“Neuro-ophthalmological emergencies constitute vision or life-threatening conditions if diagnosis and treatment are not promptly undertaken.

Even with immediate therapy, these clinical entities carry a high rate of morbidity.”

Signs of Neuro-Ophthalmological Emergencies

- Optic nerve edema or pallor
- Extraocular/intraocular abnormality
- Multiple cranial nerve palsies
- Pupil-involving CN III Palsy
- Anisocoria
- Ptosis

PEARL for Concern: If you have more than one of the following
- Pupil abnormality
- Eyelid abnormality
- EOM abnormality


Patient History in a Neuro-Ophthalmological Emergency

HPI
CC: Diplopia
CC: Vision Loss

Location
Monocular or Binocular?

Extent
Gaze dependent?
- Left vs. Right, Up vs. Down, Distance vs. Near
Central or peripheral visual field?
Left vs. right visual field?
Quadrant/location?

Onset
When did it start?
Sudden or gradual?
What were you doing?

Frequency
Is it getting better, worse or staying the same since it started?

Duration
How long does it last? (seconds, minutes, hours or days)
Intermittent or constant

Timing
Is it worse at he beginning or end of the day?

What makes it better?
Covering an eye? Blinking?

Has it happened before?
History of childhood strabismus, previous eye surgery?


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History of childhood strabismus, previous eye surgery?

Acute Vision Loss

1) Does visual acuity improve with a pinhole?
YES
NO

2) Pupils – Presence of RAPD?
YES
NO

3) Visual Field Defect?

1) Do you see double with both eyes open?
YES
NO

2) Cover the right eye, do you still see double?
YES
NO

3) Cover the left eye, do you still see double?
YES
NO

Binocular
Monocular

Giant Cell Arteritis

Epidemiology

- Most common systemic vasculitis affecting adults >50yo
- Rare in people <50yo
- Average age of onset is 74-76yo
- For each decade after 50, incidence increases from
  - 2.0 (50-60yo)
  - 11.8 (61-70yo)
  - 31.3 (71-80yo) per 100,000 persons/year
- Women affected 2-3x more than men
- More common in whites, Nordic/Northern European ancestry, and other northern latitudes

Pathogenesis

- Infectious? immune trigger in a genetically predisposed subject
- T-cell mediated granulomatous inflammation of medium- and large-vessels
  - Aorta
  - External carotid artery
  - Posterior ciliary artery → AAION
  - Temporal arteries

Why is it an emergency?

- Blindness
- Vision loss ~10% of patients
- Stroke
- Aortic aneurysm or dissection
  - The sooner GCA is diagnosed and treated, the lower the incidence of visual loss
  - However, patients presenting with poor vision have little chance of recovery despite immediate steroid treatment
  - The main goal of treatment is to prevent vision loss in the fellow eye
  - Usually occurs within days in 50% of cases of untreated GCA

Ocular Manifestations

Symptoms

- Sudden visual loss
- Most frequent symptom ~50% of cases
- Transient visual loss ~30%
- Often followed by permanent visual loss
- Diplopia ~6%
- Eye pain ~8%

Signs

- AAION (6.9%) → 1.3 per 100,000 population
  - (+) RAPD
  - Pale/dull ON edema
  - (+/-) retinal cotton wool spots
  - Accounts for 85% of cases of permanent vision loss
- CRAO (1.6%)
  - CilioRAO (0.4%)
  - Posterior ION
  - Ocular Ischemic Syndrome
- Why is it an emergency?
  - Blindness
  - Vision loss ~10% of patients
  - Stroke
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  - Usually occurs within days in 50% of cases of untreated GCA

Pathogenesis

- Persistent vessel wall inflammation → vascular damage → stenosis, occlusions, and aneurysms


Systemic Manifestations

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jaw Claudication (48%)</td>
<td>Temporal artery tortuosity, prominence, and/or tenderness</td>
</tr>
<tr>
<td>Neck pain (17%)</td>
<td></td>
</tr>
<tr>
<td>Headache (57%)</td>
<td></td>
</tr>
<tr>
<td>Scalp tenderness (20%)</td>
<td></td>
</tr>
<tr>
<td>Weight loss (40%)</td>
<td></td>
</tr>
<tr>
<td>Anorexia (31%)</td>
<td></td>
</tr>
<tr>
<td>Myalgias (28%)</td>
<td></td>
</tr>
<tr>
<td>Malaise (37%)</td>
<td></td>
</tr>
</tbody>
</table>

20% of cases with permanent vision loss from GCA may present without systemic symptoms of GCA.

