Posterior Segment

10

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10.1 Posterior Segment Examination Sequence

10.1.1 Indirect Slit-Lamp Biomicroscopy

- Introduce yourself to the patient
- Set patient up on the slit-lamp
- Focus and center a slit beam (use the brightest light intensity that the patient can easily tolerate) in the center of the corneal surface and hold a condensing lens (e.g. +90 D) stationary between your thumb and index finger approximately 5–10 mm from the patient's cornea
- Keeping the slit beam centered on both the patient's cornea and the condensing lens, pull the slit-lamp joystick with your freehand away from the patient until the patient's red reflex becomes a focused fundus image
- Ask the patient to look at your ear or shoulder to begin examining the optic disc. Proceed temporally circumferentially around the posterior pole and ending at the fovea. Examine different aspects of the peripheral retina by asking the patient to look in different directions of gaze

10.1.2 Indirect Ophthalmoscopy

- For the exam the patients will likely be lying flat on a bed with the pupils dilated
- Introduce yourself to the patient
- Ask for room lights to be dimmed (if not already dimmed)
- Begin by asking the patient to stare at a distance target on the ceiling just above and beyond your shoulder (your right shoulder when examining the patient's right eye and your left shoulder when examining the patient's left eye)
- Examine each eye in turn. Direct the indirect ophthalmoscopes light source into the center of the patient's pupil. Hold the condensing lens, and position it just in front of the patient's eye and center the pupil in it. Pull the lens slowly away from the patient's eye by flexing your wrists and by bending your fingers holding the lens until you see a focused image of the fundus
- Shift the field of view to different areas of the eye by walking around the patient and by asking the patient to look in different directions of gaze

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- Remember that the image of the patient's fundus is reversed and inverted

10.2 Stargardt Disease (Fig. 10.1 and Table 10.1)

10.2.1 Examination

- Bilateral symmetrical changes often confined to the posterior pole
- Yellowish white flecks (differ from drusen in that they are more elongated than round and they often contact each other at angles that create a branching or net-like appearance) at the level of the RPE — pisciform flecks (two adjacent flecks form an obtuse angle)
- Relative sparing of the peripapillary retina and RPE
- Circular area of RPE atrophy centered at the fovea with a metallic sheen (crystalline deposits overlying this atrophy) ± clumps of dark pigment within the atrophic lesion
- Vermillion or light-brown colour to the fundus with complete obscuration of the underlying choroidal vessels
- Bull's eye maculopathy (see Fig. 10.2 and Table 10.2): ring of atrophic RPE surrounding the fovea

10.2.2 Investigations

- FAF: loss of autofluorescence in areas of atrophy (including the atrophic fovea and bull's eye maculopathy)
- FFA: dark choroid (see Fig. 10.3) — masking of the choroidal circulation with dye filled retinal vessels lying upon a completely hypofluorescent background ± hyperfluorescence of flecks and areas of atrophy
- OCT: RPE atrophy and outer retinal loss (loss of ellipsoid zone)

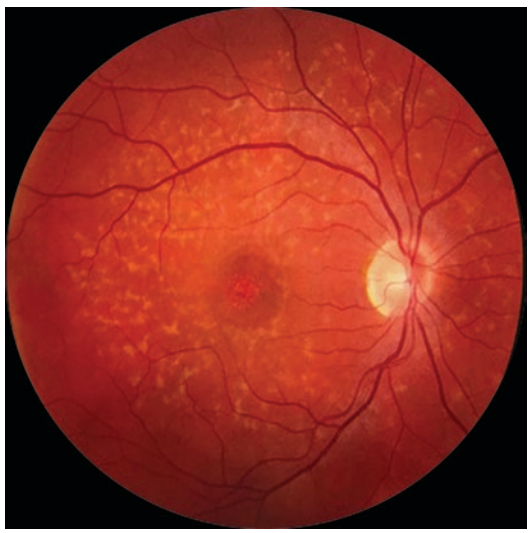

Fig. 10.1 Colour fundus image of a patient with Stargardt disease showing the typical yellowish-white flecks in the posterior pole

Table 10.1 Key facts about Stargardt disease

• AR macular dystrophy (onset in childhood)
• Mildest of the ABCA4 phenotypes

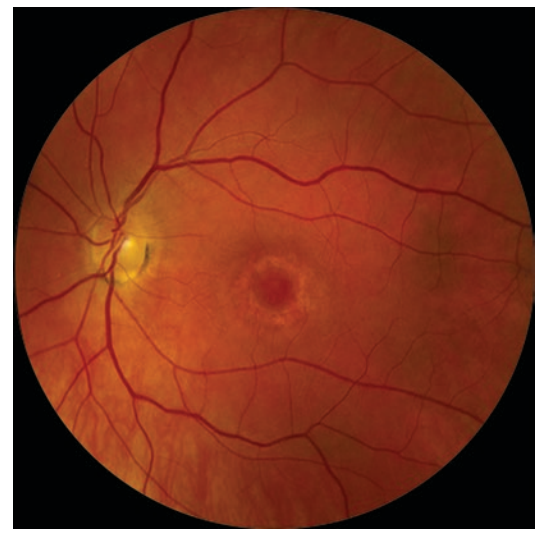

Fig. 10.2 Colour fundus image of a patient with Stargardt disease with a typical bull's eye maculopathy

Table 10.2 Causes of Bulls eye maculopathy

• Stargardt disease
• Hydroxychloroquine toxicity
• Cone dystrophy/Cone-rod dystrophy
• Batten disease (neuronal ceroid lipofuscinosis — ERG severely reduced or extinguished before age of 10 years)

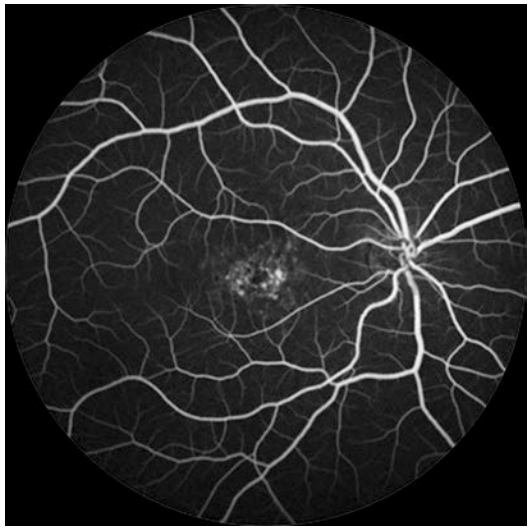

Fig. 10.3 Fundus fluorescein angiogram of a patient with Stargardt disease showing a typical dark choroid with hyperfluorescence of flecks

- Molecular testing: ABCA4 gene
- EDTs: full field ERG - typically normal in Stargardt disease, pattern ERG (PERG) — group 1 (severe PERG abnormality with normal full field ERG), group 2 (additional loss of photopic function), group 3 (additional loss of both photopic and scotopic function)

10.2.3 Treatment

- There is currently no proven treatment for ABCA4 disease

10.2.4 Other Diagnosis to Consider

- Fundus flavimaculatus: AR, adult onset, pisciform flecks, peripapillary sparing, relative preservation of vision (no macular atrophy)
- Stargardt like autosomal dominant macular dystrophy (SLDMD): positive FHx, pisciform flecks, peripapillary sparing, macular atrophy, normal ERG
- Pattern dystrophy (group of inherited conditions characterised by changes at the level of the RPE) — reticular pattern: AD (positive FHx), gene testing for PRPH2 mutations, low risk of CVN development
- Cone dystrophy/Cone-rod dystrophy
- Central areolar choroidal dystrophy: AD (positive FHx), earliest change is a fine mottled depigmentation in the macula of both eyes and gradually evolves into symmetric, sharply outlined oval or round areas of GA of the RPE, normal full field ERG

10.3 Best's Macular Dystrophy (BMD) (Fig. 10.4 and Table 10.3)

10.3.1 History

- Positive FHx (autosomal dominant)

10.3.2 Examination

- Solitary round oval yellow slightly elevated lesion centered on the fovea (egg-yolk like

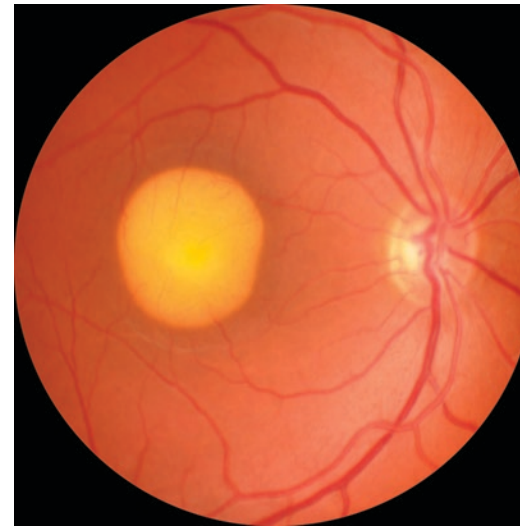

Fig. 10.4 Colour fundus image of a patient with BMD showing the typical egg-yolk like lesion

Table 10.3 Key facts about BMD

• AD macular dystrophy caused by mutations in BEST1 gene • Onset usually in childhood • BMD refers to the "classic" form of a single symmetric egg-yolk like lesion centered on the fovea of each eye

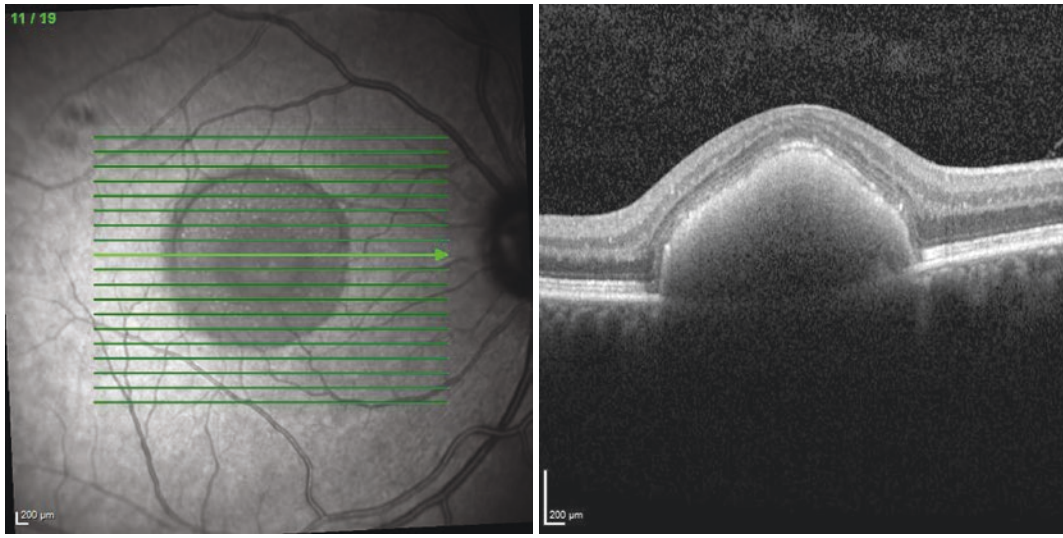

Fig. 10.5 OCT image of a patient with BMD showing the typical sub-RPE hyper-reflective lesion

lesion) ± pseudohypopyon (yellow material gravitates inferiorly in the subretinal space) ± subretinal fibrosis ± RPE atrophy ± RPE hyperpigmentation
- Subretinal haemorrhage in the macula — suggestive of a CNV membrane (20%)

10.3.3 Investigations

- OCT (see Fig. 10.5): sub-RPE hyper-reflective lesion
- FFA: look for leakage suggestive of a CNV membrane
- EDT: EOG — reduced Arden ratio (less than 1.5), ERG — normal full field ERG

10.3.4 Treatment

- Treatment of CNV with anti-VEGF therapy

10.3.5 Staging of BMD

- Stage 1: Pre-vitelliform — EOG findings only
- Stage 2: Vitelliform — yolk-like macular lesion
- Stage 3: Pseudohypopyon — partial absorption leaving level
- Stage 4: Vitellidisruptive — scrambled egg appearance with RPE atrophy + hyperpigmentation
- Stage 5: End-stage — geographic atrophy of the RPE

10.3.6 Other Diagnoses to Consider

- Adult-onset foveomacular vitelliform pattern dystrophy — smaller (less than 1/3 DD) vitelliform lesions than BMD, normal EOG

10.4 Sorsby Macular Dystrophy (Fig. 10.6 and Table 10.4)

10.4.1 History

- Positive FHx

10.4.2 Examination

- In the early stages, yellow-to-grey material (similar in appearance to drusen) is present at the level of Bruch's membrane

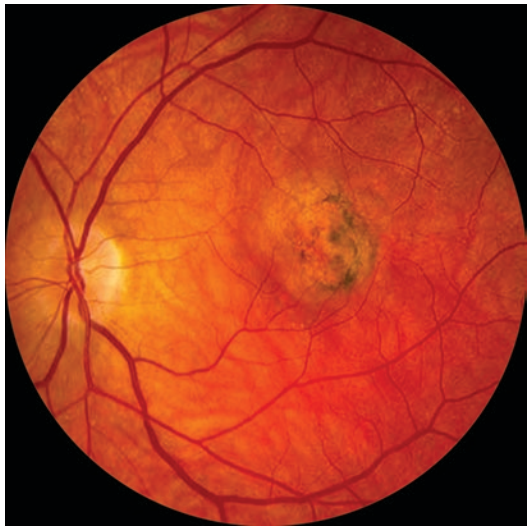

Fig. 10.6 Colour fundus image of a patient with Sorsby macular dystrophy showing subfoveal disciform scarring

Table 10.4 Key facts about Sorsby macular dystrophy

- AD condition caused by a mutation in the TIMP3 gene
- Progressive atrophy of the peripheral choroid and RPE is common
- Most commonly presents in the fourth or fifth decade of life

- Bilateral subfoveal CNV membranes with extensive disciform scarring, which can extend peripherally (unlike AMD which rarely extends beyond the temporal vascular arcades) in the late stages

10.4.3 Investigations

- Mutation in TIMP3 gene

10.4.4 Treatment

- CNV: anti-VEGF therapy

10.4.5 Other Diagnoses to Consider

- AMD

10.5 Retinitis Pigmentosa (RP) (Fig. 10.7 and Table 10.5)

10.5.1 Classification

- Based on inheritance (50% have no FHx): AD (later onset and less severe), AR, XL recessive (both AR and XL recessive RP have an earlier onset and more severe)

10.5.2 History

- Positive FHx
- Symptoms: nyctalopia, loss of peripheral visual field, loss of central vision, photopsias

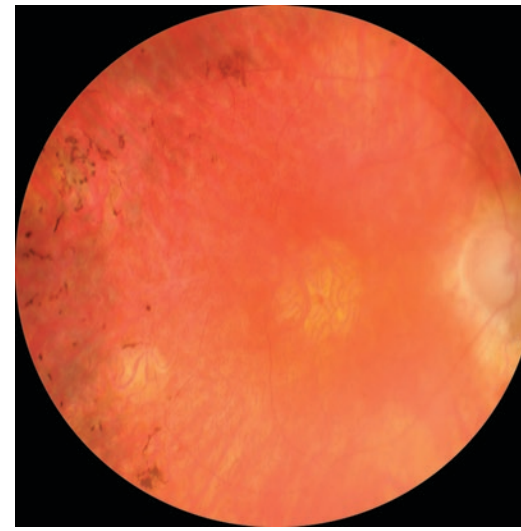

Fig. 10.7 Colour fundus image of a patient with RP showing disc pallor, attenuated retinal vessels, bone spicules and foveal and peripheral atrophy of the RPE and choriocapillaris

