KMK Optometry Board Certification
Review: NOA Quiz Bowl

Instructors:
Kyle M. Cheatham, O.D., F.A.A.O.
Christopher Wolfe, O.D., F.A.A.O.
www.kmkoptometryboardcertification.com
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KMK Contributors

Kyle M. Cheatham, O.D., F.A.A.O. graduated from Centre College with a B.S. in Biology and completed optometry school at Indiana University School of Optometry before accepting a residency at the Kansas City VA Hospital. Dr. Cheatham practices at Heartland Eye Consultants, a secondary eyecare center in Omaha, Nebraska, where he specializes in ocular disease and low vision. He serves as an adjunct faculty member of six colleges of optometry teaching optometric interns at Heartland Eye Consultants. Dr. Cheatham also provides consultative care within the ophthalmology department at Creighton University Medical Center.

Melissa A. Cheatham, MPAS, PA-C graduated from the University of Nebraska, Lincoln, with a Bachelor of Science degree in Communication Studies before receiving her Master’s in Physician’s Assistant Studies from the University of Nebraska Medical Center. She is a board certified physician assistant and has clinical experience in family practice, obstetrics, and gynecology and most recently in cardiology with the Bryan Heart Institute in Lincoln, NE.

Christopher Wolfe, O.D., F.A.A.O. graduated from the University of Nebraska at Omaha with a B.S. in Biotechnology and completed optometry school at Northeastern State University Oklahoma College of Optometry. While in school he served as president of the American Optometric Student Association. Dr. Wolfe practices at Exclusively Eyecare in Omaha, Nebraska. He specializes in anterior segment pathology with emphasis on post-surgical and corneal ectatic contact lens cases.

Kevin B. Wood, Ph.D. graduated Magna Cum Laude from Centre College with a B.S. in Chemical Physics before completing an M.S. in Biology and earning a double Ph.D. in Theoretical Physics and Physical Chemistry from the University of California, San Diego (UCSD). He is currently a Postdoctoral Research Fellow in biological physics at Harvard University, where his research focuses on nonequilibrium dynamics in stochastic and nonlinear systems.

Sarah Dougherty Wood, O.D., M.S., F.A.A.O. graduated Magna Cum Laude from University of Evansville with a B.S. in Biology and Chemistry and graduated with honors from IU School of Optometry. She completed a residency at the Kansas City VA Hospital, where she also served as director of the low vision program, VICTORS, for 3 years. Sarah completed a two year research fellowship at the Boston VAMC in 2009 and received a Master’s degree in Vision Science from The New England College of Optometry in 2010. She currently practices at Tufts Medical Center and The Dorchester House while also serving as adjunct faculty at NECO and an instructor of ophthalmology at Tufts University School of Medicine. She is a member of the Optometric Glaucoma Society and her research has been featured in The Journal of Glaucoma.
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Chapter 1

Interactive Review: NOA Quiz Bowl

1.1 Central Retinal Vein Occlusion (CRVO)

Assessment

- CRVO’s result from compression of an artery on a vein; this leads to turbulent blood flow, venous vessel wall damage, and thrombus formation. CRVO’s are usually caused by a thrombus at or near the lamina cribosa (24). Thrombus formation leads to ischemia and release of VEGF and characteristic retinal signs, including retinal hemorrhages, collaterals, dilated tortuous retinal veins, cotton-wool spots (CWS), macular edema, and optic disc edema.

- Risk factors for CRVO include (non-exhaustive list) hypertension (61%), diabetes, cardiovascular disease and open-angle glaucoma (28). Glaucoma is the ocular disease most commonly associated with CRVO’s (17).

- CRVO’s are characterized by sudden, unilateral painless vision loss in an elderly patient (90% are > than 50 years old) (19). CRVO’s are the 2nd most common vascular cause of vision loss; diabetic retinopathy is most common (30).

- In young CRVO and BRVO patients consider the following conditions for etiology: oral contraceptive pills, protein S/protein C/antithrombin III deficiency, factor XII deficiency, antiphospholipid antibody syndrome, collagen vascular disease, and AIDS (19).

- Vision threatening complications include macular disease and complications from neovascularization. VEGF stimulates neovascularization of the posterior and anterior segment and has been proven to cause capillary leakage leading to macular edema (24).

1. Macular Disease - ischemia and edema
2. Neovascularization - vitreous hemorrhage, neovascular glaucoma, tractional retinal detachment
Macular edema is the leading cause of vision loss in both ischemic and non-ischemic CRVO’s (24).

