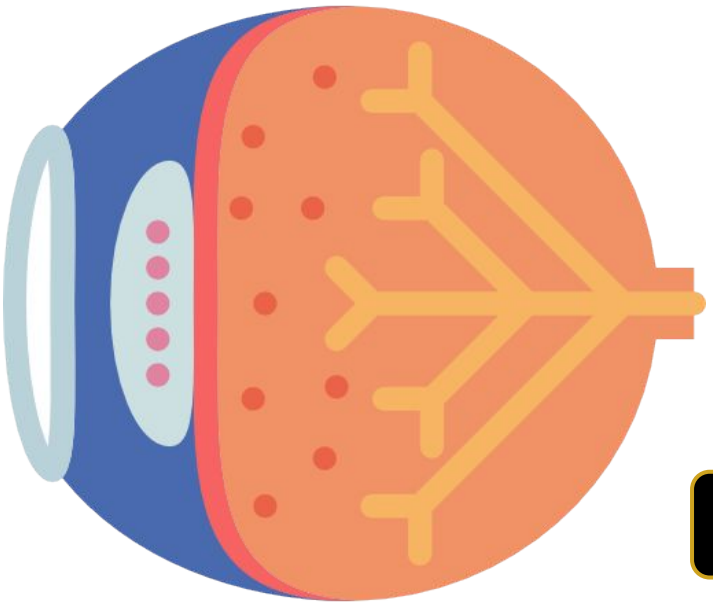
Lecture: 7



Editing file

Chronic visual loss

- **Presented by Dr. Majed Al Kharashi & Dr. Essam Osman**
- To know common conditions that present with chronic visual loss (causes, clinical manifestation and management):
  - Chronic glaucoma (causes, types, management), Senile cataract, Diabetic macular edema, macular degeneration and Hereditary retinal diseases.
- How to use ophthalmic instruments and results of investigations to differentiate between different causes.
- To know what are treatment options for different conditions.
- To know when to refer a case to a specialist.
- Causes, Medication, types are important for the exam
- All the Extra slides were not explained in the lecture and not included in the slides

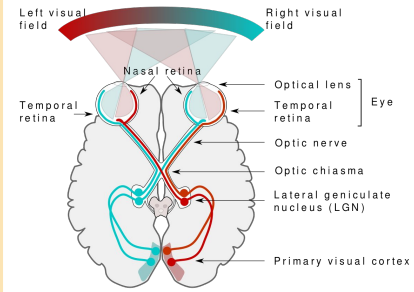
Important | Doctor's notes | Golden notes | Extra | Book

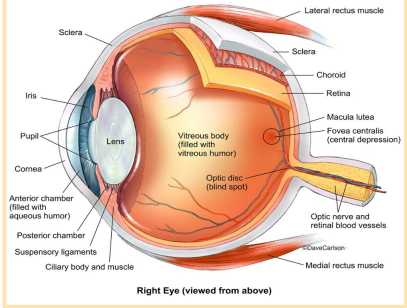
OPTHALMOLOGY TEAM

Introduction To Chronic Visual Loss

❖ Anatomy of the eye & the visual pathway

- ◇ **Phototransduction**: By photoreceptors (rods and cones).
- ◇ **Image processing**: By horizontal, bipolar, amacrine and RGCs.
- ◇ **Output to optic nerve**: Via RGCs and nerve fiber layer.





Definitions

- It's a **gradual** progressive event **(painless visual loss).**
- There is **no pain** no redness, no discharge (quite eye).
- **Duration**: > 2 weeks.

❖ Vision: So how can we assess the vision?

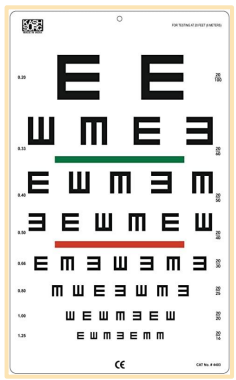
- ◇ Quantity: VA (Visual acuity).
  - ■ See the image: good quantity but bad quality.
- ◇ Quality: VF (Visual Field), clarity of vision, color vision.

❖ Causes:

- ◇ Refractive
- ◇ Cornea
- ◇ Lens
- ◇ Vitreous
- ◇ Retina
- ◇ Optic Nerve
- ◇ Neurologic

❖ Approach:

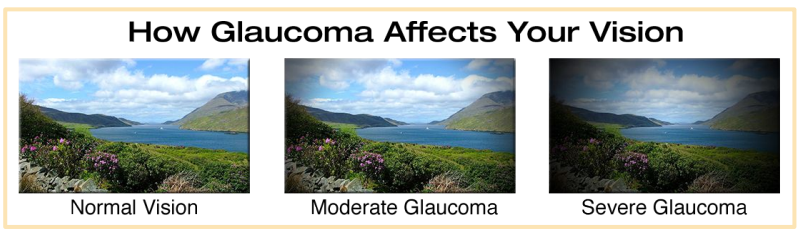
- ◇ Measure the IOP; always high in glaucoma except in **normal tension** glaucoma which is type of open angle glaucoma presented with low IOP < 21
- ◇ Evaluate the Optic nerve, normal or abnormal disc
- ◇ Evaluate the lens to rule out cataract
- ◇ Evaluate the function and appearance of macula and retina to rule out diabetes and age-related macular degeneration.



❖ Causes of slowly progressive visual loss :

From most to least common:

- ◇ Cataract commonest cause
- ◇ Chronic Glaucoma
- ◇ Diabetic retinopathy
- ◇ Macular degeneration age related
- ◇ Retinitis pigmentosa.



Causes of Chronic Visual Loss

1 Glaucoma

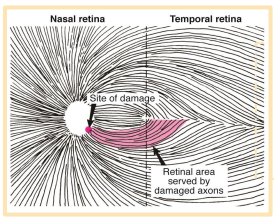
Helpful video

❖ The significance: الماء الأزرق

- ◇ A major cause of blindness, most common cause of blindness in **African Americans** MCQ!
- ◇ **Progressive optic neuropathies**, that have in common characteristic morphological changes at the optic nerve head and retinal fiber layer because of the increase in the IOP cause the damaging (compression of the nerve) in the absence of other ocular disease or congenital anomalies. **Progressive retinal ganglion cell death** and **visual field loss** are associated with these changes.
- ◇ **Second leading cause of blindness (cataract is the first)** MCQ!
- ◇ Often Asymptomatic; in early stage. Majority of patients lack pain, ocular inflammation.
  - ■ If glaucoma is detected early and treated medically or surgically, blindness can be prevented.
  - ■ **Damage is irreversible** MCQ!

❖ Classification of glaucoma:

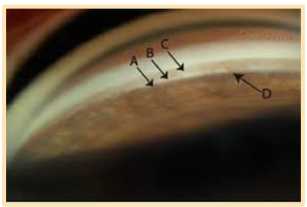
- ◇ **According to the etiology:**
  - ■ **Primary**, **secondary** and **congenital**.
    - Most common causes of secondary glaucoma: DM (neovascular glaucoma), uveitis (unilateral or bilateral).
    - Primary Vs Secondary: Primary No detectable ocular or systemic abnormality, often bilateral and often familial. Secondary predisposing ocular or systemic abnormality, often unilateral and often sporadic.
- ◇ **According to the appearance of the angle:**
  - ■ Open Angle Glaucoma.
  - ■ Acute Closed angle Glaucoma.



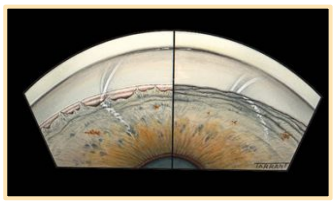
Glaucoma horizontal damage

❖ Risk factors:

- ◇ **African Americans.** MCQ!
- ◇ IOP **(most imp)**.
- ◇ Age.
- ◇ Family history. (most imp)
- ◇ DM/HTN.
- ◇ Medication "Steroids".
- ◇ Myopia.