Table 10.5 Key facts about RP

- Term used for a group of disorders that are characterised by inherited, progressive dysfunction, cell loss, and eventual atrophy of retinal tissue
- Initial involvement of photoreceptors leads to subsequent damage to inner retinal cells. Eventually there is widespread atrophy of several, if not most, layers of the retina
- RP may be seen in isolation (typical RP) or in association with systemic disease in <25% of cases (syndromic RP)

10.5.3 Examination

- In typical RP, there is a high degree of symmetry of fundus abnormalities between the two eyes
- Bone spicule intraretinal pigmentation — represent RPE migration into the retina
- Optic nerve head pallor (from atrophy ± gliosis) ± optic disc drusen
- Attenuated retinal vessels
- Mottling and granularity of the RPE
- Atrophy of RPE and choriocapillaris with fundus pallor and visible large choroidal vessels in advanced disease
- Check IOP: RP patients at risk of open angle glaucoma
- Perform an anterior segment examination: look for cataracts (posterior subcapsular cataract) and keratoconus

10.5.4 Investigations

- Refraction: high myopia and astigmatism
- Kinetic VF: scotomas in the mid-periphery which coalesce to form a ring of VF loss (ring scotoma)
- EDT: scotopic ERG is more affected than photopic ERG, ERG extinguished in advanced RP, abnormal EOG
- OCT: CMO, ERM, decreased thickness of the ONL, loss of the ELM and IS/OS junction
- Genetic testing especially for RPE65 gene mutations

10.5.5 Treatment

- Refractive error correction
- Cataract extraction when indicated
- Treatment of CMO when present: topical/oral CAI's
- Visual impairment registration and referral for low vision aids
- Voretigene neparvovec if RPE65 gene mutations are present (NICE Guidance [HST11])

10.5.6 Syndromic RP

- Usher syndrome: AR sensorineural deafness (most commonly congenital), pigmentary retinopathy indistinguishable from typical RP, ERG is profoundly abnormal to undetectable — cochlear implantation
- Kearns-Sayre syndrome: mitochondrial DNA deletion syndrome, pigmentary retinopathy, CPEO, ptosis, cardiac conduction block, cerebellar ataxia — muscle biopsy shows red ragged fibers by light microscopy
- Refsum disease (infantile or adult-onset): AR, pigmentary retinopathy, peripheral neuropathy, ichthyosis, cardiac conduction defects, ERG responses are severely abnormal or unrecordable at all ages, high phytanic acid levels in blood and urine — restriction of dietary phytanic acid (dairy products) required
- Bardet-Biedl syndrome: AR, pigmentary retinopathy, polydactyly, congenital obesity, mental retardation, hypogonadism

10.6 Sector Retinitis Pigmentosa (Sector RP) (Fig. 10.8 and Table 10.6)

10.6.1 History

- Positive FHx

10.6.2 Examination

- Bone-spicule like pigmentary changes limited to one or two quadrants with minimal or no extension of the retinal area involved with time ± chorioretinal atrophy
- Retinal arteriolar attenuation in the affected quadrants only
- Cataract: posterior subcapsular lens opacity
- Sector RP can be differentiated from acquired pigmentation by its symmetry between the two eyes

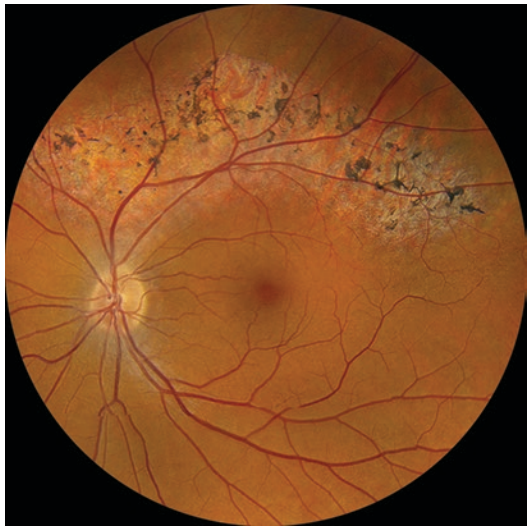

Fig. 10.8 Colour fundus image of a patient with superior sector RP

Table 10.6 Key facts about sector RP

• Specific subtype of RP: usually AD but can be AR
• Usually bilateral and symmetrical

10.6.3 Investigations

- Refraction
- VF: defects usually only in the regions of retinal pigmentation
- ERG: relatively good responses — relative preservation of amplitudes, with mild to moderate sub-abnormalities of both rod- and cone-mediated responses with normal implicit times
- Genetic testing
- OCT: look for CMO

10.6.4 Treatment

- Refractive error correction
- Cataract extraction when indicated
- Treatment of CMO when present: topical/oral CAI's
- Visual impairment registration and referral for low vision aids

10.6.5 Other Diagnoses to Consider

- Prior trauma (e.g. surgical such as a prior RD repair)
- Inflammation or infection: syphilis (congenital or acquired), congenital rubella (normal ERG, cataracts, hearing loss, no arteriolar attenuation), toxoplasmosis, DUSN
- Retinal toxicity: thioridazine, chlorpromazine, chloroquine, hydroxychloroquine, quinine
- Neoplasm (retinal or choroidal) and CAR (autoimmune paraneoplastic retinopathy)

10.7 Age-Related Macular Degeneration (AMD)

10.7.1 Classification of AMD

10.7.1.1 NICE Guidance [NG82]

- Early AMD
  - Low risk of progression
    - Medium drusen (63–124 μm)
    - Pigmentary abnormalities
  - Medium risk of progression
    - Large drusen (≥125 μm)
    - Reticular drusen
    - Medium drusen with pigmentary abnormalities
  - High risk of progression
    - Large drusen with pigmentary abnormalities
    - Reticular drusen with pigmentary abnormalities
    - Vitelliform lesion without significant visual loss (BCVA better than 6/18)
    - Atrophy smaller than 175 μm and not involving the fovea
- Late AMD (indeterminate)
  - RPE degeneration and dysfunction (presence of degenerative AMD changes with SRF or IRF in the absence of neovascularisation)
  - Serous PED without neovascularisation
- Late AMD (dry)
  - Geographic atrophy (in the absence of neovascular AMD)

  - Significant visual loss (6/18 or worse) associated with:
    Dense or confluent drusen
    Advanced pigmentary changes and/or atrophy
    Vitelliform lesion
- Late AMD (wet [neovascular] active)
  - Classic CNV
  - Occult (fibrovascular PED and serous PED with neovascularisation)
  - Mixed (predominantly or minimally classic CNV with occult CNV)
  - RAP
  - PCV
- Late AMD (wet [neovascular] inactive)
  - Fibrous scar
  - Subfoveal atrophy or fibrosis secondary to an RPE tear
  - Atrophy (absence or thinning of RPE and/or retina)
  - Cystic degeneration (persistent IRF or tabulations unresponsive to treatment)

10.7.1.2 The Age-Related Eye Disease Study (AREDS) Severity Scale

- Group 1 (No AMD): none or a few small drusen (<63 μm)
- Group 2 (Early AMD): any or all of the following — multiple small drusen; few intermediate drusen (63–124 μm); RPE abnormalities (increased pigmentation or depigmentation but not geographic atrophy)
- Group 3 (Intermediate AMD): any or all of the following — extensive intermediate drusen; at least one large drusen (≥125 μm, equivalent to the width of a major retinal vein at the optic disc edge), geographic atrophy that does not extend under the center of the macula
- Group 4 (Advanced AMD): presence of geographic atrophy extending under the center of the macula and/or presence of neovascular AMD

10.7.1.3 Rotterdam Classification System

- Grade 0 (no AMD): no signs of AMD at all **OR** hard drusen (<63 μm) only
- Grade 1 (preliminary early AMD): soft distinct drusen (≥63 μm) only **OR** pigmentary abnormalities only
- Grade 2 (early AMD): soft indistinct drusen (yellow lesions with indistinct borders and ≥125 μm in size)/reticular pseudodrusen only **OR** soft distinct drusen (≥63 μm) **AND** pigmentary abnormalities
- Grade 3 (early AMD): soft indistinct drusen (yellow lesions with indistinct borders and ≥125 μm in size)/reticular pseudodrusen **AND** pigmentary abnormalities
- Grade 4 (late AMD): atrophic, neovascular, or mixed AMD

10.7.2 History

- Ask about risk factors for AMD (NICE Guidance [NG82]):
  - Older age
  - Smoking
  - Positive FHx of AMD
  - HTN
  - Presence of AMD in other eye
  - BMI of 30 kg/m^2 or higher
  - Diet high in fat
  - Lack of exercise

10.7.3 Examination

- Non-neovascular (see Fig. 10.9): confluent pale yellow poorly defined soft drusen (intermediate 63–124 μm, large ≥125 μm [width of a retinal vein at the disc edge]) ± RPE focal hyperpigmentation (intraretinal pigment clumping from RPE migration associated with drusen) ± RPE hypopigmentation (associated with drusen) ± RPE atrophy (sharply delineated round or oval area of hypopigmentation or depigmentation with visible choroidal vessels)
- Neovascular (see Fig. 10.10): subretinal (red) or sub-RPE (grey) haemorrhage, subretinal or sub-RPE exudates, retinal or RPE detachment, subretinal fibrosis (disciform scar)

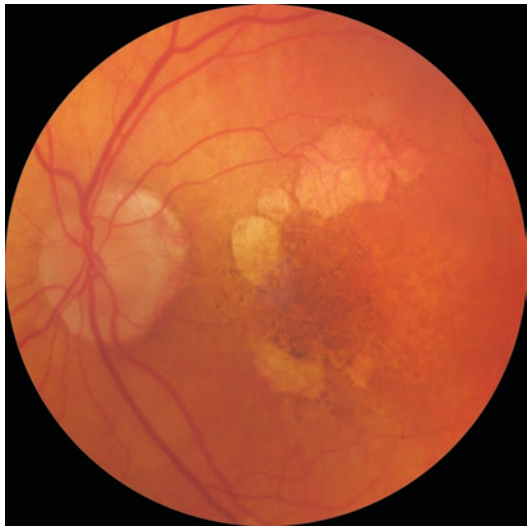

Fig. 10.9 Colour fundus image of a patient with late AMD (dry) showing the presence of geographic atrophy and advanced pigmentary changes

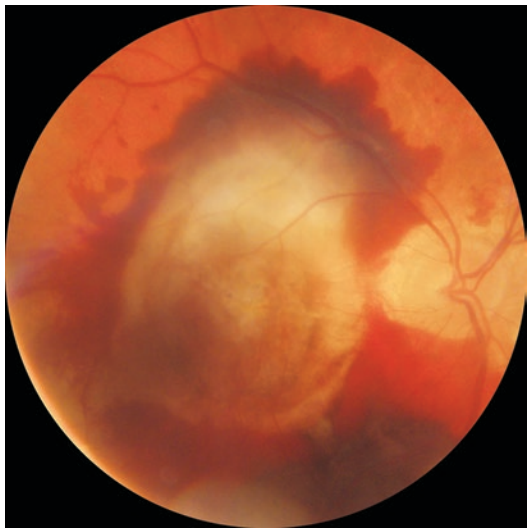

Fig. 10.10 Colour fundus image of a patient with late AMD (wet active) showing the presence of a subretinal haemorrhage with a disciform scar

10.7.4 Investigations

- OCT (see Sect. 2.1.3.5)
- FFA (see Sect. 2.3): perform if late AMD (wet active) suspected to confirm presence of CNV membrane
- ICG: branching vascular network (BVN) in PCV, "hot hyperfluorescent spot" in RAP

10.7.5 Treatment

- Non-neovascular
  - Visual impairment registration and referral to low vision aid service
  - Referral to support groups
  - Refraction with increased near add
  - Amsler grid: advise patients about new or progressive metamorphopsia
  - Lifestyle changes: smoking cessation, increased intake of food rich in macular carotenoids (spinach, broccoli, cabbage) and omega-3 fatty acids (oily fish such as salmon, mackerel, sardines), exercise
  - Vitamin supplementation (indications — advanced AMD in 1 eye, ≥1 large druse or extensive intermediate drusen in 1 or both eyes): AREDS 1 (vitamin C and E, β-carotene, zinc, copper) — 25% reduced risk of progression to advanced AMD at 5 years (The Age-Related Eye Disease Study Research Group 2001), AREDS 2 (vitamin C and E, zinc, copper, lutein, zeaxanthin) — 18% reduced risk of progression to advanced AMD at 5 years (The Age-Related Eye Disease Study 2 (AREDS2) Research Group)
  - Discharge from outpatient department
- Neovascular
  - Visual impairment registration and referral to low vision aid service
  - Referral to support groups
  - Refraction with increased near add
  - Amsler grid: advise patients about new or progressive metamorphopsia
  - Lifestyle changes: smoking cessation, increased intake of food rich in macular carotenoids (spinach, broccoli, cabbage) and omega-3 fatty acids (oily fish such as salmon, mackerel, sardines), exercise
  - Laser photocoagulation: extrafoveal or peripapillary CNV

– Anti-VEGF therapy:

Eligibility criteria (NICE Guidance [NG82]) in the UK:

- BCVA 6/12–6/96
- No permanent structural damage to the central fovea
- Greatest linear dimension of lesion size ≤12 DD
- Evidence of recent disease progression (recent VA changes or blood vessel growth on FFA)
- In eyes with BCVA worse than 6/96 consider anti-VEGF treatment for late AMD (wet active) only if a benefit in the person's overall visual function is expected (e.g. if the affected eye is the persons better seeing eye)

Ranibizumab (Lucentis, Novartis):

- Clinical trials:
  - **MARINA** (Rosenfeld et al. 2006): monthly Lucentis compared with sham injection for minimally classic or occult (see Sect. 5.4.2)
  - **ANCHOR** (Brown et al. 2006): monthly Lucentis compared with vPDT for predominantly classic CNV (see Sect. 5.4.1)
  - **PRONTO** (Fung et al. 2007; Lalwani et al. 2009): PRN dosing (see Sect. 5.4.3)
  - **PIER** (Regillo et al. 2008; Abraham et al. 2010): quarterly dosing (see Sect. 5.4.4)

Aflibercept (Eylea, Bayer):

- Clinical trials:
  - **VIEW 1** and **VIEW 2** (Heier et al. 2012): see Sect. 5.4.6

Bevacizumab (Avastin, Genentech/Roche):

- Clinical trials:
  - **CATT** (The CATT Research Group 2011): see Sect. 5.4.5
  - **IVAN** (Chakravarthy et al. 2013): see Sect. 5.4.7

Brolucizumab (Novartis)

- Not licensed by NICE at present
- Clinical trials:
  - **HAWK** and **HARRIER** (Dugel et al. 2020): see Sect. 5.4.8

10.7.6 Defining Risk for the Development of Advanced AMD

- The AREDS investigators devised a clinical scoring system defining risk categories for development of advanced AMD (The Age-Related Eye Disease Study Research Group 2005a)
- The scoring system tabulates a person score by assigning 1 risk factor to each eye of an individual for the presence of at least 1 large druse (within 2 DD of the foveal center) and 1 risk factor for the presence of any pigment abnormality (increased pigmentation thought to be attributed to AMD, RPE depigmentation, or areas of noncentral geographic atrophy)
- Risk factors are summed across both eyes on which the 5-year risk of developing advanced AMD in at least one eye can be approximated
- Risk of developing advanced AMD is as follows: total score 0, 0.5% risk; 1 factor, 3%; 2 factors, 12%; 3 factors, 25%, and 4 factors, 50%
- Modifications of the scale award persons without any large drusen 1 risk factor if medium sized drusen are present in both eyes and individuals with advanced AMD in their first eye receive a score of 2 for that eye when tabulating the person score to estimate the risk for their fellow eye

