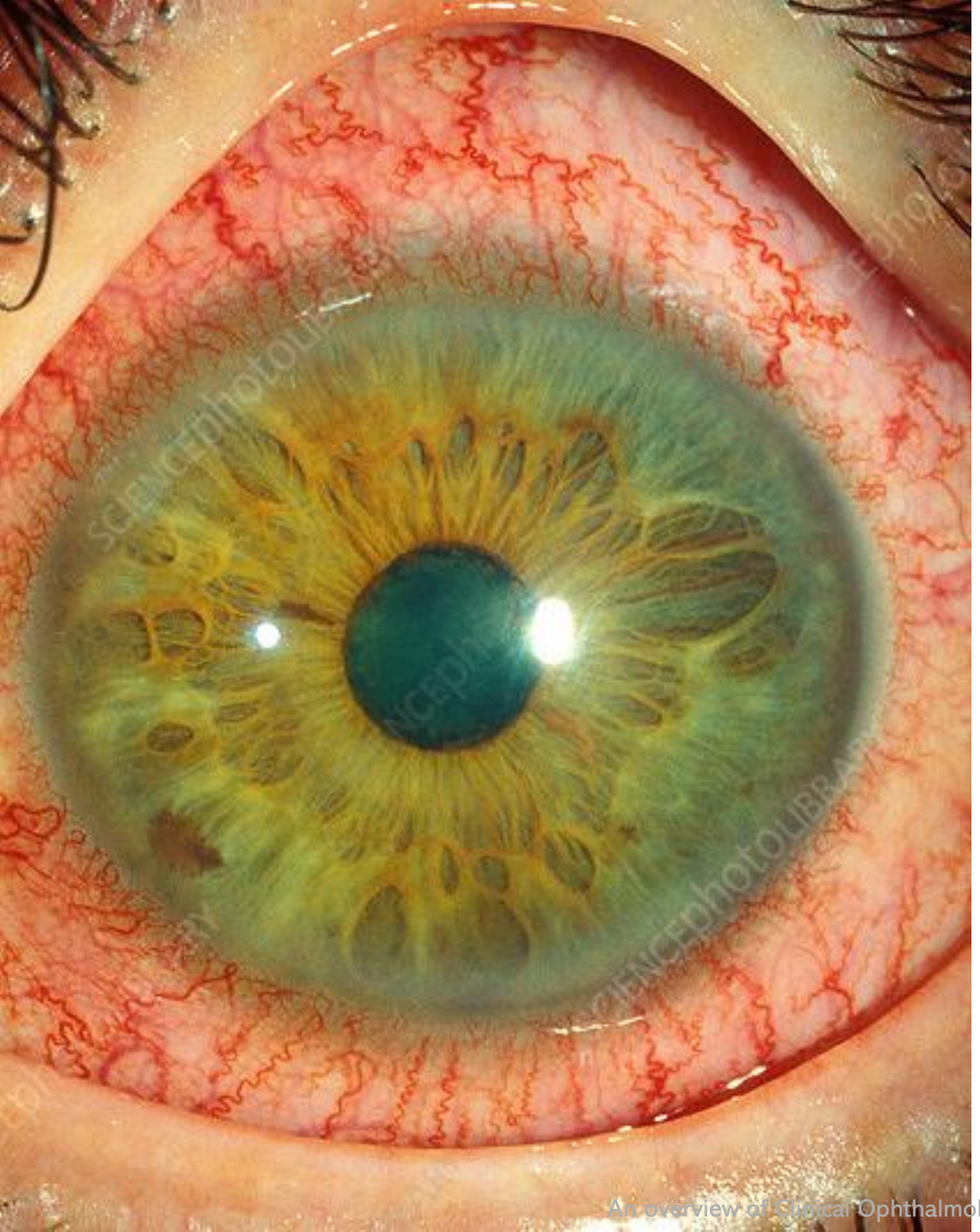
An overview of Clinical Ophthalmology for Family Doctors

Part 2

Agenda

After discussing external eye and orbital pathologies, today we shift focus to pathologies affecting structures inside the eye and the visual pathway.

- Anterior uveitis
- Acute angle closure
- Glaucoma
- Cataract
- Diabetic retinopathy
- Age related macular degeneration
- Retinal vascular disease
- Vitreoretinal disease – PVD, Retinal Tears, Retinal Detachment
- GCA
- Optic neuritis
- Diplopia



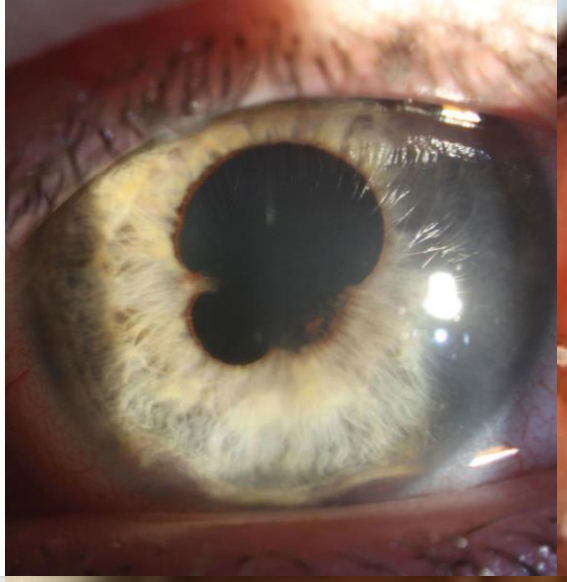
Acute Anterior Uveitis

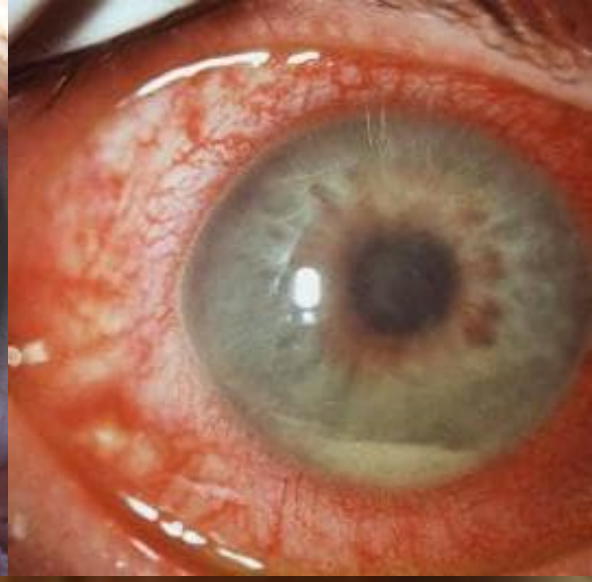
- Most common presentation for uveitis is acute anterior uveitis or AAU. Anterior means that the front portion of the uvea, the iris and ciliary body, are primarily affected by the inflammation.
- The symptoms of acute anterior uveitis are pain, redness, epiphora and photophobia, that typically develop rapidly, over a few days. Blurred vision might also be associated.
- Risk factors: HLA-B27 allele, ankylosing spondylitis, psoriatic arthritis, certain medications (bisphosphonates, sulphonamides, topical agents)

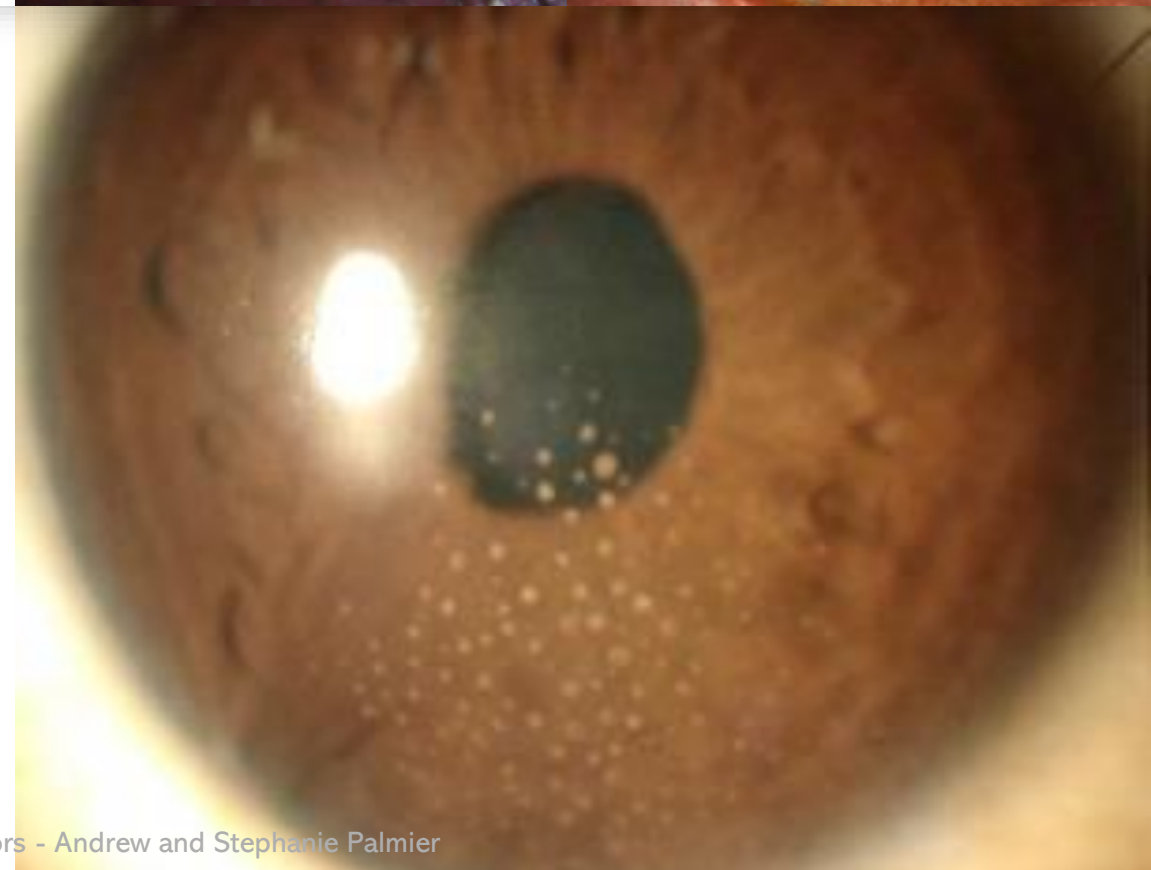
Anterior uveitis - Aetiology

Infectious	Inflammatory	Malignancy	Other
•Syphilis •Tuberculosis •HSV •CMV •Toxoplasmosis •Rubella •VZV	•HLA-B27 Associated •Inflammatory Bowel Disease •Psoriatic arthritis •JIA •Sarcoidosis •Tubulointerstitial nephritis and uveitis •Post-infectious	•Lymphoma •Retinoblastoma	•Idiopathic •Medication-induced

Anterior uveitis – Clinical Features







The slit lamp is the most important tool that is used in distinguishing the signs of AAU.

- Anterior chamber cell
- Keratic Precipitates
- Flare
- Hypopyon
- Iris nodules
- Posterior synechiae
- Pupillary miosis

Anterior uveitis – treatment and other considerations

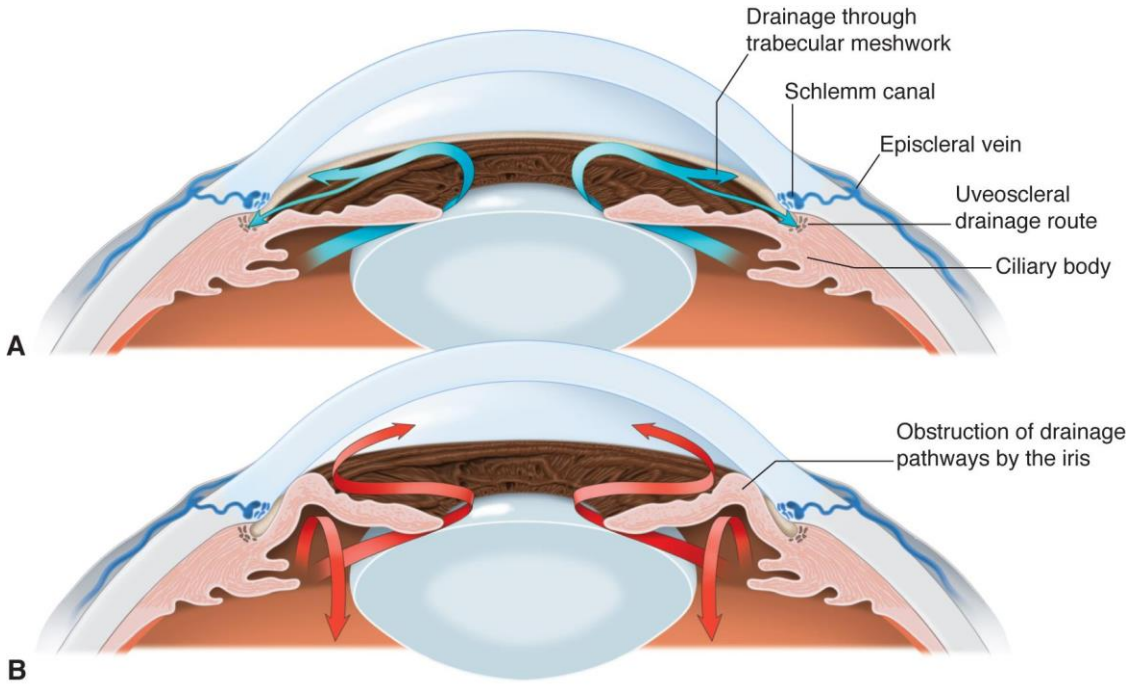
- The mainstay of therapy for AAU is topical steroid drops and dilating drops such as cyclopentolate. The steroid drop treats the underlying inflammation. The dilating drop reduces pain and helps to prevent formation of posterior synaechiae. Some forms of AAU are associated with an infection such as herpes and will also require therapy directed at the known infectious cause. Occasionally periocular or systemic corticosteroids and systemic immunosuppression are considered.
- AAU may have complications such as
  - rise in intraocular pressure, which may lead to glaucoma
  - cystoid macular oedema (CMO)
  - cataract either from inflammation or corticosteroids.

Primary angle closure – some basic principles

Aqueous humour flow.

A, Normal flow of aqueous humour from the posterior chamber, through the pupil, and into the anterior chamber. Aqueous humour exits the eye through 2 pathways in the iridocorneal angle: the trabecular meshwork and the uveoscleral pathway.

B, In primary angle closure due to pupillary block, the flow of aqueous through the pupil is obstructed, resulting in a positive pressure gradient between the posterior and anterior chambers, anterior displacement of the peripheral iris, and closure of the anterior chamber angle.



Acute angle closure

- Acute angle closure is an urgent and dramatic symptomatic event with blurring of vision, painful red eye, headache, nausea, and vomiting.
- Diagnosis is made by noting high intraocular pressure (IOP), corneal oedema, shallow anterior chamber, and a closed angle on gonioscopy. Medical or surgical therapy is directed at widening the angle and preventing further angle closure. If glaucoma has developed, it is treated with therapies to lower IOP.
- Angle closure can be primary, secondary to another eye disease, or drug induced.

Medications with potential to cause acute angle closure

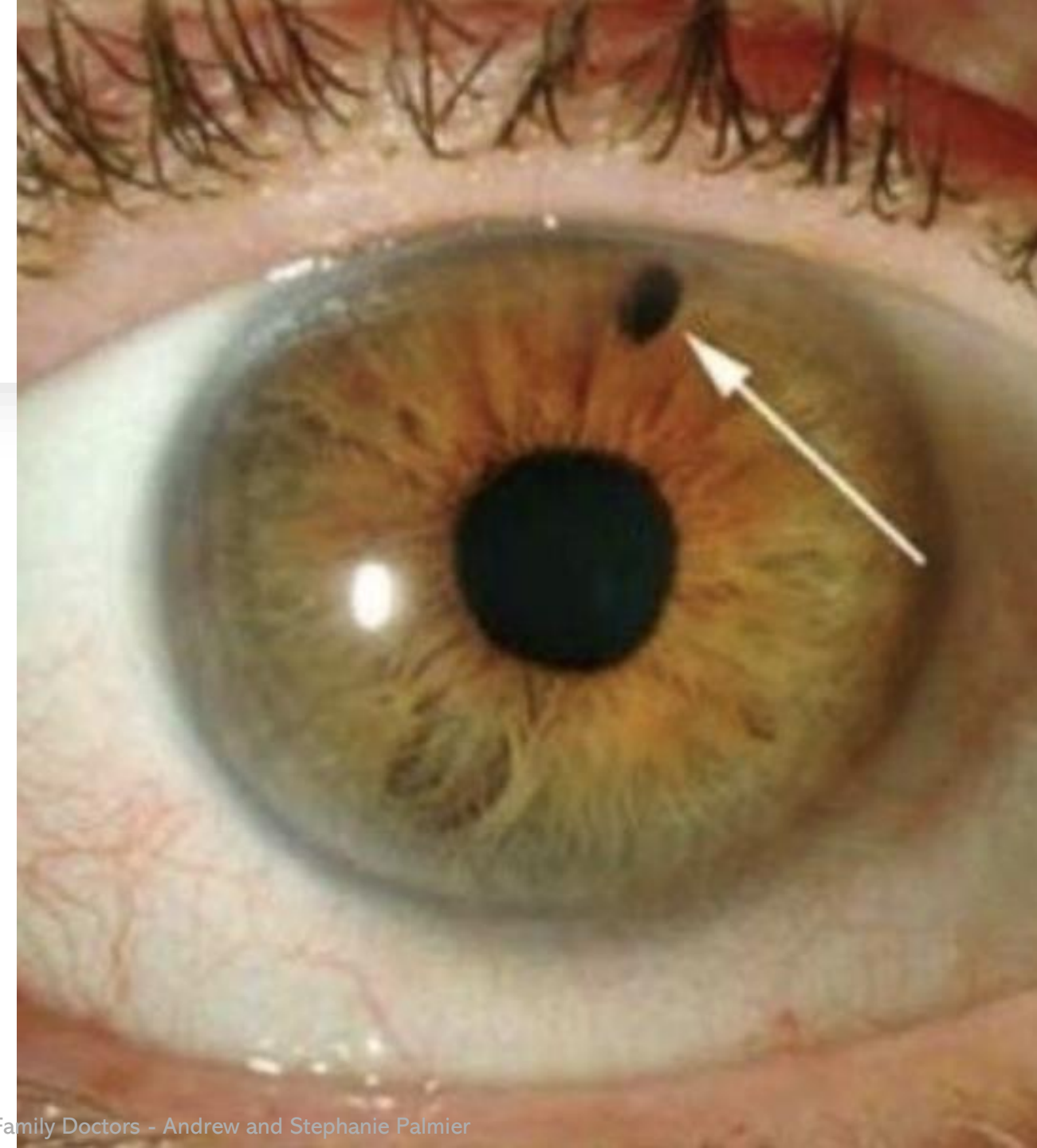
CLASSES	MEDICATIONS	COMMON USES	MECHANISM OF ANGLE CLOSURE
Ophthalmics	Mydriatics (e.g., tropicamide, atropine, phenylephrine)	Pupillary dilation	Pupillary block
	Miotics (e.g., pilocarpine)	Open-angle glaucoma; acute primary angle-closure glaucoma (low dose); pupillary miosis (Adie's syndrome); plateau iris	Forward shift of the lens-iris diaphragm; pupillary block
Psychiatric	SSRIs (e.g., escitalopram, fluoxetine, paroxetine)	Antidepressant; anxiolytic; other psychiatric conditions	Pupillary block
	SNRIs (e.g., venlafaxine)	Neuropathic pain	
	TCAs (e.g., imipramine)		
Neurologic	Topiramate	Antiepileptic; migraine	Lens-iris diaphragm displacement (ciliochoroidal effusion)
Respiratory	OTC cold medications (i.e., containing pseudoephedrine, phenylephrine)	Nasal decongestants	Pupillary block
	Oseltamivir (Tamiflu)	Influenza	Lens-iris diaphragm displacement (ciliochoroidal effusion)
	Nebulized respiratory medications (e.g., albuterol with ipratropium)	Chronic obstructive pulmonary disease; asthma	Pupillary block
Sulfa drugs	Sulfamethoxazole, acetazolamide, glipizide, topiramate, hydrocholorothiazide	Antibiotics; diuretics; insulin secretagogues; antiepileptic; migraine	Lens-iris diaphragm displacement (ciliochoroidal effusion)

Key Factors for Acute Angle-Closure Glaucoma

Presence of risk factors (eg, hyperopia, thick cataractous lens)	Halos around lights	Aching eye or brow pain	Headache
Nausea, vomiting	Reduced acuity	Eye redness	Closed angle on gonioscopy
Extremely elevated IOP	Corneal oedema	Engorged conjunctival vessels	Fixed dilated pupil

Management of an acute episode

- The immediate goal of treatment is to relieve the acute symptoms and decrease IOP, which is usually achieved with medical therapy.
- Oral or topical carbonic anhydrase inhibitors, topical beta-blockers, or alpha-2 adrenergic agents may be used as first-line therapies either alone or more typically in combination. Cholinergics such as pilocarpine are considered according to case.
- It is recommended to position the patient supine, mechanically helping to relieve the angle closure.
- Following resolution of the acute attack, definitive surgical treatment should be performed within 24 to 48 hours with the aim of achieving a persistently open angle; Laser Peripheral Iridotomy



Primary open angle glaucoma

- Glaucoma describes a group of conditions in which there is characteristic cupping of the optic disc with corresponding visual field defects, due to retinal ganglion cell loss. It is a progressive condition and is the most common cause of irreversible blindness worldwide.
- Primary open angle glaucoma (POAG) is a subset of the glaucomas defined by an open, normal appearing anterior chamber angle and raised intraocular pressure (IOP), with no other underlying disease. If there is an identifiable underlying cause for raised IOP, this is termed secondary glaucoma. If the IOP is within normal limits, this is termed normal tension glaucoma (NTG).

Risk Factors for POAG

IOP: only known modifiable risk factor. Higher IOP is associated with a higher risk of developing POAG.

Age: The prevalence of POAG increases with age

Race: more prevalent in people of African-Caribbean descent compared with Caucasians.