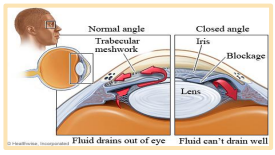


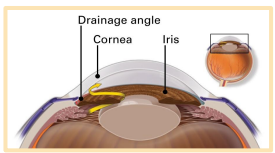
Open

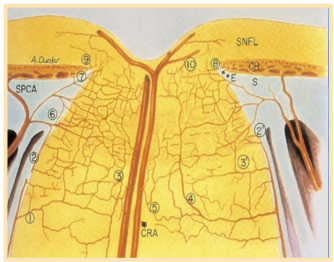


Closed

- In glaucoma, the loss will starts with peripheral vision, Why?
  - Because the increase in the IOP will start to compress the peripheral part of the retina which leads to damage the outer nerve axons first as it's will run at the peripheral part of the optic nerve so the middle axons will be pushed against the peripheral axons which will lead to being compressed against the lamella (dense connective tissue) so the peripheral axons will be damaged then the middle axons (the papillomacular bundle the one responsible for central vision will be last affected)
  - The retinal ganglion is divided into superior and inferior retina embryologically, so the axon deviated to the superior and inferior retina so if the damage is at one side that side will defect.



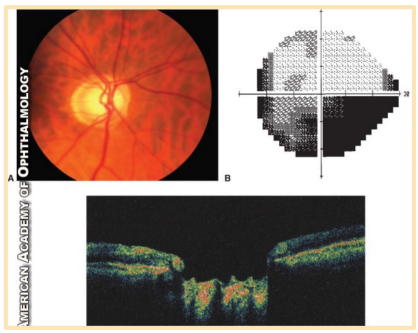




Causes of Chronic Visual Loss

❖ Symptoms:

- ◇ Initially asymptomatic.
- ◇ Glaucoma starts with peripheral (navigational) vision involvement (NO central vision involvement).
  - ■ Much peripheral vision can be lost before the patient notices visual impairment.
  - ■ Patients present late to the clinic when they have progressive loss of vision & their visual field is markedly affected.
- ◇ Usually detected on routine examination.



❖ Signs: IMP!

- ◇ Horizontal visual field defect (vertical visual field loss is more associated with neurological causes of blindness).
- ◇ Optic disc cupping - more details in slide 7
- ◇ Focal damage to the optic nerve.
- ◇ Optic disc hemorrhage
  - ■ When you shine a light to one eye the pupil for both eyes will contract at the same degree normally, but for example, if there is optic nerve damage in the right eye by 50% so the normal eye will constrict more than the left eye اذا عندك 100 رجال يعلمون العين اليسار ان فيه اضاءة واليمين 50 رجال فيصير ال response in the left will be better so it's constructed. This is when we don't alternate the light BUT if we shine the light on the left eye (normal) both eyes constrict with a variant degree, then directing the light to the right eye (the defected one) it's will dilate why? because the nerve damage on the right eye won't receive the same amount of light as the left eye do (relative afferent pupillary defect) but in a normal situation will be no variant as both eyes record the same amount of signals.
- ◇ Most of the world calls Glaucoma "blue water", however in some areas it can be called "black water" مثل السودان or even Swerek Disease مرض سويرق لأنه يسرق النظر بهدوء بدون اعراض

- **Glaucoma triad:**
  - **High IOP**
  - **Characteristic optic nerve head damage**
  - **Visual field loss.** Secondary to nerve fiber layer loss.

- **IOP is the single factor to be controlled (normal is 10-21 mmHg).**
- **Mean= 15.9mmHg ± 2 SD, IOP > 21.7 is abnormal.**
  - In end-stage glaucoma, patients lose their peripheral visual field resulting in what's called tunnel vision. Patients will tilt their heads toward you when you speak to them.

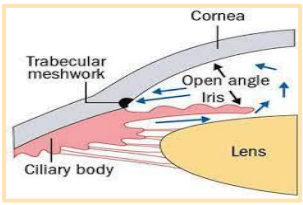
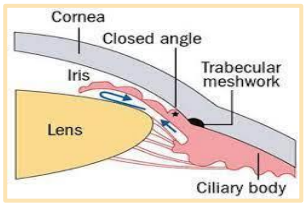
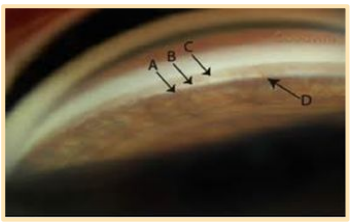
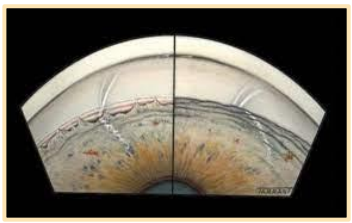
❖ Factors affecting IOP:

- ◇ Age, sex, race, hereditary, diurnal and seasonal variation, blood pressure, obesity, drugs, posture, exercise, neural, hormonal, refractive error, eye movement, eyelid closure, inflammation, and surgery.
- ◇ There is diurnal variation in IOP ranges b\w 4-5 if the variation is >8 → suggestive of glaucoma.

Causes of Chronic Visual Loss

❖ Classification of glaucoma:

According to The Angle

Open = Iris Not covering TM	Closed = Iris covering TM
• In open angel you can see the angel structure • Open angle: trabecular meshwork not occluded by peripheral iris. • The problem with the pores in the Trabecular Meshwork.	• You will not able to see the angel structures • Unless you press the lens the structures will ○ **Mcq's or SAQ:** in peripheral anterior synechiae there will be adhesion between iris and angle.
	
Laser Rx: **SLT**= selective laser trabeculoplasty→ burns TM→ capability of drainage of aqueous increased.	Laser Rx: **PI** = peripheral iridotomy→laser opening behind the iris → aqueous goes behind the iris→pressure will decrease.
	

Treatment
Treatment is aimed at reducing intraocular pressure by 3 modalities available

1. Antiglaucoma medications to decrease the secretion of the Aqueous humor or increase the draining.
2. Laser treatment.
3. If no improvement do surgery, Surgical treatment either:
   a. Cataract
   b. incisional: trabeculoplasty, iridotomy, Canaloplasty and Ahmad implant.
   c. Non incisional.

Factors that influence the retinal ganglion cell health: EXTRA 437

1. Increased intraocular pressure.
2. Excessive glutamate stimulation in the retina.
3. Increase in the inflammatory cytokines secreted by glial and microglial cells.
4. Aberrant immunity.
5. Retinal ischemia.
6. Apoptosis in the trabecular meshwork cells.

Dr. Essam Note, Extra 437.

- Generally, if you have a glaucoma patient in your clinic, you need to decide the type of glaucoma, open or closed angle. Why? For management.
- Patient in which the structures are visible with gonioscopy, but is having high IOP, optic nerve head & visual field changes → open angle glaucoma.
- Patient in which the iris is rolling on the angle structures with gonioscopy lens → angle closure glaucoma.
- Sometimes, you will have the iris covering the angle structures. In this case, press the gonioscopy lens gently on the cornea. Why? We want to force the aqueous in the anterior chamber to go to the angle and push the iris posteriorly. If the angle will open with pressure, and you can see the structures, then this is appositional angle closure. If it opened and you see adhesions, then this is a chronic process and the patient is having a primary angle closure glaucoma.
- Why does the angel close? remember we said the maximum area of resistance in between the iris & lens. Sometimes, patients who have increased anterior posterior diameter of the lens (as in cataract for example), resistance will be higher. So aqueous is accumulating behind the iris. Then it will push the iris anteriorly. Iris will roll against the angle. So, instead of 2 microliters\min coming out, 1 microliter\min is coming out which means the is an accumulation of 1 microliter\min. This will push the iris & close the angel. With chronicity, adhesions will be developed.