10.7.7 Other Diagnoses to Consider

- Polypoidal choroidal vasculopathy (PCV — see Fig. 10.11): black and asian populations, reddish-orange polypoidal lesions and is often associated with serosanguineous PEDs, branching vascular network (BVN) appear as a shallow elevation of the RPE, while the polypoidal lesions appear as sharper protuberances

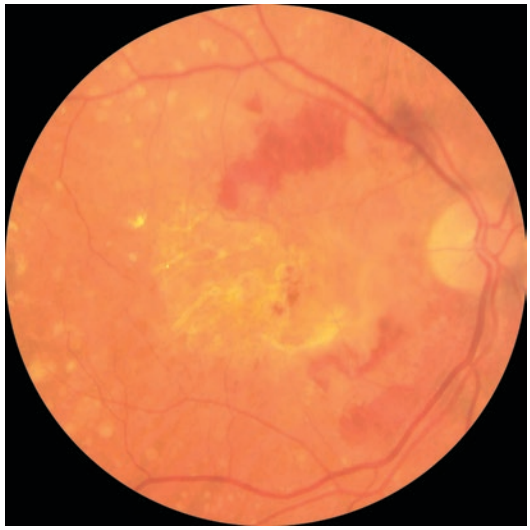

Fig. 10.11 Colour fundus image of a patient with PCV showing a peripapillary and macula subretinal haemorrhage

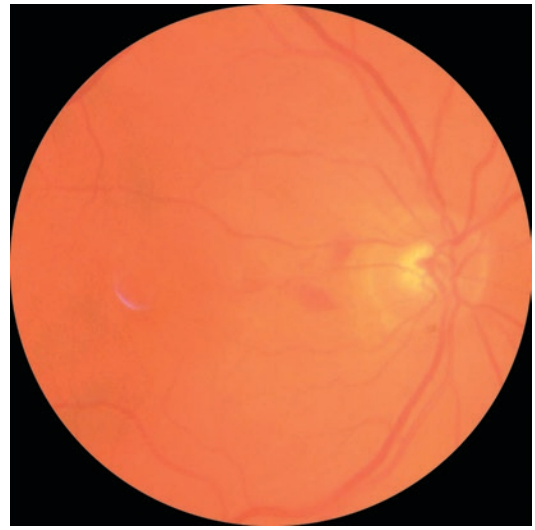

Fig. 10.12 Colour fundus image of a patient with a peripapillary CNV membrane

10.8 Peripapillary Choroidal Neovascular (CNV) Membrane (Fig. 10.12 and Table 10.7)

10.8.1 Causes of Peripapillary CNV Membrane

- Degenerative
  - AMD
  - Angioid streaks
  - Degenerative myopia
  - Laser photocoagulation scars
  - Traumatic choroidal ruptures
- Vascular
  - PCV
- Inflammatory
  - Birdshot chorioretinopathy
  - POHS
  - Sarcoidosis
  - Serpiginous choroiditis
- Optic nerve malformations
  - Optic disc drusen
  - Optic disc pits
  - Retinochoroidal colobomas
- Neoplastic
  - Choroidal naevi
  - Choroidal osteoma
- Idiopathic (up to 39% of cases)

Table 10.7 Key facts about peripapillary CNV membrane

- Peripapillary CNV membranes are defined as a collection of new choroidal blood vessels, any portion of which lies within 1 DD of the nerve head
- Account for 7–10% of all CNV membranes
- Clinical manifestations only occur if the membrane extends over the macula, if the vessels haemorrhage into the subretinal space, or fluid exudation occurs within the macula
- Two types of peripapillary CNV membranes: type 1 — CNV membrane is sub-RPE (e.g. AMD), type 2 — CNV membrane in the subretinal space (e.g. POHS)

10.8.2 Examination

- Peripapillary (within 1 DD) SRF ± subretinal haemorrhages
- Peripapillary fibrotic scar

10.8.3 Investigations

- OCT (see Fig. 10.13): look for presence of SRF
- FFA: look for leakage confirming presence of a CNV membrane
- ICG: look for PCV

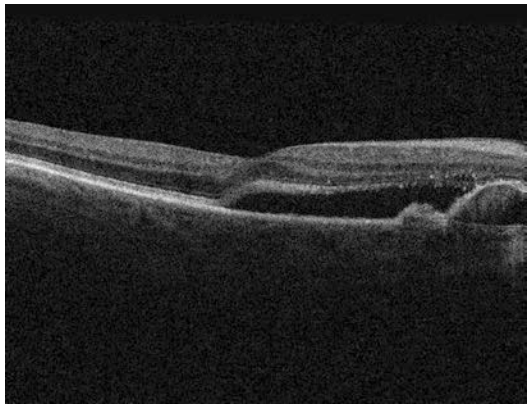

Fig. 10.13 OCT image of a patient with a peripapillary CNV membrane showing SRF tracking to the fovea

10.8.4 Treatment

- Indication: treat when fluid threatening or involving the fovea with reduction of vision
- Options:
  - Anti-VEGF agents: maintains the integrity of the papillomacular bundle
  - Laser photocoagulation: induce scar formation, thermal injury, vitreous haemorrhage, BRAO, damage to the papillomacular bundle

10.9 Myopic Macular Degeneration (Fig. 10.14, Tables 10.8 and 10.9)

10.9.1 Examination

- Tilted optic discs
- Peripapillary atrophy (predominantly temporal to optic disc)
- Patchy or diffuse chorioretinal atrophy: visibility of choroidal vessels
- Lacquer cracks (ruptures of Bruch's membrane): yellowish linear lesions
- Posterior staphyloma: outward protrusion of all layers of the posterior eye globe
- Macular CNV (macular haemorrhage) ± elevated pigmented forster fuchs spot

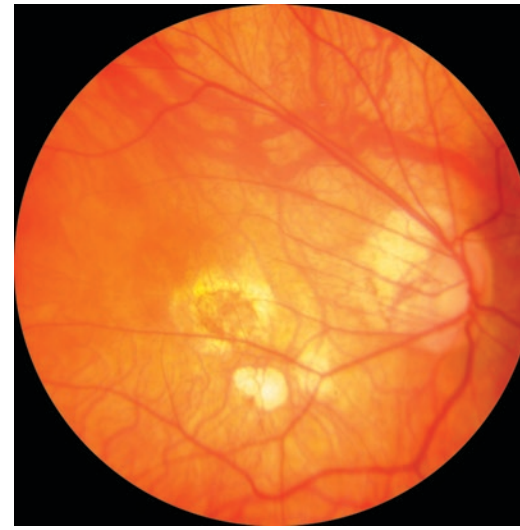

Fig. 10.14 Colour fundus image of a patient with myopic macular degeneration showing diffuse chorioretinal atrophy, tilted optic disc, peripapillary atrophy and a forster fuchs spot

Table 10.8 Myopia definitions

• High myopia is defined as myopia −6.00 D or more or an axial length ≥26.5 mm
• Pathologic myopia is defined as myopia over −8.00 D or an axial length >32.5 mm

Table 10.9 Associations of myopia

• Connective tissue disorders:
– Sticklers syndrome
– Marfan syndrome
– Ehlers-Danlos syndrome
• Infectious:
– Congenital rubella
• Chorioretinal dystrophies:
– Gyrate atrophy
• Others:
– Albinism
– Downs syndrome

10.9.2 Investigations

- Refraction
- B-scan US: posterior staphyloma
- OCT: posterior staphyloma, myopic foveoschisis (split layers have bridge columns between

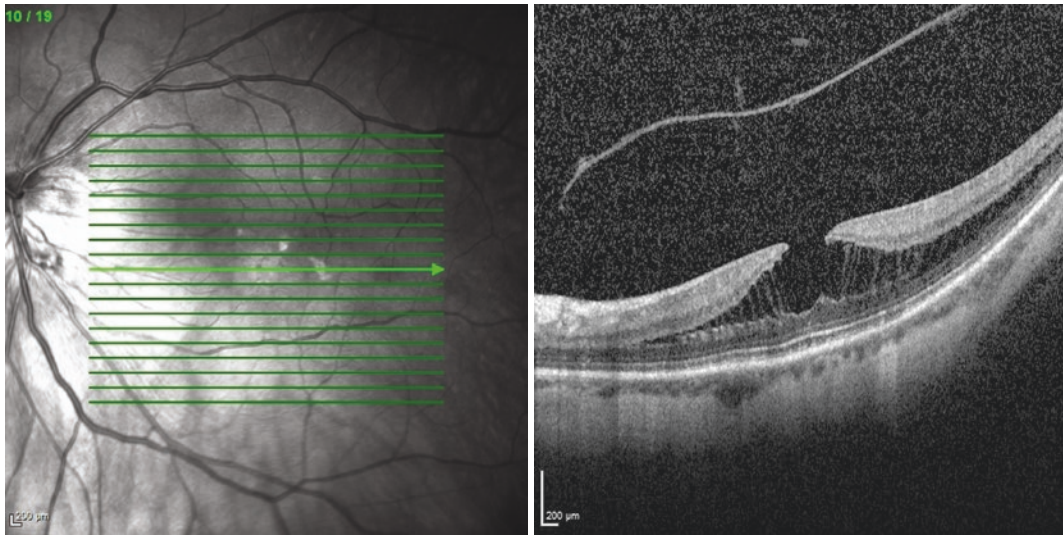

Fig. 10.15 OCT image of a patient with myopic foveoschisis

them, ILM detachment, well preserved IS/OS junction — see Fig. 10.15), macular hole
- FFA: classic CNV membrane

10.9.3 Treatment

- CNV
  - Laser photocoagulation for extrafoveal CNV
  - Anti-VEGF therapy for subfoveal CNV:
    Ranibizumab (Lucentis, Novartis):
    - NICE Guidance [TA298]: ranibizumab is recommended as an option for treating visual impairment due to choroidal neovascularisation secondary to pathological myopia
    - **RADIANCE** study (Wolf et al. 2014): comparison of vPDT and Lucentis — after 3 months of treatment Lucentis showed significantly greater gain in no. of ETDRS letters

    Aflibercept (Eylea, Bayer):
    - NICE Guidance [TA486]: aflibercept is an option treating visual impairment because of myopic choroidal neovascularisation in adults
    - **MYRROR** study (Ikuno et al. 2015): comparison of Eylea and sham injection — at week 24 a gain of 12.1 letters and loss of 2 letters was seen in the Eylea and sham group, respectively.
- Myopic foveoschisis
  - If foveal detachment is found on OCT images, macular hole formation is likely to start in the near future and surgery must be planned soon (1–2 months)
  - If retinoschisis present with no foveal retinal detachment — not an indication for surgery

10.10 Branch Retinal Vein Occlusion (BRVO) (Figs. 10.16, 10.17, and Table 10.10)

10.10.1 Risk Factors

- Arteriosclerosis: HTN, DM, smoking, hyperlipidaemia (including secondary to hypothyroidism)
- Glaucoma
- Ocular inflammatory disease: Behcet's disease, PAN, sarcoidosis, SLE
- Haematological: Protein C, protein S or anti-thrombin deficiency, activated protein C resistance, factor V Leiden, myeloma, Waldenstrom's macroglobulinaemia, antiphospholipid syndrome

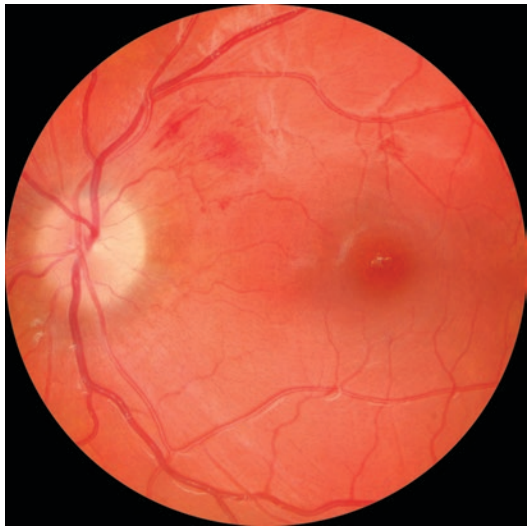

Fig. 10.16 Colour fundus image of a patient with a superior macular BRVO

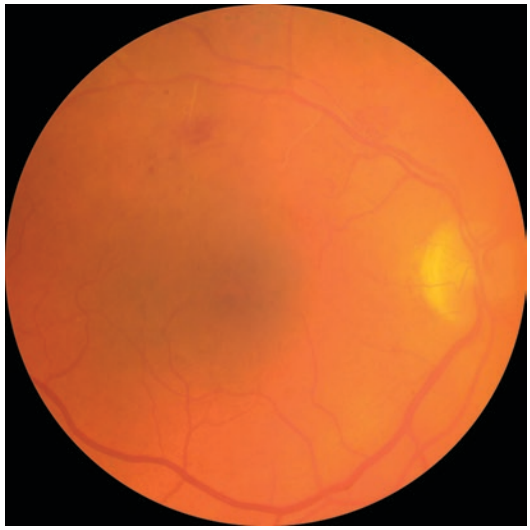

Fig. 10.17 Colour fundus image of a patient with a superotemporal BRVO with NVE

Table 10.10 Key facts about BRVO

- Mostly occurs at arteriovenous crossings
- Retinal artery and vein share a common adventitial sheath — lumen of vein compressed by arteriosclerotic artery at the crossing site
- Venous obstruction leads to elevation of venous pressure that may overload the collateral drainage capacity and lead to macular oedema and ischaemia

10.10.2 Examination

- Acute: wedge shaped segmental distribution of intraretinal haemorrhage in a quadrant of the fundus, narrowed branch retinal vein passing under a retinal artery, dilated and tortuous retinal vein, cotton wool spots
- Chronic: telangiectatic vessels (dilation of capillaries) forming collaterals that cross the horizontal raphe, microaneurysms, exudates, sclerosed retinal vein ± NVE > NVD > NVI
- Check for RAPD
- Check the IOP
- Look for NVI on anterior segment examination
- Perform gonioscopy for NVA

10.10.3 Investigations (RCOphth RVO July 2015 Guidelines)

RCOphth Retinal Vein Occlusion (RVO) Guidelines 2015:

- BP
- Bloods: FBC, ESR, glucose
- OCT: CMO
- FFA (if uncertain diagnosis): delayed filling of the occluded retinal vein, capillary non-perfusion — >5 DD is defined as an ischaemic BRVO (The Branch Vein Occlusion Study Group 1984, 1986), macular ischaemia, telangiectatic vessels forming collaterals that cross the horizontal raphe

10.10.4 Treatment

- Control of cardiovascular risk factors: HTN, hyperlipidaemia, smoking, DM
- Neovascularisation: sectoral PRP — branch vein occlusion study — BVOS (The Branch Vein Occlusion Study Group 1984, 1986) recommended that laser photocoagulation be applied only after NV is observed, which reduces the likelihood of vitreous haemorrhage from about 60% to 30%

- Macular oedema
  - Laser photocoagulation:

    BVOS (The Branch Vein Occlusion Study Group 1984, 1986):
    - An RCT that aimed to answer three questions: (1) can laser photocoagulation improve VA compared to observation in eyes with macular oedema from BRVO that reduces the vision to 6/12 or worse; (2) Can sectoral PRP prevent the development of NV; (3) Can sectoral PRP prevent vitreous haemorrhage
    - Wait for at least 3 months if VA 6/12 or worse before considering laser therapy to allow clearing of intraretinal haemorrhages to permit FFA and evaluation of macular oedema and macular ischaemia
    - If perfused macular oedema accounts for the visual loss, and vision continues to be 6/12 or worse without spontaneous improvement, consider grid macular photocoagulation (0.1 s duration, 100 μm spot size, power titrated to produce a "medium" white burn) to the leaking area demonstrated by FFA
    - After 3 years of follow-up, 65% of treated eyes gained 2 or more lines of vision compared to 37% of untreated eyes
  - Dexamethasone implant (Ozurdex, Allergan):

    NICE Guidance [TA 229]: recommend as an option for treatment of macular oedema due to a BRVO when treatment with laser photocoagulation has not been beneficial or treatment with laser photocoagulation is not considered suitable because of the extent of retinal haemorrhages

    GENEVA study (Haller et al. 2010): A RCT that evaluated the effects of a single dexamethasone implant compared to sham injection for the treatment of macular oedema secondary to BRVO and CRVO. Increase in BCVA of ≥15 ETDRS letters was achieved in 30% of the Ozurdex 0.7 mg group, 26% of the Ozurdex 0.35 mg group, and 13% of the sham group 60 days after injection (peak response) and was maintained through day 90 in BRVO patients. There was no difference between either of Ozurdex groups and the sham group at day 180.
  - Ranibizumab (Lucentis, Novartis):

    NICE Guidance [TA283]: recommend as an option for treatment of macular oedema due to a BRVO when treatment with laser photocoagulation has not been beneficial or treatment with laser photocoagulation is not considered suitable because of the extent of retinal haemorrhages

    BRAVO study (Campochiaro et al. 2010): A RCT to evaluate the efficacy and safety of Lucentis in the treatment of macular oedema from BRVO. Patients were randomised into 3 groups: (1) sham injection; (2) 0.3 mg Lucentis; and (3) 0.5 mg Lucentis. In the first 6 months, injections were given monthly. At 6 months, both Lucentis groups gained +16.6 and +18.3 ETDRS letters (0.3 mg and 0.5 mg groups, respectively) compared with a gain of +7.3 letters in the control group. At 6 months, 55.2% and 61.1% of patients receiving Lucentis 0.3 mg and 0.5 mg gained ≥3 ETDRS lines compared to 28.8% in the sham injection group
  - Aflibercept (Eylea, Bayer):

    NICE Guidance [TA305]: recommend as an option for treatment of visual impairment caused by macular oedema following BRVO

    VIBRANT study (Campochiaro et al. 2015): A RCT that evaluated the efficacy and safety of Eylea compared to macular grid laser in the treatment of macular oedema from BRVO or HRVO. After 6 months of treatment, 26.7% of the laser group gained ≥3 ETDRS lines compared to 52.7% in the

Eylea group. Mean change in letters from baseline was +6.9 letters in the laser group compared to +17.0 letters in the Eylea group.

10.10.5 Prognosis

- BVOS (The Branch Vein Occlusion Study Group 1984, 1986)
  - Only eyes with ischaemic BRVO (>5 DD of retinal capillary non-perfusion) are at risk of developing NV — 40% of these eyes develop NV, and of these 40%, 60% will experience periodic vitreous haemorrhage
  - Retinal or disc NV, or both, may develop at any time within the first 3 years after an occlusion but are most likely to appear within the first 6–12 months after the occlusion
  - Up to 10% of patients with BRVO in one eye will develop any type of RVO in the fellow eye

10.10.6 Other Diagnoses to Consider

- Macular telangiectasia
- Susac syndrome: BRAO, sensorineural hearing loss, subacute encephalopathy, hyperintense lesions in the corpus callosum on T2 MRI
- Diabetic retinopathy
- Radiation retinopathy
- Retinal inflammation: Sarcoidosis, Behcet's disease

10.11 Central Retinal Vein Occlusion (CRVO) (Fig. 10.18)

10.11.1 Risk Factors

- Arteriosclerosis: HTN, DM, smoking, hyperlipidaemia (including secondary to hypothyroidism)
- Glaucoma
- Ocular inflammatory disease: Behcet's disease, PAN, sarcoidosis, SLE

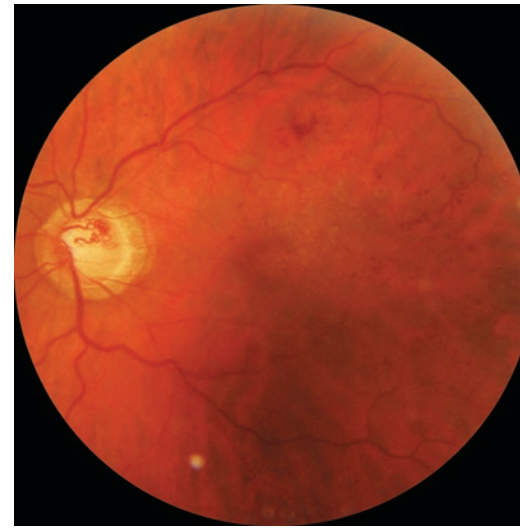

Fig. 10.18 Colour fundus image of a patient with a chronic CRVO showing the presence of telangiectatic vessels and optociliary shunt vessels

- Haematological: Protein C, protein S or antithrombin deficiency, activated protein C resistance, factor V Leiden, myeloma, Waldenstrom's macroglobulinaemia, antiphospholipid syndrome
- Pharmacological: oral contraceptive pill

10.11.2 Examination

- Acute: retinal haemorrhages (flame shaped and deep blot type) in all four quadrants of the fundus with a dilated tortuous retinal venous system, cotton wool spots, optic disc swelling, CMO
- Chronic: optociliary shunt vessels (collateral vessels connecting the choroidal and the retinal vasculature — do not leak on FFA - see Fig. 10.19 and Table 10.11), telangiectatic capillary bed, persistent dilatation and tortuosity, perivenous sheathing, NVI > NVD > NVE, CMO, glaucomatous optic neuropathy
- Check for a RAPD
- Check the IOP
- Look for NVI on anterior segment examination
- Perform gonioscopy for NVA

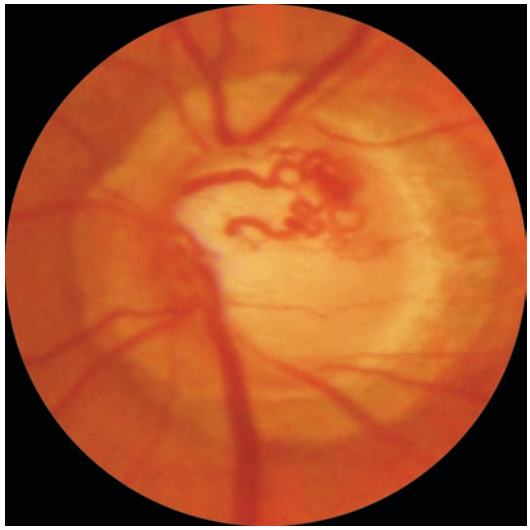

Fig. 10.19 Colour fundus image of a patient with optociliary shunt vessels from a chronic CRVO

Table 10.11 Causes of optociliary shunt vessels

• CRVO
• Chronic glaucoma
• Chronic papilloedema
• Optic nerve sheath meningioma

10.11.3 Investigations

RCOphth RVO Guidelines 2015:

- BP
- Bloods — FBC (leukaemia), ESR (myeloma), glucose
- OCT — CMO
- FFA (uncertain diagnosis) — delayed filling of the occluded retinal vein, capillary non-perfusion, macular ischaemia, telangiectatic vessels forming collaterals that cross the horizontal raphe

10.11.4 Treatment

- Control of cardiovascular risk factors: HTN, hyperlipidaemia, smoking cessation, DM
- RCOphth RVO guideline 2015 recommends that oestrogen containing HRT and OCP should not be commenced in women with history of RVO. However, the continued use in a patient who develops RVO does not appear to be associated with a higher rate of recurrence
- Neovascularisation:
  - Full scatter PRP — central vein occlusion study — **CVOS** (The Central Vein Occlusion Study Group 1997) recommended that laser photocoagulation be applied only after NV is observed, with greater resolution of NVI/NVA by 1 month after PRP in 56% of no early treatment eyes (no NVI or NVA present) compared with 22% of early treatment eyes (NVI or NVA present)
- Macular oedema
  - Dexamethasone Implant (Ozurdex, Allergan):

    NICE Guidance [TA229]: recommend as an option for the treatment of macular oedema following CRVO

    GENEVA study (Haller et al. 2010): A RCT that evaluated the effects of a single dexamethasone implant compared to sham injection for the treatment of macular oedema secondary to BRVO and CRVO. Increase in BCVA of ≥15 ETDRS letters was achieved in 29% of the Ozurdex 0.7 mg group and 9% of the sham group 60 days after injection but not at 90 or 180 days.
  - Ranibizumab (Lucentis, Novartis):

    NICE Guidance [TA283]: recommend as an option for treatment of visual impairment caused by macular oedema following CRVO

    CRUISE study (Brown et al. 2010): A RCT to evaluate the efficacy and safety of Lucentis in the treatment of macular oedema from CRVO. Patients were randomised into 3 groups: (1) sham injection; (2) 0.3 mg Lucentis; and (3) 0.5 mg Lucentis. In the first 6/12, injections were given monthly. At 6/12, both Lucentis groups gained +12.7 and +14.9 ETDRS letters (0.3 mg and 0.5 mg groups, respectively) compared with a gain of +0.8 letters in the control group. At 6 months, 46.2% and 47.7% of patients receiving Lucentis 0.3 mg and 0.5 mg gained ≥3 ETDRS lines compared to 16.9% in the sham injection group
  - Bevacizumab (Avastin, Roche/Genetech):

    Off license treatment, where licensed treatments are not available

  - Aflibercept (Eylea, Bayer):
    NICE Guidance [TA305]: recommend as an option for treatment of visual impairment caused by macular oedema following CRVO
    CORPERNICUS study (Boyer et al. 2012): comparison of Eylea and sham injection — the proportion of patients who gained ≥15 letters from baseline in the Eylea and sham groups was 56.1% and 12.3% at week 24, respectively
    GALILEO study (Holz et al. 2013): comparison of Eylea and sham injection — the proportion of patients who gained ≥15 letters from baseline in the Eylea and sham groups was 60.2% and 22.1% at week 24, respectively
- Delay in initiating treatment up to 6 months results in fewer visual gains compared to immediate initiation of treatment. Therefore, treatment should be initiated as soon as the diagnosis is established (RCOphth RVO Guidelines 2015)
- Careful consideration should be given to further therapy in such eyes that do not improve in terms of snellen VA or OCT central subfield thickness after three loading injections at monthly intervals and treatment with anti-VEGF is not recommended if no response occurs after six injections (RCOphth RVO Guidelines 2015)
- No robust data on outcomes of switching steroid to an anti-VEGF agent or switching between anti-VEGF agents or combining steroids with anti-VEGF agents for macular oedema due to CRVO (RCOphth RVO Guidelines 2015). However, eyes with suboptimal response to steroids may be switched to anti-VEGF therapy or vice versa.

10.11.5 Follow up

- Non-ischaemic CRVO (may resolve completely without any complications): initial follow up every 3 months for 6 months, follow up for at least 2 years but the development of disc collaterals and the resolution of macular oedema for at least 6 months should allow the discharge of the patient from clinical supervision (RCOphth RVO Guidelines 2015)
- Ischaemic CRVO (see Table 10.12): follow up after 6 months should be every 3 months for 1 year (RCOphth RVO Guidelines 2015)
- RCOphth RVO guidelines 2015 recommends that oestrogen containing HRT and OCP should not be commenced in women with history of RVO. However, the continued use in a patient who develops RVO does not appear to be associated with a higher rate of recurrence

Table 10.12 Features suggestive of an ischaemic CRVO

- Poor VA (CVOS — 44% of eyes with vision of <6/60 develop rubeosis)
- RAPD
- Presence of multiple dark deep intraretinal haemorrhages
- Presence of multiple cotton wool spots
- Degree of venous dilation and tortuosity
- FFA showing greater than 10-disc areas of capillary non-perfusion on 7 field FFA (CVOS)
- ERG: reduced b wave amplitude, reduced b:a ratio (negative ERG) and prolonged b-wave implicit time

10.11.6 Prognosis

- At 3 years, there was a 45% chance of developing neovascular glaucoma after onset of ischaemic CRVO — highest risk if VA <6/60 or >10 DD of non-perfusion on FFA (The Central Vein Occlusion Study Group 1997)
- Overall, 34% of initially perfused eyes converted to non-perfused status after 3 years (The Central Vein Occlusion Study Group 1997)
- Risk of CRVO in contralateral eye is 5% by 1 year (RCOphth RVO Guidelines 2015)

10.12 Central Retinal Artery Occlusion (CRAO) (Fig. 10.20)

10.12.1 Risk Factors

- Arteriosclerosis: HTN (60%), DM (25%), smoking, hyperlipidaemia
- Embolic sources: carotid artery disease, aortic disease (dissection), cardiac valve vegetations

(infective endocarditis), cardiac tumours (atrial myxoma), arrhythmias, cardiac septal defects
- Inflammatory disease: GCA, PAN, GPA, SLE
- Haematological: antiphospholipid syndrome, leukaemia, lymphoma,

10.12.2 Examination

- Acute — retinal whitening in the posterior pole (ischaemic damage to the inner half of the retina), cherry red spot (normal appearing retina and is observed in high contrast against the surrounding opacified retina), box carring of retinal arteries, retinal emboli (Hollenhorst plaque — yellow refractile cholesterol embolus frequently found at bifurcation sites, calcific emboli — white, non-refractile, proximal to optic disc, platelet-fibrin clot — elongated), optic disc oedema
- Chronic (fundus may appear featureless) — optic atrophy, retinal arterial attenuation, cilioretinal collaterals, macular RPE changes, cotton wool spots, NVD (2%), NVE, NVI (18%)
- Check for a RAPD
- Check the IOP
- Look for NVI on anterior segment examination

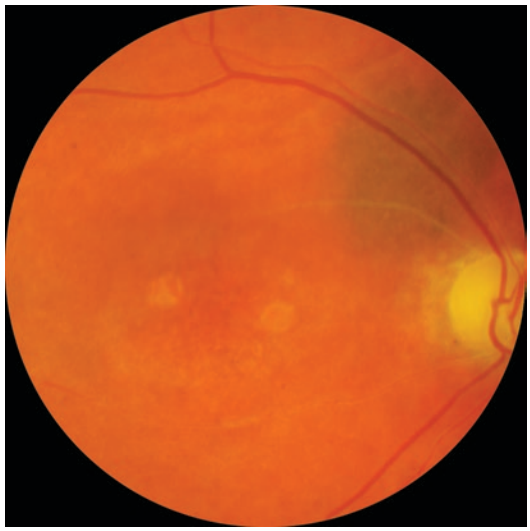

Fig. 10.20 Colour fundus image of a patient with a long standing CRAO

10.12.3 Investigations

- BP
- Glucose
- FBC, CRP, ESR ± temporal artery biopsy (TAB): rule out GCA if patient age >50 years
- OCT: inner retinal oedema in acute CRAO, inner retinal atrophy with outer retinal preservation in long standing CRAO (see Fig. 10.21)
- FFA: delayed filling of the retinal arteries (see Fig. 10.22)
- HVF: altitudinal defect
- ERG: negative ERG with the scotopic white stimulus