Refractive error: Myopia has been shown to be a risk factor

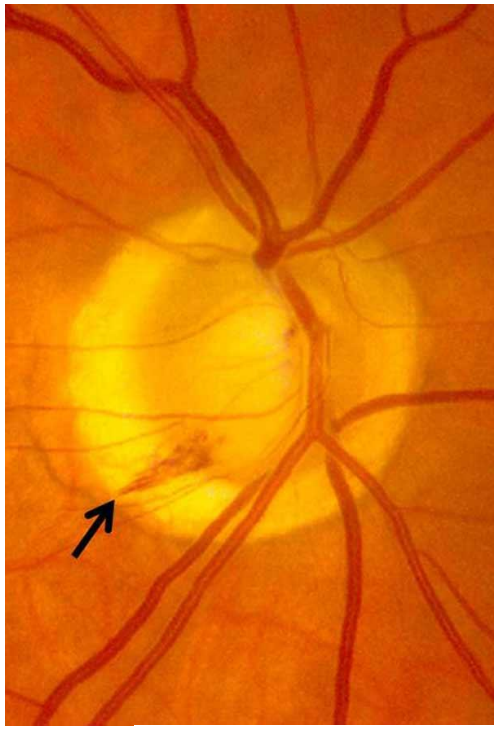
Central corneal thickness: A thinner cornea has been shown to be a risk factor for OHT patients developing POAG.

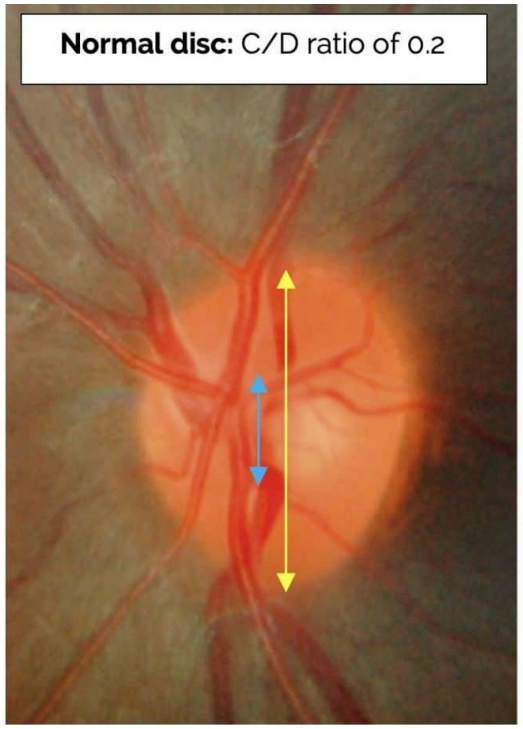
Family history: A first-degree relative with POAG is a risk factor for the development of POAG, higher still if the affected relative is a sibling.

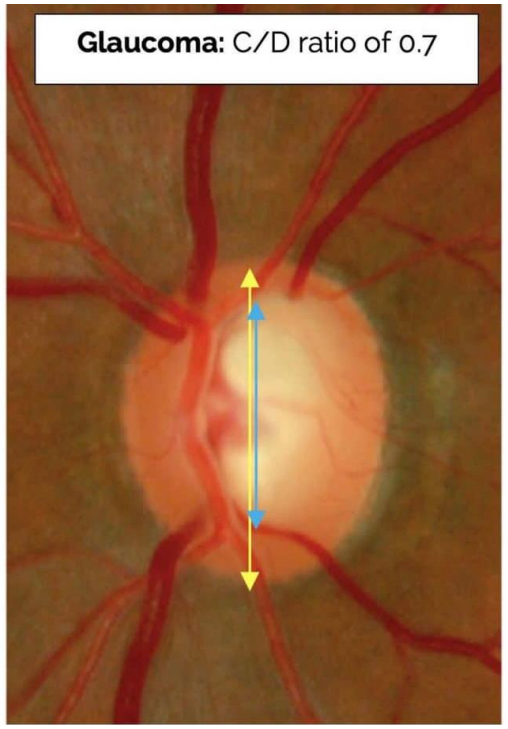
Diagnosing POAG

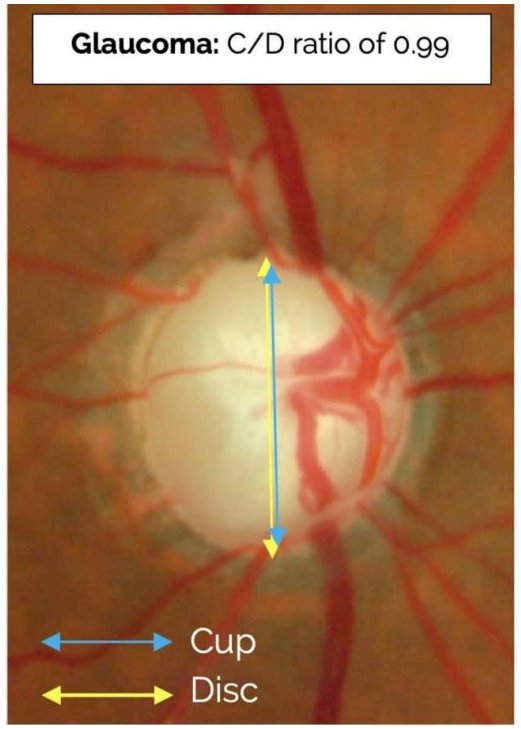
Diagnosis of POAG requires assessment of :

1. Intraocular pressure
2. Open- normal appearing anterior chamber angle
3. Characteristics signs of optic disc damage
4. Visual function loss on perimetry







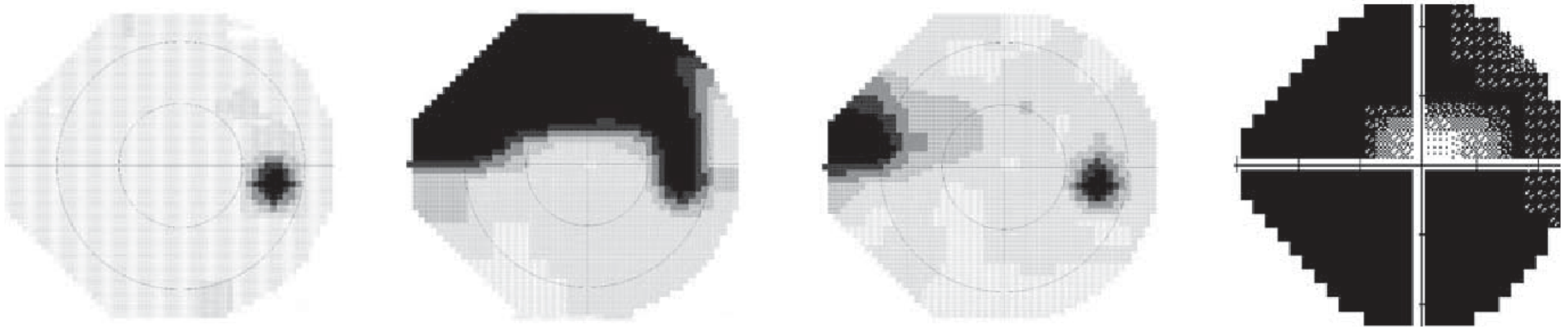


A Normal visual field

B Superior arcuate defect

C Superior nasal step defect

D End-stage glaucomatous visual field defects



All images are right eyes. Note that visual fields are from perspective of patient, with lighter areas representing higher sensitivity and darker areas representing reduced sensitivity. Blindspot is located temporally corresponding to nasal location of optic nerve.

Treatment

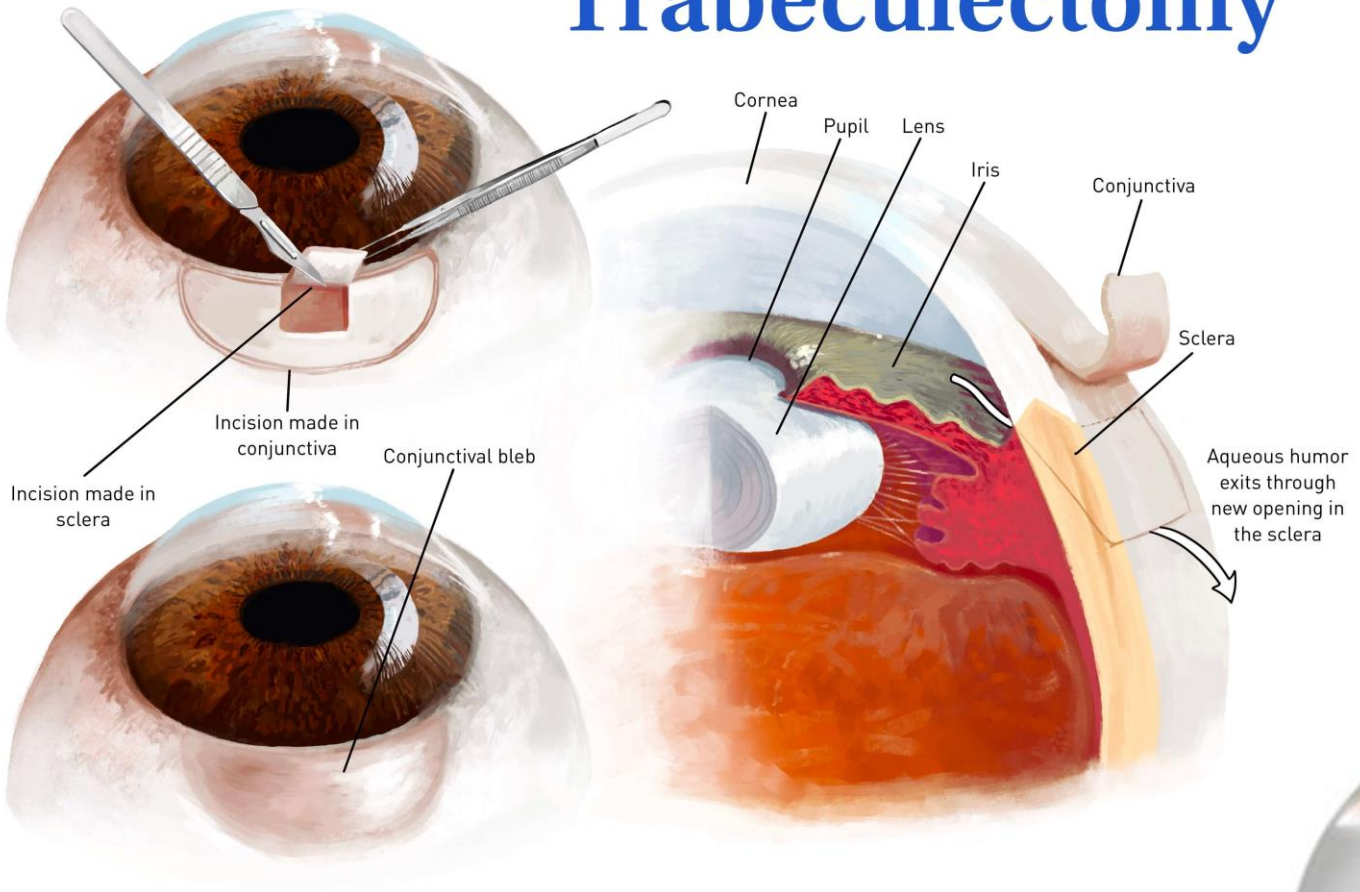
- Medical therapy:

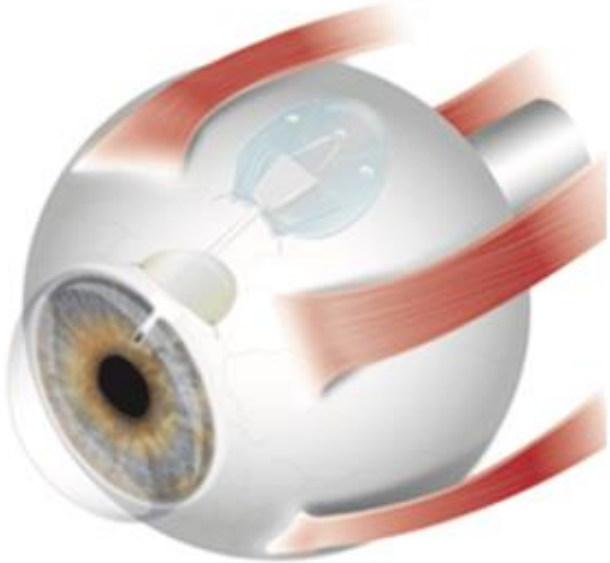
Prostaglandins (latanoprost, bimatoprost, travoprost, etc.)
Beta-blockers (timolol, betaxolol, carteolol, levobunolol,etc.)
Carbonic anhydrase inhibitors (brinzolamide, dorzolamide)
Alpha-agonists (apraclonidine, brimonidine)
Miotics (pilocarpine,etc.)
Oral Acetazolamide

- Surgical therapy:

Laser trabeculoplasty (SLT)
Trabeculectomy +/- augmentation
Non-penetrating drainage surgery
Shunt procedures (see section Glaucoma Drainage Implant Surgery)
Cyclodestructive procedures (cyclodiode)

Trabeculectomy





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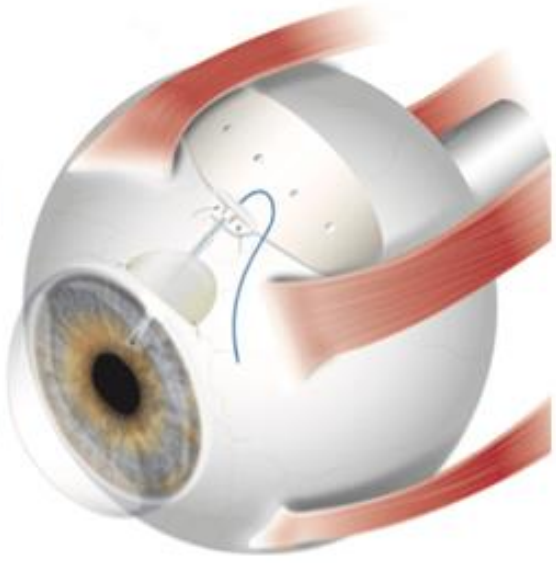

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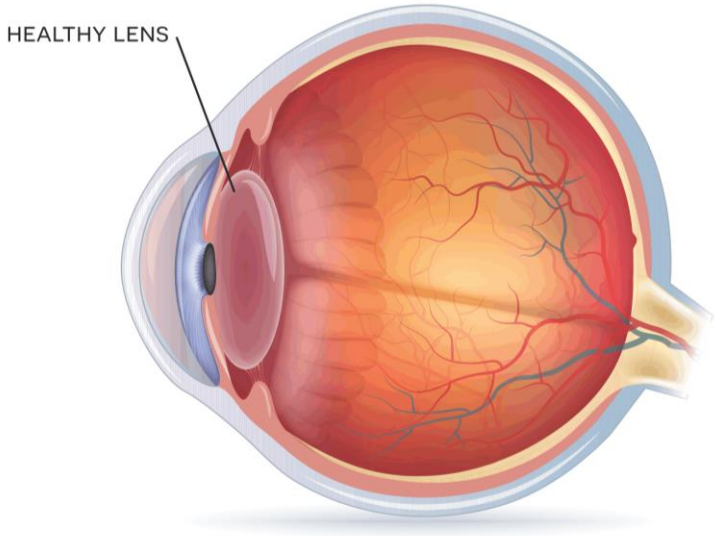
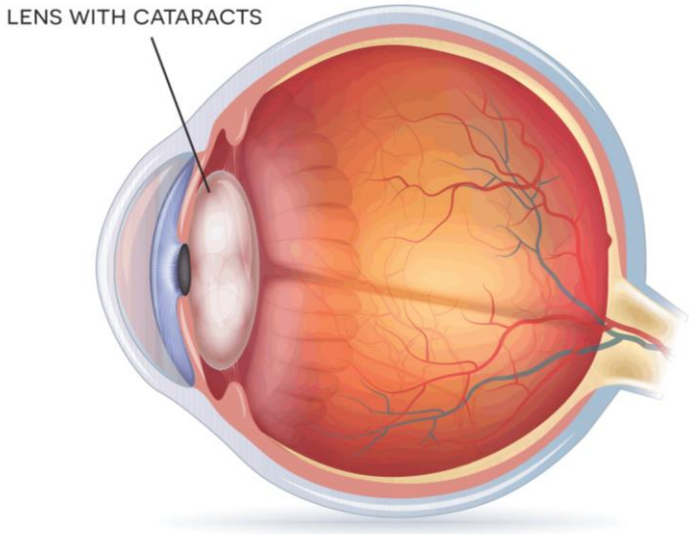
Cataract

- A cataract is a clouding of the natural intraocular crystalline lens that focuses the light entering the eye onto the retina. This cloudiness can cause a decrease in vision and may lead to eventual blindness if left untreated.
- Cataracts often develop slowly and painlessly, so vision and lifestyle can be affected without a person realizing it.
- Worldwide, cataracts are the number one cause of preventable blindness.



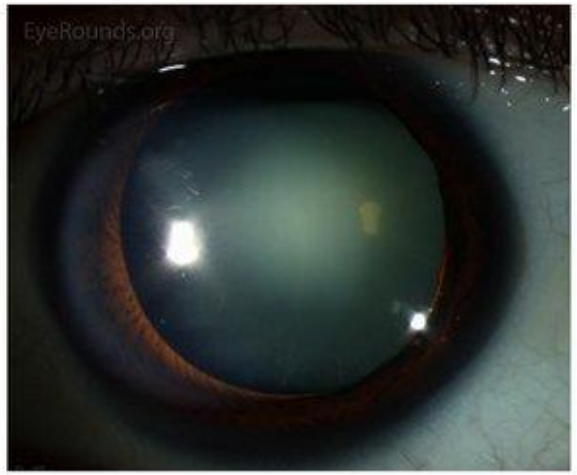


Cataract

- While most cataracts in the population are age-related, there are many types and causes of cataract.
- Cataracts can also occur in children, and may be classified according to the age of onset (congenital or infantile/juvenile cataracts)
- Cataracts may be secondary to hereditary factors, trauma, inflammation, metabolic or nutritional disorders, and exposure to radiation
- Lifestyle factors such as tobacco smoking and high alcohol intake are associated with an increased risk of developing age-related cataracts

Types of Age-Related Cataracts

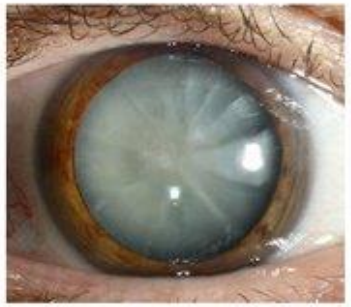
Nuclear Sclerosing Cataract



Associated with statins and miotics.

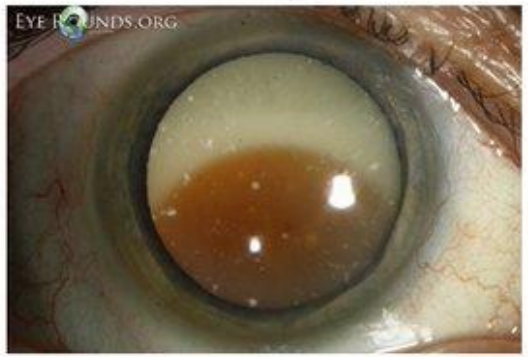
Cortical Cataract

Early Vacuoles



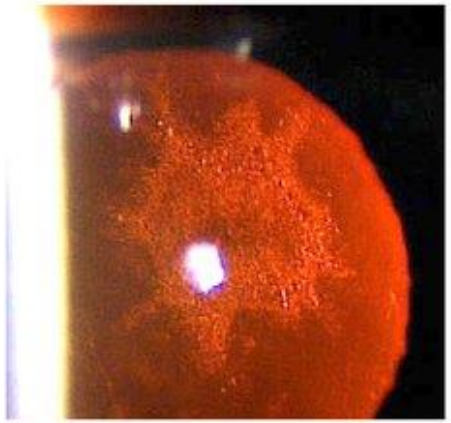
Cortical Spokes

A Morgnagnian Cataract forms when the lens nucleus sinks in liquified cortex. Cortical cataracts are associated with UV and IR radiation.

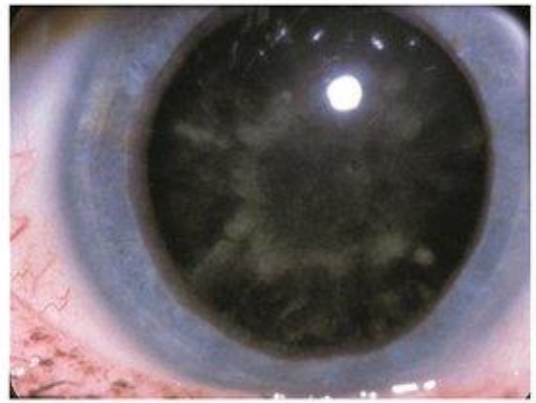


Morgnagnian Cataract

Posterior Subcapsular Cataract



Associated with steroid use



Diabetes accelerates formation and can result in a “snowflake” cataract.

@EyeFlyMD

Symptoms

- Blurred vision at distance or near (different types may affect distance greater than near or vice versa, see below)
- Glare (halos or streaks around lights, difficulty seeing in the presence of bright lights)
- Difficulty seeing in low light situations (including poor night vision)
- Loss of contrast sensitivity
- Loss of ability to discern colours
- Increasing near-sightedness or change in refractive status (including "second sight" phenomenon)

Ophthalmological referral for surgery

When patients are evaluated for cataracts, the primary objective is to determine the following:

1. Is there is a visually significant lens opacity;
2. Does the lens opacity account for the patient's level of vision;
3. Would removal of the cataract likely lead to improved vision and improved level of functioning and is the potential improvement enough to warrant the risks of surgery;
4. Would the patient tolerate the operation and be able to follow postoperative instructions and follow up care.

If the answers to these questions lead the patient and physician to agree that surgical intervention is warranted, preoperative planning must be done.

Pre-operative planning

- Measuring the visual acuity with and without spectacle correction. In patients complaining of glare, brightness acuity can be tested by asking a patient to read the eye chart while shining a bright light at the patient from the side.
- A comprehensive dilated eye exam is performed on all patients.
- Specific attention is paid to several factors impacting surgical planning including the severity of the cataract, the size of the dilated pupil (smaller pupils increase the complication rate), the clarity, thickness and health of the cornea, stability of the lens, depth of the anterior chamber, and health of the optic nerve and retina.
- Ocular biometry: To determine the IOL power needed, measurements of the axial length of the eye, the corneal refractive power, and the anterior chamber depth are taken.

Nonsurgical Treatment

No medical treatment has been shown to be effective in the treatment or prevention of cataracts, although this is an active area of research.

