A PICTORIAL GUIDE TO COMMON AND INTERESTING CONDITIONS OF THE OCULAR FUNDUS

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INTRODUCTION

Traditionally, the family physician had to rely mainly on the direct ophthalmoscope to examine the ocular fundus. Although useful, this examination method has proved difficult for some, especially if the patient's pupils are not pharmacologically dilated. More recently, the availability of the fundus cameras in many large primary care facilities such as the polyclinics, as well as in screening services in hospitals, has enabled the family physician to examine large areas of the retina and the optic disc with greater ease. This, in turn, has allowed many of them to diagnose common retinal and optic nerve disorders without difficulty.

Many eye conditions have characteristic findings in the ocular fundus. This pictorial guide aims to familiarise the family physician with some common as well as interesting conditions of the ocular fundus. The interpretation of clinical signs and their implications are emphasised. Rarer or more esoteric material has been included for clinical interest. Unlike most commercial colour atlases, the fundus photographs chosen for this pictorial guide are not all high quality textbook illustrations. Instead, they are selected to show the type of materials that are encountered in day-to-day clinical practice.

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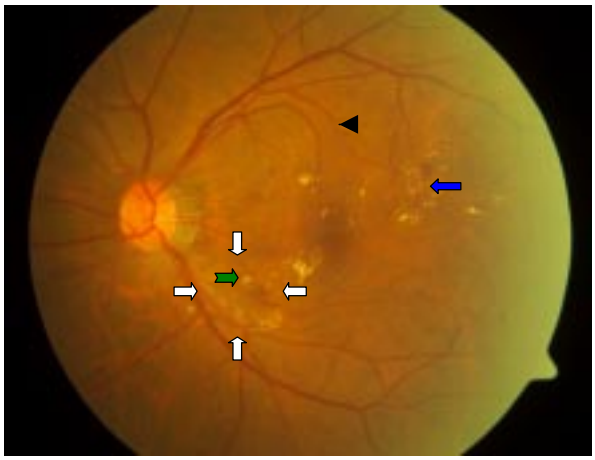

Figure 1: Diabetic maculopathy.

DIABETIC RETINOPATHY

Diabetic retinopathy is essentially a microangiopathy affecting the retina. The retinal precapillary arterioles, capillaries and venules are the usual vessels affected, although larger vessels may also become involved. The changes in the ocular fundus are caused by microvascular leakage and microvascular occlusion.

Figure 1 demonstrates both signs of microvascular leakage (haemorrhages and hard exudates) and microvascular occlusion (cotton wool spot) in the retina. Because the changes are occurring in the macula, it is termed diabetic maculopathy.

Increased vascular permeability leads to retinal haemorrhage (blue arrow) and retinal oedema. Chronic localised retina oedema due to leakage from microaneurysms leads to the deposition of hard exudates, usually at the junction of oedematous and healthy retina. These exudates are composed of lipoprotein and lipid-laden macrophages. They vary in size, have a yellow waxy appearance with relatively distinct margins, and are frequently distributed in a circinate pattern peripheral to areas of chronic focal leakage

(white arrows). Microaneurysms are usually present in the centres of rings of hard exudates (green arrow).

A cotton-wool spot (black arrowhead) is an infarct of the retinal nerve fibre layer due to capillary occlusion. It was previously also known as "soft exudate", but this is a misnomer since it is not a result of microvascular leakage. Ischaemia in the infarcted area causes an interruption of the axoplasmic flow of the retinal nerve fibres and subsequent build-up of material within the nerve axons, giving rise to the opaque white appearance.

The eye in Figure 1 has severe diabetic maculopathy and is at risk of losing vision. The patient requires a referral to an ophthalmologist for further evaluation and treatment. Diabetic maculopathy is treated with focal laser photocoagulation.