Causes of Chronic Visual Loss

❖ Aqueous humor:

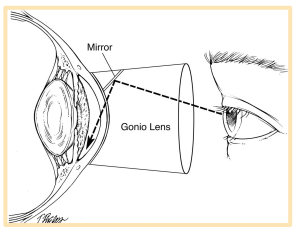
Helpful video

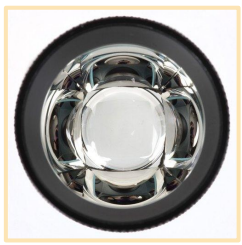
◇ Active secretion:

- Na/K ATPase.
- Cl secretion.
- Carbonic anhydrase (available within the ciliary body and corneal endothelium)
  - The endothelium pumps fluid away from the cornea to keep the stroma dry.
  - The principal physiological function of the corneal endothelium is to allow leakage of solutes and nutrients from the aqueous humor to the more superficial layers of the cornea while at the same time pumping water in the opposite direction, from the stroma to the aqueous.
  - Accumulation of fluid within the stroma will result in a cloudy cornea instead of the normally transparent cornea.
  - For example, a patient underwent penetrating keratoplasty and developed glaucoma. Give the patient medication to stop the action of the carbonic anhydrase pump to reduce the aqueous production. At the same time, the medication will stop the function of the carbonic anhydrase pump within, and fluid will accumulate in the stroma (resulting in a cloudy cornea). In such a case, carbonic anhydrase pump inhibitors are not my 1st option.

◇ Passive secretion:

- Ultrafiltration.
- Diffusion.





❖ Diagnosis:

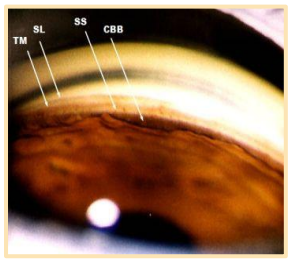
◇ Gonioscopy or Zeiss gonioscopy lens:

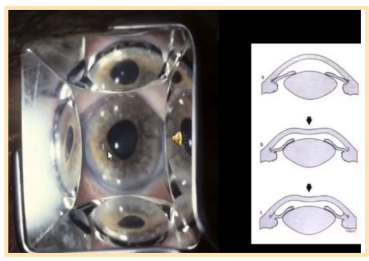
- Are we dealing with open or closed, acute or chronic?
- You should do a **gonioscopy** and check the angle as we can not see the angle by the slit lamp.
- **It is used to view the iridocorneal angle in the case of glaucoma.**
- **Instrument: gonioscope/goniolens. It is used to view the iridocorneal angle in case of glaucoma.**

◇ Normal angel structures (important):

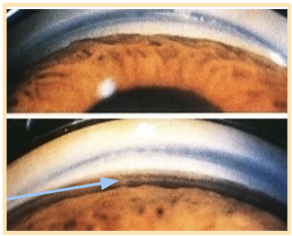
◇ Schwalbe's line (SL).

- Trabecular meshwork (TM).
  - Non pigmented.
  - Pigmented.
- Sclera spur (SS) is the attachment of the sclera and the cornea.
- Ciliary body.
  - **If you didn't see them it's close-angle.**



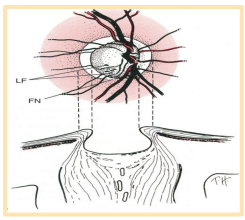


Above: no angel structure seen.
Below: when you press by the lens, it shows you that the angle is open.

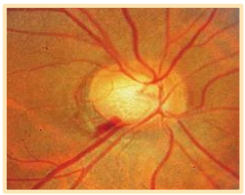


Left: When you put the gonioscopy lens in center, you can see the angle in all quadrants at the same time.
Right: "Indentation gonioscopy", when you indent by these lens you may open a narrow angle. So it can differentiate between open angle, closed angle and narrow or positional angl

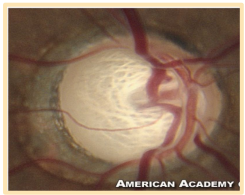
Causes of Chronic Visual Loss



- **Optic Nerve Head (ONH) complex evaluation** [Extra 437]:
  - In clinic do detailed optic nerve exam because you might have:
    - Glaucomatous optic nerve head damage.
    - Anomalous disc.
    - Disc pallor because of CNS or DM.
  - **What to evaluate:**
    - **Disc margin and disc diameter.**
    - **Cup/disc ratio: (important)**
      - How to estimate it? Take the vertical ratio; if more than 0.3 we should worry.
      - **Normal value is 0.3, bigger cup = more nerve tissue loss.**

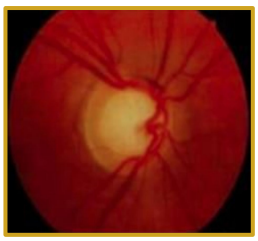


Optic disc

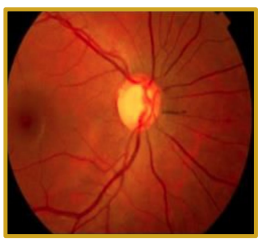


Hemorrhage

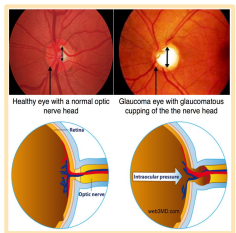
  - Neuroretinal rim: area of axon
    - The cup is almost circular. There are almost 1 million axons passing within the optic canal to go to the brain. These axons will form the optic disc. There is an empty space in the middle because 1 million axons cannot completely fill the canal. The space is called cup. When there is an enlargement of the cup, it means you are losing some axons because of glaucoma.
    - How to assess the cup?
      1. Disc margin.
      2. Neuro-retinal rim. وتناسب نسبة.
    - How much is the ratio of the cup to the disc margin vertically.
      - Disc size
  - PPA (Peripapillary atrophy).
  - Nerve fiber layer (NFL) defect.
  - Optic disc hemorrhage.

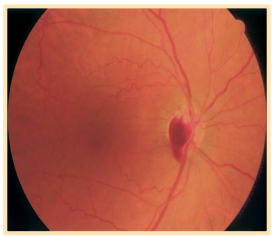


- **Diagnosis:**
  - Disc cupping
- **Investigation to confirm the diagnosis:**
  - Visual field examination.
- **2 types of visual field defect:**
  - Nasal step.
  - Arcuate scotoma.
  - Peripheral visual field defect.



- **Diagnosis:**
  - Disc cupping
- **Next step:**
  - Goldmann applanation tonometry.





- **Open angle glaucoma: Intrinsic defect in outflow tract** [Extra 437]:
  - "In case of open angle glaucoma the angle is anatomically open, but physiologically impaired, why? Because the layer of trabecular meshwork isn't functioning as required, so when the aqueous humor goes from the anterior chamber and circulate between the iris and the lens, it won't go through the trabecular meshwork"
- **To diagnose an open angle glaucoma: (Diagnosis by exclusion)**
  - Measure IOP (you should have high pressure by Goldmann applanation Tonometry)
  - Look for optic disc (optic disc cupping)
  - Visual field (defect respecting the horizontal midline)
  - Gonioscopy (the angle is widely open)

Causes of Chronic Visual Loss

2 Lens "cataract" The most common cause

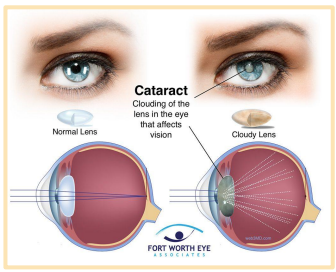
Definition

Opacity of the lens: Lens is transparent, and it contains proteins which are well-organized. When there is disorganization of lens protein = from soluble to insoluble, there will be clouding of the lens (cataract).

Helpful video

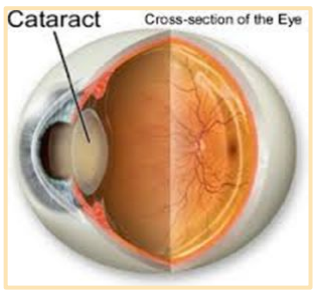
Pathophysiology:

- Disorganization of lens proteins → **opacification**.

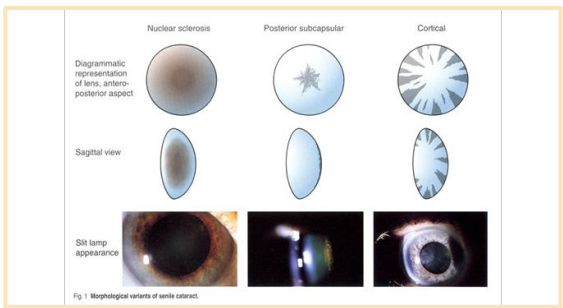


Causes: MCQ

- Age related (Most Common Cause, MCQ)
  - Subcapsular (anterior or posterior).
  - Nuclear and
  - Cortical.
- Metabolic:
  - DM
  - Galactosemia
  - Galactokinase deficiency
  - Fabry's disease
  - Lowes syndrome
  - Hypocalcemic syndrome
- Systemic diseases
  - Myotonic dystrophy
- Traumatic.
- Congenital.
- Drugs: Cataractogenic drugs, Chlorpromazine, Miotics, Myleran, Amiodarone, gold
  - **Steroid (The problem not only develop posterior subcapsular cataract they will develop Glaucoma as well).**
- Inflammation.
- Ocular.



