

Secondary Glaucoma

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CLASSIFICATION

Secondary glaucoma occurs when the intraocular pressure (IOP) is raised secondary to another ocular condition. These form a small proportion of all types of glaucomas, although frequently they produce the most difficult problems in diagnosis and management.

Although many of these glaucomas are chronic and develop glaucomatous cupping and field loss with time, these features need not be present to make the diagnosis. A patient presenting acutely with a high IOP secondary to, for example, a traumatic hyphaema will still be considered to have a secondary glaucoma (although strictly speaking they will be suffering from secondary ocular hypertension).

Increased IOP secondary to other conditions can occur from a fall in outflow facility or an increase in aqueous humour production. The types of glaucoma may be grouped accordingly.

Outflow obstruction

- (a) Pre trabecular block from iris-trabecular contact or adhesion, e.g. secondary angle closure glaucoma:
 - with pupil block,
 - without pupil block.
- (b) Trabecular block secondary to damage to the mesh-work, e.g. secondary open angle glaucoma.

- (c) Post trabecular block, e.g. from raised episcleral venous pressure.

(ii) Hypersecretion glaucoma

These are rare. An example is epidemic dropsy glaucoma which occurs secondary to contamination of cooking oil with seeds of the Mexican Poppy (*Argemone mexicana*). It produces a dramatic rise in IOP, associated with prostaglandin release in the anterior segment.

TREATMENT

Treatment of the secondary glaucomas is directed firstly at the cause of the condition: thus inflammation is suppressed, an obstructing lens is removed. This may not always succeed in IOP control as the trabecular meshwork may have been extensively damaged, so that even after removal of the cause longterm glaucoma therapy will still be required. Many secondary glaucomas present with extremely high IOPs, they require emergency treatment with carbonic anhydrase inhibitors, hyperosmotic agents and beta-blockers to reduce the IOP. Long-term strategy then depends upon the predicted natural history of the condition as the therapeutic approaches differ considerably with each individual type of glaucoma.

PRETRABECULAR OUTFLOW OBSTRUCTION: SECONDARY ANGLE CLOSURE GLAUCOMA

SECONDARY ANGLE CLOSURE GLAUCOMA WITH PUPIL BLOCK

Secondary pupil block occurs when the iris becomes adherent at the pupillary margin to the lens. Possible reasons for this are:

- (i) inflammation in the anterior segment;
- (ii) occlusion of the pupil in aphakic and pseudophakic eyes by vitreous gel or the implant;
- (iii) forward movement of the anterior lens surface (including lens growth in small eyes).

Glaucoma may persist after an attack of pupil block from any cause if a large proportion of the angle has been closed by peripheral anterior synechiae which have formed during the attack, or if the trabecular meshwork has become damaged.

Inflammation in the anterior segment

Inflammation of the anterior segment, producing posterior synechiae and occluding the pupillary aperture, is the most common cause of secondary angle closure glaucoma with pupil block, although the use of topical steroids and mydriatics have been effective in reducing its incidence. Pupil block may develop insidiously in low grade iritis with gradual occlusion of the pupil, or suddenly in acute iritis. Treatment is aimed at preventing occlusion of the pupil with topical steroids and mydriatics, but for the established condition laser or surgical iridectomy is necessary to break the pupillary block. Before the advent of YAG laser iridotomies peripheral iridectomy was performed routinely during most intraocular surgery to avoid potential pupil block post-operatively.

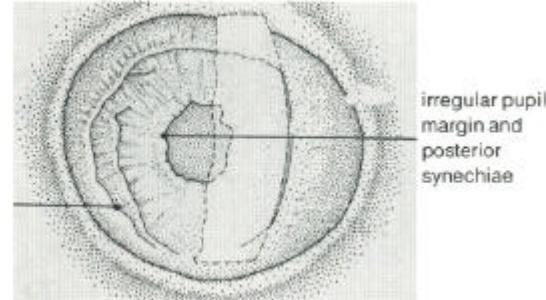
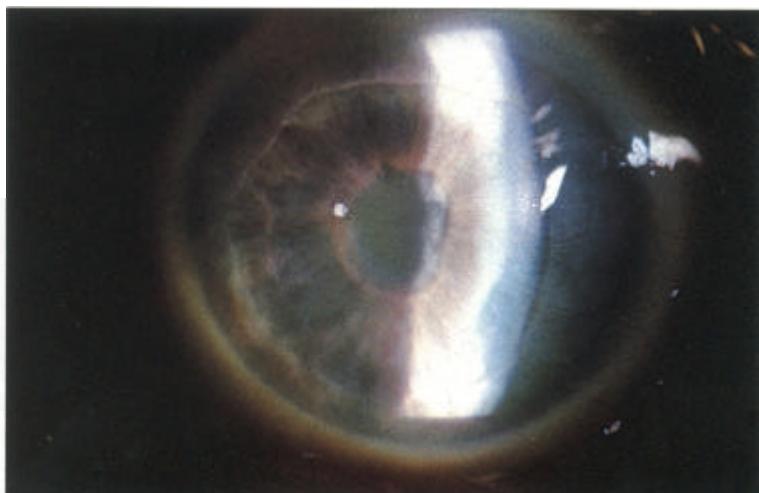


Fig. 8.1 Examination of the anterior segment of a patient with chronic uveitis reveals a diffuse scattering of light from aqueous flare and clearly visible circumferential peripheral anterior synechiae. The pupil is occluded by posterior synechiae and iris bombe is present.

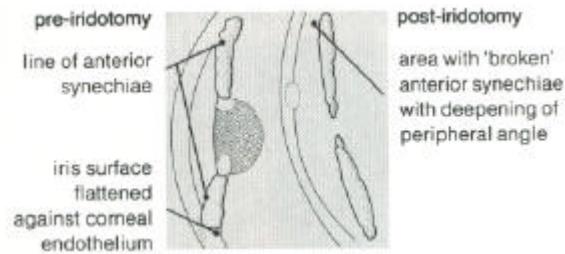
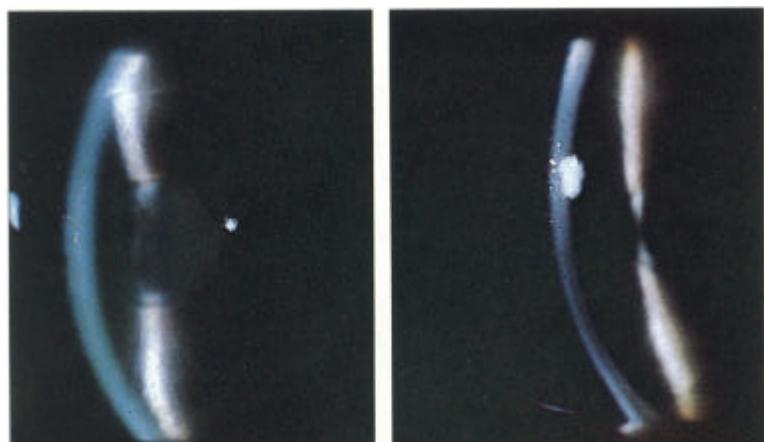


Fig. 8.2 These two slit-image photographs show the same patient before and after treatment. Peripheral anterior synechiae are evident as are pupil block and iris bombe. A deepening of the peripheral part of the anterior chamber is clearly evident following laser iridotomy (right).

Pupil block in aphakic and pseudophakic eyes

Following intracapsular cataract surgery pupil block often occurs as an acute event although sometimes the effects may not become apparent until weeks later. The diagnosis is suggested by a combination of raised IOP and peripheral shallowing of the anterior chamber. Usually there is little vitreous prolapse, the central anterior chamber may be of normal depth, and slight shallowing at the periphery is observed, although extensive prolapse of gel through the pupil may also occur.

The established condition is treated by laser iridotomy and when successful there is immediate deepening of the periphery of the chamber. Occasionally the iridotomy may become occluded with vitreous gel. When this occurs, another iridotomy should be tried or, alternatively, a core vitrectomy carried out to re-establish aqueous flow.

Pseudophakic pupil block can occur in eyes with an anterior chamber lens and a nonfunctioning iridotomy. Coexistent inflammation causes adhesions to develop between the lens and the iris. A rigid anterior chamber lens may maintain axial anterior chamber depth, with iris ballooning around the edges of the lens making the periphery of the anterior chamber shallower and occluding the angle. Treatment is to create another iridotomy. Modern techniques of extracapsular cataract surgery with posterior chamber lens implantation are sufficiently free from postoperative inflammation not to require an iridotomy and pupil block is rarely seen in these eyes. When it does occur it may be due to the implant becoming trapped in the pupillary aperture or from occlusion by posterior synechiae to the lens capsule remnants (see Chapter 11).

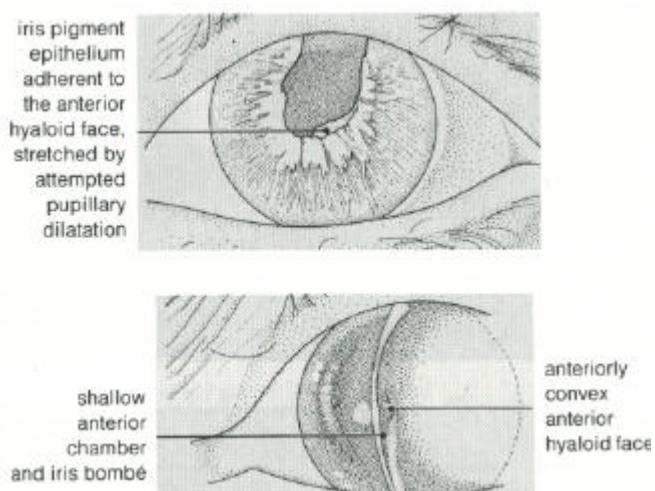


Fig. 8.3 In this patient, who has had intracapsular cataract extraction, adherence of the pigment epithelium of the iris to the anterior hyaloid face by posterior synechiae is seen following attempted mydriasis as a pigmented adhesion between the pupil margin and anterior hyaloid face (top). A slit-image photograph of the same patient (bottom) shows anterior iris convexity together with a shallow anterior chamber. The extent to which the vitreous gel fills or is prolapsed into the anterior chamber determines the central depth. Treatment is by intensive mydriasis, but if this fails to break the synechiae to the hyaloid face a posterior synechotomy, laser iridotomy or anterior vitrectomy is indicated.

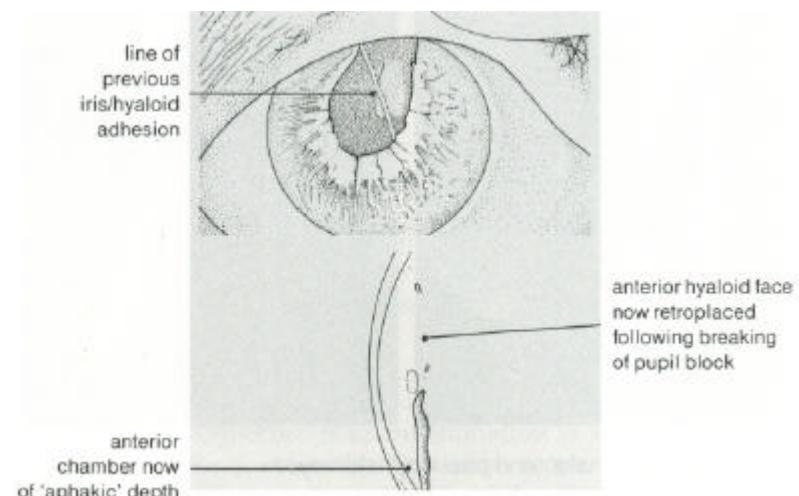
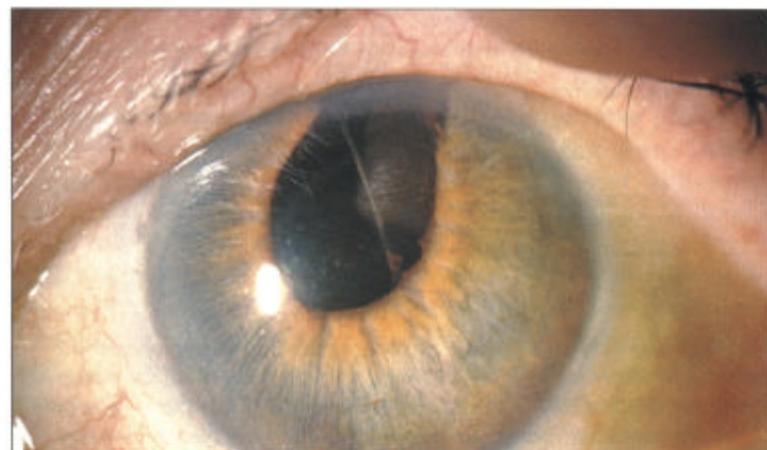


Fig. 8.4 Following a synechotomy procedure in the same patient as in Fig. 8.3, an oblique pigmented line can be seen crossing the anterior hyaloid face with opacification of one half of the face and an optically clear zone in the other half. This iris pigment remained after the division of adhesions between the anterior hyaloid face and the pupil margin. The optically clear zone represents the anterior hyaloid face which has been exposed following successful surgical treatment and subsequent mydriasis. A slit -image photograph demonstrates the deepening of the anterior chamber seen following resolution of the block.

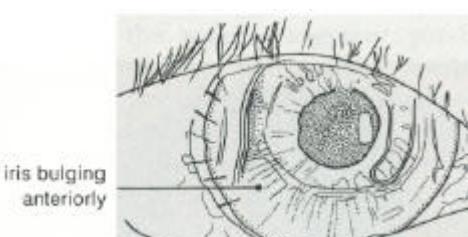
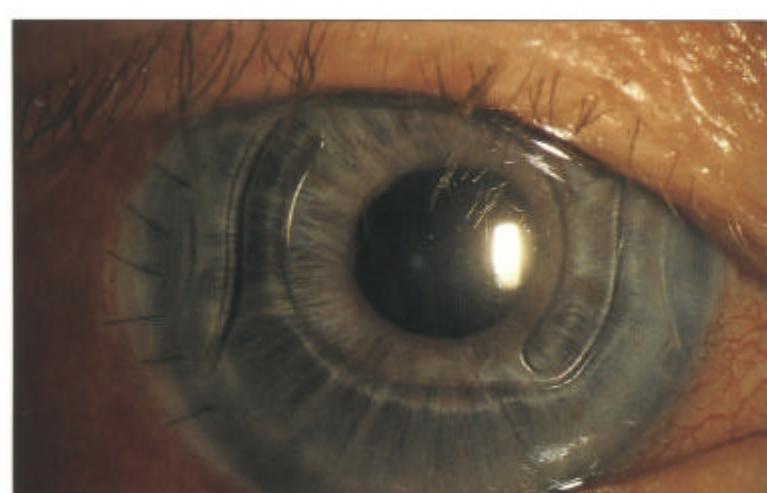


Fig. 8.5 This eye developed pupillary block immediately postoperatively following a secondary anterior chamber implant. The iris can be seen bulging forwards around the implant that maintains the AC depth. The anterior chamber angle was reopened by a laser iridotomy.

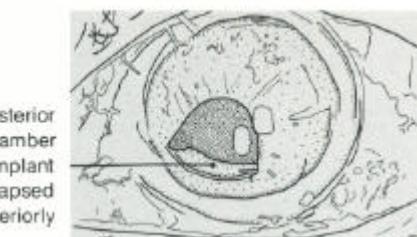
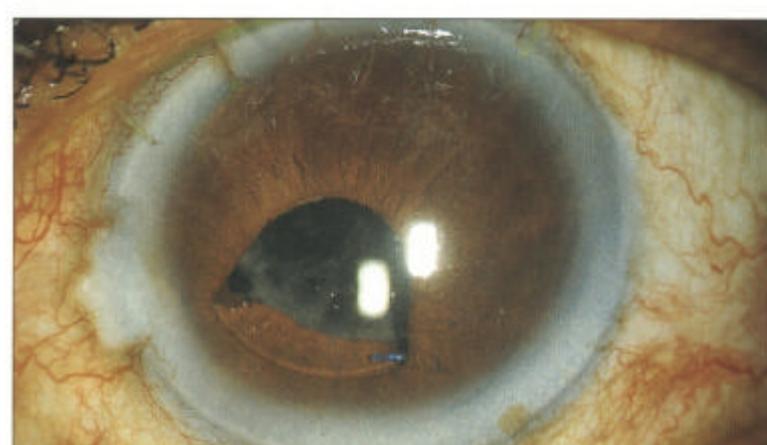


Fig. 8.6 Shallowing of the anterior chamber, for instance from wound leakage, can lead to a posterior chamber implant prolapsing forwards to block the pupil and create pupil block. In the early stages the implant can be repositioned but synechiae soon occur and in these cases pupil block is more easily reversed by a laser iridotomy.

Forward movement of the anterior lens surface

Intumescence of the lens, anterior dislocation or, in eyes with small anterior segments, age-related growth changes in the lens can all produce pupil block and angle closure by forward

movement of the anterior lens surface. In these eyes treatment may be an iridotomy or, alternatively, lens removal. With the former, angle closure may still occur from progressive lens growth.