Lifestyle advice: To slow the development of cataracts it is generally recommended that patients eat a balanced diet, prevent excessive exposure to UV radiation by using good quality UV blocking sunglasses, avoiding injuries by using protective eyewear, and if diabetic closely control blood sugar levels.

Surgical treatment

Cataract surgery is one of the most common surgical procedures performed around the world and has a very high success rate. The most common type of cataract surgery utilizes ultrasound energy to break the cataract into particles small enough to aspirate through a handpiece. This technique is referred to as phacoemulsification.

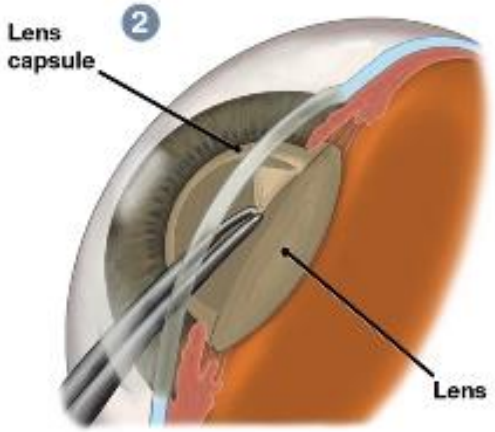
Phacoemulsification and IOL Placement

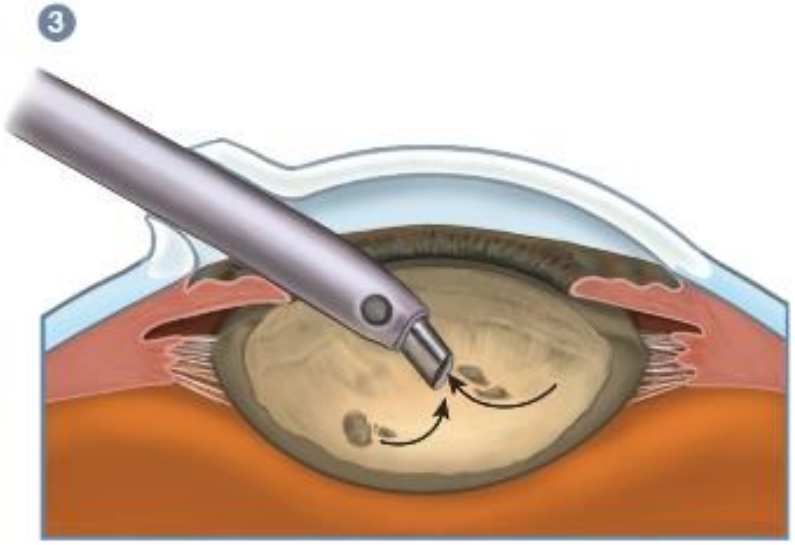
Normal lens without cataract

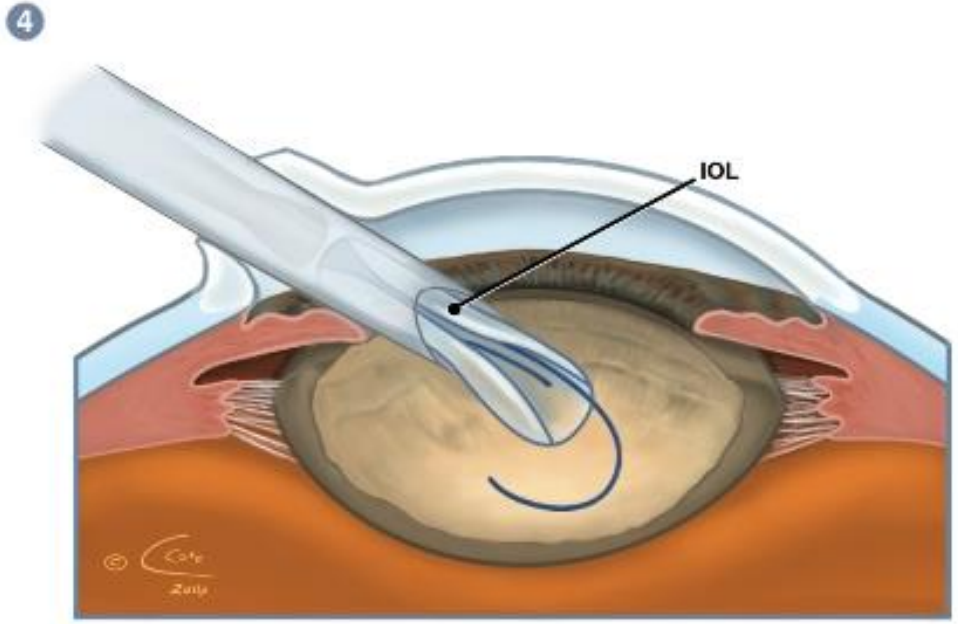
Lens with cataract

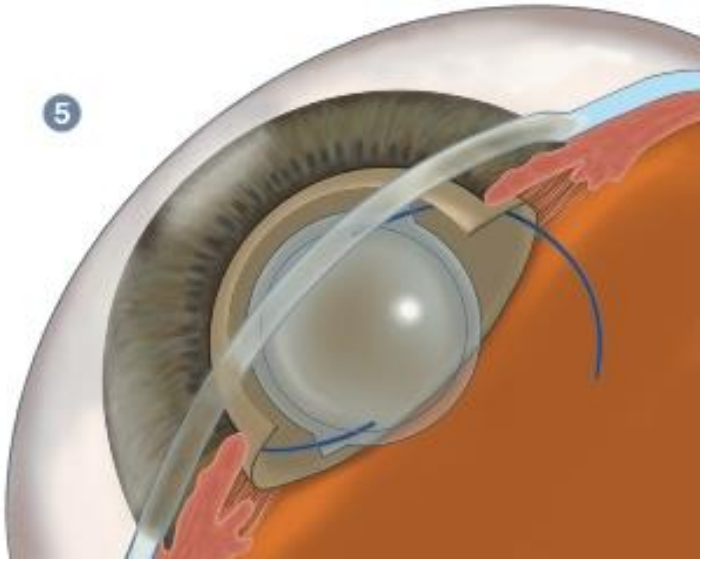
When a lens becomes cloudy, it needs to be extracted and replaced

Figure 1: Side port and cataract incision
Figure 2: Capsulorrhexis (breaking the lens capsule)
Figure 3: Phacoemulsification and irrigation
Figure 4: Initial placement of IOL (intraocular lens)
Figure 5: Final placement of IOL









Risk factors for suboptimal visual outcome

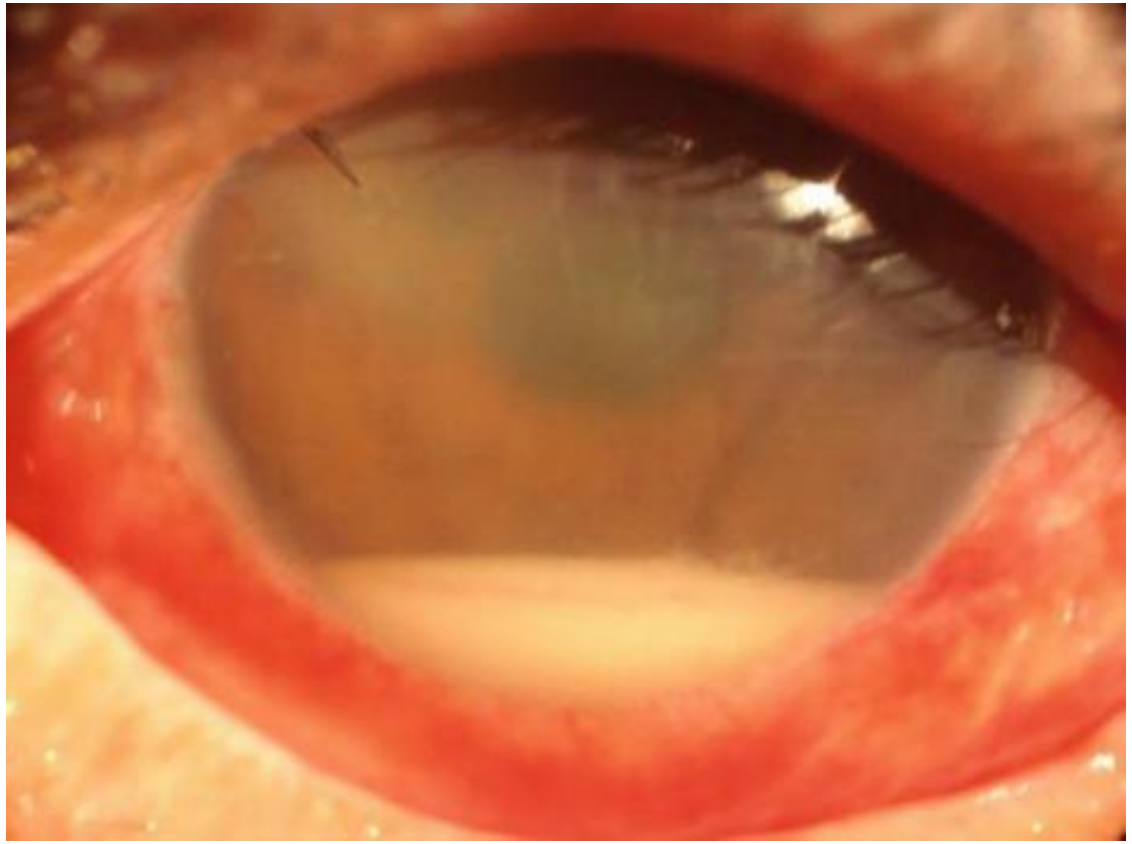
- Age-related macular degeneration
- Diabetic retinopathy
- Corneal opacity/pathology
- Older age
- Female sex
- Previous vitrectomy
- Previous retinal detachment
- Alpha blockade
- Intraoperative complications

Complications

- Posterior Capsule Rupture/Vitreous loss
- Cystoid Macular oedema
- Endophthalmitis
- Vitreous/Suprachoroidal haemorrhage
- Retinal Tears/Detachment
- Lens Dislocation

Acute Postoperative Endophthalmitis

- Refers to infectious endophthalmitis shortly after ocular surgery
- Most present within 1-2 weeks, usually 3-5 days after the surgery.
- Initial symptoms: rapidly progressive, including pain, red eye, ocular discharge, and worsening vision
- Common signs: decreased visual acuity, lid swelling, conjunctival and corneal oedema, anterior chamber cells + fibrin, hypopyon, vitreous inflammation, retinitis, retinal haemorrhages and blunting of red reflex
- Management: Vitreous Tap for MCS, Intravitreal and Intravenous Antibiotics, PPV for patients presenting with LP only visual acuity



Diabetic retinopathy

- Diabetic retinopathy represents microvascular end-organ damage as a result of diabetes. It ranges from non-proliferative diabetic retinopathy (NPDR) and its stages to proliferative diabetic retinopathy (PDR). As the disease progresses, associated diabetic macular oedema (DMO) may also become apparent.
- The Wisconsin Epidemiologic Study of Diabetic Retinopathy (WESDR) Cohort showed that after 20 years of diabetes mellitus, 99% of patients with type 1 and 60% of patients with type 2 show some degree of retinopathy.

Risk factors for DR

Diabetes duration

Hypertension

Dyslipidemia

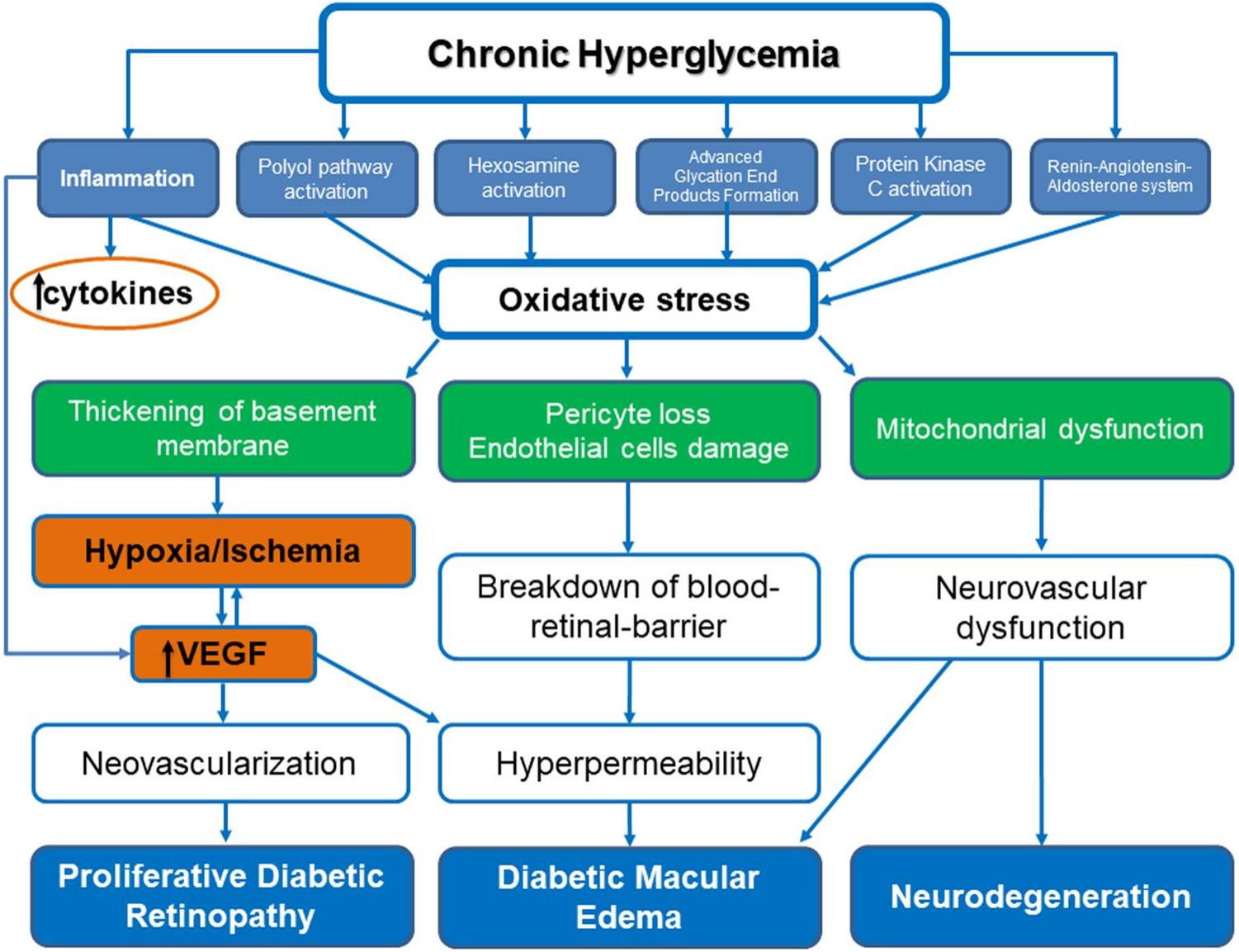
Ethnicity (South Asian, African, Latin American)

Pregnancy

Smoking

Pathophysiology

Vascular endothelial growth factor (VEGF) is secreted by the ischemic retina. VEGF leads to a) increased vascular permeability resulting in retinal swelling/oedema and b) angiogenesis or new blood vessel formation



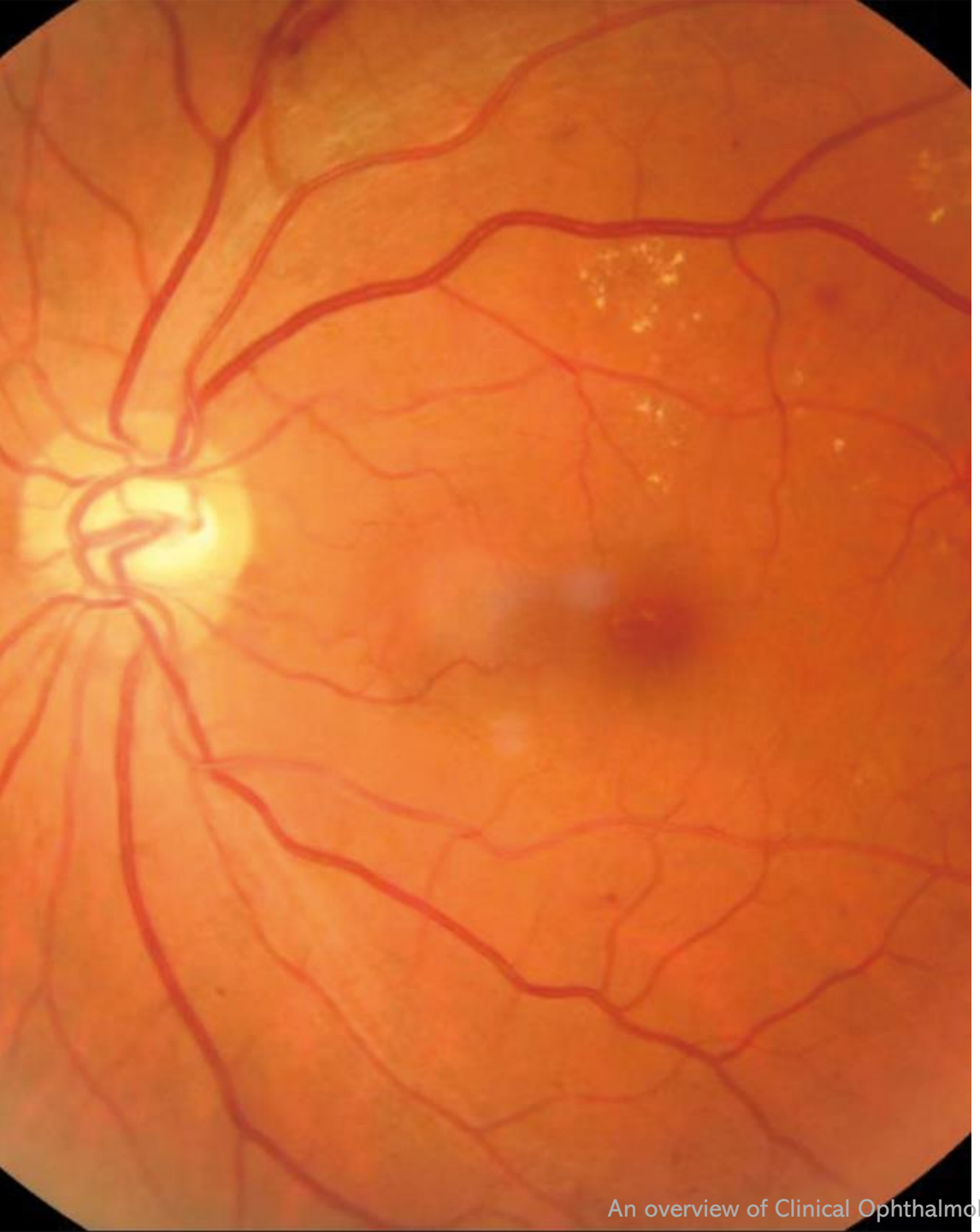
Presenting symptoms of DR

- A good proportion of patients are mostly asymptomatic and diabetic retinopathy changes are identified on fundoscopic screening.
- Patients may present with symptoms of decreased vision or fluctuating vision/metamorphopsia (lens or macular oedema), presence of floaters (vitreous haemorrhage), or visual field defects (tractional detachment).

NPDR vs PDR

The main types of diabetic retinopathy are non-proliferative diabetic retinopathy (NPDR) and proliferative diabetic retinopathy (PDR).

The distinguishing feature between these two categories is the presence (proliferative) or absence (non-proliferative) of abnormal new blood vessels (retinal, optic disc, or iris/angle neovascularization).

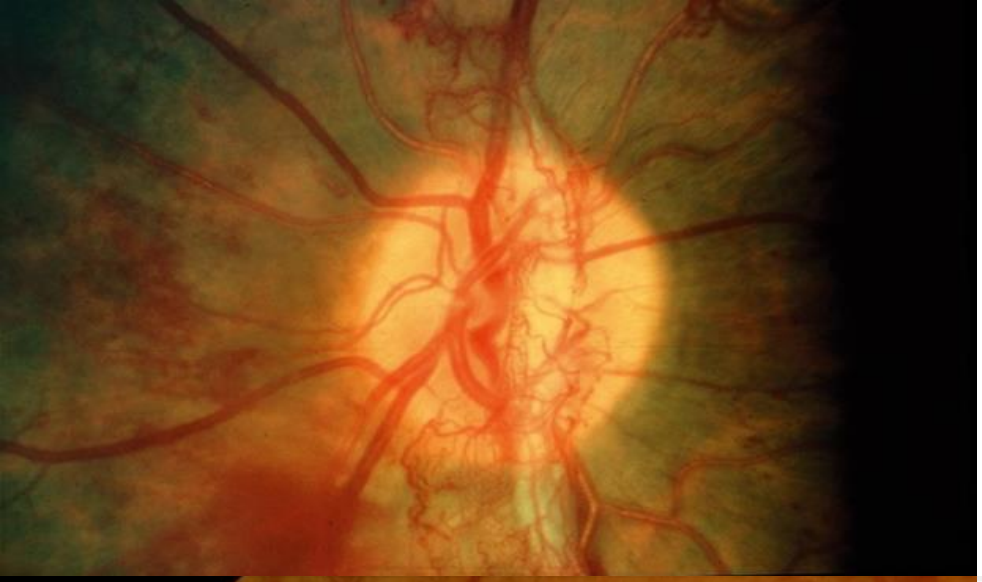


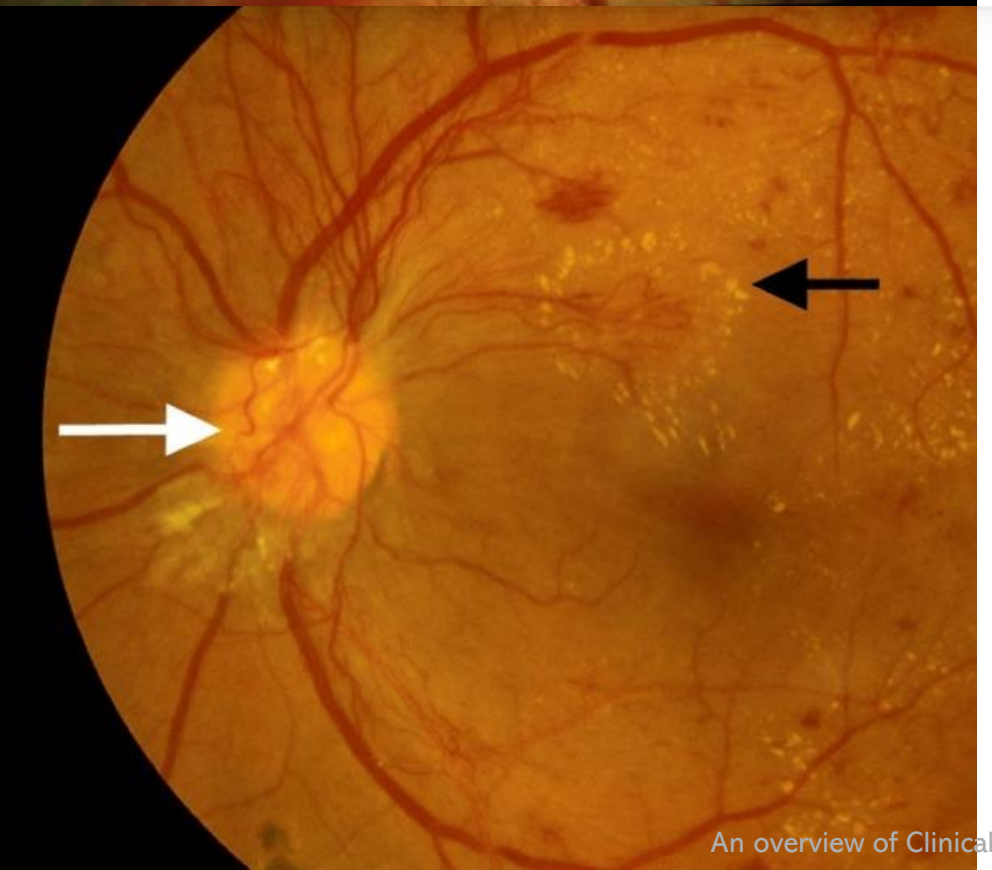
NPDR

NPDR can be classified into mild, moderate or severe stages based upon the presence or absence of retinal bleeding, abnormal beading of the venous wall (venous beading) or abnormal vascular findings (intraretinal microvascular anomalies or IRMA).

