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Age-related macular degeneration

Drusen

1. Histopathology (Fig. 17.1)
   - Discrete deposits of the abnormal material located between the basal lamina of the RPE and the inner collagenous layer of Bruch membrane.
   - Thickening of Bruch membrane compounded by excessive production of basement membrane deposit by the RPE.

   ![Fig. 17.1 Histopathology of drusen](image)

2. Signs – yellow sub-RPE excrescences distributed symmetrically at both posterior poles.
   a. Small hard drusen – less than half a vein width in diameter with discrete margins (Fig. 17.2).
   b. Large soft drusen – vein width or more in diameter with indistinct margins (Fig. 17.3a).
   c. Calcified drusen – may be hard or soft (Fig. 17.4).

   ![Fig. 17.2 Hard drusen](image)

   ![Fig. 17.3 (a) Soft drusen; (b) FA](image)
3. FA
   a. Hyperfluorescence – window defect due to atrophy of the overlying RPE and late staining (see Fig. 17.3b).
   b. Hypofluorescence – of hydrophobic (high lipid content) drusen.

4. Drusen and AMD – features associated with an increased risk of subsequent visual loss include large soft and/or confluent drusen, and focal RPE hyperpigmentation (Fig. 17.5), particularly if the other eye has AMD.

5. Prophylactic treatment – high-dose multivitamins and antioxidants can decrease the risk of progression of AMD in eyes with the following high risk characteristics: visual loss in the contralateral eye from pre-existing dry or wet AMD, and confluent soft drusen even in the absence of visual loss.

Atrophic (dry) age-related macular degeneration

1. Pathogenesis – slowly progressive atrophy of the photoreceptors, RPE, and choriocapillaris.
2. Presentation – gradual impairment of vision over months or years.
Fig. 17.6 Dry AMD

Fig. 17.7 Geographic atrophy

Fig. 17.8 Window defect in dry AMD

Fig. 17.9 (a) PED; (b) ICG
Retinal pigment epithelial detachment

1. Pathogenesis – reduction of hydraulic conductivity of the thickened Bruch membrane impeding movement of fluid from the RPE towards the choroid.
2. Presentation – metamorphopsia and impairment of central vision.
3. Signs – circumscribed, dome-shaped elevation at the posterior pole (Fig. 17.9a).
4. ICG – oval hypofluorescence with a faint ring of surrounding hyperfluorescence (Fig. 17.9b).
5. OCT – separation of the RPE from Bruch membrane (Fig. 17.10).

6. FA – well demarcated oval area of hyperfluorescence which increases in density but not in size (Fig. 17.11).
7. Course – may follow one of the following patterns:
   a. Spontaneous resolution – without residua.
   b. Geographic atrophy – following spontaneous resolution.
   c. Detachment of the sensory retina.
   d. RPE tear formation.

Retinal pigment epithelial tear

1. Pathogenesis – tearing of the RPE at the junction of attached and detached RPE due to tangential stress – may be spontaneous, or more commonly, follows laser photocoagulation, PDT or anti-VEGF therapy of CNV.
2. Presentation – sudden worsening of central vision.
3. Signs – crescent shaped RPE dehiscence with a retracted and folded flap (Fig. 17.12a).
4. FA – relative hypofluorescence over the flap with adjacent hyperfluorescence due to the exposed choriocapillaris (Fig. 17.12b–d).
5. **OCT** – loss of the normal dome-shaped profile of the RPE in the PED, with hyper-reflectivity adjacent to the folded RPE (Fig. 17.12e–h).

6. **Prognosis** – poor for subfoveal tears.

### Neovascular (wet) age-related macular degeneration

**Pathogenesis**
- CNV originating from the choriocapillaris grows through defects in Bruch membrane (Fig. 17.13).
- Initial visual loss is caused by leakage from CNV under the sensory retina and under the RPE.
- This is followed by bleeding from CNV.
- Permanent visual loss is caused by subretinal (disciform) scarring.

**Clinical features**

1. **Presentation** – metamorphopsia, positive scotoma, and blurring of central vision.
2. **Signs** – serous retinal elevation, foveal thickening, CMO, subretinal haemorrhage, and hard exudate formation (Fig. 17.14a).

**Fluorescein angiography**

1. **Classic CNV**
   - Well-defined membrane which fills with dye in a ‘lacy’ pattern during the very early phase of dye transit (Fig. 17.14b), fluoresces brightly during peak dye transit (Fig. 17.14c), and then leaks into the subretinal space and around the CNV within 1–2 min.
   - Late staining of fibrous tissue within the CNV (Fig. 17.14d).
   - Classic CNV is classified according to its relation to the centre of FAZ as extrafoveal, subfoveal, and juxtafoveal.
CNV can be further subdivided into wholly classic and predominantly classic in which 50% or less of the lesion has a classic component.