10.12.4 Treatment

- If within 4 h of onset: reduce IOP with 500 mg IV diamox, ocular massage (dislodge emboli), AC paracentesis (all treatment options described here have limited evidence for its effectiveness)

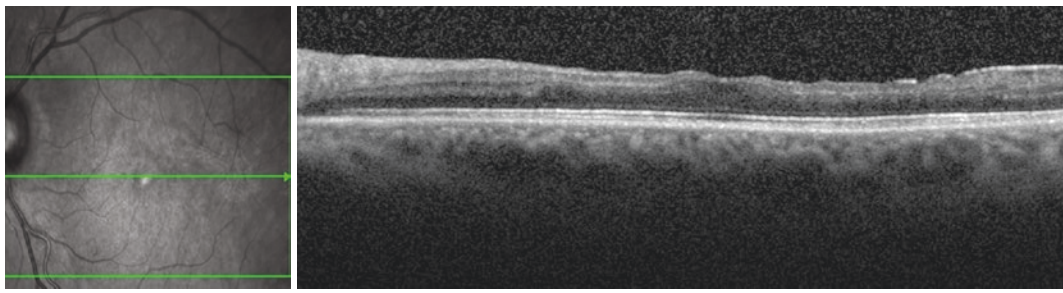

Fig. 10.21 OCT image of a patient with a chronic CRAO with inner retinal atrophy

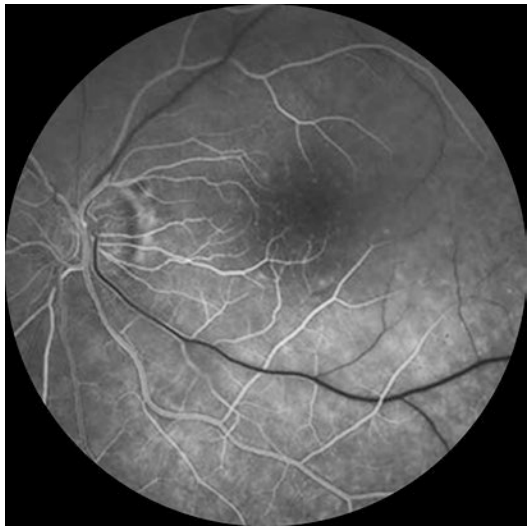

Fig. 10.22 Fundus fluorescein angiogram of a patient with CRAO showing delayed filling of the retinal arteries

- If GCA suspected: start steroids and arrange TAB
- If embolic, refer to stroke (TIA) clinic for carotid doppler ± echocardiography
- Treat NV: PRP

10.13 Diabetic Retinopathy (DR) (Fig. 10.23)

10.13.1 Proposed International Clinical Diabetic Retinopathy and Diabetic Macular Edema Severity Scales 2003

- Mild Non-proliferative DR (NPDR)
  - Microaneurysms (MAs) only
- Moderate NPDR
  - More than just MA's but less than severe NPDR
- Severe NPDR (4-2-1 rule)
  - One or more of the following:
    - More than 20 intraretinal haemorrhages in each of 4 quadrants
    - Definite venous beading in 2 or more quadrants
    - Prominent intraretinal microvascular abnormalities (IRMA) in 1 or more quadrants
- Very severe NPDR
  - Two or more features of severe NPDR present
- Proliferative DR (PDR)
  - One or more of the following:
    - Extraretinal neovascularisation
    - Vitreous or preretinal haemorrhage
- Mild DMO
  - Some retinal thickening or hard exudates in the posterior pole, distant from the center of the macula
- Moderate DMO:
  - Retinal thickening or hard exudates near the center of the macula but not involving the center
- Severe DMO:
  - Retinal thickening or hard exudates involving the center of the macula

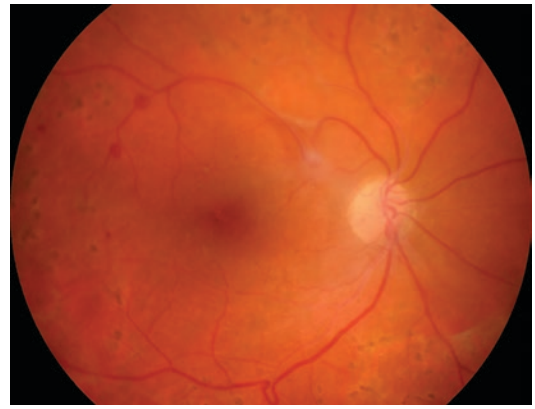

Fig. 10.23 Colour fundus image of a patient with active PDR; PRP commenced but incomplete

10.13.2 High-Risk PDR

- Defined by the Diabetic Retinopathy Study (The Diabetic Retinopathy Study Research Group 1981) with patients at higher risk of visual loss and requires prompt PRP treatment
  - Neovascularisation of the disc (NVD — new vessels on or within 1DD of the optic

disc): NVD ≥¼ disc area or any size NVD with vitreous and/or preretinal haemorrhage
- Neovascularisation elsewhere in the retina (NVE): NVE ≥½ disc area with vitreous and/or preretinal haemorrhage

10.13.3 Clinically Significant Macular Oedema (CSMO)

- Defined by the Early Treatment of Diabetic Retinopathy Study (Early Treatment Diabetic Retinopathy Study Research Group. ETDRS Report number 9. 1991a):
  - Retinal thickening at or within 500 μm of the centre of the macula and/or
  - Hard exudates at or within 500 μm of the centre of the macula, if associated with thickening of the adjacent retina and/or
  - Zone or zones of retinal thickening 1-disc area or larger, any part of which is within 1 DD of the center of the macula

10.13.4 Examination

- NPDR
  - MAs (small red dots)
  - Intraretinal haemorrhages: dot and blot haemorrhages, flame haemorrhages (located in NFL)
  - Hard exudates: sharply demarcated yellow-white deposits within the retina
  - Cotton wool spots (CWS — patches of relative ischaemia affecting the NFL of the retina): small white patches with wispy borders situated in the inner retina
  - Venous beading: localised areas of change in vessel calibre with alternating regions of relative dilation and constriction
  - Venous loops
  - IRMA (see Fig. 10.24): segments of dilated and tortuous retinal vasculature without crossing both arterioles or veins in the underlying retina
  - Subretinal fibrosis
- PDR

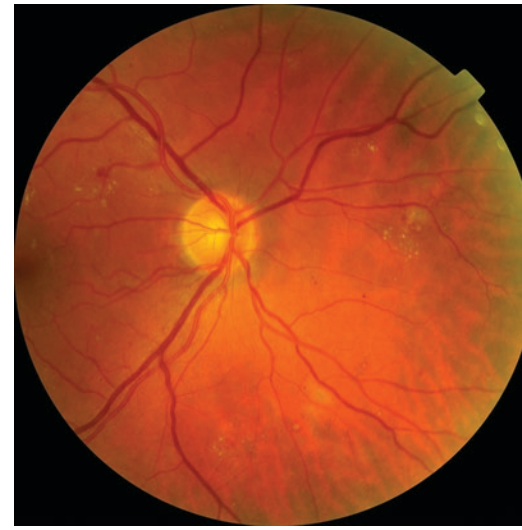

Fig. 10.24 Colour fundus image of a patient with severe NPDR showing the presence of IRMA

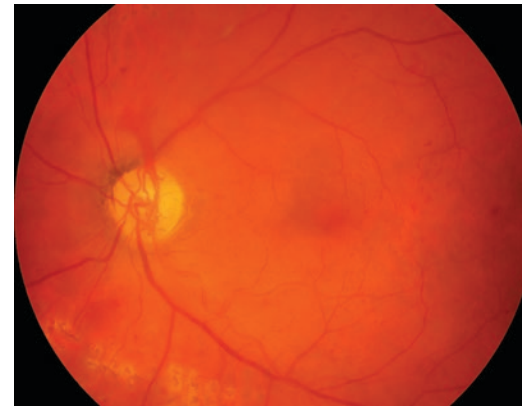

Fig. 10.25 Colour fundus image of a patient with PDR with active NVD, and incomplete PRP

  - NVD (see Fig. 10.25): wheel like network of vessels on or within 1 DD of the disc
  - NVE (see Fig. 10.26): superficial location lying over retinal veins, formation of wheel-like networks, cross both arterioles and veins in the underlying retina, regressing new vessels appear to become sheathed
  - NVI/NVA
  - Concave tractional RD (see Fig. 10.27) ± distortion or displacement (dragging) of the macula

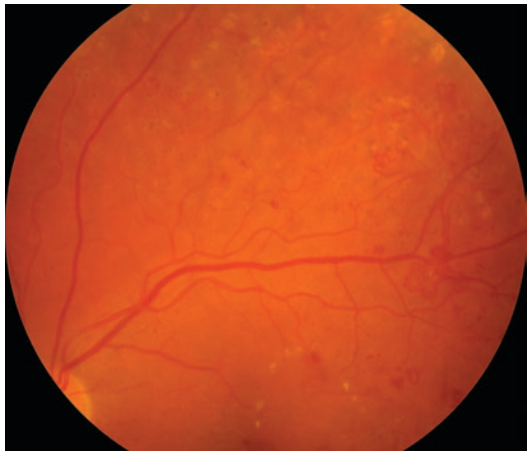

Fig. 10.26 Colour fundus image of a patient with PDR with active NVE (PRP commenced)

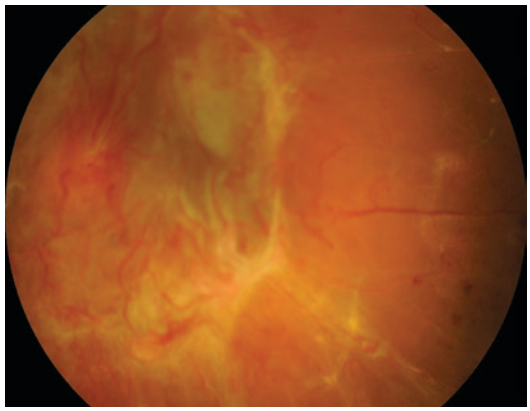

Fig. 10.27 Colour fundus image of a patient with PDR with a tractional RD involving the fovea

10.13.5 Investigations

- Venous fasting glucose ≥7.0 mmol/L ± oral glucose tolerance test (75 g anhydrous glucose) with a 2-h value ≥11.1 mmol/L ± random venous glucose ≥11.1 mmol/L
- OCT
  - Hard exudates appear as hyperreflective foci within the retina
  - Macular oedema
  - Traction from ERM or VMT causing underlying macular oedema in the absence of retinal vascular leakage demonstrable by FFA
- FFA
  - MAs appear as hyperfluorescent dots visible during the arteriovenous transit phase
  - Intraretinal haemorrhages appear hypofluorescent blocking normal fluorescence from the underlying choroid
  - Hard exudates appear hypofluorescent
  - CWS appear hypofluorescent
  - IRMA appear hyperfluorescent during the arteriovenous transit phase and are often situated at the borders of areas of capillary non-perfusion
  - Macular oedema: petaloid pattern of leakage
  - NVD/NVE: leakage

10.13.6 Treatment

- NPDR
  - Modification of life-style:
    Smoking cessation
    Weight loss
    Exercise
  - Modification of systemic risk factors:
    Hyperglycaemia control:
    - **DCCT** (Diabetes Control and Complications Trial Research Group 1995): type 1 DM — tight control of HbA1C at mean of 7.2% was associated with 76% reduction in onset of retinopathy and slowed progression of DR by 54%, 60% reduction in onset of neuropathy, and 54% reduction in onset of nephropathy at 6.5 years
    - **UKPDS** (UK Prospective Diabetes Study Group 1998): type 2 DM — tight control of HbA1C at mean of 7% was associated with a 25% reduction in the onset of microvascular disease
    - **ACCORD** (The ACCORD Study Group and ACCORD Eye Study Group 2010): type 2 DM — intensive control of HbA1C reduced progression of DR by 42% and reduced development of PDR from 10.2% to

6.5%: avoid Pioglitazone in the presence of macular oedema, personalized HbA1c target should be set, usually between 48–58 mmol/mol (6.5–7.5%)

BP control:

- **UKPDS** (UK Prospective Diabetes Study Group 1998): type 2 DM — tight control of BP at mean of 144/82 was associated with a 37% reduction in the onset of microvascular disease and 32% reduction in diabetes related deaths

Lipid control:

- **ACCORD** (The ACCORD Study Group and ACCORD Eye Study Group 2010): type 2 DM — 40% reduction in the odds of having progression of DR over 4 years in patients allocated to fenofibrate in combination with a statin, compared to simvastatin alone): consider adding fenofibrate to a statin for NPDR in type 2 DM, avoid statins in pregnancy

- CSMO
  - Focal/grid laser:

    ETDRS (Early Treatment Diabetic Retinopathy Study Research Group. ETDRS Report number 9. 1991a): focal/grid laser reduced the risk of moderate vision loss (loss of ≥15 letters from baseline) by 50% at 3 years in eyes with mild or moderate NPDR with CSMO

    Indicated if CMT <400 μm and CSMO is not center involving (RCOphth Diabetic Retinopathy Guidelines 2012)
  - Anti-VEGF therapy:

    Aflibercept (Eylea, Bayer):
    - NICE Guidance [TA346]: option for treatment of DMO if CMT ≥400 μm at the start of treatment
    - Regimen: a single injection every month for five consecutive months, followed by one injection every 2 months with no requirement for monitoring between visits for the first 12 months
    - Clinical trials:
      - **VISTA** study (Brown et al. 2015)
      - **VIVID** study (Brown et al. 2015)

    Ranibizumab (Lucentis, Novartis):
    - NICE Guidance [TA274]: option for treatment of DMO if CMT ≥400 μm at the start of treatment
    - Regimen: given monthly and continued until maximum VA is reached (VA stable for three consecutive months):
    - Clinical trials:
      - **RISE** study (Nguyen et al. 2012; Brown et al. 2013)
      - **RIDE** study (Nguyen et al. 2012; Brown et al. 2013)
      - **RESTORE** study (Mitchell et al. 2011)

    Off license bevacizumab
  - Corticosteroids:

    Dexamethasone implant (Ozurdex, Allergan):
    - NICE Guidance [TA349]: option for treatment of DMO if eye is pseudophakic and CSMO does not respond to non-corticosteroid treatment or such treatment is unsuitable
    - Regimen: A single implant is injected into the vitreous and remains in the vitreous for up to 270 days before fully dissolving
    - Clinical trials:
      - **MEAD** study (Boyer et al. 2014)
      - **BEVORDEX** study (Gillies et al. 2014)
      - **PLACID** study (Callanan et al. 2013)

    Fluocinolone implant (Iluvien, Alimera):
    - NICE Guidance [TA301]: option for the treatment of chronic CSMO that is insufficiently responsive to available therapies if an eye is pseudophakic
    - Regimen: A single 190 μm of Fluocinolone Acetonide implant is injected with daily release of 0.2 μg/day for 36 months

- Clinical trials:
  - **FAME** study (Cunha-Vaz et al. 2014)

