Chapter 5

Disorders of the Lens

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CAUSES OF ACQUIRED CATARACTS

Age related cataracts

1. Subcapsular cataract
   (a) Anterior subcapsular cataract (Figure 5.1) lies directly under the lens capsule and is associated with fibrous metaplasia of the anterior epithelium of the lens.

   Figure 5.1 Anterior subcapsular cataract

   (b) Posterior subcapsular cataract (Figure 5.2) lies just in front of the posterior capsule and is associated with posterior migration of the epithelial cells of the lens. Patients with this type of opacity are particularly troubled by headlights of oncoming cars and bright sunlight. Their near vision is also frequently diminished more than their distance vision.

   Figure 5.2 Posterior subcapsular cataract

2. Nuclear cataract (Figure 5.3) starts as an exaggeration of the normal ageing change involving the lens nucleus. It is often associated with myopia resulting from an increase in the refractive index of the lens nucleus and also with increased spherical aberration. Some elderly patients with nuclear cataracts may be able to read again without spectacles, because of induced myopia ('second sight of the aged').

   Figure 5.3 Nuclear cataract

3. Cortical cataract involves the anterior, posterior or equatorial cortex. The opacities start as vacuoles and clefts between the lens fibres (Figure 5.4). Subsequent opacification leads to the formation of the typical radial spoke-like opacities (Figure 5.5).

4. Christmas tree cataract (Figure 5.6) is uncommon. It is characterized by striking, polychromatic, needle-like deposits in the deep cortex and nucleus.

All of these types of cataract can progress to maturity so that the entire lens becomes opaque (Figure 5.7). A hypermature cataract is one in which leakage of water has resulted in shrinkage of the cataract and wrinkling of the anterior capsule (Figure 5.8). A morgagnian cataract is a
hypermature cataract in which total liquefaction of the cortex has allowed the nucleus to sink inferiorly (Figure 5.9).

Presenile cataracts

Presenile cataracts may be associated with the following systemic diseases:

1. **Diabetes** may result in osmotic overhydration of the lens and the development of bilateral white punctate or snowflake posterior or anterior opacities (Figure 5.10). In certain cases the cataract may mature in a few days.

2. **Myotonic dystrophy** is initially associated with fine polychromatic granules followed later by stellate posterior subcapsular opacities (Figure 5.11). Cataracts develop in 90% of patients, usually after the age of 20 years, but do not interfere with vision until the age of 40. The systemic and other ocular features of myotonic dystrophy are described in Chapter 15.

3. **Atopic dermatitis** is associated with two types of cataract:
(b) **Posterior subcapsular cataract**, which resembles a complicated cataract (Figure 5.13). About 10% of patients with severe atopic dermatitis develop cataracts between the ages of 15 and 30 years. Both eyes are affected in about 70% of cases and frequently the opacity becomes mature (Figure 5.14).

4. **Neurofibromatosis type 2** is associated with presenile posterior subcapsular or posterior cortical cataracts. The systemic and other ocular features are described in Chapter 15.

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**Figure 5.10** Diabetic cataract

**Figure 5.11** Mild stellate posterior subcapsular cataract in myotonic dystrophy

(a) **Shield-cataract**, consisting of a dense anterior subcapsular plaque which wrinkles the anterior capsule (Figure 5.12).

**Figure 5.12** Shield-like anterior subcapsular cataract in atopic dermatitis

**Figure 5.13** Posterior subcapsular cataract in atopic dermatitis

**Figure 5.14** Mature atopic cataract – note the skin changes of atopic dermatitis

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**Traumatic cataracts**

Trauma is the most common cause of unilateral cataract in young individuals. Lens opacities may be caused by the following types of injury:

1. **Direct penetrating injury** to the lens (Figure 5.15).
2. **Concussion**, which may lead to 'Vossius' ring, resulting from an 'imprinting' of iris pigment onto the anterior lens capsule (Figure 5.16), as well as flower-shaped opacities (Figure 5.17).
3. **Electric shock** and lightning, which are rare causes.
4. **Ionizing irradiation** to ocular tumours.
Toxic cataracts

1. Steroids, both systemic and topical, are cataractogenic.

(b) The exact relationship among the total dose, weekly dose and duration of administration of systemic steroids and cataract formation is unclear, although it is thought that patients receiving <10 mg prednisone equivalent per day or those treated for 4 years are probably immune. Although it is generally believed that children are more susceptible than adults to the cataractogenic effects of systemic steroids, there may also be an individual (genetic) susceptibility in some individuals in whom lens changes can develop after short-term therapy. There are also other individuals who do not develop cataracts following prolonged steroid administration.

(c) On the basis of these findings, it has been suggested that the concept of a 'safe' dose should be abandoned. Patients in whom lens changes develop should have their therapy reduced to the minimum consistent with the control of the disease and should, if possible, be considered for alternate-day therapy; this is because lens changes occur less frequently in patients receiving intermittent therapy. Regression of early opacities may occur when the drug is stopped or reduced, although progression may occur despite withdrawal.

2. Chlorpromazine may cause the deposition of fine, yellowish-brown granules on the anterior lens capsule (Figure 5.19). The opacities are located within the pupillary area but they are rarely dense enough to interfere with vision. The lens deposits are dose-related and may be associated with diffuse, granular
deposits on the corneal endothelial and deep stroma. Chlorpromazine may rarely also cause retinotoxicity (see Chapter 10).

3. **Miotics**, particularly long-acting cholinesterase inhibitors, if used long term, may cause tiny anterior subcapsular vacuoles and, occasionally, more advanced opacities. Cessation of medication may stop, retard or occasionally reverse their progression.

4. **Busulphan** (Myleran), a drug used in the treatment of chronic myeloid leukaemia, may very occasionally cause lens opacities.

5. **Amiodarone** is a drug used to treat a variety of cardiac arrhythmias. Visually inconsequential anterior subcapsular lens deposits occur in about 50% of patients using moderate-to-high doses. Cornea verticillata may also occur.

6. **Gold** is used occasionally to treat patients with rheumatoid arthritis. Approximately 50% of patients who have received treatment for 3 years or more develop innocuous anterior capsular lens deposits.

