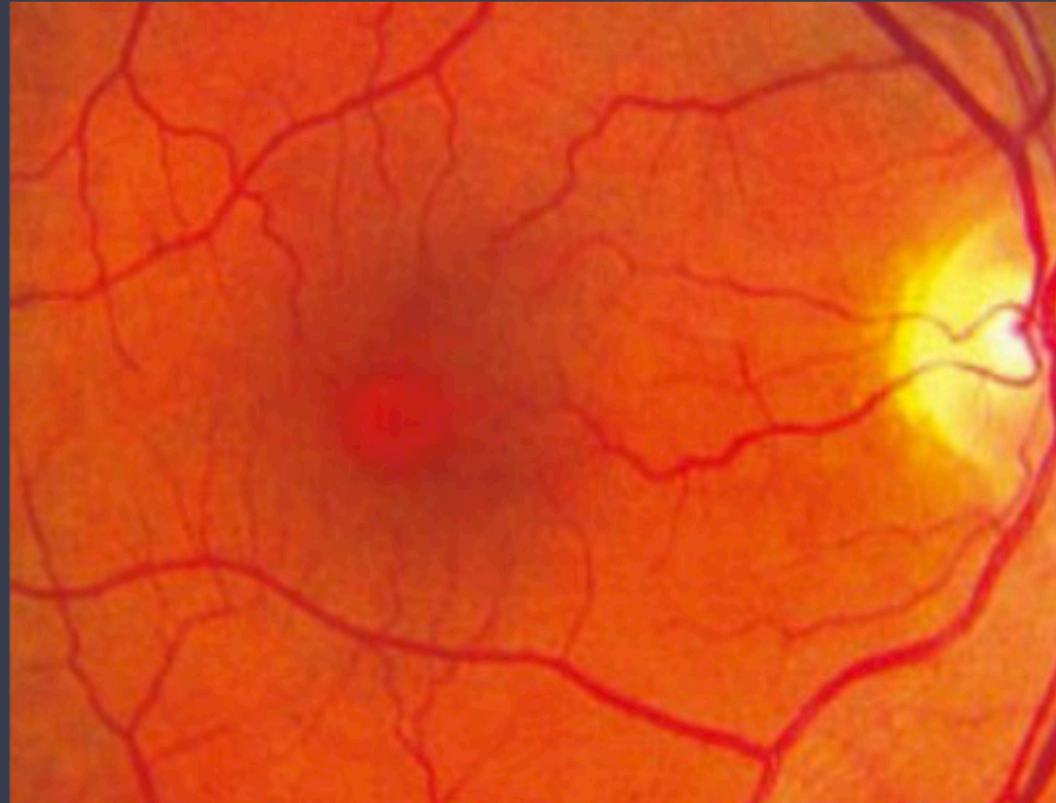


# MACULAR HOLE



EyeToday



Dr Praveena M

# MACULAR HOLE

**Definition** : A **round full thickness dehiscence** in the retina at the location of the fovea that **involves all the layers of the retina**, from the internal limiting membrane through the outer segments of photoreceptor layer.

## **Classification** :

- **Primary macular hole** - is commonly an idiopathic macular hole which is caused by abnormal vitreofoveolar traction
- **Secondary macular hole** - most common due to orbital trauma and high myopia

## **Incidence :**

- 1.7 per 1000 population according to the study in southern India
- 3.3 per 1000 population in Baltimore eye study
- 0.2 per 1000 population in Blue Mountain study
- 0.9 per 1000 population in Beijing study
- **Female predominance**

## **Risk factors:**

- Age group - more than 65 years
- Hypertension.
- Diabetes mellitus.
- Coronary artery disease.
- Previous cerebrovascular accident.
- Elevated plasma fibrinogen level



<u>Year</u>	<u>Author</u>	<u>Description</u>
1869	Knapp	First case <b>description of macular hole (traumatic)</b>
1871	Noyes	First detailed clinical description of macular hole (traumatic)
1900	Ogilvie	Published case series & proposed terminology including macular hole
1900	Kuhnt	Atraumatic theories of cystic retinal degeneration leading to macular hole
1901	Fuchs	Early histopathologic descriptions of macular hole including cystic retinal changes
1907	Coats	
1912	Zeeman	Histopathologic recognition of premacular vitreous condensation
1924	Lister	<b>Vitreous forces and “vitreous traction bands” (anteroposterior) may cause macular holes</b>
1967	Reese et al	Vitreous separation critical to macular hole formation
1982	McDonnell et al	Possible female hormonal influence on vitreous separation and macular hole formation
1983	Avila et al	Vitreous separation not necessary in formation of a macular hole
1986	Morgan & Schatz	Involutional macular thinning is a premacular hole condition
1988	Gass & Johnson	<b>Tangential vitreous traction and Gass biomicroscopic classification of macular hole</b>
1995	Gass	Centrifugal displacement of retinal receptors with umbo dehiscence Reappraisal of biomicroscopic classification of premacular hole and macular hole lesions

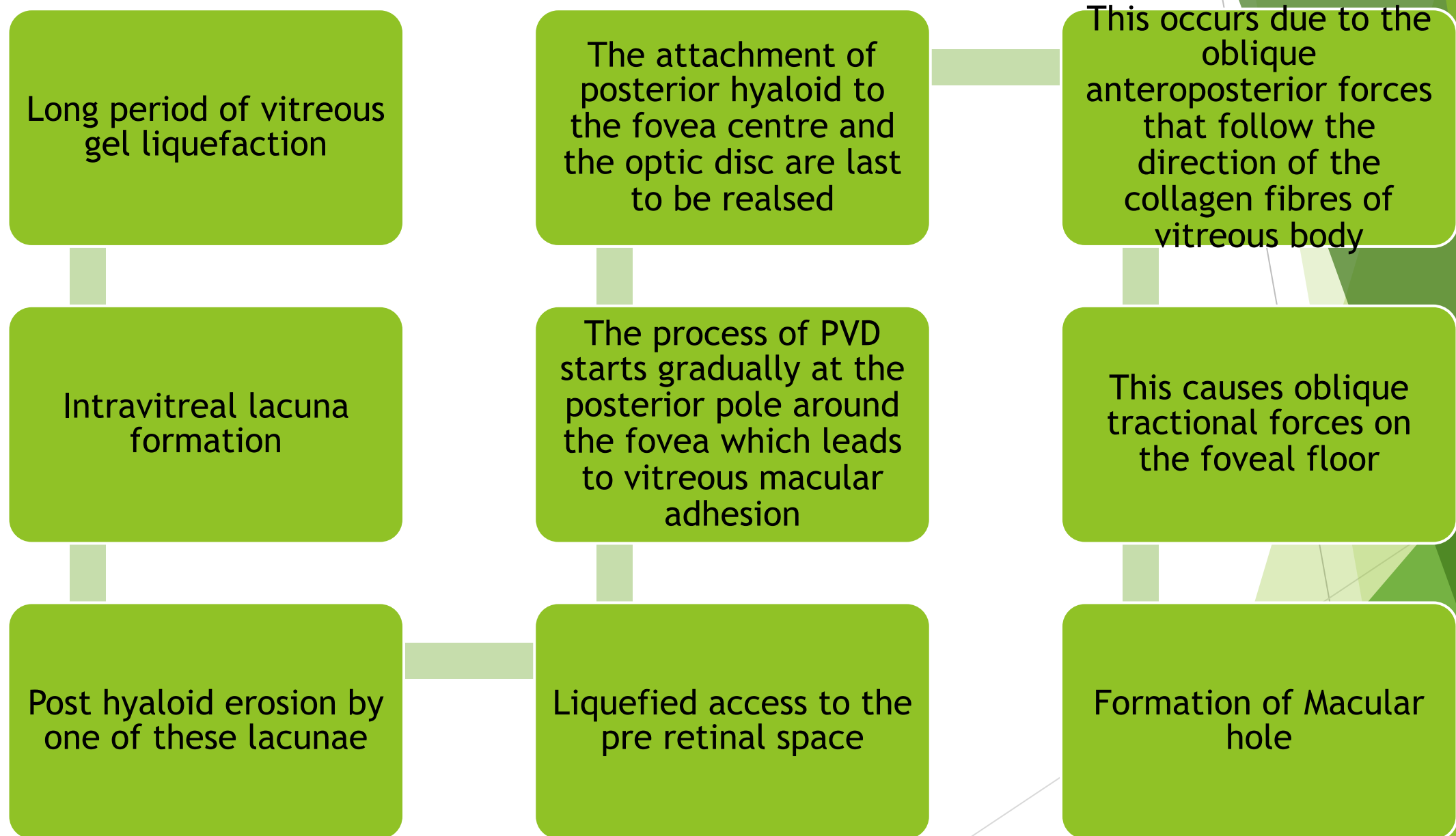
# Etiology

- **Idiopathic** : This is the most common cause and is without a known predisposition. This variety is commonly noted in middle aged females
- **Post traumatic** : This follows blunt trauma. Macular holes after injury can be associated with choroidal rupture or RPE damage in the vicinity of the macular hole.
- **Chronic cystoid macular edema** : CME of long duration of any etiology (diabetic macular edema, chronic inflammation, etc.) can lead to formation of macular hole.
- **Associated with rhegmatogenous retinal detachment** : Retinal detachment can be associated with macular hole as an isolated feature or along with other peripheral holes.
- **High myopia** : causes macular Retinal detachment
- **Others** : Hypertensive retinopathy, lightning strike or YAG laser which causes impaired choroidal perfusion. More common after YAG laser capsulotomy especially in high myopic eye.

