANAGNOSIA - the inability to perceive multiple visual objects at the same time.



PCA Visual Symptoms



Simultanagnosia

Homonymous Hemianopia

Ocular Motor Apraxia – inability to fixate

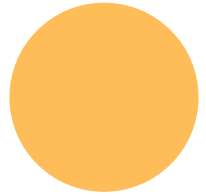
Visual Agnosia – Inability to recognize objects

Prosopagnosia – Inability to recognize faces

Visual – spatial deficits – not accurately reaching for a doorknob

INABILITY TO READ





VISION REHABILITATION



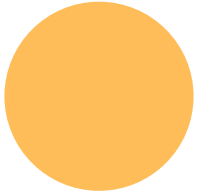
**Vision rehab difficult due to
progressive cognitive
impairment**

**Educate family to understand
and what to expect**

Focus on Text-to-speech devices

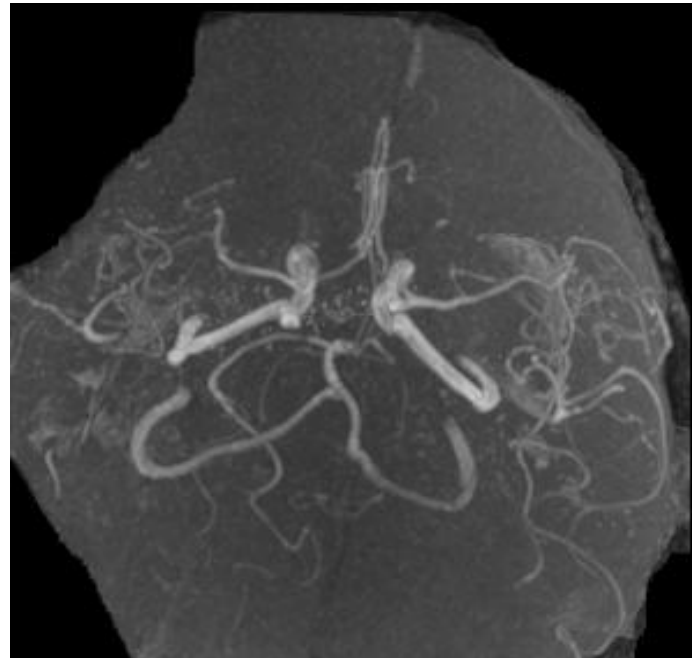


CASE



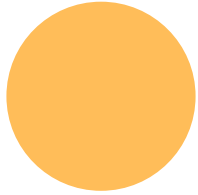
Medical Diagnosis Given

but presentation not as expected



Case History

46-year-old man



Chief Complaint

Cannot read since stroke.

Systemic History

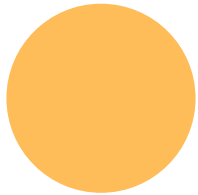
**HTN, Stroke affecting left side of brain 4 months prior
Initially - weakness in right upper extremity, inability to talk, and vision loss
Has since had significant improvement.**

Ocular History

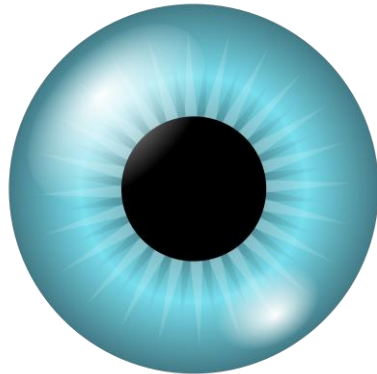
Right homonymous hemianopia since stroke



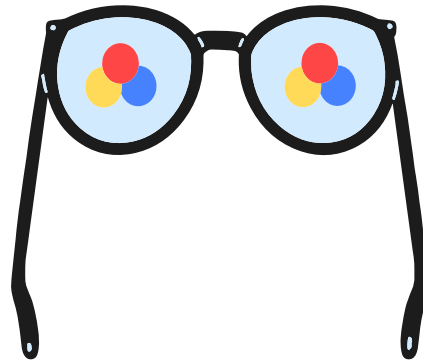
Afferent Visual System



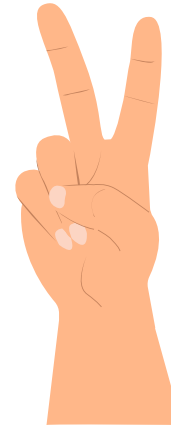
**VISUAL
ACUITY**



PUPILS



COLOR VISION



**CONFRONTATIONAL
FIELDS**



Tumbling E

OD: 20/25

OS: 20/25

**OD: round
Bright: 2.0mm
Dim: 2.5mm**

**OS: round
Bright: 2.5mm
Dim: 3.5mm**

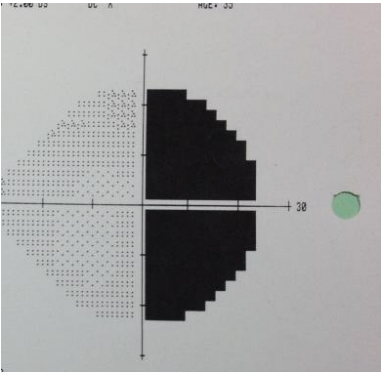
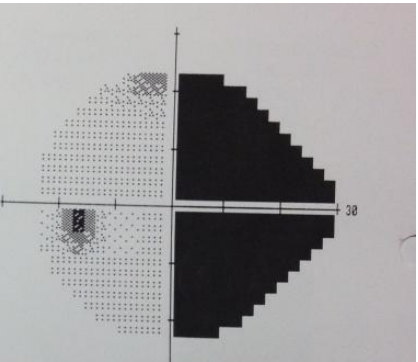
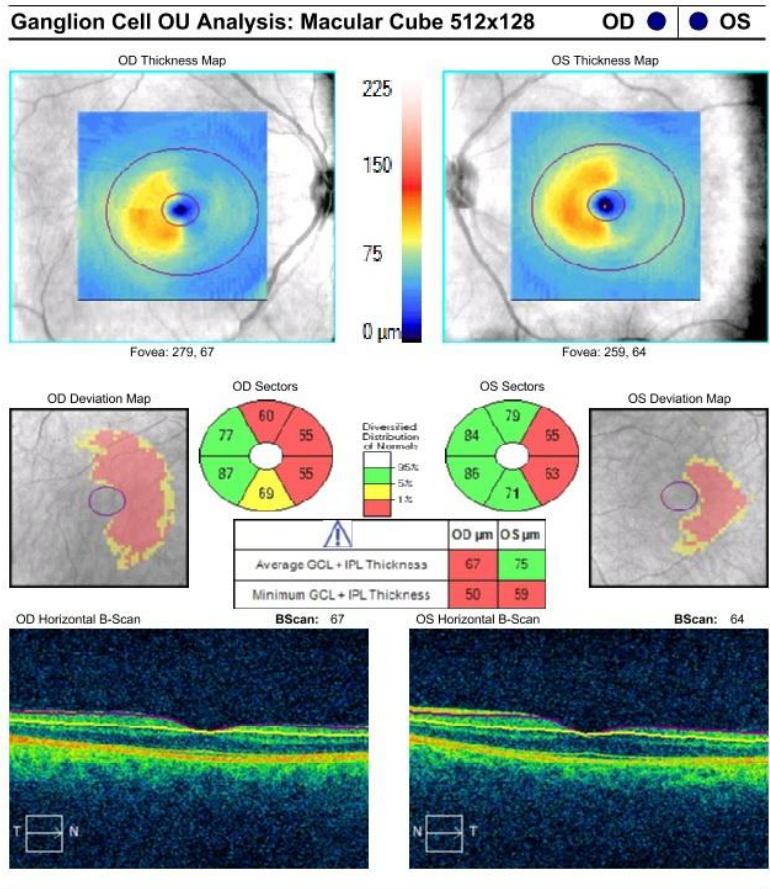
– APD

Too Frustrating

**Complete Right
Homonymous
Hemianopia**



Additional Testing



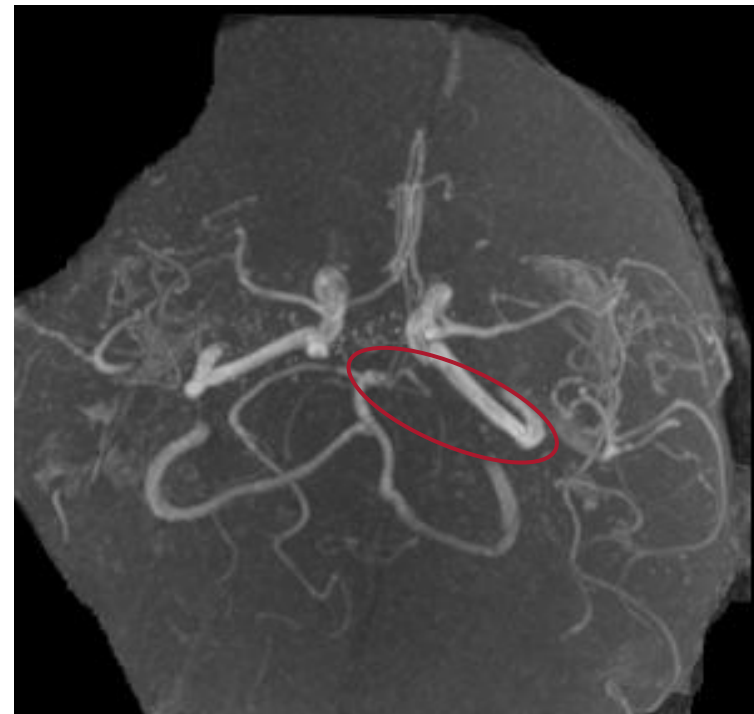
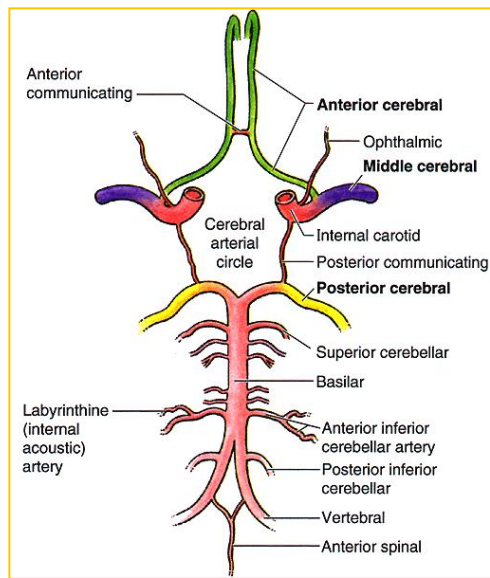
**REMAINDER OF
OPHTHALMIC EXAM
NORMAL**

Why can't he read?