- CRVO’s can be grouped into non-ischemic (67%) and ischemic categories (19). Ischemic CRVO is defined as 10 disc diameters or more of non-perfusion on fluorescein angiography. 90% of cases present with 20/200 vision or worse - prognosis is poor (14). Visual acuity is typically counting fingers or worse in these patients (31). 16% of nonischemic cases progress to ischemic.

- Neovascular glaucoma is a major concern in patients with CRVO’s (“90-day glaucoma”). 60% of ischemic cases develop iris neovascularization; up to 33% develop neovascular glaucoma (33). Overall, less than 10% of non-ischemic cases develop rubeosis or angle neovascularization (28).

Treatment/Management

Neovascularization

The Central Retinal Vein Occlusion Study (CVOS) demonstrated the following: If rubeosis (> 2 clock hours of iris), angle neovascularization, neovascular glaucoma or any posterior segment neovascularization develops, panretinal laser photocoagulation (PRP) is indicated. If no neovascularization is present (as in this case), prophylactic panretinal photocoagulation is not beneficial.

Macular edema

The Central Retinal Vein Occlusion Study (CVOS), Standard Care vs. Corticosteroid for Retinal Vein Occlusion (SCORE) study, and CRUISE trial provide evidence for how to treat CRVO-related macular edema.

- CVOS: demonstrated that focal laser photocoagulation of the macula does not improve visual acuity in patients with CRVO’s

- SCORE study: demonstrated that intravitreal steroids can improve visual acuity in patients with CRVO’s who have macular edema. After 12 months, 27% of patients with a 1mg triamcinolone injection achieved the desired result - an improvement in visual acuity of 15 or more letters; only 7% of patients in the sham (control) group achieved the desired visual outcome (34).

- CRUISE trial: demonstrated that ranibizumab (anti-VEGF agent) can improve visual acuity in patients with CRVO’s who have macular edema. After 12 months, an improvement of three or more lines was observed in 46% of the subjects in the 0.3mg ranibizumab group, 48% of the subjects in the 0.5mg ranibizumab group, and 17% of the subjects in the sham (control) group (13).

- Final visual acuity with a CRVO is typically similar to what the acuity was on initial presentation. The risk of the fellow eye eventually having a CRVO is approximately 10% (31).
1.1. CENTRAL RETINAL VEIN OCCLUSION (CRVO)

- Patients should be followed every month for 6 months, with gonioscopy at each visit (33) (28). Underlying medical conditions should be evaluated; including hypertension, elevated blood sugar, and congestive heart failure (28). Always check blood pressure in the office. Patients should be evaluated or referred for underlying systemic etiology; consider ordering blood tests to help determine the etiology.

Dry Age-Related Macular Degeneration (ARMD)

Assessment

Dry ARMD - accounts for 85 – 90% of cases of ARMD (23) (19). Most patients with dry ARMD do not have vision loss; metamorphopsia, gradual vision loss (months to years), and blurred vision are common complaints. Dry ARMD is characterized by the presence of drusen (hallmark of ARMD); associated RPE abnormalities (mottling, granularity, geographic atrophy, focal hyperpigmentation) may also be present. 12% of all dry ARMD patients will develop severe vision loss (defined as loss of >6 lines) (19); the majority of these cases result from geographic atrophy.

- ARMD is the leading cause of blindness in the U.S. population for patients over 50 years old (19). ARMD is the 2nd leading cause of blindness for patients 45-64 years old (diabetes is 1st) (22).

- Framingham Eye Study revealed that 6.4% of patients 65-74 years old and 19.7% of patients greater than 75 years old had signs of ARMD (25).

- Risk factors include increasing age (especially 75 years and older), ethnicity (Caucasians most at risk), positive family history, light iris color, cigarette smoking, hyperopia, hypertension (HTN), hypercholesterolemia, female gender, and cardiovascular disease (19). Nutritional factors and light toxicity are believed to play a role in pathogenesis (19).

Hyperopia greater than 0.75 diopters (D) increases the risk of exudative ARMD by up to 2.5 times (21). 10 – 20% of patients with ARMD have at least one first-degree family member with vision loss (21) (20).

The Macular Photocoagulation Study Group established four main risk factors that increase the risk of progression from dry to wet ARMD - they include (26):

1. Multiple soft drusen (especially if confluent)
2. Focal hyperpigmentation
3. Hypertension
4. Smoking
Treatment/Management

Management for dry ARMD involves the following:

- **Cessation of smoking.** Smoking is the number one risk factor consistently associated with all forms of ARMD (28). The Age-Related Eye Disease Study (AREDS) concluded that current smokers should NOT take beta carotene at high doses (15 mg) due to increased risk of lung cancer. Smokers should be advised to utilize formulations in which lutein has been substituted for beta carotene.