# Pathogenesis

- **Vitreous traction**- anteroposterior and tangential force plays an important role in the pathogenesis of macular hole
- **Vitreoschisis** also contributes to the formation of the macular hole
- **Gass** has postulated that the **tangential contraction of the prefrontal posterior hyaloid membrane resulted in the detachment of the central photoreceptors and then in the opening of the fovea.** The process of macular hole formation was divided in four stages that are used still today.
- A new OCT based classification has been published by the **International Vitreomacular Traction Study (IVTS)**

# Pathogenesis:



# Stage 0 macular hole

- **IVTS Classification** : Vitreomacular adhesion - VMA
- **OCT features:** Perifoveolar detachment (oblique) of posterior hyaloid with normal fovea contour
- No clinical changes occurs at this stage



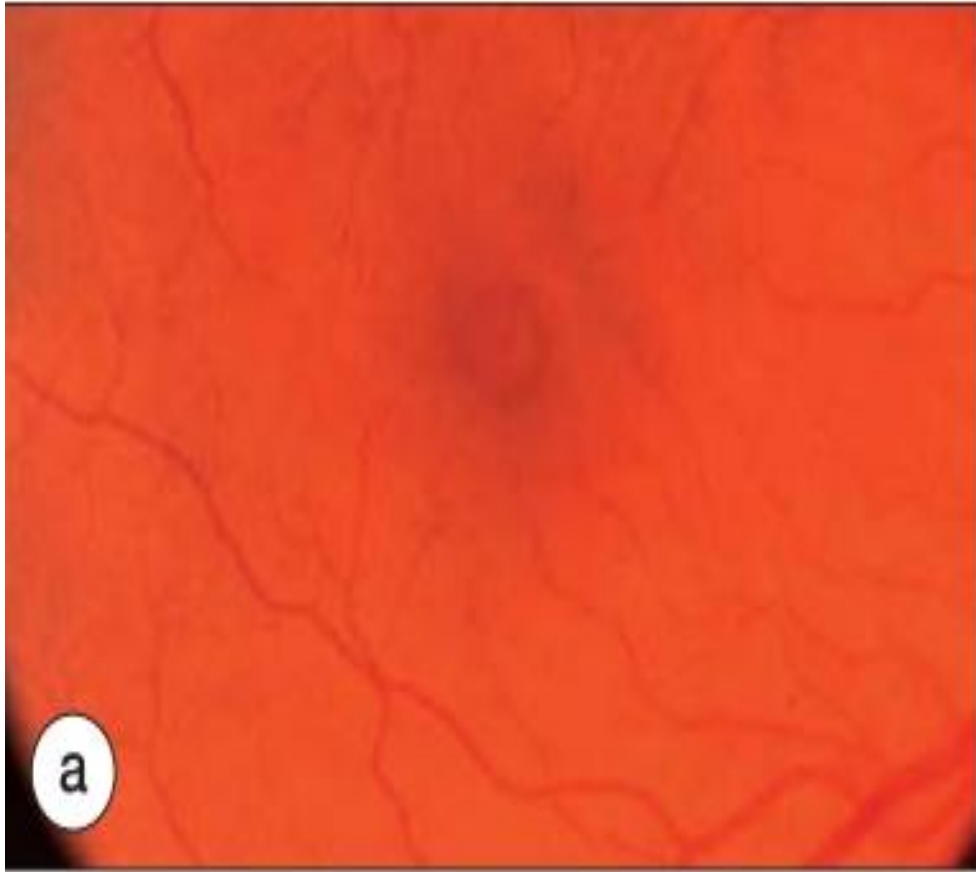
# Stage 1A

- **Gass classification** : Impending macular hole
- **Signs** : Central **yellow spot**, loss of foveolar depression, no vitreofoveolar separation
- **Pathology**: Early serous detachment of foveolar retina. The inner retinal layers detach from the underlying photoreceptor layer, often with the formation of cyst like schisis cavity
- **IVMT classification** : Vitreomacular traction- VMT
- **OCT features**: Perifoveolar detachment of posterior hyaloid. Foveal cyst in the inner fovea
- **DD for foveal yellow spot**- Adult vitelliform macular dystrophy, solar and retinal pointer retinopathy, CMO

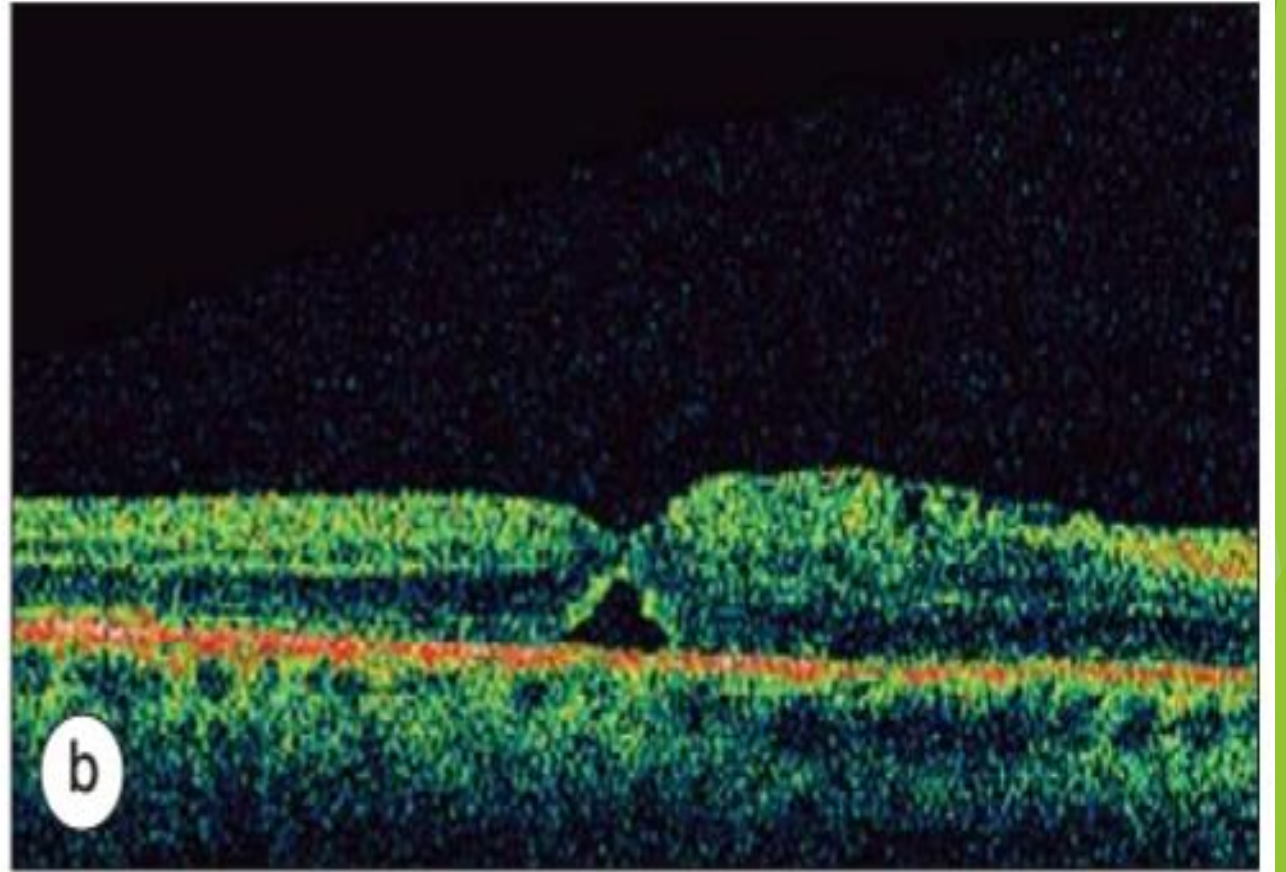
# Stage 1B

- **Gass classification** : Occult macular hole
- **Signs** : **Yellow ring** with bridging interface, loss of foveolar depression, no vitreofoveolar separation
- **Pathology**: Serous foveolar detachment with centrifugal displacement of xanthophyll or photoreceptor layer due to loss of structural support.
- **IVMT classification** : Vitreomacular traction- VMT
- **OCT features**: Perifoveolar detachment of posterior hyaloid. Foveal cyst extending in the outer retina, causing break in the photoreceptor layer.

