

Cataracts: A Literary Review and Summary

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INTRODUCTION

A strict textbook definition of a cataract is any opacity of the crystalline lens or its capsule but, as we all know, functionally this is not the case. As eye care specialists we must concern ourselves with the functional definition of a cataract and carry this one step further into proper patient management in the case of cataractous changes.

Noting that lenticular opacities exist in a patient is relatively easy with today's instruments, however, evaluating to what extent a visual loss is due to these changes is not necessarily as simple. The practitioner must be able to reasonably differentiate between macular involvement, optic nerve involvement and cataractous changes when a patient presents with decreased visual function.

The purpose of this paper is to re-familiarize the practitioner with lenticular anatomy and physiology and with the various types of cataracts which may be seen in a practice. This review is not intended to bore the reader with pages of facts but rather to make the practitioner more aware of the numerous cataract types and their probable progression, enabling him to better manage these anomalies in his office. Only by fully understanding and grading cataractous changes can the practitioner competently state that a functional visual loss is due to these lenticular changes and not a macular or optic nerve involvement or some combination of the three.

ANATOMY

The crystalline lens is a transparent bi-convex organ positioned between the iris and the vitreous, which refracts the light entering the eye and focuses it on the retina. The primary functions of the lens are to maintain its own clarity and to provide refractive power by adding to the optical system of the eye.

The lens is avascular but obtains nutrition from the surrounding aqueous and vitreous. Glucose, from the aqueous primarily, provides the lens with the chemical energy required to continue growth and maintain transparency.

The equatorial diameter of the lens is 6.5mm at birth and grows to 9.5mm in the second decade, changing little thereafter. The axial diameter of the lens varies with accommodation. Axial diameter continuously increases throughout life as a result of lens fiber build up.

As with all lenses, the crystalline lens has two surfaces, anterior and posterior, and a border where the surfaces meet called the equator. The anterior surface has a flatter curvature than the posterior surface, being 10mm and 6mm in radius respectively. Located centrally on each surface are the anatomical poles of the lens.

The equator of the lens is not smooth, but shows a number of indentations corresponding to the zonular fibers which suspend the lens from the ciliary processes. When the

ciliary muscle contracts and the zonules relax, the lens becomes thicker and more convex, increasing the refractive power of the lens. The lens becomes thinner following relaxation of the ciliary muscle.

Lens anatomy can be simplified by stating: it is entirely surrounded by a capsule, and under the anterior capsule is a layer of epithelial cells. The epithelial cells reproduce; near the equator the cells elongate toward the poles and become lens fibers. The lens fibers stretch in an arcuate fashion from the anterior to posterior pole of the lens. Lens fibers are formed throughout life with new fibers covering old.¹

When considering the structure of the lens, there are basically three elements to be addressed:

- 1.) the lenticular capsule
- 2.) the lenticular epithelium
- 3.) the lens cells or fibers

The capsule of the lens forms a transparent, homogeneous, highly elastic envelope which is actually a thick basement membrane. It is PAS-positive. The capsule is secreted by the lens epithelium at the embryonic stage. Capsular thickness varies, being thinnest at the poles and equator and with the anterior surface thicker than the posterior surface. With the electron microscope many lamellae are found, each containing numerous fine fibrils.

Although the capsule is highly elastic in nature, it contains no true elastic tissue. Theoretically, its elasticity resides in the disposition of the fine fibrils which are composed of a form of collagen-like protein. The striation period of the fibrils is different from that of the zonules, though these fuse together.

Even though the lens capsule contains enzymes, ATP, and glycolytic intermediates, it cannot be considered to have independent metabolism. The capsule depends on its contact with the lens epithelium and fibers for metabolic supplies.

Anatomically, the capsule appears to be unchanged throughout life. However, the presence of glycosidases in the capsule suggests a slow degradation of its glycoprotein, and therefore chemical turnover of its constituent materials.

The lenticular epithelium consists of a single layer of cuboidal cells on the anterior surface deep to the capsule. The cells are firmly attached to the capsule and loosely attached to the underlying fibers. There is no corresponding posterior epithelium.

Following the anterior epithelium cells towards the equator, it is noted that they gradually become columnar, elongated, and eventually converted into lens fibers. The portion of the cell in contact with the capsule becomes the posterior part of the lens fiber, while the opposite end grows into the anterior portion of the lens fiber. The nuclei form a S-shaped nuclear zone at the equator.