PREVIOUS IMAGING REPORT



- **Left Posterior Cerebral Artery infarct**
 - **Occipital lobe**
 - **Medial posterior temporal parietal lobes**
 - **Splenium of corpus callosum**
 - **Left Thalamus**



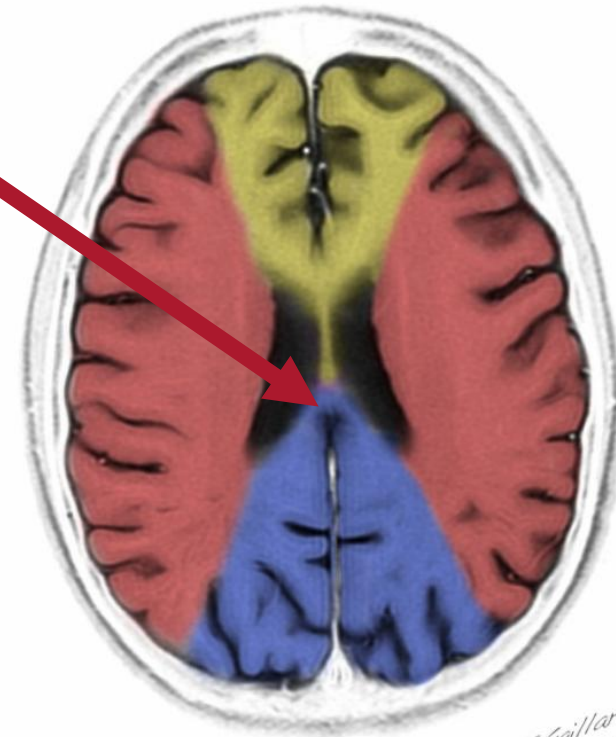
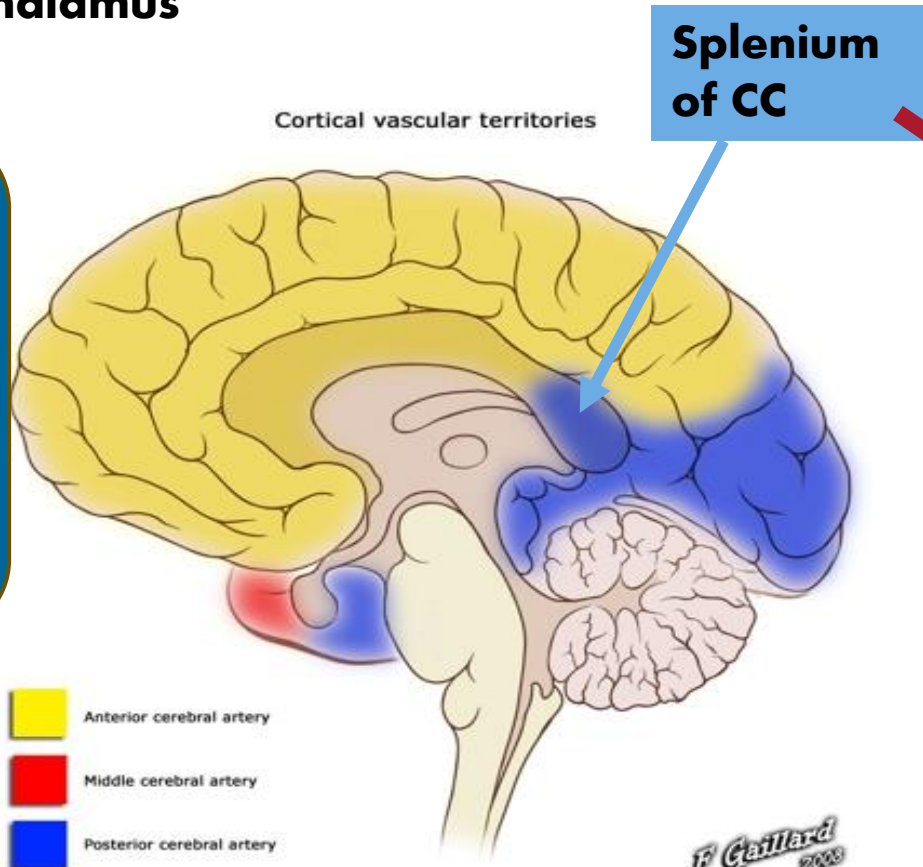
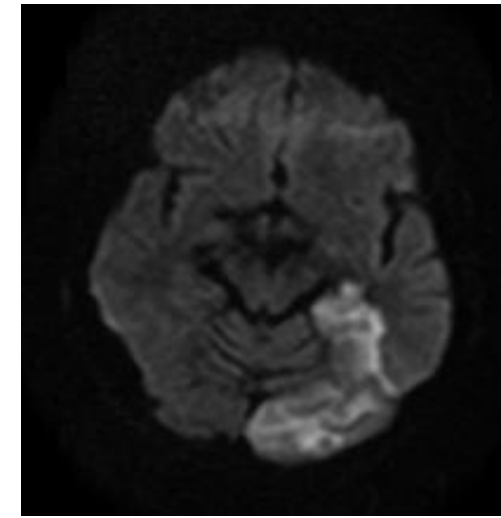
MRI Report

Left Posterior Cerebral Artery infarct

- Occipital lobe
- Medial posterior temporal parietal lobes
- **Splenium of corpus collosum**
- Left Thalamus

Splenium of CC

Allows
interhemispheric
communication
for visual
processing and
reading



F Gaillard
2003

©Radiopaedia.org

Line drawing of brain by Patrick Lynch (patricklynch.net)

F Gaillard
2010

Radiopaedia.org CC-BY-SA-BY

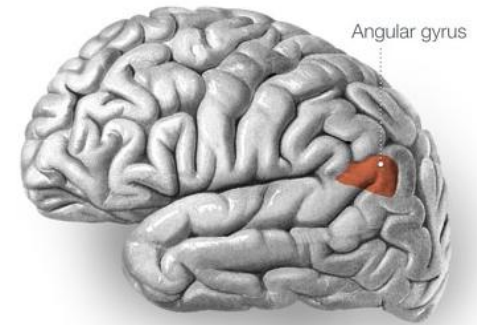
LEFT POSTERIOR INFARCT & LANGUAGE

- ~ 90% of population right-handed.
 - 99% left hemisphere dominant for language.
- Language left-handed people left hemisphere dominant
- Language is primarily localized to LEFT hemisphere

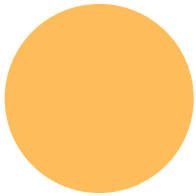


DIAGNOSIS – ALEXIA WITHOUT AGRAPHIA

- **Lesion = Left occipitotemporal region**
 - Infarct typically of left posterior cerebral artery
 - Lesion often splenium of the corpus callosum & posterior thalamus
- **Complete right homonymous hemianopia**
- **Indirect involvement of angular gyrus in dominant hemisphere**
 - Disrupts connection between intact right visual cortex and left angular gyrus (center for reading)
- **CAN NAME THINGS BUT CANNOT READ**



VISION REHABILITATION



**Text-to-speech – AI glasses
(primarily adults)**

**Immediate therapies to help develop
connections.**

Multiple Oral Re-reading techniques

**Multiple sensory input
i.e., tracing letters on patient's arm**

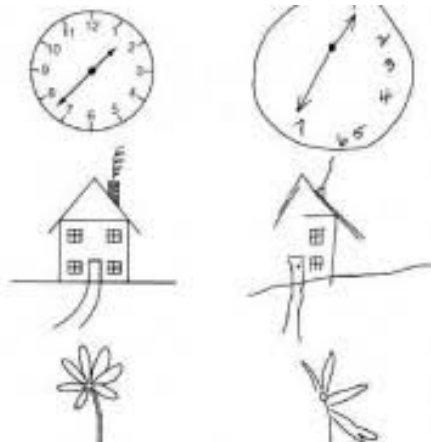
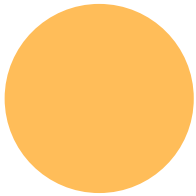
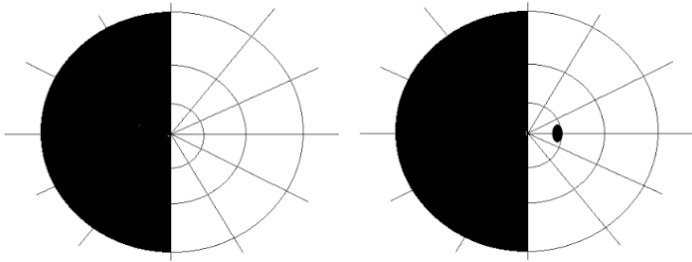


VISUAL NEGLECT / SPATIAL NEGLECT

LEFT HOMONYMOUS HEMIANOPIA

Typically Right-sided Brain Injury

Injury likely to right posterior parietal cortex including the inferior parietal lobule and the temporoparietal junction.

