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THE CRYSTALLINE LENS AND CATARACT*

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I. Basic anatomy, physiology, and biochemistry

The crystalline lens is a biconvex avascular structure suspended by thin filamentous zonules attached to the ciliary processes between the iris anteriorly and the vitreous humor posteriorly. The lens is the lesser of the two refractive diopteric elements in the eye accounting for approximately 18 D in the unaccommodated state, increasing with accommodation. It is an encapsulated multicellular organ surrounded by a basal lamina, the lens capsule, with an anterior layer of cuboidal epithelium covering concentric layers of fibers (Fig. 7.1). The lens capsule is rich with type IV collagen. The anterior lens capsule is thicker than the posterior capsule and contains another matrix protein, laminin. The epithelial cells contain nuclei, mitochondria, endoplasmic reticulum, and other cytoplasmic organelles; metabolic activity is both aerobic and anaerobic. At the equator, epithelial cells undergo mitotic division and differentiate into lens fibers. With aging and differentiation, all cells are gradually incorporated into the lens by anterior and posterior elongation to form the fiber cells of the lens. Cellular organelles are lost during differentiation. The lens sutures are formed by interdigitation of the anterior and posterior tips of the spindle-shaped fibers. Additional branches are added to sutures as the lens ages. No cells are lost from the lens. Newly laid fibers crowd and compact previous fibers; thus the oldest (embryonic and fetal) layers are the most central. The outermost fibers constituting the lens cortex are the most recently formed fibers. In lens fibers aerobic metabolic activity is absent.

Fig. 7.1. Anatomic layers of the crystalline lens. Between the fetal and adult nuclei lie the lamellae of the infantile and adolescent nuclei.

The nucleus, the innermost part of the lens, contains the oldest cells, and metabolic activity in this region is virtually nonexistent. Metabolic activity supports active transport of amino acids and cations across the epithelium as well as protein synthesis in the fibers. Cations move actively across the anterior epithelium, but passively across the posterior lens capsule—a so-called pump–leak system. The maintenance of homeostasis is essential to lens clarity. Physiologic stresses may disrupt this homeostasis and lead to cataract formation, or opacification of the lens.

II. Optics

The lens and cornea form an optical system that focuses light from a distant object on the retina (emmetropia), anterior to the retina (myopia), or posterior to the retina (hyperopia). Myopic and hyperopic refractive errors are corrected with spectacle or contact lenses. The lens has a higher refractive index than its surroundings, resulting from the high

concentration of α-, β-, and δ-crystallins in the lens fiber cytoplasm. The ability of the lens to change the refractive power of the eye and focus near objects is called ocular accommodation. The most commonly accepted mechanism of accommodation is that ciliary muscle contraction relaxes zonular tension on the lens and allows the intrinsic elasticity of the lens capsule to increase the central convexity of the anterior lens. This change reduces the focal length of the lens and moves the point of clear vision closer to the eye. When accommodation is relaxed, the equatorial edge of the lens moves toward the sclera (see Chapter 9, sec. I.B. and Chapter 14, sec. V.B.). The accommodative response resulting from the same amount of ciliary muscle contraction, or accommodative effort, may vary depending on the age of the patient. It may be expressed as the diopteric change of lens power (amplitude of accommodation) or as the distance between the far point and the near point of the eye (range of accommodation).

Infants possess great powers of accommodation; with age, this power decreases. By about age 40 years, a substantial amount of accommodative power has been lost and reading glasses are needed for clear near vision. This is known as presbyopia, which is one of the earliest age-related lenticular changes. The amplitude of accommodation is helpful to calculate the power of the add used in bifocal spectacles to correct presbyopia.

In addition to presbyopia, ultrastructural deterioration and various biochemical changes of the crystalline lens take place with aging. The lens nucleus also becomes increasingly yellow with age (nuclear sclerosis), and in some cataracts the nuclear color may be brown or black. Nuclear sclerosis per se is not associated with loss of clarity. Often it is associated with an increase in the refractive index of the lens and a myopic shift in refraction, known as lenticular myopia. A change in color perception may result from the superimposition of a yellow filter between the retina and the incident light.

III. Age-related changes

Age-related cataracts are a major cause of visual impairment in older adults. When the transparency of the crystalline lens decreases enough to disturb vision, a clinically significant cataract exists. Such a decrease is usually the result of scattering of light rays or absorption in the axial part of the lens; similar changes in the peripheral parts of the lens may exist without loss of vision. Although these changes in the periphery are strictly cataractous in nature, surgical intervention is rarely warranted in the absence of visual symptoms.

A cataract is characterized by the zones of the lens involved in the opacity: anterior and posterior subcapsular, anterior and posterior cortical, equatorial cortical, supranuclear, and nuclear. In certain congenital cataracts, the nuclear zone is further subdivided into adult, adolescent, infantile, fetal, and embryonic zones (Fig. 7.1). There is a gradual transition but no distinct morphological differentiation between the layers of a cataract. The distinctions between these regions relate primarily to potential differences in their behavior and appearance during surgical procedures.

A. Epidemiology of cataracts

Ninety-five percent of individuals older than 65 years of age have some degree of lens opacity; many have cataracts sufficiently dense to warrant cataract extraction. The Beaver Dam Eye Study reported that 38.8% of men and 45.9% of women older than 74 years had visually significant cataracts. It is estimated that more than 1 million cataract extractions are performed each year in the United States alone. Cataract accounts for more than 15 million cases of treatable blindness in the world;

extraction often leads to complete visual rehabilitation. The Baltimore Eye Survey showed that untreated cataract was the source of blindness in 27% of African-Americans and 13% of whites.

B. Optics of cataracts

Visual disturbances resulting from a cataractous lens are secondary to fluctuation in the index of refraction creating light scattering and/or a loss of transparency. In cortical, supranuclear, and subcapsular cataracts, protein-deficient fluid collects between fibers. The index of refraction of this fluid is much less than that of fiber cytoplasm, and light scattering occurs at this interface. Light scattering also occurs from large protein aggregates linked to the cell membrane by disulfide bonds. This may cause monocular diplopia. Progressive yellowing of the lens in nuclear sclerotic cataracts causes poor hue discrimination, particularly at the blue end of the visible spectrum. The myopic shift associated with nuclear cataracts may transiently enable presbyopic patients to read without spectacles, a condition referred to as “second sight.”

IV. Evaluation and management of cataracts

A. Symptoms of cataract formation

1. **Decreased vision.** Cataracts cause painless progressive decrease in vision. Clinically significant cataracts cause a decrease in distance or near visual acuity. Posterior subcapsular cataracts of even mild degree can reduce visual acuity substantially. Nuclear sclerotic cataracts cause image blur at distance but not at near. Image blur occurs when the lens loses its ability to differentiate (resolve) separate and distinct object points. When this occurs, near visual tasks, such as reading and sewing, become more difficult. Many older patients may tolerate considerable reductions in distance acuity if their night driving is minimal, but they may not be as tolerant of a blur that interferes with their indoor activities.
2. **Glare.** One of the symptomatic manifestations of light scattering is glare. When a patient looks at a point source of light, the diffusion of bright white and colored light around it drastically reduces visual acuity. The effect is akin to looking at automobile headlights at night through a dirty windshield. Posterior subcapsular opacification is responsible for much of the glare.
3. **Distortion.** Cataracts may make straight edges appear wavy or curved. They may even lead to image duplication (monocular diplopia). If a patient complains of double vision, it is essential to determine if the diplopia is binocular or monocular. If

monocular, the examiner is usually dealing with corneal, lenticular, or macular disease.

4. **Altered color perception.** The yellowing of the lens nucleus steadily increases with age. Artists with significant nuclear sclerosis may render objects more brown or more yellow than they actually are.

5. **Unilateral cataract.** A cataract may occur in only one eye or may mature more rapidly in one eye than in the other. Unless the patient is in the habit of checking the acuity of each eye, he or she may not be aware of the presence of a dense cataract in one eye. It is not uncommon for a patient to claim that the vision in the cataractous eye was lost precipitously. Because cataracts rarely mature precipitously, it is more likely that the slowly evolving lens opacity was unrecognized until the patient happened to test his or her monocular acuity.

