Pigment Dispersion/Pigmentary Glaucoma

Description
- A pathologic dispersion of pigment on the corneal endothelium (Kruckenberg’s spindle), iris surface, trabecular meshwork (TM), posterior equitorial surface of the lens, and anterior hyaloid face of the vitreous
- Mid-peripheral slit defects in the iris are pathognomonic.
- More common in young, adult, myopic men
- Results from posterior bowing of the iris causing iridozonular contact
- Subsequent mechanical rubbing releases pigment that disperses
- After exercise or dilation with epinephrine, a shower of pigment may be released into the AC and the IOP may increase.
- May lead to glaucoma if pigment clogs TM causing IOP to be elevated
- Patients have an increased incidence of lattice degeneration with vitreous adhesions and subsequent holes and tears.

Differential Diagnosis
1. Pseudoexfoliation glaucoma
   - White, flaky, dandruff-like material is located at pupil border and deposits on the endothelium and anterior lens capsule; mostly in older females.
   - Transillumination defects seen at pupil border due to a loss of pigment from posterior iris surface.
   - Pigment can accumulate in the TM causing an elevation in IOP
2. Iris melanoma
   - Raised pigmented lesion on iris or a diffusely pigmented iris
   - No iris transillumination defects
3. Phenothiazine, e.g. Thorazine, Mellaril, toxicity

Workup
- Look for Kruckenberg’s spindle.
- Transilluminate, and look for slit iris defects.
- Perform gonioscopy and look for excessive pigment in the TM.
- Look for pigment on the lens equator
- Run baseline glaucoma evaluation

Treatment
- Miotics
- Trabeculoplasty
- Peripheral iridotomy
Dural Cavernous Fistula

Description
- A carotid cavernous fistula is an abnormal communication between the internal carotid artery (ICA) and the cavernous sinus (CS).
- The fistula allows a mixing of arterial blood, within the ICA or its tributaries, with the venous blood within the CS.
- If the ICA is breached, the arterial blood under high pressure flows retrograde within the CS and enters the veins of the orbit, extra-ocular muscles, lids, conjunctiva, and globe; dramatic signs and symptoms occur.
- If a dural tributary of the ICA tears (dural cavernous fistula), arterial blood under lower pressure flows retrograde within the CS and enters the veins of the orbit, globe and adnexa; mild to moderate signs and symptoms occur.

Signs and Symptoms
- Dilated, corkscrew-like conjunctival and episcleral vessels
- Exophthalmos because the superior ophthalmic vein and EOMs are distended and push the globe forward
- Diplopia because of swollen, constricted EOMs and compression of the VI cranial nerve by the inferior petrosal sinus
- Severe lid and conjunctival edema
- Globe pulsation and bruit because the systole of the heart is translated to the arterialized superior ophthalmic vein
- Elevated IOP because the episcleral veins are arterialized, and the episcleral venous pressure is elevated

Management of Dural Cavernous Fistula
- Dural cavernous fistulas occur without antecedent cause in middle-aged females and often resolve spontaneously.
- If the dural cavernous fistulas do not resolve, arterial blood may flow into cortical veins and lead to cerebral venous infarction.
- Transvenous embolization via microcatheterization
- Treat elevated IOP with aqueous suppressants and/or prostaglandin derivatives

Vernal Keratoconjunctivitis (VKC)

Description
- A chronic, perennial, bilateral, allergic conjunctivitis involving the cornea; predominantly affects young males; characterized by intense itching, thick, ropey, mucous discharge, significant chemosis, and keratopathy (ranging from diffuse SPK to a superior-located, sterile shield ulcer)
- Most cases manifest GPC on the superior tarsal plates; limbal papillae and limbal Horner’s- Trantas’ dots are more common in darkly pigmented patients
Etiology and pathophysiology
- Type 1 immediate hypersensitivity reaction with vasoactive amine release via degranulation of mast cells and Type 4 delayed hypersensitivity reaction with cytokine release from T cells and macrophages
- Elevated levels of histamine and eosinophils
- Superior corneal irritation and epithelial breakdown from superior tarsal GPC and from enzymes released by eosinophils

Treatment
- Avoid allergen
- Stage the presentation and treat accordingly: In acute presentations utilize a brief pulse of topical steroids, e.g., Alrex, Lotemax, FML, Vexol, Pred Mild or topical cyclosporine, e.g., Restasis to deactivate T cells and macrophages.
- In preallergy season utilize mast cell stabilizers, e.g. Alomide, Opticrom.
- In early allergy season with mild signs and symptoms, utilize cold compresses and cold lubrication and topical antihistamines, e.g., Emadine; or antihistamine-mast cell stabilizers, e.g., Patanol, Zaditor, Elestat, Optivar
- If signs and symptoms suggest late-phase allergy, prescribe topicals which inhibit mast cell release and recruitment of eosinophils and other late phase inflammatory cells, e.g., Alocril, Alamast.
- Treat shield ulcers with heavy lubrication, antibiotic-steroid combination, e.g., Tobradex, cycloplegia (if needed), and remove of any mucous, fibrin, or serum that is coating the base of the ulcer.
- If VKC is refractory consider subtarsal injection of steroid, e.g triamcinolone, and continue as above.