**Secondary cataracts**

A secondary (complicated) cataract develops as a result of some other ocular disease.

1. **Chronic anterior uveitis** is the most common cause of secondary cataract.
   (a) The earliest finding is a polychromatic lustre at the posterior pole of the lens. If the uveitis is controlled, the progression of cataract may be arrested.
   (b) If the inflammation persists, anterior (Figure 5.20) and posterior subcapsular opacities (Figure 5.21) develop and then the lens may become completely opaque. The lens opacification seems to progress more rapidly in the presence of posterior synechiae (Figure 5.22).
2. Acute congestive angle-closure glaucoma is associated with the subsequent formation of glaukomflecken consisting of small, grey-white, anterior, subcapsular or capsular opacities in the pupillary zone (Figure 5.23).

3. High myopia is frequently associated with secondary posterior lens opacities as well as early development of nuclear sclerosis. Simple myopia does not, however, predispose to cataracts, although a myopic change in refraction frequently precedes the development of true nuclear sclerotic cataract.

4. Hereditary fundus dystrophies such as retinitis pigmentosa, Leber congenital amaurosis, gyrate atrophy, Wagner and Stickler syndromes may be associated with posterior subcapsular lens opacities. Cataract surgery may occasionally improve visual acuity even in the presence of severe retinal changes.

**Figure 5.23** Anterior capsular opacity following acute angle-closure glaucoma (glaukomflecken)

## MANAGEMENT OF AGE-RELATED CATARACTS

### Introduction

**GENERAL INDICATIONS FOR CATARACT SURGERY**

1. **Visual improvement** is by far the most common indication for cataract extraction, although requirements vary from person to person. For example, a librarian with a posterior subcapsular cataract may need surgery, if vision falls to N6 or less when reading in bright light, even though distance vision is still 6/12. On the other hand, a farmer with nuclear cataracts which reduce visual acuity to 6/18 may need surgery although he can still read N6.

2. **Medical indications** are those in which the presence of a cataract is adversely affecting the health of the eye, for example, phacolytic glaucoma or secondary angle closure by an intumescent lens, and diabetic retinopathy, treatment of which is hampered by a cataract.

3. **Cosmetic indications** are those in which a mature cataract in an otherwise blind eye is removed to restore a black pupil.

### OPTIMAL POSTOPERATIVE REFRACTION

The optimal postoperative refraction differs according to whether the patient requires a monocular or binocular correction.

1. **If monocular correction** is required in a patient whose fellow eye has poor visual acuity from a dense cataract or amblyopia, the best postoperative refraction is about –1D. This will enable the patient to carry out most ordinary tasks without spectacles and wear bifocals when finer visual acuity is required. Some of the most dissatisfied patients are low myopes who have been turned into hypermetropes after IOL implantation.

2. **If binocular correction** is required the difference in refraction between the two eyes should not be more than 3D. This is because, although with spectacles the patient will have excellent visual acuity in both eyes when looking straight ahead, there will be double vision on looking up or down because of induced vertical prism differences between the two eyes. If a patient has good visual acuity in the unoperated eye, the postoperative refraction in the operated eye ideally should be within 1–2D of the prescription in the unoperated eye. For example, if a patient has +4D in the unoperated eye, a postoperative refraction of –1D would produce a +5D difference between the two eyes and be unacceptable.

### Surgical techniques

The two most frequently used techniques for removal of age-related cataract are: (a) **large incision extracapsular extraction** and (b) **phacoemulsification**. Only the most important steps will be described.

#### LARGE INCISION EXTRACAPSULAR CATARACT EXTRACTION

1. A vertical groove is made in peripheral clear cornea. The cystitome is introduced into the anterior chamber and multiple small radial cuts are made in the anterior capsule for 360° (Figure 5.24a). An alternative method of performing the capsulotomy is by capsulorhexis which involves making a controlled circular tear in the capsule.
2. The full-thickness incision is completed with scissors (Figure 5.24b).
3. The nucleus is expressed by alternating pressure from above and below (Figure 5.24c).
4. The tip of the infusion–aspiration cannula is introduced into the anterior chamber and passed under the iris at 6 o'clock. Strands of cortex are engaged into the port by activating the suction mechanism (Figure 5.24d). The cortex is then dragged centrally and aspirated under direct visualization. This manoeuvre is repeated sequentially until all cortex has been removed. It is important not to aspirate the posterior capsule accidentally because this may cause it to rupture, making implantation of a posterior chamber implant (PC-IOL) difficult or impossible. A sign of imminent rupture is the appearance of fine sharp lines radiating from the aspiration port (Figure 5.24e).
5. If necessary, the posterior capsule can be polished to remove any small residual subcapsular plaques (Figure 5.24f).
6. Viscoelastic substance is injected into the capsular bag to facilitate subsequent insertion of the IOL (Figure 5.25a).
7. The IOL is grasped by the optic and its anterior surface coated with viscoelastic substance (Figure 5.25b).
8. The inferior haptic is inserted through the lips of the incision and then passed under the iris at 6 o'clock (Figure 5.25c).
9. The tip of the superior haptic is grasped with forceps and advanced into the anterior chamber. As the superior pole of the haptic is clearing the edge of the pupil (Figure 5.25d), the arm is pronated to ensure that on release of the haptic it will spring open under the iris and not out of the incision. Preferably, both haptics should be placed into the capsular bag (Figure 5.25e, bottom) and not into the ciliary sulcus (Figure 5.25e, top).
10. The IOL is dialled into the horizontal position by engaging the guide holes with a special hook (Figure 5.25f).
11. The pupil is constricted by injecting Miocoll (acetylcholine) into the anterior chamber, viscoelastic substance is aspirated, and the incision is closed.
PHACOEMULSIFICATION

The technique is constantly changing and there are many variations in the technique. The basic steps are as follows:

1. A continuous capsulorhexis is performed (Figure 5.26a).
2. Hydrodissection is performed by injecting fluid between the capsule and the peripheral lens cortex (Figure 5.26b). This loosens the lens nucleus from its capsulocortical attachments and enables it to be rotated.
3. Sculpting of the nucleus is performed with the phaco probe to form two furrows at right-angles with each other (Figure 5.26c).
4. A manipulator is introduced through a separate incision.
5. The phaco probe and the manipulator engage opposite sides of the furrow.
6. The nucleus is cracked by applying force in opposite directions (Figure 5.26d).
7. The nucleus is rotated 90° and a crack is made in the second furrow in the same manner.
8. Each quadrant of the nucleus is then fragmented and aspirated in turn (Figure 5.26e).
9. The remaining cortex is aspirated (Figure 5.26f).
10. Viscoelastic material is injected into the capsular bag.
11. The IOL is inserted.