6. **Behavioral changes**

    a. **Children** with congenital, traumatic, or metabolic cataracts may not verbalize their visual handicap. Behavioral changes indicative of a loss of acuity or binocular vision may alert the parents or teachers to the presence of a visual problem. Inability to see the blackboard or read with one eye may be one such symptom; loss of accurate depth perception, e.g., the inability to catch or hit a ball or to pour water from a pitcher into a glass, may be another.

    b. **Prepresbyopic adults.** Difficulty with night driving is frequently an early sign of **cataract.**

    c. **Presbyopic adults.** Frequently, maturation of nuclear cataracts is associated with the return of clear near vision as the result of increasing myopia secondary to the higher refractive power of the rounder, harder nuclear sclerotic lens. Reading glasses or bifocals are no longer needed. This change is called “second sight.” Unfortunately, the improvement in near vision is only temporary as the nuclear zone becomes more opaque.

B. Signs of cataract formation

1. **Reduced visual acuity.** Although it is not part of the usual general physical examination, the measurement of visual acuity will alert the examiner to the presence of cataract as well as other ocular disorders. The examiner should always inquire about monocular acuity when conducting a review of systems.

2. **Lenticular opacification.** Examination of the red reflex with the direct ophthalmoscope set on +5 (black) D at approximately 20 cm from the patient frequently will reveal a black lens opacity against the reddish-orange hue of the reflex. This is an extremely sensitive method of detecting cataractous change. If on upgaze the opacity appears to move down, the opacity is in the posterior half of the lens; if the opacity moves up with upgaze, it is located in the anterior half of the lens

or in the cornea.

3. **Leukokoria.** The white pupil is seen in mature cataracts; in certain immature cataracts, whitish patches are seen in the pupillary zone, the result of foci of light scattering, located in the anterior subcapsular or cortical zone.

C. Diagnostic tests and spectacle correction for cataract

1. **Uncorrected and spectacle-corrected Snellen visual acuity.** Distant and near acuity with and without the appropriate glasses should be tested. Some patients with cataracts complain of poor visual function despite good Snellen visual acuity. Snellen charts measure high-contrast visual acuity. Cataracts can cause decreased appreciation of contrast, leading to subjective visual dysfunction. Snellen visual acuity in a brightly lit room versus a dark room may be substantially worse secondary to glare.

2. **Non-Snellen acuity.** Tests of contrast sensitivity may be used to objectively document subjective decrease in contrast, although they have yet to be widely standardized for this purpose. Cataracts, especially posterior subcapsular and cortical, may cause debilitating glare. Several readily available instruments can document the effect of glare on visual acuity, e.g., **brightness acuity testing.** Confrontation visual fields and Goldmann and automated visual field testing may be valuable to evaluate the degree of preoperative visual field loss. **Potential acuity meter** testing can be helpful in evaluating the lenticular contribution to visual loss. Similarly, **laser interferometry** is predictive of final visual acuity in moderately dense cataracts.

3. **Flashlight examination of lens and pupil.** The direct and consensual pupillary responses are not affected by lens opacities if a bright light is used; if a dim flashlight is used, the responses may be less pronounced when illuminating the eye in the presence of a dense cataract. A flashlight may also render anterior lens opacities more visible to the examiner if pupil size is not reduced excessively.

4. **Direct ophthalmoscopy.** With the pupil dilated, nuclear cataracts often appear as a lens within a lens when viewed against the red reflex with a +5 to +10 D lens.

5. **Slitlamp biomicroscopy** allows the most detailed examination of the anterior part of the eye. The extent, density, type, and location of the cataract can be easily determined. Slitlamp examination is also helpful in determining the position of the lens and the integrity of the zonular fibers. Excessive distance between the lens and the pupillary margin may indicate lens subluxation. Slitlamp biomicroscopy may have limitations, especially in detecting oil droplet cataracts, which may be easier to detect using the direct ophthalmoscope through the 10 D lens.

6. **Refraction and retinoscopy.** Myopia induced by the early stages of nuclear cataract formation can be detected by routine refraction. Patients may be well corrected for

years with a stronger myopic distance lens and reading add. Retinoscopy will reveal the abnormal reflexes associated with lenticonus, a condition in which the anterior or posterior surface of the lens (or both) is excessively convex or conical.

7. **Fundus evaluation.** Both the direct and indirect ophthalmoscope can be used to evaluate the anatomic integrity of the posterior segment. Dilated fundus examination is necessary to evaluate the macula, optic nerves, vitreous, retinal vessels, and retinal periphery. Attention to macular degeneration, diabetic retinopathy, macular edema, retinal ischemia, vitreoretinal traction, neovascularization, optic nerve pallor, extensive cupping, and posterior capsular ruptures is important because these conditions may limit visual rehabilitation after cataract surgery.

8. **A-scan and B-scan ultrasonography** are techniques for measuring the thickness and location of a cataract. A-scan ultrasound techniques to measure the eye's axial length paired with keratometric measurement of corneal curvature allow precise calculation of appropriate intraocular lens (IOL) power, thus minimizing postoperative spherical refractive error. B-scan techniques are particularly useful in evaluating partial or total dislocation of the lens, and also provide a means of detecting abnormalities in the posterior half of the eye in the presence of a very dense cataract that precludes direct visualization. Some secondary cataracts form in response to posterior segment tumors or inflammation, thereby necessitating ultrasonography to ascertain the anatomic state of the eye behind the lens (see Chapter 1, sec. III.H.).

V. Abnormalities of the lens

A. Ectopia lentis (dislocated lens)

(see Chapter 11, sec. VII.E. and sec. VII.F., and Chapter 15).

1. **Homocystinuria.** This autosomal-recessive condition is associated with a deficiency of cystathionine beta-synthetase, an enzyme responsible for condensing homocystine and serine to cystathionine. Bilateral lens dislocation occurs in this disease; if the lens dislocates into the anterior chamber, acute pupillary block glaucoma may develop. Cataractous changes are unusual. Systemic manifestations include malar flush, mental retardation, osteoporosis, pectus excavatum, decreased joint mobility, and eczema. Abnormal physical findings are usually apparent by age 10, but may be delayed until the third decade. Surgical removal of dislocated lenses is fraught with complications, including vitreous loss, iris prolapse, and retinal detachment. **General anesthesia is to be avoided,** if possible, because of the increased risk of vascular thrombosis.

2. **Marfan syndrome.** Inheritance of this disorder is autosomal dominant. In contrast to homocystinuria, in which lens dislocation is usually inferior, in Marfan syndrome dislocation is superior and only occasionally into the anterior chamber. Surgical extraction of these lenses is complicated by many of the same problems encountered in homocystinuria. The systemic manifestations of this disease include a tall and thin

body habitus, scoliosis, arachnodactyly, elastic skin, hyperextensible joints, aortic insufficiency, and aortic aneurysm. Abnormalities in the expression of fibrillin have been found in some patients with Marfan syndrome. The diagnosis is usually established by the physical examination and the characteristic patient habitus.

3. **Weill-Marchesani syndrome** is inherited as an autosomal-dominant or -recessive trait. Patients are short with broad hands and fingers. There may be joint stiffness, prominence, and decreased mobility. Carpal tunnel syndrome may result from fibrous tissue hyperplasia. Lenses are small, spherical, and frequently dislocate anteriorly, precipitating acute glaucoma. Patients are easily distinguished from those with Marfan syndrome or homocystinuria by their characteristic body habitus.

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4. **Other heritable conditions with ectopia lentis:** hyperlysinemia, Crouzon syndrome, oxycephaly, Sprengel deformity, sulfite oxidase deficiency, Sturge-Weber syndrome, Ehlers-Danlos syndrome, dwarfism, polydactyly, and mandibulofacial dysostosis.

5. **Traumatic dislocation.** During ocular injuries, particularly blunt ocular trauma, expansion and compression of the globe equator can lead to lens dislocation or subluxation. The direction of dislocation could be vertical, horizontal, anterior (into the chamber) or posterior (into the vitreous). Very often this condition is associated with traumatic mydriasis, vitreous prolapse, rosette-shaped cataract formation, blood in the anterior chamber (hyphema), and glaucoma.

B. Congenital cataracts

(see Chapter 11, sec. VI., sec. XVI., and Chapter 15)

1. **Galactosemia** is the result of an autosomal-recessive inborn error of galactose metabolism, a deficiency of galactose-1-phosphate uridyltransferase, the enzyme that converts galactose-1-phosphate to uridine diphosphogalactose. In the presence of milk sugar (lactose), this deficiency leads to the accumulation of galactose-1-phosphate and galactose. Galactose is converted by the enzyme aldose reductase to the sugar alcohol, galactitol. The accumulation of this sugar alcohol within lens cells creates a hypertonic intracellular milieu that is neutralized by the influx of water. The entry of water leads to swelling, membrane disruption, and opacification of the lens. Cataracts are not apparent at birth, but usually develop within the first few months of life. A central nuclear opacity resembling a drop of oil appears within the lens. This opacity may progress to opacification of the fetal nucleus. The disease is manifest in **patients fed milk products** that contain the disaccharide lactose (glucose plus galactose). Mental retardation, growth inhibition, and hepatic dysfunction commonly ensue if the disease goes untreated. The diagnosis can be made by an assay for uridyltransferase in peripheral red cells.