- Mild: few microaneurysms
- Moderate: increased number of microaneurysms and dot-blot haemorrhages. Cotton wool spots and hard exudates may be present.
- Severe: "4-2-1 rule" -- 4 quadrants of diffuse retinal haemorrhages and microaneurysms, 2 or more quadrants of venous beading, or 1 or more quadrant of IRMA

No treatment is usually done at this stage.





PDR

High Risk Characteristics

- NVD > 1/4 to 1/3 disc area
- Any NVD associated with vitreous or preretinal haemorrhage
- Any NVE associated with vitreous or preretinal haemorrhage

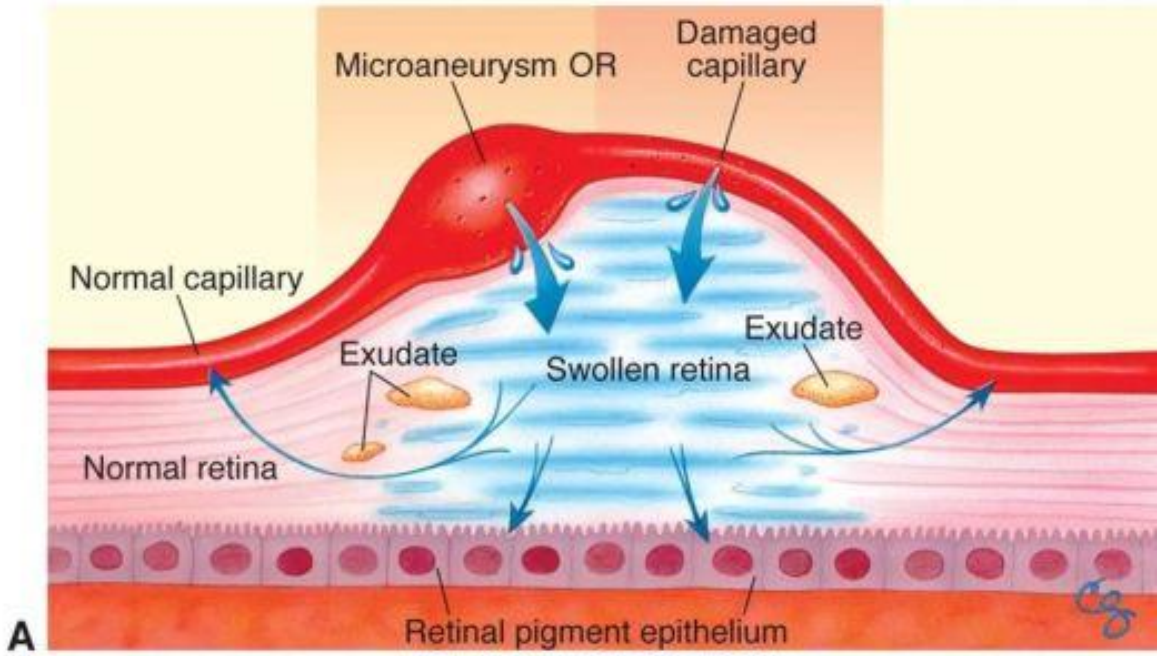
This is progressive and often requires treatment to prevent bleeding and scar tissue formation, especially in patients who meet high risk characteristics.

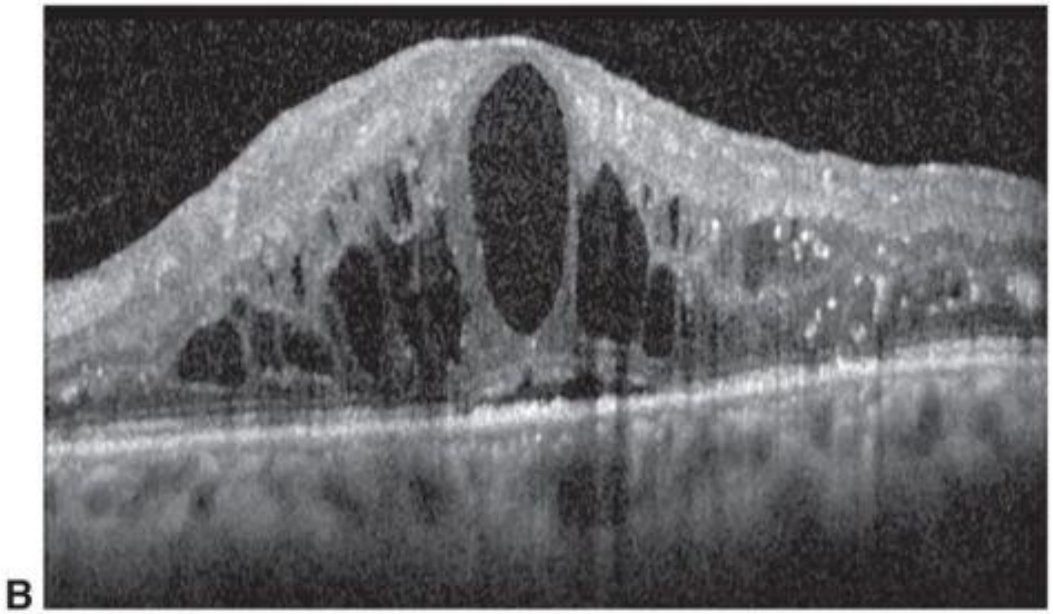
DMO

- Diabetic Macular Oedema (DMO) is suspected in patients with any level of DR who present with blurred vision or metamorphopsias.
- DMO is the accumulation of excess fluid in the extracellular space within the retina in the macular area, typically in the inner nuclear, outer plexiform, Henle's fibre layer, and subretinal space

A: Schematic diagram of DMO. Microaneurysms or damaged capillaries resulting from the breakdown of the blood-retina barrier leak fluid to the extracellular space, resulting in a swollen retina. Resorption of DMO is dependent on the adjacent capillaries and retinal pigment epithelium. Resorption of fluid may leave behind lipoprotein residues seen as exudates.

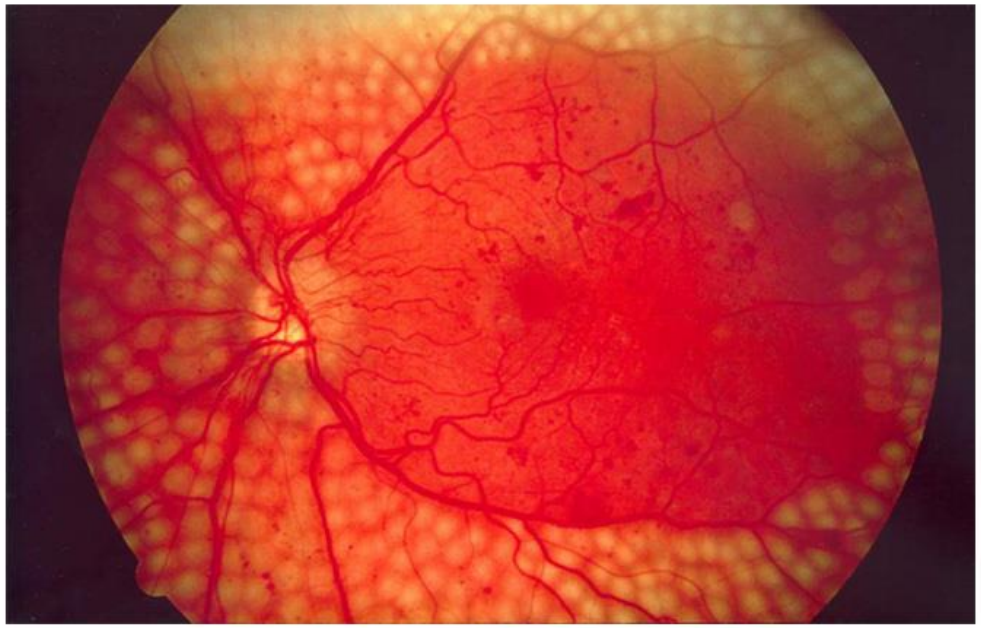
B: Optical Coherence Tomography image of DMO.



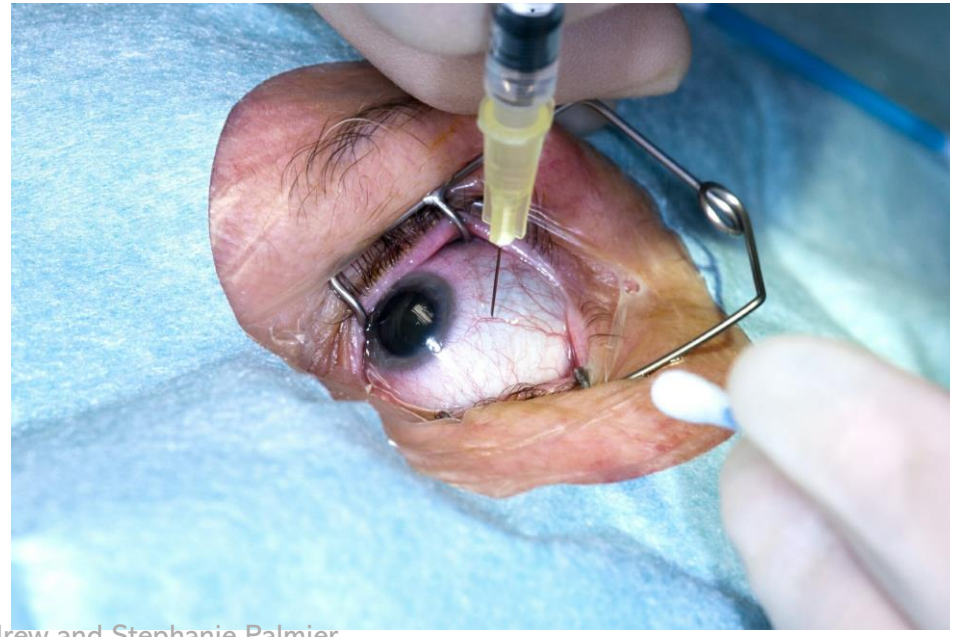


Treatment

- General Treatment: Systemic control of diabetes, hypertension, hyperlipidemia, hypercholesterolemia, nephropathy and other diseases are of paramount importance.
- Medical Treatment:
  DMO - Treatment of macular oedema is usually needed in order to prevent loss of vision or to try to improve vision. Treatment includes the use of lasers or injection of drugs (anti-VEGF therapies or corticosteroids) that decrease the retinal swelling/macular oedema
  PDR - The primary treatment option for PDR is laser photocoagulation of the peripheral retina, known as pan retinal photocoagulation (PRP). The laser is used to obliterate some of the ischemic peripheral retina to decrease VEGF release and induce regression of neovascularization.



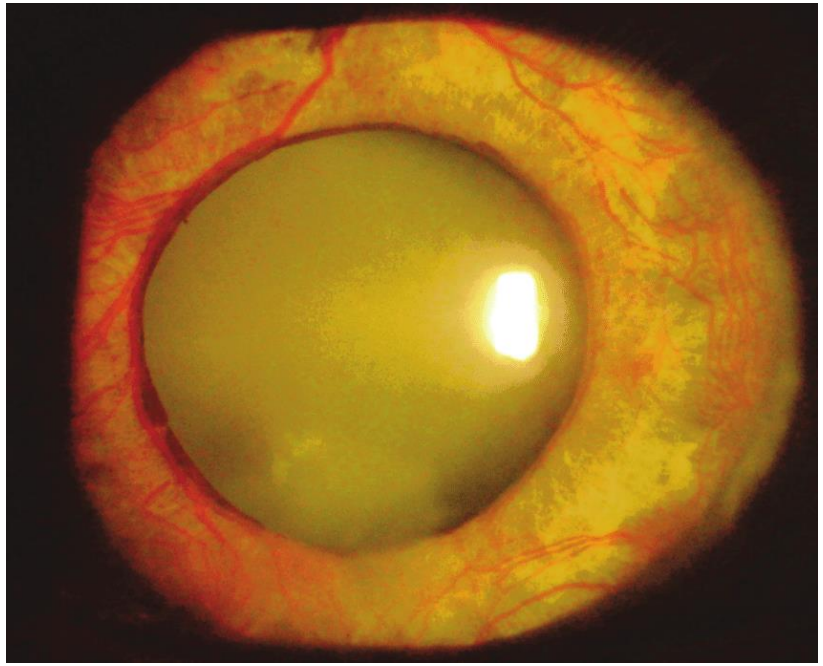
Fundus photograph after panretinal photocoagulation treatment[7]

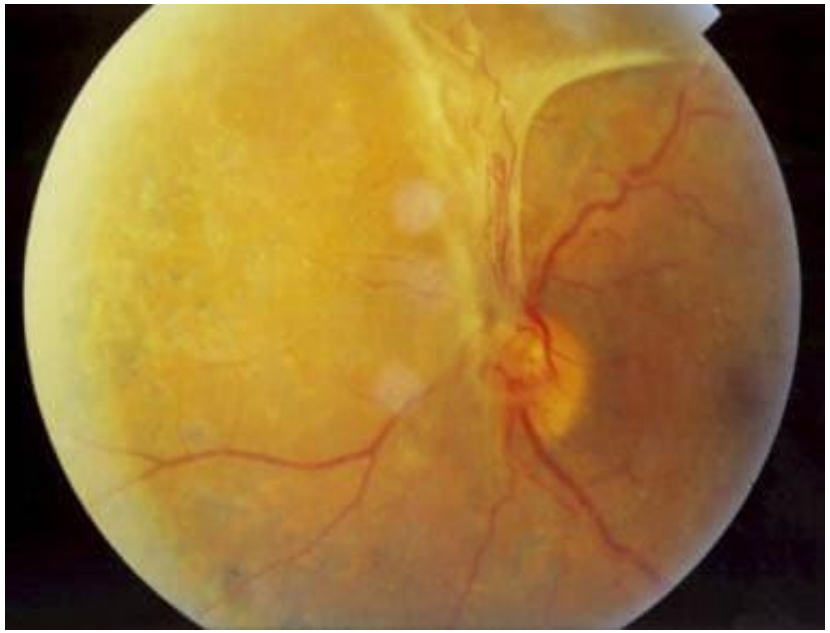


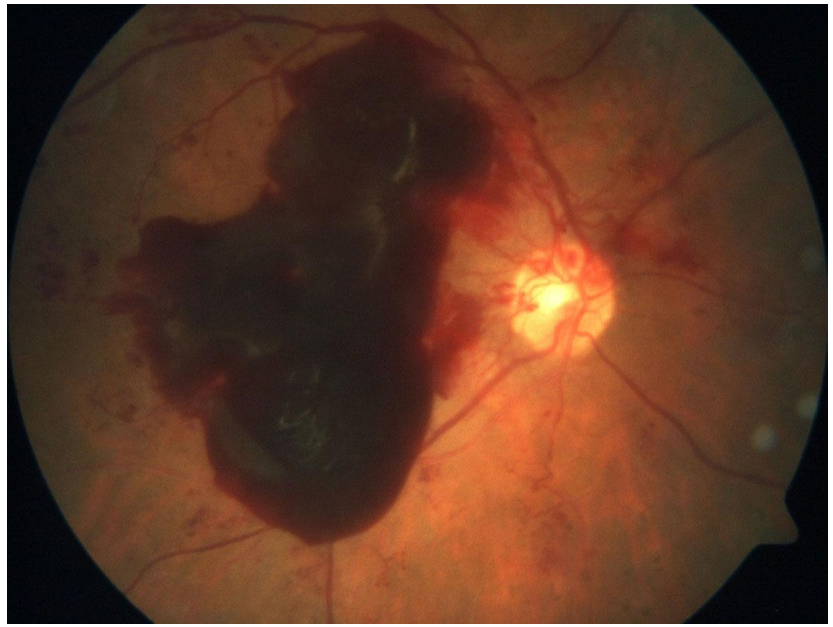
Treatment

- Surgical Treatment: Sometimes the proliferative disease is advanced and there is blood filling the eye (and preventing application of laser) or scar tissue that wrinkles the retina or pulls it off the eyewall (tractional retinal detachment). In these situations, surgery may be necessary. The goal of surgery is to remove blood and scar tissue from the retinal surface and to place laser treatment as needed.

Late stages of DR







Age related macular degeneration

- Age-related macular degeneration (ARMD) is an acquired degeneration of the retina that causes significant central visual impairment through a combination of non-neovascular (drusen and retinal pigment epithelium abnormalities), and neovascular derangement (choroidal neovascular membrane formation).
- The hallmark of age-related macular degeneration is the presence of drusen within the macula.

Risk Factors for ARMD

Age: ARMD risk increases with age.

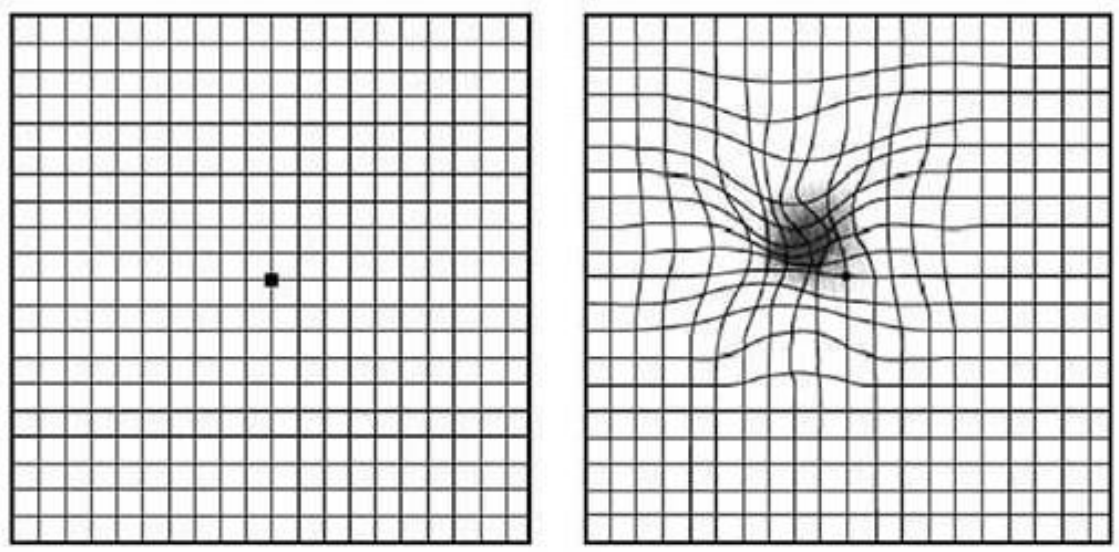
Cigarette Smoking: A ten pack-year tobacco smoking history is associated with increased development of exudative age-related macular degeneration

Genetic susceptibility

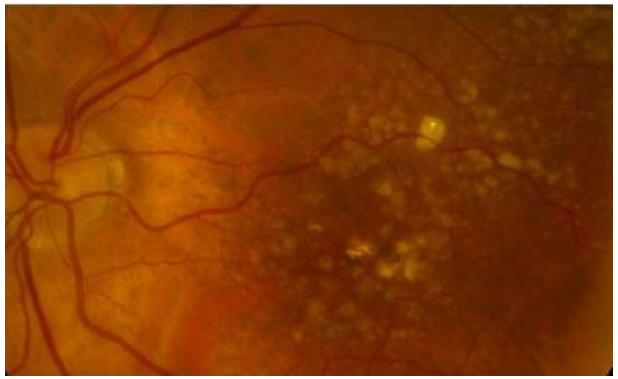
Others: Cardiovascular disease, Hypertension, Female gender, Caucasians, Hypercholesterolemia, Obesity, Hyperopia, Family History