In a flat mount preparation of anterior epithelial cells, three distinct areas are recognized: central, preequatorial, and equatorial. Cells are densely packed in the preequatorial and equatorial areas with most of the mitotic cells found in the preequatorial region. Cells in mitosis in the preequatorial region are extremely susceptible to radiation.

The lens fibers make up the bulk of the lens cortex and nucleus. Each fiber represents an elongated cell with a membrane. All but the oldest lens fibers are actually elongated, nucleated cells. The fibers formed earliest are found in the central core or nucleus of the lens. They gradually lose their nuclei and become true fibers. The membranes of the fibers have side digitations resulting in fiber interlocking. This interlocking system results in the necessary plasticity the fiber require to passively change shape during accommodation. The areas where the fibers meet anteriorly and posteriorly are the sutures of the lens. In the human lens, these sutures appear Y-shaped.

The spaces between the lens fibers account for only about 5% of the lens volume.² These spaces become enlarged in human and experimental cataracts. The vacuoles and clefts seen by slit lamp examination are result of fluid accumulation between fibers causing fiber destruction. The extra cellular spaces cannot be greatly increased unless the fiber membranes are broken.

New lens fibers are laid down throughout life resulting in a continuously growing lens. This does not, however, result in a proportional growth with the number of fibers since the older fibers shrink. At 65 years old, the lens is approximately one-third larger than at 25 years old.³

Lenticular consistency varies, with the superficial portion, or cortex, being softer than the central nucleus. The nucleus increases in size with age. The lens becomes flatter with age, but its refractive power is retained by an increase in the nuclear refractive index.

The zonules are thin, delicate filaments that maintain the lens suspended in position, and allow the ciliary muscle to act on it. The lens and zonules together act as a divider, separating the eye into an anterior and posterior portion. The anterior portion is filled with aqueous and is smaller while the larger posterior portion contains vitreous.

Other features of the zonules include their transparency, straight-stiff appearance, and inextensibility. Two basic types of zonules can be discriminated. The main fibers act as the primary connection between the ciliary body and the lens. The auxiliary fibers help to strengthen the main fibers and act as anchors for the fiber network to the ciliary body.

When viewing a normal lens in thin optic section, several distinct layers are noted which represent the various lens

nuclei and cortex. The layers are separated by bright bands called "zones of discontinuity." The brightness represents light reflected from an interface formed between adjacent layers of lens fibers which have different indices of refraction.

These zones of discontinuity are important as landmarks, which can be utilized by the examiner to more precisely localize lens abnormalities. It is possible to date the onset of an opacity from its location in the lens.

PHYSIOLOGY AND METABOLISM

The normal adult lens is approximately 65% water.⁴ This relatively dehydrated state gives the lens a refractive index different from that of the aqueous or vitreous. This index change allows the lens to act as an optical instrument of refraction at its interfaces. The percentage of water in the lens decreases with age which may be a partial explanation for the inelasticity of the lens in presbyopia. The cortex with its younger fibers contains a water content twice that of the lens nucleus.

The electrolyte composition of the lens is similar to that of other body cells with high potassium concentration and low sodium and chloride concentrations. The aqueous and vitreous, on the other hand, more closely resemble the composition of plasma with high levels of sodium and chloride, and low levels of potassium. Therefore a concentration gradient results between the inside of the lens and the aqueous.

This gradient is maintained by an active sodium-potassium pump resulting in the relatively dehydrated state of the lens. The capsule of the lens allows it to take on properties of an intact cell, such as swelling in hypotonic solution and dehydrating in hypertonic solution.

The pumping mechanism is located in the membranes of the epithelial cells primarily and is mediated by a carrier system. This pumping mechanism actively pumps potassium into the cell and sodium chloride outward to counteract the tendency of the ions to leak through the membrane in the attempt to reach equilibrium. The enzyme sodium-potassium activated adenosine triphosphatase, located within the cell, is believed to control the pumping mechanism. This mechanism is abolished by metabolic inhibitors, ouabain, or lack of sodium ion in the media.

The lens fibers hold potassium ions, amino acids and inositol in the cell against the concentration gradient. Potassium ion concentration inside the cell is twenty-five times higher than in the aqueous. The concentration of amino acids in the lens is two to six times higher than in the aqueous while levels of inositol are ninety-five times higher in the lens.⁵

Following chemical or anatomic damage to the epithelial cell membrane or fibers, there is an increased leakage of potassium ion, inositol and amino acids from the lens. Low calcium ion concentration in serum or aqueous increases the

leakage of these three substances from the lens. Corticosteroids increase potassium leakage while increasing sodium uptake into the lens.