  Current smokers are more likely to develop ARMD (2.4-2.5 times more likely) and to have a recurrence of choroidal neovascularization (CNV) (1.7-2.2 times more likely) as compared to those who have never smoked (12).

- **Monitoring of Amsler grid daily.** If distortion is noted, patients are instructed to call the office immediately.

- **Consideration of high-dose antioxidants** and vitamins for patients with category 3 (intermediate) and category 4 (advanced) ARMD; AREDS revealed a modest benefit in these patients (see box below) but no benefit in patients with category 1 (early) or category 2 (mild) ARMD.

  AREDS demonstrated an absolute risk reduction of 6% (29 – 23%) in helping patients with more advanced ARMD (categories 3 and 4); these patients lost less than 15 letters (3-lines) of visual acuity over the 5 year period as compared to placebo (35).

- **Low-dose antioxidants** (iCAPS, Centrum silver) for patients with category 1 and category 2 ARMD and patients with strong family history of the condition (19).

- Low-vision rehabilitation is warranted for patients with functional vision loss.

  The AREDS formula includes beta-carotene (15 mg), vitamin C (500 mg), vitamin E (400 International Units), zinc (80 mg) and copper (2 mg) (11). This represents 5 times the usual intake of vitamin C from diet alone, 13 times the recommended daily allowance (RDA) for vitamin E, and five times the RDA for zinc (28).
1.1. CENTRAL RETINAL VEIN OCCLUSION (CRVO)

Coats Disease

- Idiopathic peripheral vascular disease that results in unilateral (80 – 95%) telangiectatic dilated vessels that display a characteristic “light bulb” appearance. Progression of the disease can lead to marked hard exudates (classic for Coats), intraretinal hemorrhages, exudative retinal detachment, and neovascular glaucoma; the latter of which can result in a red, painful eye.

- Peak incidence is in males (85%) prior to age 20; 2/3 of cases will be diagnosed prior to age 10 (19). Children may present with poor vision, strabismus, or leukocoria. If left untreated, Coats disease will gradually progress to a total exudative retinal detachment (15) (16).

Differentials for leukocoria include Coats disease, toxocariasis, retinoblastoma, and ROP.

Myopic Choroidal Atrophy

- Pathological myopia is defined by refractive error in excess of -6.00D with an axial length greater than 26mm. Axial lengthening in the anteroposterior direction results in scleral thinning and choroidal atrophy (28).

- Condition has genetic predisposition with elongation of the globe beginning in early childhood (36). Occurs in 2% of U.S. population, most commonly in women during young adulthood (23) (19) (28).

- Characteristic signs include posterior staphylomas (hallmark of condition), oblique insertion of the optic disc, Fuch’s spots (hyperpigmented spots in the macula from RPE hyperplasia), lacquer cracks, macular holes, premature cataracts (NS and especially PSC), extensive vitreous syneresis, posterior vitreous detachment, peripheral retinal degenerations (e.g. lattice, snail-track, pavingstone), retinal breaks, and retinal detachments (32) (29).

Lacquer cracks occur in about 5% of high myopes. They appear as fine, yellow irregular lines that represent large breaks in Bruch’s membrane - choroidal neovascularization can result and lead to severe vision loss (23). Lacquer cracks frequently present in young males, and may be one of the earliest findings in pathological myopia (27).

Diabetic Retinopathy (DR)

Assessment

- DR results from damage to the blood-retinal barrier; pericytes are lost and retinal capillary basement membranes become damaged.
• DR can be divided into **Nonproliferative Diabetic Retinopathy** (NPDR) (Background DR) and **Proliferative Diabetic Retinopathy** (PDR). PDR is worse and indicates the presence of neovascularization. PDR occurs in 5% of patients with DR (28). **The 4-2-1 rule** (see below) can help determine what NPDR patients are at a higher risk for developing PDR.

• The most important risk factor for DR is **duration** of insulin-dependent diabetes. When diabetes is diagnosed before the age of 30, the risk of developing DR is about 2% per year. After 7 years - 50% will have DR; after 25 years - 90% will have DR (28).

• DR is the leading cause of new cases of blindness in the United States for adults ages 20-74 (17). DR can present with numerous signs; however, the two major concerns with DR are **macular disease** and **proliferative disease** (neovascularization). **Macular edema** is the most common reason for legal blindness in DR.