Differential Diagnosis

- Common or migraine headache
- Atherosclerosis of large vessels
- NA-AION
- Takayasu arteritis
- Other forms of vasculitis
- Polyymalgia rheumatica
  - 30-50% of patients with GCA also have PMR
  - PMR is 2.3x more common than GCA

Diagnosis

Seroological Studies
- Elevated ESR (95.7% sensitivity)
  - Normal in 9.2-14.3%
- Elevated C-Reactive Protein (97.5% sensitivity)
  - Normal in 1.7%
- ESR/CRP can also be elevated in infection and cancer
- Abnormal CBC w/differential (sensitivity <50%)
- Thrombocytosis
- Normocytic anemia
- Leukocytosis

Fluorescein angiography
- Differentiate AION from NAION
  - Choroidal hypofusion
  - Delayed choroidal filling

Temporal Artery Biopsy
Gold Standard

BUT only 48.85% of patients with GCA have a (+) TAB
May need sequential if high clinical suspicion

Treatment

- Steroids – IMMEDIATELY once AION is suspected
  - Oral (60mg po) vs. IV ASAP
  - Oral taper over months to year
  - Low dose Aspirin?
  - To reduce risk of ischemic events if no contraindications
  - Consult PCP/internist
- Monitor for steroid-related complications – hypertension, diabetes, osteoporosis, infection, etc.
- Smoking cessation
- Follow up – 2-4 weeks

- Only 4% of patients will improve visual loss with steroids
- 4% of patients lose vision within the first 5 days, even on steroid treatment

Hypertension and Visual Loss

Future

- Glucocorticoids are very effective at high doses
  - Relapses occur in up to 50% of patients when doses are tapered
  - Are not ideal for chronic management
- Adjunct immunosuppressants
  - Methotrexate
    - Reduce risk of relapses
    - Positive effects may take 6-8 months to emerge
    - Anti-TNFa (infliximab, adalimumab, etanercept)
    - No additional benefit above prednisone monotherapy
    - Increased risk of infection
  - Anti-IL-6 (tocilizumab)
    - May provide additional benefit to prednisone by inducing and maintaining remission for up to 52 weeks. Quick onset
  - Other targeted anti-inflammatory therapies
    - Abatacept – reduced risk of relapses
    - Ustekinumab – glucocorticoid-sparing response
    - Rituximab

Hornor’s Syndrome

- Jivraj Roberts J, Clifford A. Update on the management of giant cell arteritis. The Acta Ophthalmologica 2017; 45:3
- Etanercept
  - Anti-TNFα
- Adalimumab
- Infliximab
- Etanercept
- Methotrexate
- Chloroquine
- Methotrexate
- Azathioprine
- Plasmapheresis

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- Plasmapheresis
Pupil Pathway Review: Sympathetic

- Efferent (3 neuron chain)
  1. Hypothalamus → ciliospinal center of Bulge at C8-T2 (spinal cord)
  2. Through sympathetic chain (adjacent lung apex) → superior cervical ganglion located at level of carotid bifurcation
  3. Postganglionic neuron travels with the internal carotid artery until cavernous sinus, then follows CN6 → CNV1 → long ciliary nerves → iris dilator → MYDRIASIS

Abnormal: Pupil is larger in the Dark. Miotic pupil is abnormal

Pathophysiology

- CAD: Intimal wall disruption → intromural hematoma

Epidemiology of Carotid Artery Dissection

<table>
<thead>
<tr>
<th>Epidemiology</th>
<th>Etiology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Incidence of 2.6-5 per 100,000 population</td>
<td>Traumatic</td>
</tr>
<tr>
<td>Peak incidence – 5th decade</td>
<td>Spontaneous</td>
</tr>
<tr>
<td>Mild male predominance</td>
<td>Underlying vasculopathy</td>
</tr>
<tr>
<td>Responsible for 25% of strokes in young adults (&lt;45yo)</td>
<td>Fibromuscular dysplasia</td>
</tr>
<tr>
<td></td>
<td>Connective tissue disorder</td>
</tr>
<tr>
<td></td>
<td>Marfan's</td>
</tr>
<tr>
<td></td>
<td>Ehler's Danlos</td>
</tr>
</tbody>
</table>