The potential advantages of phacoemulsification, compared with standard ECCE, include more rapid wound healing, short convalescence and early stabilization of refractive error with less astigmatism. The main disadvantages are a higher incidence of complications by beginners because the technique is relatively difficult to master.

Operative complications

RUPTURE OF POSTERIOR CAPSULE

Rupture of the posterior capsule is potentially serious because it may be accompanied by vitreous loss which, in turn, may lead to postoperative complications such as updrawn pupil, uveitis, vitreous touch, vitreous wick syndrome, expulsive haemorrhage, secondary glaucoma, retinal detachment and chronic cystoid macular oedema. The management depends on the severity of the capsular tear and the presence of absence of vitreous loss.

1. Capsular rupture without vitreous loss is managed as follows:
   (a) If the tear is small, a PC–IOL may be implanted with the aid of a glide.
   (b) If the tear is large or if there is a zonular tear, an AC–IOL may need to be used. In this situation, the pupil is constricted with Miocinol and the IOL inserted with the aid of a glide.

2. Capsular rupture with vitreous loss is managed first by clearing the incision and anterior chamber of formed vitreous and then deciding whether or not lens implantation is appropriate. The following are the two main techniques of anterior vitrectomy:
   (a) Sponge vitrectomy involves the excision of vitreous by the use of small triangular sponges and scissors. The tip of the sponge is applied to the vitreous; the sponge is then retracted slightly and the vitreous which has become adherent to the sponge is excised with scissors. This has to be repeated until the desired effect has been achieved.
   (b) Automated vitrectomy can be performed with the Kauffman vitrector (Figure 5.27) or an equivalent instrument.

Following anterior vitrectomy, a decision has to be made about whether or not to proceed to implant an AC–IOL. However, AC–IOL implantation is associated with a higher risk of postoperative complications than a PC–IOL, although it is probable that retinal detachment and chronic cystoid macular oedema are more closely related to the accompanying vitreous loss than to the type of IOL. Other complications, such as bullous keratopathy, hyphaema, iris tuck and pupillary irregularities, may be directly related to imperfect positioning of an AC–IOL.
POSTERIOR LOSS OF LENS FRAGMENTS

Lens fragments may migrate into the vitreous cavity after zonular dehiscence or posterior capsule rupture. This complication is more commonly associated with phacoemulsification than extracapsular extraction. The displaced fragment may involve the entire nucleus or only a very small part. Subsequent management depends on the size of the lost fragment:

1. **Small fragments** may be observed without treatment.
2. **Large fragments** consisting of 25% or more of the lens (Figure 5.28) should be removed, usually within 2 weeks of the original cataract surgery to expedite visual rehabilitation and also to break the cycle of progressive lens-induced inflammation, and to avoid long-term glaucoma. The surgical technique involves a pars plana vitrectomy and removal of the fragment using ultrasonic fragmentation.

SUPRACHOROIDAL HAEOMORRHAGE

Massive suprachoroidal (expulsive) haemorrhage is a large bleed into the suprachoroidal space which results in extrusion of intraocular contents from the eye or apposition of retinal surfaces. It is a dreaded complication which occurs in about 1:1000 cataract extractions. The source of the bleeding is a ruptured, long, or short, posterior ciliary artery. Although the exact cause is unknown, contributing intraoperative factors include a sudden decrease in intraocular pressure, Valsalva manoeuvre, coughing, vitreous loss, a sudden rise in systemic blood pressure and administration of a retrobulbar anaesthetic without adrenaline. Patients who are elderly or have arteriosclerosis, hypertension, diabetes mellitus, blood dyscrasias, glaucoma or severe myopia are at increased risk.

1. **Presentation** is typically after lens delivery with a progressive shallowing of the anterior chamber, increased intraocular pressure and prolapse of the iris. This is followed by vitreous extrusion, loss of the red reflex and the appearance of a dark mound behind the pupil. In very severe cases, all intraocular contents may be extruded through the incision.
2. **Immediate treatment** is closure of the incision and administration of a hyperosmotic agent. Although posterior sclerotomy has been advocated, it may actually exacerbate the bleeding and result in a vicious circle and loss of the eye. Postoperatively, the patient should be treated with topical and systemic steroids to reduce intraocular inflammation.
3. **Subsequent treatment** is between 7 and 14 days later when liquefaction of the blood clot allows better drainage of the haemorrhage. It involves drainage of the blood, followed by pars plana vitrectomy and air-fluid exchange. Although the visual prognosis is grave, useful vision may be salvaged in some cases.

Early postoperative complications

IRIS PROLAPSE

1. **Cause** of iris prolapse (Figure 5.29) is usually inadequate suturing of the incision. It most frequently follows inappropriate management of vitreous loss.
2. **Complications** of untreated iris prolapse include defective healing of the incision, excessive astigmatism, chronic anterior uveitis, epithelial ingrowth, cystoid macular oedema and endophthalmitis.
3. **Treatment** involves excision of the prolapsed iris tissue and resuturing of the incision.
INTRODUCTION

1. **Causative organisms**, in order of frequency, are *Staph. epidermidis*, *Staph. aureus*, *Pseudomonas* sp. and *Proteus* sp.

2. **Source of infection**, in most cases, cannot be identified with certainty. It is thought that the patient's own external bacterial flora of the eyelids, conjunctiva and lacrimal drainage passages is the most frequent culprit. Other potential sources of infection are contaminated solutions and instruments, and environmental flora including that of the surgeon and operating room personnel.