2. **Galactokinase deficiency.** The enzyme galactokinase converts galactose to galactose-1-phosphate. In this autosomal-recessive disorder; lack of this enzyme leads to the accumulation of galactose, which is then converted to galactitol. The

same osmotic events as in galactosemia occur and lead to cataract formation. Systemic manifestations of galactosemia are absent, however. Except for cataracts, these patients usually enjoy normal health. Treatment is dietary restriction of galactose-containing foods. Patients who are heterozygous for this genetic defect are also at increased risk of cataract formation during the first year of life.

3. **Hypoglycemia.** Neonatal hypoglycemia occurs in approximately 20% of newborns. The incidence is significantly increased in premature infants. Blood sugars of 20 mg per dL or less may cause repeated episodes of somnolence, diaphoresis, unconsciousness, and convulsions. Repeated hypoglycemic episodes may lead to a characteristic lamellar cataract in which layers of cortical opacity are separated from a deeper zonular cataract by clear cortex. The cataract does not usually appear until the child is at least 2 to 3 years old; in many patients, no visual disability is encountered. Experimental evidence suggests that this cataract may be the result of an inactivation of type II hexokinase. Treatment of this condition is aimed at the restoration and maintenance of normal glucose levels in the blood.

4. **Lowe syndrome (oculocerebral renal syndrome).** Bilateral nuclear cataracts and microphakia are always found in this X-linked recessive disorder. Aspiration of these cataracts is associated with a poor prognosis for full visual recovery. Other ocular abnormalities include glaucoma and malformation of the anterior chamber angle and iris. Most striking is the **blue sclera,** a manifestation of scleral thinning. Frequently, there is associated mental retardation, failure to thrive, absence of eyebrows, and vitamin D-resistant rickets. Vomiting, glucosuria, proteinuria, renal caliculi, and convulsions are not unusual. The exact biochemical defect is unknown. Female genetic carriers have punctate cortical opacities.

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5. **Myotonic dystrophy** is inherited as an autosomal-dominant trait and is the result of a defect in the gene encoding myotonin protein kinase; the defective gene contains increased repeats of a trinucleotide sequence. Early cataracts are characteristic and consist of fine, scattered, dust-like opacities in the cortex and subcapsular region. Multicolored (especially red and green) refractile bodies are scattered among these finer dust-like opacities; this finding is commonly referred to as a “christmas tree” cataract. Later in the disease, a granular, posterior subcapsular cataract develops. Cataract extraction usually is performed in adulthood, and the visual prognosis is good if there is no serious posterior segment abnormality such as optic atrophy or retinal degeneration. Associated systemic findings include dystrophic changes in muscles, including impaired contraction and relaxation, gonadal atrophy, and frontal baldness.

6. **Rubella cataract** results from fetal infection with rubella virus before the ninth week of gestation. This virus is known to inhibit mitosis and cell division in many fetal tissues. Involvement of the lens vesicle at the time of elongation of the posterior epithelial cells leads to abnormal lens development. The cataract has a characteristic morphology: slightly eccentric, dense, white core opacity and lesser opacification of the surrounding cortex. The anterior suture may be visible. Other ocular manifestations of this disease are microphthalmos, pigmentary retinopathy,

and iritis. Because of the involvement of dilator fibers, pupillary dilation is frequently incomplete. Early referral to an ophthalmologist will optimize chances for successful cataract extraction. Surgery is frequently difficult because of poor pupillary dilation, shallow anterior chamber depth, and the small size of the eye. The newest techniques, however, with phacoemulsification and other forms of cataract aspiration, give this procedure a better prognosis. Rubella prevention, through vaccination, probably offers the safest and most effective method of reducing the incidence of this disease.

7. **Other congenital abnormalities** in which cataracts are found include Werner syndrome, congenital ichthyosis, Rothmund-Thomson syndrome, Fabry's disease, incontinentia pigmenti, Refsum's disease, gyrate atrophy of the choroid and retina, Stickler syndrome, neurofibromatosis type II, cerebrotendinous xanthomatosis, Wilson's disease, Niemann-Pick disease Type A, mannosidosis, mucolipidosis I, and Hallermann-Streiff-François syndrome (see Chapter 10, sec. IX, sec. X, sec. XI and sec. XII.).

C. Age-related cataracts

The term *senile* was previously used to describe cataracts in older adults with no specificity with regard to morphology or etiology. However, with the emergence of surgical procedures specifically suited to certain forms of cataracts, it has become more important to specify the degree of nuclear sclerosis, which often correlates with hardness, when distinguishing one form of cataract from another. Typically, cataracts are now described by their cortical, nuclear, and subcapsular components.

D. Metabolic cataract

1. **Diabetic cataract**

    a. **Osmotic cataract.** In the pre-insulin era, the acute onset of a mature cataract in an untreated or brittle diabetic was not uncommon. Because blood sugar control is now relatively easy with insulin, this form of acute osmotic cataract is extremely rare. It is possible to precipitate this cataract, however, by abruptly lowering a markedly elevated blood sugar with insulin. The intracellular hypertonicity, which results from the accumulation of sorbitol and glucose, remains after the serum osmolarity decreases precipitously with the blood sugar. A rapid influx of water leads to acute swelling and opacification of the lens.

    b. **Poorly controlled diabetics** frequently experience changes in their refractive status. Increasing blood sugar is associated with myopic change; decreasing blood sugar is associated with hyperopic change. These changes are most noticeable during periods of poor control. Restoration of control eliminates these refractive error fluctuations.

c. **The reversible appearance and disappearance of a posterior subcapsular cataract** have been documented in adult diabetics. These are believed to be a somewhat lesser response to the same osmotic stress that opacifies the entire lens of the uncontrolled juvenile diabetic.

d. **Incidence.** Cataract extraction is done more frequently and at an earlier age in adult diabetics when compared to the general population. It is not clear whether this is because of the increased incidence and more rapid maturation of cataract in diabetics, or whether it is a manifestation of the increased detection of cataracts in a population already under medical supervision. Abundant experimental evidence suggests that diabetes is a significant cataractogenic stress that, added to other age-related stresses, may lead to earlier maturation of cataracts. The development of aldose reductase inhibitors to block the conversion of glucose to sorbitol may provide a means of eliminating this cataractogenic stress. The morphology of the adult diabetic cataract is indistinguishable from that of the nondiabetic senile cataract.

2. **Hypocalcemic (tetanic cataract).** The morphology of the cataract associated with hypocalcemia varies with the age at which hypocalcemia occurs. In the infant, depression of serum calcium produces a zonular cataract with a thin, opacified lamella deep in the infantile cortex. In the adult, acquired or surgical hypoparathyroidism is associated with punctate, red, green, and highly refractile opacities occurring in the subcapsular area. In **pseudohypoparathyroidism,** lamellar opacities are found in the nucleus of the lens. Ocular involvement may include papilledema, diplopia, photophobia, and strabismus. It is believed that calcium is necessary to maintain membrane integrity and that calcium deficiency leads to membrane disruption and increased permeability.

3. **Aminoaciduria** (see sec. V.A. and sec. V.B., above; Chapter 10, sec. VII.).

4. **Wilson's disease** is an autosomal-recessive disorder of copper metabolism. Ocular involvement includes the Kayser-Fleischer ring of golden peripheral discoloration of Descemet's membrane and the formation of a characteristic sunflower cataract. The latter is due to the deposition of cuprous oxide in the anterior lens capsule and subcapsular cortex in a stellate pattern. Sunflower cataracts, however, do not generally induce significant reductions in visual acuity.

5. **Galactosemia** (see Chapter 10, sec. VII. and sec. V.B., above)

6. **Myotonic dystrophy** (see sec. V.B.5., above)

E. Drug-induced cataracts

1. **Corticosteroids.** The use of topical, inhaled, and systemic corticosteroids is associated with the appearance of axial posterior subcapsular cataracts. These cataracts frequently assume a discoid morphology and, by virtue of their axial position near the nodal point of the eye, cause significant visual disability. The

higher the dose of corticosteroid and the longer the course of treatment, the more likely the patient is to develop cataract. By reducing the dose and duration of treatment, the cataractogenic process can be slowed or stopped. All patients with diseases requiring prolonged corticosteroid therapy should be periodically evaluated by an ophthalmologist.