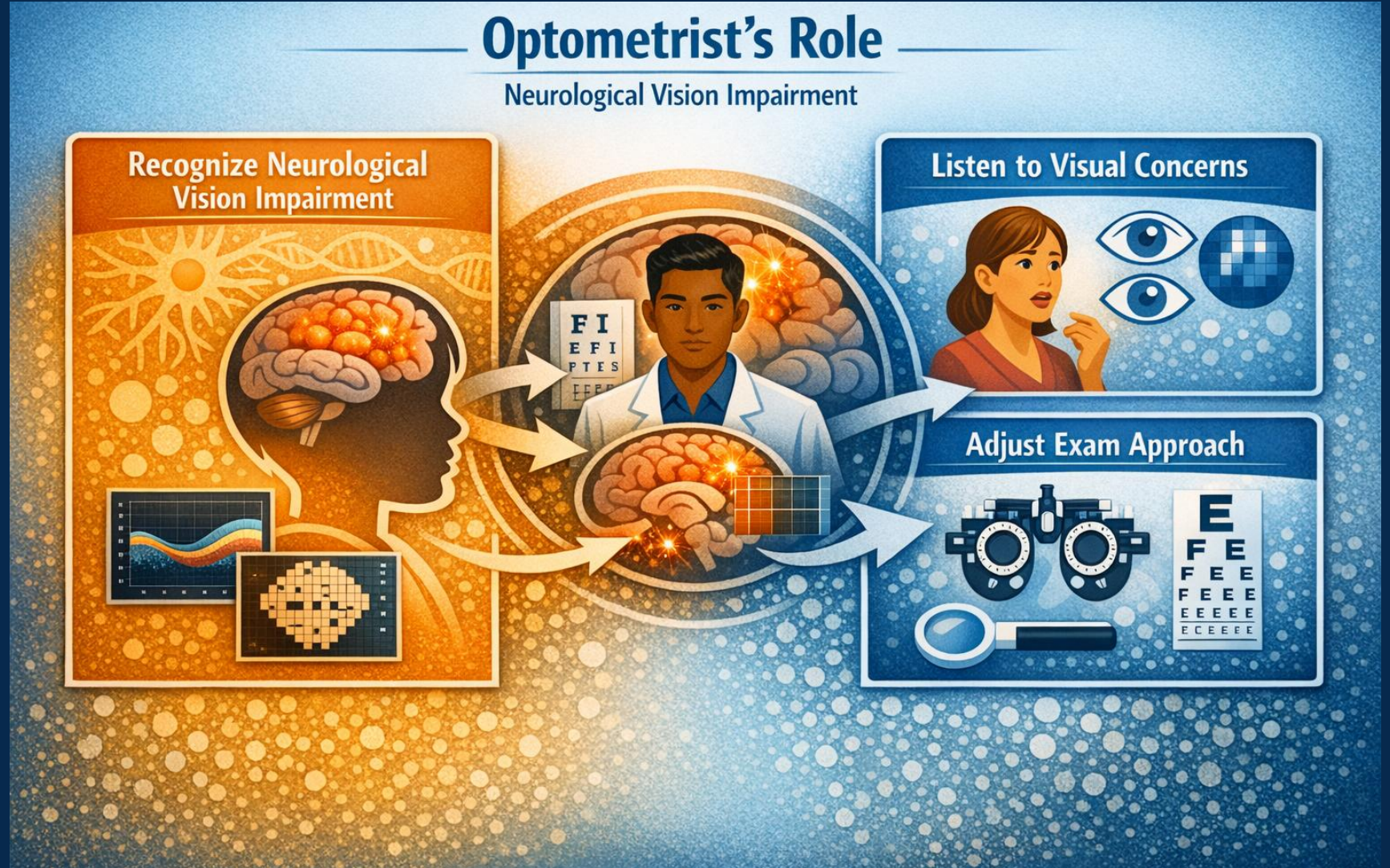


Brain Damage/Injury $\neq$ Visual Perception

Depending on where the damage is will dictate the clinical presentation/visual issues

- Keep foveas pointed at objects of interest  **Cerebellar/vestibular damage = nystagmus/oscillopsia**
- Coordinate eye movements  **Brainstem/Cerebral = diplopia, gaze palsies**
- Locating, fixating, pursuits, saccades, vergences  **Cerebellar/vestibular damage = nystagmus/oscillopsia**
Vergence issues
- Visual Recognition  **Temporal lobe = alexia, agnosias**
- Perception of motion and space  **Parietal lobe = optic ataxia, Visual-spatial apraxia**
- Coordinate body movements with vision  **Cerebral, cerebellar damage**

Optometry's Role



#1 Job

Give the Brain as clear of a message as possible



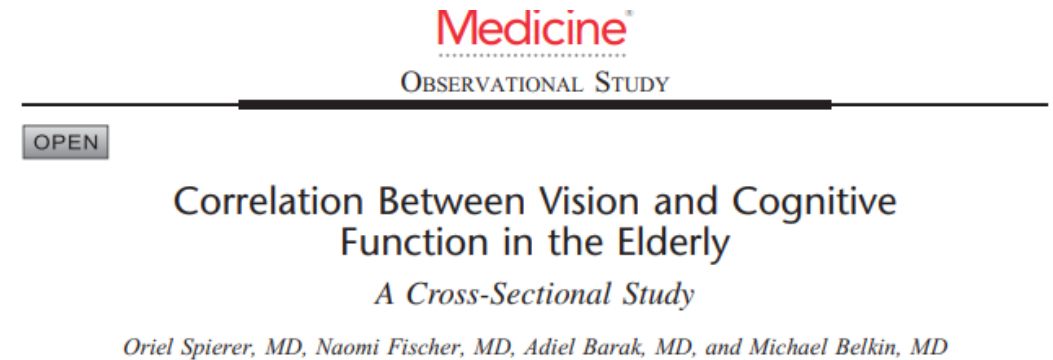
Perform a detailed refraction.

- **May need to perform outside of the phoropter**
- **Trial frame the prescription**
- **May not be verbal; use tumbling E, matching,**

#1 Job

Give the Brain as clear of a message as possible

- **Treat Any Ocular Disease**
 - **Aggressive Dry Eye Treatment**
 - **Don't Delay Cataract Surgery**
- **Prescribe Prism**
 - **Correct even small deviations**
 - **Check for convergence insufficiencies**



The association between cataract and incidence of cognitive impairment in older adults: A systematic review and meta-analysis

Zhenrong Xiong ¹, Xinming Li ², Dejiang Yang ², Chongyu Xiong ¹, Qinggang Xu ³, Qi Zhou ⁴

Affiliations + expand

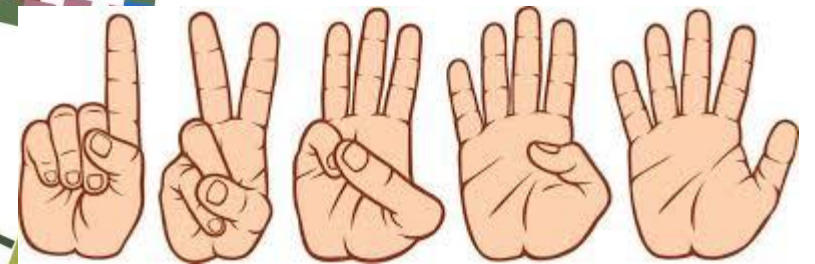
PMID: 37148915 DOI: [10.1016/j.jbbr.2023.114455](https://doi.org/10.1016/j.jbbr.2023.114455)