3. **Prevention** by the following measures may be beneficial in prevention of bacterial endophthalmitis:
   (a) **Treatment** before surgery of pre-existing infections such as staphylococcal blepharitis, conjunctivitis, dacryocystitis, or infected contralateral sockets in patients with ocular prostheses.
   (b) **Preoperative instillation of povidone-iodine.** A 5% solution is prepared by diluting the full-strength 10% Betadine aqueous solution used for skin preparation with 1:1 balanced saline solution. Two drops of the diluted solution are instilled into the conjunctival sac (Figure 5.32) and the eyelids gently manipulated to distribute the solution over the ocular surface. Following preparation of the skin and draping, the eye is irrigated with saline solution.
   (c) **Meticulous draping technique** that ensures that the lashes and lid margins are isolated from the operative field (Figure 5.33).
   (d) **Postoperative injection** of anterior sub-Tenon antibiotics.

Acute bacterial endophthalmitis

Acute endophthalmitis is a devastating complication that occurs in about 1:1000 cases. Despite early treatment, about 50% of eyes become blind.

CLINICAL FEATURES

The clinical features depend on its severity at the time of examination.
1. Severity
   (a) **Severe endophthalmitis** is characterized by pain, marked visual loss, lid oedema, chemosis, conjunctival injection, corneal haze, fibrinous exudate in the anterior chamber (Figure 5.34), hypopyon (Figure 5.35), vitritis, absent red reflex and inability to visualize the fundus with the indirect ophthalmoscope (Figure 5.36).
   (b) **Mild or early endophthalmitis** may be associated with only slight pain, absent or very small hypopyon (Figure 5.37), and preservation of some red reflex.

2. Time interval between the cataract extraction and the onset of symptoms can be useful in the prediction of probable offending organisms. For example:
   (a) **Staph. aureus** and Gram-negative organisms typically present between the first and third postoperative days with severe signs.
   (b) **Staph. epidermidis** may present between the fourth and tenth postoperative days with relatively mild signs.
3. **Differential diagnosis**
   
   (a) **Retained lens material** in the anterior chamber (Figure 5.38) or vitreous (see Figure 5.28) which may be associated with a severe anterior uveitis, although hyphopyon and pain are absent.

   (b) **Toxic reaction** to irrigating fluid or foreign material introduced into the eye at the time of surgery. Rarely, an intense fibrinous reaction may develop on the anterior surface of the IOL up to the eighth postoperative day (Figure 5.39). In these cases, treatment with intensive topical and periocular steroids is very effective, although posterior synechiae to the IOL may develop in some cases.

   (c) **Difficult or prolonged surgery** which has resulted in anterior uveitis and corneal oedema.

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**MANAGEMENT**

1. **Identification** of the causative organism from the aqueous and vitreous confirms the diagnosis. However, a negative culture does not necessarily rule out the possibility of infection. The samples should be taken in the operating room as follows:

   (a) **Aqueous samples** are obtained by making a small, bevelled, partial thickness incision in peripheral clear cornea with a razor-blade knife, and about 0.1 ml of aqueous is aspirated with a 25-gauge needle attached to a tuberculin syringe.

   (b) **Vitreous samples** are obtained by making a partial-thickness sclerotomy 3 mm behind the limbus and aspiration of approximately 0.3 ml of liquid vitreous from the mid-vitreous cavity with a 23-gauge needle attached to a tuberculin syringe. Frequently it is difficult to aspirate an adequate amount of vitreous with a needle and a far better method is to use a designed mini-vitrector (Figure 5.40). The samples are inoculated onto blood agar, chocolate agar, liquid thioglycolate and Sabouraud agar. If these culture media are not readily available, direct inoculation of blood culture bottles is a good
alternative. Several drops are also placed onto slides for Gram and Giemsa staining.

2. **Vitrectomy** is beneficial only in cases with very severe infection and a visual acuity reduced to 'light perception'. If visual acuity is 'hand movements' or better then vitrectomy is unnecessary.

3. **Antibiotics** which cover both Gram-positive and Gram-negative organisms should be administered. The currently recommended antibiotics are either amikacin or ceftazidime for cover against many Gram-positive and Gram-negative organisms, and vancomycin for coagulase-negative and coagulase-positive cocci. Amikacin acts synergistically with vancomycin but is potentially more retinotoxic than ceftazidime, which is not synergistic with vancomycin.

(a) **Intravitreal antibiotics** should be given after the culture specimens have been obtained and the eye has been softened. Amikacin (0.4 mg in 0.1 ml) or ceftazidime (2 mg in 0.1 ml), and vancomycin (1 mg in 0.1 ml) are injected slowly into the mid-vitreous cavity using a 25-gauge needle. After the first injection has been given, the syringe is disconnected but the needle is left inside the vitreous cavity so that the second injection can be given through the same needle (Figure 5.41).

(b) **Periocular injections** consist of an anterior sub-Tenon injection of vancomycin 25 mg and ceftazidime 100 mg or gentamicin 20 mg and cefuroxime 125 mg. The injections are repeated daily for 5–7 days according to the response to therapy.

(c) **Topical therapy** consists of fortified gentamicin 15 mg/ml and vancomycin 50 mg/ml drops every 30–60 minutes.

(d) **Systemic antibiotics** are not beneficial because of their relatively poor intracellular penetration. **Steroid therapy** will not interfere with the control of the infection, provided the organisms are sensitive to the antibiotics.

(a) **Periocular injections** of betamethasone 4 mg or dexamethasone 4 mg (1 ml) are given daily for 5–7 days according to response to therapy.

(b) **Systemic therapy** with oral prednisolone 20 mg, given four times daily for 10–14 days, may be considered only in very severe cases.

(c) **Topical therapy** with 0.1% dexamethasone drops is given every 30 minutes.

5. **Subsequent management** is to a certain extent governed by culture results. If resistant bacteria are cultured, antibiotic therapy should be modified accordingly but it may be too late.

**Late postoperative complications**

### OPAICATION OF THE POSTERIOR CAPSULE

Capsular opacification is the most common late complication of uncomplicated cataract extraction.