- Complicated cataract:
  - Uveitis, Retinal dystrophy, retinitis pigmentosa, High myopia and Acute glaucoma.
- Intrauterine causes: Rubella, toxo and CMV.
- Syndromes: down syndrome, werner syndrome, rothman syndrome.
- Hereditary: 1/3
- Ocular: Patient with retinitis pigmentosa.



Causes of Chronic Visual Loss

Clinically:

- **Gradual onset of painless visual loss**.
- Visual acuity:
  - Worsening of **existing myopia**
    - In cataract the lens will enlarge & the anterior-posterior diameter increases, so light rays will be focused more anterior to retina).
  - **Correction** of **hyperopia**.
    - In hyperopia, the light rays fall behind the retina; thus, some cataract patients, especially old ones who need near vision, will be happier with nuclear sclerosis, WHY? The diameter increases, light rays fall more anterior into the retina. So old patients who do not drive at night, will be happier & should be informed that they will need glasses for near vision after surgery
- Loss of contrast sensitivity in low light.المريض ما يقدر يفرق بين درجات الألوان, فيحتاج ضوء اقوى عشان يفرق
- Glare in bright light (scatter of light).
  - Classically happens with posterior subcapsular cataract during night drive. Even if the patient has a visual acuity of 20\20; this affects visual quality.
  - The posterior sub-capsular cataract will not allow equal penetration of light through the pupil. The light that passes through areas of condensation of posterior subcapsular cataract will have different intensity than the light that passes freely.

Treatment: MCQ

- **Aphakic**: normal
- **Pseudophakic**: IOL
- **Phakic**: no lens

Indications of cataract surgery:

- Vision loss and Decreased VA
- Patient need
- To prevent complications: phakomorphic glaucoma and phakolytic glaucoma
- To treat diabetic retinopathy

- Congenital: lens aspiration ± IOL
- Acquired: Know the indication only.
  - ECCE + PCIOL (posterior chamber intraocular lens) “very severe cataract”.
    - Open anterior capsule & empty the lens from the opacified proteins; After that, you do ECCE or phacoemulsification then you put an artificial lens.
    - The problem with artificial lens: it does not allow accommodation. But new lenses allow patients to look far, intermediate and near without correction, it is called multi- or tri-focal IOL.
  - Phaco (Phacoemulsification) + PCIOL (Phaco is modified ECCE: small opening and putting a foldable lens).
    - Simply you have an opacified lens proteins you need to exchange the lens with an artificial clear lens).
    - Under local anesthesia to block 3rd, 4th, 6th CN:
      - Retrobulbar anesthesia through inferior orbital rim.

✗ ICCE (Intracapsular cataract extraction) removed lens and cataract and capsule, rare now.

✗ ECCE (Extracapsular cataract extraction) removed center of lens that contains cataract, leave capsule. Bigger incision , more suture , more corneal damage, more infection risk

✗ ECCE IOL (Extracapsular cataract extraction) + (intraocular lens) keeping the capsule in the eye acts as a bag for the artificial lens

✗ **PHACO IOL** (Phacoemulsification) + (intraocular lens) small incision (2-3 mm), break cataract through ultrasound and place a foldable artificial lens. **this is now the preferred method.**

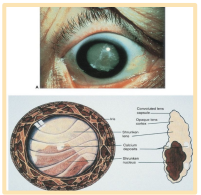
Causes of Chronic Visual Loss

❖ Cataract Clinical Classification (important):

- ◇ **Maturity**
- ◇ **Morphological**
- ◇ **Age of onset**

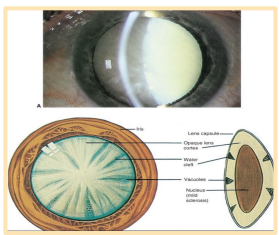
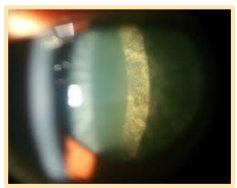
According to The Maturity

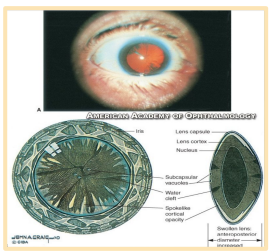
Immature	Mature	Hyper-Mature
• You can see the posterior pole (retina). • Space b/w cortex and capsule. • Counting finger.	• The whole lens is **completely white.** • You cannot see the posterior pole "retina". • The anterior chamber is shallow (narrow) → risk of glaucoma, so do a prophylaxis which is iridotomy. • Only light perception.	• The lens proteins leak through the intact capsule, Lens is composed of nucleus, cortex & capsule. When the cortical proteins start to leak through an intact capsule, the nucleus will sink down within the capsule & the capsule will wrinkle. Urgent case.


Morphologic (According to the Anatomic location)

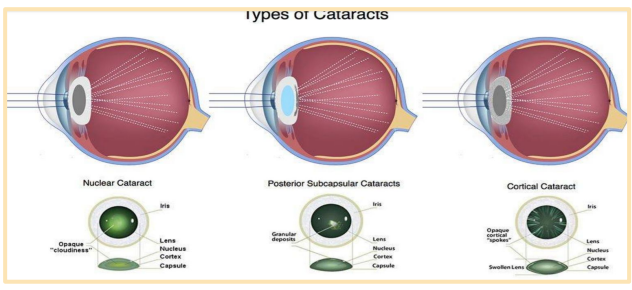
Nuclear	Subcapsular	Cortical
• The lens nucleus itself becomes opacified.	• Nucleus is not significantly affected. • It precipitates (the opacity) in the **posterior** subcapsular area. • **Steroids** mostly cause this type of cataract and **age** Cataract • In bright light patients will not see.	• Cortex is opacified.



According to the Age of onset

Congenital VERY IMP, MCQ	Infantile	Pre senile & Senile
- Hereditary ⅓ - Metabolic - **Syndromes:** Down syndrome, trisomy 13 & 18 - **Intrauterine infection:** Rubella, Toxoplasmosis, CMV		

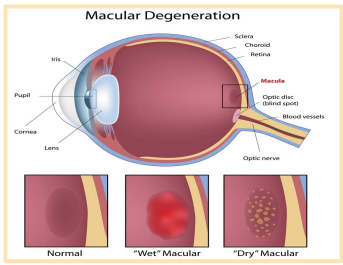


Causes of Chronic Visual Loss

3 Age Related Macular degeneration (AMD)

❖ Introduction:

- ◇ Impaired **central** vision, while, **peripheral** vision **preserved** (opposite to glaucoma).
  - ■ That is why patients with macular degeneration will complain faster than patients with glaucoma.
- ◇ A leading cause of irreversible **central** visual loss (20/200 or worse).
- ◇ Legal blindness **(20/200 or worse)**.
  - ■ It Is known **as age-related macular degeneration** because it comes with age. why in the macula? because it is highly used as all the light are spot in the macula and it's highly vascularized at the **center** to cool down the heats of the macula that why the damage at the center
  - ■ Lipid deposition under the photoreceptors -> toxic reactions. Inflammatory element.

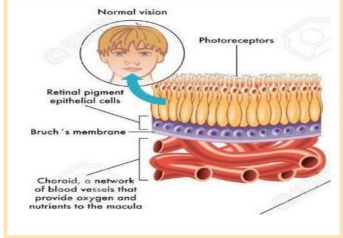


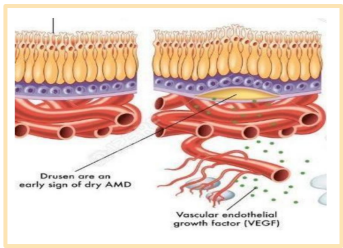
❖ Symptoms:

- ◇ **Metamorphopsia**: vision distorted (كأني أشوف الي قدامي مكسر).
- ◇ **Micropsia**: reduction of the size of objects.
- ◇ **Macropsia**: enlargement of the size of objects.
- ◇ **Scotoma**: visual field loss.