Perform Visual Fields

**May need to get
creative**



**Hold up 2 colored targets in right and left
periphery. Ask which color do they see.**

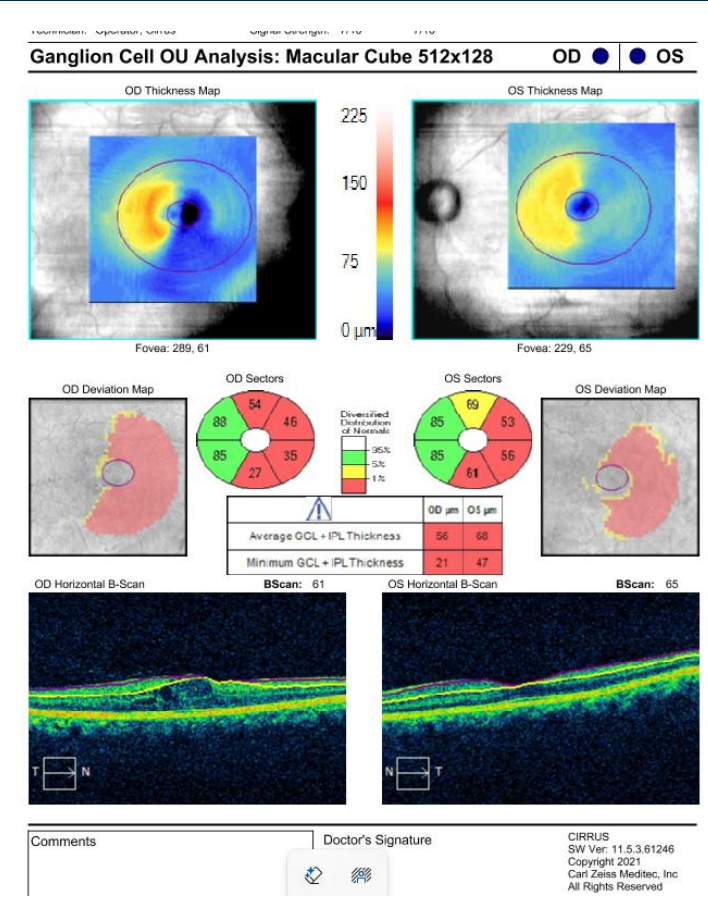
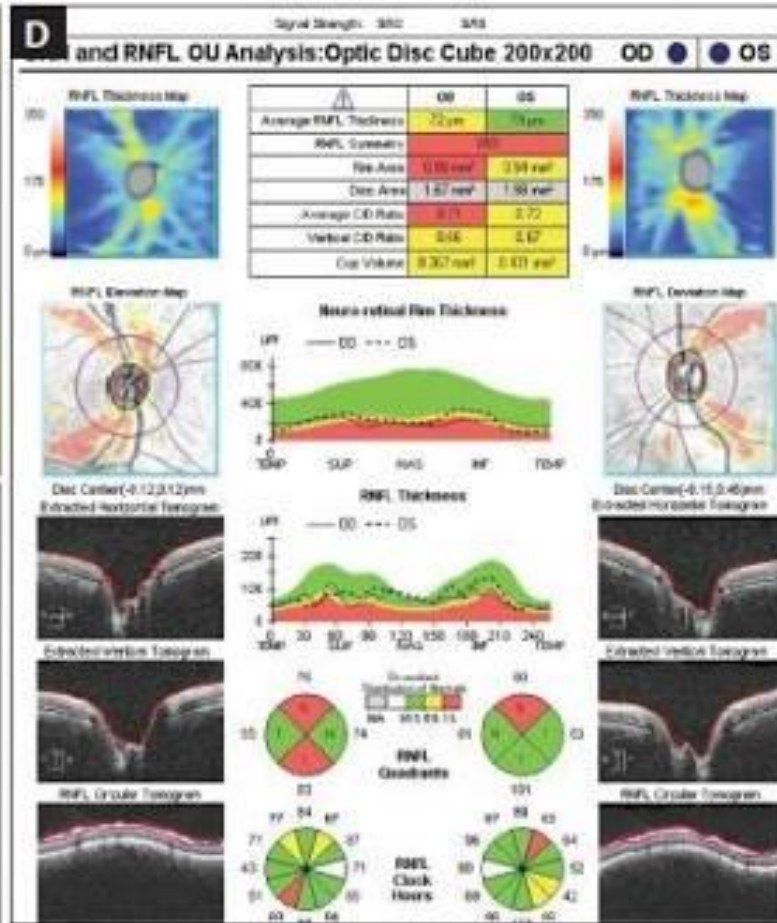
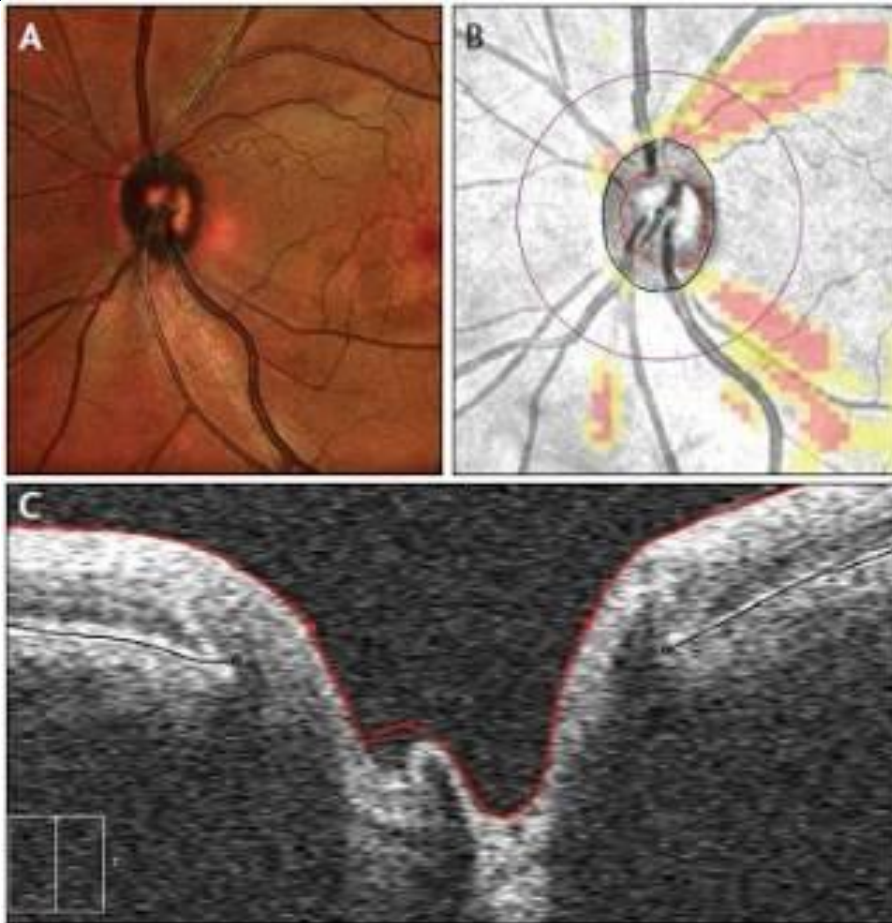


**Nonverbal patients can
mimic numbers or match.**

Helpful OD Tools:

OCT – Optical Coherence Tomography

Non-invasive measurement of neurons



Helpful OD Tools: OCT

CASE

53-year-old woman

- History of Sturge-Weber Syndrome
- Verbal but cognitively impaired
- History of seizures
- S/P corpus callosotomy

Long-term patient at clinic.

Treated for glaucoma

Not able to take a formalized visual field

STURGE-WEBER

QUICK FACTS

WHAT IS IT?

- A rare, congenital disorder that affects the skin, brain, and eyes
- Caused by a mutation in the *GNAQ* gene



PORT-WINE BIRTHMARK

A facial birthmark caused by dilated blood vessels



COMPLICATIONS

- Seizures
- Glaucoma
- Developmental delays



COMMON MYTHS

- A port-wine birthmark is "just a stain"

OCT in detecting neurological field deficits

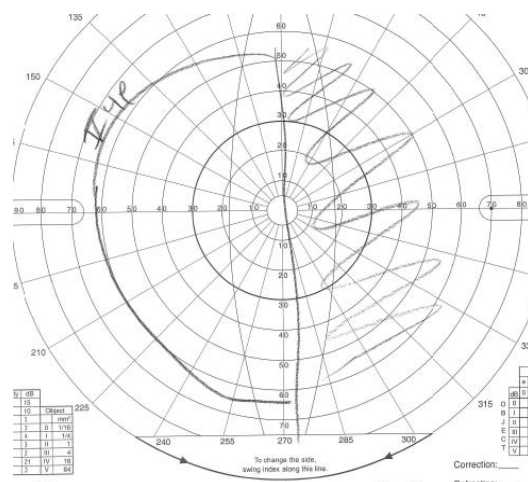
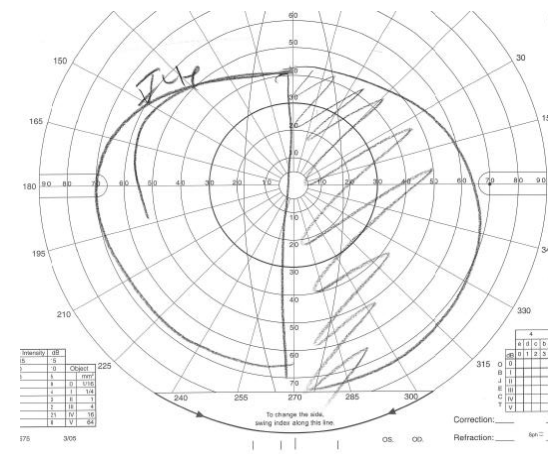
CASE