2. **Occult CNV** – poorly defined with less precise features on the early frames (Fig. 17.15b) and gives rise to late, diffuse or multifocal leakage (Fig. 17.15c,d).

3. **Fibrovascular PED** – combination of CNV and PED in which the CNV fluoresces brighter (hot spot) than the detachment; in other cases, the CNV may be obscured by blood or turbid fluid.

**Course**

The course of untreated CNV is often relentless and the prognosis very poor due to the following complications.

1. **Haemorrhagic PED**
   - Initially, the blood is confined to the sub-RPE space and appears as a dark elevated mound (Fig. 17.16).
   - The blood may then break into the subretinal space and assumes a more diffuse outline and a lighter red colour (Fig. 17.17).

2. **Vitreous haemorrhage** – when subretinal blood breaks through into the vitreous cavity; rare.

3. **Subretinal (disciform) scarring** – causes permanent loss of central vision (Fig. 17.18).
Fig. 17.15 FA of occult CNV
Fig. 17.16 Sub-RPE haemorrhage
Fig. 17.17 Subretinal haemorrhage
4. Massive subretinal exudation – due to chronic leakage from CNV (Fig. 17.19).

**Retinal angiomatous proliferation**

1. **Definition** – uncommon type of wet AMD in which neovascularization originates from the retinal vasculature and not the choriocapillaris.

2. **Signs**
   - Intraretinal and subretinal neovascularization often accompanied by haemorrhage and oedema.
   - CNV associated with fibrovascular PED and retinochoroidal anastomoses.

3. **FA** – similar to purely occult or minimally classic CNV.

4. **ICG** – hot spot in mid or late frames.

5. **Treatment** – PDT with adjunctive intravitreal triamcinolone.

**Polypoidal choroidal vasculopathy**

1. **Definition** – bilateral, choroidal vascular disease in which the inner choroidal vessels consist of a dilated network with multiple terminal aneurysmal protuberances that have a polypoidal configuration.

2. **Signs**
   - a. *Exudative* – multiple PED, serous RD, and lipid deposits (Fig. 17.20a).
   - b. *Haemorrhagic* – haemorrhagic PED and subretinal haemorrhage (Fig. 17.21a).

3. **ICG** – polypoidal dilatations beneath the RPE that fill slowly and then leak intensely (Figs 17.20b & 17.21b).

4. **Course**
   - Spontaneous resolution in 50%.
   - In the remainder occasional repeated bleeding and leakage, resulting in macular damage and visual loss.

5. **Treatment** – PDT.
Age-related macular hole

1. Pathogenesis – abnormal vitre-foveolar attachment, with resultant antero-posterior and tangential traction.

2. Presentation – in old age.

3. Staging (Fig. 17.22)
   - Stage 1a (impending) – flat umbo, yellow foveolar spot 100–200 µm in diameter with loss of the foveolar reflex.
   - Stage 1b (occult) – yellow ring with a bridging interface of vitreous cortex.
   - Stage 2 (early hole) – full-thickness defect, less than 300 µm in diameter with or without an overlying pseudo-operculum.
   - Stage 3 (established hole) – full-thickness defect more than 400 µm in diameter with an attached posterior vitreous face with or without an overlying pseudo-operculum.
   - Stage 4 – round defect more than 400 µm in diameter surrounded by a cuff of subretinal fluid and tiny yellowish deposits within its crater, and completely detached vitreous cortex (Fig. 17.23).
Fig. 17.22 Staging of macular hole

1. Normal fovea
2. Stage 1a. Impending hole
3. Stage 1b. Occult hole
4. Stage 2. Hole
5. Stage 3. Hole
6. Stage 4. Hole

4. OCT – useful in the diagnosis and staging of macular holes (Fig. 17.24).

5. FA – corresponding area of hyperfluorescence.

6. Surgery (Fig. 17.25)
   - Indicated for stage 2 and above, associated with a visual acuity worse than 6/9.
   - Following successful surgery, visual improvement is achieved in 80–90% of eyes, with a final visual acuity of 6/12 or better in up to 65%.
Acquired Macular Disorders

Macular microhole

1. **Presentation** – central scotoma or reduced reading vision.

2. **Signs** – very small, red, well demarcated intraretinal foveal or juxtafoveal defect that remains stationary with long-term follow-up.