2. **Miotics (anticholinesterase drugs including echothiophate, diisopropyl fluorophosphate, and demecarium bromide)** are used to treat glaucoma and some forms of strabismus in children. Prolonged use is associated with the appearance of anterior subcapsular vacuoles and granular opacities. Removal of the drug reduces the risk of progression and may even be associated with reversal of cataract.

F. Traumatic cataract

1. **Contusion cataract.** Lens opacification may occur in response to blunt and penetrating trauma.

2. **Infrared radiation** (glassblower and glassworker cataract). Prolonged exposure (over several years) to infrared radiation leads to exfoliation of

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the anterior lens capsule. In contrast to the pseudoexfoliation seen in elderly patients, in true exfoliation large pieces of the lens capsule flake off and may curl back on themselves in the pupillary zone of the lens. Although this is not a true cataract, prolonged exposure may lead to the appearance of a discoid posterior subcapsular opacity with many highly refractile spots. This cataract is rarely seen currently, but was common in the 19th century.

3. **X-ray radiation.** Ionizing radiation can produce a characteristic posterior subcapsular opacity. The degree of cataract formation is a function of the radiation dose. As little as 300 to 400 rad as a single dose may lead to cataract formation. These opacities do not necessarily progress. With higher doses, the cataract may progress to involve the entire posterior subcapsular zone, and rare cases may involve the entire lens. There is usually a latent period between the exposure and the onset of cataract that may be as brief as 6 months (after intensive exposures such as atomic bomb injury) or as long as several years. Neutron and alpha beams produce the greatest ionization and pose the greatest risk of cataract formation. Gamma and x-rays are the most frequently used forms of radiation in medicine; therefore, these forms are most frequently associated with cataract formation. Appropriate shielding of the lens is necessary when tumors around the eye are being treated.

4. **Microwave radiation.** Although it is possible to produce cataracts in animals exposed to high doses of microwave radiation, microwave exposure in humans has not been associated with cataract formation. Surveys of armed services personnel exposed to microwave radiation at radar installations have not revealed an increased incidence of cataracts. This remains, however, a potential cataractogenic factor.

5. **Ultraviolet (UV) radiation.** Exposure to UV radiation has been linked to human cataracts in many studies. This radiation is divided into three wavelength bands: UV-A (400 to 320 nm), UV-B (320 to 290 nm), and UV-C (290 to 100 nm). UV-A induces suntanning, UV-B induces blistering and skin cancer, and UV-C does not normally reach the earth's surface. Depletion of the ozone layer, however, is allowing more UV-B and potentially UV-C to penetrate our atmosphere. The cornea absorbs some of the UV-B (snow blindness is UV-B keratitis), but all wavelengths longer than 300 nm are transmitted into the eye. Although the lens tends, in turn, to transmit UV-A, it absorbs almost all intraocular UV-B, the wavelength shown experimentally and clinically to be most damaging to the lens, particularly in formation of cortical and nuclear cataracts. Only the retina is susceptible to damaging effects from visible light. Ocular exposure to UV-B may be reduced by 50% by just wearing a hat with a brim, 95% by wearing ordinary glasses with glass lenses, and 100% by using a UV coating or UV screener incorporated into spectacle lenses. UV screeners may protect against cataract formation.

   The ability of the natural lens to absorb UV light may have a protective effect on the retina. UV irradiation may cause macular degeneration in patients whose natural lenses have been removed because of cataracts. Because of this, manufacturers of IOLs incorporate filters that block transmission of wavelengths below 400 nm. All polymethylmethacrylate (PMMA) and many silicone IOLs now contain UV filters. Similar filters are being used in spectacles. The patient benefits from both the filtered spectacles and the implants by experiencing less glare in bright light and perhaps greater macular longevity. Darkly tinted glasses block transmission of visible light but an incorporated UV block or coating must be present to offer full UV protection.

6. **Electrical cataract.** Electrocution injury can be associated with cataract formation. The cataracts may involve both the anterior and the posterior subcapsular and cortical regions and are usually more extensive on the side with the greater electrical burn. The morphology of these cataracts

   varies, but may include punctate dust-like vacuoles as well as linear and cortical spokes. Nuclear opacification is unusual.

7. **Copper (chalcosis) and iron (siderosis) cataracts.** Intraocular foreign bodies containing copper and iron may lead to cataract formation. Both intraocular copper and iron may lead to significant loss of vision.

   a. **Copper** is extremely toxic to the eye and produces a sunflower cataract with small yellowish-brown dots in the subcapsular cortex within the pupillary zone. The petals of the sunflower may extend toward the equator.

   b. **Intraocular iron** may produce a brownish subcapsular opacity without characteristic morphology. Intralenticular iron may produce a mature cataract. The brown color may involve other parts of the eye such as the iris or the cornea.

8. **Syn- and cocataractogenic factors.** Many investigators have advocated the concept

of cataractogenesis as the result of multiple subthreshold cataractogenic stresses. Each stress acting alone is insufficient to cause cataract; however, when these stresses act in concert, a cataract may form. It is possible that age is one of these cataractogenic stresses, and that the superimposition of other toxic stresses on an aging lens may accelerate the rate of cataract formation. Conversely, the elimination of one or more subcataractogenic stresses may delay or entirely prevent cataract formation.

G. Other forms of secondary cataract

1. **Cataract secondary to ocular inflammation.** Chronic keratitis, iritis, and posterior uveitis may all lead to cataract formation. The exact mechanism of lens opacification is poorly understood, but treatment of this cataract is addressed primarily to the control of the ocular inflammation while minimizing the dose of corticosteroid used to treat the inflammation.
2. **Neoplasia.** Anterior and posterior segment tumors such as ocular melanoma and retinoblastoma may lead to cataract formation. Metastatic tumors involving the choroid or anterior segment may also cause cataract.

H. Miscellaneous lenticular abnormalities

1. **Pseudoexfoliation.**

   Pseudoexfoliation is the deposition of a dandruff-like material on the anterior lens capsule, posterior iris, and ciliary processes, and is associated with a form of open-angle glaucoma. The material does not derive from the lens capsule and is therefore called pseudoexfoliation. It is of visual significance because severe **glaucoma** and **weak lens capsule zonules** frequently coexist, which may make ciliary sulcus placement of a posterior chamber lens implant advisable at the time of cataract surgery despite a successful extracapsular procedure.
2. **Lens-induced inflammation.** A hypermature cataract may leak lens protein into the anterior chamber. These proteins may act as antigens and induce antigen–antibody formation, complement fixation, and inflammation. Although topical steroid therapy will temporarily suppress the inflammation, a permanent cure of this condition is obtained only by cataract extraction.
3. **Lens-induced glaucoma.** In a similar manner, the leakage of lens protein into the anterior chamber may elicit a macrophage response. Macrophages engorged with lens protein and/or free high-molecular-weight lens proteins obstruct the trabecular meshwork outflow tract, and aqueous humor produced within the eye cannot exit freely. An acute glaucoma called **phacolytic glaucoma** may arise. Treatment is by immediate cataract extraction.

4. **Pupillary block glaucoma.** As a mature lens swells and becomes hypermature, the disintegration of protein molecules into smaller molecules results in intralenticular hypertonicity, and swelling may obstruct the flow of aqueous humor around the iridolenticular interface. This leads to iris bombé and an acute form of **angle-closure glaucoma.** Peripheral iridectomy is inadequate treatment. The lens must be extracted to relieve

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the pupillary block. In other forms of pupillary block glaucoma in which an intumescent lens is not involved, peripheral iridectomy is sufficient treatment.

VI. Medical treatment of cataract

A. Dietary factors

Epidemiologic studies have recently indicated that patients with diets high in the antioxidant carotenoid alpha carotene had a notably lower incidence of nuclear cataract, those high in the carotenoid lycopene were lowest in cortical cataract, and those high in the carotenoids lutein and possibly zeaxanthin were lowest in posterior subcapsular cataract. Vitamins C and E had no notable effect.

B. Mydriatics

The patient with a small axial cataract may occasionally benefit from pupillary dilation (mydriasis); this allows the clear, paraxial lens to participate in light transmission, image formation, and focusing, eliminating the glare and blur caused by these small central cataracts. Phenylephrine 2.5%, one drop bid in the affected eye, may clarify vision. In the hypertensive patient, the use of a short-acting, mydriatic-cycloplegic drug such as tropicamide 1% or cyclopentolate 1% will exacerbate hypertension.