53-year-old woman

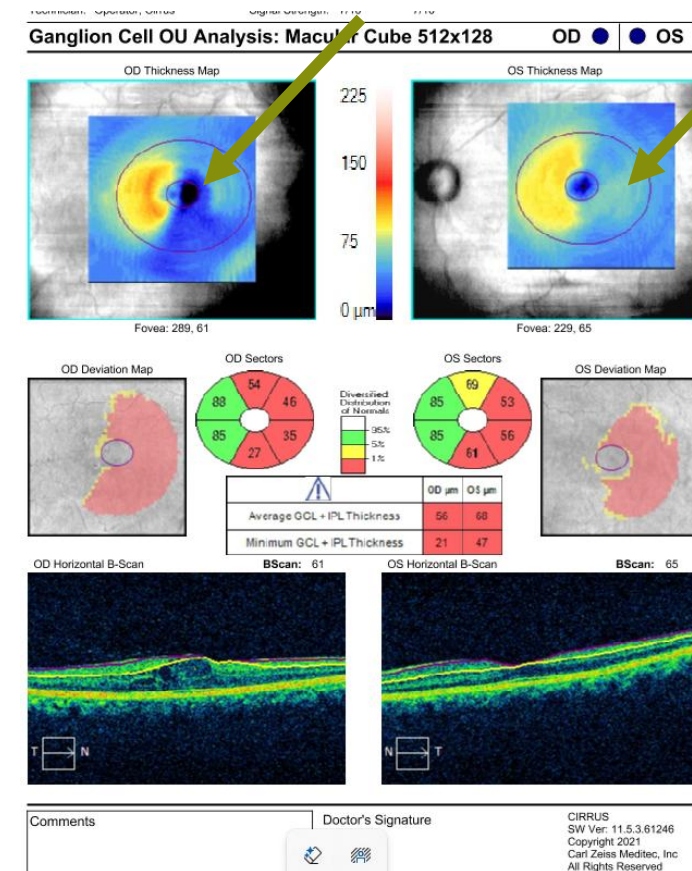
History of Sturge-Weber Syndrome

Verbal but cognitively impaired

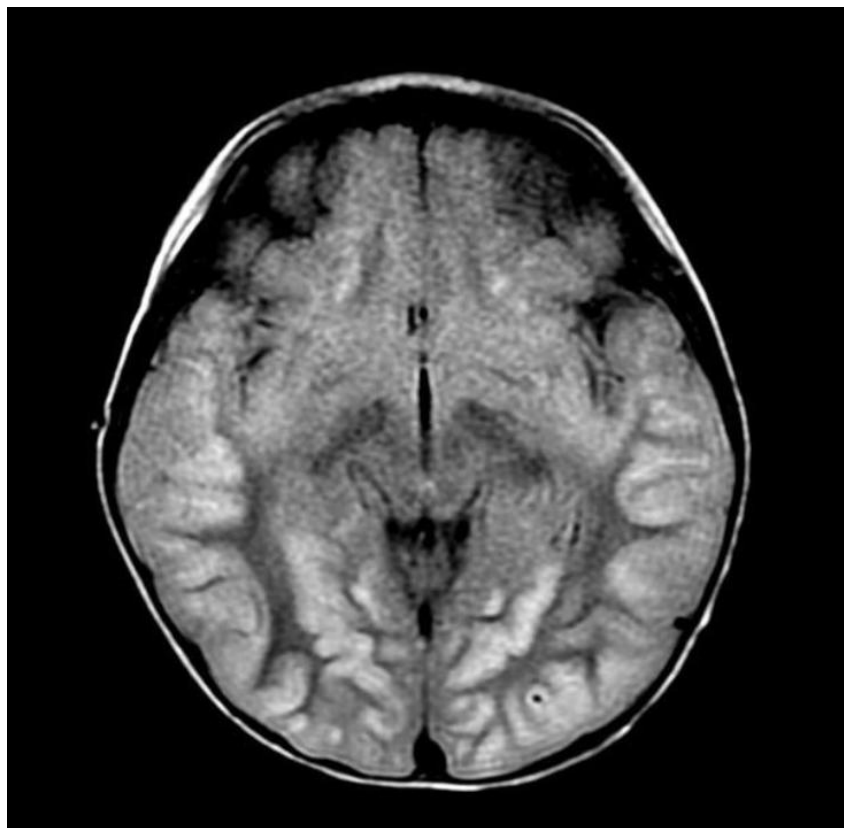
Confrontational field testing with 2 different colored targets suggesting of Right Homonymous Hemianopia



Right Homonymous Hemianopia

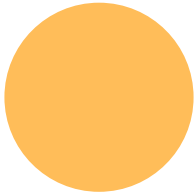


CASE



CASE HISTORY

36-year-old man



Chief Complaint

Eyes feel like they are not working together.
Blind/splotchy areas in the inferior field of both eyes

Systemic History

Anoxic brain injury 6 years prior due to cardiac arrest from heroin use.
Has defibrillator, depression, anxiety

Ocular History

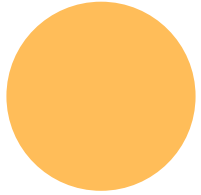
Traumatic cataract OS due to hit with rock at age 3

Social History

Quit smoking, drugs, and alcohol 6 years ago



AFFERENT VISUAL SYSTEM



VISUAL ACUITY

Snellen

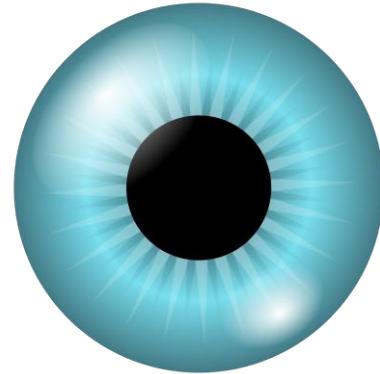
OD: 20/20

OS: 20/20-3

Refraction

OD: plano

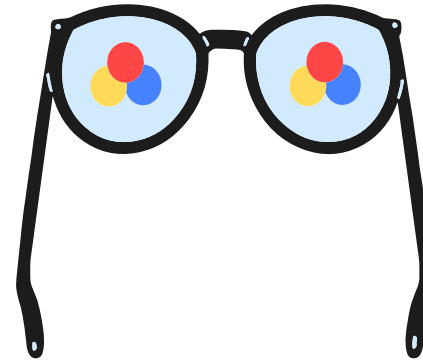
OS: +2.00 sph



PUPILS

OD: round
Bright: 4.0mm
Dim: 6.0mm

OS: round
Bright: 4.0mm
Dim: 6.0mm
– APD

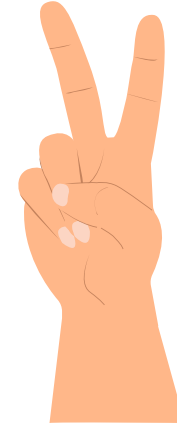


COLOR VISION

OD 14/14

OS 14/14

Ishihara

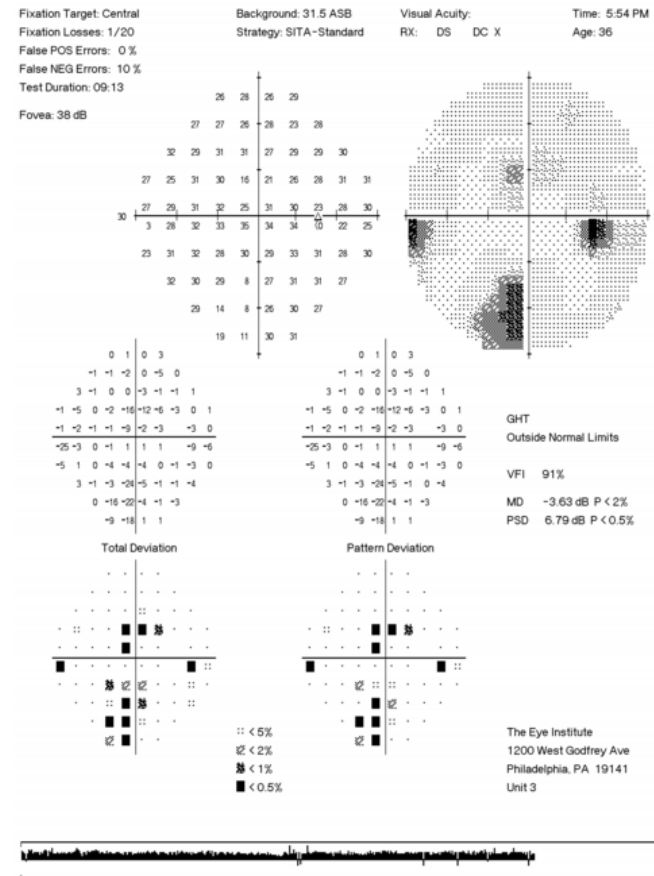
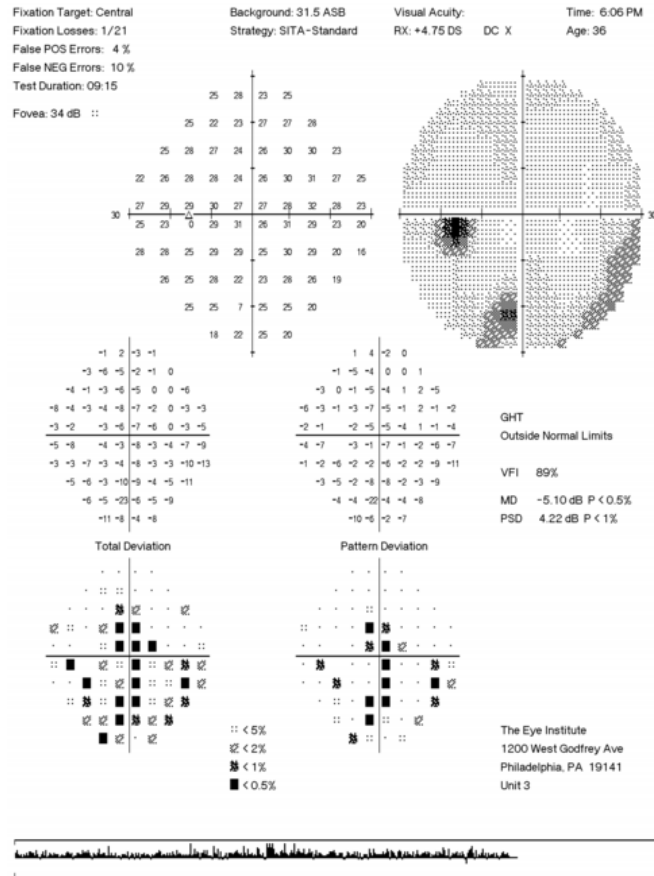


CONFRONTATIONAL FIELDS

OD: mild inf
nasal
constriction

OS: mild inferior
temporal
constriction





Chief Complaint

- Eyes feel like they are not working together.
- Blind/splotch areas in the inferior field of both eyes

Efferent System Testing Normal

Fundus Exam Normal

Chief Complaint

- Eyes feel like they are not working together.
- **Blind/splotch areas in the inferior field of both eyes**

Refraction

OD: plano

OS: +2.00 sph

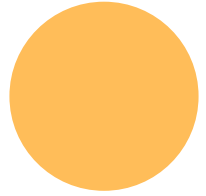


The Brain is receiving 2 different sized images.