# Stage 1



Central yellow ring

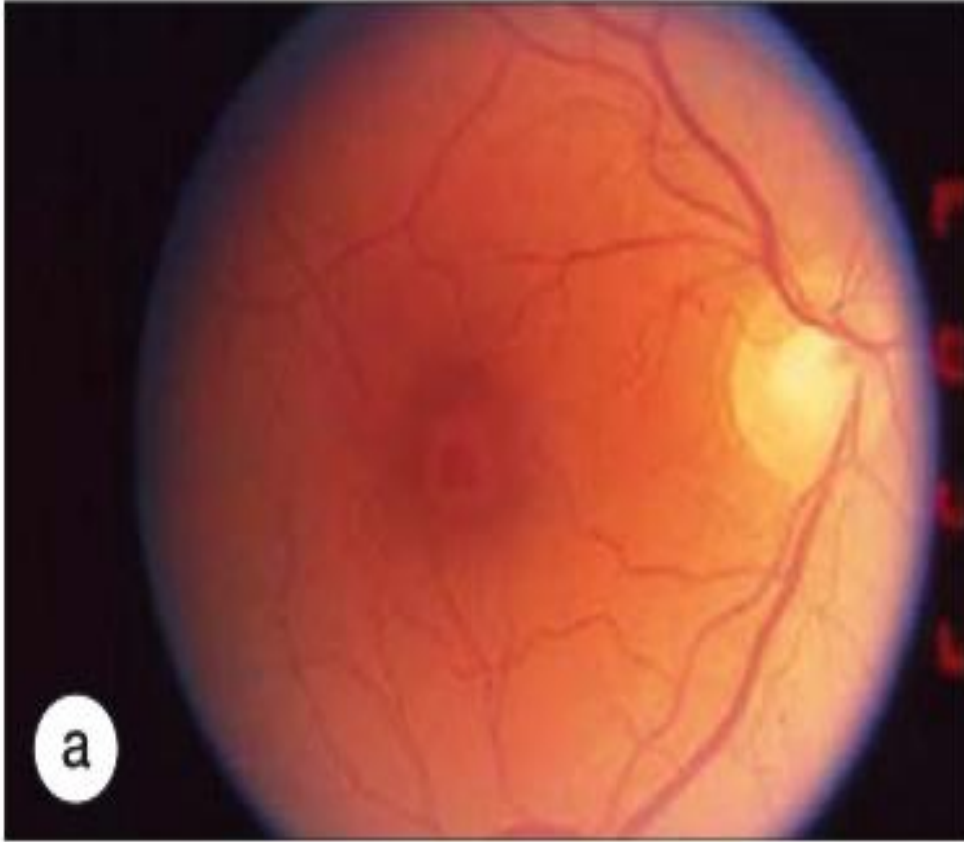


Foveolar detachment

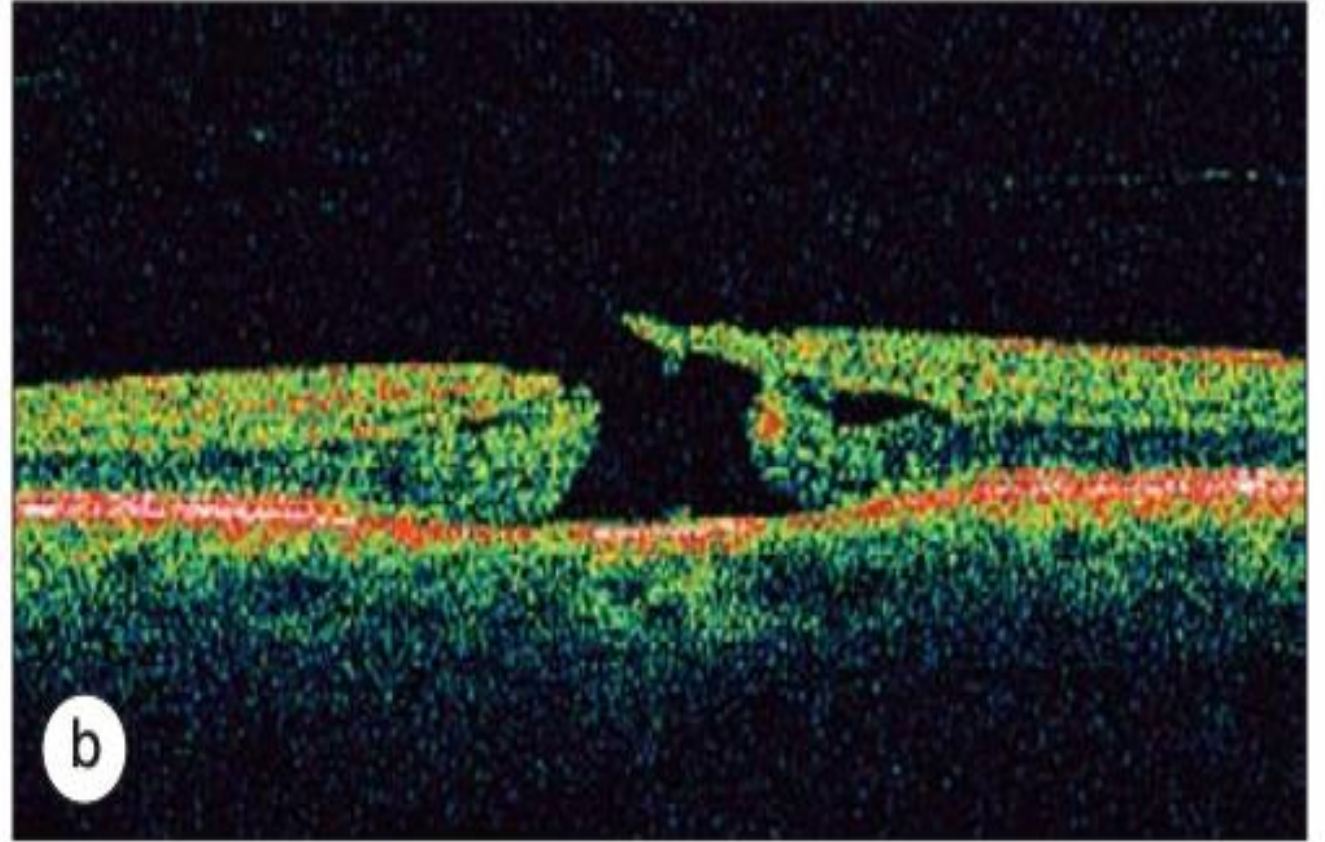
## Stage 2

- **Gass classification** : Small full thickness hole
- **Signs** : **Eccentric oval, crescent or horseshoe retinal defect** inside the edge of yellow ring. The full thickness macular hole defect is less than 400  $\mu\text{m}$  in diameter
- **Pathology**: Tear or hole in contacted prefoveolar vitreous bridging round retinal hole, no loss of foveolar retina
- **IVMT classification** : Small or medium Full thickness macular hole (FTMH) with VMT
- **OCT features**: Partial opening of the roof of the cyst, the operculum staying still attached to the edge of the hole. Partial detachment of the posterior hyaloid, which is still attached at the operculum. The operculum contains retinal elements.

## Stage 2



Full thickness hole of diameter less than 400  $\mu\text{m}$



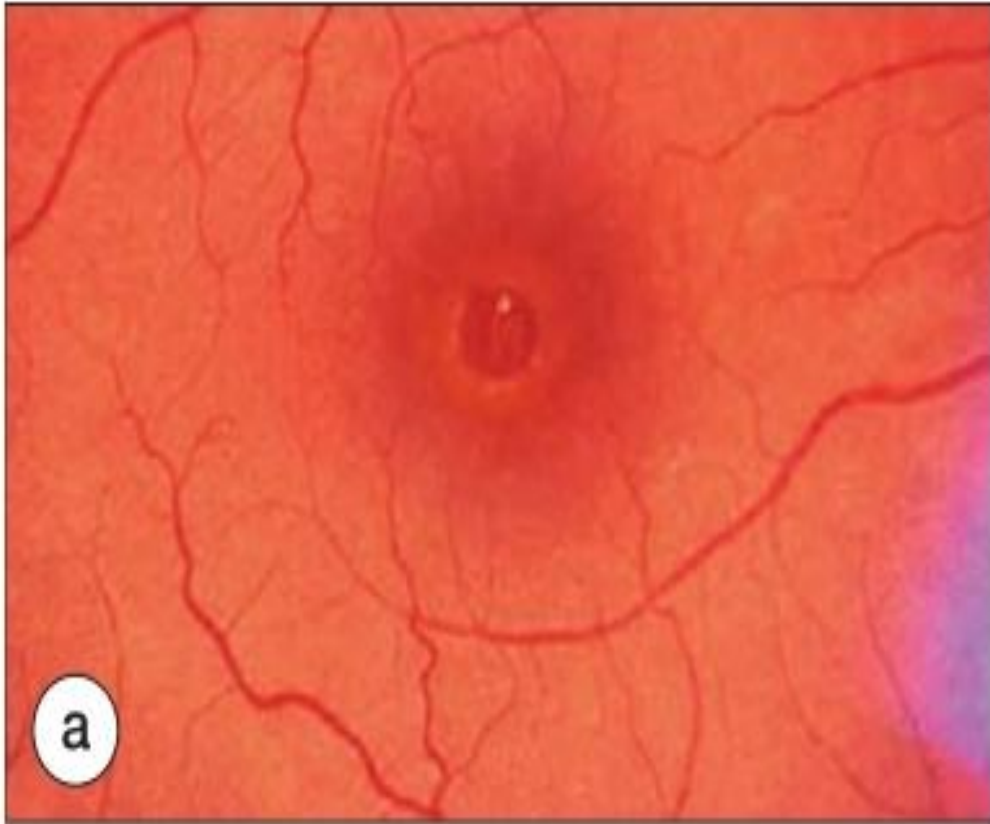
Vitreous traction on operculum which is not yet detached from the retina



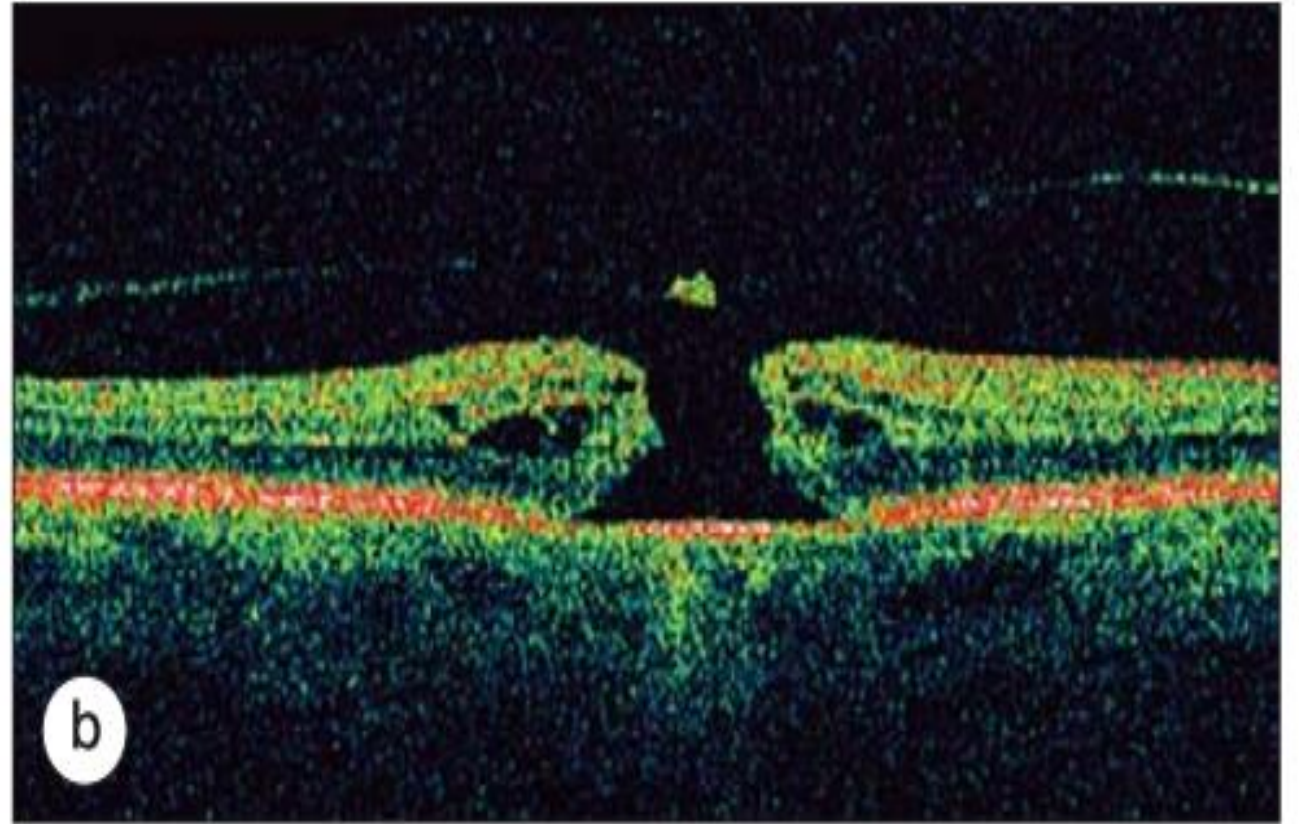
## Stage 3

- **Gass classification** : Full size macular hole
- **Signs** : Central round  $> 400 \mu\text{m}$  in diameter retinal defect with a red base in which yellow white dots are seen. A surrounding grey cuff of sub retinal fluid is present and an overlying retinal speculum (sometimes called pseudo operculum) may be visible. No Weiss ring, rim of elevated retina with or without prefoveal opacity.
- **Pathology**: Opercula consists of glial tissue and condensed vitreous cortex (40% contains photoreceptor elements), no posterior vitreous detachment
- **IVMT classification** : Medium or large FTMH with VMT
- **OCT features**: Posterior hyaloid detached from the macular surface but still attached to the optic disc, most often containing an operculum