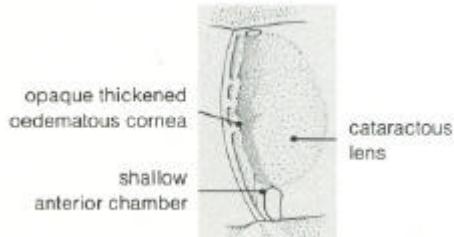
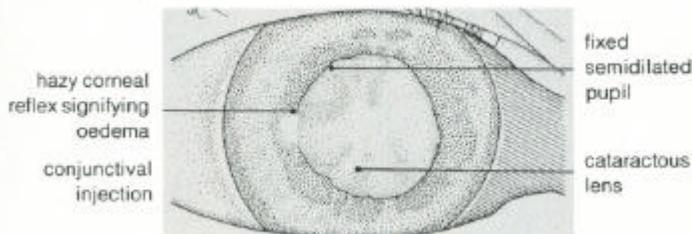
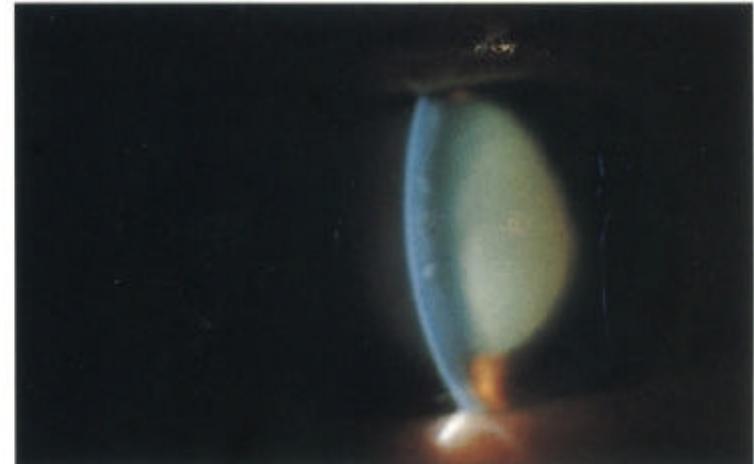


Fig. 8.7 Acute angle closure glaucoma secondary to intumescence of the lens. In addition to the swollen cataractous lens, this eye has other features which are suggestive of a raised intraocular pressure, these include a hazy corneal reflex from corneal oedema, a semidilated and fixed pupil from iris infarction and circumlimbal injection signifying inflammation.

Fig. 8.8 A slit-image photograph of the same patient shows evidence of corneal oedema and opacification of the lens. Notice the shallowing of the anterior chamber. Some uveitis may be present because of ischaemia and this must be differentiated from the large accumulations of lens material and macrophages seen in the deep anterior chamber of patients with phacolytic glaucoma.

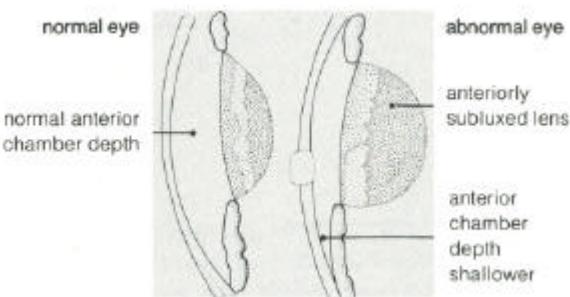
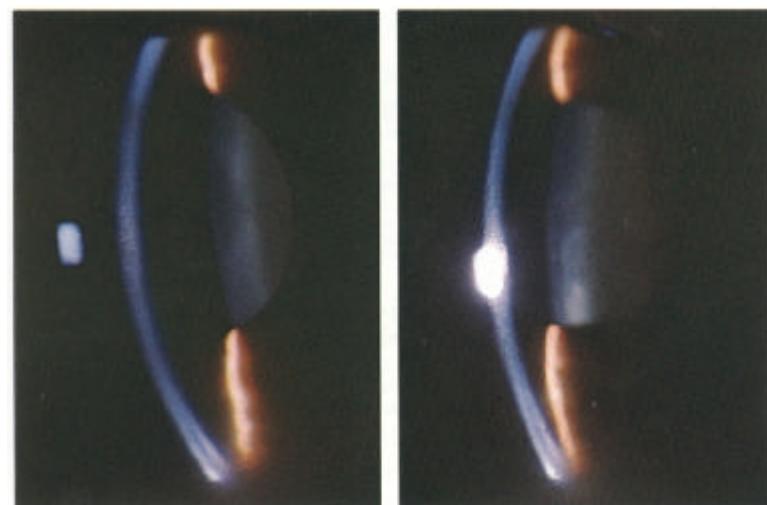


Fig. 8.9 Slit-image photography of the left and right eye of this patient demonstrates a subluxed lens in the left eye. The anterior chamber is shallow and further examination shows that the anterior lens surface is closer to the posterior corneal surface inferiorly than superiorly occluding the pupil and producing block.

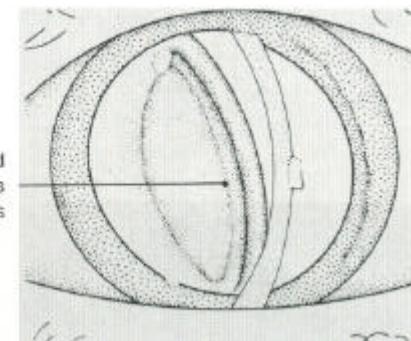
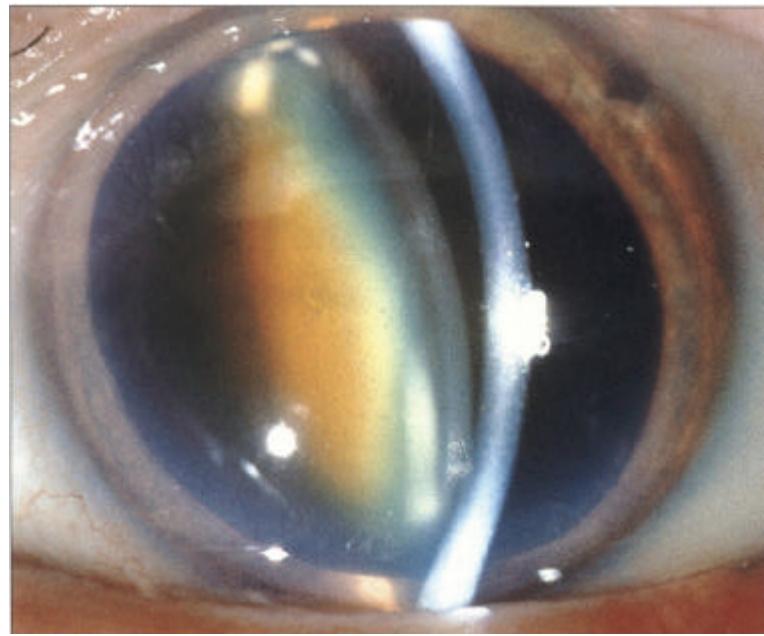


Fig. 8.10 Occasionally pupillary dilatation during routine funduscopy allows a subluxated lens to swing into the pupil and block communication between the posterior and anterior chamber. Careful positioning of the patient and use of miotics usually allow the lens to be repositioned safely. Notice the nuclear cataract in this lens.

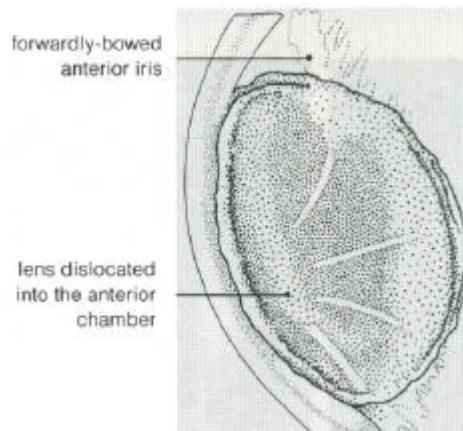
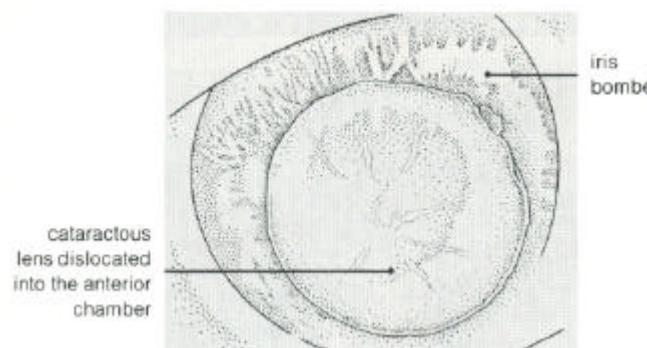
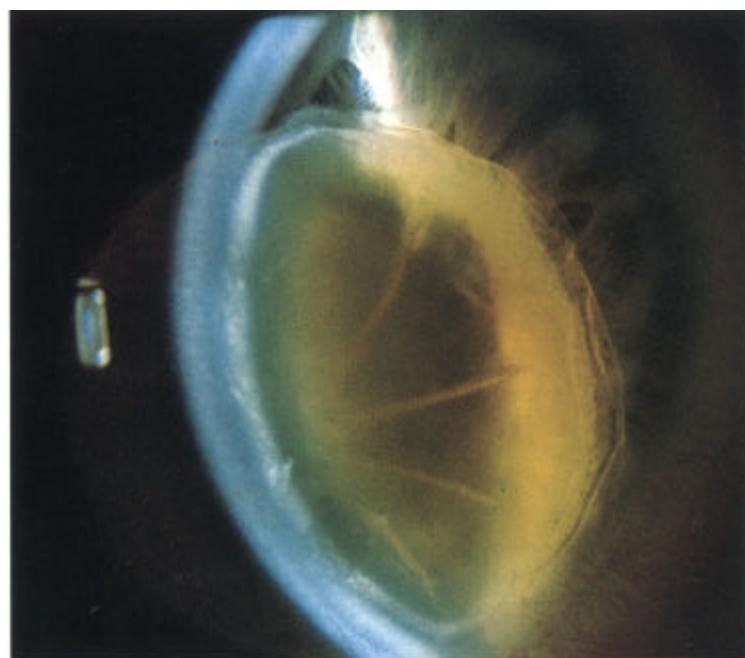


Fig. 8.11 Pupil block glaucoma following traumatic anterior dislocation of the cataractous lens. The slit-image view demonstrates how the pupil is totally blocked by the lens with the peripheral iris convex anteriorly.

Angle closure in small eyes

Angle closure glaucoma typically occurs in hypermetropic eyes which have a small anterior segment. There are a number of other ocular conditions where the eye itself is considerably smaller than normal, such as congenital high hypermetropia (*nanophthalmos*; see Chapter 10), mucopolysaccharidoses or

congenital syphilis. Angle closure glaucoma occurs frequently in these rare conditions and often responds poorly to open iridectomy because the anterior chamber fails to reform and the eye develops cilio-lenticular block (*malignant glaucoma*). Laser iridotomies is the preferred method of treatment to break the pupil block in these unusual cases.

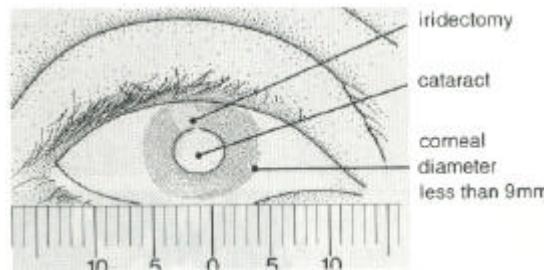
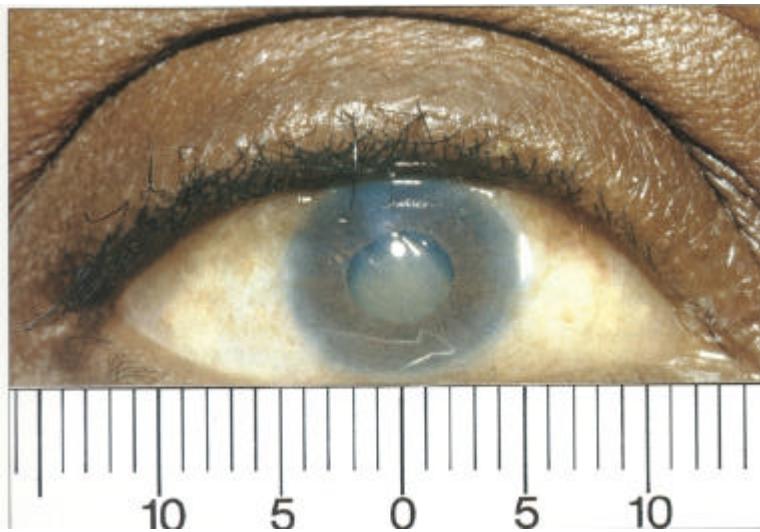


Fig. 8.12 A small shallow anterior chamber predisposes to secondary glaucoma with pupil block and this is occasionally seen in patients with congenital syphilis. In this patient the millimetre scale demonstrates a horizontal corneal diameter of less than 9mm (the eye has had an iridectomy and a cataract is also present). Measurement of the axial length confirms the small size of the eye.

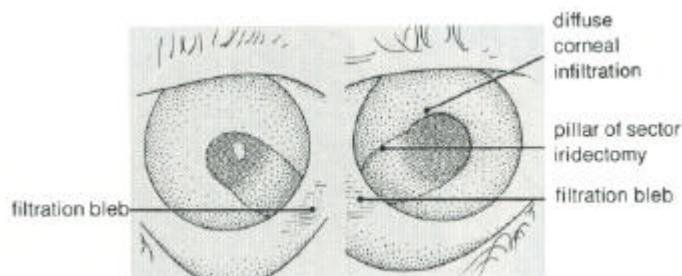


Fig. 8.13 Other conditions which result in small eyes and increase the risk of angle closure glaucoma include retinopathy of prematurity, high hypermetropia (*nanophthalmos*) and the mucopolysaccharidoses (as in this patient with the cloudy corneas of Maroteaux-Lamy syndrome who has also had bilateral broad optical iridectomies). A small eye will be at risk from progressive angle closure even after removal of pupil block by means of an iridotomies. Glaucoma control in such eyes is extremely difficult as fistulizing surgery may be followed by cilio-lenticular block. Treatment in such eyes may include elective lens extraction with a posterior chamber implant in combination with a fistulizing operation.

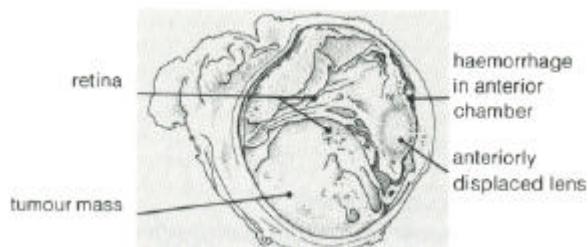
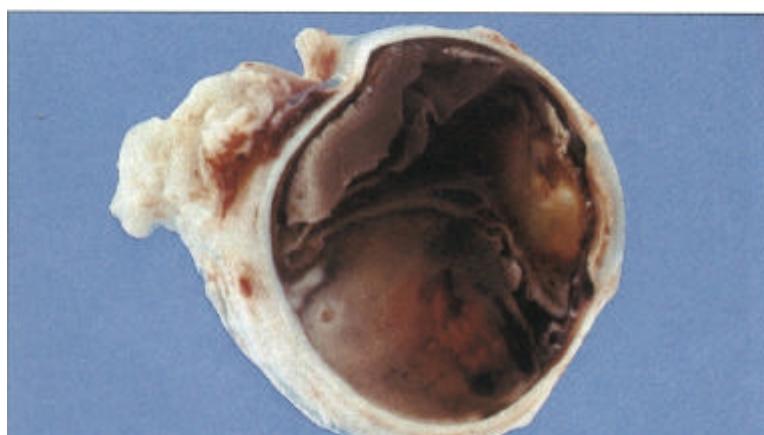
SECONDARY ANGLE CLOSURE GLAUCOMA WITHOUT PUPIL BLOCK

Angle closure may occur without pupil block in three ways:

- (i) changes in the posterior segment which push the lens-iris diaphragm forwards;
- (ii) changes in the anterior segment which result in loss of the anterior chamber and iris-trabecular contact;
- (iii) cellular proliferation within the angle of the anterior chamber resulting in iris-trabecular adhesions.

Changes in the posterior segment

Tumours form the most important (although the least common) group of conditions that are responsible for changes in the posterior segment pushing the lens-iris diaphragm forwards. Other conditions which can cause an increase in the volume of the posterior segment include choroidal effusions arising either spontaneously or secondary to intraocular surgery, encircling straps used in retinal detachment surgery or posterior scleritis. An increase in permeability following a breakdown



of the blood-retinal barrier may occur after panretinal photocoagulation or central retinal vein occlusion and lead to either a choroidal effusion or a volume increase of the posterior segment pushing the lens-iris diaphragm forwards. The 101? may not initially be raised in these eyes, despite angle closure, as detachment of the ciliary body from the effusion leads to aqueous hyposecretion.

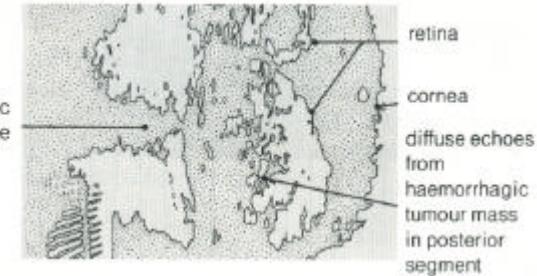


Fig. 8.14 This patient had a long-standing blind right eye and presented with recent onset of pain, conjunctival oedema, and haemorrhage into the anterior chamber. The ultrasound scan of this patient shows posterior segment echoes which are suggestive of a tumour mass.

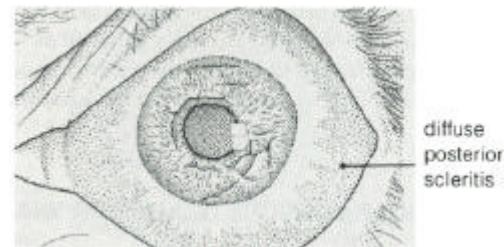
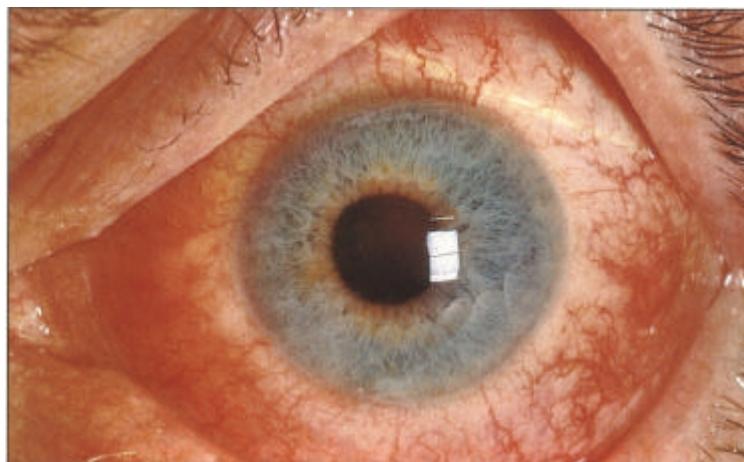


Fig. 8.16 Posterior scleritis may be associated with an annular choroidal effusion causing a forward rotation of the ciliary body about the scleral spur and a corresponding forward movement of the lens-iris diaphragm to produce angle closure glaucoma. This photograph shows an eye with diffuse posterior scleritis.