❖ Risk factors: important:

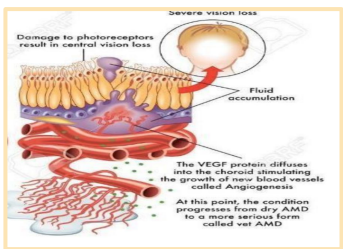
- ◇ **Drusens are a predisposing factor.**
- ◇ **Uncontrollable risk factors**
  - ■ **Age (above 60).**
  - ■ Race (Caucasian).
  - ■ Gender (females).
  - ■ **Genetics (family history).**
- ◇ **Controllable**
  - ■ Smoking, high BP, high cholesterol, poor nutrition, UV light, Unprotected exposure to sunlight, excessive sugar intake, obesity, sedentary lifestyle.





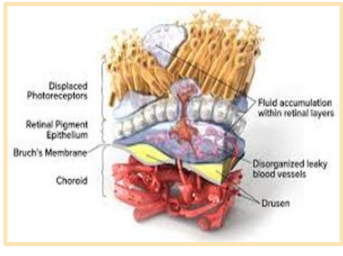
❖ Pathogenesis:

- ◇ **Macular involvement.**
- ◇ Outer retinal layer "photoreceptors".
- ◇ Retinal pigment epithelium (RPE).
- ◇ Bruch's membrane.
- ◇ Choriocapillaris.



❖ Types:

- ◇ Dry: slow progressive atrophy of retinal pigment epithelium (RPE) and photoreceptors.
- ◇ Wet: RPE detachment and choroidal neovascularization.

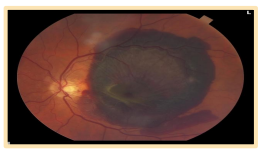


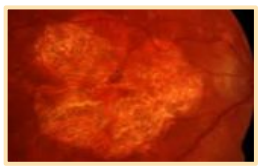
Causes of Chronic Visual Loss

Types

Atrophic = dry	Exudative = wet
• 90% **common**, Slow progressive atrophy of RPE and photoreceptors.	• 10%, RPE detachment and choroidal neova.Blood vessels leak fluid
• Often asymptomatic. • Gradual over years.	• Rapidly progressive (weeks). It can be an acute or chronic cause of visual loss.

• Signs:
  ○ Drusen.
  ○ Geographic atrophy.
  ○ Photoreceptor degeneration.
  ○ Scotoma when light adapting.
	• Signs:
	  ○ Choroidal (subretinal) neovascularization.
	  ○ Preretinal hemorrhage (shiny blood).
	  ○ Elevation of retina because of accumulation of blood
	  ○ Subretinal fibrosis.
	  ○ Metamorphopsia (a classical complain of patients with macular degeneration).
	  ○ Central scotoma (because of blood and scarring within the macular area).
	 

• Drusens are soft, mostly circular & deep.
• Its color is dark similar to that of the normal retina.
 
Dry type ; atrophic and lipofuscin accumulation.
	• Subretinal hemorrhage
	  ○ If choroidal (subretinal) neovascularization bleeds, it will appear dark grey hemorrhage. That means it is under RPE (RPE is always dark in color).
	  ○ If it is bight, that means it is above RPE.
	 

• The accumulation of drusens (lipids) for so many years will create a barrier that prevents oxygenation to reach the retina (photoreceptors) causing **atrophy of the photoreceptors & RPE** resulting in geographic atrophy. Patient then can't see.
 
	• Subretinal fibrosis
	  ○ Remember we said the neovascularization almost always comes with fibrosis.
	  ○ Once the patient has sub-retinal fibrosis, nothing can be done; thus, it needs urgent management.
	 

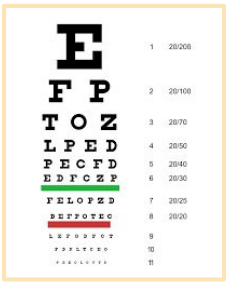
• WE can't treat the patient because photoreceptors already are destroyed. • Ends with macular scar	• We can give the patient some medications to stop these blood vessels to invade the retina like **Anti-VEGF** agents or **lasers**. • the disadvantage of the laser is **scaring.**

Causes of Chronic Visual Loss

Diagnostic Tests

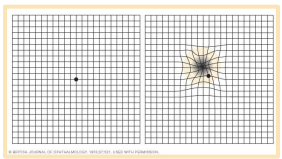
Visual acuity

- Because central vision will be affected first.



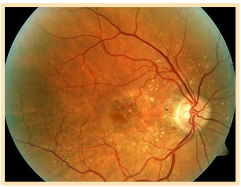
Amsler grid

- If the patient sees wavy lines (Metamorphopsia), it means there is disorganization of photoreceptors secondary to accumulation of drusens. The black spot (blind spot) is caused by accumulated drusens, localized ischemia, localized loss of photoreceptors, hemorrhage & fibrosis.



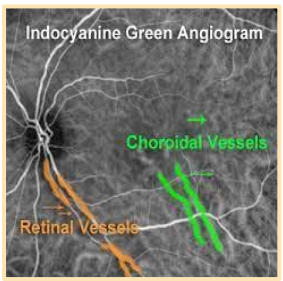
Ophthalmoscope

- Here you can see drusens, geographic atrophy, hemorrhage or fibrosis. Depends on the pt's stage.

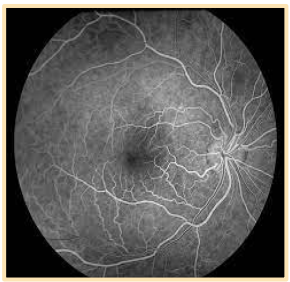


ICG (Indocyanine Green)

- It will show you the choroidal neovascular membrane.

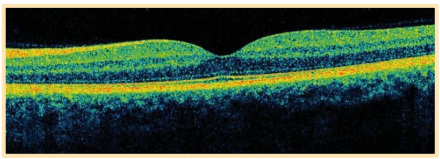


Fluorescein angiography



OCT (Optical Coher Tomography)

- To show subretinal hemorrhages and fibrosis, which is dense homogenous membrane under the retina.



❖ Treatment:

- ◇ If dry:
  - ■ Lifestyle: stop smoking, reduce UV exposure, Zinc & antioxidants.
  - ■ Low-vision aid.
  - ■ Monitoring with Amsler chart to follow up the progression (important).
  - ■ Observation.
  - ■ Laser photocoagulation, for neovascular membrane especially for the wet type.
  - ■ Anti-VEGF agents e.g: avastin, Lucentis & eylea Wet type (stop the retina from signaling VEGF & as a result will stop formation of abnormal blood vessels).
  - ■ You don't need to know this: Verteporfin photodynamic therapy (PDT):
    - Injection of photosensitizer into systemic circulation followed immediately by laser targeting new vessels in macular area.

Causes of Chronic Visual Loss

Extra

Drusens

Drusens (yellowish discoloration):

- lipid products from photoreceptor outer segments, found under retina.
- Drusens are lipid products from photoreceptor outer segment, found under retina.
- Drusens are hyaline nodules (or colloid bodies) deposited in Bruch's membrane, which separates the inner choroidal vessels from the RBE.
- Drusens may be small and discrete or larger, with irregular shapes and indistinct edges.
- Patients with drusens alone tend to have normal or near normal visual acuity, with minimal metamorphopsia
- With age certain people will develop lipid material deposition under the photoreceptors this will cause some sort of toxic reaction to the photoreceptors.
- Normally, the lipid deposits will be drained and absorbed by the choroid. If there is accumulation of lipid deposits, what will happen to the oxygenation from the choroid reaching the retina? decreased> ischemia> atrophy. So the patient won't be able to see.

Neovascularization

With ischemia new vessels from choroid grow into the subretinal space forming subretinal neovascular membrane or preaching the Bruch's membrane
any new blood vessels in abnormal place are ABNORMAL

Hemorrhage

The new vessels are very fragile can bleed easily

- New vessels is bad Either it continues to vitreous causing **vitreous hemorrhages** or it will bleed under the RPE causing sub **RPE hemorrhages** or it will bleed under the internal limiting membrane and causes **subhyaloid hemorrhage.**
- Hemorrhage into subretinal space or even through the retina into the vitreous (significant loss of vision).