Hyphema

Description
- An accumulation of blood in the anterior chamber resulting from a tear in the blood vessels at the root of the iris
- Usually results from blunt trauma to the front of the eye – contracoup injury
- The injury may tear the iris away from the ciliary body (iridodialysis).

Workup
- Rule out globe rupture - severe subconjunctival hemorrhage and edema along with restricted ocular motility.
- Rule out orbital floor fracture - displaced globe, restriced elevated gaze, reduced sensation in the distribution of the infraorbital nerve, emphysema of the periorbital tissues, positive CT scan.
- Quantify degree of hyphema (if greater than 50%, there is an increased risk of glaucoma and endothelial decompensation leading to blood staining of the corneal stroma).
- Applanation tonometry QD until hyphema resolves
- Dilate pupil and evaluate fundus if patient has not sustained a concussion (with concussion, you want to make sure there is no subsequent compression on CN III).
- No gonioscopy or scleral depression so as not to disturb clot
- Rule out sickle cell disease if patient is black. (sickled cells may block TM causing IOP to rise; reduced perfusion to the optic nerve may occur at lower IOPs).
- Attempt to prevent rebleed within 2-5 days post trauma so as to prevent increase in IOP and decompensation of endothelium.

Management and Treatment
- Attempt to encourage clotting for at least 3 days; then encourage reabsorption of the clot.
- Hospitalize patient if he is too young to cooperate or if systemic medications will be administered.
- Advise bed rest with head elevated 30 degrees (lowers venous pressure so as to reduce chance of rebleed, decreases IOP, facilitates settling of the blood).
- Advise against bending or lifting of heavy weights.
- Utilize Fox shield to prevent accidental jarring of eye.
- Recommend acetaminophen for pain (aspirin may induce rebleed).
- Utilize atropine 1% BID to immobilize pupil and to stabilize traumatic iritis; utilize Pred Forte to reduce the inflammation of traumatic iritis.
- Consider aminocaproic acid (Amicar) systemically to preserve the clot.
- Reduce IOP if above 30 mm Hg in a caucasian or if above 24 mm Hg if patient has sickle cell trait (slight elevations of IOP are beneficial as a tamponade and tend to encourage clotting)
- Consider irrigation and aspiration if elevated IOP is nonresonsive, hyphema is greater than 50% and corneal staining is beginning to occur, and if hyphema has not begun to clear in 4 days.
- Gonioscopy and scleral depression 30 days post hyphema and then yearly thereafter; evaluate for angle recession.

Optic Neuritis

Description
- Optic neuritis is an acute, rapidly progressing, but short-lasting inflammation of the optic nerve.
- It usually presents unilaterally and affects patients between the ages of 18-45; it may occur in children and present bilaterally.
- There is usually some degree of ocular pain that is exacerbated on eye movement. Vision loss, dyschromatopsia, decreased contrast sensitivity, decreased light-brightness sensitivity, and an afferent pupillary deficit are present to a variable degree.
Clinical Presentation
- Papillitis – the intraocular form of optic neuritis in which there is pink, disc edema with an occasional flame-shaped hemorrhage and a few vitreous cells in front of the disc; it is often bilateral and occurs more frequently in children and young adults
- Retrobulbar optic neuritis – the optic nerve, fundus, and vitreous appear unchanged because the inflammation is behind the lamina; it is usually unilateral

Etiology
- Idiopathic
- Multiple sclerosis (MS)
- Childhood viral diseases e.g., measles, mumps
- Mononucleosis
- Herpes zoster
- Granulomatous infiltrations e.g., syphilis, sarcoidosis, TB
- Intraocular inflammations e.g., scleritis, uveitis, retinitis
- Inflammations of the orbit and sinuses