Fig. 8.15 Hemisection of the enucleated eye shows a large haemorrhagic choroidal melanoma together with an anteriorly displaced lens and loss of the anterior chamber.

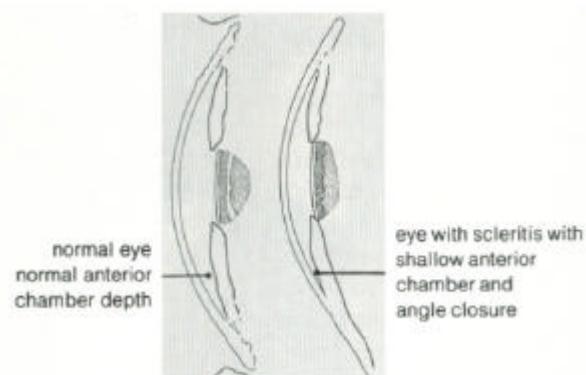
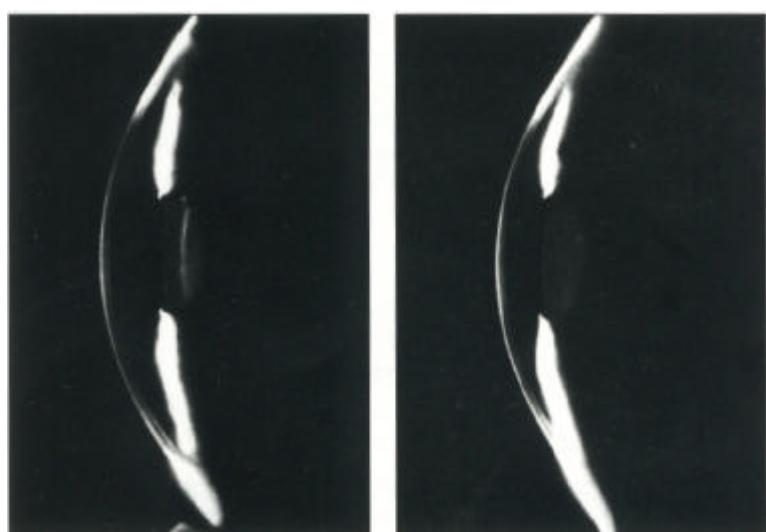


Fig. 8.17 Slit-image photographs of each eye of the same patient show a considerable difference in anterior chamber depth between the two eyes with angle closure in the affected eye.

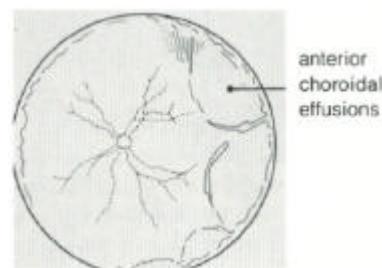
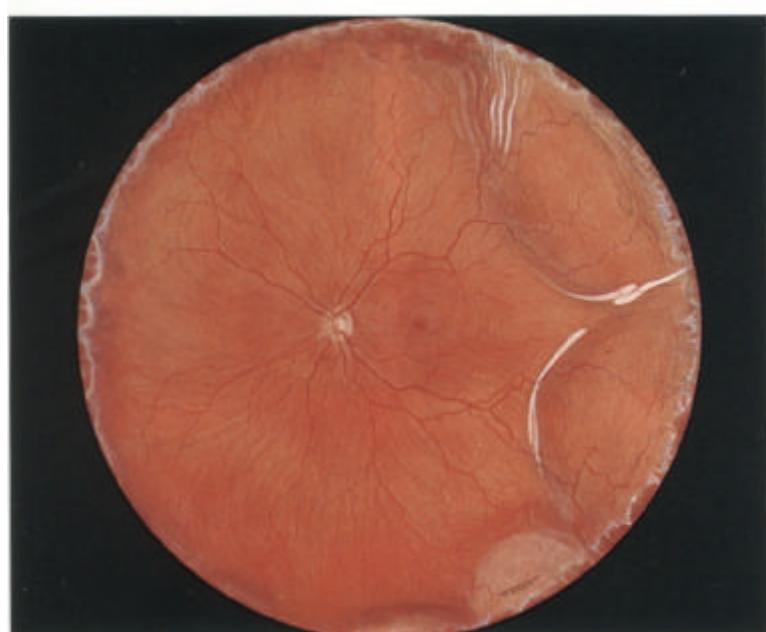


Fig. 8.18A fundus painting of the same patient as in Fig. 8.16 reveals an annular choroidal effusion which was confirmed by a B-scan ultrasonography (Fig. 8.19). Such effusions are easily missed unless the peripheral fundus is carefully inspected under full mydriasis.

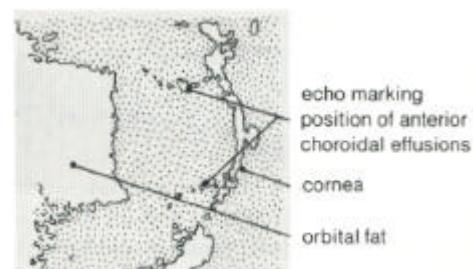


Fig. 8.19B-scan ultrasound of the same patient showing choroidal effusion. Management requires investigation for the cause of scleritis and anti-inflammatory therapy such as systemic nonsteroidal anti-inflammatory drugs or steroids. Treatment of the scleritis will be followed by spontaneous resolution of the effusions with deepening of the anterior chamber. Antiglaucoma therapy involves acetazolamide and topical beta-blockers for the raised intraocular pressure, and cycloplegics to dilate the pupil and deepen the anterior chamber, together with topical steroids. Should the angle remain closed for longer than a few days a laser iridotomy will help in limiting the development of peripheral anterior synechiae.

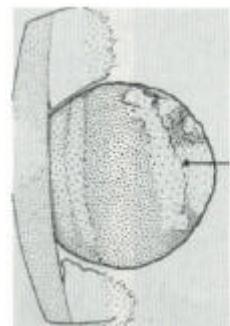
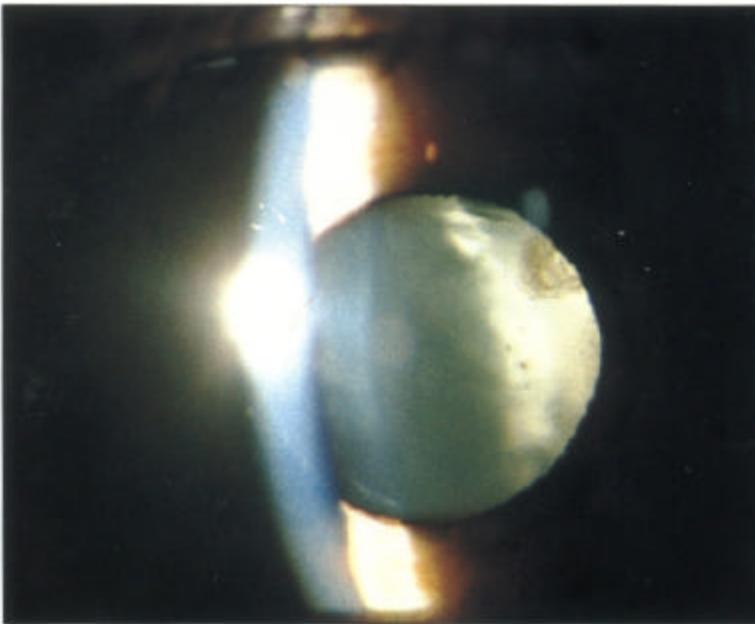


Fig. 8.20 Persistent hyperplastic primary vitreous can produce a contracting retrobulbar mass with forward rotation of the ciliary body and lens-iris diaphragm, pushing the iris forwards to occlude the angle. The anterior chamber is often shallow in these eyes making angle occlusion more likely. This slit-image photograph shows a shallow anterior chamber and retrobulbar mass.

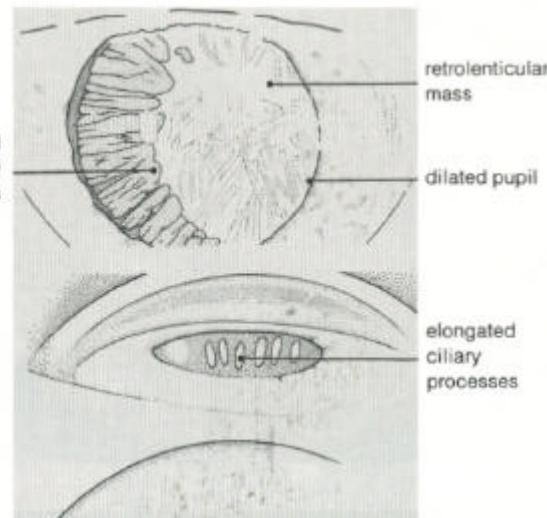
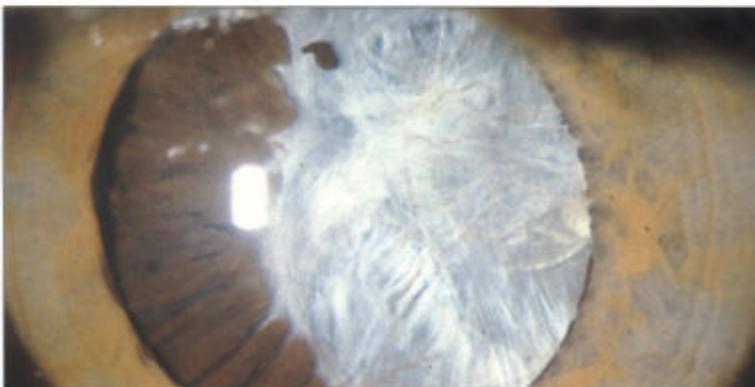


Fig. 8.21 Following mydriasis, elongated ciliary processes may be seen adhering to the mass. Gonioscopy further demonstrates the traction on the ciliary body with elongation of the ciliary process. The treatment of this condition has been revolutionized by vitreoretinal surgery. Useful visual results may be obtained in a number of these patients.

Changes in the anterior segment

Loss of the anterior chamber from leakage of aqueous will cause iris-trabecular contact and if the rate of loss exceeds the capacity to reform aqueous the anterior chamber will flatten and in time peripheral anterior synechiae (hastened by coincidental inflammation) will develop.

Cilio-lenticular block (malignant glaucoma)

This term is used to describe eyes with very high intraocular pressures, absent or shallow anterior chambers and a retrobulbar accumulation of aqueous humour in the absence of pupil block. The most common cause of this rare condition is fistulizing surgery on an eye with a shallow anterior chamber. Surgery on the fellow eye may be followed by the same result.

The primary process appears to be an obstruction to forward movement of aqueous humour in the presence of a shallow anterior chamber causing misdirection of aqueous into the posterior segment with pooling in the vitreous gel and forward movement of the lens-iris diaphragm to occlude the angle.

The old term for cilio-lenticular block was malignant glaucoma which was originally used because the eye did not respond to, and appeared to be made worse by, treatment with pilocarpine. The treatment for cilio-lenticular block should initially be topical atropine (to relax the ciliary muscle and pull the lens-iris diaphragm posteriorly) together with acetazolamide, beta-blockers, and hyperosmotic agents (to lower intraocular pressure, dehydrate the vitreous and reduce its volume). Laser

treatment can be used in the phakic eye; by directly applying argon laser burns to the ciliary processes they may be reduced in size and allow aqueous to percolate around the equator of the lens into the posterior chamber. YAG laser treatment has a very specific role in the aphakic eye (see below). Surgical treatment involves decompression of the retro-lenticular aqueous pool by pars plana vitrectomy combined with perforation of the anterior hyaloid face. This is usually successful. The older operations of vitreous aspiration and reformation of the anterior chamber (Chandler's operation) or cataract extraction have now been largely superceded.

A process similar to that described above can occur in eyes that have had the natural lens removed. Cilio-vitreal block can occur if adhesions exist between the anterior hyaloid face and

the ciliary processes which may be difficult to distinguish from aphakic pupil block. The diagnosis is confirmed, however, if following YAG laser iridotomy the condition is not resolved and vitreous is seen to be occluding the iridotomy. Surgical or YAG laser rupture of the anterior hyaloid face is curative in this condition. Cilio-pseudophakic lenticular block may also occur. Physical signs are essentially the same as for straightforward cilio-lenticular block with the exception that a posterior chamber intraocular lens with an intact posterior capsule is present. For these eyes the treatment of first choice appears to be YAG laser vitreolysis and posterior capsulotomy. This photo disruption allows aqueous to percolate from the loculated pools within the anterior vitreous into the posterior chamber and thus relieve the block to aqueous flow.

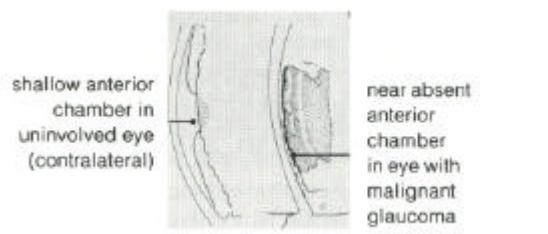
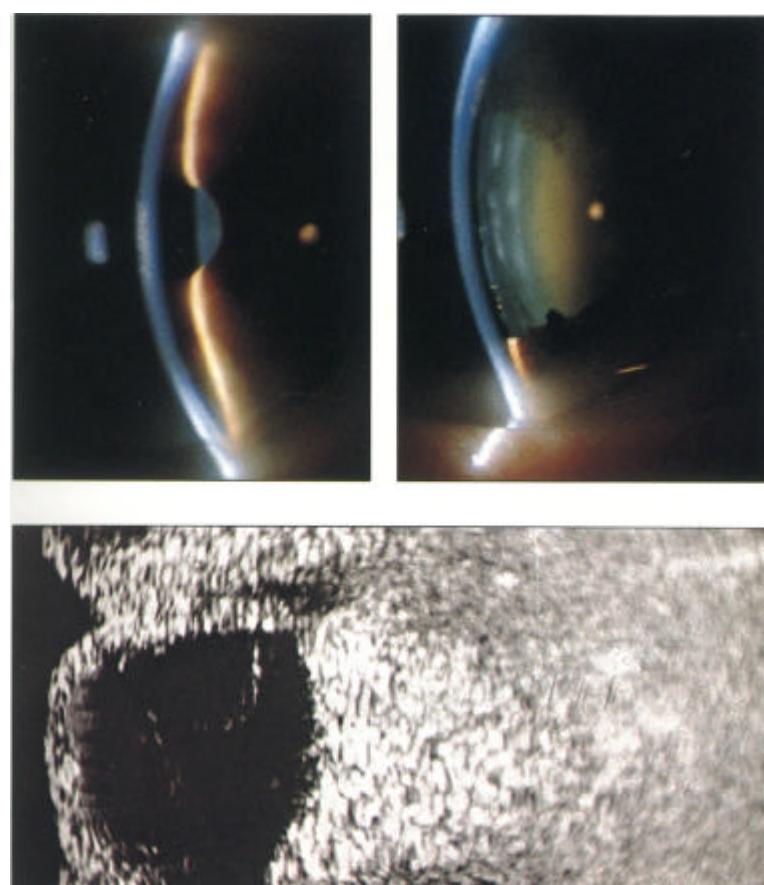


Fig. 8.22 These two slit-image photographs of the anterior segments of both eyes from one patient demonstrate a shallow anterior chamber in one eye while the fellow eye has no anterior chamber. This particular eye has recently undergone filtering surgery which led to the loss of the anterior chamber and the development of cilio-lenticular block.



Fig. 8.23 The ultrasound B scan demonstrates retro-lenticular accumulation of aqueous humour in a neglected case of cilio-vitreal block.

Cellular proliferation with angle closure

Different cell types may be responsible for cellular proliferation within the angle of the anterior chamber angle:

- # neovascularization e.g. thrombotic glaucoma;
- # endothelium e.g. iridocorneal endothelial syndromes;
- # epithelium e.g. epithelialization of the anterior chamber.

Neovascularization

Neovascularization (rubeosis) of the iris and angle, with accompanying fibrous tissue formation, is the most common type of cellular proliferation causing angle occlusion. It is seen frequently in diabetic patients, or in eyes having suffered a central retinal vein occlusion with extensive loss of retinal perfusion (see Chapter 14). Iris neovascularization can occur less commonly with diffuse retinal vascular disease, ocular arterial insufficiency, long standing retinal detachment or intraocular tumours. In all these conditions the stimulus for neo-

vascularization would appear to be retinal hypoxia, with the release of neovascular factors which stimulate new vessel growth on the iris and in the angle. New vessels appear initially around the pupil margin and in the angle. Contraction of the fibrovascular tissue on the anterior iris surface causes the fixed dilated pupil seen in the late stages of the condition. New vessels in the angle are usually followed by PAS formation, outflow obstruction and raised IOP follow. The extent and rapidity of this process varies with the disease. It may become apparent within weeks, in eyes with severe retinal vein occlusion causing extensive retinal hypoxia, although it occurs less rapidly in diabetic patients. During the process of neovascularization there may be a massive and sudden breakdown of the blood-aqueous barrier with inflammation in the anterior chamber. The patient experiences severe pain in the eye, and by the time of presentation the IOP may have risen to very high levels from the development of PAS.