Figure 2 shows an ocular fundus that has been treated previously with focal laser photocoagulation for diabetic maculopathy. There are multiple areas of well-circumscribed hypo- and hyperpigmented scars in the macula secondary to the laser photocoagulation. Hard exudates which were previously present have resolved, following laser ablation of the leaking microanerysms. A cotton-wool spot is present superonasal to the fovea (blue arrow).

Figure 3 shows new blood vessels (neovascularisation) at the optic disc (white arrow) and fibrovascular membranes causing traction on the retina along the superotemporal vascular arcade. Note the distortion of the vasculature due to the traction (green arrows). This patient has previous laser photocoagulation treatment as indicated by the laser scars in the peripheral fundus. He requires additional laser treatment (panretinal photocoagulation). There is also a ring of hard exudates in the macula.

Figure 4 shows tractional retinal detachment as a result of progressive contraction of fibrovascular membranes (white arrows) over large areas of vitreoretinal adhesions along the vascular arcades. This is a devastating complication of proliferative diabetic retinopathy. Vitreous surgery (vitrectomy, peeling of fibrovascular membranes and endolaser photocoagulation) is indicated. However, even with aggressive surgical intervention, the visual prognosis is guarded at this late stage of the condition.

RETINAL VASCULAR DISEASES

Acute central retinal artery occlusion (CRAO) is an ophthalmic emergency. The sudden occlusion of the retinal arterial circulation may be due to arteriosclerotic thrombosis, embolism or vasculitis.

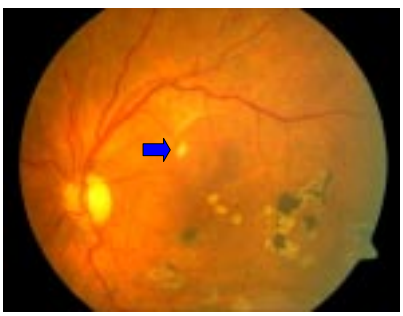

Figure 2: Diabetic maculopathy treated with focal laser photocoagulation.

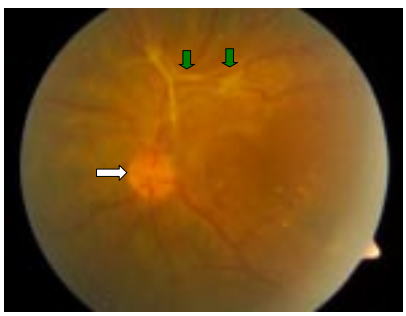

Figure 3: Proliferative diabetic retinopathy.

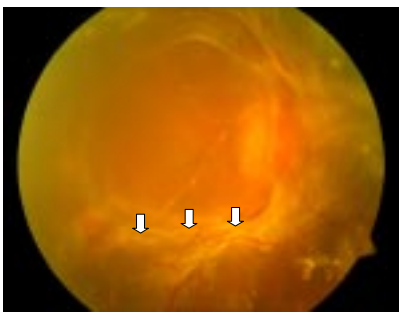

Figure 4: Advanced diabetic retinopathy with tractional retinal detachment.

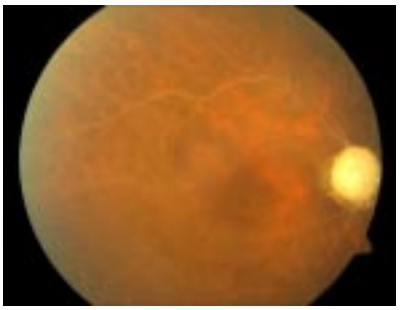

Figure 5: Old central retinal artery occlusion (CRAO).

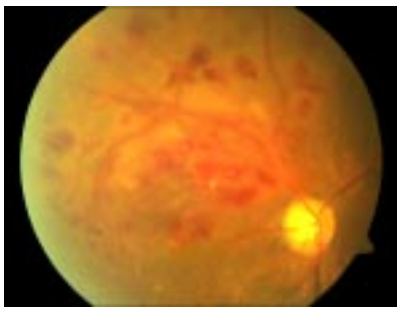

Figure 6: Superotemporal branch retinal vein occlusion (BRVO).