C. Diabetes

1. **Age-related cataracts,** as stated earlier, are widely believed to occur more frequently and mature more rapidly in diabetics. Just as careful control of blood sugar levels can minimize the troublesome changes in refractive error that occur in patients with poorly controlled diabetes, some mild cataracts can be reversed through diabetic control. Advanced cataracts are not benefited by better control of diabetes.
2. **Aldose reductase inhibitors** have been used successfully in animals to prevent “sugar cataract” (diabetic and galactosemic) formation; such drugs may be beneficial in human diabetics. Blocking the conversion of glucose to sorbitol by aldose reductase might delay or prevent the adverse osmotic stress resulting from the intracellular accumulation of sorbitol, a sugar alcohol.

D. Removing cataractogenic agents

Irradiation (infrared and x-ray radiation) as well as **drugs** (corticosteroids, phenothiazines, cholinesterase inhibitors, and others) can cause cataracts. Conversely, their removal may delay or prevent further progression of the cataract. Any drug or agent with known cataractogenic properties should be used as briefly, at as low a dose as possible, or both. Ophthalmologic evaluation before and during treatment can alert the physician to signs of cataract formation.

VII. Surgical treatment of cataract

A. Timing of surgery

1. **Visual considerations.** The mere presence of a cataract is insufficient reason for its removal: It is important to establish the patient's specific visual needs before undertaking surgery. If the cataract is uniocular, surgery may be delayed until the cataract is mature, as long as visual function in the fellow eye is sufficient for the patient's needs and the patient does not need stereoscopic vision. If bilateral cataracts are present, extraction of the cataract from the eye with the worse visual acuity may be done when the patient regards the visual handicap as a significant deterrent to the maintenance of his or her usual lifestyle.

B. Preoperative evaluation and considerations

1. **The preoperative ophthalmologic evaluation** should include a complete examination to rule out comorbid conditions, such as longstanding amblyopia, pseudoexfoliation, retinal tears or holes, macular lesions, or optic nerve abnormalities that may affect the visual or surgical outcome. An accurate refraction of both eyes, measurement of corneal refractive power with a keratometer, and measurement of axial length with an A-scan ultrasound are all necessary to calculate the appropriate IOL power. Some surgeons measure macular acuity using devices that project either Snellen letters or gratings onto the macula through a relatively clear part of the lens (potential acuity meter). These measurements give the surgeon and patient an indication of the visual acuity that can be obtained

postoperatively, but they are not foolproof. These tests tend to underestimate postoperative visual acuity in some situations, while overestimating visual acuity in some macular diseases.

2. **Preoperative medical evaluation.** Each patient should be evaluated by an internist or general practitioner before surgery for any conditions that may affect the patient's surgical or postsurgical course. The necessary testing depends on the patient's age and prior medical history.

3. **Preparation for surgery** should include a full explanation of the potential risks and

benefits of proposed surgery and anesthesia, as well as the technique for administering eye drops and ointments and other postoperative care. Any blepharitis, dacryocystitis, or other ocular surface disease should be treated and resolved before proceeding with intraocular surgery.

4. **Both outpatient and inpatient** surgical facilities are used for cataract surgery, with the latter reserved for patients at risk for medical complications. Well-designed, certified outpatient surgical facilities offer the patient the briefest possible surgical experience and reduce to a minimum the disruption of the patient's normal living routine. Such facilities offer the surgeon an opportunity to deliver state-of-the-art surgical care in an efficient outpatient environment at minimal cost.

C. Preoperative medications

1. **Mydriasis.** For planned extracapsular cataract extraction (ECCE) and phacoemulsification, it is crucial that the pupil be widely dilated throughout most of the procedure. This is most often achieved with a preoperative combination of an adrenergic agent (such as phenylephrine), an anticholinergic agent (such as cyclopentolate or tropicamide), and a cyclooxygenase inhibitor (such as flurbiprofen). The cyclooxygenase inhibitor is believed to contribute to maintaining and preventing formation of intraoperative mydriasis. Intraoperative mydriasis may also be maintained with the use of dilute epinephrine in the irrigating solution.

2. **Anesthesia options.** Cataract extraction may be performed under local, topical, or general anesthesia. Local anesthesia minimizes the risk of wound rupture, a complication frequently associated with coughing during extubation and postoperative nausea and vomiting. The use of 1:1 mixed 2% to 4% xylocaine and 0.75% bupivacaine in facial and peribulbar or retrobulbar blocks achieves rapid anesthesia, akinesia, and postoperative analgesia for several hours. Care to avoid intravascular injections of anesthetic is essential, because refractory cardiopulmonary arrest may result from an inadvertent intravenous or intraarterial injection. Many patients express dread of the facial and retrobulbar injections; proper preoperative sedation and good rapport with the surgeon make them quite tolerable.

   Topical anesthesia, in conjunction with intravenous sedation and clear corneal incisions, has been used with increasing frequency in very cooperative patients and does not carry the risks of local anesthesia. This is becoming the most commonly used approach for managing uncomplicated soft cataracts. It permits very early postoperative use of the eye, because there is no lid ptosis, diplopia, or amaurosis. Although topical anesthesia for cataract surgery is a relatively new approach, it will be some time before safety comparable to retro- or peribulbar techniques can be demonstrated in patients with dense nuclei. Patients who are extremely apprehensive, deaf, mentally retarded, unstable, or cannot communicate well with the surgeon are discouraged from having topical anesthesia.

3. **Intraocular pressure (IOP) lowering.** Preoperative IOP reduction can prevent such

operative complications as vitreous loss, expulsive choroidal hemorrhage, and shallowing of the anterior chamber. This can be accomplished by mechanical pressure (digital massage or Honan balloon) or osmotic means (intravenous mannitol).

4. **Preoperative prepping–antibiosis** is designed to prevent postoperative endophthalmitis, a condition that is devastating but rare. Many surgeons prescribe a topical antibiotic such as tobramycin preoperatively to eradicate conjunctival bacterial flora. Most surgeons prepare the lids and facial

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skin with 10% povidone–iodine. Many surgeons will also place one drop of 5% povidone–iodine into the conjunctival cul-de-sac.

D. Surgical techniques

1. **Intracapsular cataract extraction (ICCE).** Removal of the entire lens (with capsule intact) is performed with a forceps or cryoprobe. Usually the supporting zonules are dissolved with the enzyme alpha-chymotrypsin. This procedure was the most widely used surgical technique of cataract extraction for nearly 60 years, but has been almost entirely replaced by extracapsular and phacoemulsification techniques.

2. **ECCE** (Fig. 7.2). This technique is designed to remove the opaque portions of the lens without disturbing the integrity of the posterior capsule and anterior vitreous face. Compared to ICCE, there is a significantly lower incidence of postoperative cystoid macular edema (CME) and retinal detachment, improved prognosis of subsequent glaucoma filtering surgery or corneal transplantation, reduced incidence of vitreocorneal touch and bullous keratopathy, and reduced secondary rubeosis in diabetics. In ECCE, the anterior capsule is opened widely, the nucleus expressed through a 9- to 10-mm incision, and the residual equatorial cortex aspirated, using either automated irrigation–aspiration machines or manual handheld devices. The posterior capsule may be polished, but is otherwise undisturbed, and serves as the resting site for posterior chamber lens implants. Some posterior capsules may opacify within a few months or years of surgery. These are easily opened on an outpatient basis using the infrared neodymium:yttrium, aluminum, and garnet (Nd:YAG) laser mounted on a slitlamp delivery system (see sec. VII.E.9., below).

Fig. 7.2. Planned extracapsular cataract extraction by irrigation–aspiration technique. **A:** Posterior limbal groove, 10 to 11 mm, 2/3 scleral depth. **B:** 3.5-mm entry into anterior chamber, viscoelastic substance in anterior chamber. **C:** 360-degree round cystotome opening of anterior capsule. **D:** Removal of anterior capsule. **E:** Corneal scissors angled at 45 degrees extend wound to 10 mm. **F:** 10-0 nylon sutures 7 mm apart displaced from wound and nucleus expressed by gentle pressure at 6 o'clock (muscle hook) and 12 o'clock (lens loop). Assistant lifts cornea and rotates and “teases” nucleus out with 19-gauge needle. **G:** Irrigate anterior chamber, tie sutures, and place third suture at 12 o'clock before irrigation–aspiration of residual lens. **H:** Polishing posterior capsule. (Adapted from Hersh P. *Ophthalmic surgical*

procedures. Boston: Little, Brown, 1988:91–93.)