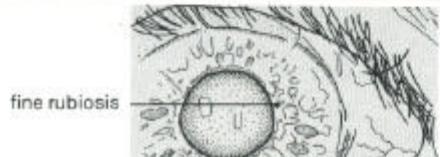
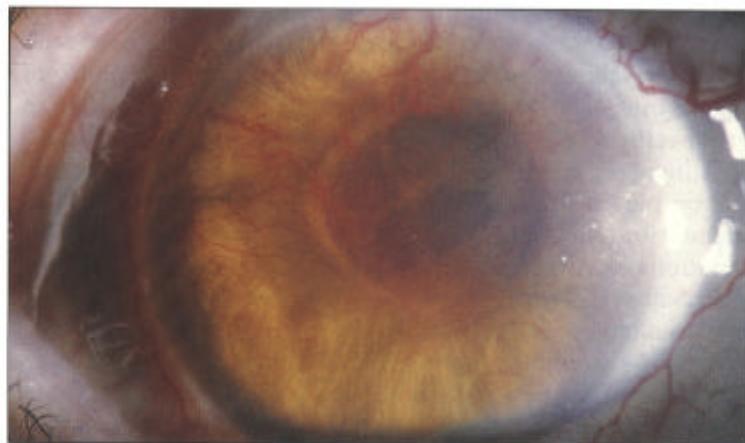
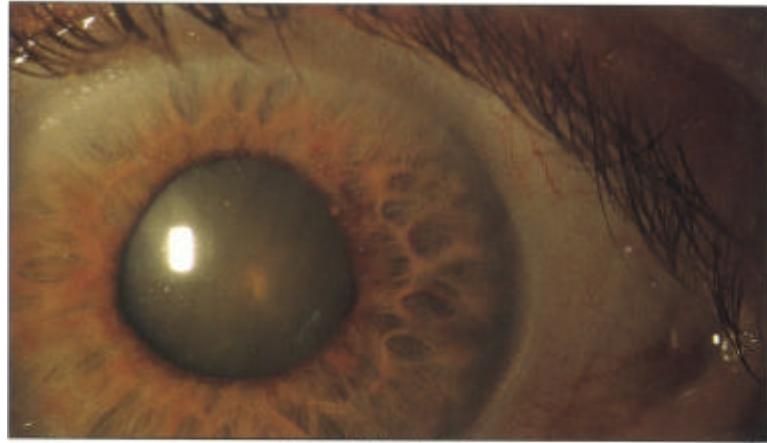


Fig. 8.24 Early and late changes of thrombotic glaucoma. In the early stages neovascularization can be seen around the pupil margin where irregular, mild ectropion uveae is present (left). In a more advanced case gross neovascularization is present, the cornea is hazy from oedema and there is a marked aqueous flare (right). The eye was blind and painful. Pain in these eyes is usually due to inflammation rather than the high IOP and the eye can be kept comfortable, and enucleation prevented, with topical atropine and steroids.

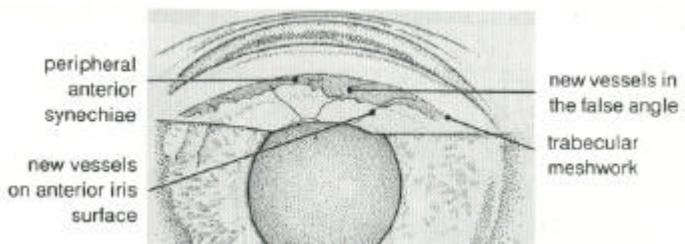
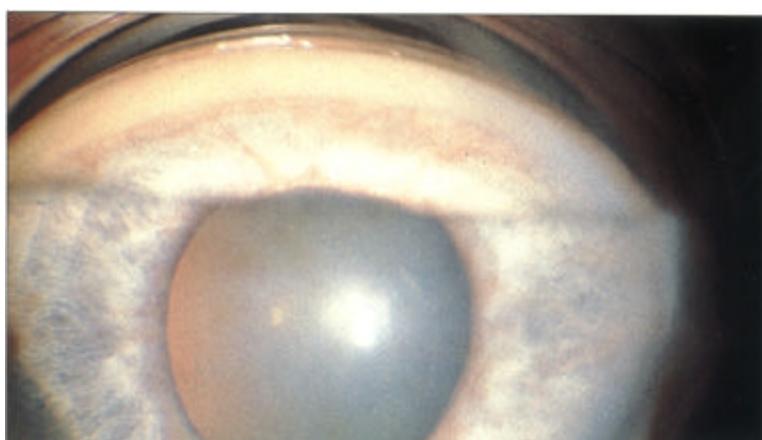


Fig. 8.25 This goniophotograph shows new vessels on the anterior iris surface and in the 'false' angle of a patient in which the angle has been closed by peripheral anterior synechiae formation.

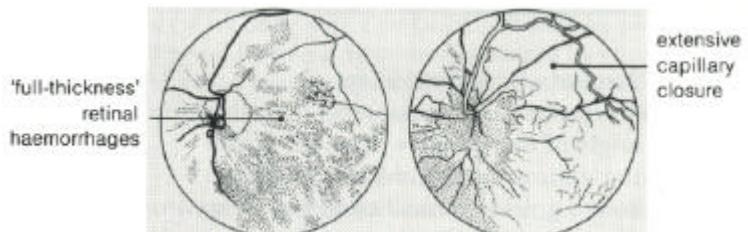
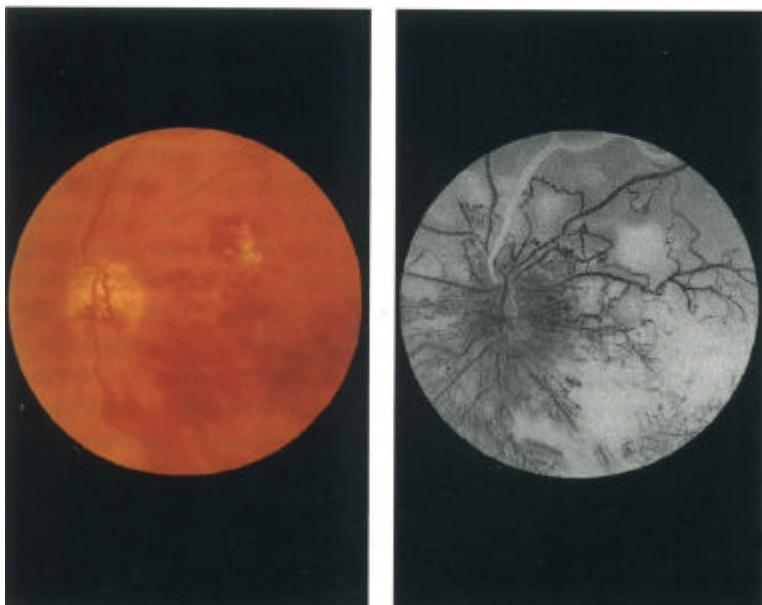


Fig. 8.26 Following a central retinal vein occlusion, this fundus shows resolving retinal haemorrhages and disc swelling. The extensive full-thickness retinal haemorrhage indicates the likelihood of massive capillary closure and non-perfusion of the retina. This is confirmed by the fluorescein angiogram, which demonstrates closure of the normal retinal capillary bed with leakage from the retinal vessels in the hypoxic retina. Such an eye has a substantial risk of developing rubeosis within three months of the initial venous occlusion. Peripheral panretinal ablation at the stage of early rubeosis with laser or cryotherapy can prevent progression to neovascular glaucoma. Conventional drainage surgery is usually unsuccessful in these eyes because of fibrosis in the drainage bleb.

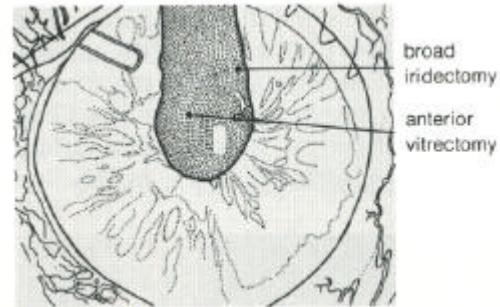
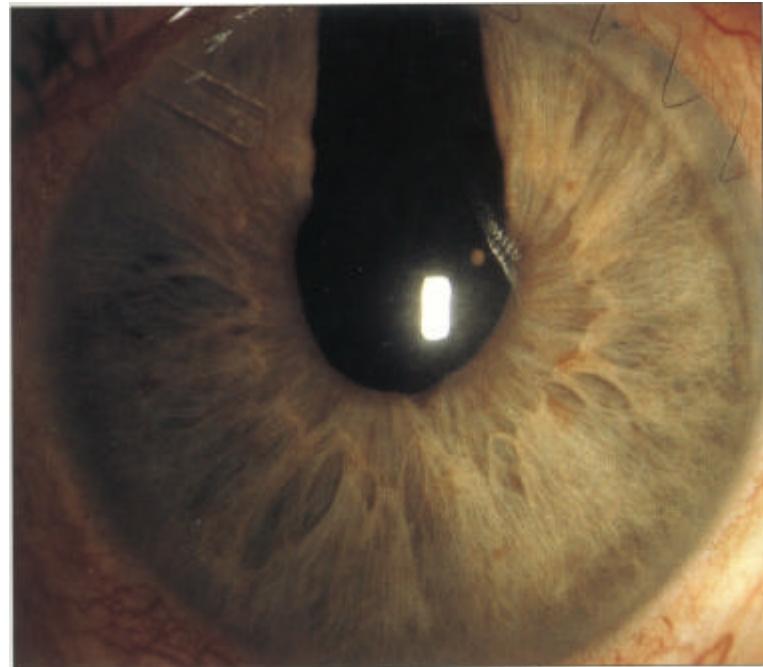


Fig. 8.27 Some eyes with useful vision can be salvaged by using a silicone drainage tube device. Alternatively mitomycin C used at surgery or 5-fluorouracil given subconjunctively after surgery can be helpful in reducing fibrosis of the drainage bleb. This eye had multiple surgical procedures for postoperative endophthalmitis and glaucoma, and was eventually successfully controlled by a tube implant device.

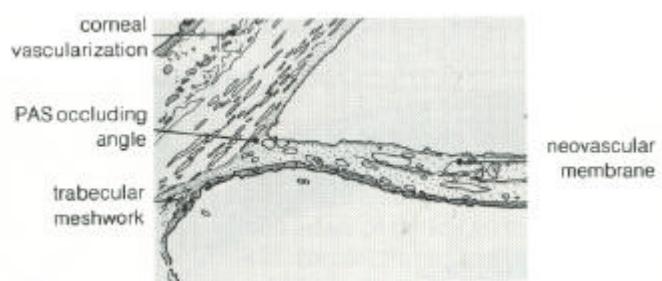
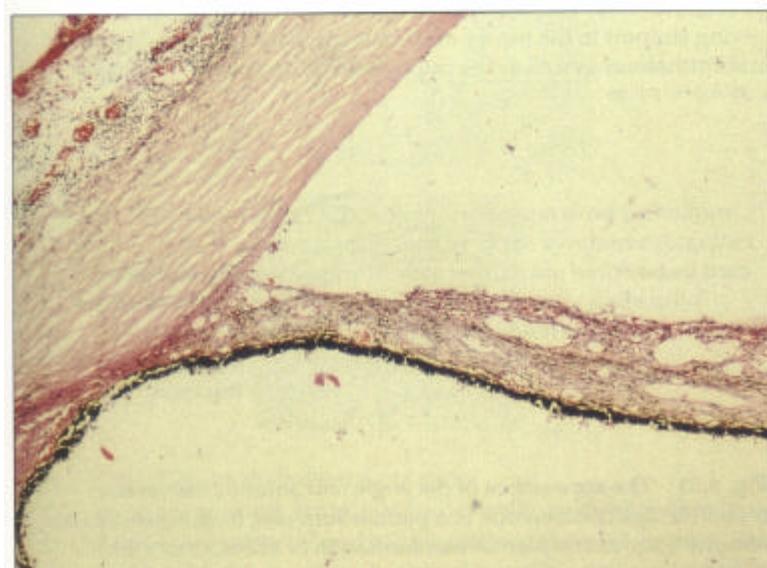
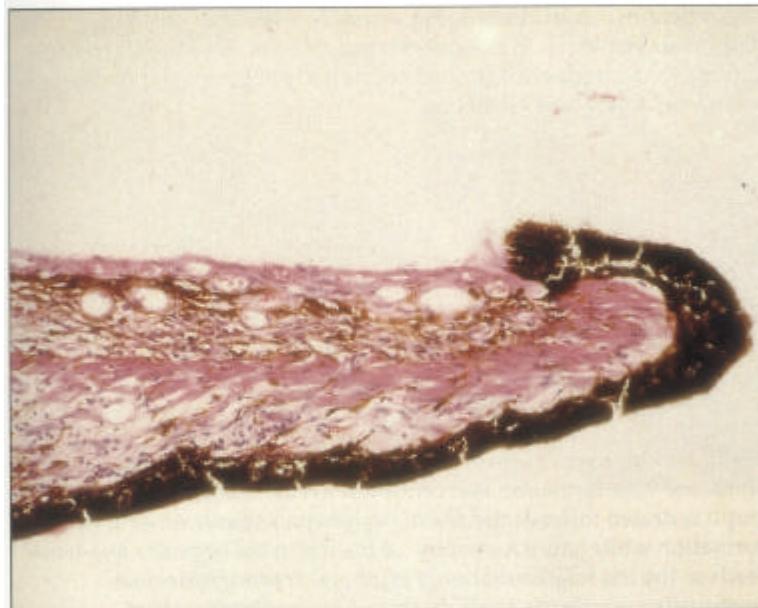


Fig. 8.28 Histology of the iris in neovascular glaucoma demonstrates a fibrovascular membrane on the anterior iris surface (top). Following occlusion of the angle, endothelial slide occurs from the corneal surface over the false angle formed on the iris surface creating a new basement membrane across the angle (bottom).

The iridocorneal endothelial (ICE) syndromes

Essential iris atrophy, Chandler's syndrome, and the irisnaevus (Cogان-Reese) syndrome all result from a primary disorder of the corneal endothelium, and may, in fact, be different parts of the same disease process. The basic defect appears to be a cellular proliferation of the corneal endothelium with formation of Descemet's membrane spreading across the angle and onto the anterior iris surface (see Chapter 6).

Chandler's syndrome is considered a variant of essential iris atrophy. It is unilateral, occurs mainly in females, and

is characterized by corneal endothelial cell decompensation, which causes early presentation with haloes. The glaucoma produced is often quite mild: peripheral anterior synechiae and corectopia (eccentric pupil) are not major features. The iris-naevus syndrome is also unilateral with heterochromia. Iris nodules, which are really foci of remaining normal iris stroma surrounded by endothelial cells, are seen on the anterior iris surface. This endothelial traction also produces ectropion uveae and peripheral anterior synechiae.

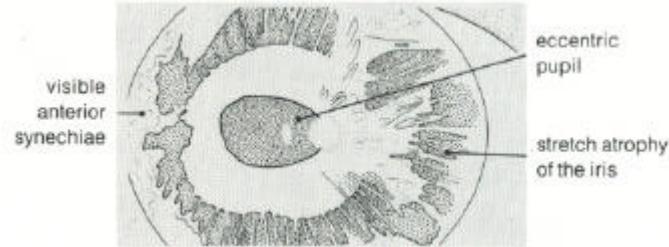


Fig. 8.29 Essential iris atrophy is a rare syndrome which is characterized by unilateral glaucoma, corectopia, pseudopolyopia, and peripheral anterior synechiae formation. The corneal endothelium has an abnormal appearance with large pleomorphic cells (see Chapter 6). In the early stages, there is thinning of the iris stroma in the midperiphery and eccentricity of the pupil progressing to full-thickness hole formation.

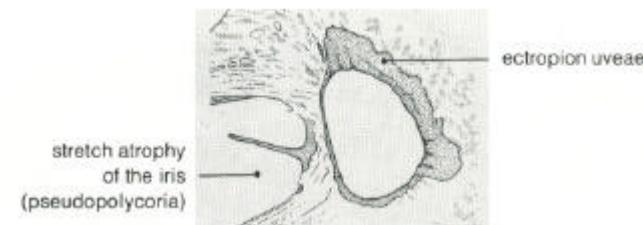


Fig. 8.30 In severe cases of essential iris atrophy, there is full-thickness hole formation and ectropion uveae. Characteristically, the pupil is drawn towards the site of peripheral anterior synechiae formation while 'stretch atrophy' of the iris in the opposite quadrants leads to the iris hole formation. Peripheral anterior synechiae eventually encircle the angle. In this photograph notice that pseudopolyopia and ectropion uveae lie in opposite quadrants, giving support to the theory of iris stretch 'pulling' the pigmented iris epithelium as well as the pupil towards one part of the angle.

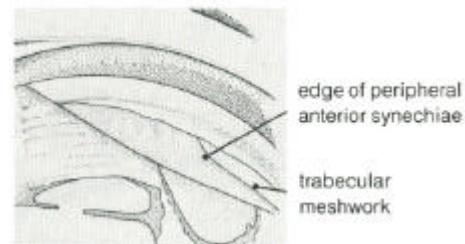


Fig. 8.31 The appearance of the angle and anterior iris surface under the light microscope in a patient suffering from essential iris atrophy shows ectropion uveae, formation of a Descemet's-like membrane on the anterior iris surface, and peripheral anterior synechiae.

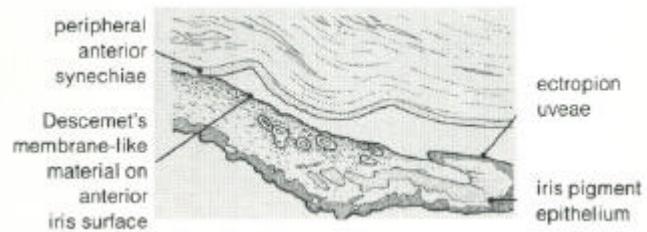
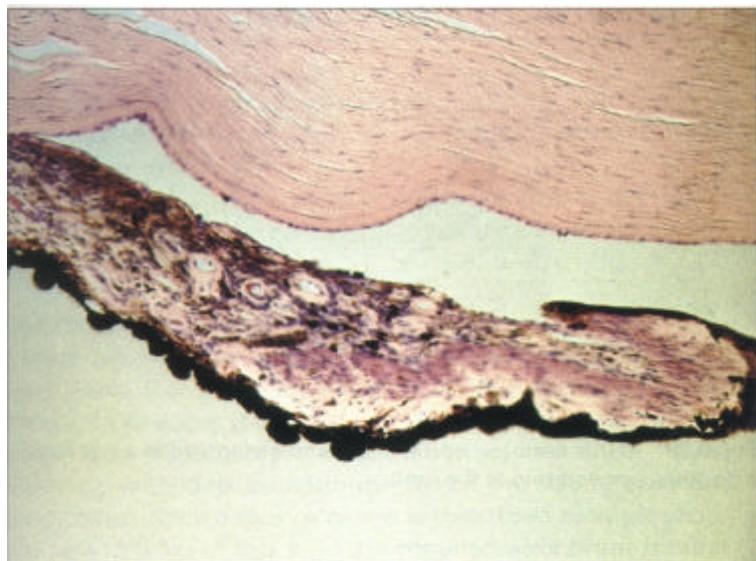


Fig. 8.32 A goniophotograph of a patient with essential iris atrophy illustrates the formation of the peripheral anterior synechiae. The glaucoma is usually out of all proportion to the extent of peripheral anterior synechiae formation and probably reflects endothelial cells covering the trabecular meshwork in those areas where the angle still appears to be open.