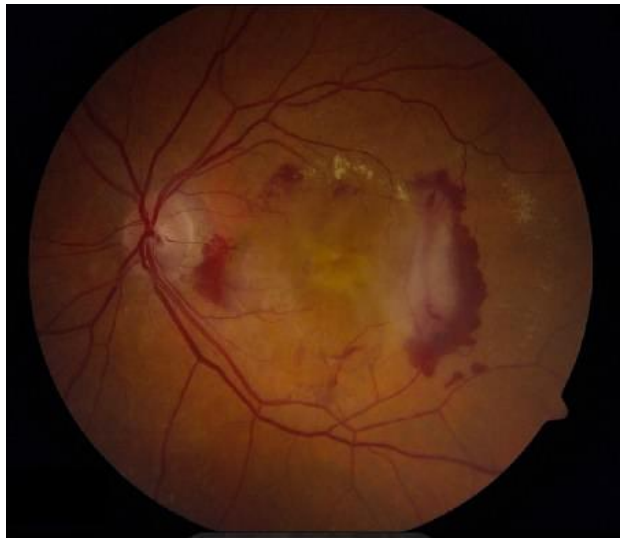
Clinical presentation

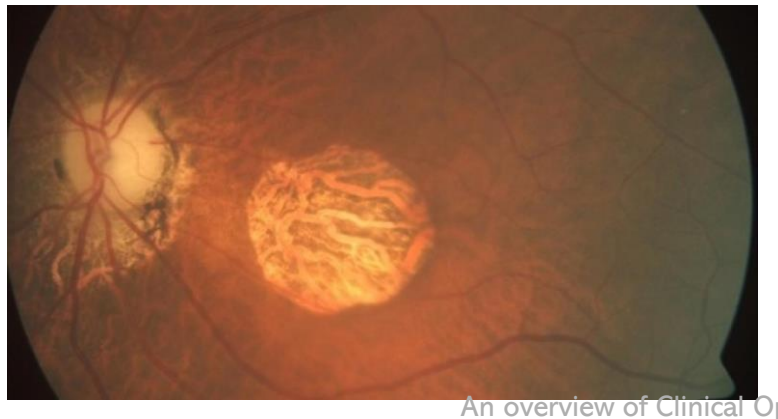
- History: Decreased central visual acuity may not be present in ARMD, especially in early stages, and is not a reliable indicator of disease severity. Exudative ARMD changes may be associated with acute to subacute drops in vision, or perception of visual distortion such as metamorphopsia.
- Physical examination: Periodic dilated fundus exams are warranted to identify patients who progress to neovascular ARMD without having symptoms. Importance of the use of an Amsler grid.
- Signs: Drusen, Geographic atrophy, Subretinal fibrosis, RPE changes, Subretinal fluid or haemorrhage/hard exudate



Stages of ARMD







- In early disease, the macula shows yellowish-coloured subretinal deposits called “drusen” and/or increased pigment. Drusen are thought to be byproducts of retinal pigment epithelium dysfunction. (dry ARMD)
- Late disease (atrophic and exudative) can lead to significant loss of vision. Exudative disease occurs in only 10 percent of patients with age-related macular degeneration, but it is responsible for 80 to 90 percent of cases of severe vision loss related to the disease. (wet ARMD)

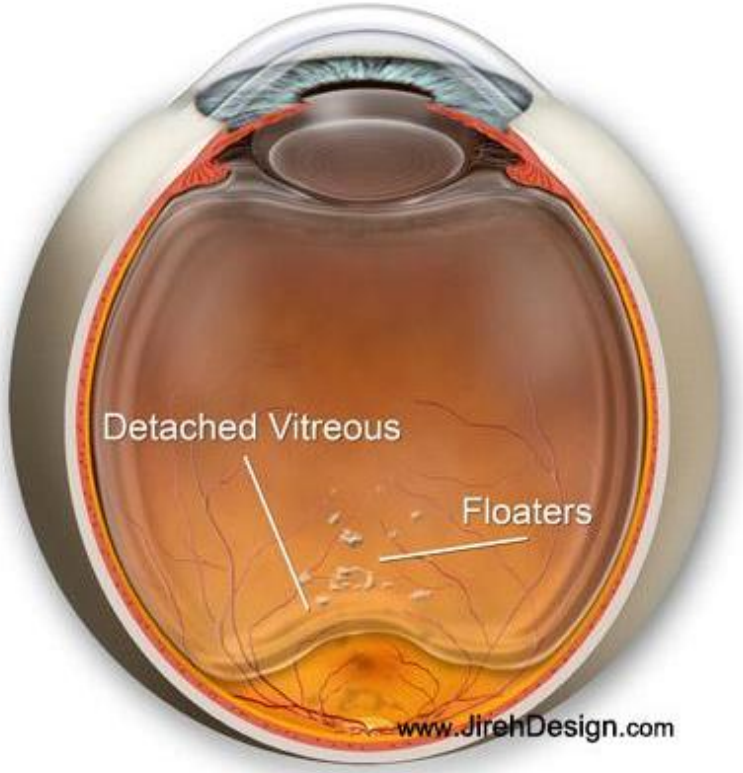
Treatment

- Medical therapy: Antioxidant and mineral supplementation
- Treatment for neovascular ARMD: anti-VEGF injections

Treatments targeting biochemical pathways implicated in the pathophysiology of ARMD are being tested actively in clinical trials.

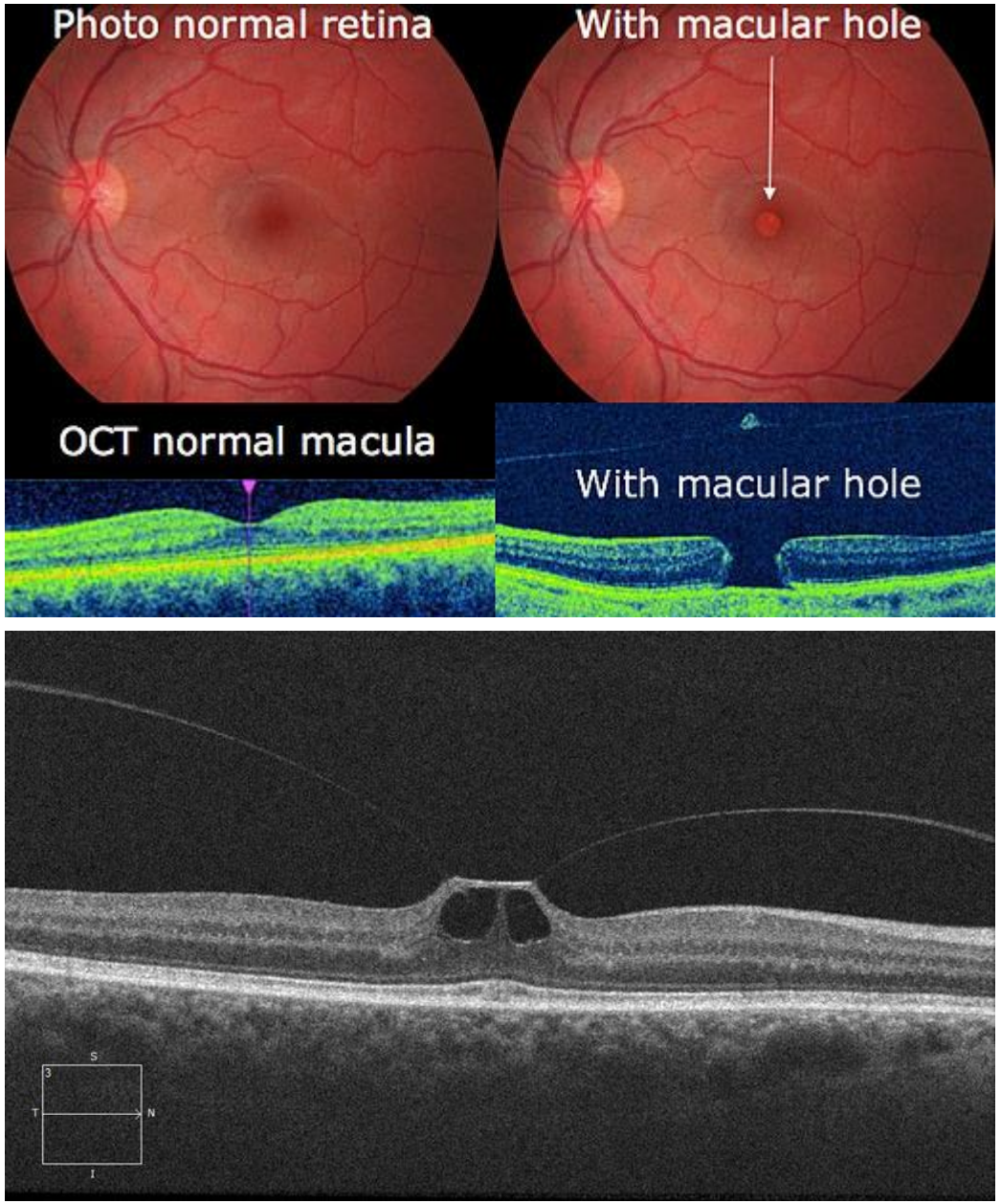
Posterior Vitreous Detachment

- The vitreous is strongly attached to the retina at the vitreous base, a ring-shaped area encircling the ora serrata. The vitreous is also adherent to the optic disc margin, macula, main retinal vessels and some retinal lesions such as lattice degeneration.
- The initial event is liquefaction and syneresis of the central vitreous. A rupture develops in the posterior hyaloid (or vitreous cortex) through which liquefied vitreous flows into the retro-vitreous space, separating the posterior hyaloid from the retina. It typically starts as a partial PVD in the perifoveal region and is usually asymptomatic until it progresses to the optic disc, when separation of the peripapillary glial tissue from the optic nerve head occurs, usually with formation of a Weiss ring and accompanying symptoms.
- Vitreous traction at sites of firm adhesion may result in a retinal tear with or without subsequent rhegmatogenous retinal detachment.
- PVD occurs earlier in myopic eyes, in eyes with inflammatory disease and following blunt trauma or cataract surgery



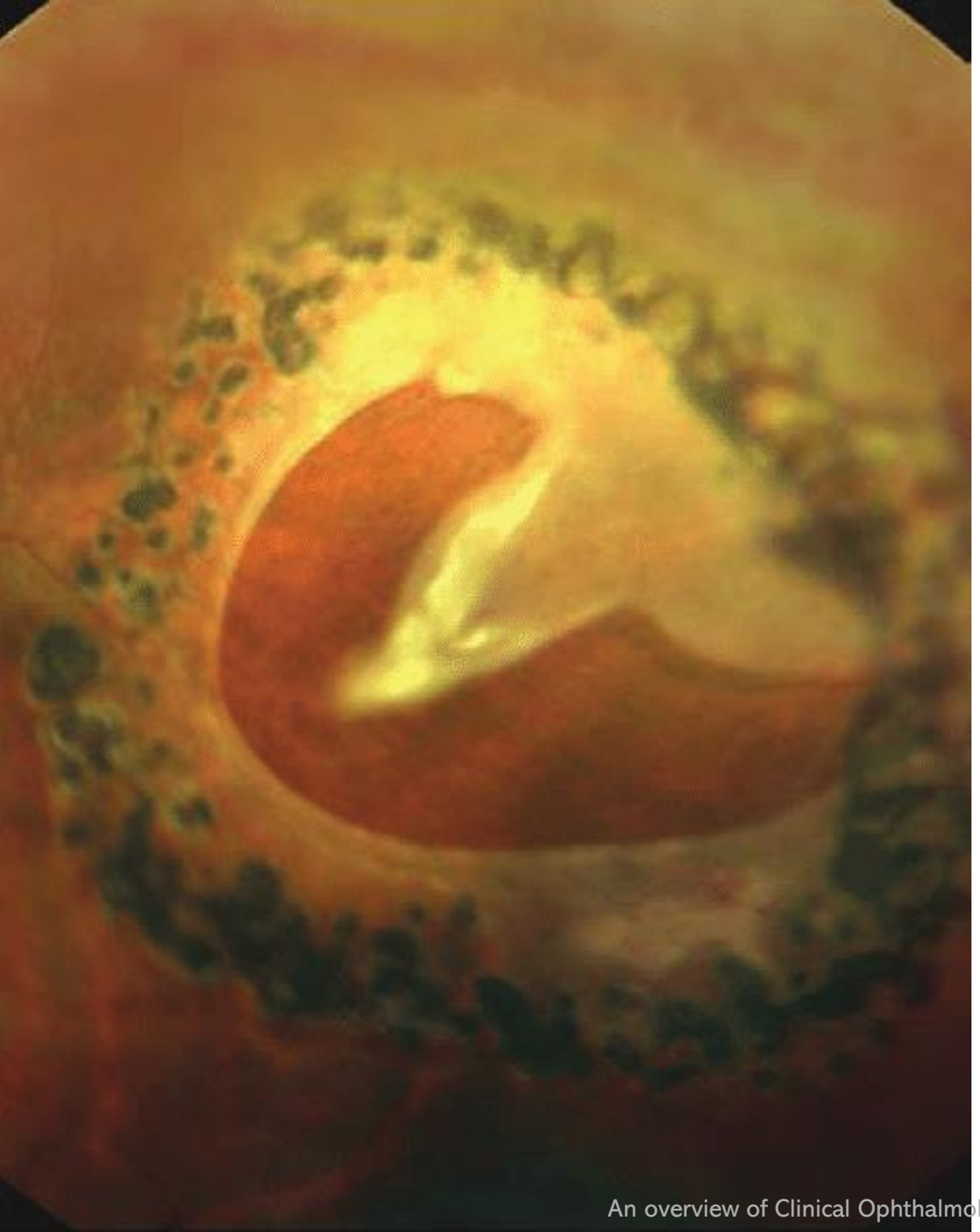
Presenting symptoms

- Floaters are the most common complaint and result from vitreous opacities such as blood, glial cells or aggregated collagen fibers torn from the margin of the optic disc. They move with vitreous displacement during eye movement and scatter incident light, which casts a shadow on the retina that is perceived as a grey structure resembling "hairs", "flies" or "spiderwebs". Floaters may continue indefinitely although they usually gradually diminish over time.
- Photopsias are caused by physical stimulation of the retina from vitreoretinal traction. They reportedly occur in 50% of symptomatic PVD and are usually vertical and temporally located.
- Observation with strict retinal detachment precautions and follow up exam to rule out retinal breaks.



Complications of PVD

- PVDs are occasionally associated with vitreous haemorrhage, retinal tear and retinal detachment.
- These should be ruled out during dilated fundus examination of patients with a PVD. Long term complications may include vitreo-macular traction, lamellar macular holes, full-thickness macular holes and epiretinal membranes.



Retinal Tears

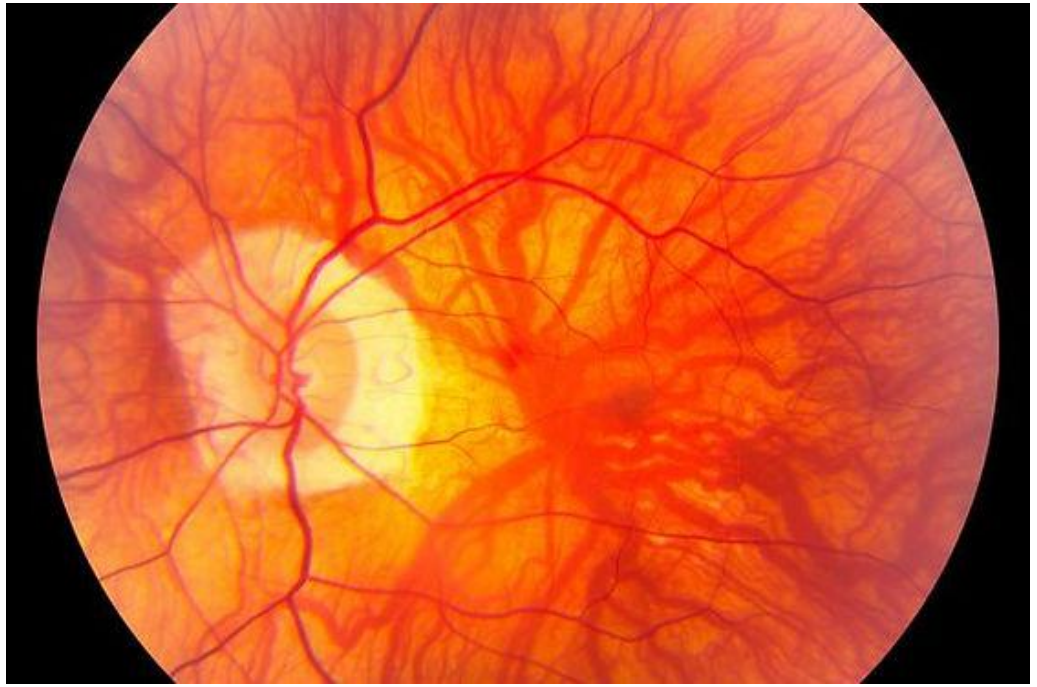
- Horseshoe tears, also referred as flap or U-shaped tears, are full thickness breaks in the neurosensory retina that occur secondary to vitreo-retinal traction. The apex of the flap is pulled anteriorly into the vitreous cavity while the base remains attached to the retina.
- The most common cause of a horseshoe tear is a posterior vitreous detachment (PVD). Horseshoe tears are more common in the superotemporal quadrant.
- Risk Factors: Age, Myopia, Lattice Degeneration, Ocular trauma, Family history of retinal tears or retinal detachment, Previous ocular surgery
- Treatment: retinopexy, either laser photocoagulation or cryotherapy.

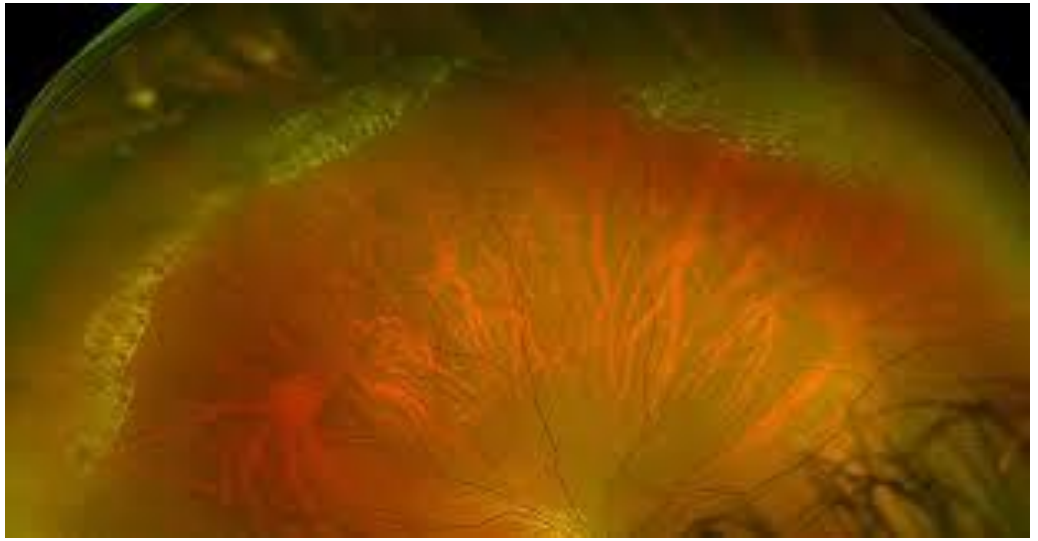
Retinal Detachment

- Retinal detachment is a sight threatening condition with an incidence of approximately 1 in 10000. In the last 50 years techniques in scleral buckling, pneumatic retinopexy and vitrectomy have made the repair of retinal detachments significantly more manageable with better visual outcomes.
- Retinal detachment occurs when subretinal fluid accumulates between the neurosensory retina and the retinal pigment epithelium. One mechanism involves occurrence of a break in the retina allowing vitreous to directly enter the subretinal space. This is known as a **rhegmatogenous retinal detachment**. Rhegmatogenous retinal detachments are often due to retinal tears associated with posterior vitreous detachment or trauma.

Risk Factors for RRD

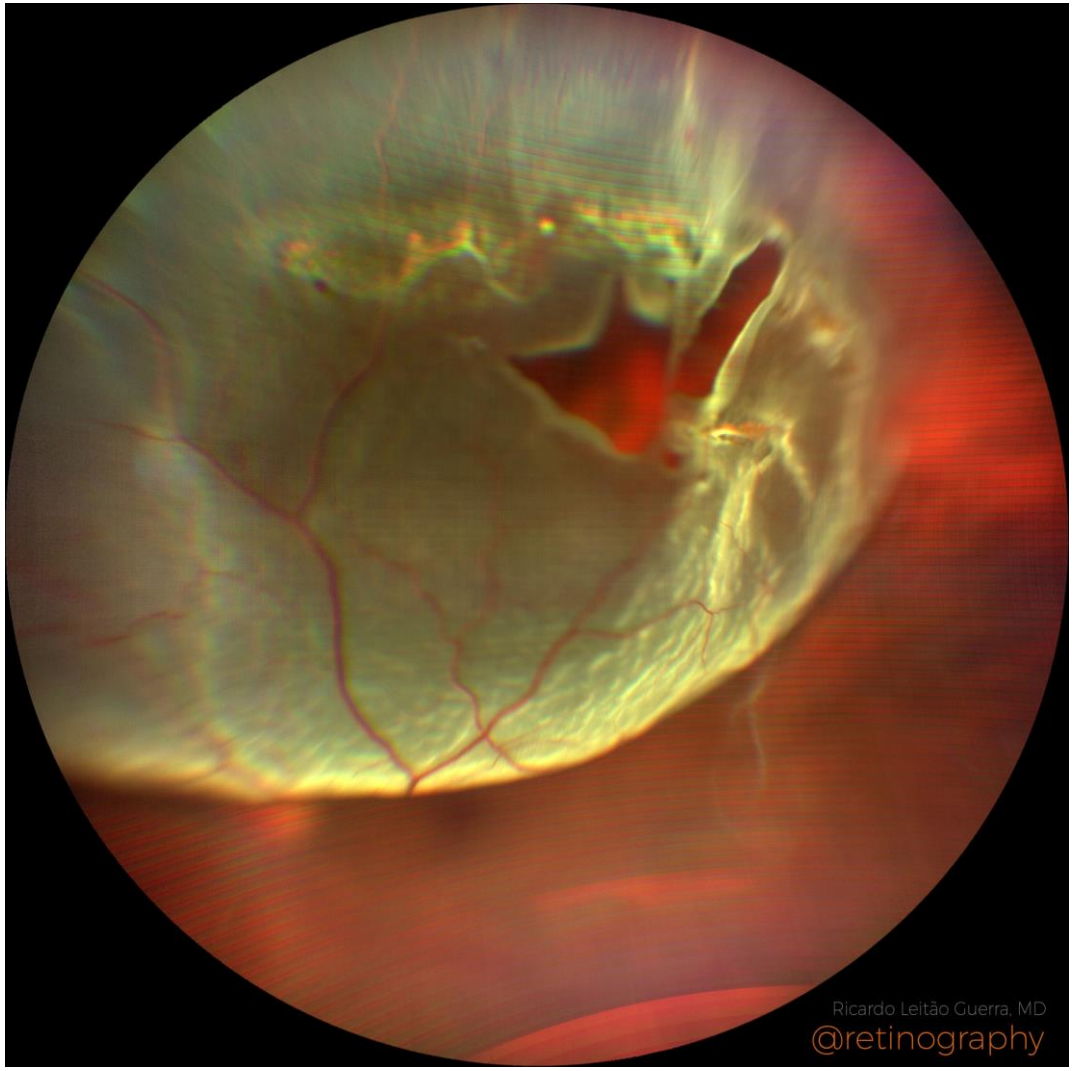
- Lattice degeneration
- Peripheral retinal breaks
- Pathologic myopia
- Previous intraocular surgery
- Trauma
- Previous retinal detachment
- Family history





Clinical presentation

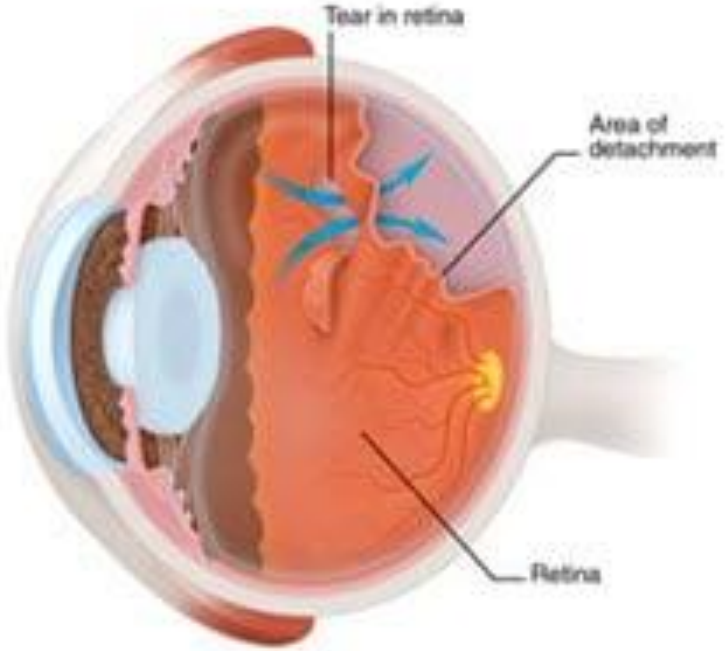
- Patients who present with symptoms of new onset significant photopsias and/or persistent new floaters should be suspected of having a retinal tear, which could lead to a retinal detachment. A patient with constant fixed or slowly progressive visual field loss should be suspected of having a detachment until proven otherwise.
- Important information in the history includes onset of symptoms, presence and duration of decreased central visual acuity, prior trauma, prior surgery, haemorrhage, and a complete past medical history and review of systems.
- On fundoscopy, a rhegmatogenous retinal detachment has a corrugated appearance and undulates with eye movements. In most cases, a retinal break will be identified with proper examination.