This can be corrected best with Contact Lenses

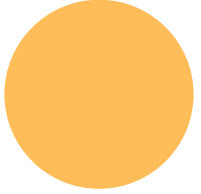


Diagnosis – Anoxic Brain Injury



- **Anoxic brain injury, also known as global hypoxic-ischemic injury**
- **Lack of oxygen to entire brain**
- **If the oxygen supply is interrupted, consciousness will be lost within 15 seconds and damage to the brain begins to occur after about four minutes without oxygen.**

Diagnosis – Anoxic Brain Injury



Factors Influencing Extent of Injury

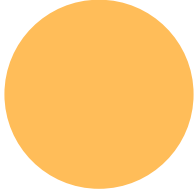
- **Age of patient**
- **Severity of hypoxic insult**
- **Duration of hypoxic-ischemia**

Causes of Anoxic Brain Injury

- **Cardiac or respiratory arrest**
- **Very low blood pressure (shock or hemorrhage)**
- **Suffocation/choking/strangulation**
- **Very severe asthma attack**
- **Complication of general anesthesia**
- **Near drowning**
- **Exposure to high altitudes**
- **Smoke/carbon monoxide inhalation**
- **Poisoning**
- **Drug overdose**
- **Electric shock**



Anoxic/Hypoxic Brain Injury and Vision

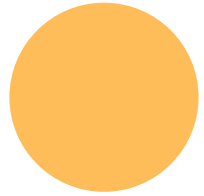


The occipital lobe is particularly susceptible to ischemia

Anton Syndrome can occur:

- **Bilateral occipital lobe infarcts/damage – Cortical blindness**
- **NLP but the patient thinks they can still see**
- **Denial of blindness**
- **Pupil light responses are intact**

**Our patient did not
have this**



VISION REHABILITATION

**Each patient will
be different**

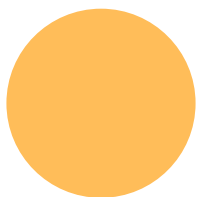


**Correct aniseikonia with
contacts**

Educate on field loss

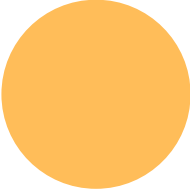
**Recommend continuing with
speech and cognitive
therapies**





CASE





CASE HISTORY

29-year-old woman



Chief Complaint

**Has been noticing little specks of light in her vision for past 3 years.
Started after a night of heavy partying
Now also light sensitive and notice trailing of images**

Systemic History

Anxiety (started after visual changes) and migraines

Ocular History

Dry eyes, near-sighted (wears glasses), does not like driving at night

Medications

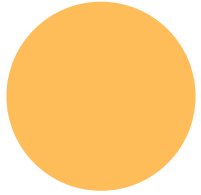
Venlafaxine, B₁, B₁₂, Iron, Magnesium

Social History

Previous alcohol and drug use



AFFERENT VISUAL SYSTEM

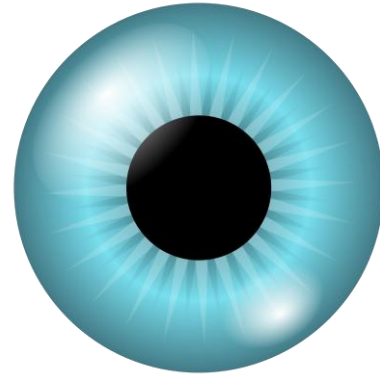


VISUAL ACUITY

Snellen Acuity

OD: 20/20

OS: 20/20

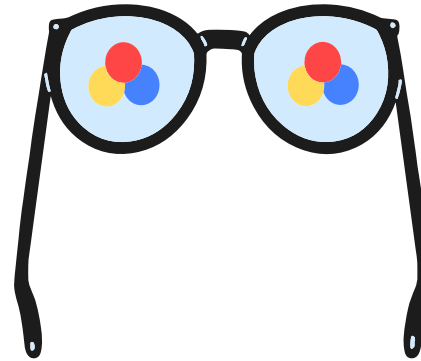


PUPILS

OD: round
Bright: 5.0mm
Dim: 7.0mm

OS: round
Bright: 5.0mm
Dim: 7.0mm

– APD

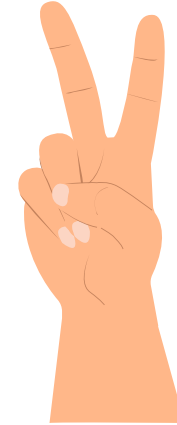


COLOR VISION

OD: 14/14

OS: 14/14

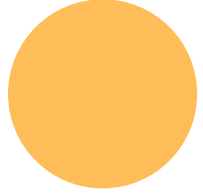
Ishihara



CONFRONTATIONAL FIELDS

FTFC OU



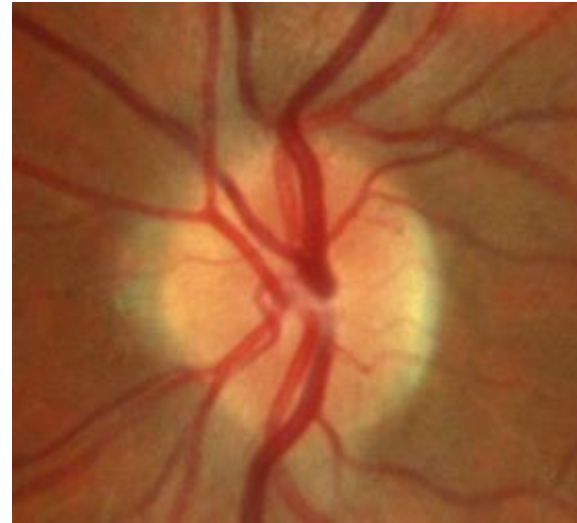
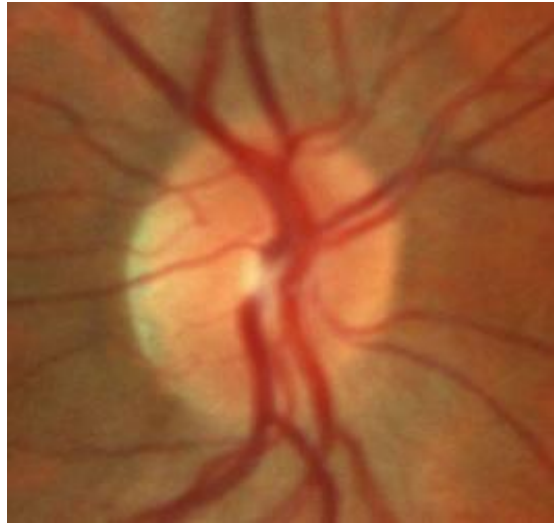


Efferent visual system normal

IOPs 17/19 mmHg

BP 120/80 mmHg

Fundus exam normal





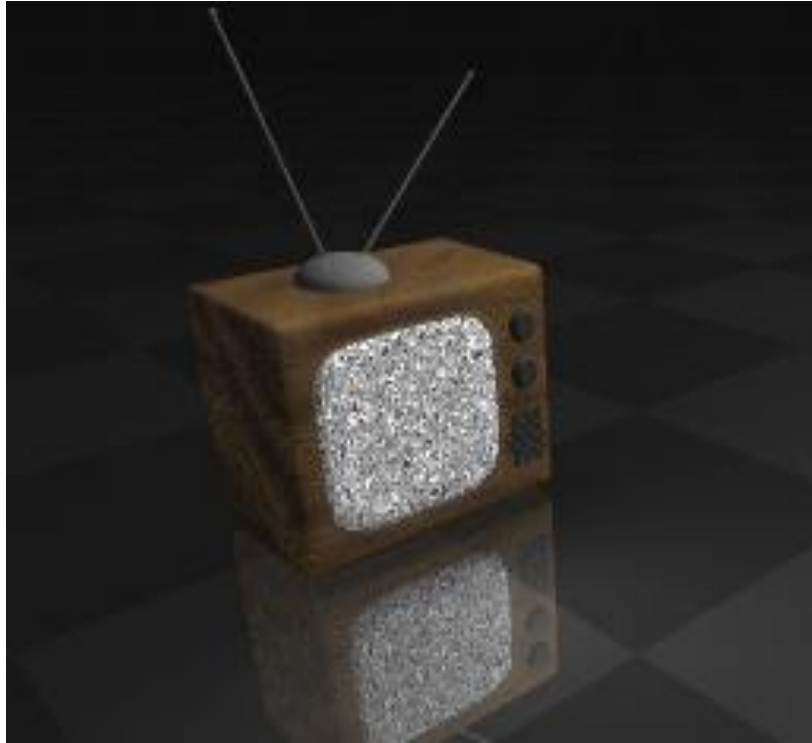
Her exam is normal

**Why the specks in her
vision?**

DIAGNOSIS – VISUAL SNOW SYNDROME

Description of Visual Snow

- flickering and moving dots
- similar to the static of analogue televisions
- persistent and occupy the entire visual field
- can be black and white, transparent, flashing, colored, or a combination of all of these.



Visual snow simulator: <https://visionsimulations.com/visual-snow.htm>

DIAGNOSIS – VISUAL SNOW SYNDROME

Causes of Visual Snow

- Spontaneous
- Commonly with migraine
- Concussion or mild TBI
- Hallucinogenic drugs
- Drugs/alcohol can induce/worsen

Associated Symptoms

- Migraines ✓
- Tinnitus
- Photophobia* ✓
- Palinopsia* – trailing images ✓
- Enhanced entopic phenomena*
 - Lines moving across vision
 - Rain running down a window
 - Excessive floaters
- Nyctalopia* (poor night vision) ✓

✓ = our patient

*need at least

2 of these
additional
symptoms

present for
at least

3 months

for diagnosis
of VSS

Visual snow simulator: <https://visionsimulations.com/visual-snow.htm>

VISION REHABILITATION



Blue light blocking to decrease migraines

FL 41 tints for light sensitivity + migraines

Vitamins, nutraceuticals

Transcranial Magnetic Stimulation?