1. **Types of opacification**
   (a) **Elshnig pearls** (Figure 5.42) are caused by the proliferation of lens epithelium on to the posterior capsule at the site of apposition between the remnants of the anterior capsule and the posterior capsule. This is the most frequently seen type of opacification and is related to the patient's age. It is extremely common in children and occurs in about 50% of adults after 3–5 years.

   (b) **Capsular fibrosis** usually appears within 2–6 months after surgery. It may involve the posterior capsule (Figure 5.43), or remnants of the anterior capsule. The latter may be associated with contraction of the capsule in front of the implant (phimosis, Figure 5.44).

2. **Indications for treatment**
   (a) Diminished visual acuity.

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**Figure 5.41** Mini-vitrector and needle in the vitreous cavity

**Figure 5.42** Elshnig pearls
Figure 5.43 Fibrosis of the posterior capsule

Figure 5.44 Fibrosis and phimosis of the anterior capsule

Figure 5.45 Technique of Nd:YAG laser capsulotomy (see text)

(b) Impaired visualization of the fundus for diagnostic or therapeutic purposes.

(c) Monocular diplopia or severe glare caused by wrinkling of the posterior capsule.

3. Nd:YAG laser capsulotomy

The key to safe and successful laser capsulotomy is accurate focusing and using the minimal amount of energy required to puncture the capsule. With a Q-switched laser, the power setting is between 1 and 2.5 mJ/pulse and, with a mode-locked laser, between 3 and 5 mJ/pulse. The capsulotomy can be performed by applying a series of punctures in a cruciate pattern, with the first puncture aimed at the visual axis (Figure 5.45a-c). An opening of about 3 mm is usually adequate (Figure 5.45d), but larger capsulotomies may be necessary for adequate retinal examination or retinal photocoagulation.

4. Potential complications

(a) Damage to the IOL may occur if the laser is poorly focused. Although undesirable, the presence of a few laser marks on the IOL does not alter visual function or change the ocular tolerance of the IOL.

(b) Cystoid macular oedema is an occasional complication the incidence of which appears to be less when capsulotomy is delayed for 6 months or more after cataract extraction.

(c) Retinal detachment is rare, except in very myopic patients. Its incidence appears to be less when capsulotomy is delayed for 1 year or more after cataract extraction. As with cystoid macular oedema, the interval between capsulotomy and development of retinal detachments is usually many months.

Most patients who develop complications have no identifiable risk factor. The number of laser pulses and the energy level are probably not related to the risk of complications. As a result of the long interval between capsulotomy and the subsequent development of some of the complications, such as retinal detachment and cystoid macular oedema, it is advisable to keep the patient under review for several years so that potentially serious complications can be detected and treated.

MALPOSITION OF INTRAOCULAR LENS

Malposition of an IOL is uncommon but when it occurs it may be associated with both optical and structural problems.

(a) Tilting of the IOL induces astigmatism.

(b) Decentration of the IOL (Figure 5.46) may occur if one haptic is inserted into the sulcus and the other
RETINAL DETACHMENT

The main risk factors for the development of retinal detachment after cataract extraction are:

1. **Disruption of the posterior capsule**, which may occur either at the time of surgery or if capsulotomy is performed, within the first year of extraction.
2. **Vitreous loss**, particularly when management has been inappropriate, is associated with an approximate 7% risk of retinal detachment. In the presence of myopia >6D the risk is increased to 15%.
3. **Lattice degeneration** carries an increased risk of retinal detachment after cataract extraction. If possible, it should be treated prophylactically before cataract surgery or laser capsulotomy, or as soon as possible thereafter.

**SUNSET SYNDROME**

This very rare complication occurs months or years after implantation of a PC-IOL in which the implant dislocates into the vitreous, probably as a result of zonular rupture during implantation (Figure 5.48). Treatment is unnecessary if the IOL is completely dislocated and not causing problems. Partially dislocated IOLs located in the anterior vitreous can be removed and, if necessary, replaced by an AC-IOL.

CHRONIC ENDOPTHALMITIS

Chronic indolent endophthalmitis occurs when an organism of low virulence becomes trapped in the capsular bag. The two most common causative organisms are *Propionibacterium acnes* and *Staphylococcus epidermidis*.

1. **Clinical features**
   
   (a) Presentation is with late-onset, persistent, low-grade inflammation which may sometimes have
granulomatous features such as mutton fat keratic precipitates (Figure 5.49).

(b) The inflammation usually responds well to topical steroids (Figure 5.50).

(c) After the cessation of treatment the inflammation soon recurs (Figure 5.51).

(d) A sign that is highly suggestive of infection by *P. acnes* is an enlarging white plaque on the posterior lens capsule (Figure 5.52).

(e) The diagnosis can be confirmed by vitreous cultures and growth of the organism on thioglycolate broth.

2. Treatment strategy

(a) Topical and periocular steroids, and antibiotics may be tried but the response is frequently transient.

(b) Intravitreal injections of vancomycin are required in most cases.

(c) Removal of the IOL, remaining cortex and the entire capsular bag may eventually be required.

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**CONGENITAL CATARACTS**

**Morphological types**

1. **Central pulverulent cataract** (*cataracta centralis pulverulenta*) is a small spheroidal opacity, 1–4 mm in diameter, within the lens nucleus, with a clearer centre (Figure 5.53). It is dominantly inherited and non-progressive.

2. **Nuclear cataract** consists of an opacity of the central zone, between the anterior and posterior Y sutures (Figures 5.54 and 5.55). Nuclear cataracts are bilateral in two-thirds of cases and are commonly associated with microphthalmos and microcornea.

3. **Lamellar cataract** is characterized by an opacity which is sandwiched between clear nucleus and cortex and which may be associated with ridges (Figure 5.56 and 5.57). In some cases it may progress to a
Figure 5.53 Congenital central pulverulent cataract

Figure 5.56 Congenital lamellar cataract

Figure 5.54 Congenital nuclear cataract

Figure 5.57 Congenital lamellar cataract

Figure 5.55 Congenital nuclear cataract

Figure 5.58 Congenital sutural cataract
nuclear cataract. Systemic associations include galactosaemia, hypocalcaemia and hypoglycaemia.