**Vision threatening complications of DR** include **macular disease** and **neovascularization** complications:

1. **Macular Disease** - ischemia, edema

2. **Neovascularization** - preretinal/vitreous hemorrhage, neovascular glaucoma, tractional retinal detachment

**Macular ischemia** can look normal or edematous; a fluorescein angiogram is required to differentiate from macular edema. There is no treatment for macular ischemia.

**Treatment/Management**

The Diabetic Retinopathy Study (DRS) examined when to treat neovascularization. The **DRS** defined high risk characteristics (HRC’s) as any of the following (28) (18):

**HRC’s**

1. Neovascularization of the Disc (NVD) greater than 1/4 Disc Diameter (DD).

2. Any NVD or Neovascularization elsewhere (NVE) with a vitreous or preretinal hemorrhage.

**The DRS** demonstrated that PRP reduced the risk of severe vision loss by 50 – 60% in patients with HRC’s.
1.1. CENTRAL RETINAL VEIN OCCLUSION (CRVO)

The Early Treatment Diabetic Retinopathy Study (ETDRS) determined when to treat macular disease and when PRP should be utilized for treatment in patients with NPDR (whether to treat at moderate NPDR, severe NPDR, etc).

The ETDRS defined Clinically Significant Macular Edema (CSME) and Severe NPDR (“4-2-1 rule”):

CSME

1. Retinal thickening within 500 um (1/3 DD) of the foveal center
2. Hard exudate within 500 um of the foveal center, with adjacent thickening
3. Retinal thickening of at least 1 DD, within 1 DD of the foveal center

To be diagnosed with CSME, the patient only needs to have one of the three criteria.

The ETDRS demonstrated that focal argon laser treatment of patients with CSME reduced the risk of moderate vision loss by 50% or more (18); thus, patients with CSME should be treated. The ETDRS found that patients with mild to moderate NPDR should not be treated with PRP. Patients with severe NPDR and early PDR received minimal benefit from treatment (18).

Severe NPDR (“4-2-1 rule”)

1. 4 quadrants of hemorrhages/microaneurysms
2. 2 quadrants of venous beading
3. 1 quadrant of intraretinal microvascular abnormalities (IRMA)

Patients should be diagnosed with severe NPDR (pre-proliferative DR) if they meet one of the three criteria within the 4-2-1 rule. 10 – 50% of patients with severe NPDR will develop PDR within one year (28). If the patient meets 2 of the 3 criteria of the 4-2-1 rule, they have VERY severe NPDR and their likelihood of progression to PDR is even higher.

Preretinal / Vitreous hemorrhage

Assessment

- Preretinal and vitreous hemorrhages result from the same etiology - conditions that cause retinal neovascularization (e.g. diabetic retinopathy, retinal vein occlusion, sickle cell retinopathy, retinopathy of prematurity
(ROP), ocular ischemic syndrome). In each of these cases, the neovascularization is preretinal and the newly formed vessels lack endothelial tight junctions. The location (preretinal) and strength (leaky) of these vessels creates a situation where vitreous traction can cause shearing of the vessel, resulting in hemorrhage formation.

- A **preretinal hemorrhage** is located between the retina and an intact posterior vitreous face. A **vitreous hemorrhage** is located anterior to the posterior vitreous face (within the vitreous).

- Vitreous hemorrhages are characterized by sudden, painless vision loss and/or black spots that can have corresponding flashing lights. Mild cases of vitreous hemorrhage will reveal blood that obscures part of the fundus view; severe cases will not allow any view of the fundus, while chronic cases cause the fundus to appear gray-white and/or yellow in appearance.

Diabetic retinopathy is the most common cause (31% – 54%) of spontaneous vitreous hemorrhage (31).

**Treatment/Management**

- Vitreous hemorrhages can be treated with **pars plana vitrectomy** for non-clearing diabetic vitreous hemorrhages for >1 month and persistent (>6 months) idiopathic vitreous hemorrhages (19).

- **B-scan** is indicated if no fundus view can be obtained. Bed rest, with head elevation, is recommended for 2-3 days and aspirin, warfarin (Coumadin), and other anti-clotting agents are discontinued (if systemic health allows).