Ocular Manifestations of Horner's Syndrome in Carotid Artery Dissection

- Painful third-order Horner Syndrome ~60%
  - Ptoea ~1.2mm
  - "Inverse ptosis" – when lower lid is slightly elevated
  - Miosis (anisocoria of 1-1.5mm)
  - Dilation Lag
  - WITHOUT facial anhidrosis or partial
- Less common ocular signs of CAD
  - Transient monocular vision loss
  - NAION
  - PION
  - CRAO
  - Ocular ischemic syndrome
  - Ocular motor nerve palsies

Systemic Manifestations

- Ipsilateral headache ~70%
- Ipsilateral neck pain (10-30%)
- Ipsilateral ear pain (10-30%)
- Less common
  - Lower cranial nerves affected
  - Pulsatile tinnitus
  - Vertigo
  - Dysgeusia (foul taste in the mouth)
- Diseases that affect the brainstem, spinal cord, chest or neck can present with purely ocular symptoms
- Ophthalmologic signs/symptoms can proceed ocular or cerebral infarction in 33% of patients 6-14 days
Diagnosis

Horner’s Syndrome

Pharmacologic diagnosis/localization
- Cocaine 10%
- Apraclonidine 1%
- Hydroxyamphetamine 1%

- (+) = failure of pupillary dilation after one hour (ie anisocoria >1mm remains)

Caveats
- Can’t perform cocaine and hydroxyamphetamine test on the same day (within 24-48 hours)
- Hydroxyamphetamine test may yield a false-negative, and is not commercially available

Carotid Artery Dissection

- MRI/MRA
- 85% sensitivity
- 95% specificity for dissection
- CTA of head/neck or carotid doppler ultrasound
- CT of chest
- CBC with differential

Risk factors for Aneurysmal Rupture
- Increasing age (peak 6th decade)
- Female gender
- Smoking
- Hypertension
- Heavy alcohol consumption
- (+) Family history of intracranial aneurysm or subarachnoid hemorrhage
- Aneurysm size>10mm
- Genetic disorders– polycystic kidney disease
- Aneurysm location
- PCA and basilar tip have higher rupture risk at 5 years

Epidemiology

- 9-36% of CNIII palsies are caused by an intracranial aneurysm
- Posterior communicating artery (PCA) aneurysms present with a CNIII palsy 30-60% of the time
- 40% of aneurysms are located at the level of the PCA, ophthalmic artery, and cavernous sinus

Ocular Manifestations

- Pain
- Mid-dilated pupil
- Poor or absent light reaction
- Complete or partial external CNIII palsy

Rule of the Pupil

- Complete CN III
  - Pupil-sparing → likely compressive
  - Pupil-involving → likely compressive
- Incomplete CN III
  - Pupil-sparing →???
  - 14% will have aneurysm, just present to office in an early phase before the pupillary fibers are involved
  - Pupil-involving → compressive

Compressive lesions:
- Most common – aneurysm
- Less common – tumor, trauma, congenital, uncal herniation, cavernous sinus mass, pituitary apoplexy, orbital disease, varicella zoster virus, ischemia, leukemia
Topographical localization of CNIII Palsy
- Brainstem
- Hemiparesis
- Hemisensory loss
- Other cranial neuropathies
- Subarachnoid space
- Meningeal signs
- Severe headache
- Other cranial neuropathies
- Carotid sinus
- CN – 4, 5, or 6 involvement
- Homer syndrome
- Orbit
- Proptosis
- Chemosis
- Optic neuropathy

Diagnosis and Management

Diagnosis
- Digital subtraction angiography (DSA)
- 1-2% morbidity risk
- Gold standard
- MRA/CTA
- Noninvasive
- Can detect 95% of aneurysms
- Aneurysm needs to be >5mm
- MRI/CT/LP
- ESR/CRP/CBC w differential

Management
- 70-100% of surviving patients make a complete or partial recovery of the oculomotor deficit
- Usually starting with resolution of ptosis
- Pupillary and EOM abnormalities may persist
- Sometimes aberrant regeneration