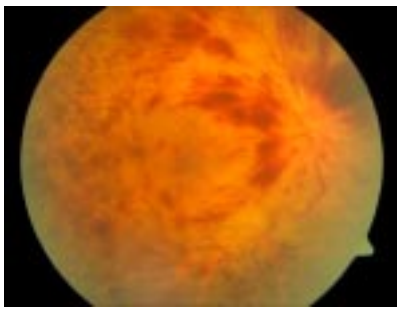

Figure 7: Central retinal vein occlusion (CRVO).

In the acute stage, the retina appears whitish due to oedema of the nerve fibre and ganglion cell layers. As these are absent in the fovea, the still perfused choroidal vasculature (the eye has a dual circulation) produces a characteristic "cherry red spot" against the opaque retina surrounding the fovea[1].

Figure 5 shows a pale optic disc (optic atrophy) associated with attenuated and sclerosed retinal vessels due to a previous CRAO.

In general, CRAO is associated with a poor visual prognosis, despite aggressive acute management (ocular massage, anterior chamber paracentesis and intravenous acetazolamide) that aims to reduce the intraocular pressure and increase retinal perfusion. A recent advancement in the management of acute occlusion of the central retinal artery is local intra-arterial fibrinolysis.[2] However, its efficacy has not been proven by any large randomised controlled trial.

Occlusion of the retinal venous circulation may involve part of the circulation i.e. branch retinal vein occlusion or the entire circulation i.e. central retinal vein occlusion.

Systemic risk factors for venous occlusion include advanced age, systemic hypertension, hyperlipidaemia, diabetes mellitus and hypercoagulable states. Certain drugs (e.g. oral contraceptives) may also predispose one to venous occlusion. Ocular risk factors include glaucoma, optic disc drusen and periphlebitis.

Potential complications include neovascular glaucoma, vitreous haemorrhage secondary to neovascularisation, chronic macular oedema and ischaemic maculopathy.

Figure 6 shows extensive retinal haemorrhages and tortuous retinal veins involving the superotemporal vascular arcade. The macula is also involved. This patient will require a complete ophthalmic review and a systemic work-up to exclude any predisposing ocular or systemic conditions.

Figure 7 shows diffuse massive flame and blot haemorrhages involving all 4 quadrants of the ocular fundus. The retinal vessels are tortuous and the optic disc is swollen. The clinical picture is that of a central retinal vein occlusion.

Central retinal vein occlusion may be classified as either ischaemic or non-ischaemic. The former has a poorer prognosis and are more likely to develop complications such as neovascular glaucoma. A fundal fluorescein angiography may be useful to assess areas of capillary non-perfusion and neovascularisation. Panretinal laser photocoagulation will have to be performed if there are extensive areas of capillary non-perfusion or if neovascularisation is present[3].

AGE-RELATED MACULAR DEGENERATION (AMD)

Age-related macular degeneration (AMD) is a common cause of severe irreversible visual loss among the elderly population. With the ageing population in Singapore, AMD is expected to become an even more important cause of visual impairment in the near future.

Although the exact aetiology of AMD is unknown, a number of risk factors have been identified. They are advanced age, female, smoking, family history, white race and co-existing cardiovascular risk factors such as hypertension and hyperlipidaemia.

There are 2 main forms of AMD. One is known as non-exudative (dry, atrophic or non-neovascular) and the other is called exudative (wet or neovascular) AMD. Both types tend to be progressive and bilateral[4,5].

Figure 8 shows an eye with early non-exudative AMD with multiple small, discrete, yellow-white, slightly elevated lesions called drusen (blue arrow). There are also some pigmentary changes in the fovea. At this early stage, the visual may be normal or slightly impaired.

Figure 9 shows an eye with advanced non-exudative AMD with confluent areas of atrophy of the retinal pigment epithelium in the macula (geographical atrophy). Because the atrophic area involves the fovea, the associated visual loss is severe. Numerous drusen surrounding the geographic atrophy are also present.