**Stargardt’s Disease**

- Autosomal recessive inherited condition that presents between the ages of 6 and 20 with symptoms (rapid vision loss) that do not correlate with signs (minimal fundus irregularities). Acuity is typically 20/200 by the third decade and stable or slowly progressive thereafter (19). Stargardt’s disease is the most common hereditary macular dystrophy (19). There is no sex predilection.

stARgardt’s = AR condition

- As condition progresses, bilateral **yellow flecks** appear scattered in a **pisciform (fish-tail) configuration** throughout the posterior pole and mid-periphery. Non-specific RPE mottling of the macula may also be apparent. Color vision and electroretinography (ERG) also start to become
abnormal. In late stages a classic “beaten-bronze” macular pattern is apparent (bull’s eye maculopathy) and “salt and pepper” pigmented changes may appear in periphery.

- **Fundus flavimaculatus** and Stargardt’s Disease are considered variants of the same disorder. Fundus flavimaculatus diagnosis is reserved for patients without macular dystrophy signs; it often presents later in life (4th, 5th decade) and patients are commonly asymptomatic (19) (23). Vision loss can still occur if fleck-lesions involve the macula.

- No treatment available. Schedule consult with low-vision specialist for patients with functional vision loss.

Similar to pain in the early stages of *acanthamoeba* that does not correlate with corneal findings, *Stargardt’s* begins with decreased vision that is out of proportion with retinal findings.

### Macular Hole

#### Assessment

- Condition results from **posterior vitreous traction** on the macula. 83% of cases are associated with aging (50-70 years old) and **women** (3:1) are more commonly affected (31) (19). Less common causes include trauma (10%), surgery, cystoid macular edema (CME) or inflammation (19) (18).

- Patients typically report decreased vision and/or metamorphopsia. 20/200 acuity and worse is associated with full-thickness macular holes.

- Condition is characterized by round, red, well-delineated spot in macula. Can be classified according to the following stages:
  
  1. **Stage 1** - Impending hole, loss of foveal depression with **yellow spot** or ring.
  2. **Stage 2** - Round, small full-thickness hole with pseudo-operculum present.
  3. **Stage 3** - Large, full-thickness hole present with operculum. Positive **Watzke-Allen** sign now apparent (19) (18).
  4. **Stage 4** - Stage 3 plus PVD.

A positive **Watzke-Allen** test is characterized by a complete break in the middle of a thin line of light projected within the macular area (28).
Treatment/Management

- **Stage 1 holes** - no treatment because spontaneous hole closure can occur. However, 50% progress and will subsequently warrant surgical intervention.

- **Stage 2-4 holes** - If recent onset (<1 year) and acuity within the range of 20/40 - 20/400, surgery has better likelihood of success (19). Surgical intervention consists of **pars plana vitrectomy**, peeling of the posterior hyaloid, and injection of gas. Peeling of the internal limiting membrane may also be performed. Patients must maintain **face-down positioning** for 1-2 weeks following the procedure to allow the gas bubble to tamponade the retina (31) (19).

The risk of developing a macular hole in the fellow eye is 10 – 20% (very small risk if PVD is present) (28). Condition has bilateral onset in 25 – 30% of cases (18).

Lattice Degeneration

- Area of peripheral retinal thinning that is typically circumferential (often cigar-shaped) and concentric with the ora serrata (31).

- The inner portion of the lesion is atrophic (thin), while the outer margins of the lesion have a firm adhesion to the vitreous. The majority of lesions do not contain the criss-cross pattern of white lines (sclerosed vessels); only 12% of patients have this classic appearance (18).

- Lattice is present in 6 – 10% of patients; 20 – 33% of patients with rhegmatogenous retinal detachments will have lattice degeneration; however, only 1% of patients with lattice degeneration develop a retinal detachment (28).

- Lattice degeneration is often bilateral (33 – 50%) (19) and more commonly located temporal (than nasally) and superior (than inferiorly) (23). In 25% of cases, lattice degeneration will contain an atrophic hole; retinal tears can result from vitreoretinal traction on the atrophic, thinned retina (19).

- Patients with Marfan’s syndrome, Stickler syndrome, and Ehlers-Danlos syndrome can have lattice-like lesions (“atypical lattice”) that increase the patient’s risk of a RD (23).

**Retinal detachments (RD’s)** result from separation of the sensory retina from the underlying RPE.
Central Serous Choroidopathy

Assessment

- Condition that results in RPE and/or choroidal dysfunction; this creates a route for fluid accumulation under the macula and subsequent complaints of blurred vision (20/20 to 20/200) and/or metamorphopsia (if macula involved). **Acute onset** with **unilateral** presentation is typical.

- Most common in **young to middle-aged men (20-50) with a type A personality**. Unknown etiology, but associated with (non-exhaustive list) stress, pregnancy, steroid use, hypochondriacal and hypertension (19). History of similar episodes is common - recurrences occur in 40% of cases (31).