- PDR
  - Modification of life-style:
    Smoking cessation
    Weight loss
    Exercise
  - Modification of systemic risk factors (same as for NPDR)
  - Pan-retinal photocoagulation (PRP):
    Indications (The Royal College of Ophthalmologists Diabetic Retinopathy Guideline 2012): presence of NVD/NVE/NVI/NVA — PRP performed on same day or within 2 weeks of diagnosis, consider PRP for severe/very severe NPDR in older patients with type 2 DM, where retinal view is difficult, prior to cataract surgery, in only eye where first eye lost to PDR, where regular clinic attendance is likely to be poor, difficult to examine patient for other reasons
    Prognosis (The Diabetic Retinopathy Study Research Group 1981): PRP reduces severe vision loss (<5/200 at two consecutive visits) from high risk PDR by 50% at 2 years
  - Vitrectomy:
    Indications (The Royal College of Ophthalmologists Diabetic Retinopathy Guideline 2012): non-clearing (within 3 months for type 2 diabetic and 1 month for a type 1 diabetic) severe vitreous haemorrhage (confirmation of attached retina not possible with ophthalmoscopic examination), significant recurrent vitreous haemorrhage despite maximal PRP, tractional RD involving or threatening the fovea, combined tractional rhegmatogenous RD which involves or threatens to involve the fovea, diffuse CSMO associated with posterior hyaloidal traction
    Prognosis (The Diabetic Retinopathy Vitrectomy Study Research Group 1988): early vitrectomy (for those with severe vitreous haemorrhage with VA 5/200 or worse) increased chance of 20/40 vision from 12% to 36% in type 1 diabetics

10.13.7 Diabetic Retinopathy in the Context of Cataract Surgery

- Coexisting PDR should be treated with laser PRP preoperatively where possible if visualisation is allowed otherwise indirect laser treatment may be performed at the conclusion of the cataract surgery

10.13.8 Diabetic Maculopathy in the Context of Cataract Surgery

- Maculopathy should ideally be fully treated (macular laser treatment or anti-VEGF treatment) with resolution of oedema prior to cataract surgery
- For cases in which macular oedema persists despite treatment at the time of cataract surgery, it is reasonable to consider adjunctive treatment with intravitreal triamcinolone injected at the completion of cataract surgery

10.14 Von Hippel Lindau (VHL) Syndrome (Fig. 10.28 and Table 10.13)

10.14.1 History

- Positive FHx (AD)

10.14.2 Examination

- Peripheral capillary haemangioblastoma (red nodular lesion) with the majority located in the ST midperipheral retina ± optic nerve capillary haemangioblastoma
- Normal to dilated and tortuous afferent and efferent vessels (paired retinal arteriole and venule) that may extend all the way posterior to the disc

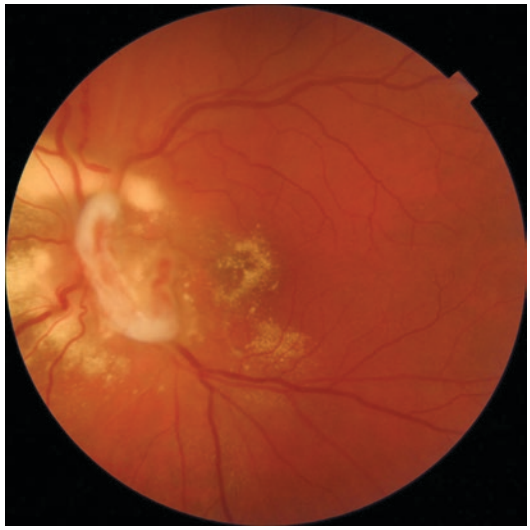

Fig. 10.28 Colour fundus image of a patient with VHL syndrome with an optic nerve capillary haemangioblastoma with subsequent intraretinal and subretinal exudation

Table 10.13 Key facts about VHL syndrome

- Retinal capillary haemangioblastomas are benign hamartomas of the retinal (or optic disc) vasculature, consisting of capillary-like vessels — may be isolated or may be multiple and bilateral as part of a systemic disease (VHL syndrome)
- VHL is an AD, multisystem, familial cancer syndrome caused by germline mutations in the VHL tumour suppressor gene
- VHL is characterised by ocular retinal capillary haemangioblastomas as well as systemic tumours such as cerebellar or spinal cord haemangioblastomas, renal cell carcinoma, phaechromocytomas, and islet cell carcinoma
- Diagnosis of VHL can be made in an individual with multiple tumours or isolated tumours with a positive FHx

- Intraretinal and subretinal exudation: formation of circinate retinopathy and exudative RD — exudates may accumulate preferentially in the macula (centripetal flow of subretinal fluid)

10.14.3 Investigations

- Standard colour fundus photography: assist with follow up to confirm growth, stability or regression following treatment
- FFA: rapid sequential filling of artery, haemangioma, and vein with diffuse leakage of fluorescein in the mid and late frames
- BP: HTN from phaechromocytomas
- 24 hour urine collection: vanillylmandelic acid (VMA) and metanephrines
- Abdominal US scan: renal cell carcinoma, islet cell carcinoma
- MRI brain/spinal cord: cerebellar or spinal cord haemangioblastomas

10.14.4 Treatment

- Refer to geneticist, physician or oncologist for systemic evaluation
- Laser photocoagulation (most suited for small tumours <1 DD): burns of large size (>500 μm), low intensity, and long duration (0.2–0.5 s) are applied directly to surface of the angioma ± feeder vessels
- Trans-scleral cryotherapy (for larger lesions, when media clarity poor, tumours located to anterior equator): 2–3 freeze-thaw cycles
- Radiotherapy

10.14.5 Other Diagnoses to Consider

- Retinal vascular disorders: acquired vasoproliferative retinal tumours, Coats disease, retinal artery macroaneurysm, racemose haemangioma, retinal cavernous haemangioma
- Tumours: retinal astrocytoma, retinoblastoma, malignant melanoma
- Inflammatory disorders: toxoplasmosis, toxocariasis

10.15 Angioid Streaks (Fig. 10.29 and Table 10.14)

10.15.1 Causes of Angioid Streaks

- Pseudoxanthoma elasticum (PXE or Gronbald-Strandberg syndrome — AR connective tissue disorder, optic disc drusen)
- Ehlers-Danlos syndrome
- Paget's disease
- Sickle cell retinopathy/β-thalassaemia
- Idiopathic
- Acromegaly

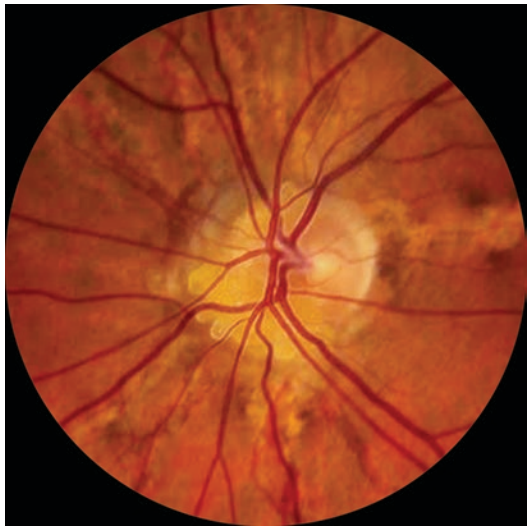

Fig. 10.29 Colour fundus image of a patient with angioid streaks radiating from the optic disc as well as optic disc drusen

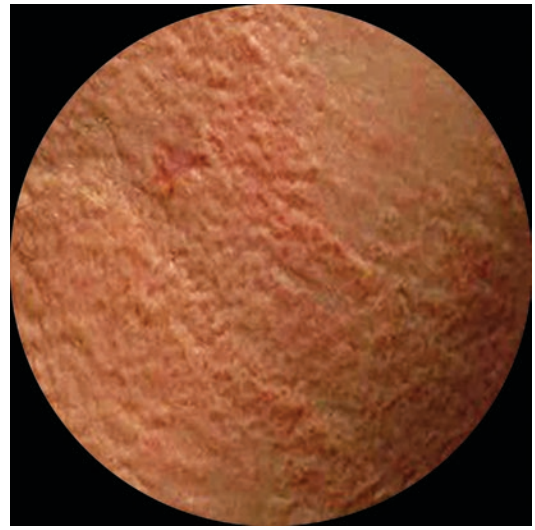

Fig. 10.30 Colour photo of the neck of a patient with PXE showing the typical papular skin lesions

Table 10.14 Key facts about angioid streaks

• Angioid streaks are breaks in an abnormally thickened and calcified Bruch's membrane

10.15.2 Examination

- Irregular reddish brown subretinal lines of varying widths that radiate out from the optic nerve to the retinal periphery
- CNV membrane ± subfoveal haemorrhage ± choroidal rupture (after minor trauma)
- Optic disc drusen
- Perform a systemic examination to look for papular skin lesions and skin laxity of the neck (see Fig. 10.30), axilla, and antecubital fossa

10.15.3 Investigations

- FFA: look for leakage for CNV membrane, hyperfluorescence of angioid streaks in the early phase with late staining

10.15.4 Treatment

- Avoid contact sports
- CNV: subfoveal — anti-VEGF therapy, extrafoveal — laser photocoagulation

10.16 Optic Disc Pits (Fig. 10.31 and Table 10.15)

10.16.1 Examination

- Small hypopigmented yellow or whitish oval or round excavated defect, most often within the inferior temporal portion of the optic cup

10.16.2 Investigations

- OCT: serous macular detachment

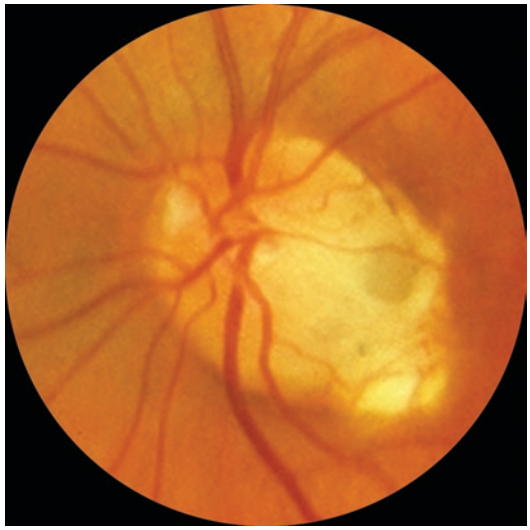

Fig. 10.31 Colour fundus image of a patient with an optic disc pit

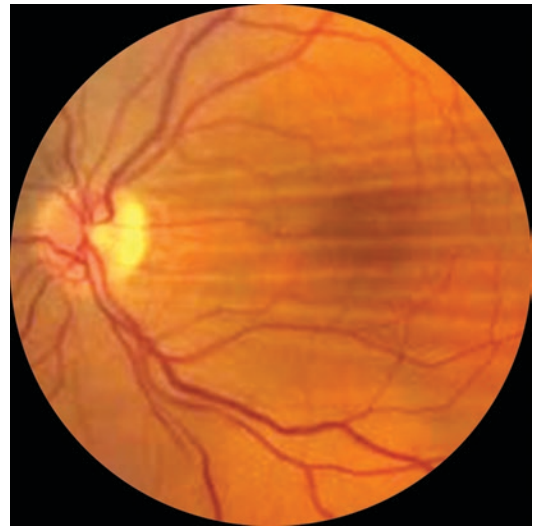

Fig. 10.32 Colour fundus image of a patient with choroidal folds

Table 10.15 Key facts about optic disc pits

• 40–60% of patients with optic disc pits develop non-rhegmatogenous serous macular detachments

10.16.3 Treatment

- Serous macular detachment:
  - Conservative: observation — 25% of optic disc pit maculopathies resolve spontaneously
  - Laser photocoagulation along temporal disc margin
  - Surgical: PPV ± ILM peeling ± Endolaser + long acting gas tamponade

10.17 Choroidal Folds (Fig. 10.32)

10.17.1 Causes of Choroidal Folds

- Ocular
  - Hypotony
  - Posterior scleritis
  - Uveitis
  - Choroidal lesions — tumours, disciform scars
- Orbit
  - TED
  - Retrobulbar mass
  - Idiopathic orbital inflammatory disease
- Intracranial:
  - Raised ICP
- Idiopathic:
  - Hypermetropia — bilateral in asymptomatic individuals

10.17.2 Examination

- Parallel alternating yellow and dark bands on the posterior pole

10.17.3 Investigations

- OCT (see Fig. 10.33)

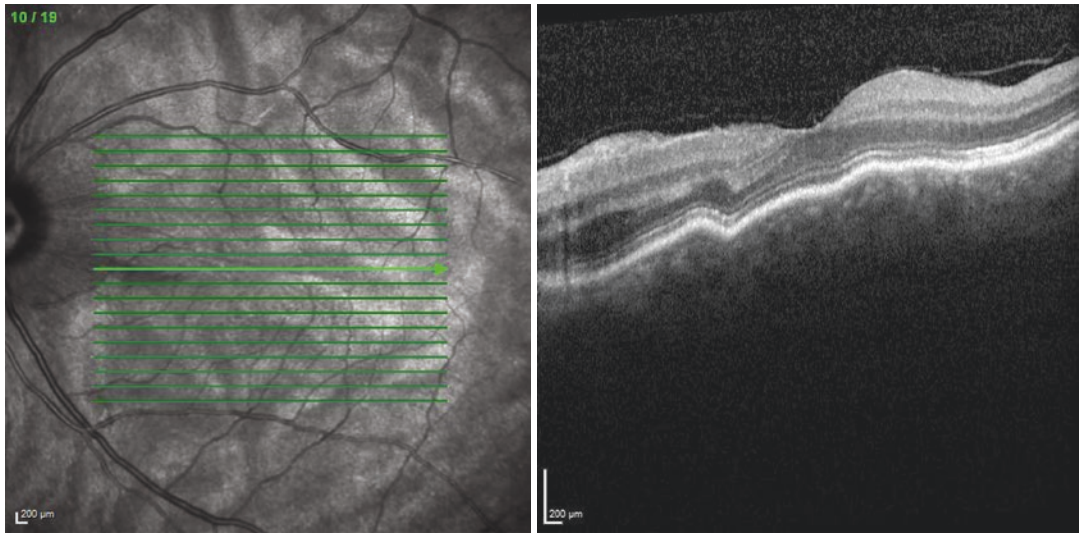

Fig. 10.33 OCT image of a patient with choroidal folds

10.17.4 Treatment

- Treat underlying cause

10.18 Choroideraemia (Fig. 10.34 and Table 10.16)

10.18.1 Examination

- RPE and choroidal atrophy with exposure of bare sclera: may get preservation of the choroid and RPE in the macula
- Relative sparing of retinal vessels and optic disc

10.18.2 Investigations

- VF — constricted
- Genetic testing
- ERG — reduced or absent scotopic (affected first) and photopic responses

10.18.3 Treatment

- There is currently no licensed treatment available for choroideraemia

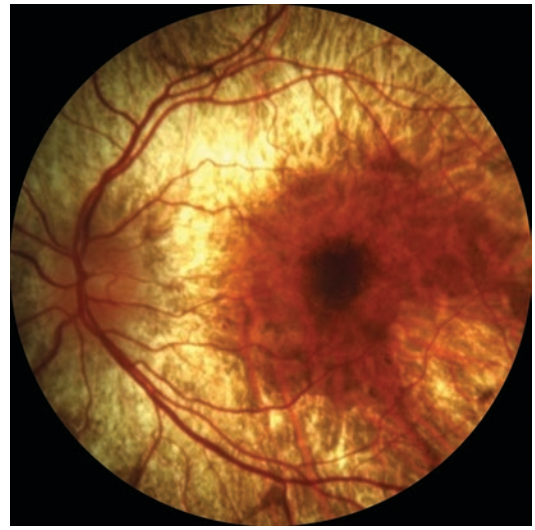

Fig. 10.34 Colour fundus image of a patient with choroideremia showing diffuse chorioretinal atrophy of the peripheral retina with preservation at the central macula