There is currently no treatment available for non-exudative AMD.

Figure 10 shows an eye with exudative AMD. There is a large subretinal haemorrhage (green arrow) surrounding a choroidal neovascular membrane. The haemorrhage is subretinal in location because the normal retinal vessels can be

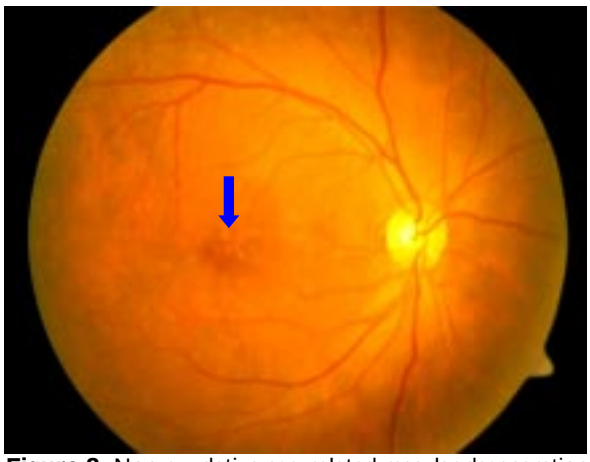

Figure 8: Non-exudative age-related macular degeneration.

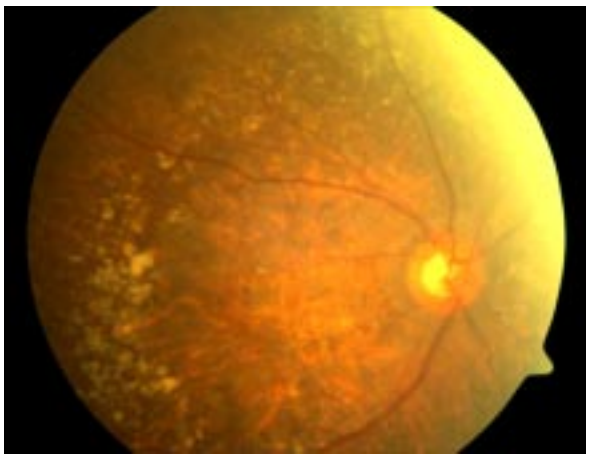

Figure 9: Non–exudative AMD.

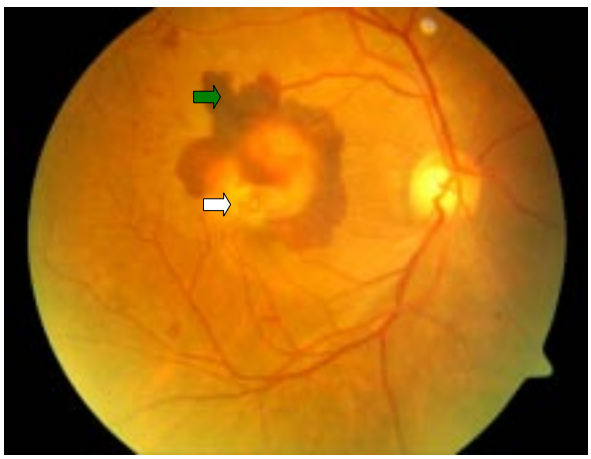

Figure 10: Exudative age-related macular degeneration.

seen to course over it undisturbed. Contraction of an associated epiretinal membrane has produced retinal striae radiating from the epicentre of contraction (white arrow). The choroidal neovascularisation can be readily demonstrated using fundal fluorescein angiography.

Figure 11 shows another eye with exudative AMD with a haemorrhagic pigment epithelial detachment (haemorrhage beneath the retinal pigment epithelium) (green arrow) and choroidal neovascularisation. Numerous drusen (blue arrow) are also present.