- Characterized by a localized, shallow, **macular serous detachment**; 3% of cases will have an RPE detachment as well (19). Fluorescein angiography will reveal a gradual pooling of fluorescein into a pigment epithelial detachment (90% of cases) or **“smokestack” appearance** (10% of cases) (19); optical coherence tomography (OCT) is being used more frequently (as compared to FA) for diagnosis and monitoring of condition. After resolution of condition, patients often have permanent residual RPE changes within the macula.

Treatment/Management

- Set expectations appropriately. Some sources report a recurrence rate as high as 50% (18).

- Most patients improve, without treatment, by **1 to 3 months**; 94% of patients will regain > 20/30 acuity (19). 66% of patients achieve 20/20 vision (18).

- In most cases, the condition is observed, but laser should be considered for treatment in certain scenarios, including: persistent detachment after 4 months, previous CSR episode that resulted in permanent vision reduction, and patient demand for more immediate visual recovery. Laser treatment can expedite recovery process, but does not result in better final acuity (18).

1.2 The Swollen Optic Nerve

The concept of axoplasmic flow

Axonal transport along the ganglion cell axons occurs in orthograde (from eye towards brain) and retrograde (brain towards eye) directions. The net flow is in the orthograde direction. If this is reversed in a pathologic state, swelling of the optic nerve and spillover of fluid into the nerve fiber layer occur.
The importance of a bilateral versus unilateral presentation in determination of etiology

- For papilledema to occur: Cerebrospinal fluid (CSF) pressure is elevated in the subarachnoid space which causes axoplasmic stasis of both nerves.

- For unilateral disc edema to occur: Presence of pre-chiasmal disruption of the axoplasmic flow such as compression or ischemia.

Bilateral disc edema versus true papilledema

- Bilateral disc edema occurs when a pre-chiasmal process causes edema of each nerve separately. An example could be bilateral non-arteritic ischemic optic neuropathy (NAION). A person develops NAION in one eye and then at a point shortly thereafter, develops NAION in the other eye. Both nerves may appear swollen but it is not true papilledema because these were two separate processes and no increase in intracranial pressure is present.

- True papilledema is, by definition, due to increased intracranial pressure which has to affect both nerves. The swelling may be asymmetric but will both be swollen. This is considered a medical emergency.

Possible clinical findings in papilledema

- Paton’s folds, lack of spontaneous venous pulsation (SVP), enlarged blind spot on visual field testing, acuities can be normal initially but can diminish over time, splinter hemorrhages, cotton wool spots, blurred disc margins, nerve fiber layer edema

Most common causes of papilledema:

Post-chiasmal tumor, pseudotumor cerebri, malignant hypertension, an infiltrative process, compromised or obstructed venous outflow

- Post-chiasmal tumor (space occupying lesion)
  - The cranium is a fixed size and the addition of a tumor or other space occupying lesion increases the CSF pressure
  - Diagnosis based on CT/MRI imaging
  - Special case- Foster Kennedy Syndrome
    * A frontal lobe tumor presses on one optic nerve and causes atrophy. As the tumor grows, it starts to increase the intracranial pressure and therefore causes swelling of the other optic nerve. Very rare (10)

MRI is better for imaging soft tissue, CT for imaging bone, and MRA for imaging blood vessels, often using contrast.
1.2. THE SWOLLEN OPTIC NERVE

• Pseudotumor cerebri (Idiopathic Intracranial Hypertension)
  - Demographic: most commonly an overweight female of childbearing age
  - Signs and symptoms: CN 6 palsy, transient visual obscurations (TVO), headache, visual field defect (enlarged blind spot), papilledema, nausea and vomiting
  - Possible contributers: CANT mnemonic: Contraceptives, Vitamin A, Naladixic acid (antibiotic) and Tetracycline and minocycline
    Can also occur with oral steroid use or steroid withdrawal.
  - Diagnosis of exclusion (Modified Dandy criteria) (6): normal MRI/MRV (magnetic resonance venography), normal CSF composition (no infection, infiltration or neoplasm found), high opening pressure on lumbar puncture (LP), awake and alert patient, no localizing findings on neurological exam (except CN 6 palsy), no other cause of increased intracranial pressure found

High opening pressure on LP:
> 200 mm of water for a normal weight individual
> 250 mm of water for an obese individual (8)

Anterior ischemic optic neuropathy

Two types: Arteritic and Non-arteritic

• The anterior portion of the optic nerve is perfused by the posterior ciliary arteries. In arteritic AION (A-AION), occlusion of these arteries occurs which results in poor perfusion. In non-arteritic AION, the exact causal mechanism of poor perfusion is unknown.