4. **Sutural cataract** follows the anterior or posterior Y suture. It may occur in isolation or in association with other opacities (Figure 5.58).

5. **Coronary (supranuclear) cataract** consists of round opacities in the deep cortex which surround the nucleus like a crown (Figure 5.59). They are usually sporadic and only occasionally hereditary.

6. **Polar cataract**
   - *Anterior polar cataract* may involve only the capsule (Figure 5.60) or it may be pyramidal and project into the anterior chamber (Figure 5.61). Occasional ocular associations include: persistent pupillary membrane (Figure 5.62), anterior lenticonus, Peters anomaly and aniridia.
   - *Posterior polar cataract* may involve only the capsule or it may form a plaque (Figure 5.63). Occasional ocular associations include: persistent hyaloid remnants (Mittendorf dots), posterior lenticonus and persistent hyperplastic primary vitreous.

7. **Focal blue dot opacities** (Figure 5.64) are extremely common and innocuous and may co-exist with other types of congenital cataracts.
METABOLIC CAUSES

1. **Galactosaemia** is a severe impairment of galactose utilization which is caused by absence of the enzyme galactose-1-phosphate uridylic transferase (GPUT). Inheritance is autosomal recessive.
   (a) **Systemic features**, which become manifest during infancy, include failure to thrive, lethargy, vomiting and diarrhoea. Reducing substance is found in the **urine after drinking milk**. Unless galactose, in the form of milk and its products, is withheld from the diet, hepatosplenomegaly, renal disease, anaemia, deafness and mental handicap occur subsequently. The disease is ultimately fatal.
   (b) **Cataract**, characterized by a central ‘oil droplet’ opacity, develops within the first few days or weeks of life in a large percentage of patients. The exclusion of galactose (in milk products) from the diet will prevent the development of cataract and early lens changes may be reversible.

2. **Galactokinase deficiency** is a reduction or absence of galactokinase, which is the first enzyme in the metabolic pathway of galactose use. Inheritance is autosomal recessive.
   (a) **Systemic features** are absent although a reducing substance is present in the **urine after drinking milk**.
   (b) **Cataract**, consisting of lamellar opacities, may develop in the fetus or in early infancy, and some presenile cataracts may also result from galactokinase deficiency. Galactose is only indirectly cataractogenic as a result of its reduction to dulcitol within the lens. Dulcitol accumulation within the metabolizing lens cells leads to an increase in intralenticular osmotic pressure with disruption of the lens fibres and opacification.

3. **Mannosidosis** is caused by a deficiency of the enzyme alpha-mannosidase leading to the accumulation in the tissues of mannose-rich oligosaccharides.
   (a) **Systemic features** may resemble the mucopolysaccharidoses with mild-to-severe (Hurler-like) facial coarseness, mental handicap, short stature, skeletal changes and hepatosplenomegaly.
   (b) **Cataract**, characterized by ‘spike-like’ posterior opacities, is common. The absence of corneal changes may aid in the clinical differentiation from Hurler disease.

4. **Neonatal hypocalcaemia**
   (a) **Systemic features** include seizures, failure-to-thrive and irritability.
   (b) **Cataract** begins as fine, punctate, cortical opacities which may progress to lamellar cataracts.

5. **Hypoglycaemia** during the perinatal period may result in lens opacities, which are frequently reversible.

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**Causes**

**INHERITED WITHOUT SYSTEMIC ABNORMALITY**

About one-third of patients fall into this group. The mode of inheritance may be autosomal dominant, autosomal recessive, or X-linked recessive traits. Autosomal dominant is the most frequent mode of inheritance. The morpholgy of the opacities and also frequently the need for surgery are usually the same in parent and offspring.
INTRAUTERINE INFECTIONS

1. Congenital rubella is associated with cataract in about 15% of cases. After the gestational age of about 6 weeks, the virus is incapable of crossing the lens capsule so that the lens is immune. Although the lens opacities (which may be unilateral or bilateral) are usually present at birth, they may occasionally develop several weeks or even months later. The opacity can involve the nucleus and have a dense pearly appearance or it may present as a more diffuse opacity involving most of the lens. The virus has been shown to be capable of persisting within the lens for up to 3 years after birth.

2. Other intrauterine infections that may be associated with neonatal cataract are toxoplasmosis, cytomegalovirus, herpes simplex and varicella.

SYSTEMIC SYNDROMES

There are many syndromes associated with neonatal cataract. The most important are:

1. Lowe (oculocerebrorenal) syndrome, which is a rare inborn error of amino acid metabolism which predominantly affects boys. It is one of the few conditions in which congenital glaucoma and congenital cataract may coexist. Inheritance is X-linked recessive.
   (a) **Systemic features** include mental handicap, renal dwarfism, osteomalacia, muscular hypotonia and frontal prominence.
   (b) **Congenital cataract** is universal. The lens is small, thin and disc-like (microphakia), and may show posterior lentiglobus. The lens opacities may be capsular, lamellar, nuclear or total. Mothers of affected children may also show multiple punctate lens opacities.
   (c) **Congenital glaucoma** is present in 50% of cases.

2. Hallerman-Streiff-Francois syndrome
   (a) **Systemic features** include dyscephaly with a small thin nose, hypotrichosis, dental anomalies, and postnatal growth retardation.
   (b) **Congenital cataract**, which may be membraneous, is universal.
   (c) **Other ocular features** include blue sclera, strabismus and disc coloboma.

3. Chromosomal disorders: Down syndrome (trisomy 21), Patau syndrome (trisomy 13), Edward syndrome (trisomy 18), Cri-du-chat syndrome (deletion of chromosome 5) and Turner syndrome.

4. Other syndromes include Nance-Horan, Rubenstein-Taybi and Marinesco-Sjögren.

Investigations

**OCULAR EXAMINATION**

Since visual acuity cannot be obtained in neonates, greater reliance has to be placed on the density and morphology of the opacity, other ocular associated findings, and visual behaviour of the child, in order to ascertain whether or not the cataract is visually significant.