Table 10.16 Key facts about Choroideraemia

• X-linked recessive condition characterised by generalised degeneration of the retina and choroid

10.19 X-Linked Retinoschisis (Fig. 10.35 and Table 10.17)

10.19.1 History

- Positive FHx — X-linked recessive inheritance

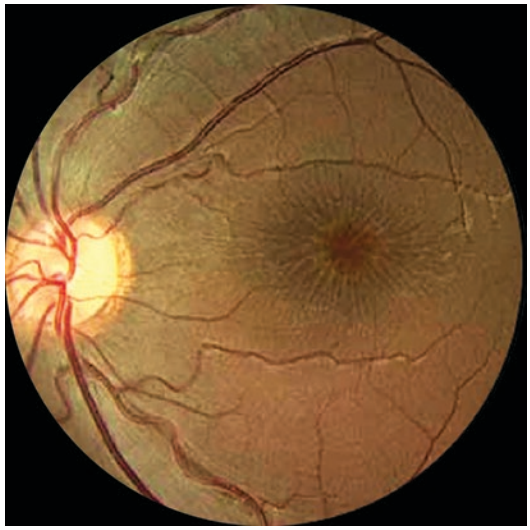

Fig. 10.35 Colour fundus image of a patient with X-linked retinoschisis showing the typical spokewheel pattern of folds radiating out from the fovea

Table 10.17 Key facts about X-linked retinoschisis

• Inherited retinal degenerative disease • Most common form of juvenile onset retinal degeneration in males

10.19.2 Examination

- Foveal schisis: spokewheel pattern of folds radiating out from the fovea
- Peripheral retinoschisis: often in the inferotemporal region

10.19.3 Investigations

- OCT — splitting of the foveal neurosensory retina in the NFL
- EDT — negative ERG — reduced b-wave amplitude with preservation of a-wave amplitude
- FFA — absence of leakage — differentiates foveal schisis from CMO

10.19.4 Treatment

- Genetic counselling
  - Carrier women have 50% chance of transmitting the mutation in each pregnancy
  - Males with the mutation will be affected and females with the mutation will be carriers
  - Affected males pass the disease-causing mutation to all of their daughters and none of their sons
- Medical — carbonic anhydrase inhibitors
- Surgical — if tractional or rhegmatogenous RD or vitreous haemorrhage present

10.19.5 Other Diagnosis to Consider

- CMO — late hyperfluorescence in a petaloid pattern
- Degenerative retinoschisis — bilateral, hypermetropes, smooth convex elevation, inferotemporal region, laser take up, absolute scotoma in schitic areas, split retina within OPL
- Goldmann Favre vitreoretinal degeneration — AD inheritance, foveal schisis, marked VF loss, pigmentary clumping, absence of vitreous veils, markedly reduced a-waves and b-waves

10.20 Macular Telangiectasia (MacTel) Type 2 (Fig. 10.36 and Table 10.18)

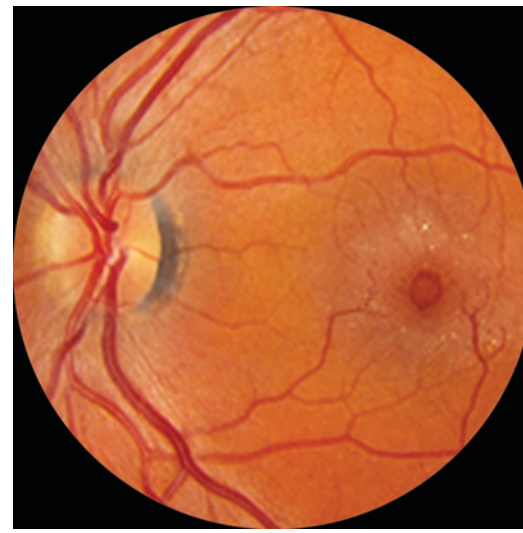

Fig. 10.36 Colour fundus image of a patient with MacTel type 2 showing dilation of capillaries in the temporal parafoveal region

Table 10.18 Key facts about MacTel type 2

- Disease characterised by telangiectatic vessels in the Juxtafoveal region, which tends to be bilateral but asymmetrical
- Middle aged and older persons
- All lesions begin temporal to the fovea but may subsequently involve the entire parafoveolar area
- Other diagnoses to consider:
  - MacTel type 1 — developmental or congenital, usually unilateral vascular anomaly (may be part of the larger spectrum of Coats disease)
  - Diabetic retinopathy
  - Vascular occlusion
  - Eales disease
  - Radiation retinopathy
  - ROP
  - Sickle cell retinopathy

10.20.1 Examination

- Subtle loss of retinal transparency in the perifoveal region, beginning temporally
- Dilation of the parafoveal capillaries in the temporal parafoveal area, which may extend to surround the fovea
- Crystalline deposits at the vitreoretinal interface
- Right angled venules ± RPE hyperplasia and intraretinal pigment migration
- CNV or associated sequelae — subretinal or intraretinal haemorrhage, intraretinal oedema, retinal hard exudates, fibrovascular disciform scar

10.20.2 Investigations

- OCT (see Fig. 10.37) — subfoveal cystoid cavities (retinal cavity formations) ± photoreceptor disruption (IS/OS junction) ± outer retinal atrophy
- FAF — loss of normal hypofluorescence at the fovea with abnormally increased autofluorescence at the fovea
- FFA — telangiectatic vessels starting predominantly temporal to the fovea with eventual involvement of entire parafoveal area with early hyperfluorescence and late leakage

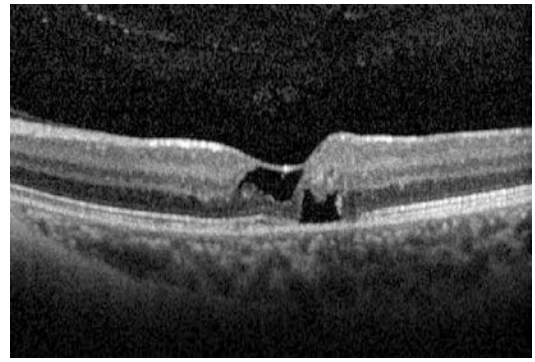

Fig. 10.37 OCT image of a patient with MacTel type 2 showing subfoveal cystoid cavities

10.20.3 Treatment

- CNV — anti-VEGF therapy

10.20.4 Other Diagnoses to Consider

- BRVO — segmental capillary changes, involves an area distal to an arteriolar-venular crossing, and does not cross the horizontal raphe unless there is already collateral formation
- Radiation retinopathy — involves larger area of retina, cotton wool spots, preretinal NV, hx of radiation to the eye, orbit or head
- Neovascular AMD — drusen, RPE changes, uncommon to have retinal capillary disease

10.21 Birdshot Chorioretinopathy (Fig. 10.38 and Table 10.19)

10.21.1 Examination

- Multiple small (1/4–1/2 DD) oval or round cream coloured lesions scattered around the optic disc that radiate out to the equator — lesions become atrophic but not pigmented
- Optic disc oedema leading to atrophy
- Vitritis (≤2+ vitreous haze)
- Retinal vasculitis (phlebitis)

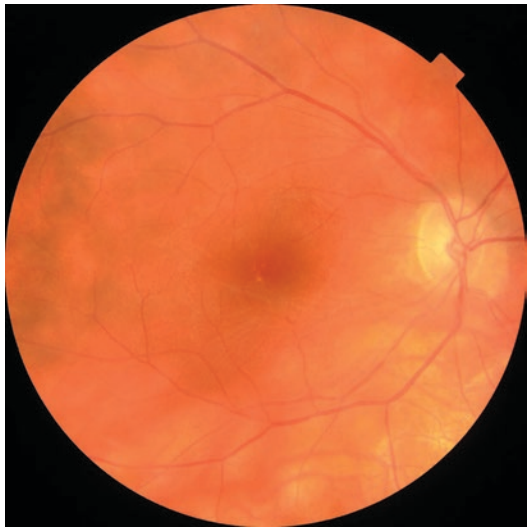

Fig. 10.38 Colour fundus image of a patient with birdshot chorioretinopathy with multiple scattered cream coloured oval lesions and an ERM

Table 10.19 Key facts about birdshot chorioretinopathy

• Bilateral condition • Middle aged Caucasian adults with a slight female preponderance • Chronic inflammation with natural course of 6–10 years

- CMO
- ERM
- CNV (5% of eyes)

10.21.2 Investigations

- OCT — CMO, ERM
- FFA — look for CNV and retinal vasculitis
- ICG — hypofluorescent (choroidal ischaemia or blockage from inflammatory infiltrates) birdshot lesions during active disease with diffuse late hyperfluorescence
- HLA-testing — positive HLA-A29
- VF — peripheral constriction, enlarged blind spot, central or paracentral scotomas
- ERG — negative ERG (reduced B wave amplitude and latency), delay in implicit time of 30 Hz flicker (to assess progression)

10.21.3 Treatment

- CMO — corticosteroids + long term immunosuppresants
- CNV — anti-VEGF therapy

10.21.4 Other Diagnoses to Consider

- Sarcoidosis
- Syphilitic chorioretinitis
- Multifocal choroiditis and panuveitis
- Sympathetic ophthalmia/VKH
- Intraocular B-cell lymphoma

10.22 Presumed Ocular Histoplasmosis Syndrome (POHS) (Fig. 10.39 and Table 10.20)

10.22.1 Examination

- Discrete focal atrophic (i.e. punched out) choroidal scars in the macula or periphery

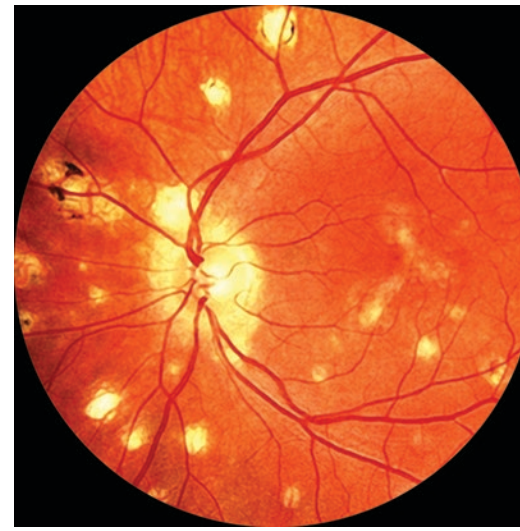

Fig. 10.39 Colour fundus image of a patient with POHS with multiple punched out atrophic chorioretinal spots and peripapillary atrophy

Table 10.20 Key facts about POHS

- An inflammatory disorder that has been postulated to result from systemic infection with the dimorphic fungi *Histoplasma capsulatum*
- Only presumed, not proven
- Histoplasma occurs mainly in Mississippi Valley; not identified in UK
- A clinical diagnosis is based on the presence of at least two of the following fundus lesions in one or both eyes in the absence of ocular inflammation

- Peripapillary chorioretinal scarring (i.e. peripapillary atrophy)
- CNV or associated sequelae — haemorrhagic RD, fibrovascular disciform scar
- Absence of anterior and posterior inflammation

10.22.2 Investigations

- FFA — look for leakage to confirm presence of a CNV

10.22.3 Treatment

- Steroid therapy if inflammation active, and vision threatening
- CNV — anti-VEGF therapy

10.22.4 Other Diagnoses to Consider

- Multifocal choroiditis with panuveitis: multiple chorioretinal scars, significant anterior and posterior inflammation present in the active phase ± CNV, bridging scars, subretinal fibrosis, clustering of lesions (e.g. macula, equator), narrow or sheathed vessels, progressive growth of lesions
- Sarcoidosis — usually accompanied by anterior and posterior inflammation, no peripapillary atrophy or CNV, elevated ACE, hilar adenopathy on CXR or CT chest
- Punctate inner choroidopathy (PIC) — predominantly seen in women, no anterior/posterior inflammation, no peripapillary atrophy, atrophic scars, CNV
- Myopic degeneration — peripapillary atrophy, CNV, small white focal areas of chorioretinal atrophy along with linear atrophic areas (e.g. lacquer cracks) in the posterior pole

10.23 Toxoplasmosis (Fig. 10.40 and Table 10.21)

10.23.1 Examination

- Acute lesions — intensely white focal lesions with overlying vitreous inflammatory haze adjacent ("headlight in the fog") to old hyperpigmented scars (satellite lesions), periphlebitis
- Chronic lesions — hyperpigmented scars with an atrophic center devoid of all retinal and

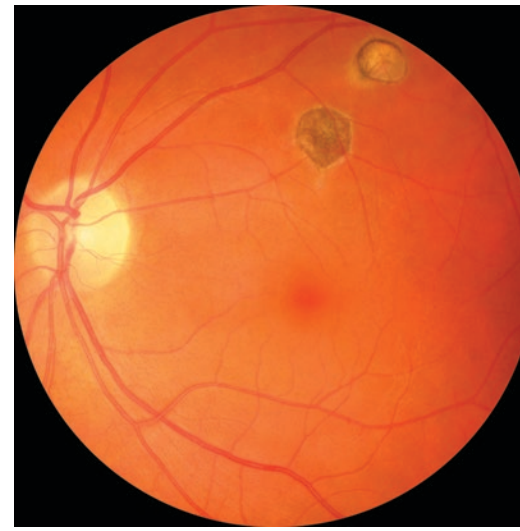

Fig. 10.40 Colour fundus image of a patient with congenital toxoplasmosis with multiple hyperpigmented scars

Table 10.21 Key facts about toxoplasmosis

- Caused by T. gondii
- Acquired from ingestion of oocyte from contaminated soil or the cyst form in undercooked infected meat — high risk of contamination from contact with cat faeces, litter boxes, or potentially contaminated outdoor sand and soil

choroidal elements — the underlying sclera gives the lesion its white center
- Check the IOP
- Perform an anterior segment examination to look for any visually significant cataract

10.23.2 Investigations

- VDRL — rule out syphilis
- ACE, CXR — rule out sarcoidosis
- HIV serology — rule out HIV

10.23.3 Treatment

- Inflammation
  - In immunocompetent patients, the disease is self-limiting and does not require treatment unless sight threatening
  - Indications for treatment:
    Lesions involving the disc, macula, or papillomacular bundle
    Lesions threatening a major vessel
    Marked vitritis
    Any lesion in an immunocompromised patients
  - No corticosteroids if patient is immunocompromised
  - Triple therapy: pyrimethamine, sulfadiazine, folinic acid, corticosteroids
  - Co-trimoxazole (trimethoprim + sulfamethoxazole) + corticosteroids
  - Intravitreal dexamethasone + clindamycin
  - Oral clindamycin
  - Spiramycin or atovaquone if maternal infection acquired during pregnancy (15% in the first trimester and 60% in the third trimester risk of fetal transmission if acquired during pregnancy)
- Cataract — extraction once uveitis is inactive for ≥3 months

10.23.4 Other Diagnoses to Consider

- Infectious — TB, syphilis, rubella, CMV retinitis, herpes simplex, toxocariasis
- Non-infectious — sarcoidosis