• Importance of distinguishing between the two: If A-AION is not treated promptly with steroids, the other eye can become involved and the patient can go bilaterally blind. The typical time interval of vision loss between the first and second eye ranges from 1 to 14 days (7). High doses of oral steroids can have severe side effects and require a long taper. Therefore, the diagnosis of A-AION should be fairly certain before beginning this treatment.

• Arteritic AION
  - Cause: Most often Giant Cell Arteritis (GCA) aka Temporal Arteritis. Other etiologies include: polyarteritis nodosa, systemic lupus erythematosus, and herpes zoster (37).
  - GCA is a systemic condition which can obstruct the medium to large sized blood vessels throughout the body. Acute visual loss occurs in approximately 7% to 60% of patients with GCA (5).
  - Who is at risk: >55 years of age (9), 3 times more common in women (37)
– Symptoms and presentation: Sudden, unilateral loss of vision with chalky, white disc edema. May also have systemic symptoms such as: temporal headache, scalp tenderness, malaise, anorexia*, neck pain*, jaw claudication*, tenderness or reduced pulsation of temporal artery, and/or polymyalgia rheumatica (*= most common). One out of 5 patients with GCA at risk for blindness never have any systemic symptoms (37).

– Testing: Must perform Westergren sedimentation rate (sed rate), complete blood count and CRP (C reactive protein) STAT. The sed rate and CRP, when both elevated, are 97% specific to GCA (37). A temporal artery biopsy may be done if the blood tests are equivocal or the clinical picture is not clear. Skip lesions on temporal artery biopsies can lead to a false negative result.

– Treatment and management: High dose IV or oral steroids which need to be initiated ASAP if A-AION is diagnosed.

– Prognosis: Vision will not return to the affected eye.

<table>
<thead>
<tr>
<th>Important association:</th>
<th>Elevated Westergren Sedimentation rate (mm/hr):</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Men: &gt; age/2, Women: &gt; (age + 10)/2 Elevated CRP: &gt;0.8mg/dL(4)</td>
</tr>
</tbody>
</table>

### Non-arteritic AION

– Who is at risk: >50 years of age (9), no gender predilection, HTN (about 50% of the time) (8), DM or other vascular disease, sleep apnea, disc at risk (<0.4 c/d), possible risk with erectile dysfunction drugs but this is not clear

– Symptoms and presentation: sudden, unilateral, painless loss of vision which often is noticed after awakening. Swollen nerve, significant vision loss, and an APD.

– Testing: The classic visual field defect is inferior altitudinal but other defects are possible. Normal sed rate and CRP.

– Treatment and management: No treatment, must rule-out arteritic AION

– Prognosis: Typically non-progressive

### Papillophlebitis

– Who: young, healthy individuals

– Presentation: Retinal venous stasis and hemorrhage, disc edema, vision no worse than 20/30, no APD, enlarged blind spot on visual field. Essentially a CRVO in a young, healthy patient.

– Etiology: unknown

– Prognosis: Will recover completely after months to 1 year (8)
1.2. THE SWOLLEN OPTIC NERVE

Oculomotor Nerve, (CN III)

Assessment and Differential Diagnosis of CN III palsy

Symptoms: Depending on the severity of the ptosis, the patient may or may not notice diplopia due to blocking of the vision in one eye.

Signs:

• A complete palsy will have ipsilateral ptosis with an eye that is down and out.

• Incomplete is anything less than complete, examples include:
  – A right inferior oblique palsy will present with a right hypotropia in primary gaze, vertical diplopia worse on gaze up and to the left and worse with head tilt left. The two other main conditions that can present this way are an inferior oblique muscle entrapment and Brown’s syndrome. The differential between all three is forced duction testing. The inferior oblique palsy will have a negative test (will comply) while Brown’s and entrapment will have a positive forced duction test.
  – Right medial rectus palsy - horizontal diplopia worse on left gaze, negative forced duction.
  – Right superior rectus palsy - right hypotropia primary gaze, worse on gaze right and head tilt right, negative forced duction.

Etiology: 25% will be idiopathic (2), most common known causes for a CN III palsy are microvascular infarct, trauma, and aneurysm.

Pupil involvement: Pupil involvement is an important characteristic that can guide clinical management. A compressive lesion (tumor or aneurysm) will almost always involve the pupil (fixed and dilated) due to the superficial location of the pupil fibers along the nerve. An ischemic palsy (likely due to microvascular disease) will likely not have pupil involvement.