## Stage 3



Full thickness hole of diameter more than 400  $\mu\text{m}$  and has an overlying pseudo operculum



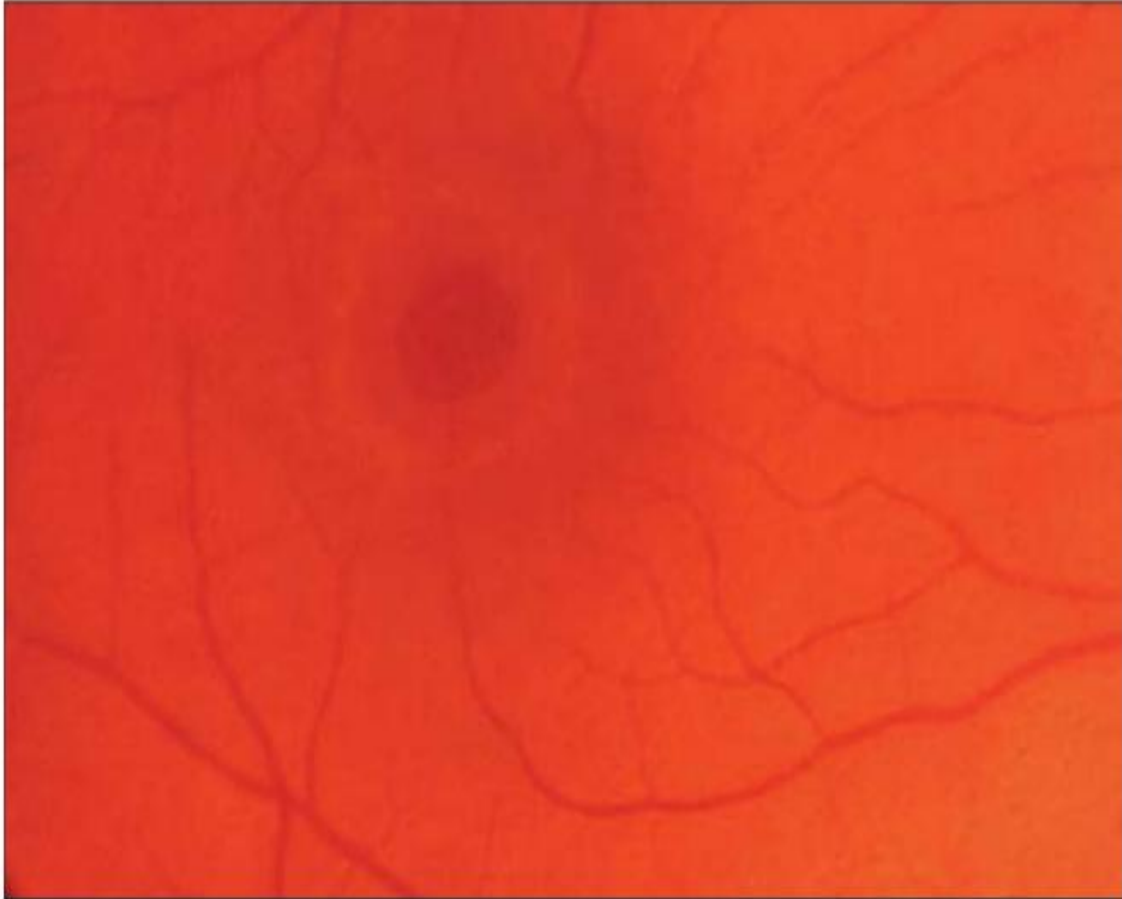
The operculum is detached from the retinal surface and attached to the posterior vitreous

# Stage 4

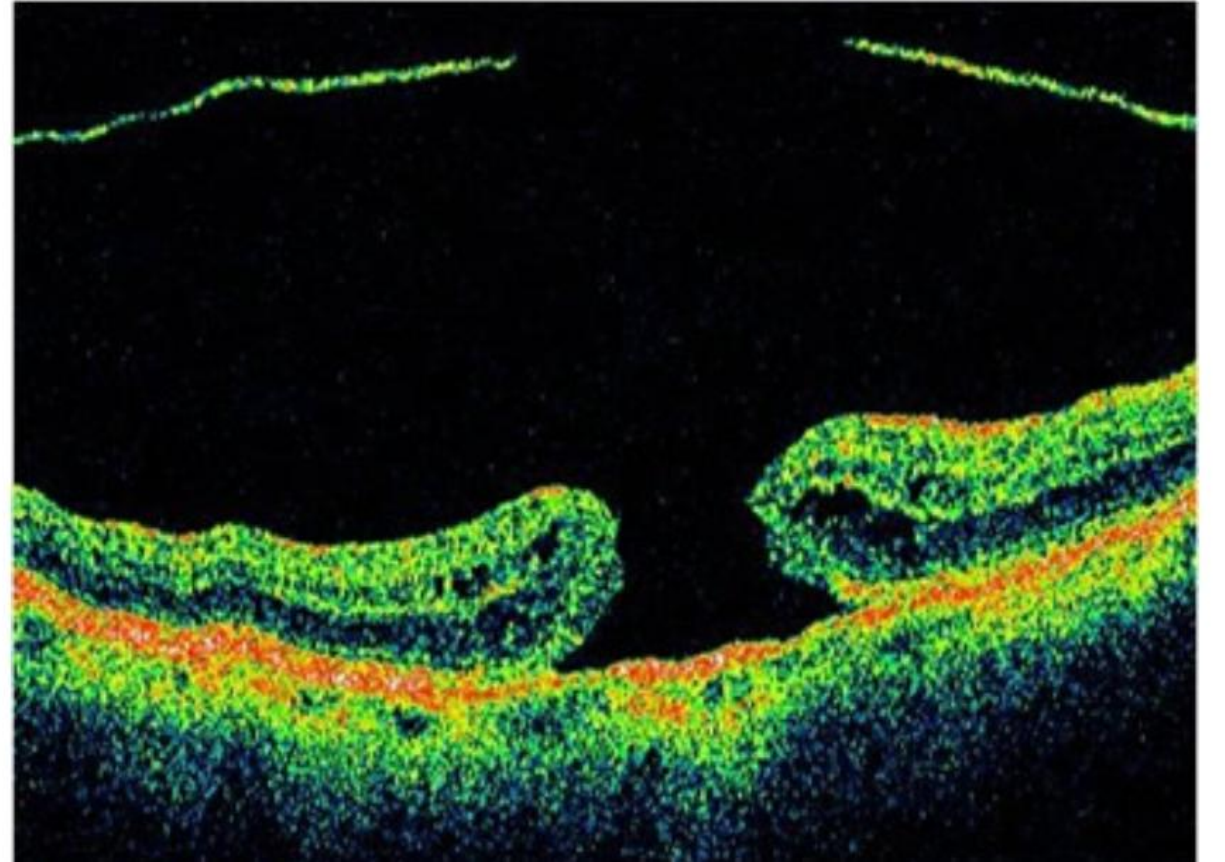
- **Gass classification** : Full size macular hole with complete PVD
- **Signs** : **Central round defect, rim of elevated retina.** Weiss ring with prefoveal opacity, may be associated with epiretinal membrane
- **Pathology**: Hole with pseudo operculum and posterior vitreous detachment from optic disc and macula
- **IVMT classification** : Small, medium or large FTMH without VMT
- **OCT features**: Posterior hyaloid is not visible



