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### Sudden Visual Loss XXII World Congress of Neurology Santiago, Chile October 31 – November 5, 2015

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## **Disclosures**



### Financial Disclosures

- Editor Journal of Stroke and Cerebrovascular Diseases
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# Unlabeled/Unapproved Uses Disclosure Nothing to disclose

### **Objective & Key Message**



A comprehensive discussion of the differential diagnosis of visual loss requires thorough understanding of neuroanatomical circuitry, vascular supply of the visual pathways, and detailed understanding of the broad range of etiologies accounting for sudden visual loss. In this presentation, we will focus on the differential diagnosis of important causes of acute visual loss as they present in emergency rooms.

### **Think Circuitry**

- Retina
- Optic nerve
- Optic chiasm
- Optic tract
- Lateral geniculate body
- Optic radiations
- Primary visual cortex



#### Vascular Supply to Visual Pathways



Retina/orbital portions of optic nerve	Ophthalmic Artery
Anterior portion of optic nerve	Posterior Ciliary Arteries
Intracranial portion of optic nerve	ICA, ACA, Anterior Communicating Artery
Optic chiasm	ICA, ACA, Anterior Communicating Artery; Posterior Communicating Artery, PCA, BA
Optic tract	Anterior Choroidal Artery
Lateral geniculate body	Anterior Choroidal Artery (lateral); Posterior Choroidal Artery (medial)
Optic radiation	MCA (rostrally); PCA (caudally)
Primary visual cortex	PCA (majority)

### **History**



- Onset (sudden or gradual)
- Painful or painless (symptoms worse with eye movements?)
- Unilateral (monocular) or bilateral (binocular)
- Duration (seconds, minutes, hours, persistent)
- Entopic phenomena (floaters, phosphenes, after images)
- Other associations (auras, scalp tenderness, jaw claudication, malaise, fever, weight loss, corneal ulcers or abrasions)
- Past medical history (hypertension, heart disease, myocardial infarction, atrial fibrillation, diabetes mellitus, dyslipidemia, strokes, peripheral arterial disease, headaches, motion sickness)
- Family history (glaucoma, macular degeneration)
- Medications

### **Physical Examination**



- Visual acuity (VA)
- Visual fields (VF)
- Fundus examination
- Pupils (RAPD)
- Ocular motility
- Color vision
- Anterior and posterior segments

### Etiologies of RAPD

- Optic nerve disease
- Large amount of retinal disease
- Optic Tract (contralateral)
- Contralateral RAPD may occur with pretectal lesions without VF loss
- Rarely amblyopia or vitreous hemorrhage

Adapted from: PW Brazis, JC Masdeu, J Biller. Localization in Clinical Neurology, 6<sup>th</sup> Edition. Wolters Kluwer Health. LWW 2011.

### **Selective Disorders**



- Optic Neuropathies
  - Demyelinating (MS, NMO)
  - Inflammatory non-demyelinating
  - Autoimmune
  - Infectious
  - Ischemic
    - AION, PION
    - Non-arteritic v arteritic
  - Traumatic
  - Toxic
  - Nutritional
  - Compressive
  - Hereditary
  - Infiltrative
  - Radiation
  - Paraneoplastic

- ↓ VA
- ↓ Color vision
- VF defect
- Ipsilateral RAPD in unilateral or bilateral, asymmetric cases
- Pupillary light-near dissociation in bilateral and symmetric cases
- Optic disc edema or optic disc atrophy

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### **Selective Disorders**



- Amaurosis fugax
- Central retinal artery occlusion
- Branch retinal artery occlusion
- Central retinal vein occlusion
- Branch retinal vein occlusion

### Papilledema

- TVOs
- Bilateral
- Enlarged blind spot
- No APD
- Idiopathic intracranial hypertension (IIH)



### Clinical Definition of IIH (Modified Dandy Criteria)



- Signs and symptoms of increased ICP
- No localizing neurologic findings (CN VI palsies allowed)
- Normal neuroimaging (with exception of an empty sella)
- Opening pressure on LP > 250 mm water, with normal CSF (protein content may be low)
- Other causes of increased ICP excluded

### **Miscellaneous**



- Pituitary apoplexy
- Unilateral and bilateral homonymous hemianopia
- Cortical blindness
- Non-organic visual loss

A 43-year-old man with history of ulcerative colitis (on sulfasalazine), came to the ED complaining of severe bilateral temporo-parietal headaches associated with nausea, vomiting, photophobia and blurred vision. MRI is shown.







# **Pituitary Apoplexy**



- Acute hemorrhage or infarct in pituitary gland
- Multiple predisposing conditions (trauma, pregnancy, radiation, dopaminergic agonists, etc)
- Sudden severe headache
  - stretching/irritation of meningeal branches of CN V in dura matter on the walls of sella turcica
- Visual impairment (↓ VA, VF, ophthalmoplegia, Horner)
- Nausea, vomiting
- Altered mental status
- Adrenal dysfunction

# **Pituitary Apoplexy**



- CT/MRI
- Lytes, blood glucose, endocrinologic evaluation of pituitary hormones
- Medical stabilization
- High dose corticosteroids
- NS evaluation

21-year-old woman status post left kidney transplant on *Tacrolimus*, presented to the ED with new onset headaches, seizures, and visual loss. Witnessed to have 3 GTC seizures.

On examination: Afebrile. HR 112. BP 151/112 mmHg. WBC 21.6 K Hemoglobin 9.8 Platelet count 64 K BUN/Cr 61/4.84 Bicarbonate of 15 Calcium 6.5 K 4.1 Alcohol level undetected







**Posterior Reversible Encephalopathy Syndrome** 



- Reversible syndrome
- Headaches, altered mental status, seizures, visual loss
- Posterior (P-O) symmetric white matter changes on neuroimaging
- Calcarine and paramedian occipital lobe structures usually spared
- Numerous causes/associations, including RCVS

### **Causes of Cortical Blindness**



- Trauma
- Schilder's disease
- Cerebral angiography
- CO poisoning
- Meningitis
- Air embolism
- Neoplasm
- Tentorial herniation
- Cardiac arrest
- SLE
- Dialysis disequilibrium
- PRES



Three hours ago, a 67 year-old man suddenly lost vision in his right eye. He had no eye pain or headache, and could only see hand movements with that eye. The figure shows the fundus of the right eye.





When a 74 year-old woman awoke this morning, she could only detect light with her left eye. She had no eye pain, but for the past two months had severe generalized headaches, tenderness of her scalp, aching of her jaws when she chewed, myalgias and fatigue. The figure shows the left optic disk.



For the last six weeks a 25-year-old obese woman has had headaches, horizontal binocular diplopia, and transient, brief, bilateral visual gray-outs precipitated by postural changes. Examination showed esotropia of the OD and a paretic right lateral rectus muscle. Visual fields showed enlargement of the blind spots. The figure shows her fundi.







Two days ago, a 30-yearold woman had left monocular painful visual loss associated with left eye pain increasing with eye movements. Visual acuity 20/70 OS & 20/20 OD. Left central scotoma. Left RAPD. Normal funduscopy.

- Acute, usually unilateral painful loss of vision
- ↓ VA (variable visual loss)
- VF (variable ON visual field defects)
- RAPD in unilateral or bilateral but asymmetric cases
- Normal or swollen optic nerve head

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### Neuromyelitis Optica

#### Autoimmune Aquaporin-4 Channelopathy

- >90% relapsing course
- ON most frequently unilateral
- Higher propensity for more posterior parts of ON
- Bilateral simultaneous or sequential
- 5 years after disease onset
  - Blind in one eye
  - Blind in both eyes







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