10.24 Multifocal Choroiditis and Panuveitis (MFC) (Fig. 10.41 and Table 10.22)

10.24.1 Examination

- Acute phase — yellow round or oval lesions in the posterior pole, peripapillary region and midperiphery ± subretinal fluid, vitritis ± anterior uveitis, periphlebitis
- Chronic phase — punched out atrophic scars with variable hyperpigmentation, peripapillary subretinal fibrosis, bridging subretinal fibrosis between atrophic scars, peripheral linear streak chorioretinal scars parallel to the ora, optic disc atrophy, CNV membrane (25–30%), CMO

10.24.2 Investigations

- OCT — CMO
- FFA — early hypofluorescence and late staining of lesions, leakage for CNV membrane
- ICG — hypofluorescent round spots with staining of edges

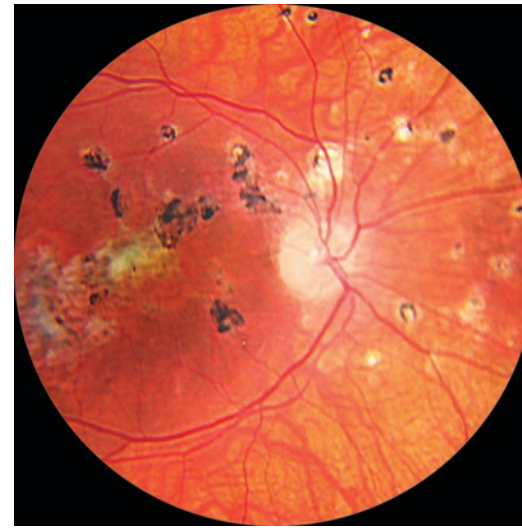

Fig. 10.41 Colour fundus image of a patient with MFC showing multiple hyperpigmented punched out atrophic scars in the posterior pole and peripapillary region

Table 10.22 Key facts about MFC

• Typically affects myopic caucasian women between the second and sixth decades of life (most affected patients are in their 30s)

10.24.3 Treatment

- CMO — topical, periocular, intraocular, and systemic corticosteroids
- CNV — anti-VEGF therapy

10.24.4 Other Diagnoses to Consider

- Inflammatory — POHS (no vitritis), PIC (posterior pole, no vitritis), Sarcoidosis, VKH syndrome/Sympathetic ophthalmia, Serpiginous choroidopathy, Birdshot chorioretinopathy
- Infectious — TB, syphilis
- Degenerative — Myopic degeneration maculopathy
- Neoplastic masquerading diseases — intraocular lymphoma

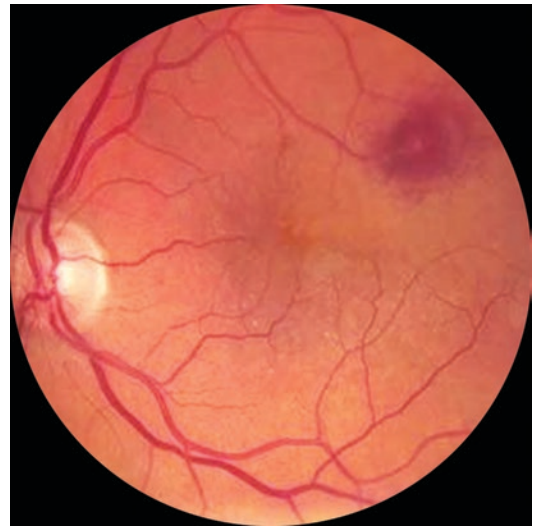

Fig. 10.42 Colour fundus image of a patient with a retinal arterial macroaneurysm

10.25 Retinal Arterial Macroaneurysm (Fig. 10.42)

10.25.1 Risk Factors

- HTN
- Diabetes

10.25.1.1 Examination

- Fusiform or round dilatations of the retinal arterioles that occur in the posterior fundus within the first three orders of arteriolar bifurcation
- Hourglass retinal haemorrhage (subretinal, intraretinal, sub-ILM, vitreous) ± retinal oedema ± retinal exudation

10.25.2 Investigations

- FFA — hyperfluorescence of macroaneurysm in the early arterial phase with leakage in the late stage

10.25.3 Treatment

- Conservative: most spontaneously thrombose and undergo involution with clearing of exudates
- Laser photocoagulation (if lipid exudation involves or threatens the fovea): treat around the macroaneurysm with a single confluent laser barrier — if leakage persists, repeat procedure with direct laser of the lesion
- Vitrectomy: non-clearing vitreous haemorrhage

10.25.4 Other Diagnoses to Consider

- Diabetic retinopathy
- Retinal telangiectasia
- Retinal capillary angioma
- Cavernous haemangioma
- Haemorrhagic PED of AMD

10.26 Choroidal Naevus (Fig. 10.43)

10.26.1 Examination

- Flat or minimally elevated slate-grey lesion with irregularly defined margins ± surface drusen

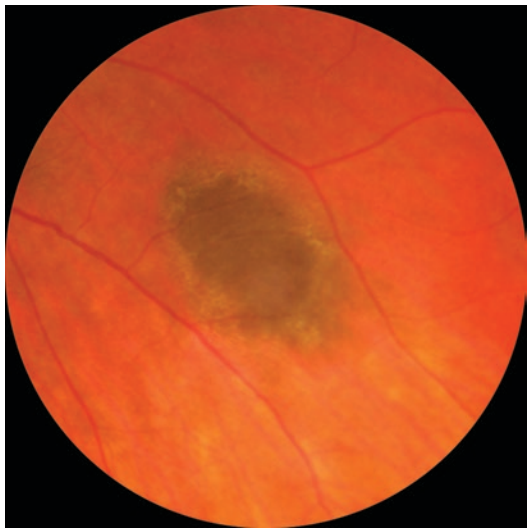

Fig. 10.43 Colour fundus image of a patient with a flat choroidal naevus with surface drusen

Table 10.23 When to refer a patient with a choroidal lesion (The RCOphth Clinical Guideline on Referral pathways for adult ocular tumours 2019)

Refer patients with a suspicious choroidal melanocytic tumour having:
- Any one of the following:
  (a) Thickness more than 2 mm
  (b) Collar stud configuration
  (c) Documented growth

OR
- Any two of the following:
  (a) Thickness more than 1.5 mm
  (b) Orange pigment
  (c) Symptoms
  (d) Serous RD

10.26.2 Investigations

- Colour fundus photography — documentation
- B-scan US — determine tumour thickness

10.26.3 Treatment

- If no suspicious features are present, these lesions do not require regular ophthalmic review. Naevus photographed and a copy is given to the patient to permit their own optometrist to monitor the lesion (e.g. annually) as part of their routine optometric review
- Refer patients if suspicious features are present in accordance with the RCOphth guidelines (see Table 10.23)

10.26.4 Other Diagnoses to Consider for a Pigmented Choroidal Lesion

- Choroidal melanoma (see Table 10.24) — more elevation, medium-low internal reflectivity on A-scan US, irregular elevated acoustically hollow choroidal mass on B-scan US

Table 10.24 Features suggestive of a choroidal melanoma (Shield et al. 2009)

"To Find Small Ocular Melanomas Using Helpful Hints Daily"
- **T**hickness: more than 2 mm
- **F**luid: presence of SRF (serous RD)
- **S**ymptoms: photopsias
- **O**range pigment: lipofuscin
- **M**argin: tumour margin within 3 mm of the optic disc
- **U**ltrasonography **H**ollowness
- **H**alo absence (in halo choroidal naevus the lesion is surrounded by a depigmented yellow ring)
- **D**rusen absence

Tumours that display a single factor have a 38% chance for transformation

Tumours with two or more factors show transformation in over 50% of cases at 5 years

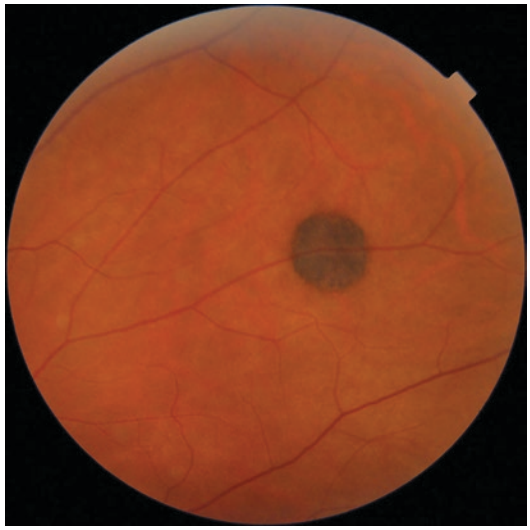

Fig. 10.44 Colour fundus image of a patient with a solitary CHRPE with intralesional lacunae (or loss of RPE hyperpigmentation)

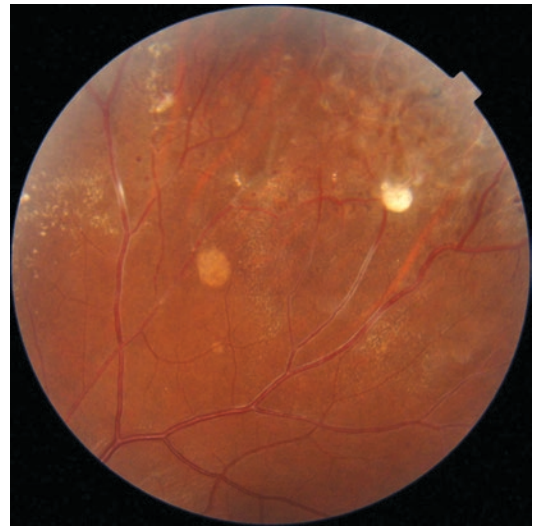

Fig. 10.45 Colour fundus image of a patient with Coats disease showing intraretinal and subretinal exudates, venous sheathing and arteriolar aneurysmal dilations

- Congenital hypertrophy of the RPE (CHRPE — see Fig. 10.44) — typically unilateral, solitary or grouped ("bear-track"), darkly pigmented, round, well circumscribed, flat lesions at the level of the RPE, intralesional lacunae, CHRPE-like lesions associated with FAP (Gardner syndrome — AD) are similar in appearance but are bilateral multiple and pisciform in shape
- Choroidal metastasis — indistinct or irregular margins, creamy-yellow colour, often associated with a serous RD out of proportion to size of tumour, B-scan US for an area of mild choroidal elevation with f A-scan, positive hx of prior malignancy, if no prior history of malignancy — CT chest (lung Ca) and breast imaging (breast Ca)

10.27 Coats Disease (Fig. 10.45 and Table 10.25)

Table 10.25 Key facts about Coat's disease

- Idiopathic condition characterised by telangiectatic and aneurysmal retinal vessels with intraretinal and subretinal exudation and fluid
- Disease affects males 3× as often as females
- Unilateral in 80–95% of cases

10.27.1 Examination

- Localised, yellow, intraretinal and subretinal exudation ± exudative RD associated with adjacent vascular anomalies, including venous sheathing and beading, telangiectasia (irregular dilation of the capillary bed), tortuosity, arteriolar aneurysmal dilatation ("light bulb" aneurysms) ± NVD ± NVE
- Check IOP (neovascular glaucoma)
- Perform an anterior segment examination to look for NVI and cataract

10.27.2 Investigations

- FFA — capillary non-perfusion between dilated telangiectatic vessels, leakage of dye from telangiectatic vessels and disc neovascularisation

10.27.3 Treatment

- Retinal telangiectasia present only: follow conservatively

- Subretinal exudation or fluid present: focal laser photocoagulation (first line treatment), or cryotherapy (if extensive subretinal exudation or RD) to areas of leaking telangiectatic vasculature and capillary non-perfusion ± adjuvant anti-VEGF injection (helps reduce subretinal fluid and macular exudation)
- Surgery for eyes with advanced exudative RD abutting the crystalline lens or those with a rhegmatogenous component to the detachment

10.27.4 Other Diagnoses to Consider

- BRVO
- Peripheral vasculitis (pars planitis)
- Eales disease
- Ocular toxocariasis
- FEVR — retinal dragging, positive FHx, bilateral
- Persistent fetal vasculature

10.28 Choroidal Osteoma (Fig. 10.46 and Table 10.26)

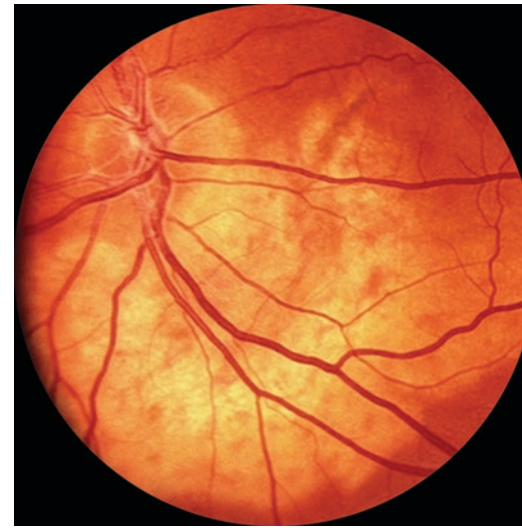

Fig. 10.46 Colour fundus image of a patient with a choroidal osteoma

Table 10.26 Key facts about a choroidal osteoma

• Benign tumour of the choroid composed of mature bone (mature bone replaces choroid with damage to overlying RPE and retina) • Typically found in healthy young females in the second or third decade of life • Tumour usually occurs as a sporadic trait and is usually unilateral

10.28.1 Examination

- Yellow-white well defined elevated or excavated oval or round geographic lesion located in the juxtapapillary or peripapillary area ± extension into the macula (rarely tumour is confined to the macular region only without involvement of the juxtapapillary area)
- Subretinal haemorrhages — suggestive of CNV
- Overlying retinal vasculature and optic disc is characteristically unaffected by the osseous tumour and there are no associated vitreous or anterior segment abnormalities

10.28.2 Investigations

- OCT — serous SRF ± retinal thinning over macular portion of the osteoma
- FFA (if SRF or haemorrhage present) — look for leakage suggestive of CNV membrane, osteoma produces a mild patchy hyperfluorescence of the tumour that evolves to a diffuse intense late staining
- A-scan US — high intensity echo spike from the inner surface of the tumour
- B-scan US — slightly elevated highly reflective choroidal mass with acoustic shadowing just posterior to the choroidal mass (gives appearance of a pseudo-optic nerve)

10.28.3 Treatment

- Treatment of CNV — extrafoveal (laser photocoagulation or PDT), subfoveal (anti-VEGF therapy)

- Induction of decalcification of tumour — if osteoma in extrafoveal region, argon laser photocoagulation or PDT to prevent growth of osteoma towards foveola (decalcified osteoma does not grow in the direction of the decalcification)

10.28.4 Other Diagnoses to Consider

- Amelanotic choroidal melanoma: less well-defined margin, more elevation, medium-low internal reflectivity on A-scan US, irregular elevated acoustically hollow choroidal mass on B-scan US
- Metastatic carcinoma to the choroid — indistinct or irregular margins, creamy-yellow colour, often associated with a serous RD out of proportion to size of tumour, B-scan US for an area of mild choroidal elevation with medium-high internal reflectivity on A-scan, positive hx of prior malignancy, if no prior hx of malignancy — CT chest (lung Ca) and breast imaging (breast Ca)
- Disciform macular degeneration — older patients with AMD, usually centered in the macula with AMD
- Circumscribed choroidal haemangioma — characteristically dome shaped with smooth regular margins with overlying serous SRF with cystoid degeneration of retina, orange-red in colour, high internal reflectivity on A-scan, acoustically full dome-shaped elevated mass
- Posterior scleritis — indistinct margins, US evidence of thickened sclera and choroid with retrobulbar oedema (T-sign)

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