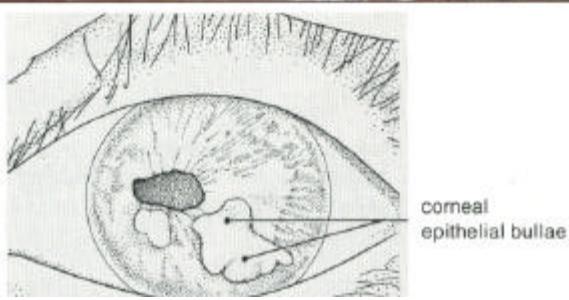


Fig. 8.33 This patient with Chandler's syndrome shows a bullous keratopathy (which is a prominent feature of the syndrome) together with corectopia and iris atrophy (both of which are less marked than in essential iris atrophy). The glaucoma is frequently quite mild. Peripheral anterior synechiae tend to be less extensive than in essential iris atrophy.

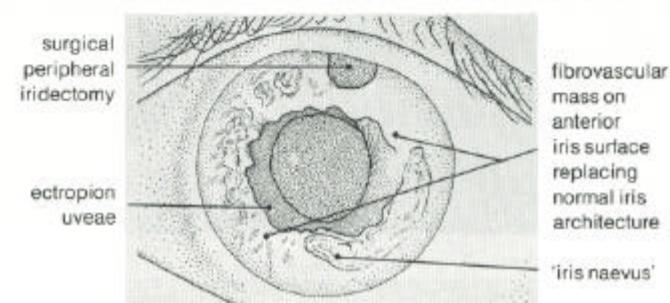
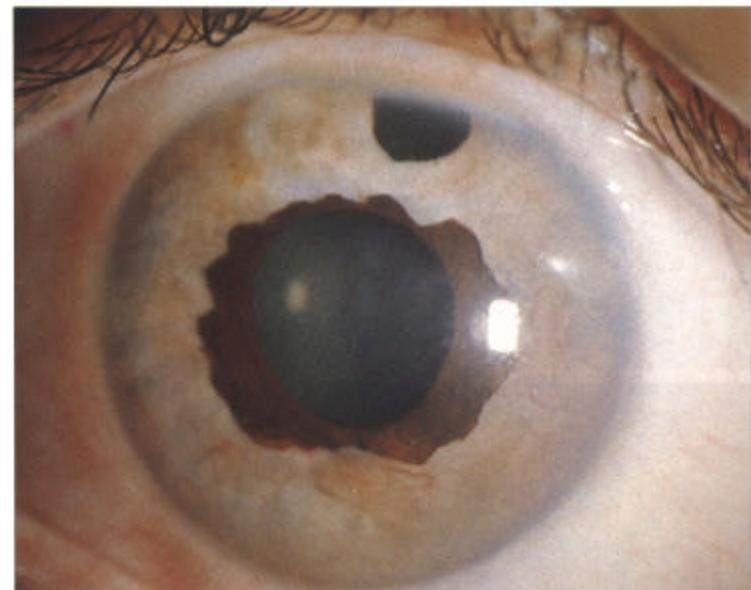


Fig. 8.34 In the iris-naevus syndrome, iris atrophy is less visible than in the typical case of essential iris atrophy while there is more corneal oedema. The anterior surface of the iris is covered by a sheet of Descemet's membrane-like material through which normal nodules of iris tissue protrude. These have been mistaken for iris melanomas. Ectropion uveae is common. In this photograph, note the ectropion uveae together with loss of the usual appearance of the anterior iris.

Epithelialization of the anterior chamber

Glaucoma may result from epithelial cells gaining entry into the anterior chamber. Conditions within the eye needed for epithelial ingrowth are a poorly closed anterior segment wound, or implantation of epithelial cell nests into the iris stroma

from injury or surgery. Should these conditions occur, proliferation of epithelial cells either as a sheet within the chamber or as a slowly growing iris cyst can occur. Either can cause angle closure and secondary glaucoma.

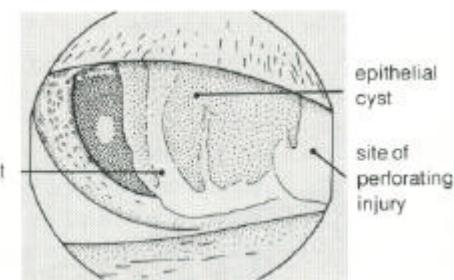
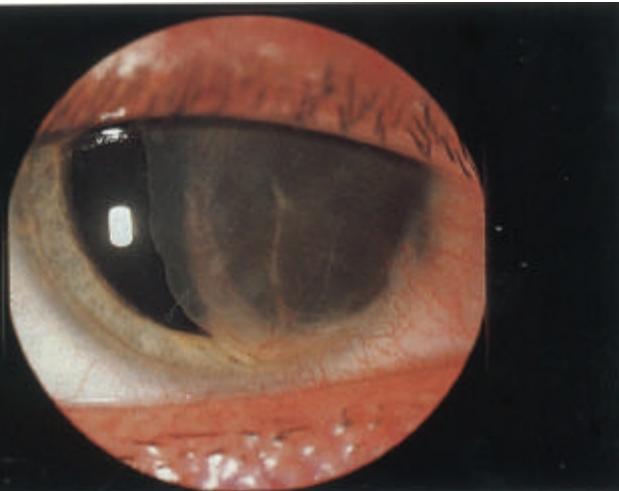


Fig. 8.35 In this example, epithelialization has spread as a cyst from a traumatic perforation at the limbus.

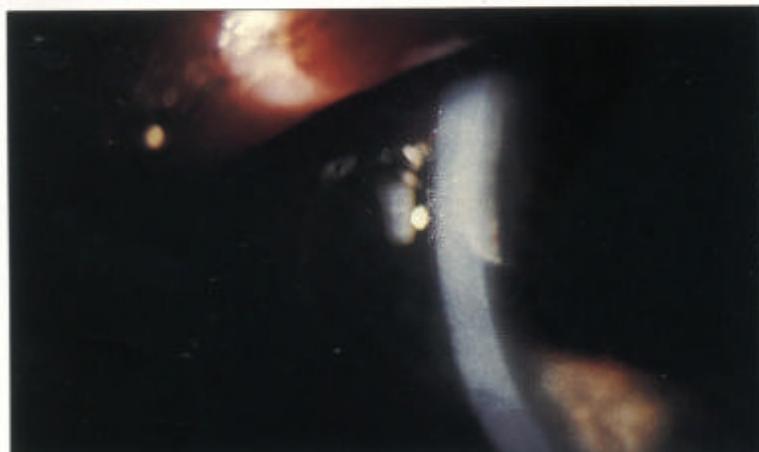


Fig. 8.36 Epithelium can be seen growing as a membrane from a corneal wound in this eye. Notice the corneal oedema and injection from raised IOP. The slit photograph shows the membrane on the corneal endothelium.

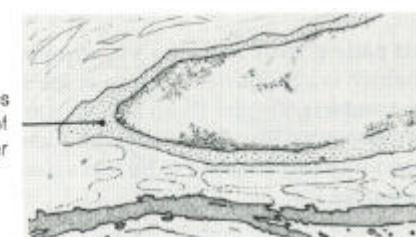
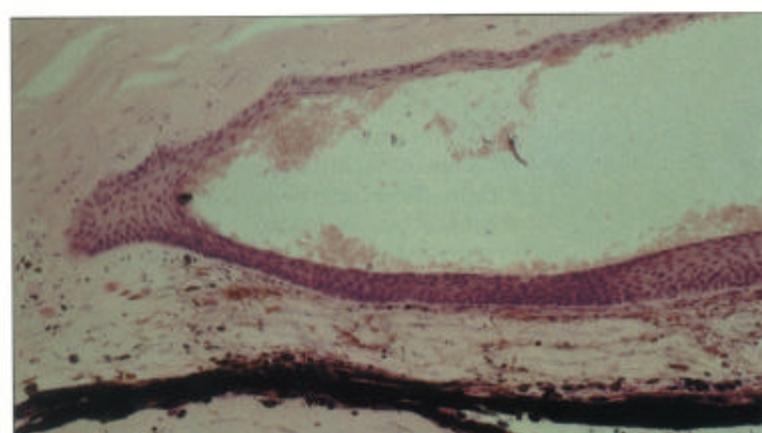


Fig. 8.37 Glaucoma results from occlusion of the angle by direct spread of epithelial cells over the inner surface of the trabecular meshwork. Treatment involves total removal of the epithelial cells, although in practice this can be difficult to achieve.

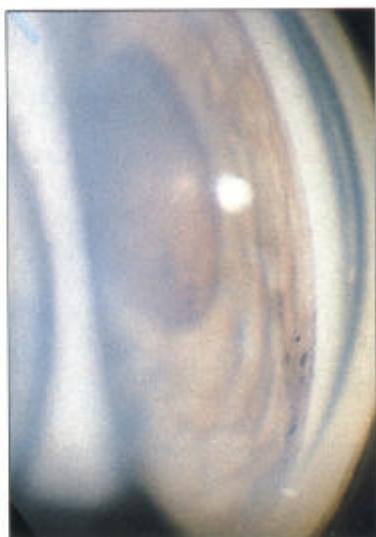
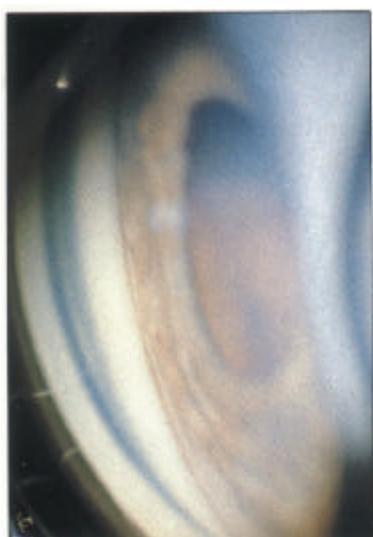
TRABECULAR OUTFLOW OBSTRUCTION: SECONDARY OPEN ANGLE GLAUCOMA

Secondary open angle glaucoma may occur if:

- (i) the meshwork has been disrupted by injury (traumatic angle recession);
- (ii) the meshwork becomes occluded by cells or debris;
- (iii) the meshwork is involved with an inflammatory process.

TRAUMATIC ANGLE RECESSION

A blunt injury to the eye compresses the globe and produces a shock wave. This shock wave is transmitted posteriorly through the eye. In so doing it can disinsert the ciliary muscle from the scleral spur. Gonioscopic examination of the angle shows widening with cleft formation between the scleral spur and ciliary band. Such a cleft or recess is described as angle recession and injuries of this kind are common after blunt trauma. Recession may be the only visible sign of a previous contusion



injury or may co-exist with other signs of ocular injury. Within the anterior segment these include iris sphincter rupture, lens subluxation or dislocation and, within the posterior segment, include commotio retinae, choroidal rupture and retinal dialysis.

Glaucoma occurring after a contusion injury may be immediate or late. Early onset glaucoma is usually related to concurrent uveitis or hyphema. The effect of hyphae are particularly severe in patients with the sickle cell trait. Acidosis within the anterior chamber causes sickling of the red blood cells which obstruct the meshwork and take longer to absorb. This can cause very high IOP levels. Extensive angle recession may be followed years later by the development of glaucoma. Eyes sustaining a marked disruption of the anterior segment at the time of initial injury have a significant risk of developing secondary open angle glaucoma years after the event and this has important medico-legal implications.



Fig. 8.38 Gonioscopy in a patient with angle recession can show a widening of the ciliary band (iris root) or, on occasion following a complete tear of the ciliary muscle, widening of the angle together with a white strip of exposed sclera. These goniophotographs show the temporal aspect of the anterior chamber in both the normal and involved eye. An artificially wide angle with pigmentation can be seen at the recessed site in the affected eye.

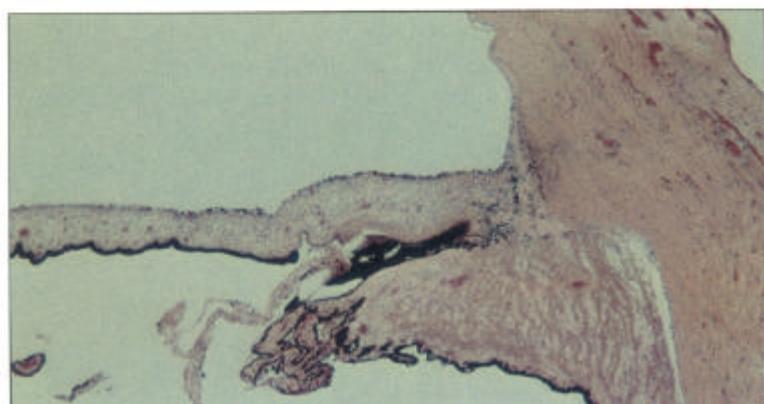


Fig. 8.39 The light microscopic appearance shows recession of the angle with inflammation of the sclera and pigmentation of the trabecular meshwork. Pigment granules are also seen on the anterior iris surface.

TRABECULAR OCCLUSION BY CELLS OR DEBRIS Pigment dispersion glaucoma

Pigment dispersion syndrome (PDS) is a descriptive term for the deposition of pigment granules derived from the pigment epithelium of the iris onto the structures within the anterior chamber. Thus they may be found on the anterior iris surface,

the corneal endothelium (Krukenberg's spindle) or the trabecular meshwork (as a midtrabecular band of pigment); less frequently pigment is seen on the lens equator and zonules following mydriasis. The pigment release occurs from abrasion of the posterior iris surface by zonular fibres and for this to

happen the midperiphery of the iris must be posteriorly concave. This concavity occurs most often in young adult myopic males with a deep anterior chamber. Cellular abrasion is followed by radial slit-like defects in the pigment epithelium visible on iris transillumination. If sufficient pigment is released to compromise outflow facility the IOP rises (PDS with ocular hypertension), and if this persists for long enough then glaucoma develops.

An uncommon form of acquired pigmentary glaucoma has been seen with posterior chamber intraocular lens implants. In these cases the loops rub against the posterior iris surface producing pigment release. Glaucoma is uncommon.

Treatment is conventional, complicated only by the patient's intolerance to the miotic and myopic effects of pilocarpine. In theory pharmacologically induced miosis would halt the condition.

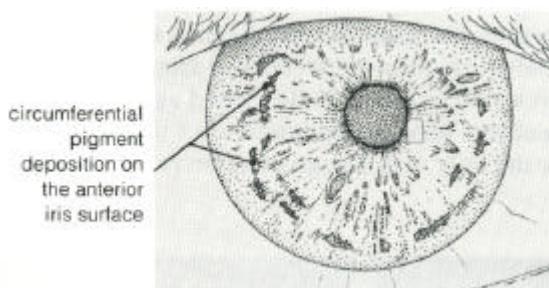
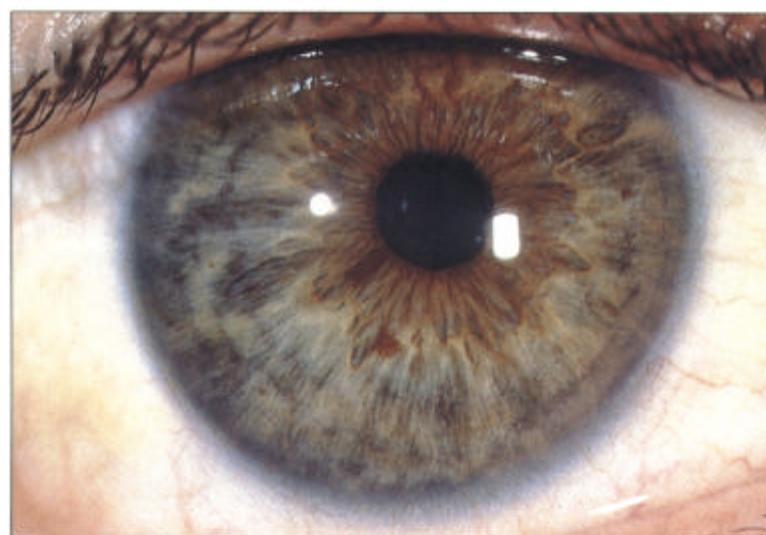


Fig. 8.40 In this patient, extensive pigment deposition is seen on the anterior surface of the iris, rather like dust particles, lying in the wrinkles formed by iris contraction.

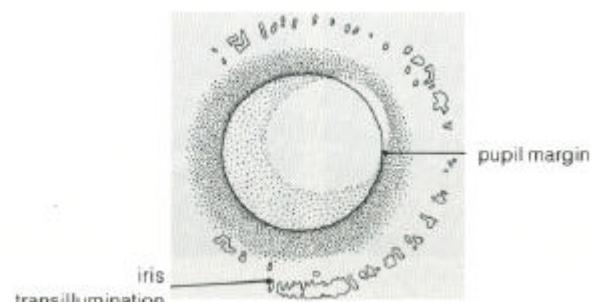
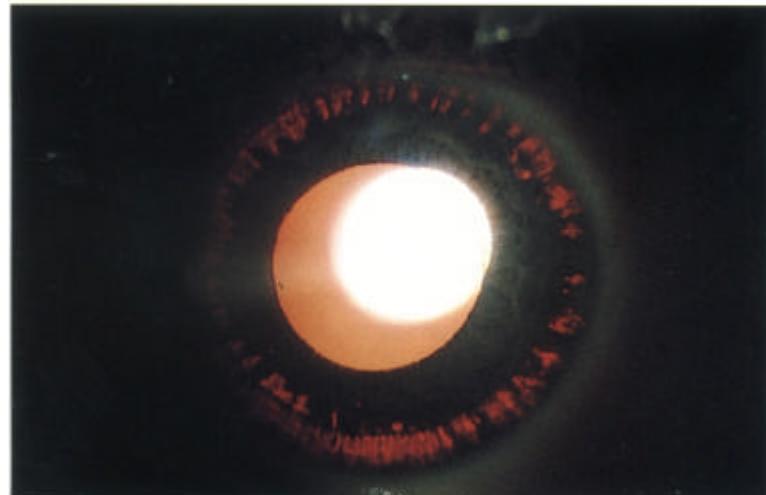


Fig. 8.41 Retroillumination of the iris of the same patient shows multiple slit-like defects in the pigment epithelium of the peripheral iris.