MS
- A chronic, inflammatory, demyelinating disease of the CNS most commonly affecting females between ages 20-40
- Non-ocular symptoms include paresthesias, clumsiness, ataxia, incontinence
- Ocular manifestations include: optic neuritis, ocular motor disturbances e.g., diplopia, nystagmus, gaze palsy, impaired pursuits and saccadics, internuclear ophthalmoplegia (lesion is in the medial longitudinal fasiculus - on attempted lateral gaze there is weakness or absence of adduction in the eye ipsilateral to the lesion and nystagmus in the contralateral abducting eye
- Diagnosis of clinically-definite-MS requires at least 2 neurologic events that have separate anatomic etiology in the CNS and that occur at different times. (separated in both space and time)
- Neuronal conduction is subsequently reduced.
- Relapsing-remitting MS occurs in 80% of cases. (Symptoms occur; patient stabilizes; patient improves; and then patient relapses.)
- Primary progressive MS occurs in 20% of cases. Symptoms are progressive and non-remitting, and the patient continues to deteriorate.

Diagnosing MS
- MRI with gadolinium dye reveals multifocal, demyelinating white plaques and scarring around the ventricles (the hallmark of chronic MS); MRI may also show demyelinating plaques within the optic nerve, brainstem, cerebellum, and spinal cord.
- Spinal fluid analysis - elevated IgG; oligoclonal bands
- VER of the optic nerve and spinal cord reveals a delay in conduction.
Optic Neuritis Treatment Trial
- After optic neuritis, 50% of patients reveal MRI evidence of demyelination; many of these patients are initially asymptomatic.
- Within 5-10 years of the initial optic neuritis episode, approximately 50% of patients with 2 or more plaques that are larger than 3mm in size will manifest clinically significant MS.
- 13%-16% of patients who demonstrate no white matter plaques after optic neuritis will be diagnosed with MS in 5 years.
- If patients revealed white matter plaques after optic neuritis, utilizing IV methylprednisolone for 3 days followed by oral prednisone for 11 days accelerated the recovery of vision and delayed the onset of MS.

Current Management approaches to MS
- If patients with optic neuritis have white plaques on MRI, treatment with IV methylprednisolone for 3 days followed by oral prednisone for 11 days, delays the onset of new clinical manifestations of MS.
- Systemic interferon Beta-1a (Avonex), interferon Beta-1b (Betaseron), or glatiramer acetate(Copaxone), reduce exacerbations and slows progression

Superior Limbic Keratitis

Description
- Bilateral episodes of superior bulbar conjunctival and limbal thickening and inflammation
- Fluorescein and rose bengal staining of superior conjunctiva and corneae
- Superior pannus
- Dense papillae on superior tarsal plates
- Corneal filaments

Epidemiology
- Middle-aged females
- Fifty percent manifest hyperthyroidism and associated lid retraction

Treatment
- Mild cases – lubrication; moderate cases – topical steroids; severe cases - apply .05% silver nitrate solution to suprabulbar conjunctiva via cotton swab
- Occlude superior puncta (1997 AJO)

Ocular Sarcoidosis

Description of Sarcoidosis
- A chronic, non-infectious, granulomatous, multisystem disease of unknown etiology occurring mostly in blacks
- Lung tissue and hilar lymph nodes are most commonly involved.
- Ocular manifestations may include conjunctival nodules, infiltrations within the lacrimal gland, trabecular meshwork, and optic nerve head, granulomatous or non-granulomatous anterior uveitis, vitreous opacities resembling snow balls, peripheral retinal venule nodules (periphlebitis) resembling candle wax drippings, and disseminated chorioretinitis

**Workup**
- History of dyspnea along with red nodules along shins
- Biopsy any suspicious nodules
- Order blood tests and blood chemistry (eosinophils, angiotensin converting enzyme, calcium, and lysozyme may be abnormally elevated)
- Order chest x-ray (lung infiltrations and hilar adenopathy) and Gallium scan (abnormal uptake in the lacrimal glands, parotid glands, submaxillary glands and hilar lymph glands)

**Treatment of Ocular Manifestations**
- Treat anterior uveitis with topical steroid, e.g., Pred Forte, at a loading dose, and cycloplegia e.g., atropine TID, .5 % homatropine BID-TID, .25% scopolamine BID-TID

**Treatment of Sarcoidosis**
- 50% resolve spontaneously; 10% die
- Systemic steroids
- Immunosuppressants e.g., methotrexate, azathioprine, chlorambucil, cyclosporin A, cyclophosphamide