# Stage 4

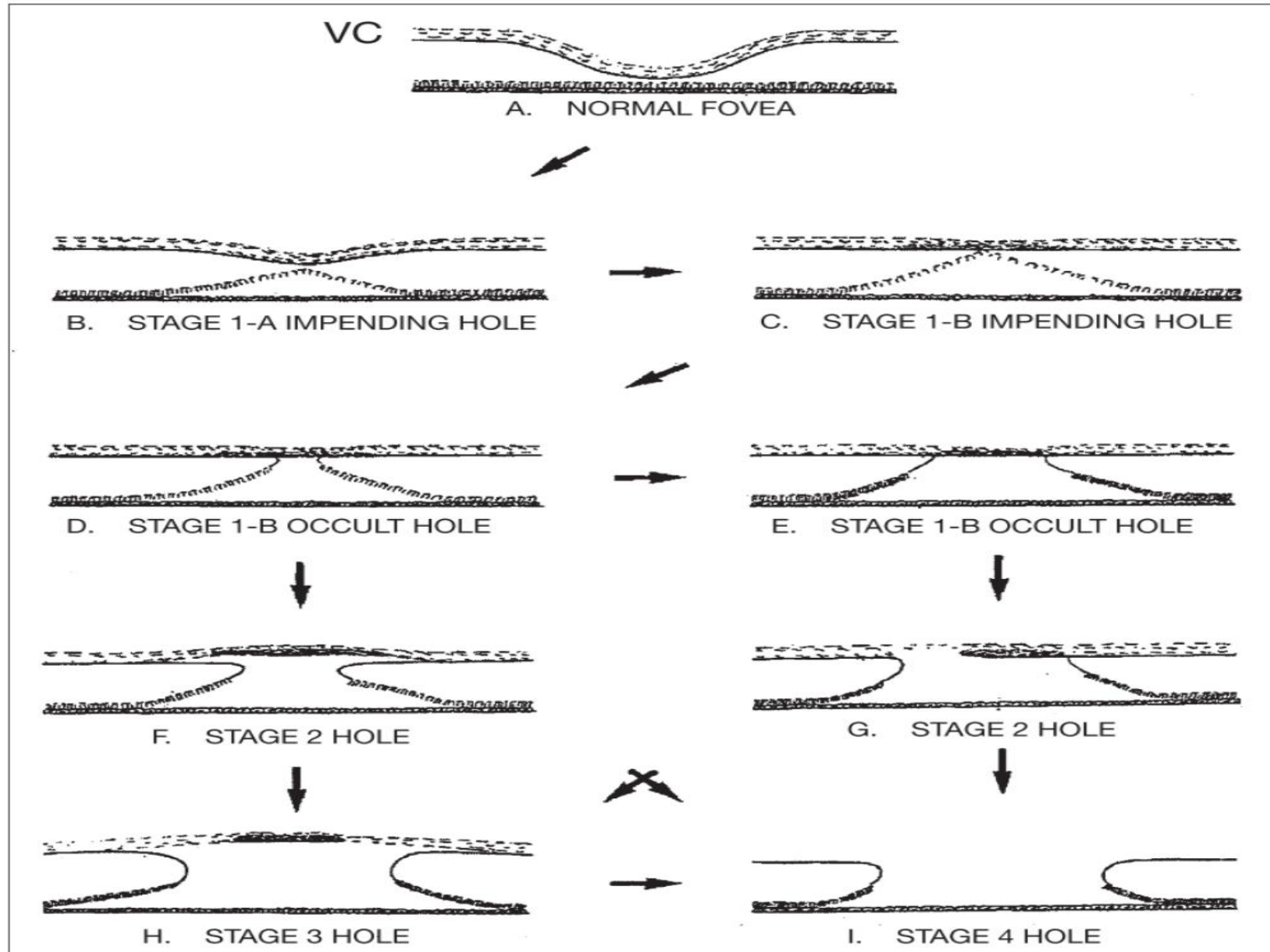


Central round defect



The posterior hyaloid face has detached off the surface of the retina

# Schematic diagram of staging of macular hole



# Clinical features

- Impairment of central vision
- Metamorphopsia

## Visual acuity:

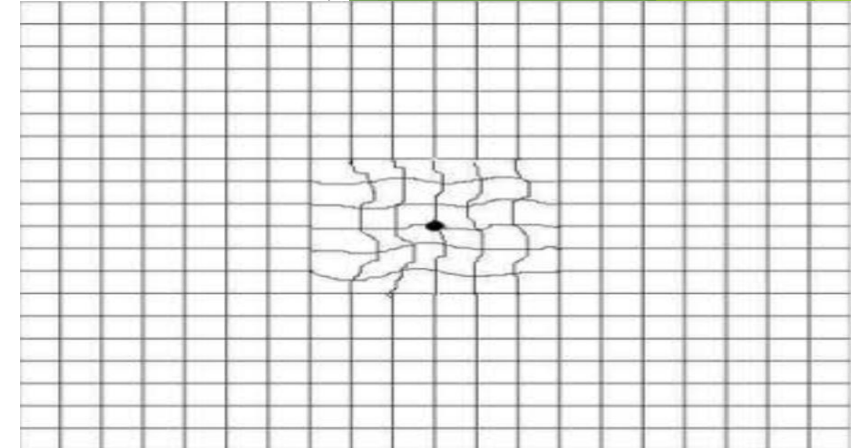
- Stage 0 - normal visual acuity
- Stage 1 and stage 2 - 6/9 to 6/12
- Stage 3 - 6/24 to 6/60
- Stage 4 - 6/60



# Investigations

## Amsler grid test:

- This usually shows non specific central distortions rather than a scotoma



## Watzke-Allen Test: -

- Performed by projecting a narrow slit beam over the centre of the hole both vertically and horizontally with a 90D or 78D lens.
- Patients with macular hole will report that the beam is broken or thinned.



## Laser aiming beam test:

- Performed by projecting a 50  $\mu$  spot of a laser aiming beam (e.g. He-Ne) at the centre of the hole.
- A patient with macular hole will report that the spot has disappeared

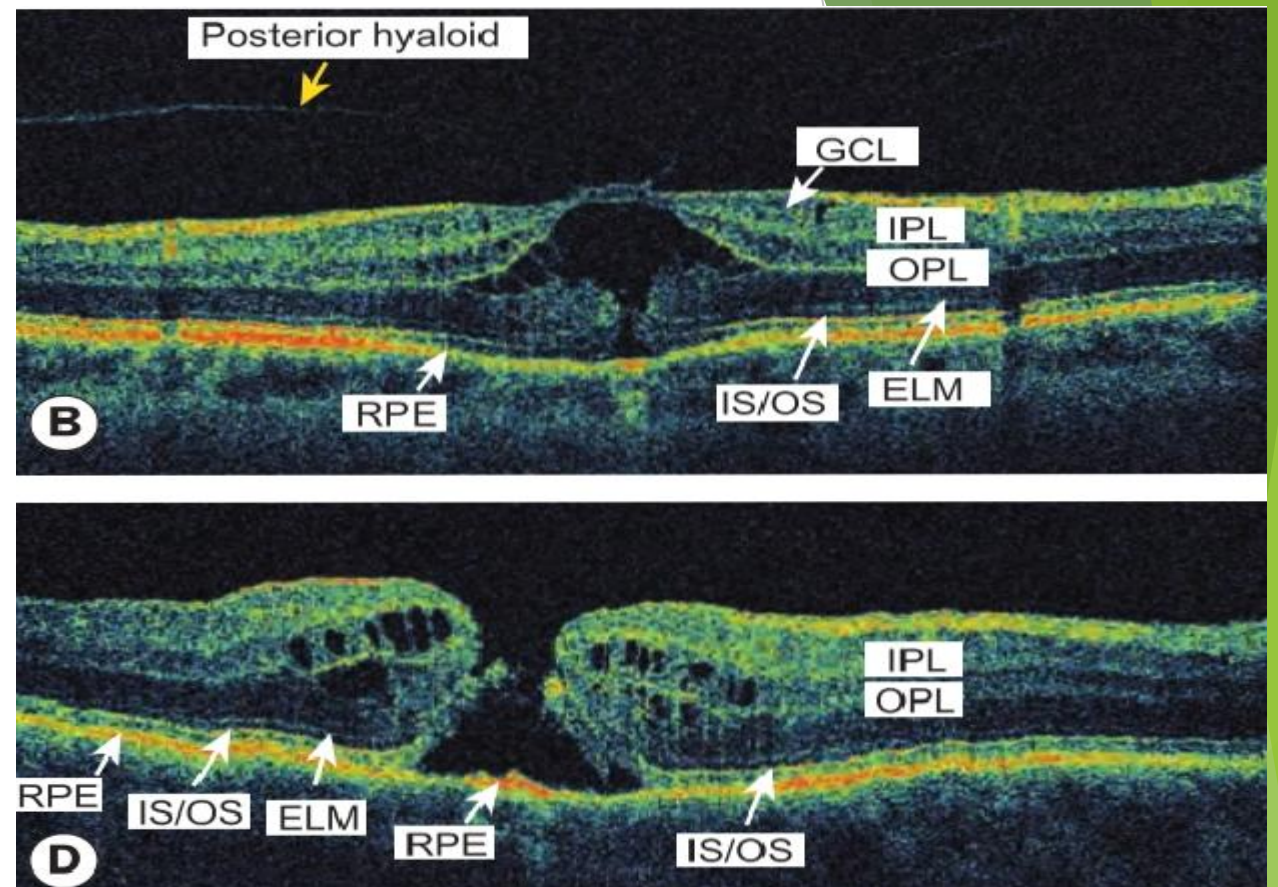
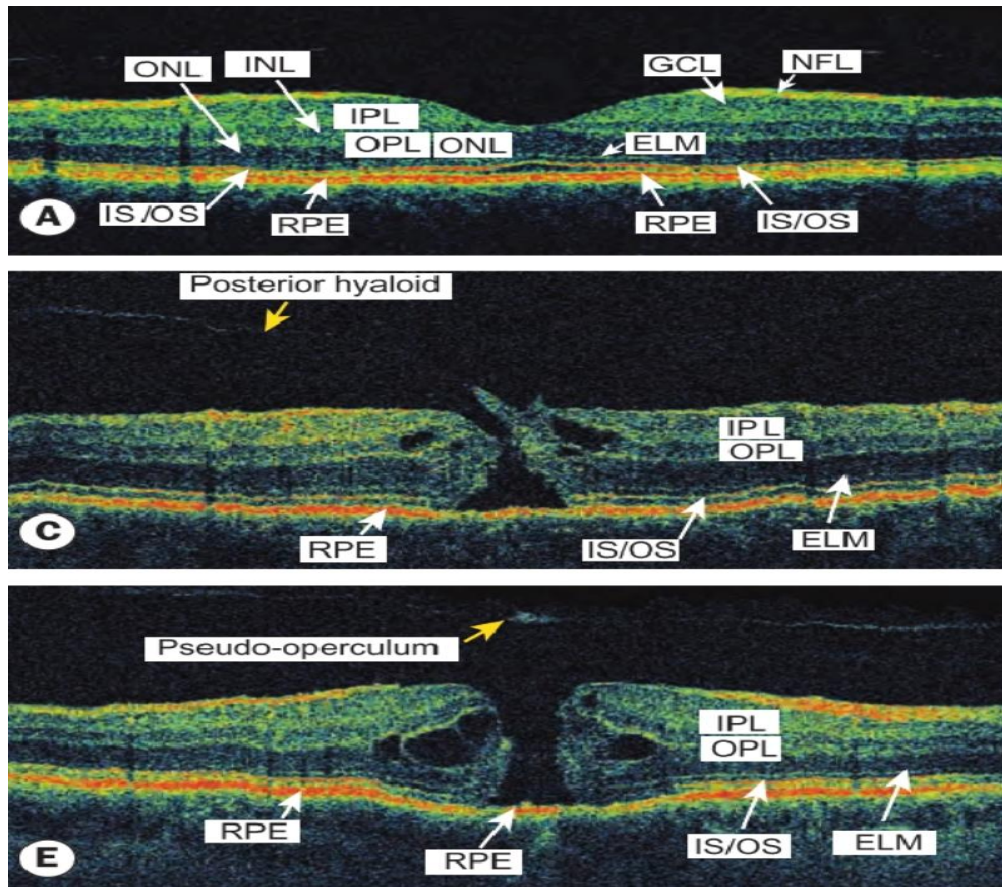
## **Optical Coherence Tomography :**