3. **OCT** – well localized subtle defect that probably indicates the presence of a gap in the photoreceptors, and/or the RPE.

Central serous retinopathy

1. **Pathogenesis** – localized detachment of the sensory retina at the macula secondary to focal RPE defects.

2. **Presentation** – unilateral relative positive scotoma, micropsia, metamorphopsia, and occasionally macropsia.

3. **Signs** – round detachment of the sensory retina at the macula that may be associated with small precipitates on its posterior surface (Fig. 17.26)

4. **OCT** – elevation of the sensory retinal layer from the highly reflective RPE layer by an optically empty zone (Fig. 17.27)

5. **FA**
   - Small hyperfluorescent spot that enlarges and ascends vertically and laterally until the entire area

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Fig. 17.25 Unsuccessful surgery for macular hole. Pre-operative appearance (above); postoperative appearance (below)

Fig. 17.26 CSR

Fig. 17.27 OCT of CSR
is filled with dye (smoke stack appearance – Fig. 17.28)
• Less frequently the hyperfluorescent spot enlarges centrifugally (ink-spot appearance).

6. ICG – early phase shows dilated choroidal vessels, mid phases show multiple areas of hyperfluorescence due to choroidal hyperpermeability.

7. Course
• Spontaneous resolution within 3–12 months is the rule.
• Occasionally the course is protracted and results in progressive widespread RPE changes (chronic retinal pigment epitheliopathy).

8. Treatment
b. Argon laser photocoagulation – in eyes with extrafoveal leaks achieves speedier resolution and lowers the recurrence rate but does not influence the final visual outcome.
c. PDT – in acute CSR with subfoveal leaks and in chronic disease.

Cystoid macular oedema

1. Definition
• Accumulation of fluid in the outer plexiform and inner nuclear layers with the formation of fluid-filled cyst-like changes (Fig. 17.29a).
• Lamellar hole formation in longstanding cases (Fig. 17.29b).

2. Signs – loss of the foveal depression, retinal thickening, and multiple cystoid areas in the sensory retina (Fig. 17.30).
3. **OCT** – collection of hyporeflective spaces within the retina, with overall macular thickening and loss of the foveal depression (Fig. 17.31).

4. **FA** – late phase shows a ‘flower-petal’ pattern of hyperfluorescence (Fig. 17.32).

5. **Vascular causes**
   - Diabetic retinopathy, retinal vein occlusion, hypertensive retinopathy, idiopathic retinal telangiectasis, retinal artery macroaneurysm, and radiation retinopathy.
   - Treatment – laser photocoagulation in selected cases.

6. **Inflammatory causes**
   - Chronic anterior uveitis, intermediate uveitis, and certain forms of posterior uveitis.
   - Treatment – control of uveitis with anti-inflammatory agents; systemic carbonic anhydrase inhibitors may be beneficial in CMO associated with intermediate uveitis.

7. **Following cataract surgery**
   - Risk factors – posterior capsular rupture, vitreous loss and incarceration into the incision site, anterior chamber and secondary IOL implantation, diabetes, and CMO in the other eye.
   - Treatment – correction of the underlying cause, if possible; persistent cases may require systemic carbonic anhydrase inhibitors, topical and periocular steroids, topical NSAIDs, intravitreal triamcinolone, and pars plana vitrectomy.
8. **Drug-induced** – topical adrenaline 2% (especially in the aphakic eye), topical latanoprost, and systemic nicotinic acid.

9. **Retinal dystrophies**
   - RP, gyrate atrophy, and dominantly inherited CMO.
   - Treatment – systemic carbonic anhydrase inhibitors in RP.

10. **Vitreomacular traction syndrome** (see below).

11. **Macular epiretinal membranes** (see below).

12. **CNV** – CMO is an adverse prognostic factor.

13. **Tumours** – retinal haemangioblastoma and choroidal haemangioma.

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### Macular epiretinal membrane

**Pathogenesis**

- Proliferation of retinal glial cells at the vitreoretinal interface that have gained access to the retinal surface through breaks in the internal limiting membrane.
- May be idiopathic or secondary to RD surgery and cryotherapy, retinal vascular disease, intraocular inflammation, and trauma.

### Cellophane maculopathy

1. **Presentation** – mild metamorphopsia although frequently the condition is asymptomatic and is discovered by chance.

2. **Signs**
   - Irregular light reflex or sheen at the macula.

- The membrane is translucent and best detected with ‘red-free’ light (Fig. 17.33).

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Fig. 17.33 Cellophane maculopathy

### Macular pucker

1. **Presentation** – metamorphopsia and blurring of central vision.