Although there are currently several treatment options available for exudative AMD, each has its own limitations[5]. Laser photocoagulation to obliterate choroidal neovascular membranes situated outside the fovea has been shown to be effective. However, many patients are unsuitable for this treatment modality because the choroidal neovascular membranes are beneath the foveal centre. Laser ablation of choroidal neovascular membranes also destroy the overlying retina. As a result, laser photocoagulation of subfoveal choroidal neovascular membranes is associated with an immediate decrease in vision and is rarely performed. Photodynamic therapy is a new treatment modality for certain exudative AMD. It involves infusion of a photosensitising dye followed by application of a non-thermal laser to the target tissues. Activation of the photosensitive dye in the choroidal neovascular membranes closes the choroidal neovascular membranes. However, recurrence is common and multiple re-treatments are often necessary.

Other potential treatments for exudative AMD include submacular surgery and macular translocation.

Exudation and subretinal haemorrhage from wet AMD leads to subretinal scarring and destruction of photoreceptors. Figure 12 shows a large disc-like whitish scar with areas of pigmentation in the macula secondary to exudative AMD.

OTHER RETINAL ABNORMALITIES

Severe myopia may be associated with myopic chorioretinal degeneration (Figure 13). A ring of chorioretinal degeneration (peripapillary chorioretinal degeneration) surrounds the optic disc and appears whitish because the underlying sclera is more visible. Elsewhere, the large choroidal vessels are unusually prominent because of atrophy the overlying retinal pigment epithelium and choriocapillaris secondary to the generalised chorioretinal degeneration.

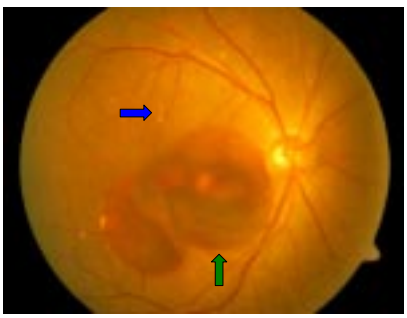

Figure 11: Exudative age-related macular degeneration.

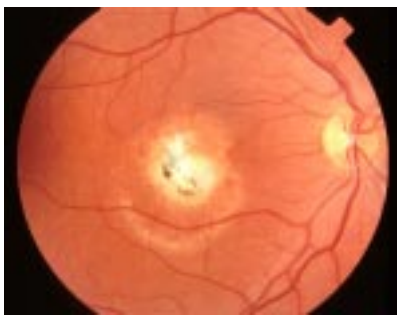

Figure 12: Disciform scar secondary to exudative age-related macular degeneration.

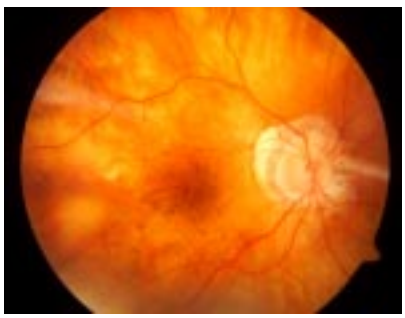

Figure 13: Myopic degeneration.

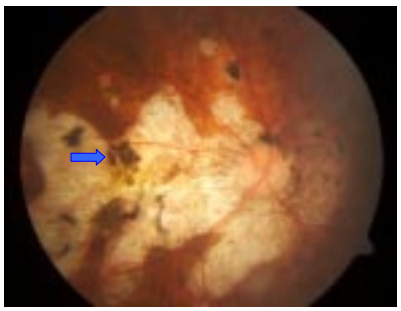

Figure 14: Severe myopic macular degeneration.

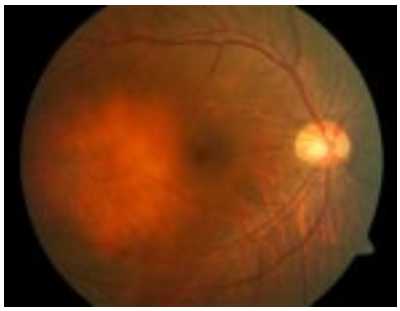

Figure 15: Choroidal haemangioma.