CVI Often misinterpreted as:

- **Intellectual disability or cognitive impairment**
- **Behavioral problems**
- **Emotional disorders**
- **Learning disabilities**
- **Reading challenges**
- **Dyslexia**
- **ADHD**
- **Laziness, lack of motivation, defiance, stubbornness, disinterest**



Case Study

Puzzle Piece that Doesn't Fit



11 yo boy

Family from Dominican Republic

Referred for evaluation of supraduction deficits

Declining Academic Performance



Ocular History

Annual Eye Exams

Low myopia OU

LV 1 year prior



Medical History

Asthma

ADHD

Autism

Currently seeing neurologist



Family History

Multiple family members
with early onset Parkinson's
Disease

Died in early/mid 30s

Review of Prior Neurology Notes (6 months prior)



Abnormal involuntary muscle movements

- Shoulder shrugging
- Jerking of upper extremities
- **"Tics": stimulant induced?**
 - **Started at 8 yo prior to psych meds**



Episodes of irritability and aggression

- Tantrums
- **Suicide attempt at 10 yo**
- Was on Prozac, methylphenidate, and clonidine
- Considering antipsychotic treatment



Additional History

- Normal pregnancy and delivery
- walked age 1
- started Speech Therapy at 3-4 yo
- Autism diagnosis at age 7

Exam Findings



- **BCVA** 20/25 **OD** 20/25 **OS**
- **Color** 14/14 **OD** 14/14 **OS**
- **Pupils** reactive; 4mm **Bright**/ 6mm **Dim**
OU; no APD
- **Confrontational Fields** **Full OD and OS**

UPGAZE



EOMS

Grossly full horizontal and infraduction;

No supraduction OU

Difficulty initiating conjugate eye movements on command

Intact Oculocephalic Reflex

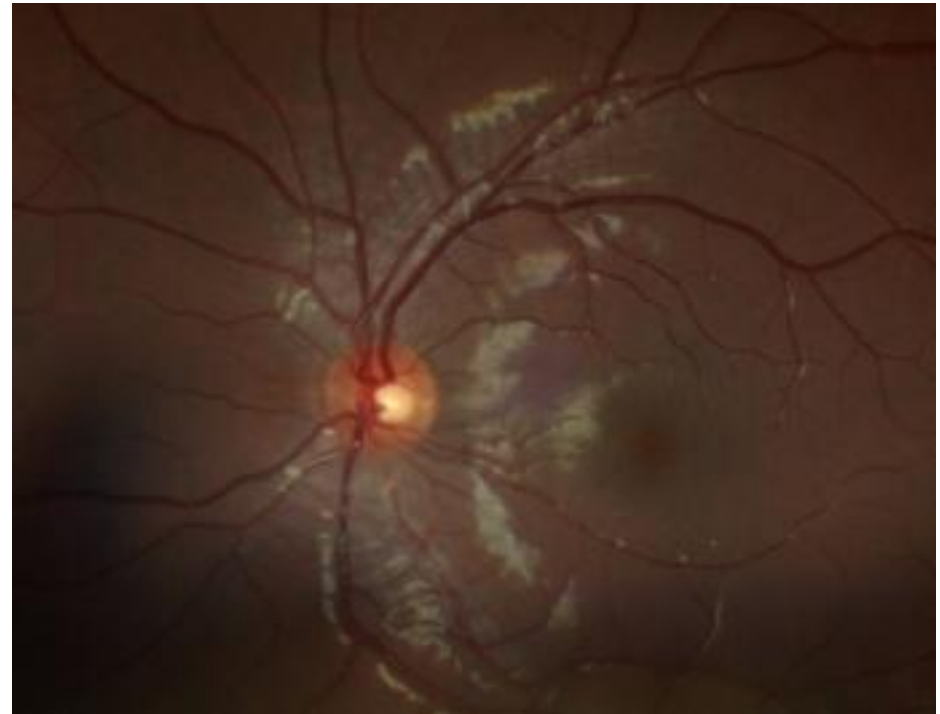
No supraduction but full with doll's head movement

Had chin down posture

Worth 4 Dot – Fusion

Ocular Health

Anterior & Posterior segment → normal OU



Mother provides additional history

Had 2nd opinion with neurology last week and MRI brain completed 2 days ago.

Brought MRI report with her.

Not yet reviewed by care team.

Results

EXAMINATION: MRI BRAIN W/O CONTRAST

CLINICAL HISTORY: New left-sided weakness, dysarthria, difficulty swallowing, cognitive changes.

IMAGING TECHNIQUE: Multiplanar, multisequence MRI of the brain was performed without administration of intravenous contrast per department protocol. Imaging was obtained on a 3 Tesla magnet.

COMPARISON: None

FINDINGS:

Midline structures: Normal, including corpus callosum and pituitary gland.

Extra axial spaces: Normal in size and configuration.

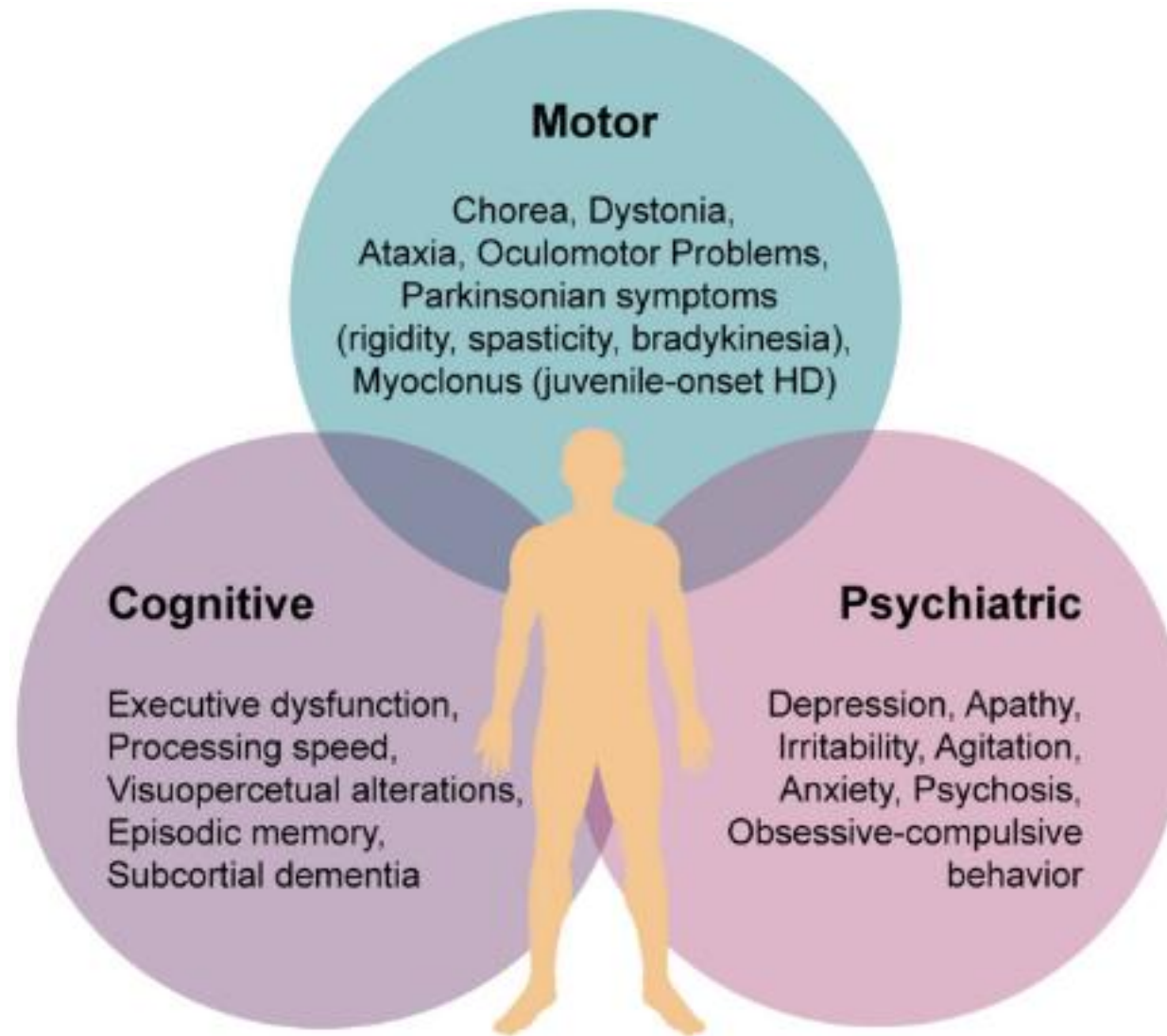
Ventricular system: There is ex vacuo dilatation of the frontal horns of the lateral ventricles related to caudate atrophy. No hydrocephalus.

IMPRESSION:

Symmetric atrophy and signal abnormality involving the caudate heads and putamina. Features are highly concerning for juvenile Huntington's disease. Genetic testing for CAG repeats is recommended.

atrophy and FLAIR hyperintensity which also demonstrate associated atrophy. There are no areas of restricted diffusion.