Diagnosis: age-related macular degeneration.
2 risk factors: age & smoking.
Name of investigation in pic B: Fundus fluorescein angiography.

Causes of Chronic Visual Loss

Extra

4 Refractive

Causes:

- Myopia, hyperopia or astigmatism.
  - Image will be anterior to Retina.
  - Hyperopia: image will be posterior to Retina.
- Amblyopia (كسل العين)
  - Occurs when the cornea is irregularly shape.

Signs:

- Normal exam
  - Sometimes patients with astigmatism have corneal scar on examination.
- Refraction needed to show errors
  - Everything will be normal except for visual acuity.

Treatment:

- Glasses, Contact Lenses, Refractive surgery
  - There are different types of refractive surgery:
    - Lasik and epilasik: we are maintaining the epithelium & applying it again.
    - PRK: there is scraping of the epithelium by focusing the laser emission on the corneal stroma.
- NB: lenticular causes need cataract surgery.

Occurrence:

- Mostly in young adults.

5 Cornea

Causes:

- Scar
  - Trauma, infection
- Hereditary
  - Corneal dystrophies, **keratoconus** القرنية المخروطية

Signs:

- Corneal scar.
- Bulging cornea.
- Stromal opacities.
- Patients might have some conjunctival injection with chronicity

Causes of Chronic Visual Loss

Extra

Treatment:

- Refraction
- Contact lens (soft or hard)
- Corneal cross linking.
- Keratoplasty زراعة القرنية
  - A cornea is taken from a donor & is implanted in a recipient eye,
    - There are 3 types:
      - Penetrating keratoplasty: implanting all layers of cornea.
      - Lamellar Keratoplasty: transplanting only the stromal and bowman layers.
    - Indications:
      - Keratoconus without scaring or with anterior stromal scarring.

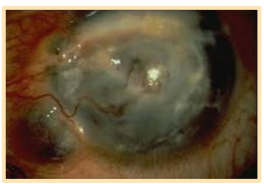
Extra 436

Keratoconus:

- What is the sign? **Munson's sign** is a V-shaped indentation observed in the lower eyelid when the patient's gaze is directed downwards Develop In advance keratoconus
- It is impossible for someone with keratoconus to have hyperopia
- When the lid margin bulges down when someone looks down, this is a sign called Munson's sign that occurs with advanced Keratoconus.
- Advanced keratoconus:
  - Irregular myopic astigmatism.
  - Munson's sign.
  - Apical scarring.

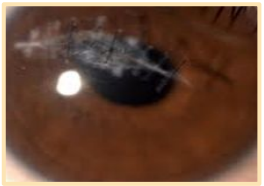
Chronic inflammatory process →chronic vascularization→scaring = Stromal opacities

- Usually the cornea is Avascular tissue so once you have vascularity reaching the cornea you will have leakage of inflammatory mediators→scaring and lipid deposit



Corneal scar

- Penetrating globe injury →underwent primary repair→suturing the wound→ ended up by corneal scar.
- The scaring isn't the issue here you need to maintain the integrity of globe.
- Sometimes the scar is central, within the pupillary area, so corneal haze will obscure the entry of light rays within the pupil, resulting in astigmatism and a barrier toward full light entry through the pupil.
- Also, infection. If you are a contact lens wearer, you might have a corneal ulcer (microbial keratitis). The most common organism in contact lens wearer is pseudomonas. It causes a very rapid corneal perforation

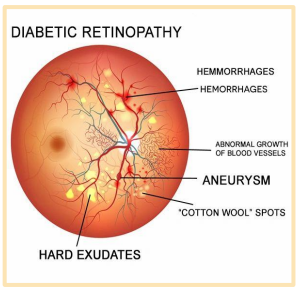


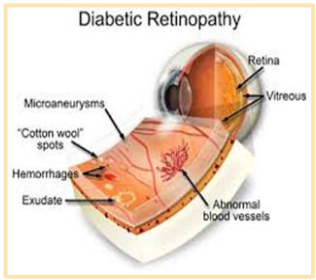
Causes of Chronic Visual Loss

6 Diabetic retinopathy (Important)

❖ Pathophysiology:

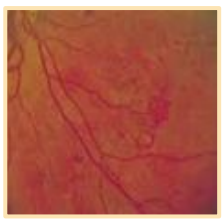
- ◇ **Microangiopathy** which involves precapillary arterioles, capillaries and postcapillary venules.
- ◇ **Two Mechanisms:**
  - ■ **Microvascular occlusion.**
  - ■ **Microvascular leakage.**



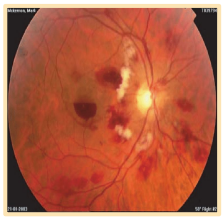


Microvascular occlusion

- **Thick capillary basement membrane**
  - It means lumen is smaller.
  - Abnormal blood vessels (fan-shape)

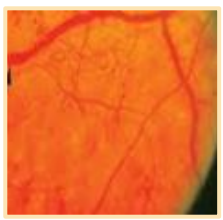


- **Capillary endothelial cell damage with changes in RBCs → Retinal ischemia**
  - → **AV shunt and neovascularization of the retina and around the optic disc.**
    - So the cause of neovascularization of the retina which is seen in proliferative type is related to microvascular occlusion.



- Change in red blood cell
  - and increased viscosity in diabetic patients.

Abnormal blood vessels



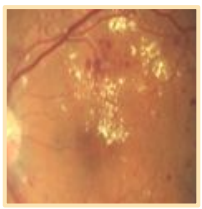
- All of these will lead to decreased blood supply > retinal ischemia > neovascularization and AV shunt formation → vitreous hemorrhage → traction retinal detachment.
- The retinal surface is dry (there should be no fluid within the retina).

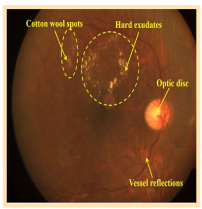
Causes of Chronic Visual Loss

Microvascular <u>leakage</u>

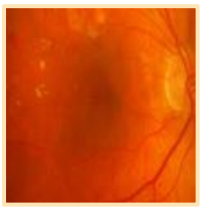
- **Loss of pericytes between endothelial cells**
  - Then there will be gap → plasma leakage into retina (retina should be dry) (with plasma there will be lipid leakage) → exudates and edema

- Exudate of lipid in Retina.
- Ill-defined exudate, no pattern.
- More superficial.



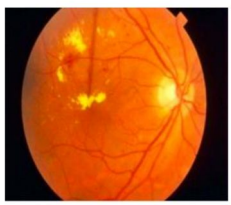


- Retinal Edema



❖ Risk factors:

◇ **Duration of the disease** is the most important factor and not the high blood sugar, pregnancy, nephropathy, poor metabolic control, smoking, HTN, obesity and hyperlipidemia, anemia.



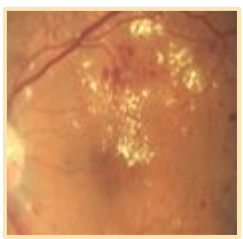
Diagnosis: non-proliferative diabetic retinopathy.
Treatment: Laser photocoagulation.

❖ Types:

◇ Proliferative with or without macular edema
◇ Non-proliferative with or without macular edema.

❖ Clinical classifications

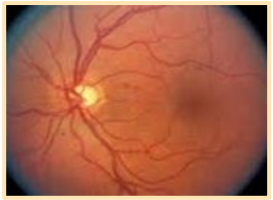
◇ Remember! The hallmark feature of non-proliferative diabetic retinopathy is micro aneurysms; while in proliferative diabetic retinopathy, it is neovascularization.