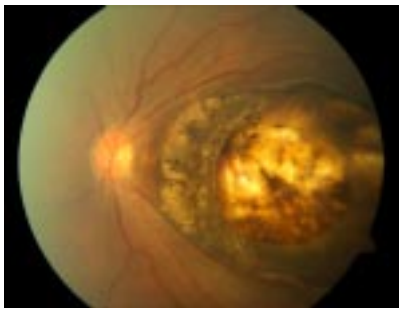

Figure 16: Healed toxoplasma retinochoroiditis involving the fovea.

Figure 14 shows very severe myopic chorioretinal degeneration involving the macula (myopic macular degeneration). There is atrophy of both the retinal pigment epithelium and the underlying choroidal vasculature, causing the white of the sclera to be more visible. Areas of hyperpigmentation known as Fuch's spots (blue arrow) represent areas of previous subretinal haemorrhage.

Figure 15 shows a large, slightly raised red-orange choroidal haemangioma occupying the temporal part of the macula. Asymptomatic lesions require no treatment.

Toxoplasmosis is an infestation by the obligatory intracellular protozoan parasite *Toxoplasma gondii*. It may be acquired or congenital. Most cases of congenital systemic toxoplasmosis are subclinical, and bilateral healed chorioretinal scars (Figure 16) may be discovered later in life either by chance on a routine fundus examination or when the child is found to have defective vision. Recurrence of old healed congenital ocular toxoplasmosis with retinitis and vitritis may occur.

Figure 17 shows an old shallow inferior retinal detachment with subretinal demarcation lines ("high water marks")(blue arrows) and pigmentary changes in the retinal periphery. These "high water marks" develop at the junction of attached and detached retina and take about three months to develop. The retinal detachment has involved the fovea, causing mild impairment of vision and a relative superior visual field defect.

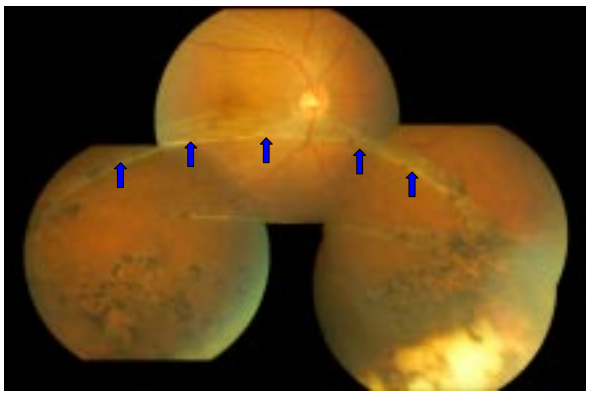

Figure 17: Chronic retinal detachment.

Cytomegalovirus (CMV) retinitis is the most common intraocular infection in patients with acquired immune deficiency syndrome (AIDS).[6] If left untreated, it will result in blindness. With the introduction of Highly Active Anti-Retroviral Treatment (HAART), there is a marked improvement in the prognosis of AIDS patients and a reduced risk of CMV retinitis[7]. The treatment of CMV retinitis includes systemic ganciclovir or foscarnet and local (intravitreal) therapy.

Figure 18 shows an area of CMV retinitis involving the inferior retina associated with retinal haemorrhages and vasculitis. This gives a "cheese and tomato ketchup" appearance.

Retinitis pigmentosa (RP) is a generic name for a group of inherited diseases characterised by night blindness and constricted visual fields. The classic triad of RP consists of bone-spicule pigmentation, arteriolar attenuation and waxy disc pallor. Figure 19 shows bone-spicule pigment clumps, some of which are paravascular, and mild arteriolar attenuation. There is atrophy of the macula.

Other ocular findings associated with RP include posterior subcapsular cataract, keratoconus, open angle glaucoma and optic disc drusen. Systemic associations include deafness, ataxia and mental deficiency. There is no known treatment for this condition.