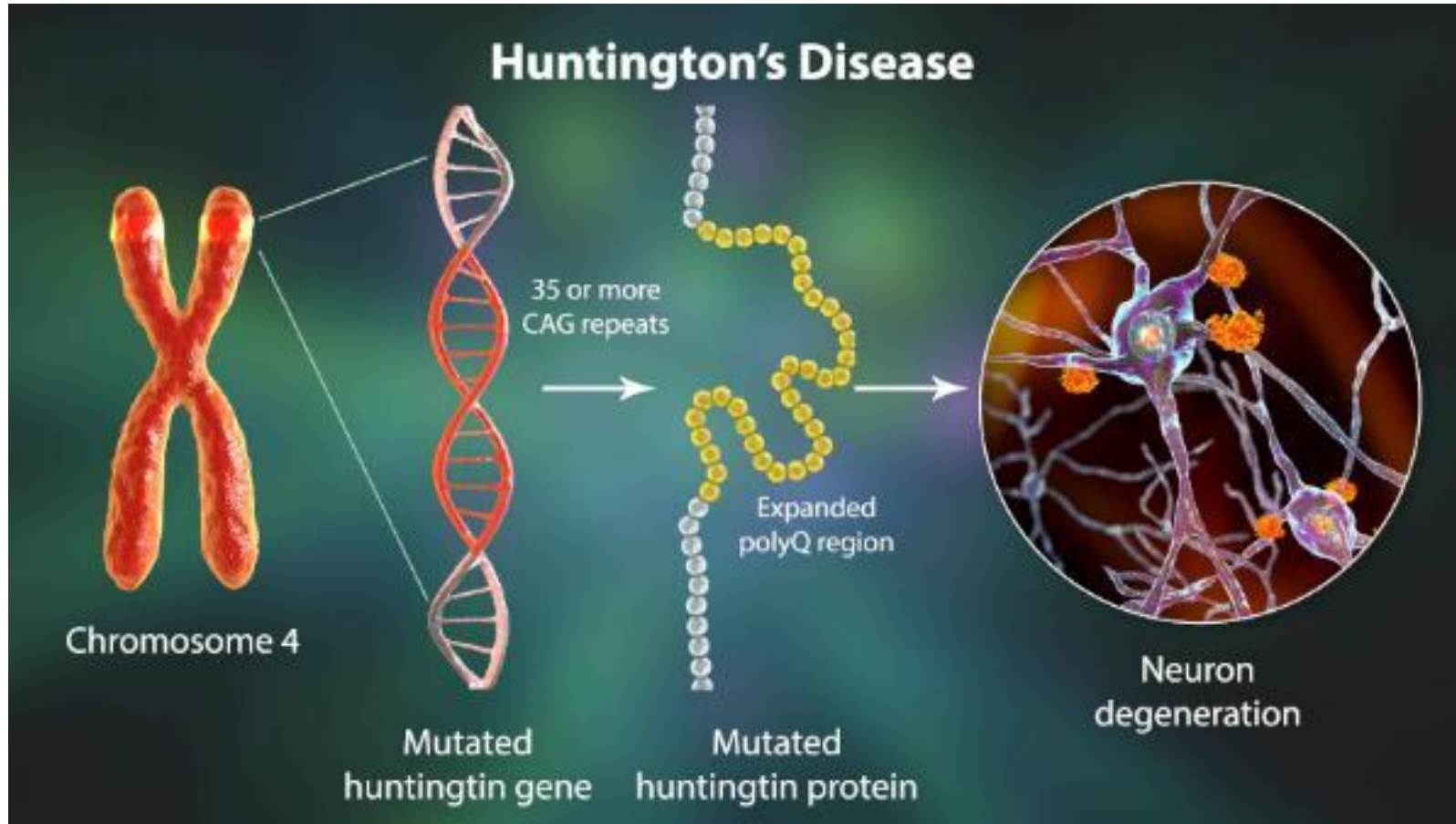
Cerebellum: Normal in signal and morphology.



Huntington's Disease

Classic Triad of Symptoms

Fernández-Valle, T., Murueta-Goyena, A. (2024). Huntington's Disease, Cognition, and Biological Markers. In: Martin, C.R., Preedy, V.R., Patel, V.B., Rajendram, R. (eds) Handbook of the Behavior and Psychology of Disease. Springer, Cham. https://doi.org/10.1007/978-3-031-32046-0_56



<https://www.lgcstandards.com/GB/en/Resources/Articles/research-huntington%27s-diseas> (accessed 9/4/2025)

- Autosomal Dominant
- **Typically travels through Paternal lineage**
- Mutation of *HTT* Gene on Chromosome 4
- > 35 repeats of CAG
- Expanded polyglutamine sequence
- **Typical onset of symptoms 30-50 years old**

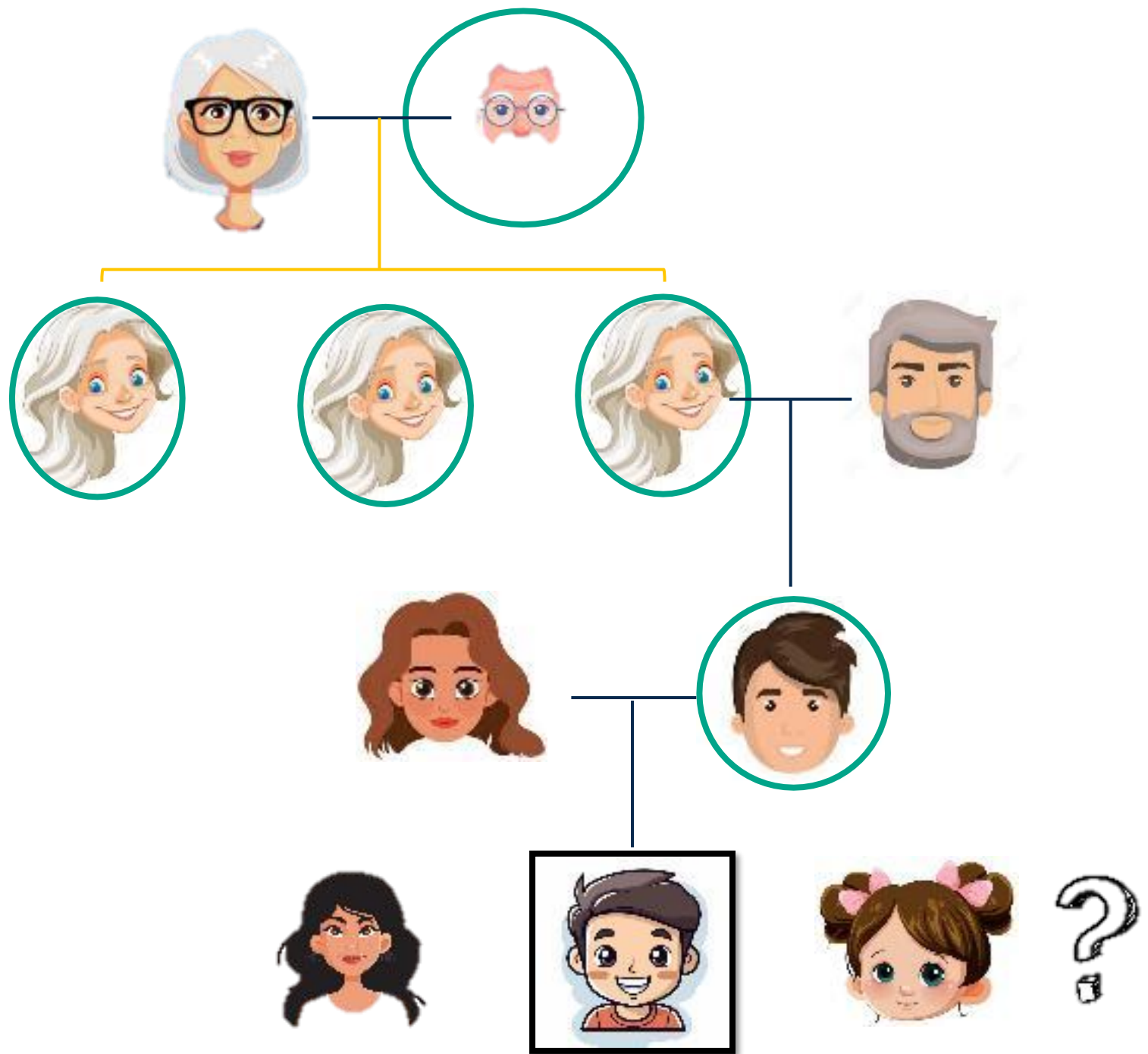
Family History

Early Onset

"Parkinson's Disease"

PGM, P aunts, PGGF

Died in early/mid 30s



Juvenile Huntington's Disease Characteristics Onset < 10 years old

Diagnostic Criteria if onset 0-10 years old

Family History

Two or more of the following:

- Declining school performance
- Seizures
- Oral motor dysfunction
- Rigidity
- Slurred speech & gait disturbance in first 2 years.



MOTOR

Speech
Fine Motor
Gait
Rigidity
Bradykinesia
Dystonia
Abnormal Saccades
Spasticity



BEHAVIOR

Irritability
Aggression
Obsessive-compulsive
Disorders
Depression

Behavioral symptoms
often misdiagnosed as
ASD or ADHD



COGNITION

Declining school
performance
Regression of
language skills
Inability to properly
develop reading skills
(Better preserved
compared to adult-
onset HD)

**Control of eye
movements &
visual
information
processing
involves all
parts of brain**



NEURODEGENERATIVE DISEASES CAUSE NVI

Communicating with ODs:

Give a call and provide records in advance

Follow-up communication

Need patient to sign release

Which ODs to refer to:

Pediatric (even adults)

Low Vision

TBI (neuro-optometry)

Thank You

Erin Draper

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