3. **Phacoemulsification** (Fig. 7.3)

Fig. 7.3. Scleral tunnel wound construction (front and side views).

a. **Technique**

1. **Wounds.** Numerous wound configurations have been developed for use with phacoemulsification. In one popular technique, a partial-thickness scleral groove long enough to accommodate the width of the IOL is made perpendicular to the sclera, 2 mm posterior and tangential to the limbus. A 2.8- to 3-mm scleral tunnel is then fashioned, with entry into the anterior chamber occurring in clear cornea. The length of the anterior chamber entry wound is initially kept just long enough for the diameter of the phacoemulsification probe and is extended after phacoemulsification to accommodate the IOL. This type of wound has a triplanar configuration and is usually self-sealing. Another method involves making the entire stepped wound through clear cornea. With any wound configuration, one or two paracenteses are made 90 degrees from the primary

wound and a viscoelastic substance is injected into the anterior chamber before entry through the primary wound. This paracentesis provides entry for a second instrument useful for handling the nucleus during phacoemulsification.

2. **Capsulotomy versus capsulorrhexis.** Most surgeons performing phacoemulsification now use the capsulorrhexis technique to create a small opening in the anterior capsule. This involves making a smooth, continuous, circular tear in the anterior capsule. The “beer can” capsulotomy technique is used exclusively with ECCE, making multiple small tears in the anterior capsule that are joined just before the removal of the capsulotomy flap. Capsulorrhexis produces a small opening in the anterior capsule that is less likely to tear than the beer can capsulotomy, possibly producing a loss of capsular support for the IOL. The small opening also allows “in-the-bag” placement of IOLs with much greater certainty.

3. **Lens removal.** After hydrodissection and hydrodelineation (injection of balanced salt solution into the lens to delineate its structures and provide for easier nucleus rotation), removal of the lens nucleus is

accomplished with the phacoemulsification probe. The nucleus is emulsified by a titanium needle vibrating at ultrasonic frequencies (28 to 68 kHz) and aspirated by the probe that also passes irrigating fluid into the eye through a concentric soft or rigid sleeve. Many different techniques are used to accomplish phacoemulsification. The “chip and flip” technique involves using the probe to sculpt the nucleus into a bowl, the superior pole of which is then flipped anteriorly so that the probe can work from below it. Phacoemulsification can be carried out in the anterior chamber as well with the nucleus being prolapsed there after capsulorrhexis. Most surgeons use the “divide and conquer” supranuclear phaco, or the “chopping” method.

The former method, in which a cross is fashioned in the nucleus with the phacoemulsification probe, produces four fragments that are then manually split and phacoemulsified separately. Supranuclear phaco requires a large capsulorrhexis opening and elevation of the nucleus above the anterior capsular rim for emulsification. The chopping technique can be used after deep nuclear sculpting to create a vertical trough. A second instrument designed for chopping is inserted through the paracentesis and the nucleus is cracked. The phaco tip is used for stabilization and the phaco chop instrument is inserted below the anterior capsule to be embedded in the periphery of the epinucleus, creating a free wedge that is easier to emulsify with the phaco tip. The size of the nuclear wedges can vary depending on the nuclear consistency; hard nuclei require smaller wedges than softer nuclei.

4. **Irrigation–aspiration.** After phacoemulsification of the nucleus, removal of the soft cortex is accomplished with an automatic or manual irrigator–aspirator.

5. **IOL placement.** After removal of the cortex, viscoelastic material is injected to expand the capsular bag and deepen the anterior chamber. The wound is extended, if needed, to accommodate the IOL, but this may not be necessary when using a foldable IOL. The lens is placed between the posterior capsule and the remaining anterior capsule if possible, because this in-the-bag placement results in a more stable IOL.

6. **Wound closure.** After insertion of the IOL, scleral tunnels sometimes may need to be closed with a single horizontal suture that helps reduce the astigmatism associated with radial sutures. Failure to use sutures to close large cataract wounds may be associated with an increased risk of endophthalmitis, but this has not

been conclusively demonstrated. For small self-sealing phacoemulsification wounds, sutures are needed only when the security of the wound is not ascertained.

b. **Benefits.** Although there is no significant difference in the final visual acuity outcome among patients with 3.5-, 7.0-, and 10-mm incisions, there are a number of advantages of phacoemulsification over ECCE: earlier visual recovery, earlier return to usual activities, decreased astigmatism, greater wound stability, decreased risk of wound rupture, and the use of a safer, closed fluidics system during surgery that minimizes IOP fluctuations, and perhaps the chance of debilitating expulsive choroidal hemorrhage.

c. **Risks.** The difficulty of phacoemulsifying hard brunescent cataracts has deterred ophthalmic surgeons from offering phacoemulsification to patients with this type of cataract. Despite new titanium needle designs (thin-walled, oval, 15-, 30-, and 45-degree tip angles) and more powerful sonicators, which facilitate emulsification of brunescent nuclei, many surgeons believe that excessive energy release may limit its safety. Emulsification of the nucleus in the posterior chamber (behind the iris plane) has reduced the risk of damage to the corneal endothelium and, in certain patients, has reduced the importance of corneal endothelial dystrophy as a contraindication to phacoemulsification. In inexperienced hands, however, there may be an increased risk of posterior capsular rupture compared to ECCE. With the advent of synthetic hyaluronic acid and chondroitin sulfate (viscoelastic substances), topical nonsteroidal antiinflammatory drugs (NSAIDs) (e.g., flurbiprofen), and iris retractor hooks, it has become possible in many cases to deal with shallow chambers and poorly dilated pupils—two additional conditions that dissuaded surgeons from considering this operation. Because the number of contraindications to phacoemulsification has decreased dramatically over the past few years as the result of improved instrumentation and techniques, phacoemulsification has become by far the preferred method of cataract extraction for most ophthalmologists.

4. **Pars plana lensectomy and phacofragmentation.** Cataracts frequently obscure vitreous pathology. To deal surgically with such pathology, the cataract may have to be removed. This may be accomplished with an ultrasonic needle introduced via an anterior approach or through the sclera and pars plana. The emulsified lens is aspirated through the same needle; irrigation fluid enters the eye through a separate portal. Pars plana techniques are not used for cataract extraction alone, because the posterior capsule cannot be preserved.

E. IOLs (intraocular lens implants)

In the last 20 years, the replacement of a cataractous crystalline lens with a clear lens of PMMA has become an integral part of almost every cataract procedure. Of these lenses, the vast majority are the posterior chamber type (PC IOL). Although the vast majority of IOLs used to be made of PMMA, silicone and acrylic lenses now account for the majority of the IOL market. Iris-fixation and iridocapsular IOLs are no longer used because of a higher complication rate. The number of implants today largely reflects new cases of visually significant cataracts. The rapid increase in IOL implants and their popularity is a function of

many factors such as increasing acceptance by physicians and patients, a growing appreciation of the improved visual rehabilitation and quality of vision compared to aphakia corrected by spectacles or the lesser convenience of contact lenses, the lowered postoperative complication rate of CME and retinal detachment with the extracapsular techniques, improved instrumentation, better lens design, and the advent of viscoelastic substances.

1. **Anterior chamber intraocular lenses (AC IOLs)** are relatively easy to insert after ICCE or ECCE, and most are easy to remove if so indicated (Fig. 7.4A, Fig. 7.4B). The first AC IOLs to gain acceptance were either rigid, vaulted, four-footed lenses, or rigid, closed-loop lenses, some of

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which had rough edges that induced **uveitis, glaucoma,** and **hyphema** (blood in the anterior chamber) **(UGH syndrome)** and had a higher incidence of corneal endothelial decompensation and CME than the newer flexible, open lenses. Although the former IOLs are no longer available, large numbers are still implanted in patients with well-functioning eyes.

Fig. 7.4. A: The nucleus is first sculpted in a cruciate configuration to approximately 75% depth (steps 1 to 8). Following this, gentle cracking is performed (steps 9 to 12). The second instrument is inserted either through the scleral tunnel or a corneal paracentesis three to four “hours” away in clock position. **B:** Position of instruments for cracking.

To reduce postoperative complications and to avoid sizing problems that resulted from rigid, oversized lenses eroding into and painfully inflaming intraocular structures, or too-small lenses “propellering” in the AC, manufacturers developed AC IOLs with flexible supporting loops that rest gently against the angle structures (Fig. 7.5A, Fig. 7.5B). It has been shown that all closed-loop and open broad-loop AC IOLs have varying angle contact during flexion of the globe. This results in chronic irritation, secondary angle synechiae, endothelial cell loss, CME, or all of these. Current evidence indicates that the optimal AC IOLs are the three- and four-footed flexible open design (Kelman style) that maintain a small but constant area of angle contact with external ocular pressure. In general, visual results are excellent, complications are low, and surgical outcomes may be comparable to those obtained with PC IOLs. Even with flexible loops, an occasional patient may have ocular tenderness upon touching the eye postoperatively. This may be an indication for removal of the IOL only if symptoms do not abate when the patient stops rubbing the eye.