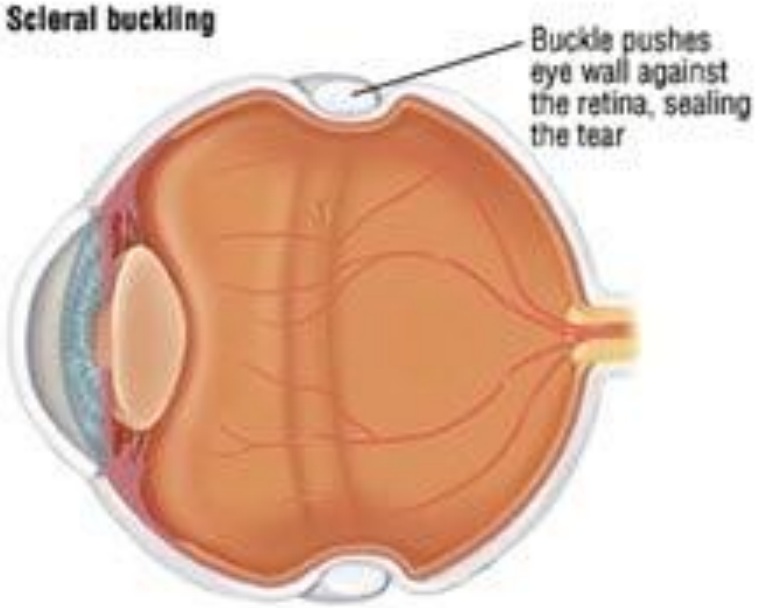


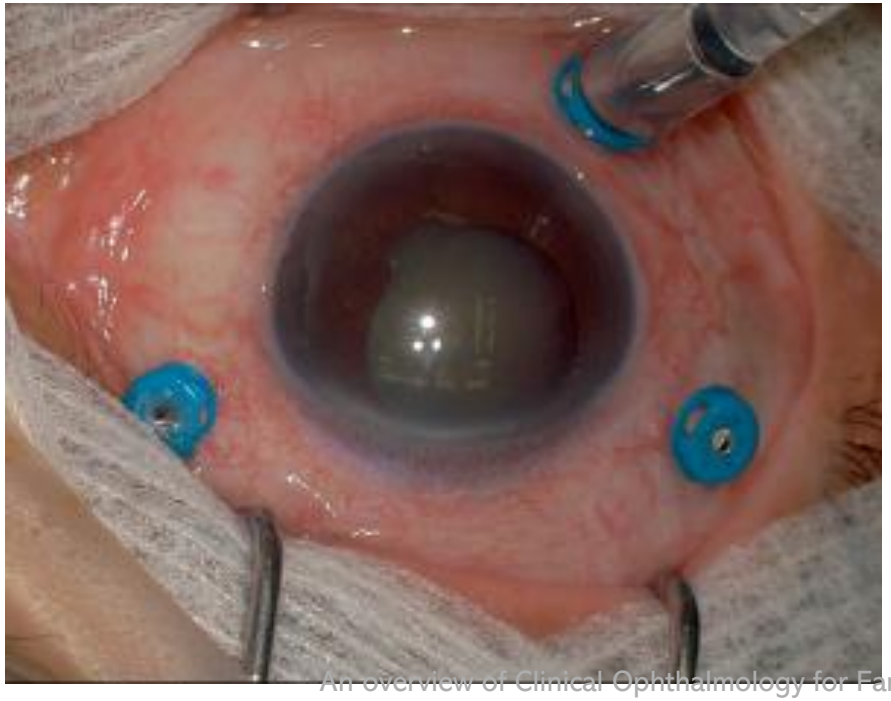
Surgical Treatment

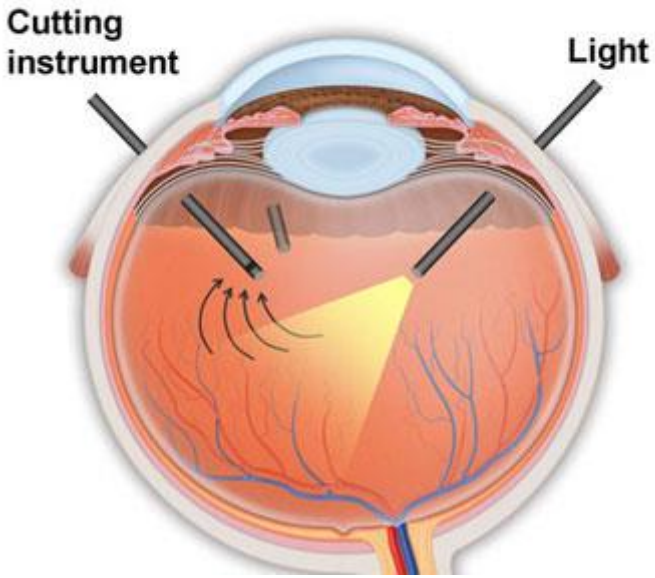
For rhegmatogenous detachments, all retinal breaks should be identified, treated and closed. Techniques for repair include:

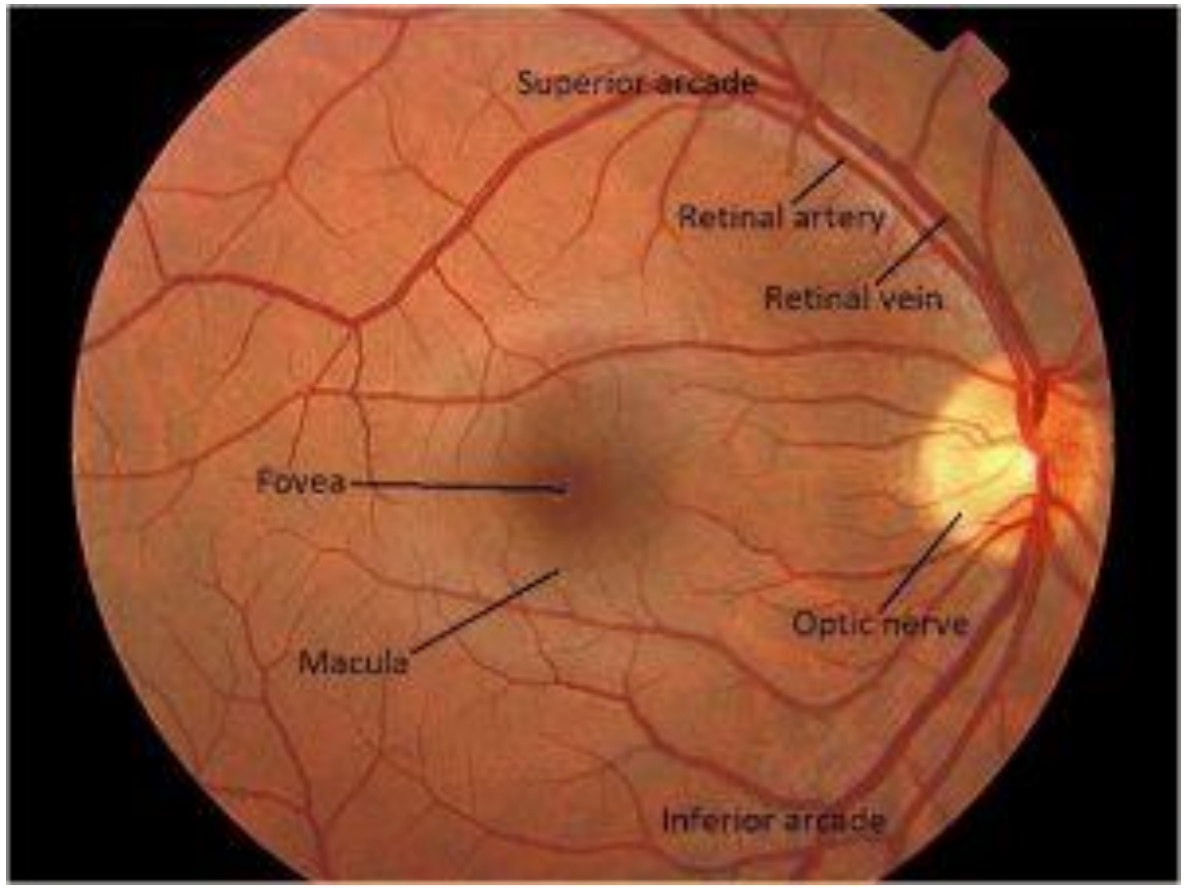
1. **Pneumatic retinopexy** involves the injection of an intraocular gas bubble along with retinopexy using cryotherapy or laser, typically in a clinic setting. An important part in the successful repair of retinal detachment with pneumatic retinopexy involves head positioning so that the gas bubble tamponades the retinal tear. Pneumatic retinopexy is typically only used with retinal detachments due to retinal tears in the superior eight clock hours and involving a single break less than one clock hour
2. **Scleral buckles** are silicone bands permanently placed around the outside of the globe under the extraocular rectus muscles to relieve any traction and support retinal tears. Scleral buckling is combined with retinopexy, typically cryotherapy. This is the oldest method of repair and still has excellent results in well-trained hands.
3. **Pars plana vitrectomy** involves removal of the vitreous by way of cutting the vitreous strands with a vitrectomy machine/handpiece and flattening of the retina through a direct intraocular process with insertion of tamponade. SF6 (lasting 2-3 weeks) and C3F8 (lasting 6-8 weeks) gas are most used although there are indications for silicone oil (lasting permanently until it is taken out) if longer tamponade is needed or in patients who are monocular, must fly, or cannot position.


Tear in retina
Area of detachment
Retina


Scleral buckling
Buckle pushes eye wall against the retina, sealing the tear




Cutting instrument
Light



Vascular retinal pathology

- Retinal Vein Occlusions (RVO)
- Retinal Artery Occlusions (RAO)

CRVO/BRVO

Both CRVO and BRVO are related to occlusion of the retinal vein, however the cause of the occlusion differs based on location.

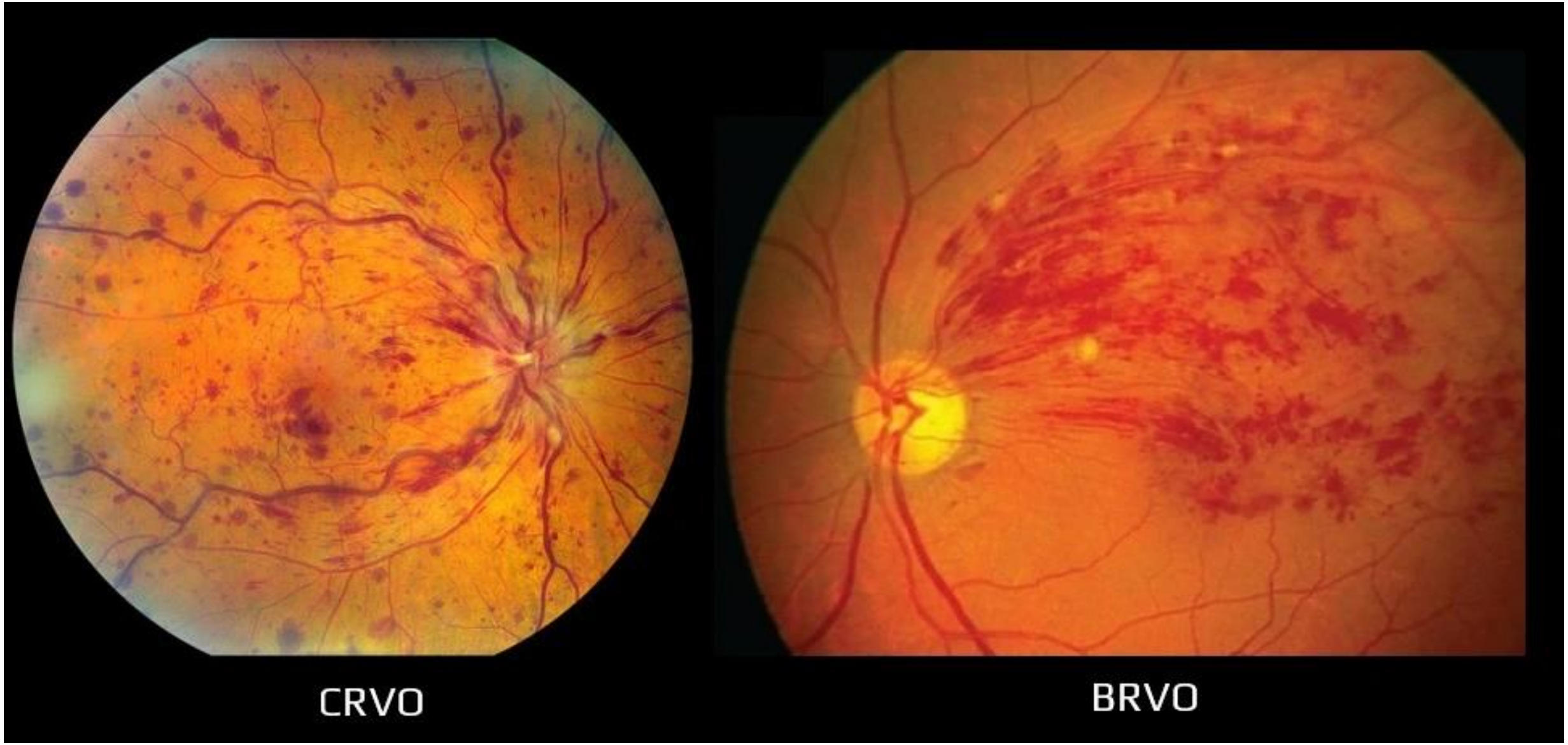
- Central Retinal Vein Occlusion (CRVO) occurs when a thrombus occludes the central retinal vein near the lamina cribrosa (Green, 1981)
- Branch Retinal Vein Occlusion (BRVO) occurs when a thrombus occurs at the arteriovenous crossing point secondary to atherosclerosis of the retinal artery causing compression of the retinal vein. (Frangieh, 1982)

Risk Factors for RVO

- Hypertension
- Open angle glaucoma
- Cardiovascular disease (BRVO)
- High body mass index (BRVO)
- DM (CRVO)

Clinical Picture

- Patients often present with acute vision loss, metamorphopsia.
  Common: Central or peripheral monocular vision loss.
  Less common: Transient visual obscurations or asymptomatic
- Diagnosis is based upon the retinal examination findings of intraretinal haemorrhages, dilated veins, and often cotton wool spots that has been described as a "blood and thunder appearance" for CRVO. Macular oedema may also be present.
- In older patients with cardiovascular risk factors, no laboratory tests are needed. In atypical cases such as younger patients and bilateral or recurrent retinal vein occlusions, laboratory tests such as SPE, thrombophilic screening may be considered.


CRVO
BRVO

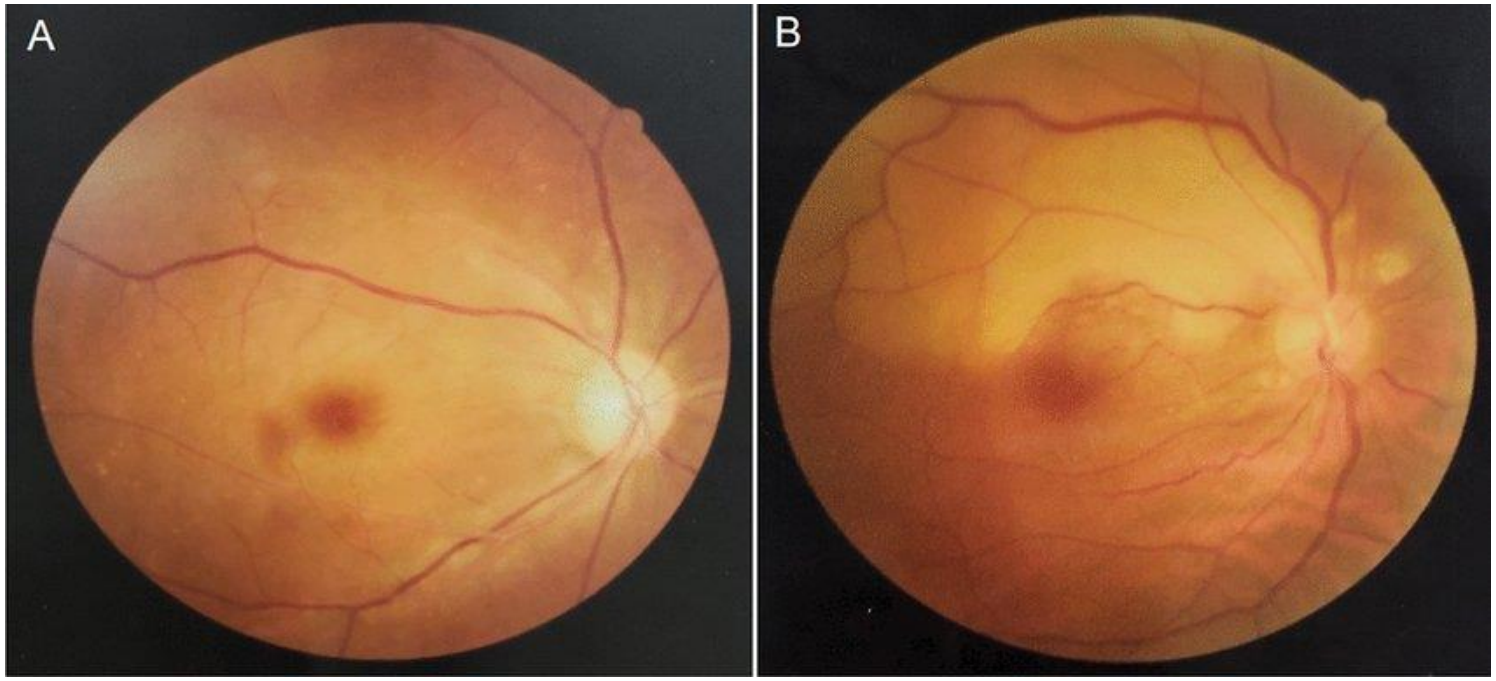
RVO - Treatment

No treatment is available to reverse retinal vein occlusions.

However, macular oedema may be managed with anti-vascular endothelial growth factor (VEGF) or corticosteroid injections.

Iris and retinal neovascularisation requires pan retinal photocoagulation.