**Fuchs’ Endothelial Dystrophy**

**Description**
- A bilateral, slowly progressive, inherited corneal endothelial disease usually affecting women over the age of 50 and resulting in corneal edema
- Begins as a guttata - a disruption to the endothelial mosaic appearing as dark spots with an associated fine pigment dusting
- Endothelium decompensates and loses its pumping ability to keep cornea relatively fluid-free
- As edema progresses forward in the stroma, folds develop in Descemet’s membrane (striae), stroma thickens, stromal haze occurs, and epithelial vesicles (bullae) form; when bullae rupture, pain ensues
- Chronic stromal edema can lead to vascularization
- Primary open angle glaucoma often occurs

**Differential diagnosis**
1. Polymorphous dystrophy - a rare congenital defect occurring early in life; endothelial vesicles appear in a linear pattern surrounded by a haze; stromal
edema may occur; many cases are associated with anterior cleavage syndromes i.e., posterior embryotoxin, iridocorneal adhesions, and glaucoma.

2. Congenital hereditary endothelial dystrophy - bilateral congenital corneal edema with no association with congenital glaucoma.

**Treatment of Fuchs’ Endothelial Dystrophy**
- Reduce IOP
- Use hair dryer in the AM to dehydrate the edematous stroma
- Hypertonic saline to reduce epithelial edema
- Full thickness corneal transplant or DSAEK

**Subclavian Steal Syndrome**

**Blood supply to the posterior portion of the brain**
- The vertebral arteries branch off the subclavian arteries and travel upward and posteriorly through the transverse lamina of the spinal column.
- At the foramen magnum, the vertebral arteries fuse into a single midline vessel, the basilar artery.
- The basilar artery sends branches to supply the mesencephalon, pons, cerebellum, medulla, and occipital cortex.

**Pathophysiology**
- An atherosclerotic plaque is located in the subclavian artery, proximal to the point where the vertebral artery branches off.
- When the patient exercises the ipsilateral arm, blood flows retrograde in the vertebral artery and is “stolen” from the from the vertebro-basilar system.

**Clinical Manifestations (Partial List)**
- Unilateral, bilateral or alternating ophthalmoplegia
- Unilateral or bilateral Horner’s syndrome
- Bilateral hemianopsia (bilateral dimming of vision)
- Vertigo and ataxia
- Difficulty in swallowing (dysphagia)
- Difficulty in articulation (dysarthria)
- Headache, vomiting, syncope

**Management**
- Percutaneous transluminal angioplasty and/or stenting
- Carotid-subclavian by pass graft
Ocular Ischemic Syndrome

Description of Carotid Insufficiency/Ocular Ischemic Syndrome
- A usually unilateral, generalized ischemia to the anterior and posterior ocular segments occurring in patients over the age of 60
- Anterior segment: orbital pain, dilated, tortuous, episcleral arteries, corneal edema, mild flare and cells, poorly responsive pupil, neovascularization of the iris and/or angle often leading to neovascular glaucoma
- Posterior segment: reduced IOP, dilated, non-tortuous retinal veins, neovascularization of the optic disk and/or other parts of the retina, mid-peripheral blot and dot hemorrhages, arteriole plaques, artery occlusions (CRA or BRA)

Etiology of Ocular Ischemic Syndrome
- Ipsilateral atherosclerotic carotid stenosis that is greater than 80%
- Associated with diabetes mellitus, hypertension, ischemic heart disease, and cerebrovascular disease

Workup of Carotid Insufficiency/Ocular Ischemic Syndrome
- History
  1. Consider the risk factors of hypertension, smoking, hypercholesterolemia, diabetes mellitus, obesity and sedentary life style, cardiac dysfunction, vasculitis, and hyperviscosity syndromes.
  2. TIAs?
  3. Light-induced amaurosis?
- Slit lamp, ophthalmoscopy to evaluate anterior and posterior segments
- Auscultation of the carotid vasculature
- Perform visual fields.
- Palpate the facial pulses.
- Determine the blood pressure in each arm.
- Order Duplex Doppler scanning/and or MRA.
- Obtain CBC and blood chemistry data; rule out coagulopathies.
- Obtain ESR and C-reactive protein data to rule out giant cell arteritis.

Treatment
- Reduce risk factors; antiplatelet therapy; Coumadin
- PRP and/or antiVEGF therapy
- Carotid endarterectomy if the stenosis is greater than 70% and the patient is symptomatic

Case Macular Pseudo-hole

Description
- An area of normal retina within a zone of significant macular epiretinal
membrane formation; the epiretinal membrane pulls the inner portion of the fovea toward the center causing the foveal contour to steepen into a hole-like configuration.
- The outer nuclear layer and the photoreceptor layer are preserved.