Fig. 7.5. Common intraocular lens styles.

2. **Secondary IOLs.** For those patients who did not have an IOL implanted at the time of primary cataract extraction, a **secondary IOL** may be put in at a later date, thus avoiding the need for contact lenses or aphakic spectacle correction. Preoperative evaluation for suitability should include specular microscopy (endothelial cell count should be greater than 1,200 mm^2), no active intraocular inflammation, an intact posterior capsule (for PC IOLs), and a sufficiently intact iris and anterior chamber angle for the AC IOL feet to rest on with stability.

3. **Rigid PC IOLs** are by far the IOLs most commonly inserted after ECCE with the posterior capsule relatively intact (a small defect may still allow use of a PC IOL in many cases) (Fig. 7.5). The loops fit either within the capsular bag of the lens or in the ciliary sulcus, a ridge just anterior to

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the ciliary processes. The loops prevent the lens from moving by becoming enmeshed in a fibrous cuff. The optic rests against the posterior capsule. In certain patients with insufficient capsular support, iris or scleral fixation with 10-0 Prolene sutures may be utilized. The advantages of this technique over AC IOLs have not yet been conclusively demonstrated.

   a. **Design modifications.** The first PC IOLs had simple J-loop Prolene haptics and PMMA optics all in one plane. Since then, modifications in designs in Fig. 7.5 include (a) a gentler J-curve or broadening of the haptic all the way to a C-curve to increase the fixation contact, with sizes ranging from 10 to 14 mm, (b) anterior angulation of the loops to minimize iris capture, (c) reduction or elimination of fixation holes that might cause glare in an inordinately large pupil, (d) variable optic sizes from 5.0 to 7.0 mm, (e) variable haptic-to-haptic diameters to allow for in-the-bag or sulcus fixation, (f) incorporation of UV chromophores to screen out harmful UV-B rays, (g) use of PMMA for both haptic and optic construction to reduce potential inflammatory reaction to Prolene degradable haptics, (h) use of single-piece construction, (i) biconvex optics to decrease posterior capsule hazing, (j) laser ridges, and (k1) introduction of multifocal and soft foldable IOLs (see sec. VII.E.5. and sec. VII.E.6., below).

      All of the above are generally aimed at various levels of IOL improvement, but each lens design must be selected on the basis of its suitability for a given patient, and not all modifications invariably achieve the desired end point.

   b. **Disadvantage.** Although the reduced incidence of cystoid macular edema and retinal detachment following ECCE with PC IOL implantation is well documented, the technical difficulty of removing a PC IOL remains a major potential disadvantage of this type of implant. Fortunately, the need for explantation of PC IOLs has been far less than for all other styles of implants, and it is the hope of all ophthalmologists that this risk will remain a potential one.

4. **Complications of AC and PC IOLs** are rare, often self-resolving or resolved with

medical therapy, and usually occur in fewer than 1% or 2% of patients. In order of approximate decreasing frequency, the most common complications are macular edema, secondary glaucoma, hyphema, iritis, corneal edema, pupillary block, retinal detachment, vitritis, endophthalmitis, and cyclitic membrane.

5. **Foldable PC IOLs** made of hydrogel or silicone are now the most commonly used lens after phacoemulsification surgery.

    a. **Advantages** of such lenses are (a) insertion through a small incision (3 to 4 mm); (b) good to excellent tolerance and flexibility; (c) hydrophilic properties that reduce endothelial damage through low interfacial energy; (d) autoclavability; (e) reduced astigmatism and faster rehabilitation due to smaller incisions; and (f) a thicker lens (silicone), which fills and distends the posterior capsular bag more fully, possibly decreasing the incidence of capsular haze (however, see sec. VII.E.9., below).

    b. **The disadvantages** of the current generation of foldable PC IOLs occasionally reported are (a) a tendency to decenter, particularly after YAG posterior capsulotomy; (b) imperfect surface finishing, especially with silicone lenses (possible potential for UGH syndrome); (c) severe iris chafing; (d) weak tensile strength with lens tears and increased susceptibility to damage; (e) groove marks from insertion instruments; (f) tissue damage when lens is released from folding instrument; (g) visual acuity possibly decreased; (h) suboptimal insertion instruments and difficulty inserting lens without tissue damage; (i) pitting during YAG capsulotomy; (j) lack of UV filters in some models; and (k) unknown long-term effects on ocular tissues.

6. **Multifocal PC IOLs.** These lenses focus both near and far objects, placing images on the macula simultaneously. The brain decides which image to

concentrate on via selective vision. Success is determined by whether the patient can distinguish between the two objects. For “best-case patients” in a U.S. Food and Drug Administration (FDA) study, 82% had near vision of J-1 to J-3 with distance correction only. A variety of styles are undergoing clinical trials. Thousands of multifocal lenses have been implanted worldwide, but adequate follow-up is needed to produce reliable safety and efficacy data.

7. **Contraindications to IOL implantation** include active, uncontrollable uveitis (see sec. VII.G., below) or proliferative diabetic retinopathy, glaucoma with progressive visual field loss (although with PC IOLs this may be a relative contraindication), and youth. The lower limit of age at which it is safe to implant a lens is not known. Ophthalmologists in some centers are implanting lenses in children, and they are doing so according to carefully designed and monitored research protocols. Most centers regard 18 years of age as the limit below which patients may be too young for IOL implantation. There is considerable difference of opinion regarding this lower limit; some surgeons believe it to be too low, others too high. Other contraindications include aniridia and a history of IOL intolerance in a fellow eye, unless judged the result of suboptimal lens style or intraoperative complication. Over the years, the list

of contraindications has decreased in size as surgical techniques and implant quality have improved.

8. **Results of IOL implantation** have been generally excellent, with final visual acuity of 20/40 or better in greater than 90% of AC and PC IOLs. PC IOLs may give slightly better results regardless of age by currently available studies (94% versus 90%). There is evidence, however, that ECCE surgery coupled with an AC IOL or suture-fixated PC IOL may give equally good results and should be considered in certain patients, e.g., pseudoexfoliation with weak zonules.

9. **YAG capsulotomy.** In patients over 65 years of age, the posterior capsule usually remains clear behind the optic. In younger patients, and in some older patients, the capsule often opacifies and must be opened. Before the advent of the YAG laser, the posterior capsulotomy with a PC IOL in place was a formidable surgical challenge. The YAG laser has simplified this procedure, however, and in fact has prompted lens manufacturers to place small ridges on the posterior surfaces of the optic to keep the capsule away from the surface of the optic and avoid inadvertent damage to the optic when the laser light is used to cut the posterior capsule. The laser ridge is probably not essential to successful lens design. If the lens dislocates sideways during normal postoperative fibrosis, however, otherwise-clear vision may be blocked by the ridge. It is possible to fracture or pit the optic with the YAG laser beam, but fortunately these small imperfections are usually insignificant. Photodisruptive powers used range from 0.6 mJ for thin capsules to 4.0 mJ for dense fibrous bands (usually posttraumatic).

F. Postoperative care

1. **Wound healing** occurs slowly over a 4- to 8-week period, but small refractive changes from further healing of the incision occur up to 9 months postoperatively. As corneal anesthesia decreases, contact lenses can be fitted if the patient did not receive an IOL. Nonabsorbable radial sutures may be cut or removed if protruding or inducing astigmatism usually between 6 and 8 weeks from ECCE wounds, and as early as 3 to 4 weeks from scleral tunnel wounds used for phacoemulsification; some are not removed. Topical corticosteroids used to control postoperative inflammation may retard healing slightly, but patients with sedentary jobs are often back to work in 3 to 5 days.

2. **Postoperative medications.** Patients undergoing general or local anesthesia often receive a subconjunctival injection of an antibiotic such as cefazolin (100 mg) and a steroid such as methylprednisolone (50 mg). A topical antibiotic ointment is instilled at the end of the procedure after topical anesthesia. If elevated pressure is anticipated, an oral carbonic

anhydrase inhibitor such as sustained-release acetazolamide 500 mg may be given immediately postoperatively, administered at bedtime, or both. A common postoperative regimen also includes antibiotic–steroid combinations initially qid and

tapered over 3 to 4 weeks.

3. **Dressings.** An eye patch is used for only a few days postoperatively if topical anesthesia is not performed; a protective shield is worn for a few weeks at night to avoid injury to the eye during sleep.