2. **VA** – 6/12 or worse.

3. **Signs**
   - Severe vascular distortion, retinal wrinkling and white striae (Fig. 17.34).
   - Macular pseudo-hole (Fig. 17.35) and occasionally CMO.
4. **OCT** – highly reflective (red) layer on the retinal surface associated with thickening (Fig. 17.36).

5. **FA** – highlights the vascular tortuosity and may show hyperfluorescence if leakage is present (Fig. 17.37).

6. **Treatment** – removal of the membrane improves or eliminates distortion, and improves visual acuity in about 50% of cases.

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**Degenerative myopia**

1. **Definition**
   - Refractive error > −6 D and axial length >26 mm.
   - Affects approximately 0.5% of the general population and 30% of myopic eyes.

2. **Signs**
   - Pale tessellate (tigroid) fundus with visibility of large choroidal vessels (Fig. 17.38).
• ‘Lacquer cracks’ consist of ruptures in the RPE–Bruch membrane–choriocapillaris complex (Fig. 17.39).
• Focal chorioretinal atrophy with visibility of the sclera (Fig. 17.40).
• Staphylomas due to expansion of the globe and scleral thinning.

3. Maculopathy
• CNV associated with ‘lacquer cracks’.
• Subretinal ‘coin’ haemorrhages from lacquer cracks without CNV (Fig. 17.41).
• Fuchs spot – pigmented lesion after absorption of a macular haemorrhage (Fig. 17.42).

4. FA – filling of large choroidal vessels but not of the choriocapillaris (Fig. 17.43).
5. Ocular complications
- Rhegmatogenous RD.
- Cataract, which may be posterior subcapsular or early-onset nuclear sclerosis.
- Increased prevalence of primary open-angle glaucoma, pigmentary glaucoma, and steroid responsiveness.

6. Systemic associations
- Stickler syndrome.
- Marfan syndrome.
- Ehlers–Danlos syndrome.
- Pierre–Robin syndrome.

Angioid streaks

1. Definition – crack-like dehiscences in thickened, calcified and abnormally brittle collagenous and elastic portions of Bruch membrane.

2. Signs
- Mottled pigmentation (‘peau d’orange’).
- Grey or dark-red linear lesions with irregular serrated edges intercommunicate around the optic disc and then radiate outwards (Fig. 17.44).
- Associated RPE hyperplasia in longstanding cases (Fig. 17.45).

3. FA – hyperfluorescence over the streaks associated with variable hypofluorescence corresponding to RPE hyperplasia.

4. Prognosis – visual loss in 70% of cases due to CNV (Fig. 17.46), traumatic choroidal rupture, or foveal involvement by a streak.

5. Systemic associations – in 50% of patients:
Hypotony maculopathy

1. **Causes** – very low IOP (usually <6 mmHg) following filtration surgery, particularly when adjunctive antimetabolites are used, trauma and chronic anterior uveitis.

2. **Signs** – irregular chorioretinal folds (Fig. 17.48).

3. **Treatment** – depends on the cause.

![Fig. 17.48 Hypotony maculopathy](image-url)
Vitreomacular traction syndrome

1. Pathogenesis
   - Vitreous cortex is attached to the fovea and the optic disc but detached temporal to the fovea and the area of the papillomacular bundle.
   - This incomplete posterior vitreous detachment exerts persistent anterior traction on the fovea, which leads to macular changes, notably CMO.

2. Signs
   - Partial posterior vitreous detachment with persistent attachment of vitreous to the macula.
   - The macula may show retinal surface wrinkling, distortion, an epiretinal membrane or CMO.

3. OCT
   - is used to confirm the diagnosis (Fig. 17.49).

4. Treatment
   - pars plana vitrectomy.

Solar maculopathy

1. Pathogenesis
   - retinal injury caused by photochemical effects of solar radiation is caused by directly or indirectly viewing the sun (eclipse retinopathy).

2. Presentation
   - within 1–4 hours of solar exposure with unilateral or bilateral impairment of central vision and a small central scotoma.

3. Fundus
   - Small yellow or red foveolar spot which fades within a few weeks.
   - The spot is replaced by a sharply defined foveolar defect with irregular borders or a lamellar hole (Fig. 17.50).

4. Treatment
   - not possible.

5. Prognosis
   - good with improvement of vision within 6 months.

Idiopathic choroidal neovascularization

Idiopathic CNV is an uncommon condition which affects patients under the age of 50 years. It carries a better visual prognosis than that associated with AMD and in some cases spontaneous revolution may occur. The CNV is of type 2 and lies predominantly above the RPE.