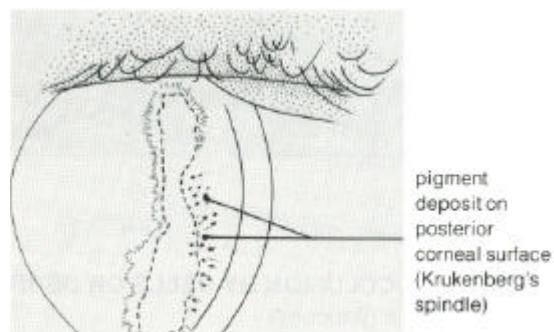


Fig. 8.42 Krukenberg's spindle is the term given to pigment deposition on the posterior corneal surface. The pigment is usually deposited in a vertical band.

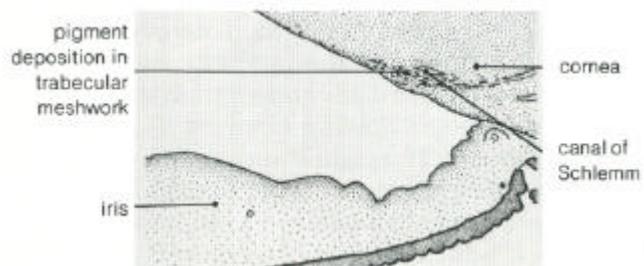
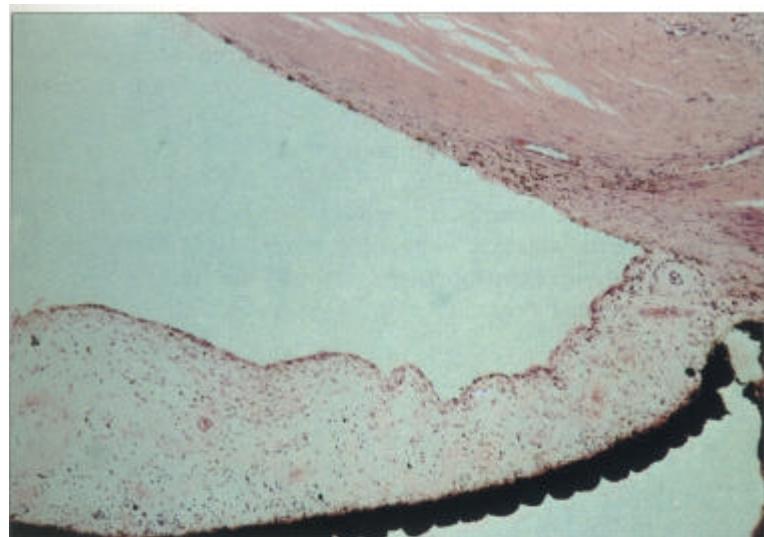


Fig. 8.43 Histological section shows that the angle has extensive pigment accumulation. Pigment granules lie freely between the trabecular plates, are phagocytosed within the endothelium, or in macrophages obliterating the normal pattern and obstructing the outflow. The gonioscopic appearances of the angle in pigmentary glaucoma have been demonstrated earlier.

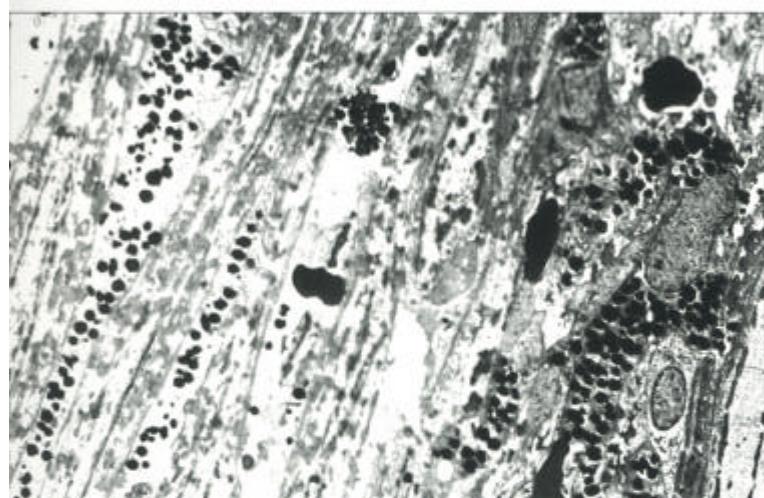


Fig. 8.44 Electron micrograph of the angle demonstrates a characteristic accumulation of intertrabecular and intracellular pigment granules which is not seen in age-matched normal eyes.

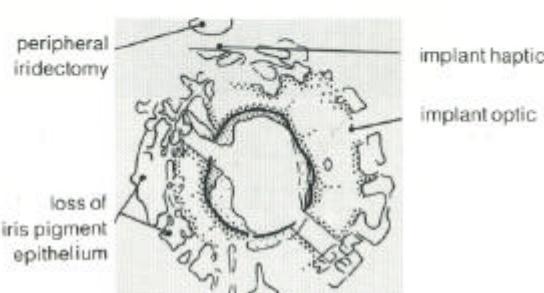
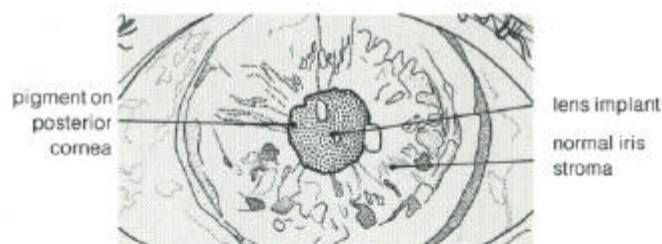


Fig. 8.45 These photographs show extensive loss of iris pigment epithelium from abrasion by a sulcus placed posterior chamber lens implant. The patient presented with blurred vision and a microscopic hyphema from rupture of iris vessels.

Pseudoexfoliation glaucoma

Pseudoexfoliation (PXE) is the descriptive term given to dandruff-like material found on the pupil margin, the anterior lens surface and occasionally the posterior corneal surface. The term 'pseudo' is used to distinguish it from true exfoliation of the lens capsule seen following exposure to infrared light (Glassblowers' cataract). Its origin is unknown but it seems to arise from the anterior lens capsule, the zonules and the inner layer of ciliary epithelium, probably as a result of a disorder of cell metabolism. If sufficient material is produced then outflow

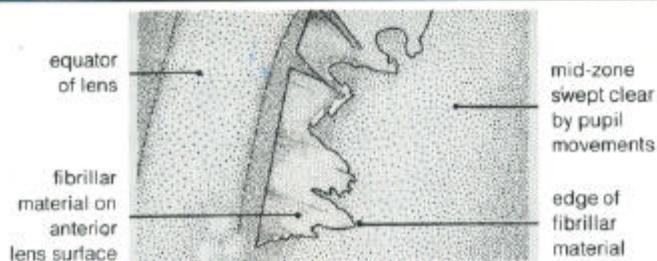
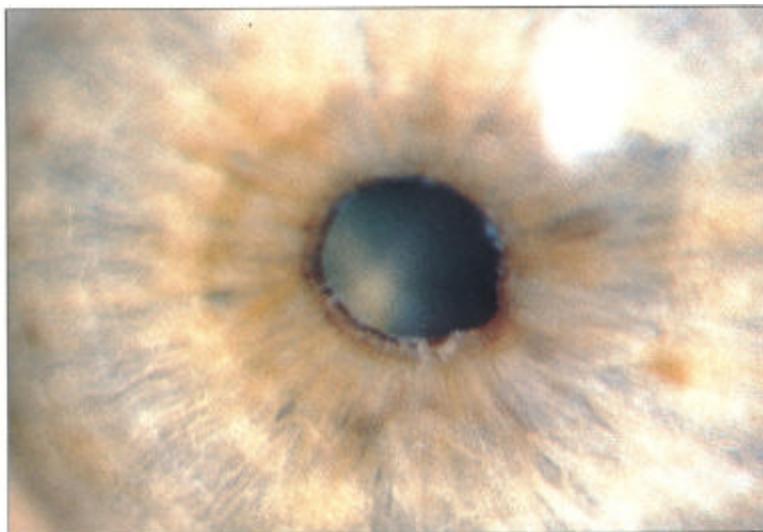


Fig. 8.47 Following mydriasis, the appearance of the abnormal material can be seen on the anterior lens surface.

facility may be compromised and IOP increased. Persistent elevation of IOP leads to glaucoma. The condition is often bilateral, although asymmetric. The response to topical glaucoma medication is often poor.

Patients with PXE are usually elderly and have coexistent cataract. Surgical treatment in these patients can usefully be combined with cataract extraction. Care needs to be taken, however, with an extracapsular extraction because the zonules are weak in these eyes.

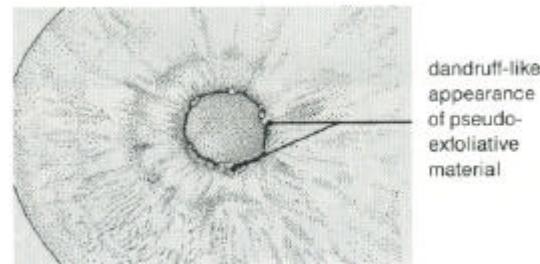


Fig. 8.46 The clinical diagnosis of pseudoexfoliation glaucoma is made by observing the dandruff-like appearance of the fibrillar material on the pupillary margin.

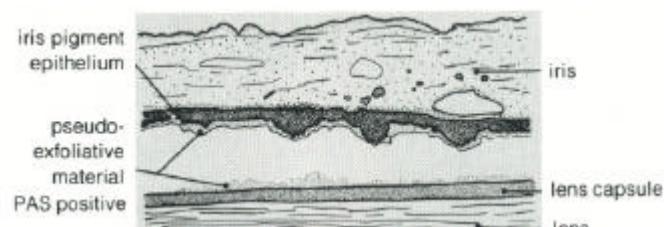
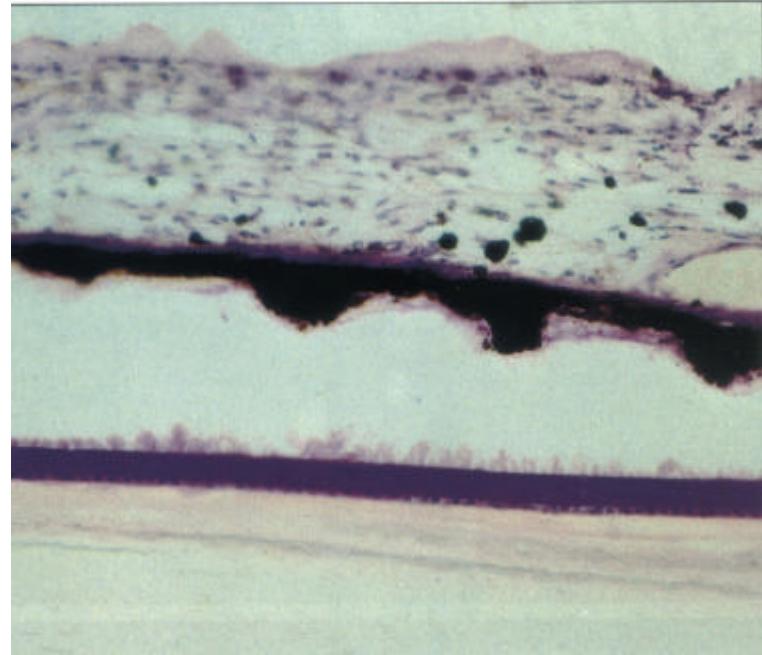


Fig. 8.48 Histology of an eye with pseudoexfoliation glaucoma shows the presence of PAS positive eosinophilic material on the anterior surface of the lens and the posterior surface of the iris.

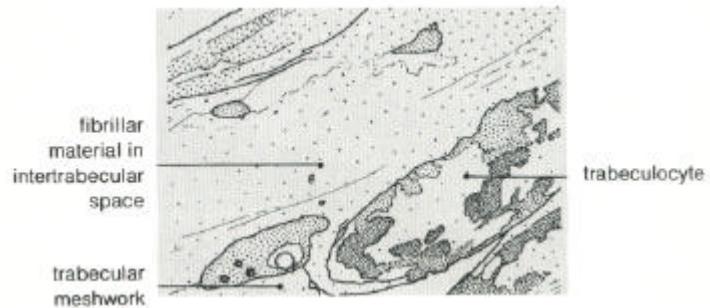
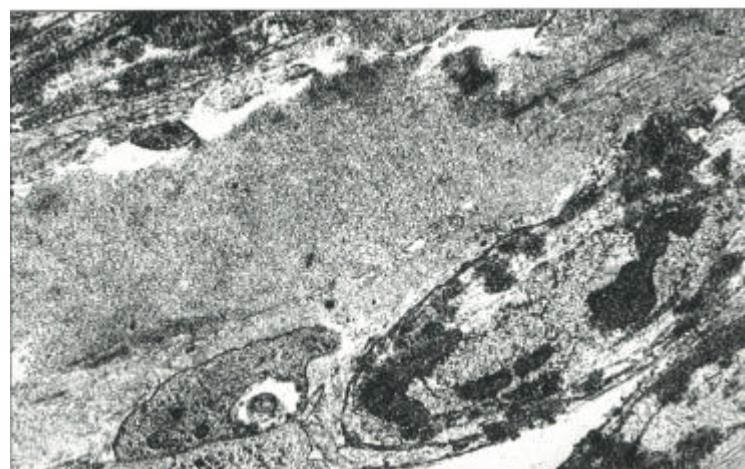
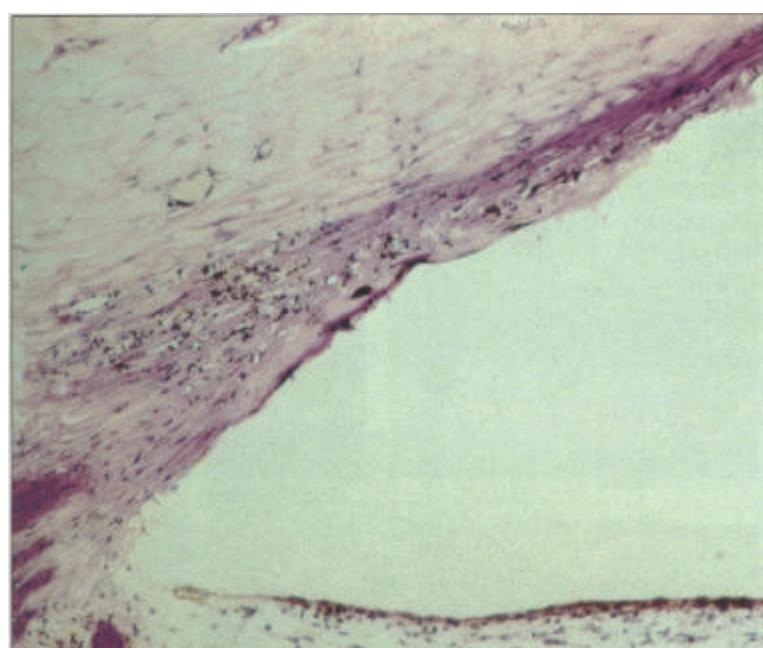


Fig. 8.49 The angle is open in pseudoexfoliation glaucoma but the meshwork contains more pigment than is usual, together with the fibrillar material caught up in the meshwork. This material can also be found histologically around the conjunctival vessels.

Lens induced glaucoma (phacolytic glaucoma)

Two different types of cellular response are seen in eyes with a degenerate lens. Under certain conditions, the lens capsule of a hypermature cataract leaks denatured cortical material. This is particularly common with a Morgagnian cataract and these

cataracts should be removed to forestall this complication (see Chapter 11). Leakage in turn excites a macrophage response and the macrophages, gorged with lens material, accumulate and clog the trabecular meshwork causing a secondary open angle glaucoma known as phacolytic glaucoma.

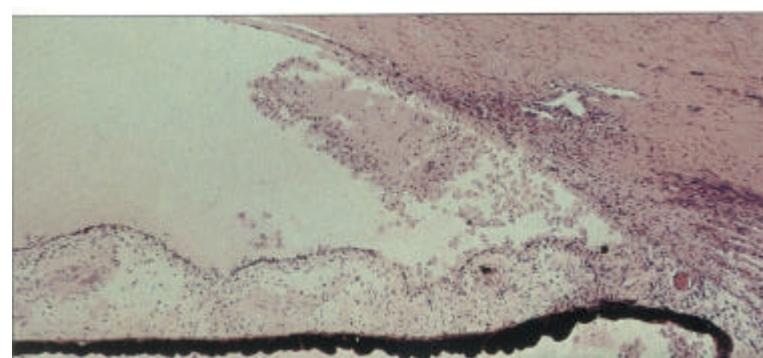


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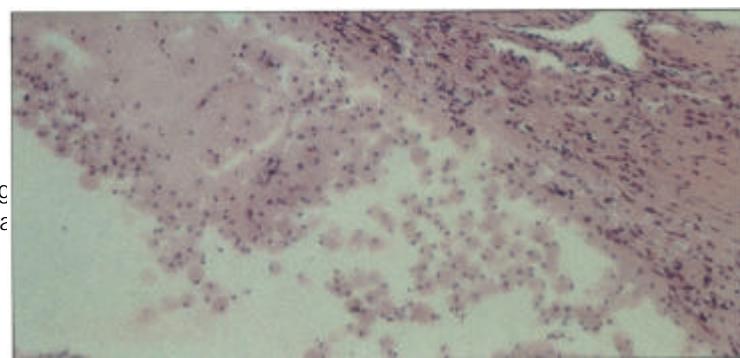
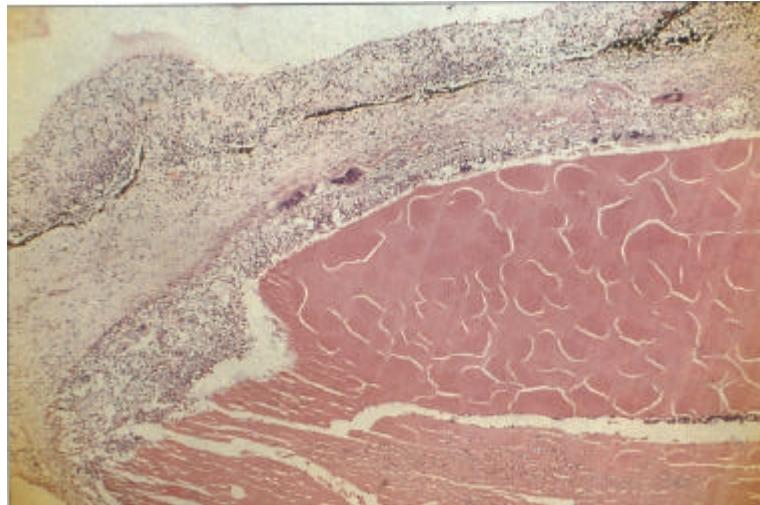


Fig. 8.51 Photomicrographs demonstrate the swollen macrophages obstructing the trabecular meshwork. Clinically large accumulations are seen floating in the anterior chamber of an eye with a cataractous lens and acute glaucoma, an open angle and a deep anterior chamber.