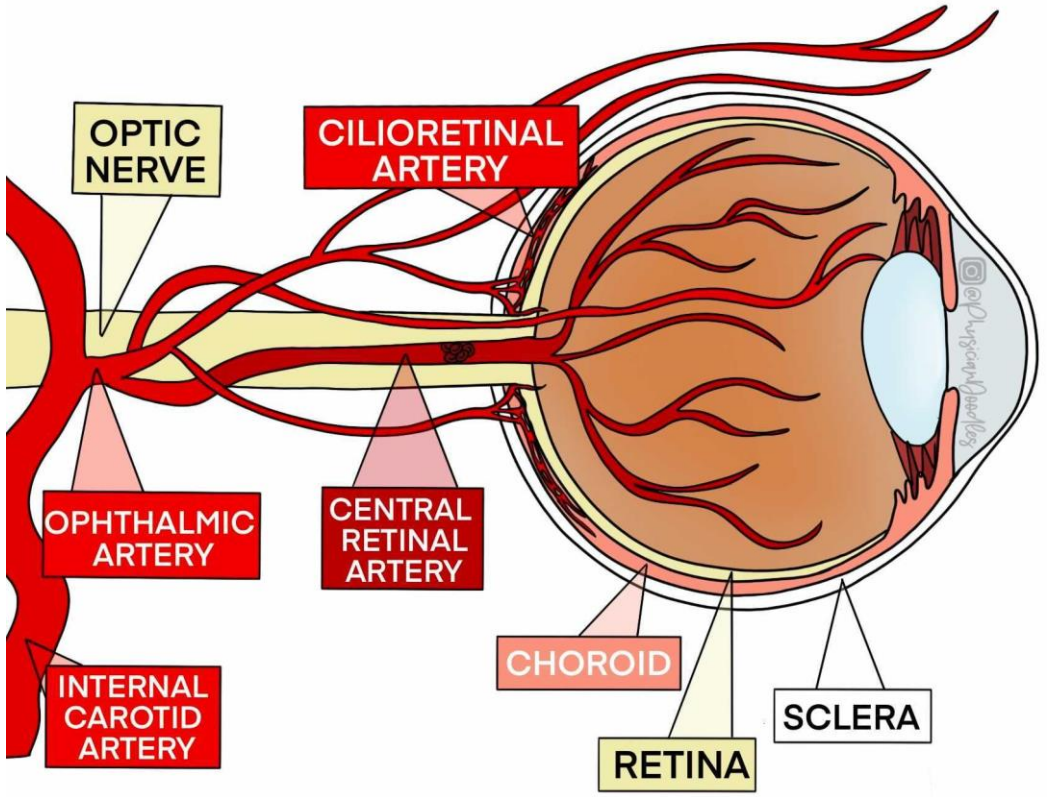
CRAO/BRAO

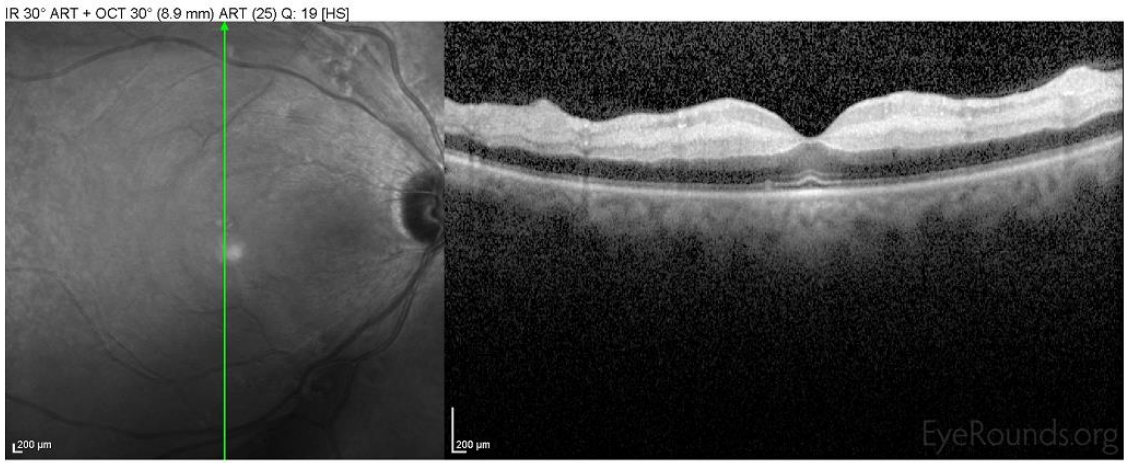


A symptomatic retinal artery occlusion is an ophthalmic emergency that requires immediate evaluation and transfer to A&E. It is an obstruction of retinal blood flow that may be due to an embolus causing occlusion, thrombus formation, vasculitis causing retinal vasculature inflammation.

CRAO/BRAO

- Retinal artery occlusion may occur in any of the vessels supplying the eye. The main artery that supplies the eye and surrounding structures is the ophthalmic artery.
- The central retinal artery, the first branch of the ophthalmic artery, is the main blood supplier of the inner layers of the retina. After entering the eye, the central retinal artery divides into superior and inferior branches.
- In addition, the cilio-retinal artery is a branch of the short posterior ciliary arteries, which is a separate branch of the ophthalmic artery. This artery supplies the choroid and the outer retinal layers.





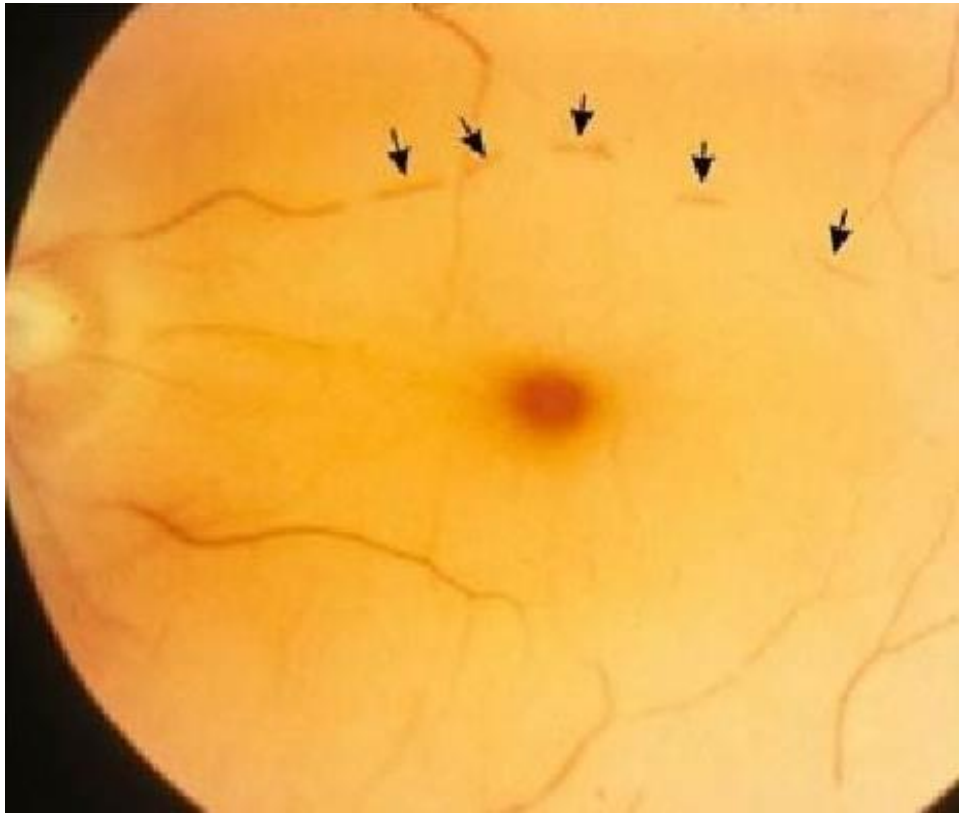
Risk Factors

The risk factors and demographics of retinal artery occlusion are similar to ischemic stroke and include several modifiable risk factors:

- Older age
- Male gender
- Smoking
- Hypertension
- Obesity
- Diabetes
- Hyperlipidemia
- Cardiovascular disease
- Coagulopathy

Clinical presentation of RAO

- Patients typically describe sudden, painless, vision loss that occurs over seconds. Patients with central retinal artery occlusion (CRAO) complain of visual loss over the entire field of vision, while those with a branch retinal artery occlusion (BRAO) complain of hemifield defect.
- Visual loss may have been preceded by transient loss of vision in the past (amaurosis fugax) in the case of embolic sources.



Clinical presentation

- Examination may show RAPD in CRAO.
- **Acute**: Initially fundoscopy may be normal. In CRAO the classic findings of retinal whitening and a cherry red spot are due to opacification of the nerve fibre layer as it becomes oedematous from ischemia. Sectoral whitening in the path of a branch retinal artery is pathognomic for a BRAO. Segmental blood flow, classically described as boxcarring or cattle trucking.
- **Chronic**: Pale optic disc, otherwise fundoscopy is mostly normal as anastomoses form and reperfusion of vessels occurs.

Management

- ESR, CRP and CBC in patients over the age of 50 who have symptoms of GCA. Patients younger than 50 should have a hypercoagulable workup including antiphospholipid antibody syndrome, autoimmune conditions, inflammatory disorders, and other hypercoagulable state.
- In older individuals, atherosclerosis and emboli are the most likely cause of the ischemia. Evaluation of the heart with echocardiography and ECG should be performed. Carotid artery stenosis should be evaluated with carotid ultrasound.
- **Retinal artery occlusion is an emergency that requires emergent systemic evaluation for cerebral vascular accident (CVA).** Patients with acute CRAO, BRAO and amaurosis fugax should be referred to hospital for further immediate management.
- Recent studies have shown a high risk of cerebrovascular event in the ensuing days, weeks and months after retinal artery occlusions. History of hypertension, non-stroke cerebrovascular disease, hyperlipidaemia, and smoking in a patient with RAO, increases the risk of stroke.
- There are no evidence-based therapies that have demonstrated efficacy in improving visual outcomes. Some therapies include ocular massage, lowering IOP, HBOT.

GCA

- Giant cell arteritis (GCA) is the most common systemic vasculitis, affecting medium and large arteries, occurring almost exclusively **above 50 years of age**. Tell-tale features include new onset headache, jaw claudication, pulseless superficial temporal artery, inflammatory girdle stiffness/pain, any acute visual/ocular symptoms, fever, and malaise.
- GCA is a medical and ophthalmic emergency. Untreated, **30-50%** of cases go on to develop anterior ischaemic optic neuropathy or retinal artery occlusion, with consequent permanent loss of vision. Glucocorticoids (GC) should be administered immediately when there is a strong suspicion. Late treatment is the main negative prognostic factor.
- The incidence of biopsy-proven GCA in the Maltese population is 3.82 per 100,000 person-years over age of 50. This is comparable to other Mediterranean countries. Women are affected **two to four times** more often than men. The highest incidence of GCA is in 75- 85 years old patients and Caucasians of North European descent.

2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EUROPEAN ALLIANCE OF ASSOCIATIONS FOR RHEUMATOLOGY

CLASSIFICATION CRITERIA FOR **GIANT CELL ARTERITIS**

AMERICAN COLLEGE of RHEUMATOLOGY
Empowering Rheumatology Professionals

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify the patient as having giant cell arteritis when a diagnosis of large-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

CRITERIA ABSOLUTE REQUIREMENTS

Age ≥ 50 years at time of diagnosis	

ADDITIONAL CLINICAL CRITERIA

Morning stiffness in shoulders/neck	+2
Sudden visual loss	+3
Jaw or tongue claudication	+2
New temporal headache	+2
Scalp tenderness	+2
Abnormal examination of the temporal artery[1]	+2

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter[2]	+2
Positive temporal artery biopsy or halo sign on temporal artery ultrasound[3]	+5
Bilateral axillary involvement[4]	+2
FDG-PET activity throughout aorta[5]	+2

Sum the scores for 10 items, if present. A score of ≥ 6 points is needed for the classification of GIANT CELL ARTERITIS.

Sensitivity 87%
Specificity 95%

Diagnostic investigations

Test	Result
› ESR	ESR ≥50 mm/hour by the Westergren method
› C-reactive protein (CRP)	elevated
› FBC	typically, patients have a normochromic, normocytic anaemia with a normal WBC count and elevated platelet count; mild leukocytosis may occur
› LFTs	transaminases and alkaline phosphatase are often mildly elevated
› temporal artery biopsy	histopathology typically shows granulomatous inflammation; in about 50% of cases, multinucleated giant cells are present; inflammatory infiltrate may be focal and segmental

The combination of elevated ESR and CRP provided better specificity than either test alone and was associated with greater odds of a positive TAB.
Among GCA patients with a positive biopsy, normal ESR and CRP was observed in 4% of cases.
It is also important to note that biopsy of the temporal artery carries a significant false negative rate (5% to 9%) due to skip lesions.

Ophthalmic manifestations of GCA

- Acute visual loss in one or both eyes is by far the most feared complication of GCA. If untreated, contralateral eye involvement within 1–2 weeks occurs in around a third of cases. Once established, visual impairment is usually permanent with severe optic atrophy.
- GCA can affect anywhere from the retina to the occipital lobe. The most common cause of vision loss is **arteritic anterior ischaemic optic neuropathy** (AAION), which occurs as a result of inflammatory occlusion of the short posterior ciliary arteries, which are the main source of blood supply to the optic nerve head.
- CRAO can also be a ophthalmic manifestation of GCA. Amaurosis fugax is an important visual symptom because it precedes permanent visual loss in 44% of cases. Consider GCA in cases of ?TIA.
- Transient or constant diplopia is a frequent visual symptom related to GCA. Common findings include upward gaze palsy and VIth cranial nerve palsy. Signs also include: RAPD, pallid/waxy optic disc swelling (highly indicative), cotton wool spots, visual field defects.

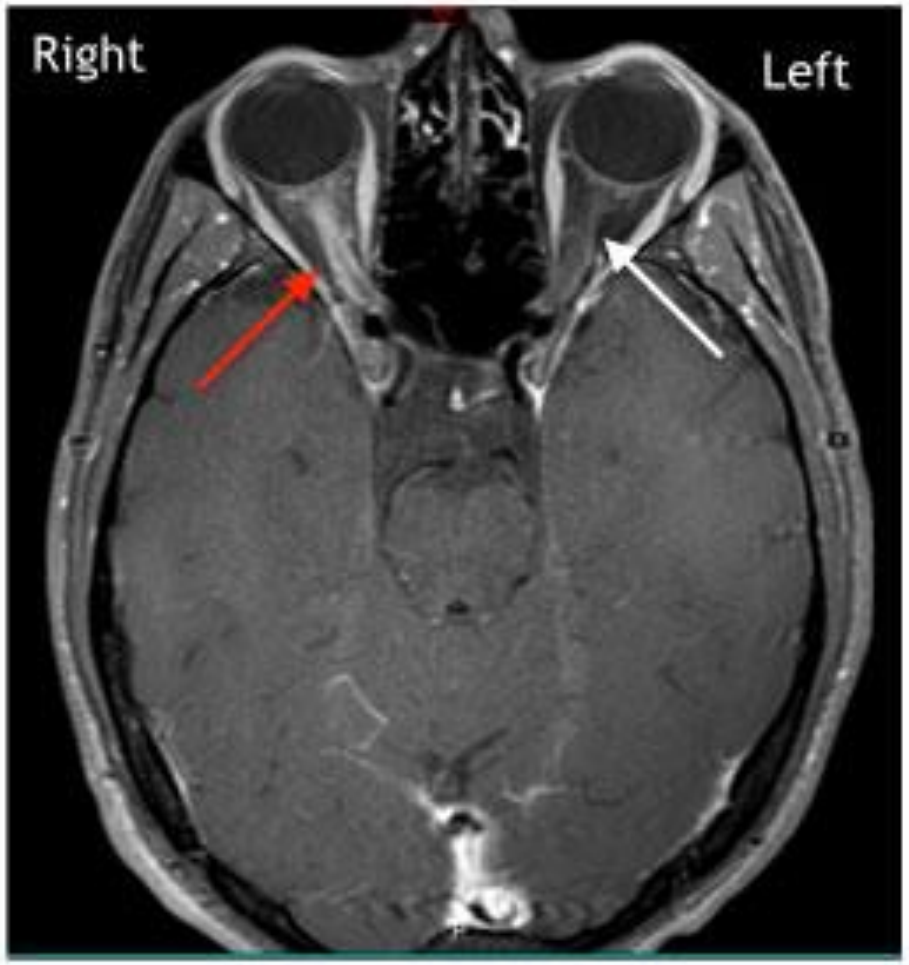
Typical findings of a patient with arteritic ischemic optic neuropathy. Note the pallid disc oedema, associated haemorrhages, and adjacent cotton wool spot.

Treatment – ‘Time is Sight’

- Urgent referral to ophthalmology in case of visual symptoms, and rheumatology for all GCA patients, is recommended. This should ideally be done prior to GC initiation but should not delay treatment.
- Early high-dose GC treatment is essential for rapid symptom control and to prevent further visual loss. Improvement of symptoms often begins within hours to days after commencing GC.
- British Society of Rheumatology guidelines recommend the following starting doses for steroids:
  - Prednisolone 40–60mg daily for uncomplicated GCA (no jaw claudication or visual disturbance)
  - IV methylprednisolone 500–1000mg for 3 days before oral steroids for evolving visual loss (recent onset of visual symptoms over 6–12 hours) or amaurosis fugax.
  - Prednisolone 60mg daily for established visual loss, to protect the contralateral eye
- Guidelines recommend the use of aspirin, given that ischaemic complications are the biggest source of morbidity and mortality in GCA.
- The initial high dose GC should be continued for 3–4 weeks and then gradual tapering, provided there is no relapse. All patients should be assessed for need of bone protection Most patients are able to discontinue GC after two years of treatment.

Optic neuritis

- Demyelinating optic neuritis may be related to MS. Presenting symptom in approximately 20% of MS patients and >50% of MS patients experience ON.
- Diagnosis is largely based on history and physical examination –prompt referral is essential for treatment and to expedite visual recovery.
- Inflammation of the optic nerve and it's lining results in inflammation of the extraocular recti muscles that surround the optic nerve. Recti muscle involvement results in ocular pain, especially with eye movements.
- The characteristic white matter lesion on MRI brain is the most important predictor in the development of MS.

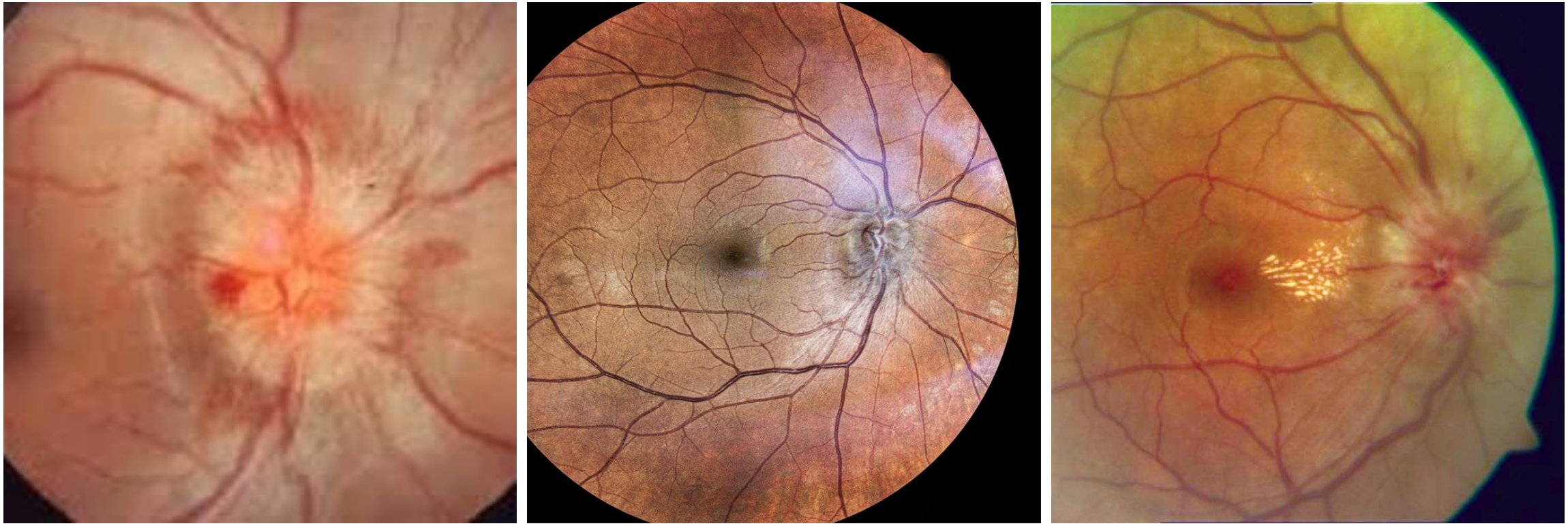


Risk Factors for Demyelinating Optic Neuritis

- 3:2-female:male ratio
- Young age (20-45 years old)
- A prodromal flu like illness commonly accompanies the event but does not always occur
- Patients with MS often have ON (up to 75% of patients with MS will have at least one episode of ON in their lifetime).

Optic neuritis clinical signs

- Unilateral decreased visual acuity
- Decreased colour vision (red desaturation)
- RAPD
- Other efferent lesions may be present in MS (e.g., ocular dysmetria or internuclear ophthalmoplegia)
- Any nerve fibre layer or central scotoma on confrontation visual fields or formal visual field testing
- Typically normal fundus but optic disc oedema can be seen in up to 35%. Thus lack of optic disc oedema is the rule rather than the exception for ON.
- Retinal vascular sheathing, pars planitis



EXAMPLES OF OPTIC NEURITIS ON FUNDOSCOPY

Retrobulbar ON

- Retrobulbar ON, where the inflammation occurs behind the optic nerve head, is more typical of idiopathic demyelinating disease and occurs in two-thirds of cases.
- In retrobulbar ON, the optic nerve looks normal but may develop pallor after 8 weeks from the onset of ON.
- A young female with vision loss, pain with eye movements and a normal looking optic nerve is likely to have retrobulbar ON.

Treatment: Interesting outcomes from the ONTT

- **Combination drugs speed recovery**. ONTT helped define the role of corticosteroids in the treatment of acute optic neuritis. When the study originated, many doctors were treating the condition with oral corticosteroids. The study looked at oral prednisone vs. high-dose intravenous methylprednisolone vs. placebo and found that the IV methylprednisolone followed by a tapering course of oral prednisone accelerated visual recovery by a few weeks.
- **Recovery of vision occurs with or without treatment**. The investigators found that the choice of regimen has no effect on final visual outcome. Most patients in the placebo group recovered vision, on average, in six to eight weeks.
- **Oral prednisone alone was no better than placebo with respect to visual recovery and, in fact, was associated with twice the risk of recurrent optic neuritis**. It is no longer recommended for an initial episode of typical, presumed demyelinative optic neuritis. The therapy should include either high-dose methylprednisolone or nothing.

Prognosis

The vast majority (94%) of patients recover vision to 6/12 or better at 5 years. Only 3% of patients had 6/60 or worse visual outcomes at 5 years (based on the ONTT).

Visual recovery tends to occur by 1 month after onset and the majority recover within 1-3 months.