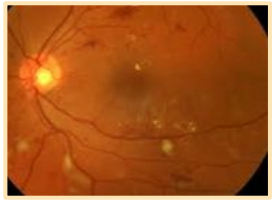
Clinically significant macular edema (CSME)

1. Non proliferative diabetic retinopathy (NPDR):

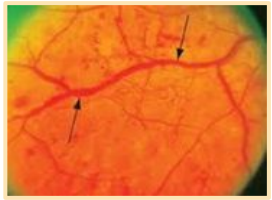
a. **Mild** (micro-aneurisms, the only patients developed).
b. **Moderate** (exudates,hemorrhages as well as micro-aneurysms).
   - Hemorrhage in 1-3 quadrants is considered moderate.
   - Venous bleeding in 1 quadrant is considered moderate.
   - Hemorrhage in 3 quadrants along with venous bleeding in 1 quadrant is still considered moderate.



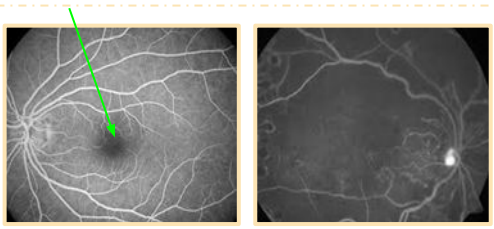
Mild



Moderate



c. **Severe** (hemorrhages in 4 quadrantes, venous bleeding or slugging (means diameter of the vein is increasing) in 2 quadrants, intra-retinal malformation of blood vessels in 1 quadrate).
   - Memorize it like this: "HIV" H: hemorrhages, I: intra-retinal vascular malformation, V: venous bleeding.
   - The intra-retinal vascular malformation is deep within the retina (deep to the arcade).
   - Intra-retinal vascular malformation is also known as intra-vascular micro-vascular abnormalities (IRMA).



Fovea or FAZ (foveal avascular zone)

Fundus fluorescein angiography: Delineation of the vascularity of retina.
A. Normal.
B. Ischemic retina (dye will not pass through the ischemic area).

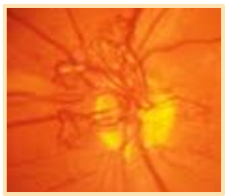
- **Symptoms:**
  - Asymptomatic
  - Decreased visual acuity: clinically significant macular edema & macular ischemia.
  - Causes of visual loss of pts w/ non-proliferative is macular edema or macular ischemia.

2. Proliferative diabetic retinopathy (PDR):

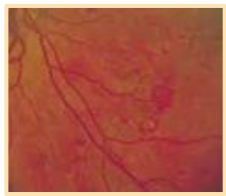
a. **Early** (neovascularization, fan-like and disorganized).
b. **Advanced** (when it progresses & bleeds, it will form fibrosis which is attached to the retina. When it contracts it will pull on the retina causing tractional retinal detachment).
   - Advanced proliferative diabetic retinopathy = there should be tractional retinal detachment.
   - The neovascularization forms on the retinal surface (superficial).

- **Symptoms:**
  - Symptomatic.
  - It can also cause macular edema & macular ischemia.

- **Neovascularization:**
  - A. NVD: neovascularization of the disc.
  - B. NVE: neovascularization elsewhere.

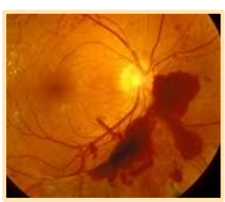


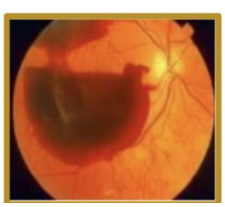
NVD



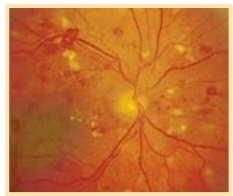
NVE

- Fragile (intra-retinal or vitreous hemorrhage).
- Associated with fibrous proliferation which can cause at a later stage tractional retinal detachment (TRD)





Vitreous hemorrhage.
Cause: Diabetic retinopathy.



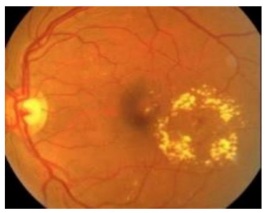
Early

Advanced

Causes of Chronic Visual Loss

❖ Diabetic Macular Edema:

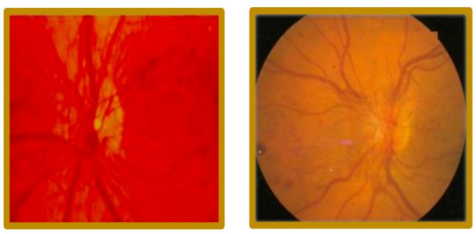
- ◇ Retinal edema threatening or involving the macula.
- ◇ Evaluate: location of retinal thickening relative to the fovea and the presence and location of exudates.
- ◇ In clinically significant macular edema:
  - ■ We have 2 classes because the management is different.
    a. Focal macular edema:
      - ◆ Circinate ring, exudate in a circular fashion.
      - ◆ Treated with focal laser.


Circinate ring in focal macular edema.

    b. Diffuse macular edema:
      - ◆ Haphazardly diffuse exudates and thickening → diffuse macular edema.
      - ◆ Treated with grid laser or injection.
- ◇ Treatment:
  - ■ Laser > For PDR → pan retinal photocoagulation (treat the hole retina except the macula).
  - ■ Intravitreal steroid injection.
  - ■ Intravitreal anti-VEGF injection. (Avastin, Lucentis, Eylea).
  - ■ Pars plana vitrectomy: For patients having vitreous hemorrhage or tractional retinal detachment

❖ Management of diabetic retinopathy:

- ◇ NPDR > Observation (no treatment expect control the blood sugar).
- ◇ PDR > Pan retinal photocoagulation (PRP). The laser changes the ischemic retina to anoxic retina and this will decrease the vascular endothelial growth factor responsible for the formation of new vessels > stops the neovascularization.
- ◇ Macular edema > Focal or Grid laser, injections of anti-VEGF can decrease the edema



Diagnosis: proliferative diabetic retinopathy (neovascularization at the optic disc).
Management: Pan-retinal photocoagulation.

Causes of Chronic Visual Loss

7 Retinitis pigmentosa العشى الليلي

Definition

A group of genetic disorders affecting the retinal ability to respond to light.

- Most are legally blind (central visual field of less than 20 degrees) by 40s. Legal blindness is 20/200 vision & worse

Helpful video

Intro:

- **X-linked recessive:**
  - Males: more often and more severe
  - Females: carry the genes and experience vision loss less frequently.
- **Target photoreceptors** and associated with pigmentary changes in the RPE, which may be primary or secondary to the photoreceptor loss.
- The retina has 2 types of cells that gather light: rods and cones. The rods are around the outer ring of the retina and are active in dim light. Most forms of RP affect the rods first. So night and peripheral vision will be affected. Cones are mostly in the center of the retina, they help in seeing color and fine details. When RP affects them, there'll be slow central vision loss and inability to see colors

Symptoms:

- **Slow loss of vision:**
  - Nyctalopia: loss of night vision, sometimes with progression > tunnel vision.
  - Tunnel vision "loss of peripheral vision".
  - Blindness.
  - Photosensitivity.
  - Low or blurry vision at night especially while driving.
  - Very slow dark-light adaptation.

Signs:

- Visual acuity varies from: 20/20 to no light perception (NLP) at all.
- +-APD (afferent pupillary defect).
- **PSCC** (posterior sub-capsular cataract).
- RPE hyperpigmentation (bone spicules) it is the dark part of the eye, alternate with atrophic regions.
- Attenuation of the arterioles "thinning".
- Waxy pallor of the optic nerve head.
- Cystoid macular edema (CME) in severe cases of RP.