1. The density of the cataract is assessed using both the direct and indirect ophthalmoscopes. A very dense cataract occluding the pupil will preclude any detailed view with either. A less dense opacity will allow the fundus to be seen with the indirect, but not with the direct, ophthalmoscope. In the presence of an insignificant opacity, the fundus can be visualized with both instruments.

2. The morphology of the opacity can give important clues to the possible aetiology, as already described. In addition, the degree of visual impairment is related to the location of the opacity. Generally, the more posterior and central the opacity, the more visually incapacitating it will be.

3. Associated ocular pathology may involve the anterior segment (corneal clouding, microphthalmos, glaucoma, persistent hyperplastic primary vitreous) or the posterior segment (chorioretinitis, Leber amaurosis, rubella retinopathy, foveal hypoplasia). Occasionally, examination under anaesthesia may be required and repeated examinations may be necessary to document the possible progression of cataract or associated disease.

4. Other features indicative of severe visual impairment are absence of central fixation, and the presence of nystagmus or strabismus.

5. Special tests such as forced choice preferential looking and visually evoked potentials can also provide helpful information but they should not be relied on exclusively since they can give spurious results.

EVALUATION OF THE PATIENT

Unless there is a definite hereditary basis for the cataracts, the investigation of infants with bilateral cataracts should include the following:

1. Laboratory investigations
   (a) **Serological tests** for intrauterine infections (TORCH = toxoplasmosis, rubella, cytomegalovirus and herpes simplex). If there is a history of maternal rash during pregnancy, varicella-zoster antibody titres should also be assayed.
   (b) **Urinalysis** for reducing substance after drinking milk.
   (c) **Urine chromatography** of amino acids for Lowe syndrome.
(d) **Other investigations** include fasting blood sugar, serum calcium and phosphorus, red blood cell transferase and galactokinase levels.

2. **Paediatric evaluation** and chromosome analysis should be sought for evidence of other systemic diseases and dysmorphic features.

### Surgery

1. **Timing** is crucial. Visually significant cataract should be removed immediately.

2. **Lenssection–vitrectomy** is a small incision technique by which the cataract is removed using a vitreous cutting instrument. The procedure is performed through either a limbal incision or the pars plana (or pars plicata). In infants the limbal incision is preferred because the pars plana is not fully developed anatomically and the risk of disinsertion of the ora serrata at the time of instrument insertion is less. Because of the high incidence of postoperative capsular opacification, a part of the posterior capsule is also removed as well as the anterior vitreous. The main steps are as follows:
   
   (a) The soft lens matter is removed by aspiration and cutting (Figure 5.66a).
   
   (b) The posterior capsule is excised (Figure 5.66b).
   
   (c) A shallow anterior vitrectomy is performed (Figure 5.66c).
   
   (d) The anterior capsule is excised (Figure 5.66d).
   
   (e) Intraocular scissors (Figure 5.67a) and forceps (Figure 5.67b) may be required to excise thick capsular material or retrolenticular plaques in eyes with associated persistent hyperplastic primary vitreous. Figure 5.68 (a) shows the preoperative appearance of a dense congenital cataract and Figure 5.68 (b) is the appearance following lenssection–vitrectomy.

![Figure 5.66 Lenssection–vitrectomy (see text)](image)

![Figure 5.67 Use of intraocular scissors and forceps (see text)](image)

![Figure 5.68 a: advanced congenital cataract; b: appearance after lenssection)](image)

### POSTOPERATIVE COMPLICATIONS

The incidence of complications is greater than that in adult eyes.
Visual rehabilitation

Although the technical difficulties of performing cataract surgery in infants and young children have mostly been resolved, visual results continue to be disappointing because of severe and irreversible amblyopia. In general, for the purpose of selecting the appropriate mode of optical correction for the aphakic child, the two main considerations are age and laterality of aphakia.

1. **Spectacles** are useful for older children with bilateral aphakia, but are not appropriate in patients with unilateral aphakia because of associated anisometropia and aniseikonia. In infants with bilateral aphakia, they may also be inappropriate because of their weight, unpleasant appearance, prismatic distortion and constriction of the visual field.

2. **Contact lenses** provide a superior optical solution for both unilateral and bilateral aphakia, and tolerance is usually reasonable until the age of about 2 years. After this period, problems with compliance may start as the child becomes more active and independent. The contact lens may become dislodged or lost, leading to periods of visual deprivation and an increased risk of amblyopia. In bilateral aphakia, the solution is simply to prescribe spectacles, although in unilateral cases lens implantation may have to be considered.

3. **Intraocular lens implantation**, in young children, is still controversial because of the difficulty in obtaining adequate parameters and the subsequent growth of the eye after implantation. Lens implants are, however, being used with increased frequency in children. The aim is to render the eye mildly hypermetropic, after lens implantation, to induce mild myopia later in life. However, the risk of complications is still significant.

Abnormalities of lens shape

1. **A lens coloboma** is characterized by notching of the inferior equator of the lens (Figure 5.71). Ocular associations—colobomas of the iris and choroid, and giant retinal tears.

2. **Posterior lenticus** is a very rare condition which is characterized by a round or oval bulge of the posterior axial zone of the lens (Figures 5.72 and 5.73). With age, the bulge progressively increases in size and the lens cortex may become opaque. Systemic associations—most unilateral cases are sporadic, whereas bilateral cases may be familial or they may be associated with Lowe syndrome.
3. **Anterior lenticous** is an axial projection of the central 3–4 mm of the lens (Figure 5.74).
   (a) **Systemic associations** – Alport syndrome which is a disorder of basement membranes character-
   ized by progressive hereditary nephritis, and sensorineural deafness.
   (b) **Other ocular features of Alport syndrome** include cataract, retinal flecks and posterior polymorphous corneal dystrophy (see Figure 4.123).

4. **Lentiglobus** is a very rare, usually unilateral, generalized hemispherical deformity of the lens which may be associated with posterior polar lens opacity.

5. **Microphakia** is a lens with a smaller than normal diameter (Figure 5.75).