Diplopia

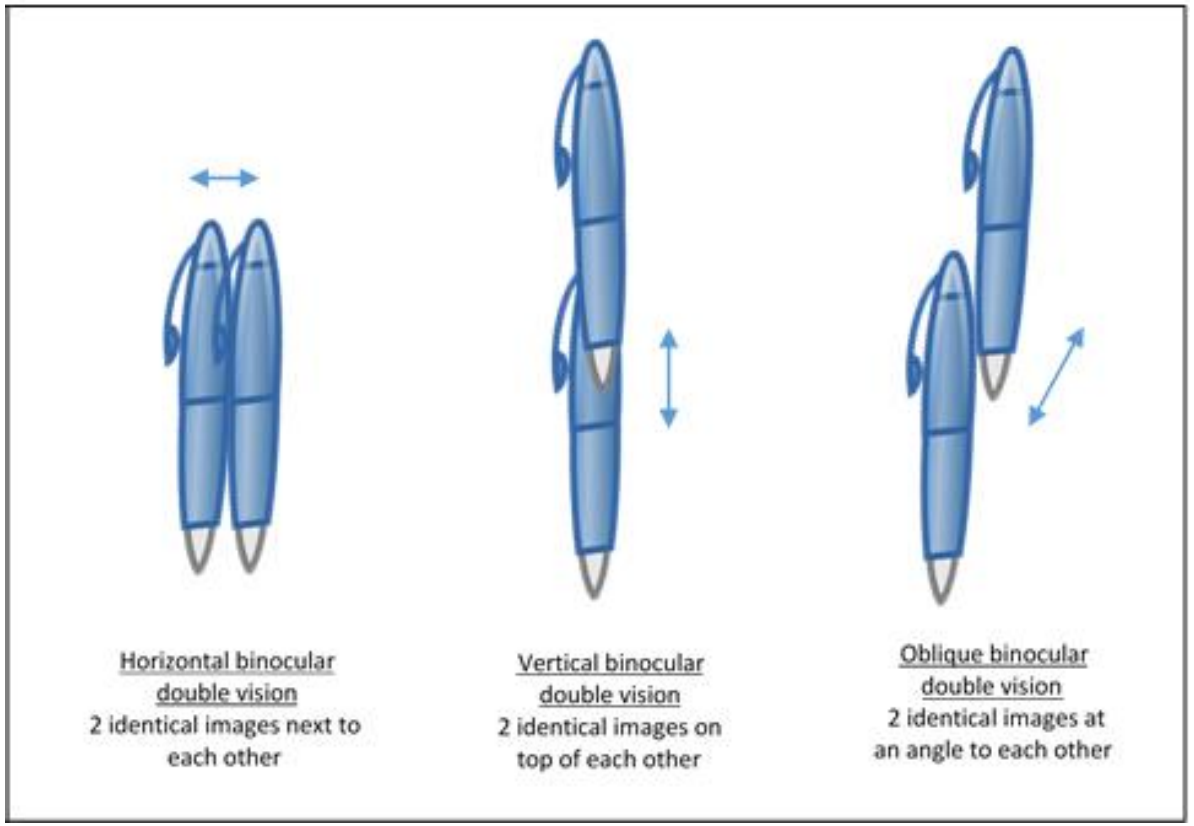
The initial stage of diplopia workup is to identify whether it is monocular diplopia or binocular diplopia.

- Monocular diplopia persists when the unaffected eye is closed but will resolve when the affected eye is closed. Typically, the cause is ocular.
- Binocular diplopia resolves with either eye being closed and indicates ocular misalignment as an underlying problem.

Binocular Diplopia – what clinical questions to ask

1. Are the images separated horizontally, vertically, or obliquely/diagonally? How does distance affect diplopia?

- Binocular horizontal diplopia is usually due to disease of the medial or lateral rectus muscle, the neuromuscular junction, or the nerves supplying these muscles (e.g., cranial nerves III or VI). Diplopia worse with distance is more typical of sixth nerve palsy because of difficulty with divergence at distance of the eyes while diplopia worse at near is more suggestive of medial rectus palsy because of the need for convergence of the eyes at near.
- Vertical diplopia can be due to involvement of extraocular muscles, neuromuscular junction (e.g., myasthenia gravis), or cranial nerves (e.g., CN III, IV). Myogenic involvement can occur with disease of the superior rectus, inferior rectus, superior oblique, or inferior oblique muscles alone or in combination. The three-step test is utilized to isolate the vertically acting weak muscle.



Binocular Diplopia – what clinical questions to ask

2. Which field of gaze provokes / worsens diplopia? Which field(s) of gaze are images closest to each other?

- The worst position of gaze will typically represent the field of action of the paretic muscle.
- However, if there is muscle restriction (e.g., thyroid eye disease, orbital fracture, orbital myositis) then the diplopia may be worse in the opposite field of action of the restricted muscle.

Binocular Diplopia – what clinical questions to ask

3. Is there pain or any associated neurological symptoms?

- Localized pain in or behind the eye or orbit may suggest intra-orbital pathology and headache may suggest intracranial pathology. Sudden onset of severe headache may be suggestive of SAH. A pupil involving third nerve palsy may occur from SAH from ruptured posterior communicating artery aneurysm. Pain in the V1 and V2 divisions of the trigeminal nerve can suggest an intracranial (e.g., cavernous sinus) or intra-orbital lesion.
- Associated neurological symptoms can also assist with narrowing the differential diagnosis and can help to topographically localize the lesion.

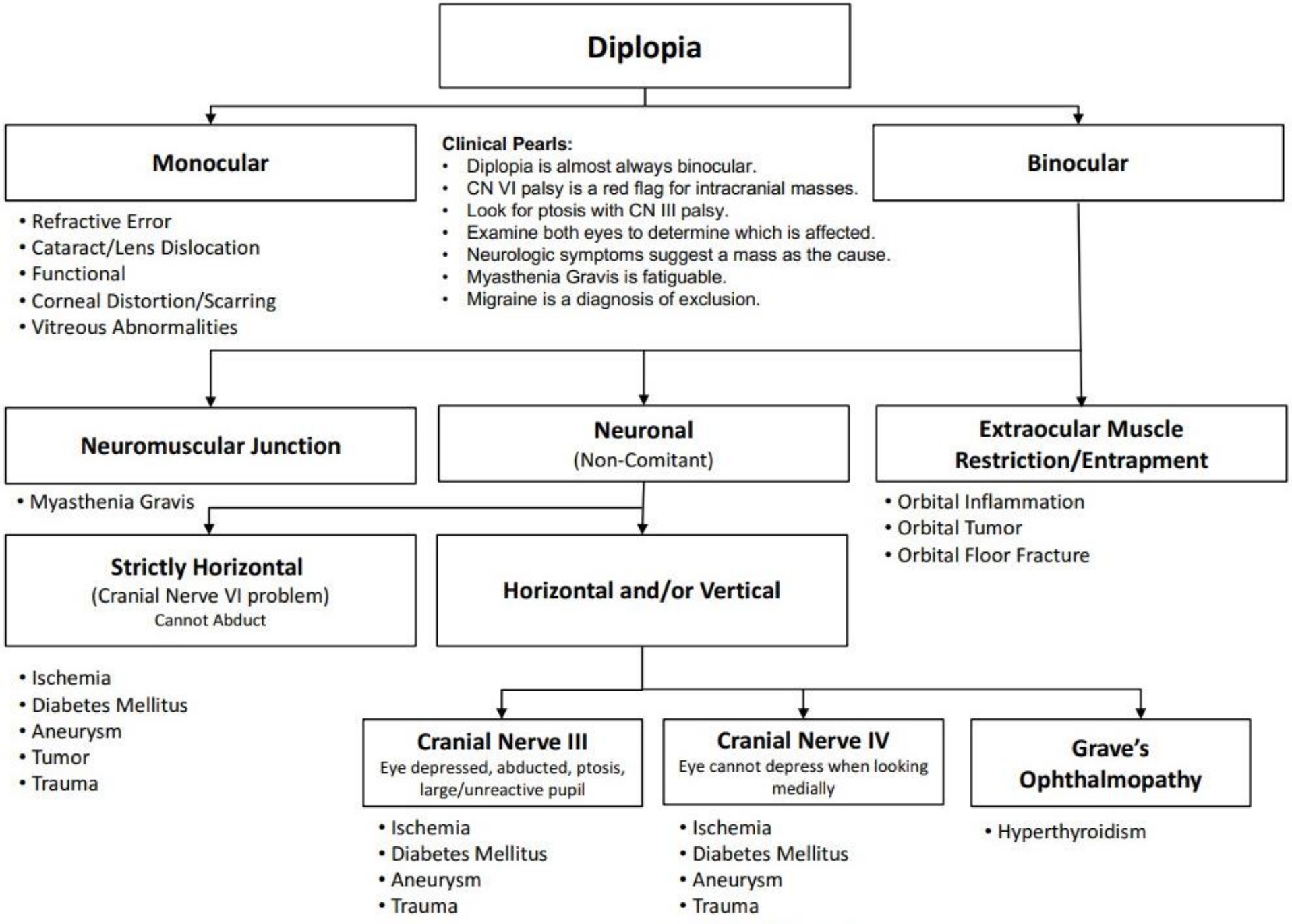
Localising the cause for diplopia

The examination should localise to supranuclear, nuclear, or infranuclear pathways.

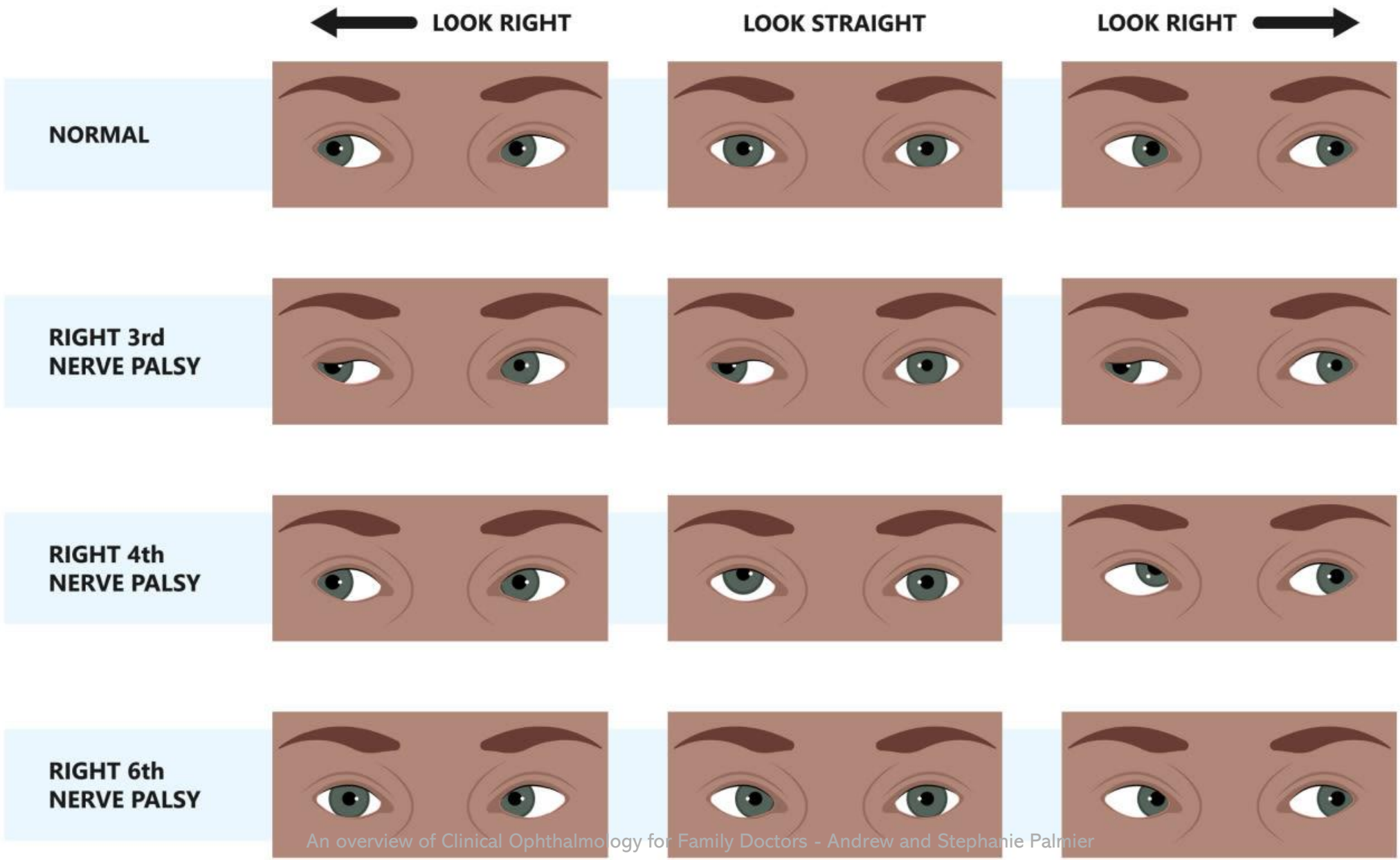
The most common supranuclear causes of diplopia include convergence insufficiency, neurodegenerative disease.

The most common internuclear ophthalmoplegia is an INO and involves the medial longitudinal fasciculus.

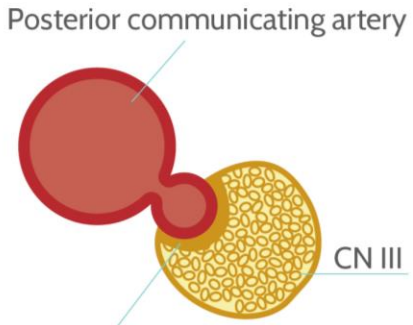
The infranuclear pathway includes the peripheral ocular motor cranial nerves (CN III, IV, VI), the neuromuscular junction (e.g., MG), and the extraocular muscles themselves (e.g., thyroid eye disease).

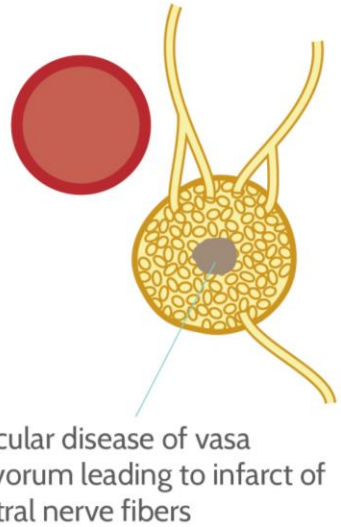

Diplopia
Monocular
• Refractive Error
• Cataract/Lens Dislocation
• Functional
• Corneal Distortion/Scarring
• Vitreous Abnormalities
Clinical Pearls:
• Diplopia is almost always binocular.
• CN VI palsy is a red flag for intracranial masses.
• Look for ptosis with CN III palsy.
• Examine both eyes to determine which is affected.
• Neurologic symptoms suggest a mass as the cause.
• Myasthenia Gravis is fatiguable.
• Migraine is a diagnosis of exclusion.
Binocular
Neuromuscular Junction
• Myasthenia Gravis
Neuronal
(Non-Comitant)
Extraocular Muscle Restriction/Entrapment
• Orbital Inflammation
• Orbital Tumor
• Orbital Floor Fracture
Strictly Horizontal
(Cranial Nerve VI problem)
Cannot Abduct
• Ischemia
• Diabetes Mellitus
• Aneurysm
• Tumor
• Trauma
Horizontal and/or Vertical
Cranial Nerve III
Eye depressed, abducted, ptosis, large/unreactive pupil
• Ischemia
• Diabetes Mellitus
• Aneurysm
• Trauma
Cranial Nerve IV
Eye cannot depress when looking medially
• Ischemia
• Diabetes Mellitus
• Aneurysm
• Trauma
• Subdural Hemorrhage
Grave's Ophthalmopathy
• Hyperthyroidism

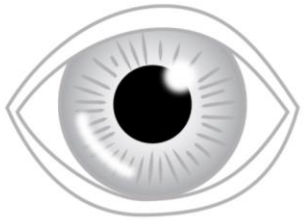
CRANIAL NERVE PALSY - EXAM FINDINGS



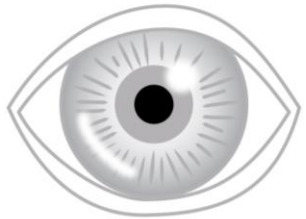
Pupil sparing vs pupil involving CNIII palsy



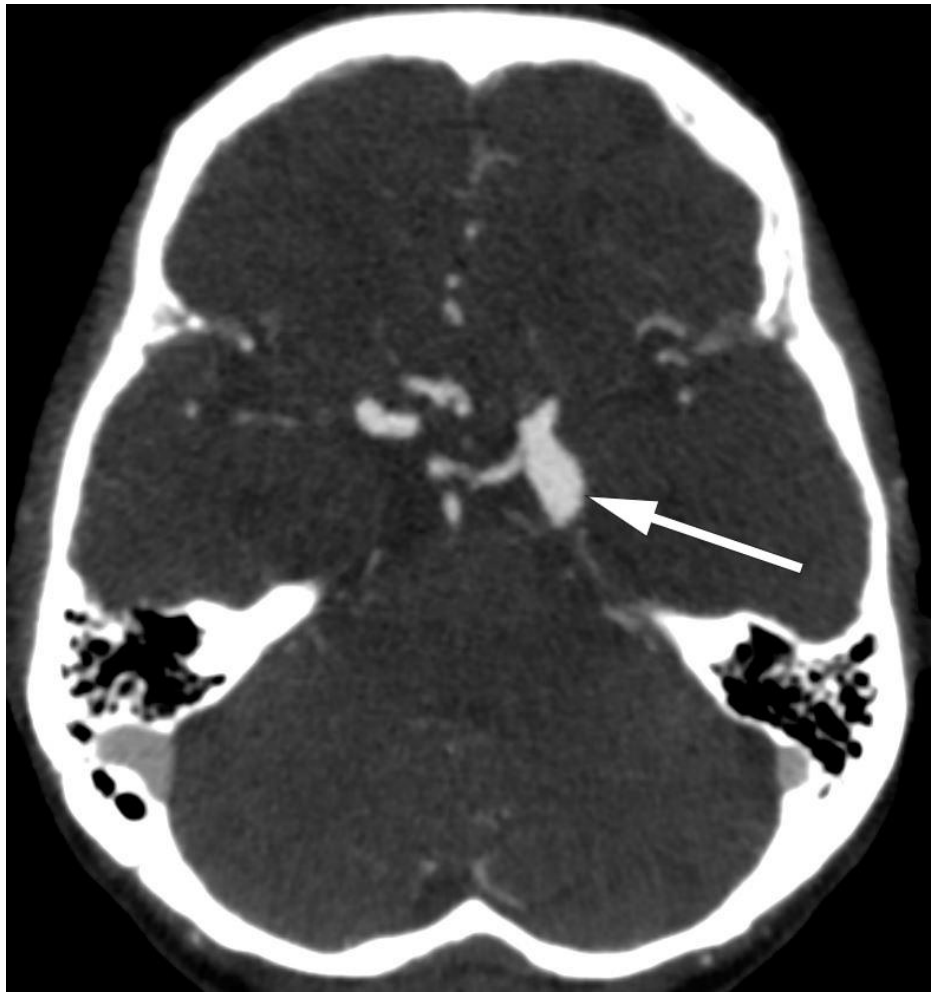




Pupillary dilation

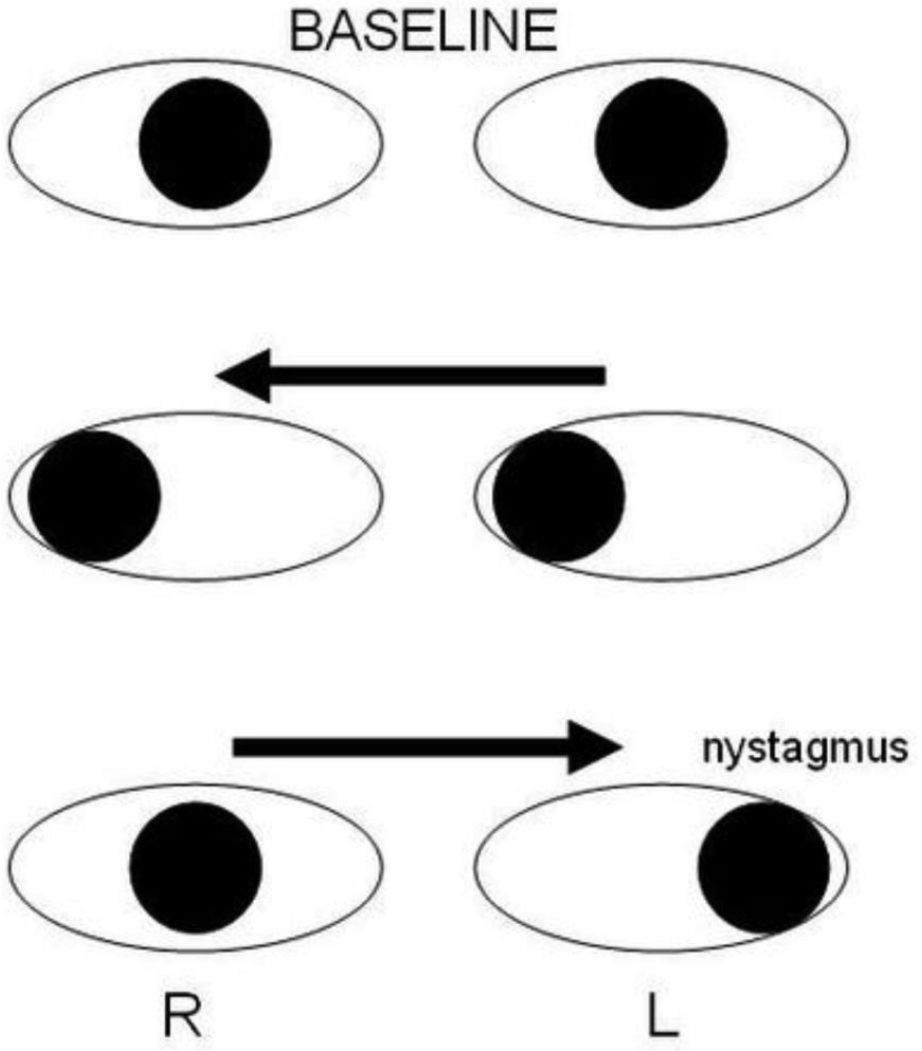


Pupil sparing



Internuclear ophthalmoplegia

- Internuclear ophthalmoplegia (INO) is an ocular movement disorder that presents as an inability to perform conjugate lateral gaze and ophthalmoplegia due to damage to the interneuron (MLF) between two nuclei of cranial nerves VI and III.
- Symptoms of INO may vary in severity. Symptoms range from, horizontal diplopia, difficulty in tracking high-speed objects, or dizziness on lateral gaze. Horizontal diplopia is caused by the limitation of adduction in the ipsilateral eye. The diplopia becomes more prominent on looking at objects on the opposite side of the lesion and diplopia is usually not seen in the primary gaze.




"That's all Folks!"