- OCT is helpful for evaluating the vitreoretinal interface and determining the degree of traction from vitreous or ERMs.
- It is also useful in staging macular holes, differentiating from pseudo- or lamellar holes, and evaluating for progression

## **Fluorescein Angiography:**

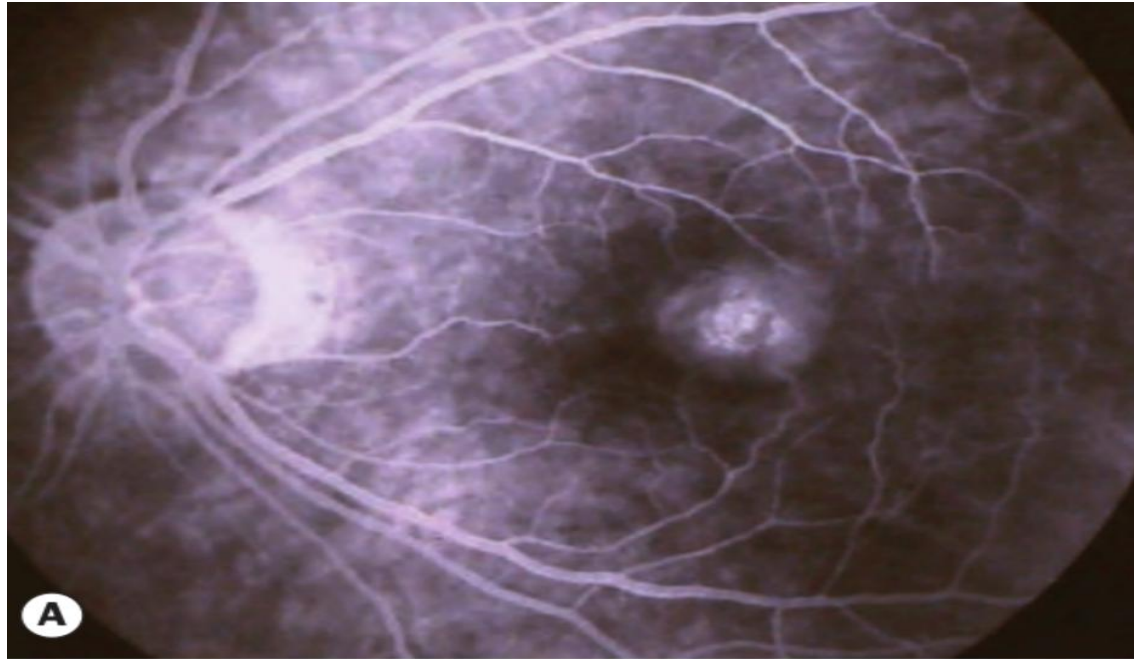
- Shows early early well defined window defect.
- This result from unmasking of background choroidal fluorescence caused by a window defect in xanthophyll due to centrifugal displacement.



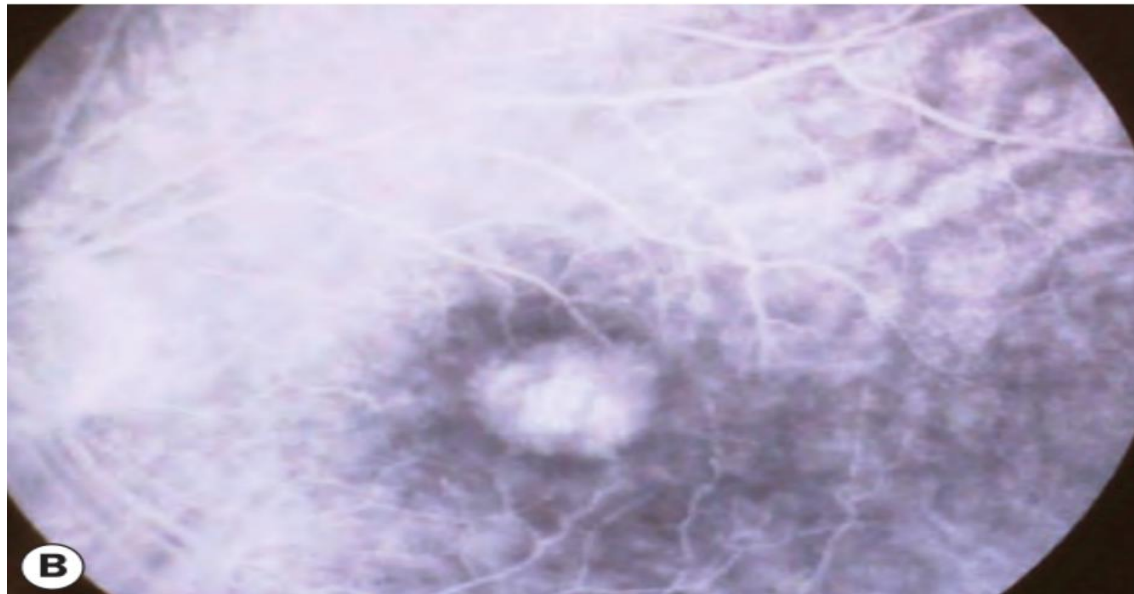


(A) Normal; (B) stage 1b - vitreomacular traction - shows attachment of the posterior hyaloid to the fovea, separation of a small portion of the sensory retina from the RPE in the foveolar region and intraretinal cystic changes; (C) eccentric stage 2 - small FTMH with vitreomacular traction (VMT) - shows attachment of the vitreous to the lid of the hole and cystic change; (D) stage 3 - medium or large FTMH with VMT - with intraretinal cystic spaces; (E) stage 4 - large FTMH with no VMT - shows a full-thickness macular hole with intraretinal cystic spaces and an overlying operculum (sometimes termed a pseudo-operculum);

## FA of stage 4 macular hole



(A) Early-phase window defect with early surrounding pooling



(B) surrounding cuff of subretinal fluid demonstrated in later shots



# Fundus Autofluorescence:

- Has been used as indication of the level of lipofuscin in the retinal pigment epithelium.
- Stage 2 shows punctate fluorescence
- Stage 3 and 4 shows hyperfluorescence foveolar spot
- Normally, autofluorescence of the retinal pigment epithelium underlying the fovea is decreased because the overlying macular yellow pigments absorb the exciting light.
- Without the overlying absorptive pigments, a macular hole creates a window defect so that autofluorescence is increased.
- Thus, this change in visible autofluorescence can be used as an indication of a full-thickness macular hole or of failure of surgical repair.



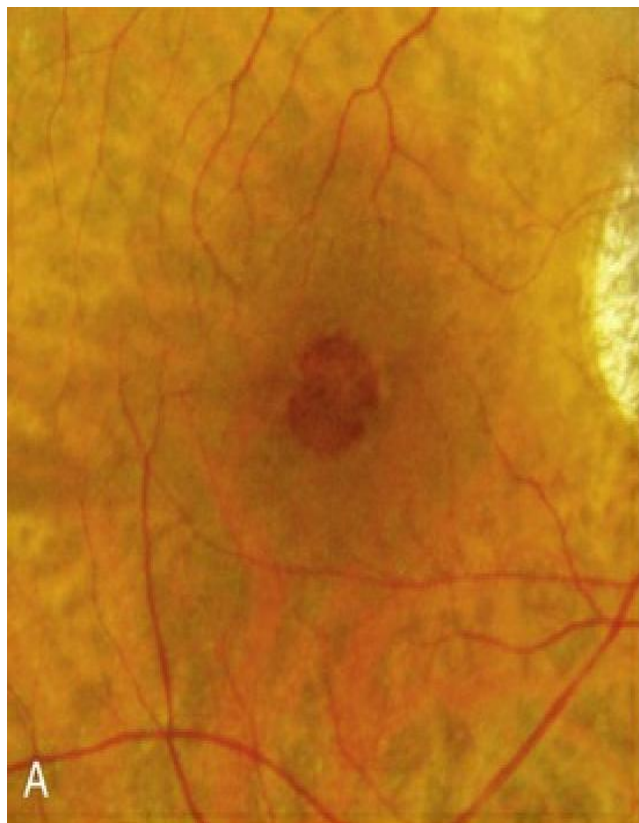
# Differential Diagnosis

- Epiretinal membrane and pseudohole
- Lamellar hole
- Cystic macular edema
- Central serous chorioretinopathy with central yellow spot
- Solar retinopathy
- Macular druse
- Pseudo-operculum