Epidemiology
- Incidence – 0.4-5 cases/100,000 per year
- Prevalence – 0.5-30 cases/100,000 per year
- Age
  - Juvenile MG
    - 10-15% of Caucasians (28)
    - 52% of Chinese cases (29)
  - Early-Onset MG (EOMG) <50yo
    - Female predominance (60-70%)
    - Between 50-60yo
    - No gender difference
  - Late-Onset MG (LOMG) >60yo
    - Male predominance

Pathophysiology
- Neuromuscular junction dysfunction leads to painless, fatigable weakness of voluntary muscles
- Autoantibodies
  - Anti-AChR
  - Anti-MuSK
- Role of the Thymus?
  - Pathogenic inflammatory triggers → chronically inflamed thymus → autoreactivity to AChR → autoreactive T Cells

Ocular Manifestations
- Ptosis
  - variable, worse with fatigue
- Diplopia/EOM involvement/Ophthalmoplegia
  - variable, worse with fatigue
- Orbicularis weakness
- Normal pupils
- Cogan’s Lid Twitch

- 15% of MG patients have ocular MG only
- Majority of patients have ocular manifestations in first year of onset
- Required to have 2 years without further generalization of MG to be diagnosed as having purely ocular form

Myasthenia Gravis
- Incidence – 0.4-5 cases/100,000 per year
- Prevalence – 0.5-30 cases/100,000 per year
- Age
  - Juvenile MG
    - 10-15% of Caucasians (28)
    - 52% of Chinese cases (29)
  - Early-Onset MG (EOMG) <50yo
    - Female predominance (60-70%)
    - Between 50-60yo
    - No gender difference
  - Late-Onset MG (LOMG) >60yo
    - Male predominance

### Systemic manifestations
- 85% of MG patients
- Skeletal Muscle Fatigue
- Facial muscles
- Proximal limb muscles
- Muscles for swallowing/breathing
- Other autoimmune disease
- Thyroid disorders (Hashimoto’s or Basedow diseases)
- Thymus problems
- Thymic follicular hyperplasia ~70%
- Thymoma (thymic epithelial cell tumor) ~10-15%

### Medical Treatment
- **Long-Acting Acetylcholinesterase inhibitor** (Pyridostigmine)
  - Improves ptosis, less effective in resolution of EOM involvement/diplopia
  - Does not affect the course of the disease
- **Immunomodulatory Therapy**
  - Oral corticosteroids
  - Effectively controls diplopia AND ptosis symptoms
  - Lowers risk of progression from OMG to MG
- Other: Azathioprine, Mycophenolate Mofetil, Methotrexate, Tacrolimus, Cyclosporine, Rituximab
- Thymectomy
- IV Ig
- Plasma Exchange

### Why is it life threatening?
- **Myasthenic crisis**
  - Respiratory failure due to muscle weakness
  - Severe weakness of respiratory muscles, upper airway muscles, or both
  - Usually due to poor control of generalized disease
- Other triggers for crisis
  - Concomitant use of certain antibiotic (aminoglycosides, quinolones, antimalarials), muscle relaxants, anti-consultants antipsychotics, botox, Beta-blockers (including topicalis), and iodinated radiocontrast agents
  - Systemic infection involving respiratory tract, aspiration, and surgery
  - Emotional stress, hot environment, sudden elevation of body temperature
  - Hypothyroidism
  - Requires immediate ventilatory assistance
  - Pre-immunotherapy era
    - Myasthenic crisis had significant mortality rate (up to 75%), but has fallen to <5% in recent years
    - 20-30% life-time prevalence of myasthenic crisis in patients with MG
    - Usually occurs during the course of first symptomatic presentation in the young and later in the course of disease in the elderly
    - White patients more likely to respond poorly to treatment then black patients
- **Pregnancy is known to aggravate MG**
  - Increased intracranial pressure
  - Increased intracranial mass
  - Increased intracranial pressure
  - Increased intracranial pressure

### Intracranial Space Occupying Lesions
- **Tumor**
- **Inflammation**
- **Infection**
- **Ischemic infarct**
- **Increased intracranial pressure**
  - Mass
  - Pseudotumor LH