Treatment and Management Options of CN III palsy

Treatment will depend on the extent of EOM and pupil involvement.

• If the palsy is incomplete or the pupil is involved (dilated), an immediate MRI/CT is indicated.

• If the palsy is complete, the pupil is NOT involved, the patient is over the age of 40, they have known vascular disease (such as diabetes, hypertension or smoking), there are no other neurological abnormalities and no new symptoms on follow-up;
  – It is assumed the palsy is ischemic in nature
  – Fresnel prism or patching if symptoms dictate
  – Monitor for resolution - if no resolution by 3 months → MRI/CT
  – Aberrant regeneration never occurs in ischemic CN III palsies.
**CHAPTER 1. INTERACTIVE REVIEW: NOA QUIZ BOWL**

**Pathological Causes of Anisocoria**

**Assessment and Differential Diagnosis**

- If the abnormal pupil is miotic (bigger difference in the dark):
  1. Horner’s syndrome
  2. Uveitis (posterior synechiae) or acute episode
  3. Argyll Robertson

- If the abnormal pupil is fixed and dilated (bigger difference in the light)
  1. Adie’s Tonic pupil
     - If the pupil constricts with .125% pilocarpine, the patient has Adie’s. If there is no response with .125% then proceed to 1% pilocarpine.
  2. CN III (will also have ptosis and diplopia)
     - If 1% causes constriction, then the patient most likely has a 3rd nerve palsy. If 1% fails to constrict the pupil than the origin is due to trauma or a pharmacological agent (patient used an oral or topical medication that caused dilation) (1).
  3. Pharmacologic
  4. Trauma (to sphincter muscle - usually after intraocular surgery)

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Cluster

Cluster headaches typically affect men in between 20-50 years of age.

Assessment and Differential Diagnosis

Symptoms:

- F: 1-2 times per day for 1 month, followed by up to years without an episode
- L: Frontal, periorbital, or temporally located with severe pain
- O: Commonly wake the patient from sleep
- P: Alcohol, smoking, nitroglycerin
- D: 1-2 hours
- R: Cold, oxygen, analgesics

Signs:

- Can cause an ipsilateral transient or permanent Horner's syndrome (2)
  - Facial redness
  - Conjunctival injection

Treatment and Management Options

- If Horner’s is present confirm it is postganglionic (1% hydroxyamphetamine), if preganglionic or other atypical symptoms, consider neuroimaging.
- See treatment options for migraines.

VF defects that respect the horizontal midline are found anterior to the chiasm; glaucomatous defects are the number one culprit. VF defects that respect the vertical midline are found posterior to the chiasm. Strokes are by far the most common reason for a post-chiasmal VF defect. In fact, 90% of homonymous hemianopsias are caused from strokes (1). However, the one main exception to this rule occurs with macular only homonymous hemianopsias – these are rarely caused by strokes (1).
Trochlear Nerve (CN IV)

Contains two nuclei (one on each side), in the midbrain, at the level of the inferior colliculus. CN IV leaves the midbrain dorsally and crosses over. The right superior oblique is innervated by the right CN IV, which has its nucleus on the left side. Because CN IV has the longest course, it is susceptible to trauma.

Assessment and Differential Diagnosis of CN IV palsy

**Symptoms:** Vertical diplopia without ptosis

**Signs:** Head tilt and turn away from the involved eye, limited depression on adduction

**Etiology:**

1. Congenital
   - Long history of head tilt - old pictures
   - Fuller face on the opposite side of the head tilt (same side as the paretic muscle)
   - No torsional diplopia complaints - may not see a torsional component with Maddox rod
   - Large vertical vergence ranges

2. Acquired - trauma, vascular, iatrogenic (sinus surgery, orbital surgery), or tumor
   - No facial asymmetry
   - Acute onset
   - Torsional Diplopia

In a study of 190 patients with superior oblique palsy, 72% were congenital, 28% were acquired. Of the acquired, 54% were due to trauma (history), 23% were iatrogenic, 13% were vascular, and 10% were caused by a tumor. So of the 190 patients with a superior oblique palsy only 2.8% were due to a tumor. (3)

**Treatment and Management Options of CN IV**

- Blood pressure, consider blood sugar (BS) and hemoglobin A1C if microvascular disease is suspected.
- If large vertical fusional vergence ranges or old pictures show head turn, consider prescribing vertical prism based on associated phoria findings.
- If progressive, not isolated (other neurological findings present), younger than 40 and no head trauma, or 40-55 with no microvascular risk factors, neuroimaging should be considered.
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