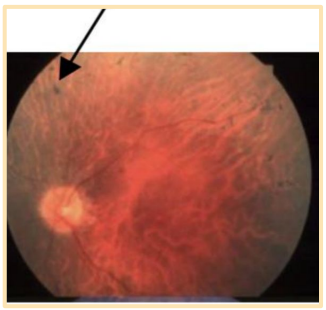
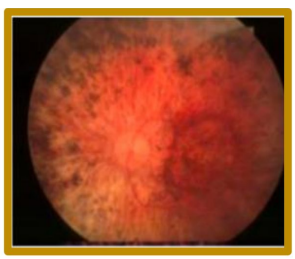
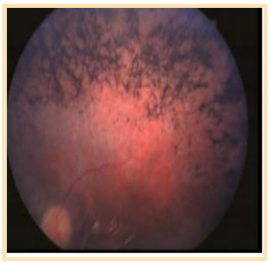
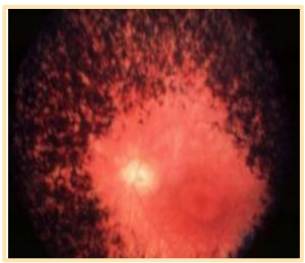
Treatment:

- Unfortunately, nothing can be done to prevent the progression of the disease, but associated ocular problems can be treated:
  - CAI: CME (carbonic anhydrase inhibitor for CME).
  - Vitamins.
  - Cataract: surgery.
  - Low vision aids.
  - Gene therapy

Causes of Chronic Visual Loss

Extra

RP stages and signs

Early stage	2nd stage	3rd stage	4th stage "severe"
• Black spots in peripheral "bone spicules, why? RPE clumping" (black arrow. 	• Bone spicules Increased more and more. • Start to loss photoreceptors. • Waxy disc appearance. • Attenuation of arteriole. • **Diagnosis: RPE hyperpigmentation (bone spicules) & waxy disc appearance.** • **Test: VF, color testing.** • **Treatment: carbonic anhydrase inhibitors.** • **Retinal changes secondary to photoreceptors loss.** 	• More loss of photoreceptor • More clumping of RPE 	• Severe photoreceptors loss • Tunnel vision • Severe RPE loss and clumping • Bone spicules • Waxy pallor • Circumferential involvement of retina. It will spare the macula. • Cystoid macular edema. 

Most Imp complications are:

- **Cataract**
- **Glaucoma**

Causes of Chronic Visual Loss

Extra

Workout:

- Visual field test (it shows constricted visual field & in advanced stage tunnel vision)
- Color testing (mild blue-yellow axis color defects).
- Dark adaptation study (reduced contrast sensitivity relative to VA).
- Genetic subtyping to confirm the presence of the particular gene-defects.
- Optical Coherence Tomography (OCT), to look for (CME), to confirm damage of photoreceptors and to show the extent of their loss. .
- FFA florican angiography.
- ERG electroretinography IMP is an eye test that detects function of the retina including photoreceptors, to confirm the dysfunction or loss of functionality of photoreceptors.
- EOG electrooculography to record eye movement.

Systemic associations:

- **Systemic associations: rule out systemic association**
- Hearing loss & RP:
  - Usher syndrome **(Important)** a condition characterized by partial or total hearing loss and RPE (common).
  - Alport syndrome is a genetic condition characterized by RP, kidney disease, and hearing loss.
  - Refsum disease is an autosomal recessive neurological disease that results in the over- accumulation of phytanic acid in cells and tissues.
- Kearns-Sayre Syndrome **(Important)**
  - External ophthalmoplegia (thus restricted ocular motility).
  - Lid ptosis (unilateral or bilateral).
  - Heart block.
  - Pigmentary retinopathy.
- Abetalipoproteinemia is a disorder that interferes with the normal absorption of fat and fat- soluble vitamins)↓vitamin A →RP
- Mucopolysaccharidoses.
- Bardet-Biedl syndrome genetic disorder characterized principally by obesity, RP, pigmentosa, hypogonadism, and kidney failure in some cases.
- Neuronal ceroid lipofuscinosis lysosomal storage disorders characterized by dementia, RP, and epilepsy.

Causes of Chronic Visual Loss

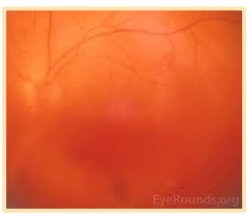
Extra

8 Vitreous

Causes

Vitreous hemorrhages

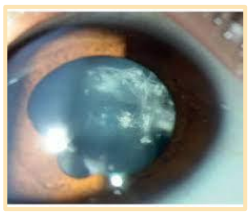
- Trauma
- Uveitis
- PDR*
- PR**



Vitreous hemorrhage, you cannot see the posterior pole

Vitreous **condensation, opacification**

- Chronic inflammation
- PVD***



Vitreous condensation (flying fly)

Vitritis

- Uveitis
- Treatment: treat the cause
- if Uveitis: steroids
- if Vitreous hemorrhage: observation if not then vitrectomy

* **PDR** (Proliferative diabetic Retinopathy) DM is the most common cause
** **PR** (Proliferative retinopathies in general)
*****PVD** (Posterior vitreous detachment)

Extra

Dr. Essam Note 437

Imp for the exam

- **What other symptoms will appear to a patient with chronic visual loss?**
  - Increase in severity
  - Loss of visual acuity or field
  - Involves both eyes -> loss of daily function
  - HE WILL NOT HAVE :
    - NO PAIN (ACUTE)
    - NO RED EYE (ACUTE)

- **How do we see a candle or a car?**
  - Light rays reflected off of the candle in divergence fashion and it will pass freely the cornea -> lens-> focused in the macula.
    - Divergence: light will fan out or spread (in nature, light is always divergence)
    - Convergence : is the physical joining together of light rays. Light rays tend to come together at a point (called point of convergence) from different directions. ex. magnifier

- **What provides the eye with refraction?**
  - Cornea 40 diopters
  - Lens 20 diopters (More important than cornea. Due to accommodation, it can provide up to 40 diopters)

- **What is the average dioptric power of the eye?**
  - 60 diopters

- **Name conditions that can disturb the ocular medium (increases opacity)?**
  - Cataract

- **What are the indications of cataract surgery?**
  1. The patient needs. For example, an engineer or an IT whose work got affected by his condition.
  2. Visual loss.
  3. Therapeutic indications, which means we remove the lens to treat other diseases. Like age-related macular degeneration or diabetic retinopathy. For example, in diabetic retinopathy we might not be able to see the retina bc of the lens so we remove them to treat the retina.
  4. When the lens complicate (Cataract causing Phacomorphic or Phacolytic Glaucoma)

- **How can the cataract cause glaucoma?**
  - When the lens increase in size, it pushes the iris and make pupillary block and angle closure, it's called (Phacomorphic Glaucoma).
  - In leaking mature or hypermature cataract, the protein leaks from the lens into the anterior chamber, it causes anaphylactic reaction, so the macrophages will block the trabecular meshwork >reduce the drainage > leading to secondary open angle glaucoma, (Phacolytic Glaucoma).

Extra

Dr. Essam Note 437 Imp for the exam

- **What is the most important risk factor for Diabetic retinopathy?**
  - Duration of diabetes, (Controlling the diabetes has its effect on the development but not as the duration).

- **What are the types of diabetic retinopathy? And what is the treatment?**
  - Proliferative with or without macular edema.
    - Treatment: Pan-retinal Photocoagulation (PRP).
  - Non-proliferative with or without macular edema.
    - Treatment: observation/no treatment.
    - Treatment of macular edema: Focal & Grid laser, anti
  - VEGF injections (Avastin, Lucentis, Eylea).

- **How does the PRP work?**
  - The laser changes the ischemic retina to anoxic retina and this will decrease the vascular endothelial growth factor responsible for the formation of new vessels > stops the neovascularization.

- **Scenarios:**

35 YO female presented to the clinic with gradual painless decrease in vision, IOP 45, optic disc show total cupping, both eyes. what is the Dx?

- CAG
- **OAG**
- Neovascular glaucoma
- Pigmentary glaucoma

(if they mention afferent pupillary defect (APD) in the scenario usually it is optic neuritis, but it could also be in advanced glaucoma → you can differentiate by IOP if high? glaucoma, low? optic neuritis)

65 YO male complaining of gradual decrease in VA. on examination 20/200 both eyes IOP 15 and pupil is reactive with no fundus view. what is the <u>most likely</u> Dx?

- ARMD (can't determine, no fundus view)
- Diabetic retinopathy (not diabetic)
- Glaucoma (could be normal tension)
- Cataract (could be also :))

85 YO male presented to the clinic with gradual decrease in VA. on examination the vision is 20/200 or CF IOP is normal pupil is normal fundus shows pigmented changes in the macula.
What Imp test to do? Amsler grid
What Imp complaint of Pt?

- **Metamorphopsia**
- Night blindness
- Blurred vision
- Watery eye

OPTHALMOLOGY TEAM

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