Phacoanaphylactic uveitis

Under normal conditions, lens protein is sequestered during foetal development from the systemic immune system. Sensitization of the immune system by the lens protein may cause phacoanaphylactic endophthalmitis. In this situation, a granulomatous inflammation occurs secondary to rupture of the lens capsule and a chronic inflammatory response ensues, often accompanied by raised intraocular pressure. Clinically this most commonly follows some weeks after extracapsular cataract surgery or a perforating injury. Histological examination



reveals invasion of the lens cortex and uvea by these inflammatory cells with a granulomatous infiltrate. Resolution follows removal of the lens material if that is possible. Phacoanaphylactic uveitis has been recognized as a clinical entity since the turn of the century but the aetiology is undergoing a complete reappraisal since it has recently been shown that many of these eyes have a low grade bacterial infection with propionobacteria or staphylococcus epidermidis and that appropriate antibiotic treatment or debridement of infected tissue leads to resolution.



Fig. 8.52 A massive inflammatory response with extensive fibrosis may be seen between the posterior iris and anterior lens surface. The anterior lens capsule has been ruptured and inflammatory cells are present beneath the capsule adjacent to, and interspersed with, the lamellae of the anterior stroma.

Haemolytic glaucoma

Blood cells and their products of degeneration may cause secondary open angle glaucoma. Under normal conditions, healthy red blood cells pass through the trabecular meshwork to enter Schlemm's canal. A hyphema results when the accumulation of red blood cells occurs at a faster rate than they can be removed by the normal process. If the trabecular meshwork becomes clogged by these cells, intraocular pressure will rise. With a healthy angle, elimination of the hyphema is followed by the return of the intraocular pressure to normal.

Haemolytic glaucoma should be suspected when open angle glaucoma is discovered in an eye with a long-standing vitreous haemorrhage and minimal uveitis with yellowish discolouration within the anterior and posterior chambers, especially if it is aphakic. Phase contrast microscopy may be used to identify these degenerate red blood cells (ghost cells) from fluid removed at the time of an aqueous tap. It is considered that the lack of malleability of ghost cells renders them more difficult to be removed through the meshwork.

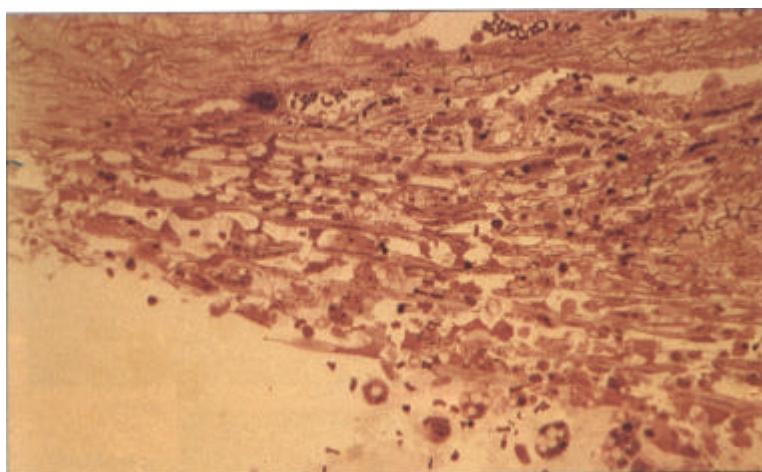


Fig. 8.53 The histological appearance of the trabecular meshwork in haemolytic glaucoma shows degenerate red blood cells (ghost cells) on the trabecular spaces and within macrophages. By courtesy of Dr I Grierson.

Siderosis

Siderosis results from the widespread deposition of iron throughout the eye which diffuses from a retained ferrous ocular foreign body. Glaucoma occurs secondary to sclerosis of the trabecular meshwork, and is thought to result from a

direct toxic effect. Clinically, eyes with siderosis demonstrate mydriasis, heterochromia, retinal degeneration with optic atrophy, as well as raised intraocular pressure. The ERG shows specific changes depending on the degree of retinal damage.

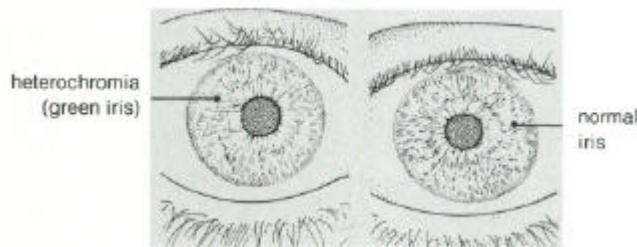
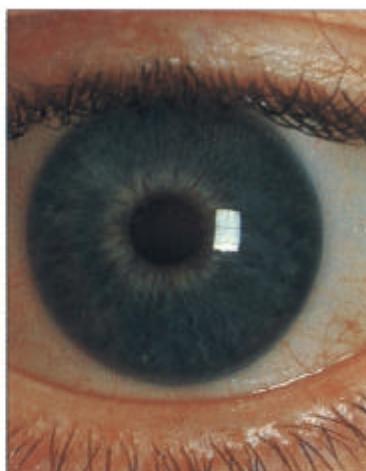
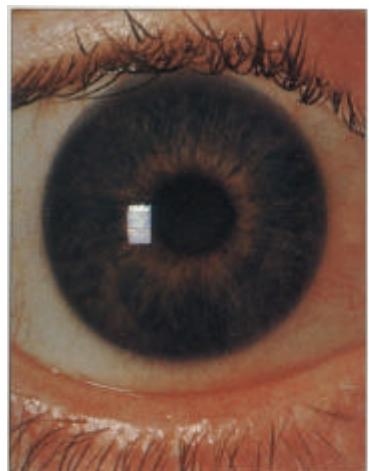


Fig. 8.54 The right eye of this patient shows evidence of siderosis with brown-green discolouration of the iris from iron deposition.

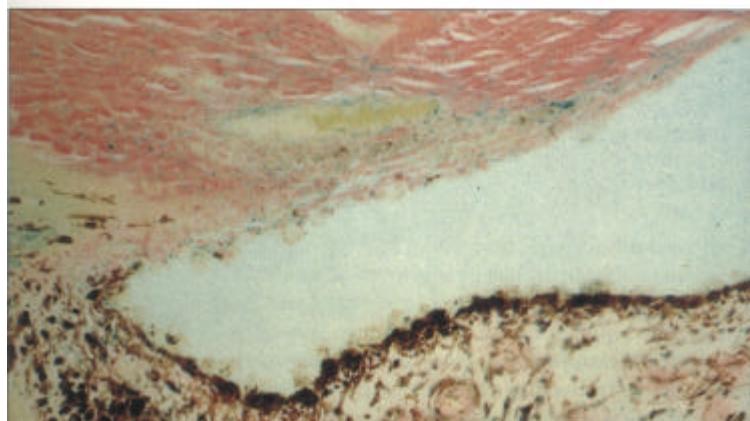


Fig. 8.55 Histology shows iron deposition within the meshwork and Schlemm's canal, associated with loss of normal trabecular architecture. There is reduction in trabeculocytes together with trabecular collapse.

Tumour infiltration

Secondary open angle glaucoma can occasionally occur with tumours of the anterior segment. It may be seen with malignant melanoma cells or other tumour cells which are liberated into the aqueous from the tumour surface and block the angle mechanically by seeding into it. Less commonly, a malignant melanoma arising from the iris root and ciliary body may in

vade the angle directly to produce glaucoma. On rare occasions 'melanomalytic' glaucoma arises secondary to trabecular obstruction from macrophages engorged with melanin from the tumour but pigmentation in the angle in the presence of an iris pigmented melanoma has to be considered to be due to tumour cells in most instances.

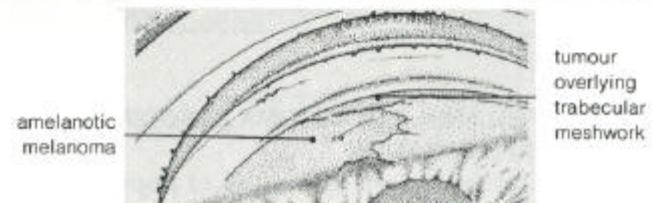
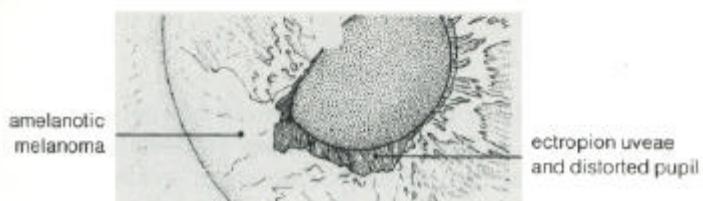
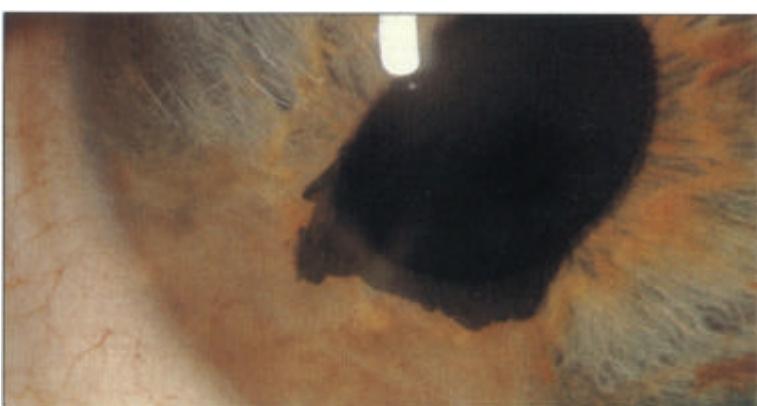


Fig. 8.56 This eye has an amelanotic iris melanoma which, on gonioscopy (bottom) is seen to be infiltrating the angle.

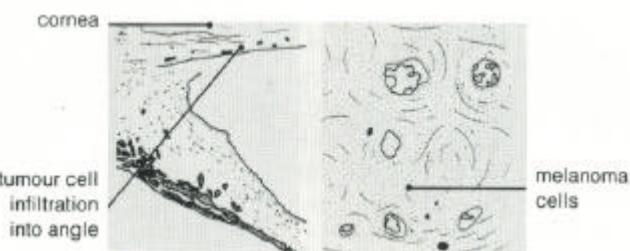
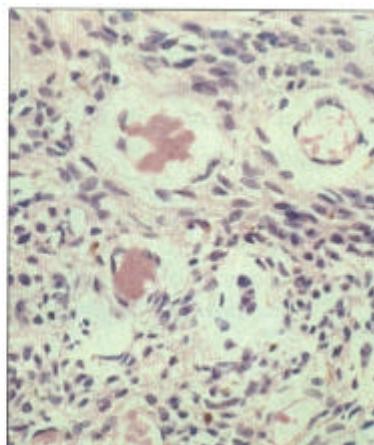
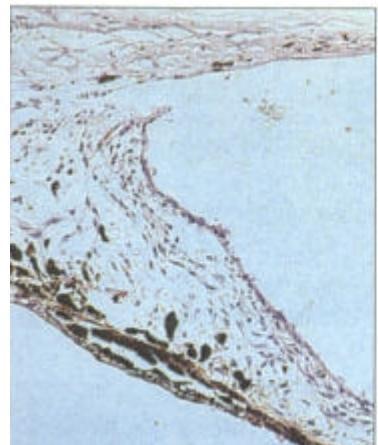


Fig. 8.57 Light microscopy of the angle in a patient with glaucoma secondary to iris melanoma shows that malignant melanoma cells have blocked the angle.

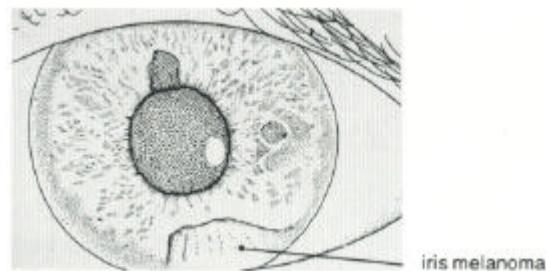
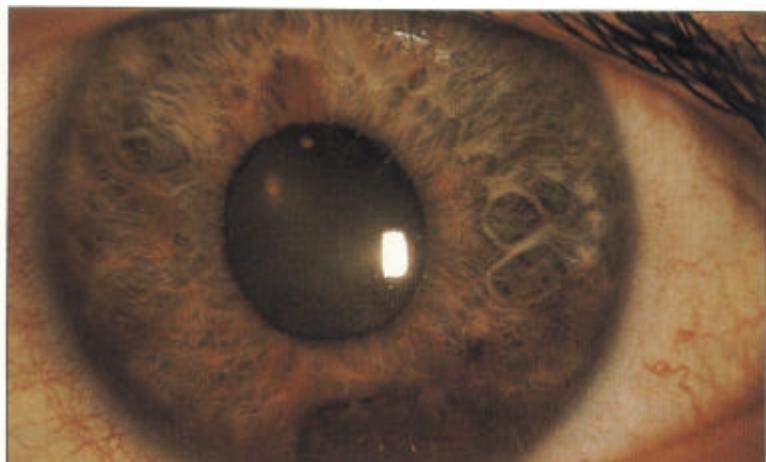


Fig. 8.58 Melanomalytic glaucoma is a very rare condition arising secondary to trabecular obstruction by macrophages engorged with melanin released by a localized tumour. Clinically it is difficult to differentiate from direct tumour invasion and the diagnosis is made pathologically.

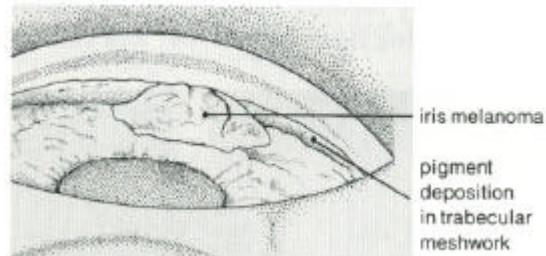
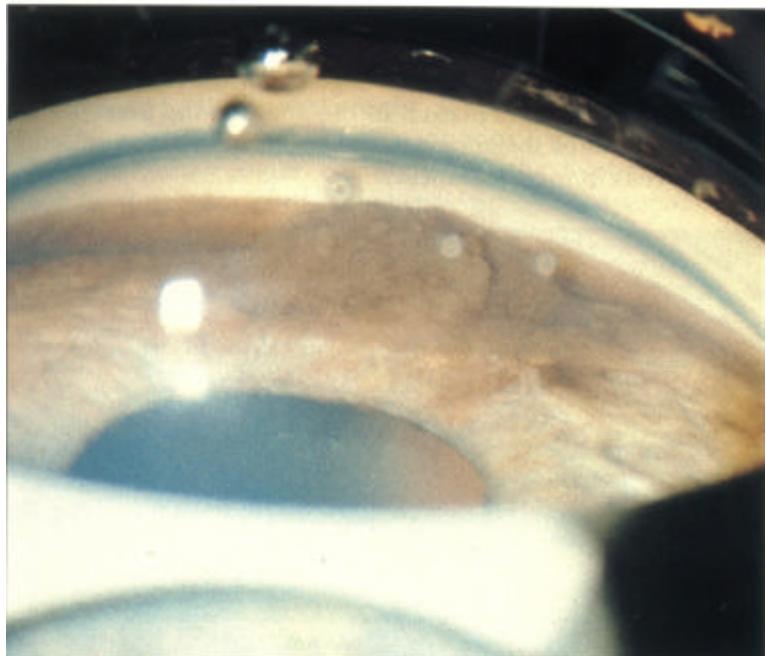


Fig. 8.59 Gonioscopic view of the same patient shows heavy pigmentation and tumour within the angle. Local excision of the tumour in this case was followed by resolution of the coincidental glaucoma.

SECONDARY OPEN ANGLE GLAUCOMA ASSOCIATED WITH OCULAR INFLAMMATION

Uveitis has already been seen as a cause of pupil block (see Figs 8.1 and 8.2). In addition it may cause chronic angle closure glaucoma from PAS. Outflow facility may also be reduced in uveitic eyes from obstruction of the open angle by inflamma

tory cellular debris or direct involvement of the trabecular meshwork in the inflammatory process (trabeculitis). Two specific syndromes, the Posner-Schlossman syndrome and Fuchs' heterochromic cyclitis, are also particularly associated with open angle glaucoma. Finally prolonged treatment with topical steroids may induce an open angle glaucoma.