# Lamellar macular hole

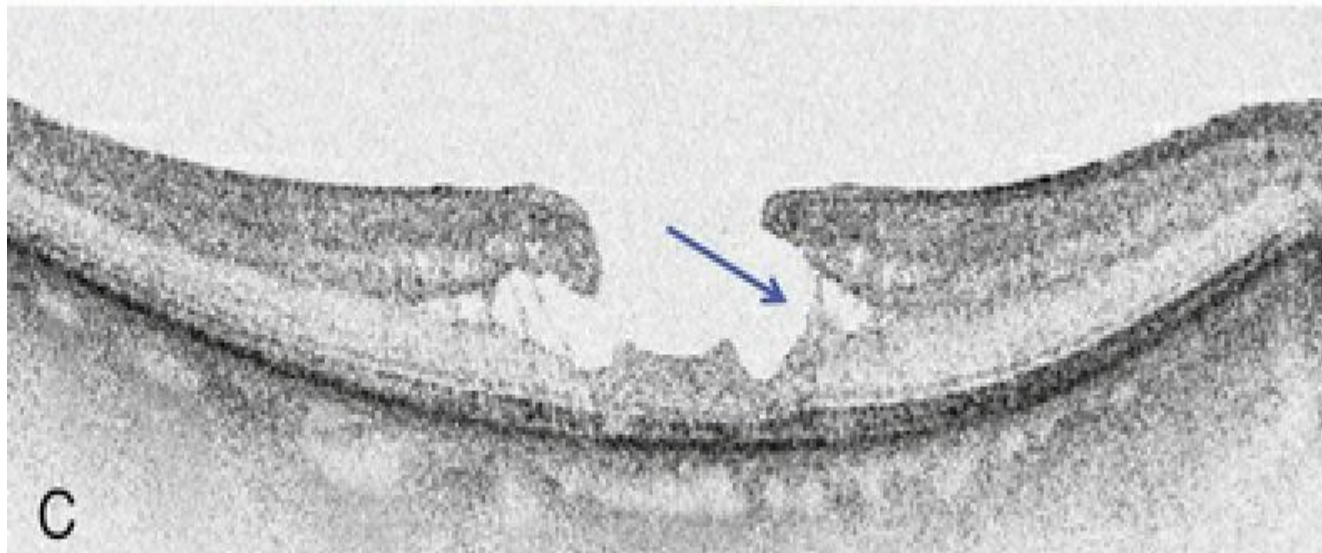
- Coined by **Gass in 1975**
- A partial thickness defect of the inner retina at the fovea but maintenance of the intact photoreceptor layer.
- The term lamellar hole was used to describe both the **endstage of a cystoid macular edema and the aborted process of formation of a macular hole**
- **OCT**-irregular thinning of the foveal floor, a cleavage between the inner and outer retina at the lamellar hole edge, and the absence of a contractile ERM.

FTMH	Lamellar MH
Round	Bi or trilobulated
Edge is thick and elevated	Edge is thinner



(A) Color photograph showing a roundish, lobulated, red central area corresponding to the thinning of the fovea after avulsion of the roof of a foveal cyst.

(B) En face optical coherence tomography (OCT) image of the same case. The dark area (white arrow) indicates the extent of the central defect and corresponds to the cleft between the inner and outer retina inside the edge of the lamellar hole.

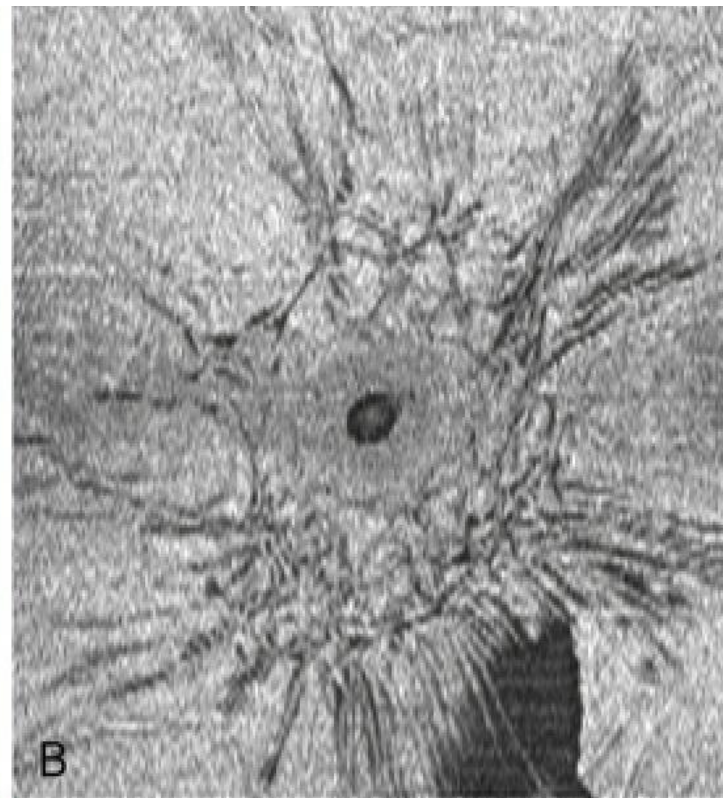


(C) OCT scan showing the irregular base of the lamellar hole and the cleft between the inner and outer retina inside the edge of the lamellar hole (blue arrow).

# Macular pseudoholes

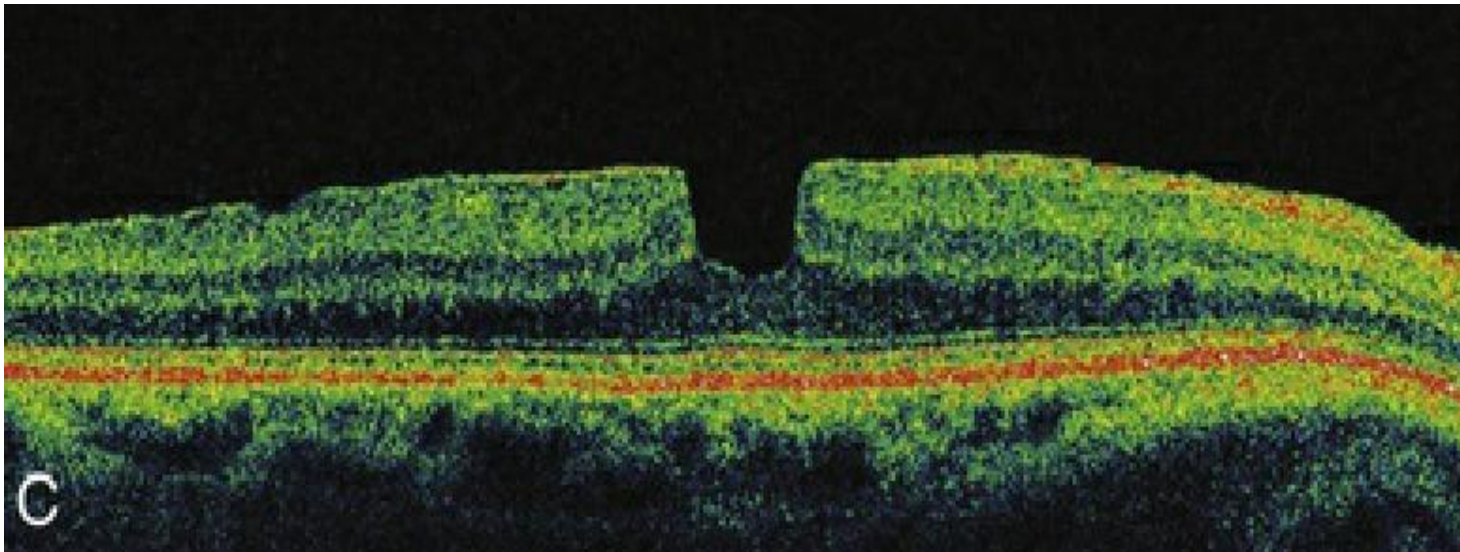
- The term of macular pseudohole was coined by **Allen and Gass in 1976** to designate a roundish centrofoveal image seen on biomicroscopy, which was due to the **centripetal contraction of an ERM**.
- This contraction induces the verticalization of the edge of the foveal pit.
- Vision may remain relatively good, and there are **no microscotomas**.
- **OCT**-thickening of the macula contracted by an ERM, and the U or V shape of the fovea. There is no loss of retinal tissue at the umbo of the fovea





(A) Color photograph showing a roundish, red, central area surrounded by the reflex of an epiretinal membrane (ERM).

(B) En face optical coherence tomography (OCT) image focused on the retinal surface showing the retinal folds induced by the ERM.



(C) A horizontal OCT scan shows the typical appearance of a U-shaped foveal pit with vertical edges, due to the contraction of the ERM.