### Diagnostic Management
- **OD Diagnostic Tests – In Office**
  - Ptosis → Fatigue, Rest, Ice Tests
- **MD Diagnostic Tests (Neurologist/Neuro-Ophthalmologist)**
  - Tension Test (Droprohonium Chloride), 95% sensitivity
  - Side Effects: Bradycardia, arrhythmia, hypotension
  - Serologic Testing for Autoantibodies, Thyroid function
  - 5% of patients with myasthenia gravis also have hypothyroidism
  - CT/MRI of Thora (rule out Thymic abnormalities)
  - Neurophysiological Specific Tests (RNS – Repetitive Nerve Stimulation)
  - SFEMG – Single Fiber Electromyography

### Serological Testing
- **Generalized Myasthenia – 96% sensitivity, high PPV**
  - 87% have autoantibodies
  - 80% - AChR
  - 40-70% MuSK
- **Ocular Myasthenia – 44% sensitivity, low NPV**
  - 50-80% have autoantibodies
  - (+)Autoantibody assay highly suggests MG
  - BUT (-)Autoantibody assay = inconclusive
- **Seronegative – (-)AChR, (-)MuSK**
  - High prevalence of cranial and bulbar muscle involvement
  - Repeated serological tests
  - 15% seroconversion rate over 1 year period
  - Patients with OMG may persistently test seronegative

### Other triggers for crisis
- **Respiratory failure due to muscle weakness**
- **Severe weakness of respiratory muscles, upper airway muscles, or both**
- **Usually due to poor control of generalized disease**

### Seroconversion
- **Increase in disease activity**
- **MOG involvement/diplopia**
- **Efﬁciency in resolution of**
- **Severe weakness of**
- **Usually due to poor control of generalized disease**

### References
Swollen Optic Nerve(s)

Papilledema

- "Disc swelling from elevated intracranial pressure"

Differential Diagnosis

- Congenital Anomaly
- Optic disc drusen
- Infection
- Inflammation
- Ischemia

Until lumbar puncture – better to diagnose “optic nerve head edema”.

Optic Nerve Edema – EVALUATION

1) SLOW YOURSELF DOWN AND BREATHE

Until lumbar puncture – better to diagnose “optic nerve head edema”.

Papilledema

Symptoms

- Headache
- Nausea
- Tinnitus
- Transient visual obscurations
- Double vision
- Other neurological deficits

Signs

- Bilateral disc edema
- - Initially superior-inferior swelling
- - Disc Hyperemia (its pink!)
- - (+) Obscuration of retinal vessels over disc margin
- - Cup is preserved
- - (+) CWS/hemorrhages over time
- - (-) Spontaneous Venous Pulsation
- - Normal VA
- - Normal Color Vision
- - (+) Visual field defect

Optic Nerve Edema

Diagnosis

- Immediate Neuroimaging
- Lumbar puncture
- Elevated intracranial (CSF) pressure

Etiology

- Mass lesion
- Severe cerebral edema
- Venous thrombosis
- Hydrocephalus
- Pseudo-tumor Cerebri

Pseudo-Tumor Cerebri

Idiopathic Intracranial Hypertension

- Normal neuroimaging
- MRI/CT brain
- Normal CSF examination
- Elevated opening CSF pressure

Associations/Risk Factors

- Obesity
- Recent weight gain
- Obstructive sleep apnea
- Anemia
- h/o medication use:
  - Glucocorticoids
  - Vitamin A products
  - Tetracycline derivatives
  - Synthetic growth hormones
  - Female predilection

IIH - Management
- Acetazolamide
  - May improve papilledema, visual complaints, headache
  - OR other diuretics/CAIs
- Weight loss
- Baseline automated VFs after treatment initiated
  - Progressive or severe vision loss may need more aggressive therapy
  - Ventriculoperitoneal shunt
  - Optic nerve fenestration

Optic Disc Pallor
If its progressing... IMAGE
Longstanding compression of optic nerve, chiasm or tract by a tumor or aneurysm can cause progressive optic disc pallor

Cavernous Sinus Lesions

Cavernous Sinus Syndromes
Complete
- Ophthalmoplegia (Diplopia)
- Ptosis
- Mydriasis
- Hypesthesia of V1/V2 (facial numbness)
- Orbital Pain
Partial
- Depends on the location of the lesion