**Differential Diagnosis**
- Cystoid macular edema – often follows cataract surgery
- Full thickness macular hole

**Pathophysiology of Macular Epiretinal Membrane**
- Develops at the vitreoretinal interface; consists of proliferating glial cells that have gained access to the retinal surface through breaks in the ILM (ILM)
- Breaks in the ILM may be created when the posterior vitreous detaches.
- Macular epiretinal membranes are divided into cellophane maculopathy and macular pucker.

**Cellophane Maculopathy**
- A thin transparent membrane of epiretinal cells
- Patient may be asymptomatic or may present with mild VA deficit or mild metamorphopsia
- Best detected with red-free light
- In time the membrane may thicken and contract causing fine striae on the retinal surface and distortion of the blood vessels.

**Macular Pucker**
- Caused by thickening and contraction of the epiretinal membrane
- Retinal wrinkling and distortion of the blood vessels are more manifest.
- Vision deficit and metamorphopsia are more manifest.
- White striae may obscure the underlying blood vessels.
- A macular pseudo-hole may form within the epiretinal membrane.

**Treatment of Epiretinal Membrane**
- Vitrectomy and membrane peel

**Macular Hole**
- Develops as a result of progressive vitreoretinal traction at the fovea
- Progresses through four stages
  - **Stage 1a** is an impending hole that appears as a yellow foveolar spot with a loss of foveal depression.
  - **Stage 1b** results from centrifugal displacement of the foveola; characterized by a yellow ring around the fovea and mild VA deficit and mild metamorphopsia
  - **Stage 2** is an early, small full-thickness hole that appears as an oval or crescent.
  - **Stage 3** is a larger full-thickness hole about the size of 1/3 of a disc diameter; has a small cuff of subretinal fluid; the posterior cortical vitreous remains attached.
Stage 4 is a larger full-thickness hole with a complete PVD; the hole is surrounded by a larger cuff of subretinal fluid; often has tiny yellow flecks at bottom of the hole

Diagnosis of Macular Hole
- Watske-Allen test – a narrow slit beam is projected vertically and horizontally over the center of the hole. Patients with a true hole will note that the slit beam is broken; patients with a pseudo-hole will note that the slit is thin but not broken.
- Optical coherence tomography (OCT)

Treatment of Macular Hole
- If the VA is less than 20/60 and the duration has been less than one year, vitrectomy and removal of ILM followed by injection of gas into the vitreal cavity; patient must maintain a face-down position for one to two weeks to allow the gas bubble to tamponade the hole.
- Hole can be closed and the VA improved in 80%

Central Serous Chorioretinopathy (CSR)

Description
- An exudative chorioretinopathy characterized by a neurosensory retinal detachment
- Appears as a thin, clear, blister-like lesion underneath the macula; the foveal reflex is lost; there are often sub-retinal precipitates.
- There may be an associated retinal pigment epithelium detachment.
- A positive scotoma, i.e. metamorphopsia and/or micropsia, is present.
- An increase in hyperopia is present.
- Most cases occur in men between ages 20-50; however, pregnancy is a known risk in females.
- CSR is associated with Type A personality features in individuals who have elevated stress and endogenous stress hormones, i.e. corticosteroids and epinephrine.
- Other etiologies include topical steroids, steroid inhalers, or systemic steroids.

Pathophysiology
- The inner choroidal vasculature undergoes vasoconstriction; some of the overlying RPE cells decompensate leading to a focal break between the RPE cells; a malfunction of the RPE pump mechanism occurs, and fluid accumulates in the space between the RPE and the sensory retina.
- Fluid accumulating between Bruch’s membrane and the RPE results in an RPE detachment which has a yellow-orange appearance and is more solid-looking and thicker than the serous neurosensory detachment.
Workup
- Amsler grid
- Optical Coherence Tomography (OCT)
- Intravenous fluorescein angiography (IVFA)
  - With CSR, a small, hyperfluorescent, diffuse spot appears in the choroidal phase and spreads out like an ink blot or spreads upward in a column resembling a smoke stack.
  - With an RPE detachment, there is an early, even leakage with sharp borders. The leakage remains localized, increases in intensity, but does not increase in size.
  - Rule out other causes of exudative retinal detachment, e.g. choroidal melanoma, metastasis to the choroid, renal disease acute hypertension, choroidal hemangioma, renal disease, chorio-retinal inflammations.

Treatment of CSR
- CSR usually resolves in 3-4 months; 90% of patients recover VA to 20/20; 30% have recurrences.
- Discontinue all steroid medication
- Diamox in a tapering dose
- Low-dose photodynamic therapy (PDT)
- Laser photocoagulation of RPE leakage sites that have been identified via IVFA
- Intavitreal anti-VEGF injections