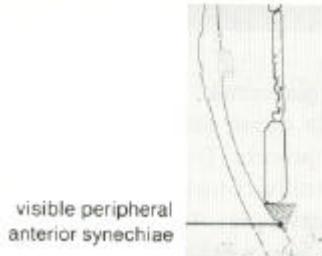
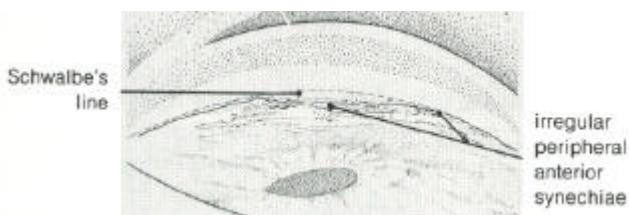
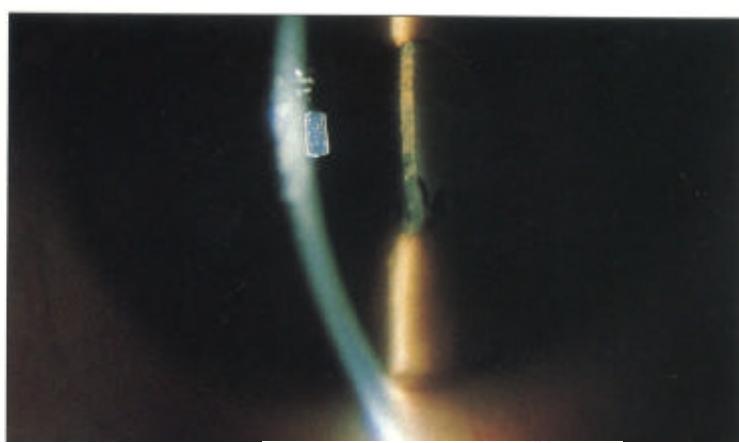
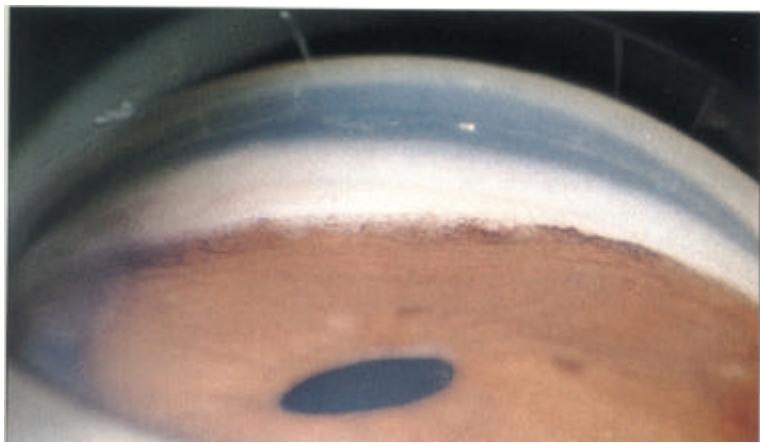


Fig. 8.60 Slit-image and gonioscopic photographs demonstrate the development of peripheral anterior synechiae in chronic uveitis. Although usually confined to the inferior angle, such synechiae can extend circumferentially. With sarcoidosis trabecular granulomas form as focal lesions around the circumference of the angle and, if untreated, these granulomas produce small areas of peripheral anterior synechiae (see Chapter 10).

The Posner-Schlossman syndrome (glaucomatocyclitic crisis) This unusual condition is typically seen in young adult males who develop very high levels of IOP with minimal anterior uveitis. The syndrome is usually uniocular but either eye can be involved in the attacks. Patients present with symptoms from corneal oedema due to intraocular pressures in the 40–60 mmHg range. The uveitis is minimal, posterior synechiae do not form, and optic nerve damage does not usually occur. Occasionally, following many attacks, chronic elevation of IOP

occurs; it is then thought that permanent damage to the meshwork has occurred either directly from the disease process or secondary to raised IOP causing degenerative changes in the meshwork. Whatever the mechanism, a condition clinically indistinguishable from primary open angle glaucoma develops. Typically acute attacks last days to weeks. In between such attacks, the occasional 'sentinel' keratic precipitates (KP) can be found.

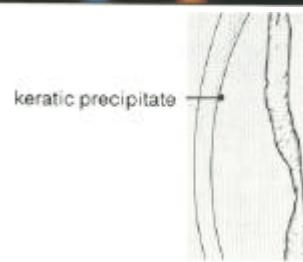
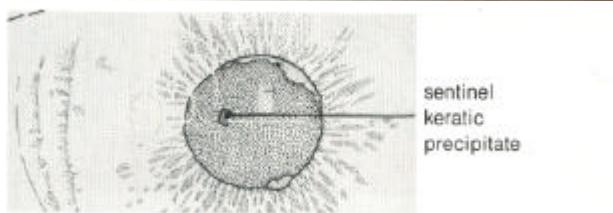
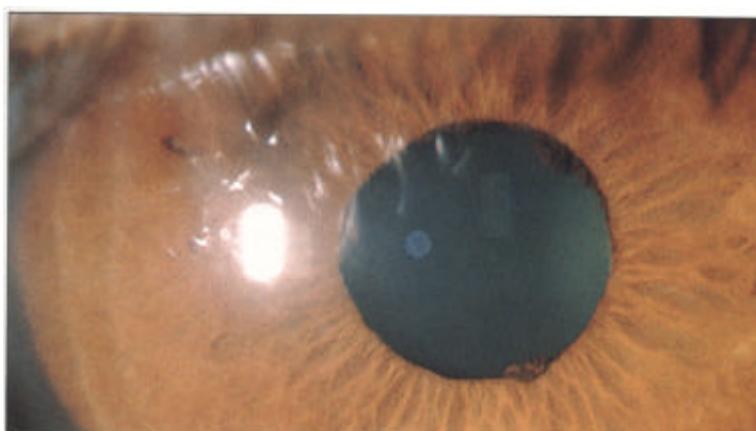


Fig. 8.61 The 'sentinel' keratic precipitates seen during the quiescent phase. There is some evidence of crises being associated with high levels of aqueous humour prostaglandins.

Fuchs' heterochromic cyclitis

About 70% of cases of Fuchs' heterochromic cyclitis are unilateral. Iris atrophy (the affected iris showing anterior stromal atrophy and a lighter colour), fine widespread KP, cataract, glaucoma, and occasionally a fine neovascularization of the angle are all seen in an established case (see Chapter 10). Neither anterior nor posterior synechiae develop. The glaucoma responds to conventional medical or surgical treatment and the eye responds well to cataract surgery should it be indicated.



Fig. 8.62 A patient with heterochromic cyclitis of the right eye. The iris is lighter than the left and a cataract is present.

Steroid induced glaucoma

Topical steroids cause raised intraocular pressure in a small proportion of patients. There is some evidence that this population is genetically controlled and may be similar to those at risk from primary open angle glaucoma. The glaucoma usually appears after a few weeks of treatment and this complication must be taken into consideration with any patient on topical steroid therapy. Different steroid preparations vary in their

ability to produce this phenomenon but this may be related to their potency and penetration of the cornea. The mechanism is uncertain. In some cases the inter trabecular spaces have been found blocked by a fibrillar material of unknown aetiology. The fact that in most cases intraocular pressure falls to normal on cessation of the topical steroid suggests that the drug plays an active part in the cellular pathology.

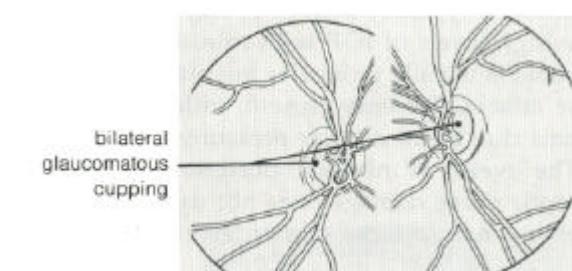
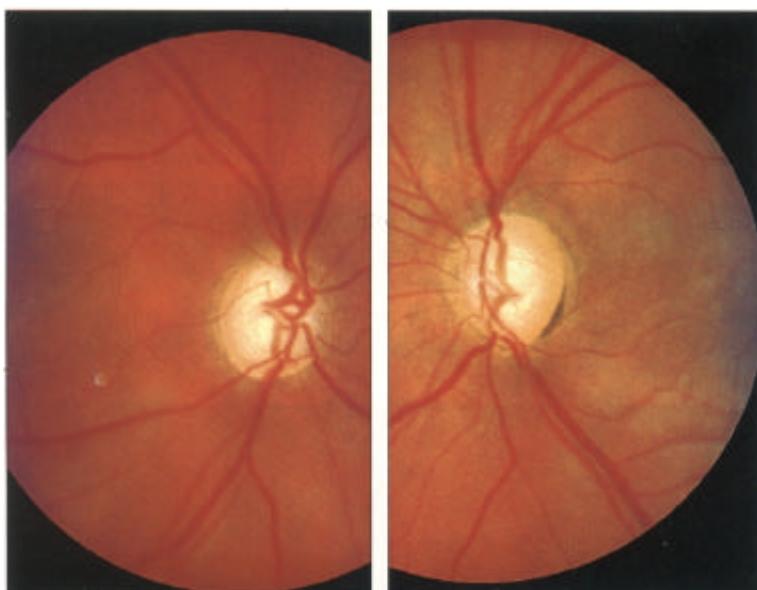


Fig. 8.63 Gross glaucomatous cupping is seen in this patient who had used topical steroids without supervision for treatment of mild ocular irritation associated with contact lens wear.

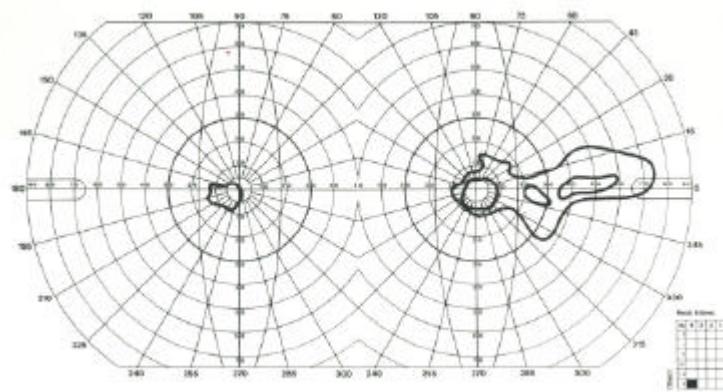


Fig. 8.64 The visual fields of the same patient show the gross field loss which caused her to present. Visual acuities were 20/30 in each eye.

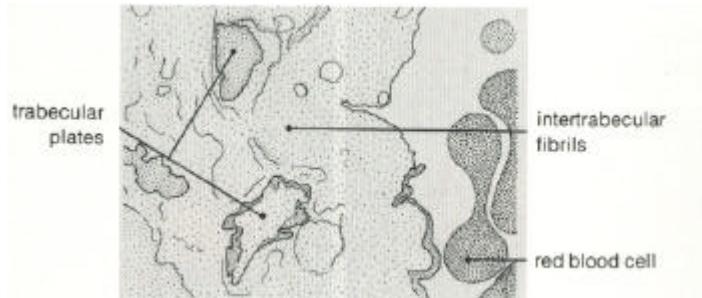
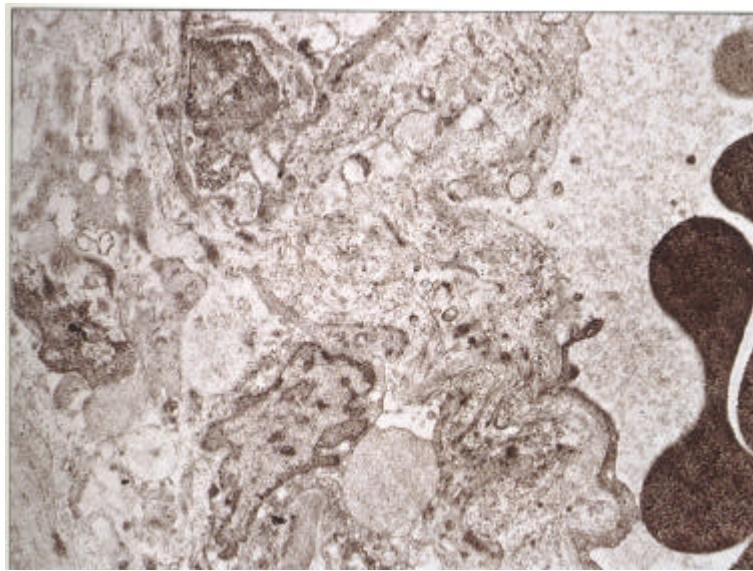


Fig. 8.65 Electron micrograph of the trabecular meshwork in the patient whose optic discs were shown in Fig. 8.62. Fibrillar material occludes the intertrabecular spaces and it is speculated that this may produce glaucoma.

POST-TRABECULAR OUTFLOW OBSTRUCTION: RAISED EPISCLERAL VENOUS PRESSURE

Raised episcleral venous pressure is usually caused by a shunting of arterial blood to the orbital veins by a caroticocavernous fistula, but is occasionally seen with gross cor pulmonale, the Sturge-Weber syndrome or superior vena cava obstruction. Any increase in episcleral venous pressure will cause an increase in intraocular pressure so as to maintain the pressure gradient across the trabecular meshwork.

With fistulae between the carotid artery and cavernous sinus where the shunt is usually of high flow, the diagnosis is obvious from the dramatic neuro-ophthalmic signs (see Chapter 20). However, coexistent ocular hypoxia complicates this picture and even in the presence of rubeosis iridis (which is not infrequently present in these eyes) the intraocular pressure may be low. In contrast arteriovenous communications within the dural vessels are frequently of a low flow type and these patients present with red eyes, arterialized conjunctival

vessels, and glaucoma without the other overt signs of bruits or proptosis. Apart from the glaucoma, these dural shunts usually have a benign prognosis and may resolve spontaneously. The glaucoma may vary in severity: those cases with high intraocular pressure respond poorly to medical treatment. The Sturge-Weber syndrome is the commonest cause to require surgery. This may be in infancy - associated with buphthalmos, in childhood or in early adult life. Despite the youth of these patients fistulizing surgery is quite successful. Preoperatively a coexistent choroidal haemangioma should be sought. If this is found or suspected, surgery should be performed under hypotensive anaesthesia to minimize intraoperative choroidal haemorrhage. The dissection of the scleral flap may be prejudiced by pressure of abnormal scleral vessels and meticulous haemostasis is required.

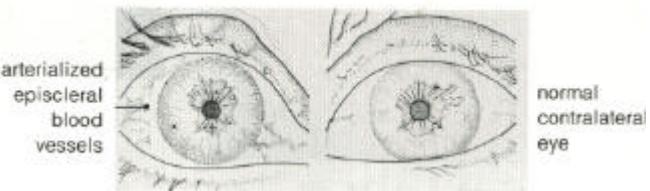


Fig. 8.66 Arterialized vessels are seen in the conjunctiva of a patient with a dural arteriovenous fistula.

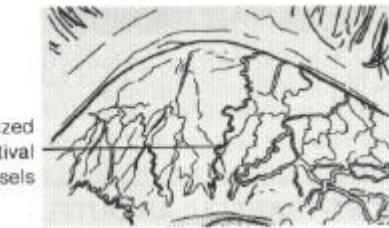
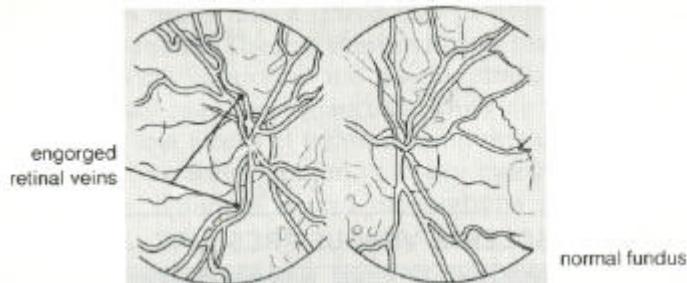
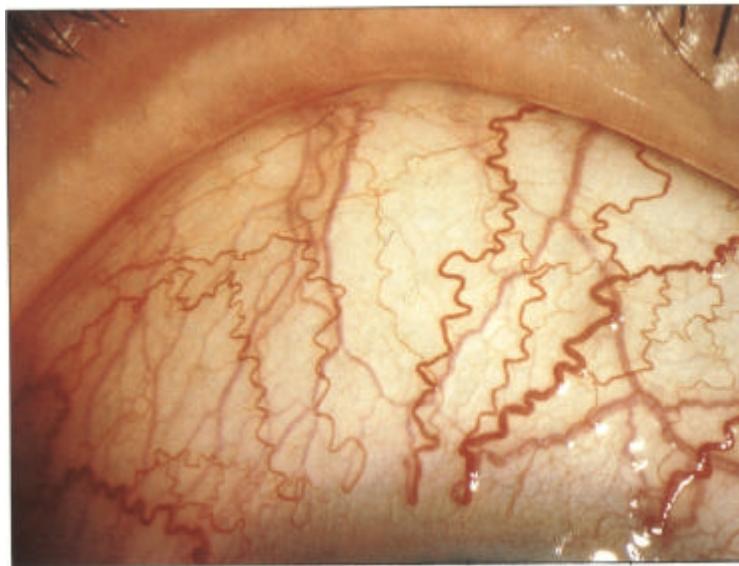


Fig. 8.67 Retinal veins are engorged in the right eye but at this stage the optic discs do not show glaucomatous changes.

STURGE-WEBER SYNDROME

Glaucoma occurs in about 30% of patients with the Sturge-Weber syndrome and is said to be more common if the upper lid is involved. Most cases present with buphthalmos within

the first two years of life but some patients may not present until young adulthood. Unilateral or marked asymmetrical ocular involvement is typical.



Fig. 8.69 Neonate with marked facial angioma involving the upper eyelid and maxilla and extending to the midline. This child later suffered epilepsy from an intracranial angioma. Both eyes were buphthalmic but the right was worse.

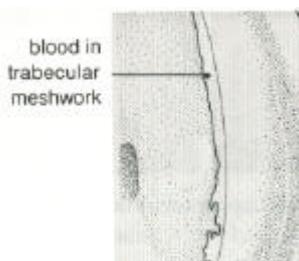
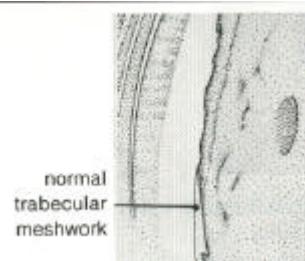


Fig. 8.70 These goniophotographs compare the normal and abnormal eyes of a patient with the Sturge-Weber syndrome. Blood can be seen within Schlemm's canal of the affected eye and this is a common finding in the presence of raised episcleral venous pressure.

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