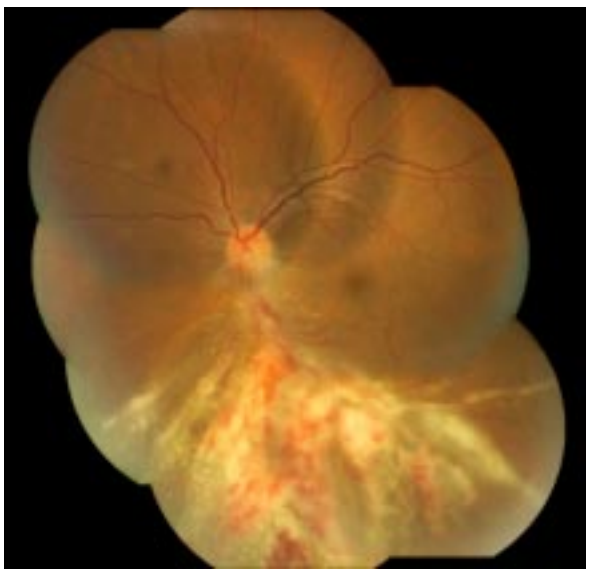

Figure 18: Cytomegalovirus retinitis.

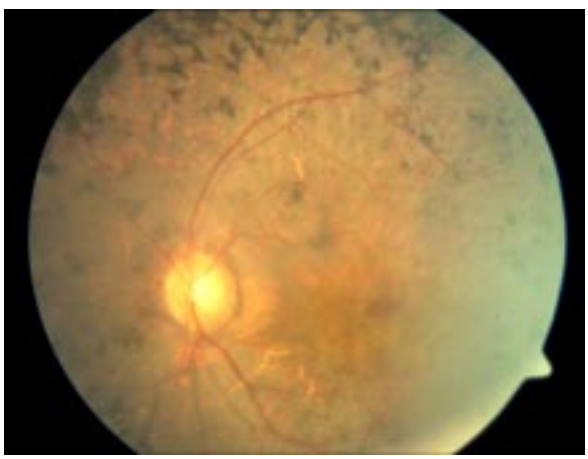

Figure 19: Retinitis pigmentosa.

OPTIC DISC ABNORMALITIES

Figure 20 shows a right optic disc with a cup-disc ratio of approximately 0.7. The edge of the cup is outlined by the angulation of the blood vessels as they dive backwards into the cup (white arrows). This patient requires additional assessment, including the measurement of intraocular pressure (tonometry) and visual field examination (perimetry) to exclude glaucoma. There is also peripapillary chorioretinal degeneration as well as diffuse chorioretinal atrophy, as evident by the unusually prominent large choroidal vessels.

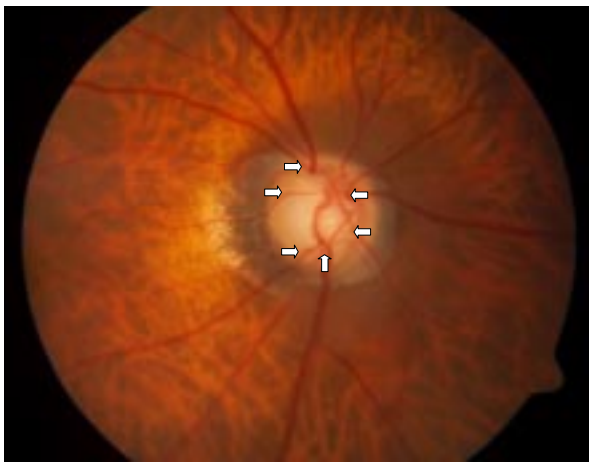

Figure 20: Increased cup-disc ratio.

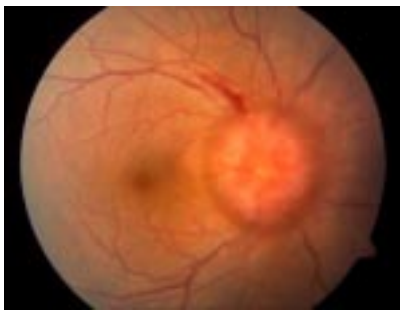

Figure 21: Optic disc swelling.