4. **Activities and limitations.** Because of the advances in wound construction and suture materials, postoperative wound strength is sufficient to allow resumption of quiet daily activities with little risk of wound rupture. It is still prudent, however, to avoid contact sports, vigorous exertion, excessive bending from the waist, and heavy lifting for approximately 3 weeks after phacoemulsification and 6 weeks after ECCE.

5. **General medical considerations.** Constipation, coughing, and wheezing should be avoided. Any stressful diagnostic procedure, such as sigmoidoscopy, barium enema, pulmonary function studies, and exercise tolerance tests, should be delayed if possible for 3 to 6 weeks. Anticoagulant therapy can be resumed 1 or 2 days postoperatively if no bleeding is observed. Most topical medications used postoperatively (topical steroid drops and antibiotics) have minimal and easily recognizable systemic side effects. Carbonic anhydrase inhibitors used to treat postoperative glaucoma may lead to potassium depletion, depression, and cardiac arrhythmias. The control of diabetes mellitus is usually easily reestablished, but sometimes ocular and nonocular (headache, nausea, and vomiting) complications of cataract surgery may interfere with this.

G. Cataract surgery in uveitis patients

Posterior subcapsular cataracts are common complications of uveitis, particularly those with cyclitis. It is possible, however, to control the uveitis and place an **IOL** successfully in many of these patients through judicious use of perioperative antiinflammatory therapy.

Inflammation should be suppressed to 0 to 2 leukocytes per 0.2-mm-high slit beam in the aqueous or anterior vitreous for at least 3 months preoperatively. This may be done through the use of topical, periocular injected, and systemic corticosteroids, NSAIDs, and, where indicated, immunosuppressive agents. For the few days before surgery, patients may be treated with 1 mg/kg/day of oral prednisone with breakfast, diflunisal 500 mg p.o. bid, and topical 1% prednisolone or 0.1% dexamethasone qid along with continuation of any immunosuppressive therapy on which the patient may have been placed. If an IOL is to be placed, a posterior chamber IOL in the capsular bag is recommended to minimize uveal tissue contact. IOLs coated with heparin, a hydrophilic substance, have been shown to be less inflammatory in animals and may cause less inflammation in human uveitic patients. At the end of the surgical procedure, 80 mg of subconjunctival methylprednisolone is injected as well as an antibiotic such as 100 mg of cefazolin. Postoperatively, topical 1% prednisolone or 0.1% dexamethasone is used four to eight times daily, depending on the anterior chamber inflammation. Systemic prednisone is tapered and discontinued after the inflammation has cleared (unless the patient is maintained on this drug for systemic disease). Diflunisal is continued for 2 months postoperatively along with an immunosuppressive indicated for the primary disease.

H. CME therapy

A leading cause of visual loss after cataract extraction, occurring in about 3% to 5%, is aphakic or pseudophakic CME. Although the majority of affected eyes will ultimately regain good vision, CME prolongs the postoperative recovery period and causes concern for physician and patient alike.

1. **Etiology.** CME is a nonspecific response, which may result from a variety of ocular conditions, leading to a disruption of the blood–ocular barrier as well as clinical and histologic evidence of inflammation, particularly following cataract surgery. These factors include excessive or prolonged postoperative uveal inflammation, intraoperative vitreous loss, ICCE, and

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   ECCE with primary capsulotomy. Prostaglandins are unsaturated fatty acid derivatives formed from arachidonic acid. Their effects on the eye include disruption of the blood–ocular barriers, dilation of iris vessels, miosis, and alteration of the IOP. Prostaglandins are found in increased concentrations in the aqueous during cataract extraction and are hypothesized to be an etiologic factor in the development of postoperative CME.

2. **Aphakic CME** frequently undergoes transient periods of relapse and recovery and, although not entirely benign, is generally considered self-limited regardless of treatment regimen.

3. **Pseudophakic CME** is more persistent and often requires therapeutic intervention, although the natural history of this process is that in the absence of obvious anatomic precipitations (vitreous touch, IOL capture, etc.), most cases usually resolve within 6 months.

4. **Medical therapy** of CME is still a controversial topic, because there are few well-controlled studies. Corticosteroids (topical, periocular, or oral) have been used to treat CME even though there have been no prospective randomized trials demonstrating efficacy. Two prospective randomized trials have demonstrated improvement in visual acuity in chronic CME patients treated with ketorolac, one drop qid (Acular), an NSAID. (NSAIDs block production of prostaglandins.) One regimen that has been successful in numerous patients is flurbiprofen 0.03% (Ocufen), one drop to the affected eye q2h for 1 week followed by one drop q4h for 2 weeks. If improved, patients are then continued on flurbiprofen three to six times daily for 1 to 3 months, then bid for 8 to 12 months or for at least 2 months after fluorescein angiogram has shown resolution of CME. Systemic NSAIDs, e.g., naproxen 250 mg or indomethacin 25 mg p.o. tid, may also have a role in cases refractory to topical therapy. No prospective study has compared corticosteroids with NSAIDs in the treatment of CME. In addition, acetazolamide 250 mg p.o. qd and hyperbaric oxygen have both been shown to increase visual acuity in small uncontrolled studies of patients with aphakic or pseudophakic chronic CME.

I. Optical correction of aphakia

1. **Temporary cataract glasses** are dispensed to patients who have not received an IOL immediately postoperatively if the vision in the unoperated eye is poor. Although vision with these glasses is not perfectly clear, acuity is usually sufficiently good that patients accept them. Aphakic spectacles contain thick convex lenses; unavoidable optical aberrations with these lenses are (a) 30% to 35% magnification of objects in the field of view; (b) a dramatic restriction of the width of the visual field; (c) a circular zone around the central field in which the patient sees nothing; and (d) the annoying “jack-in-the-box” phenomenon, an apt term describing the manner in which objects in the blind circular paracentral field suddenly pop into view. If the patient's unoperated eye has good vision, usually he or she will reluctantly wear temporary or permanent cataract glasses for ambulation and driving because of these optical problems. It is not possible to use an aphakic lens over only the operated eye, because this would lead to intolerable diplopia based on image size discrepancy. It would also lead to tilting of the glasses, because the weight of the cataract lens is greater than the phakic lens. To many patients, monocular aphakia is a visual handicap far greater than the blurred but otherwise normal vision before cataract surgery. It is therefore advisable to defer cataract extraction of a unilateral cataract unless the patient has a mature lens, a lens-induced complication, or a willingness to use contact lenses or IOLs.

2. **Contact lenses** (soft, hard, gas permeable). Without going into the specific advantages and disadvantages of each form of contact lens, it is important to understand that contact lenses offer many advantages to the postoperative cataract patient who did not receive an IOL. Image magnification is usually less than 7% and does not lead to image size diplopia. Binocular fusion is possible, so depth perception is normal. Visual field

size is unrestricted, and none of the other optical disadvantages of aphakic spectacles is present. Contact lenses may be more difficult to insert and remove, however, and the rate of contact lens failure increases with age. **Continuous-wear lenses** are available as hard, semisoft, and soft lenses. They may be worn continuously, being removed for cleaning only every 6 months in the ideally fitted patient. It is difficult, however, to predict the likelihood of a successful fit. The success rate decreases with increasing age, climate (patients in dry, dusty areas do poorly), and intercurrent illness (e.g., keratitis sicca, arthritis, senility, seborrheic blepharitis). Approximately 70% of all patients are successful with continuous-wear aphakic lenses (see Chapter 5, sec. XI., and Chapter 14, sec. VIII.).

3. **IOLs,** which are placed in almost all patients who undergo cataract extraction, spare patients the challenge of wearing contact lenses and the optical aberrations associated with aphakic spectacles. Image magnification is usually less than 3% and visual field size is unrestricted. It is necessary, however, to measure corneal curvature and axial length of the eye accurately to select an IOL based on A-scan ultrasound calculations that will yield a refractive error postoperatively that is similar

to the unoperated eye. It is not advisable to insert an “average” IOL, because many patients will have significant inequality of refractive errors postoperatively. Several techniques have been described to suture PC IOLs to the sclera in aphakic patients. This approach may offer several advantages over using AC IOLs, but the learning curve may be steep. The use of 10-0 Prolene sutures and modified IOLs with islets in the haptics has facilitated the surgical technique.

Footnote

* Updated from Gregory JK, Talamo JH. The crystalline lens and cataract. In: Pavan-Langston D. *Manual of ocular diagnosis and therapy,* 4th ed. Boston: Little, Brown, 1996:131–154; Pavan-Langston D. The crystalline lens and cataract. In: Pavan-Langston D. *Manual of ocular diagnosis and therapy,* 3rd ed. Boston: Little, Brown, 1991:125–148.