Ocular fundus: other retinal vascular disorders

Hypertensive retinopathy
Various classification systems have been proposed to grade the ophthalmoscopic findings in hypertension. The retinal changes mirror the systemic circulation, and their severity correlates well with the development of the systemic complications of hypertension and with survival.

Retinal vascular changes in hypertension comprise:
- the vasospastic reaction to an acute pressure rise (the true hypertensive response)
- the arteriolosclerotic response to chronic elevation.

Clinical features

Generalised arteriolar narrowing
Arteriolosclerosis occurs through medial hyperplasia and fibrosis resulting from chronically elevated pressure. Slowly progressive arteriolosclerosis is a feature of normal ageing. On ophthalmoscopy, there is broadening of the arteriolar light reflex (‘burnished copper’, ‘polished silver’) and venous nipping at arteriovenous crossing points.

Focal arteriolar narrowing
A vasospastic effect occurring in response to an acute pressure rise.

Flame haemorrhages
These are located in the nerve fibre layer (Fig. 1) and result from capillary damage. Dot and blot haemorrhages can also develop.

Cotton wool spots
Small feathery white spots consisting of swollen axonal endings (Fig. 1) are caused by focal ischaemia.

Exudates
Well-defined yellow-white intraretinal collections of lipid are derived from vascular leakage and vary in size. At the macula, a ‘star’ may develop, consisting of exudates arranged in a bicycle spoke-like pattern radiating from its centre (Fig. 2).

Optic disc swelling
This is thought to be caused by local ischaemia. Rarely there is raised intracranial pressure (true papilloedema).

Arteriolar macroaneurysms
These localised arteriolar dilations (Fig. 3) are strongly associated with hypertension and arteriolosclerosis. Macroaneurysms are prone to leak blood and serous fluids. Symptomatic lesions at the macula are ablated using laser.

Microaneurysms
These are similar to the lesions occurring in diabetic retinopathy and are well-defined red dots.

These features are summarised in Table 1.

Table 1
<table>
<thead>
<tr>
<th>Grade</th>
<th>Features</th>
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<tbody>
<tr>
<td>Hypertension</td>
<td>Generalised arteriolar narrowing</td>
</tr>
<tr>
<td>2</td>
<td>More marked generalised narrowing with irregular points of focal constriction</td>
</tr>
<tr>
<td>3</td>
<td>Generalised and focal narrowing plus cotton wool spots, retinal haemorrhages, hard exudates</td>
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<tr>
<td>4</td>
<td>As grade 3 but with swelling of the optic disc</td>
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Arteriolosclerosis
- Decreased venular visibility at arteriovenous crossing points
- Slight broadening of the arteriolar light reflex
- Deflection of the venule at arteriovenous crossing points
- ‘Copper wire’ arterioles, marked venular narrowing and deflection at crossing points
- ‘Silver wire’ arterioles, extreme crossing changes

Malignant hypertension
Malignant hypertension is the clinical syndrome of an accelerated rise in blood pressure. Untreated, mortality at 1 year is 90%. In the retina, it is characterised by Grade 4 hypertensive changes (Table 1).

Retinopathy of prematurity (ROP)
This fibrovascular retinal proliferative disorder occurs in premature and low birth weight babies. Its development is associated with high inhaled oxygen concentration during the neonatal period. Other less well-defined risk factors, such as maternal smoking, neonatal sepsis and blood transfusion, have been implicated.

Normal retinal vascularisation is not complete until full-term gestation. It is thought that in the premature baby, normal retinal vascularisation ceases, perhaps because of adequate oxygenation from inspired air, then either continues normally (resolved ROP) or proceeds in an abnormal manner, vessels growing forwards into the vitreous cavity. These vessels may bleed. The associated fibrous component can contract, detaching the retina.

Fig. 1 Flame haemorrhages and cotton wool spots in hypertensive retinopathy.

Fig. 2 Macular exudates (‘star’) in hypertensive retinopathy.

Fig. 3 Haemorrhage from a macroaneurysm.
Clinical features
Active ROP may progress through five stages of increasing severity (Fig. 4), culminating in total retinal detachment. Spontaneous regression of the earlier stages occurs without intervention in a majority, but sequelae may still cause sight loss. The more anterior the limit of vascularised retina at the time of the development of active ROP (the ‘zone’ of the ROP), the better the prognosis.

Other changes that have poor prognostic significance are also recognised (‘plus’ disease): dilated veins and tortuous arteries at the posterior pole, vitreous haemorrhage and iris vascularisation.

Premature babies are also susceptible to strabismus and myopia, and to intracranial haemorrhage causing cortical blindness.

Screening
Screening is recommended for babies born at or less than 31 weeks’ gestational age and for those weighing less than 1500 g at birth.

Dilated fundal examination is carried out regularly from 6 or 7 weeks after birth until the nasal retina is fully vascularised. A very premature subgroup are screened more intensively.

Treatment
Treatment is indicated for ‘threshold’ disease (defined as extensive stage 3 changes). This is the severity of disease that is likely to progress to visual loss and at which the benefits of treatment outweigh the risks. The avascular area is ablated using either cryotherapy or laser to induce regression of abnormally growing new vessels. These babies often have other medical problems that make treatment hazardous. Surgery for retinal detachment has had only very limited success.

Sickle-cell retinopathy
Mutant haemoglobin, such as sickle haemoglobin S, causes red blood cells to behave abnormally, in particular making them less flexible and unable to pass freely through small blood vessels. Hypoxia exacerbates this tendency. Combinations of abnormal haemoglobins occur: the most common, and the least severe, is haemoglobin S combined with normal haemoglobin A (sickle trait). Haemoglobin S is common in blacks but extremely rare in whites.

Sickle-cell retinopathy is caused by the impaction of deformed red cells in the retinal vasculature, leading to occlusion and ischaemia. Paradoxically, sickle cell disease (pure haemoglobin S) does not cause the most severe retinopathy.

Clinical features
Systemic manifestations include anaemia and sickle crises.

The eye shows two types of change:

- **Proliferative retinopathy.**
  Peripheral proliferative changes develop after vascular occlusion and arteriovenous anastomosis. The new vessels resemble a fan (‘sea-fan’ neovascularisation). Vitreous haemorrhage may occur. Progressive contraction of fibrovascular tissue may lead to tractional retinal detachment or to the formation of retinal tears.

- **Non-proliferative retinopathy.**
  Black ‘sunburst’ scars and ‘salmon-patch’ retinal haemorrhages are probably a result of infarction. Venous tortuosity is common. Retinal artery or vein occlusion may occur.

Management
Patients with sickle disease should be screened for retinopathy at regular intervals and observed more frequently if signs develop. Laser photocoagulation is performed for neovascularisation.

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**Ocular fundus: other retinal vascular disorders**

- Hypertensive retinopathy involves an arteriosclerotic and a vasospastic response.
- Retinopathy of prematurity:
  - can resolve naturally or progress through five stages
  - premature and low birth weight babies should be screened
  - ‘threshold’ ROP (extensive stage 3) is treated with laser or cryotherapy to the avascular area
  - high incidence of myopia and strabismus.
- Threshold (ROP) disease (extensive stage 3) is treated with laser or cryotherapy to the avascular area.
- Sickle-cell retinopathy involves proliferative (‘sea-fan’ neovascularisation) and non-proliferative (‘black sunburst’; ‘salmon-patch’ haemorrhages, venous tortuosity, vascular occlusion) changes. The former are treated with laser photocoagulation.