Etiology
Chronic Causes
- Meningiomas
- Metastases of head and neck cancers
- Slow-growing carotid aneurysm
Acute Causes
- Cavernous sinus thrombosis
  - Extension of facial/sinus infection
  - Carotid cavernous fistula
- Painful red eye + chemosis + pulsatile exophthalmos
- Inflammatory reaction (Toxosa-Hunt syndrome)
- Pituitary apoplexy

Cavernous Sinus Thrombosis
Ocular Manifestations
- Most common 80-100% presentation
  - Proptosis
  - Chemosis
  - Ptosis
  - CN III, IV, and/or VI palsies
  - Less common 50-80% presentation
    - Periorbital edema
    - Optic disc edema
    - Venous engorgement
  - Least common <50%
    - Decreased visual acuity (due to ION, CRAO, CRVO, or corneal ulceration)
    - Sluggish/dilated pupils
    - Periorbital and corneal sensory loss (CNV)
Systemic Manifestations
- Most common 80-100% presentation
  - Acute onset fever
  - Less common 50-80% presentation
  - Headache
  - Lethargy
  - Altered sensorium
  - Least common <50%
  - Meningismus
  - Seizures
  - Hemiparesis


Image for: Cavernous Sinus Lesions
Cavum
Internal Carotid Artery
External Carotid Artery
Inferior Orbital Fossa
Sphenoid Sinus
Ethmoid Sinus

e:366-370

Image for: Cavernous Sinus Syndromes

Image for: Etiology

Image for: Cavernous Sinus Thrombosis

Image for: Cavernous Sinus Syndromes

**Differential Diagnosis**

**Orbital Cellulitis**
- Painful ophthalmoplegia
- Chemosis
- Fever
- Decreased vision
- UNILATERAL

**Direct High-Flow Carotid Cavernous Fistula**
- Periorbital edema
- Ophthalmoplegia
- Increased IOP
- Decreased vision
- (+) supraorbital bruit
- Arterialized conjunctival vessels

---

**Pituitary Apoplexy**

**Epidemiology**
- Between 2-12% of patients with adenoma experience apoplexy
- Diagnosis of pituitary tumor unknown at time of apoplexy in ¾ of cases
- Presentation for 0.6-9.0% of surgically managed pituitary adenomas
- Male predominance ~60%
- Peak incidence in 5th decade

**Pathophysiology**
- Hemorrhage or infarction of a pituitary tumor causes a sudden enlargement of the gland due to ischemia and/or necrosis
- 2/3 are spontaneous
- 1/3 precipitating factor:
  - Hypotension
  - Surgery
  - Malignant hypertension
  - Anticoagulant treatment
  - Dopaminergic agonist treatment

---

**Ocular Manifestations**
- Visual Field Loss
  - Bitemporal
  - Junctional scotoma
- Unilateral and/or bilateral ophthalmoplegia ~50%
  - CNIII> CNVI> CNIV

---

**Systemic manifestations**
- Sudden severe headache ~80%
  - Retroorbital
  - Bifrontal
  - Diffuse
  - Assoc. with vomiting/nausea
- Neck stiffness
- Brain stem/hypothalamus compression
  - Reduced consciousness
  - Thermoregulatory dysfunction
  - Cardiorespiratory dysfunction
- Pituitary dysfunction
  - Thyrotropic deficiency
  - Hypothyroidism
  - Corticotrophic deficiency
  - Hypopituitarism
  - Hyperpituitarism
  - Hypercortisolism

Symptoms evolve from hours to 2 days after onset of apoplexy


**Summary**

- Neuro-ophthalmological emergencies can present to primary care optometry
- The best way to prepare is to have a systematic process for examination and workflow
- Sometimes as an optometrist you get to save a life

**Thank You!**

Jacqueline.theis@berkeley.edu

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**References**

Glauser A, Broner MD. Pituitary apoplexy: pathophysiology, diagnosis, and management. Arch Endocrinol Metab. 2015;59(3):239-44


Grose JS, McColl CM, Lee VS. An approach to examination and workflow. Primary care optometry. Sometimes as an optometrist you get to save a life. The best way to prepare is to have a systematic process for examination and workflow. Thank You! Jacqueline.theis@berkeley.edu

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**References**

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