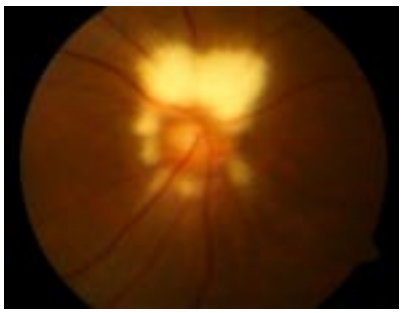

Figure 22: Myelinated nerve fibres.

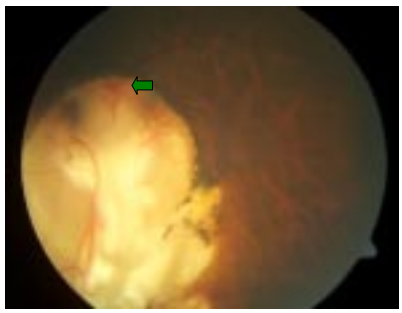

Figure 23: Optic disc coloboma.

Figure 21 shows a swollen optic disc with the disc surface elevated above the plane of the retina and is therefore out of focus. The disc margins are indistinct and there is a streak of peripapillary retinal haemorrhage superotemporal to the disc. Any patient with unilateral or bilateral swollen discs should be referred for further assessment.

Papilloedema is swelling of the optic nerve head produced by raised intracranial pressure. Symptoms suggestive of raised intracranial pressure are headache (made worse by coughing or straining), vomiting, changes in level of consciousness and diplopia. The disc swellings are usually bilateral in papilloedema.

Other causes of optic disc swelling include optic papillitis (optic neuritis) and malignant hypertension.

Figure 22 shows several peripapillary white patches of myelinated nerve fibres with frayed and feathery edges radiating from the optic disc.

Myelination is usually limited to the optic nerve. However, some retinal nerve fibres may assume a myelin sheath from oligodendrocytes. Myelinated nerve fibres are usually asymptomatic unless the macula is involved. An absolute or relative scotoma corresponding to the areas of myelination may be present[8].

Figure 23 shows a large coloboma involving the optic disc and the inferior retina. The optic disc is found at the top of the coloboma (green arrow). This is a rare condition due to incomplete closure of the fetal fissure. Eyes with colobomas often have decreased vision and a superior visual field defect.

Figure 24 shows a dysplastic coloboma of the optic disc resembling a morning glory flower. The optic nerve head is enlarged and excavated and is surrounded by an elevated annulus of chorioretinal pigmentary disturbance with retinal vessels radiating outwards like the spokes of a wheel.

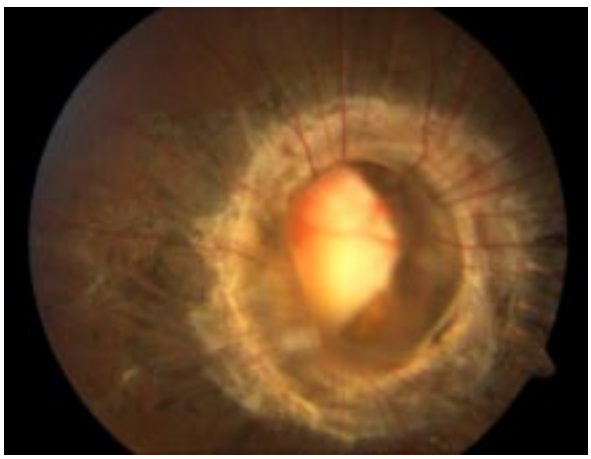

Figure 24: Morning glory syndrome.

SUMMARY

This pictorial guide serves to familiarise the family physician with the more common conditions affecting the ocular fundus, as well as a few rarer but easily recognisable conditions. Although one may not be able to view large areas of the ocular fundus readily with the direct ophthalmoscope, the easy accessibility of a fundus camera can overcome this problem.

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