Centripetal Traction

ERM

**Macular Pseudohole**

ERM Progression

**Normal retina**

Disruption of Foveal Cyst

Or,

AP Traction

Vitreomacular traction,  
via formation of inner retinal cysts

Or,

Tangential Traction

ERM/Partial PVD with eccentric  
vitreoretinal adherence

**Lamellar Macular Hole**

Dehiscence of  
Outer Retinal  
tissue

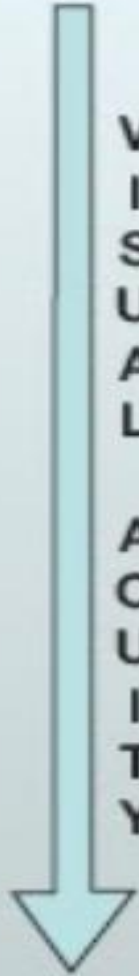
**Full Thickness Macular Hole**

AP Traction

Vitreofoveal  
traction with  
partial PVD

via formation  
of foveal  
detachment

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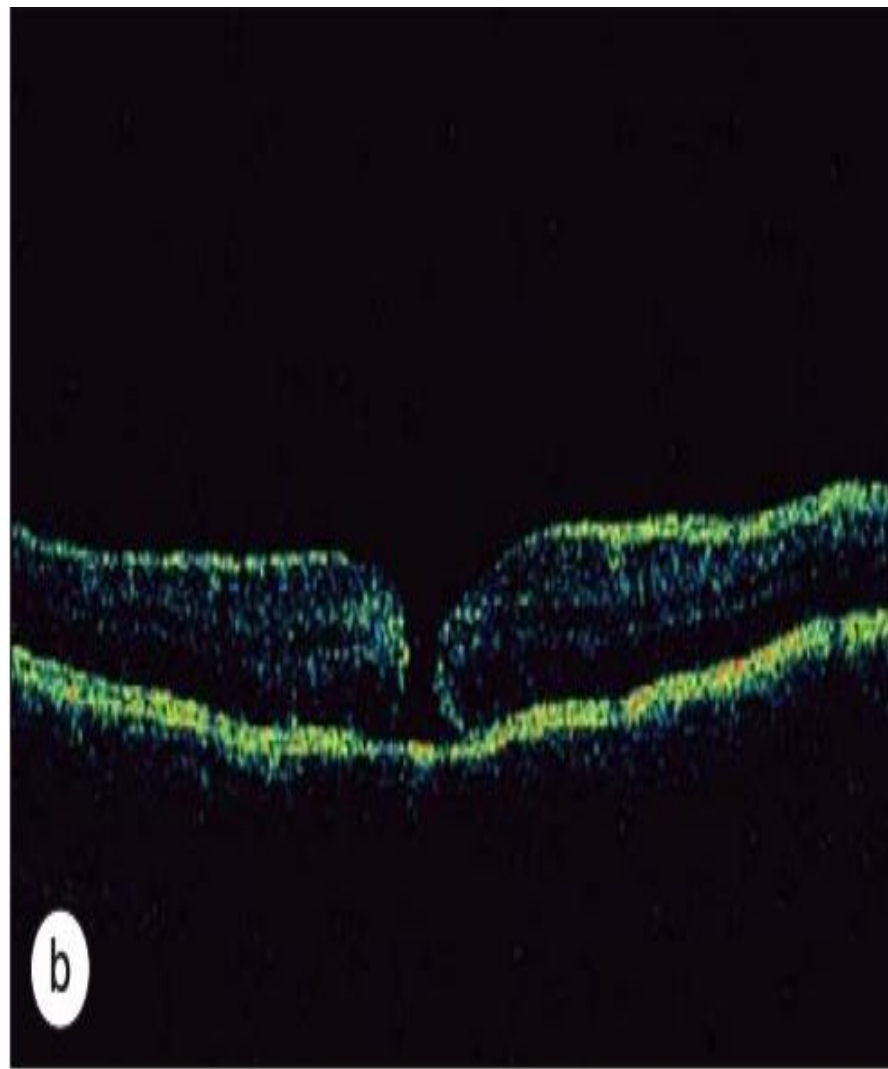


# Secondary macular hole

## Traumatic macular hole :

- Accounts for **less than 10% of macular hole** cases, occurs in children and young male adults as accidents at work in the home, or during ball games
- Traumatic macular holes typically ranges in size from **0.2 to 0.5 disc diameter**
- They are due to **sudden axial compression of the eye resulting in equatorial expansion and retinal rupture of the fovea.**
- The hole may be combined with other fundus lesions such as choroidal or Bruch's membrane disruption, commotio retinae, sclopetaria, or peripheral breaks.
- The visual prognosis depends not only on the closure of the hole but also on the topography of the other lesions.
- Vitrectomy with or without ILM peeling** can result in hole closure (90% of cases) and improvement in visual acuity
- Progression to retinal detachment is unlikely
- Spontaneous closure of traumatic macular holes has been reported, suggesting a role for initial observation





(a) Color fundus photograph and (b) ocular coherence tomography revealing small full-thickness macular hole less than 500  $\mu$ m in diameter caused by contusion injury to the eye. (c) Anterior segment photograph shows zonular fiber dialysis.



# High myopia

- Macular holes are one of the **complications of high myopia** together with posterior staphyloma and choroidal atrophy.
- Their pathogenesis may be different from that of nonmyopic eyes because the vitreous cortex often remains adherent to the retinal surface.
- Some of these Macular hole occur after a progressive decrease in vision due to the worsening of foveoschisis

# Other causes of secondary macular hole

- Alport syndrome
- Behçet disease
- Best macular dystrophy
- Cat scratch disease
- Central retinal artery occlusion
- Drusen
- Electrical shock injury
- Fungal endophthalmitis
- Idiopathic parafoveal telangiectasia
- Laser pointer
- Nd:YAG posterior capsulotomy
- Retinal arterial macroaneurysm
- Retinitis pigmentosa
- Stargardt disease
- Syphilis
- Toxoplasmic choroiditis
- Valsalva retinopathy
- Vitrectomy
- Vogt-Koyanagi-Harada disease
- X-linked juvenile retinoschisis

# Management

- **Observation** : Surgery not recommended in **stage 1**. About 50% of stage 1 holes resolve following spontaneous vitreofoveolar separation, so these are managed conservatively
- **Pharmacological vitreolysis**
  - ☆ Pharmacological vitreolysis with **ocriplasmin** is a newer treatment that may be suitable for small earlier-stage holes
  - ☆ Intravitreal injection of Ocriplasmin (**0.125 mg/0.1ml**) , a recombinant form of human serine protease plasmin that retains its proteolytic enzymatic properties.
  - ☆ It primarily targets fibronectin, laminin and collagen fibres that adheres vitreous to the retina.
  - ☆ The presence of **ERM is a poorer prognostic indicator** for ocriplasmin treatment

# Contraindications for surgery

- Choroidal rupture
- Traumatic RPE damage in macular area
- Chronic CME
- Optic nerve disorders

# Surgical procedure

## Pars plana vitrectomy

- Standard three port pars plana vitrectomy is performed.
- The **posterior vitreous is detached and the vitreous base is debulked**
- Induction of posterior vitreous detachment can be done by suction of the cutter, silicone tipped suction cannula or even a forceps that is used to grasp the vitreous fibers close to the disk. The best place to initiate the separation is **near the disk**.
- Direct aspiration of the vitreous fibers attached to the Weiss ring appears to be the most effective way of lifting the vitreous cortex en bloc and gradually extending its detachment to the equator in all the quadrants of the fundus.
- Although one can use **intravitreal triamcinolone acetonide or fluorescein** to visualize the undetached clear vitreous

# ILM peeling

- ILM denuded area of the retina becomes slightly whitish and hence may aid the identification of area with intact ILM but staining the ILM definitely makes the removal easier by improving the visualization.
- Agents used for this purpose include **triamcinolone acetonide, indocyanin green (ICG), trypan blue, brilliant blue and infracyanine green.**
- Use of **VINCE (vitreoretinal internal limiting membrane color enhancer)** enables selective painting of ILM that needs to be removed without unnecessarily staining the perifoveal retina and the peripheral retina.
- **Indocyanine green is associated with toxicity to the RPE .**
- Usually the membrane is removed as a circular disk around the macular hole for at least two disk diameters in size and called as **maculorrhexis**

# Internal tamponade

- A nonexpansile mixture of **C3F8 and air of 12 to 14 percent** is usually used and the patients are encouraged to lie **prone** for at least **14 to 16 hours a day** for the first ten days.
- **Silicone oil tamponade** has been used instead gas in cases where the patient is unable to assume the prone position for prolonged periods. Obviously this procedure would need removal of oil as a second procedure.

# Complications of surgery

- **Cataract**
- Iatrogenic retinal tears
- Rhegmatogenous Retinal detachment
- Visual-field defect
- Ocular hypertension
- Endophthalmitis
- Angle-closure glaucoma
- Transient rise in intraocular pressure



# Adjunctive Agents

## Thrombin:

- A known **stimulator of RPE & glial cells.**
- Failed to show marked increase in the successrate of macular hole closure.
- **Increased post-op inflammation.**

## Gelatin plugs:

- Preliminary results of vitrectomy followed by placement of an absorbable cross-linked gelatin plug in stages 3 & 4 have been encouraging.
- Recommended **use in high myopes with posterior staphyloma or recurrent macular hole.**

## TGF $\beta$ -2:

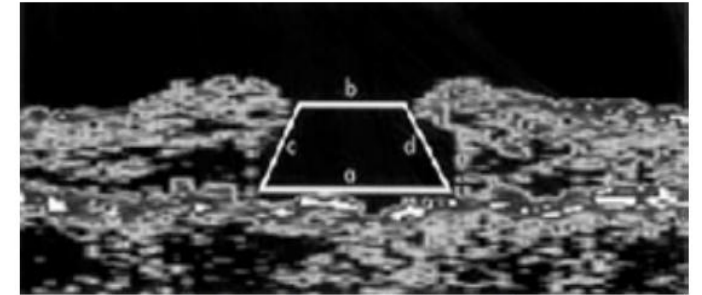
- It appears to promote **wound healing by stimulating collagen fibril growth, glycoproteins & proteoglycans.**
- Studies have suggested a potential benefit using human recombinant TGF  $\beta$ -2 for persistent FTMHs following failed primary surgery & for traumatic holes.

## Autologous platelet concentrate:

- It induces a **localized fibrocellular response that seals the retinectomy edge,** involving a mixed population of glial, RPE & fibroblastic cells.
- It has a beneficial effect in primary FTMH surgery & also increased the surgical success rate in reoperations combined with rigorous epiretinal membranectomy.

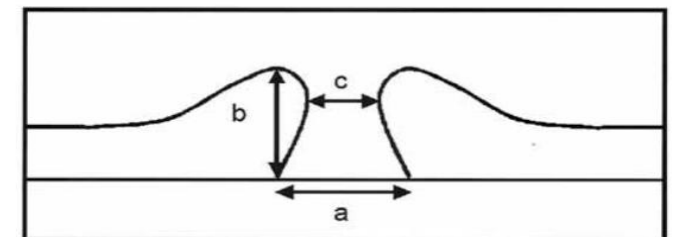
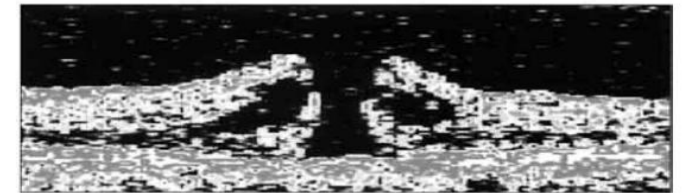
# Prognostic factors

**Hole Form Factor (HFF)** - indirect indicator of the size of hole. The higher the value of HFF ( $>0.9$ ) the better are the results.



Hole form factor (HFF) =  $c + d/a$   
 $a$  = base diameter  
 $b$  = minimum diameter  
 $c$  = left arm length  
 $d$  = right arm length

**Macular hole Index (MHI)** - is basically the ratio of hole height with base diameter. Higher MHI ( $\geq 0.5$ ) correlates with better visual outcomes.



Macular Hole Index (MHI) =  $b/a$

**Thank  
You**