6. **Microspherophakia** is a lens with a small diameter and spherical shape.
   (a) **Complications** include lenticular myopia, displacement inferiorly or into the anterior chamber (Figure 5.76), and pupil-block glaucoma.
   (b) **Systemic associations** – familial (dominant) microspherophakia which is not associated with systemic defects, Marfan syndrome,
Ectopia lentis

Ectopia lentis refers to a displacement of the lens from its normal position. The lens may be completely dislocated (luxated) from the pupillary space or partially displaced (subluxated), but still remain in the pupillary space. Ectopia lentis may be hereditary or acquired. Acquired causes include trauma, a very large eye (i.e. high myopia, buphthalmos), anterior uveal tumours and a hypermature cataract. Only the first group will be discussed.

MARFAN SYNDROME

1. **Systemic features**: Marfan syndrome is a widespread disorder of connective tissue characterized by dystrophia mesodermalis hypoplasia. Inheritance is autosomal dominant due to mutations in the fibrillin gene on chromosome 15q. However, in about 15% of cases, no other family member shows any of the stigmata of the disorder. In its classic form, Marfan syndrome is characterized by the following:
   (a) **Skeletal anomalies** – the patient’s limbs are inappropriately long compared with the trunk (Figure 5.77), fingers are long, spider-like (arachnodactyly, Figure 5.78); there is mild joint laxity, pectus excavatum, high-arched palate, scoliosis and kyphosis.
   (b) **Muscular underdevelopment**, which leads to a high incidence of hernias.
   (c) **Cardiovascular anomalies** (Figure 5.79) include aortic dilatation with or without dissecting aneurysms of the ascending aorta, aortic regurgitation and mitral valve prolapse. These
frequently are the cause of death, with the mean age of survival being in the early forties.

2. Ocular features
   (a) **Lens subluxation**, which is bilateral, symmetrical, non-progressive and upward, is present in 80% of cases (Figures 5.80 and 5.81). As the zonule is frequently intact, the ability to accommodate is retained. In some cases the lens is microspherophakic.
   (b) **Angle anomaly** is present in 75% of eyes. It is characterized by dense iris processes and thickened trabecular sheets, and it may be responsible for glaucoma.
   (c) **Retinal detachment** is associated with lattice degeneration, and is the most serious ocular complication.
   (d) **Other features** include hypoplasia of the dilator pupillae, which makes the pupil difficult to dilate, flat cornea, blue sclera and axial myopia.

WEILL–MARCHESANI SYNDROME

The Weill–Marchesani syndrome is a rare systemic connective tissue disease characterized by dystrophy mesodermalis hyperplasia. Inheritance is autosomal recessive.

1. **Systemic features** include short stature, small, short, stubby fingers (brachydactyly, Figure 5.82) and mental handicap.
2. **Ocular features**
   (a) **Microspherophakia**.
   (b) **Lens subluxation**, which is bilateral and inferior, occurs in about 50% of cases during the 'teens' or early twenties.
   (c) **Angle anomaly** associated with mesodermal dysgenesis.
   (d) **Retinal detachment**.

![Figure 5.82 Brachydactyly in Weill–Marchesani syndrome](image)

HOMOCYSTINURIA

Homocystinuria is an inborn error of metabolism in which decreased hepatic activity of cystathionine synthetase results in accumulation of homocystine and methionine. Inheritance is autosomal recessive.

1. **Systemic features**
   (a) **Skeletal anomalies** – a tall, slim build which on cursory examination may be confused with Marfan syndrome. Arachnodactyly may occur but is less frequent than in Marfan syndrome. Osteoporosis is a common problem.
   (b) **Vascular complications** are secondary to an increase in platelet stickiness, giving rise to thrombosis of intermediate-sized arteries and veins, particularly after general anaesthesia.
   (c) **Other features**, which are common, include a malar flush, fine and fair hair (Figure 5.83), and mental handicap.

2. **Ocular features**
   (a) **Lens subluxation**, which is typically downwards (Figure 5.84), usually occurs by the age
2. **Sulphite oxidase deficiency** is a very rare, recessively inherited, disorder of sulphur metabolism which is characterized by ectopia lentis, progressive muscular rigidity, decerebrate posturing, mental handicap and death usually before the age of 5 years.

3. **Stickler syndrome** is associated with ectopia lentis in about 10% of cases. The systemic features are described in Chapter 11.

4. **Ehlers-Danlos syndrome** is occasionally associated with ectopia lentis. The systemic features are described in Chapter 10.

5. **Familial ectopia lentis** is a recessively inherited condition which is not associated with systemic abnormalities. In some patients the pupil is also displaced in the opposite direction to the lens (ectopia lentis et pupillae, Figure 5.85).

6. **Aniridia** is occasionally associated with ectopia lentis.

**MANAGEMENT OF ECTOPIA LENTIS**

The main complications of ectopia lentis are: (a) optical distortion produced by lenticular myopia, (b) astigmatism and/or lens edge effect, (c) glaucoma and, rarely, (d) lens-induced uveitis. The following are three treatment options:

1. **Spectacle correction** may correct an induced astigmatism resulting from lens tilting or edge effect in eyes with mild subluxation. Aphakic correction may also obtain good visual results if a significant portion of the visual axis is aphakic as viewed through a dilated or undilated pupil.

2. **Nd:YAG laser zonulysis** may be performed to displace the lens out of the visual axis.

3. **Surgical removal** of the lens is indicated for associated cataract, lens-induced glaucoma, uveitis, endothelial touch, or if the other methods are inappropriate.

**MISCELLANEOUS ASSOCIATIONS OF ECTOPIA LENTIS**

1. **Hyperlysinaemia** is a very rare inborn error of metabolism caused by a deficiency in lysine alpha-ketoglutarate reductase. Inheritance is autosomal recessive.

   (a) **Systemic features** include lax ligaments, hypotonic muscles, seizures and mental handicap.

   (b) **Ocular features** — microspherophakia and occasionally subluxation.

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**Figure 5.83** Fair hair and malar flush in homocystinuria

**Figure 5.84** Inferior lens subluxation in homocystinuria

**Figure 5.85** Familial ectopia lentis et pupillae