"The protein content of the lens is higher than any other organ in the body. Protein accounts for 33% of the total lens weight. The perfect physiochemical assortment of lens proteins, living in an optimum environment of water, electrolytes and sulfhydryl groups, gives transparency to the lens."⁶

The physical state of lens proteins is an important factor in the maintenance of transparency. The proteins were originally subdivided into two groups, soluble crystalline and insoluble albuminoid. The crystallins are further subdivided according to their behavior in an electric field or by sedimentation, into alpha, beta, and gamma fractions. Most of the insoluble proteins are found in the nucleus of the lens, while most of the soluble proteins are in the lens cortex.

Lens protein has antigenic properties, but unlike other body proteins, it is organ specific rather than species specific. In the normal lens, the membrane of the lens fibers and lens capsule do not allow passage of protein molecules from the lens to the aqueous. If lens protein does escape into the ocular fluids, the body reacts to it as a foreign body resulting in an anaphylactic reaction, leading to inflammation of the uveal tissues.

Alpha crystalline is the largest molecule of the three soluble proteins and is closely related with the insoluble albuminoid fraction. In the ageing process or in cataracts, the albuminoid portion increases while the alpha crystalline decreases. Direct evidence supporting the interconversion of the two proteins has been demonstrated.

Glutathione, a tripeptide, is synthesized by the lens. It consists of the amino acids glutamic acid, cysteine, and glycine and is present in concentrations 1,000 times greater than in the aqueous.⁷ It is continuously formed in the lens in both the oxidized and reduced forms, with the reduced being the primary form. Glutathione has two major roles in the lens:

- 1) to preserve the physiochemical equilibrium of lens proteins by maintaining high levels of reduced sulfhydryl groups.
- 2) to maintain transport pumps and the molecular integrity of lens fiber membranes.

Reduced sulfhydryl groups are needed for the enzyme sodium-potassium adenosine triphosphatase involved in lens cation transport.

In the human lens, glutathione levels drop slightly with age. There is a comparatively large loss of lens glutathione in human senile cataracts and all experimental cataracts. The decrease of lens glutathione is always associated with or precedes disruption of the lens fiber membrane.

Lipids make up a relatively small but important percentage of the lens. Phospholipids are the main constituents of cell membranes and therefore are major components of the lens fiber membrane. High concentrations of cholesterol are found in the human lens with the concentration increasing with age. Cholesterol is the other major component of the lens fiber membrane.

Ascorbic acid is not synthesized in the lens but is present in concentrations exceeding that of the aqueous. The highest concentration lies under the capsule and decreases inwardly towards the nucleus. It is found almost exclusively in its reduced form and may participate as a hydrogen donor in the oxidative reactions of metabolism.

The lens is restricted in energy production by the environmental factors surrounding it. Being a transparent tissue, it cannot have high concentrations of pigmented respiratory enzymes. Since it has no blood supply all of its nutrients and waste products must be exchanged with the surrounding aqueous. Uniquely, it performs no work, therefore energy is required only for maintenance of transparency and the growth of cells.

The oxygen consumption of the adult lens is extremely low. The epithelium consumes the greatest amount, followed by the cortex. The capsule and nucleus consume essentially no oxygen.

Glucose is the primary substrate metabolized for energy production. Glucose from the aqueous and vitreous diffuses into the lens and is rapidly metabolized through four main pathways:

- 1) the glycolytic pathway
- 2) the Krebs cycle
- 3) the hexose mono phosphate (pentose) shunt
- 4) the sorbitol pathway

The oxygen present in the aqueous and available to the lens is only a small fraction of the amount present in the blood. With this restricted oxygen supply, the majority of glucose metabolism is through anaerobic glycolysis, the Embden-Meyerhof Pathway. Pyruvic acid is the end point of glycolysis and is further converted to lactic acid, which diffuses out into the aqueous.

Anaerobic glycolysis is relatively inefficient in terms of energy yield, however it can maintain lens transparency in the absence of oxygen, as long as adequate glucose is available. Approximately 85% of glucose in the lens is metabolized anaerobically.⁸

The Krebs Cycle requires oxygen and is very inactive in the lens as there are a paucity of mitochondria and oxidative enzymes. Cytochrome containing mitochondria are located primarily in the epithelium, but are present in comparatively small numbers than in metabolically active tissues of the body.

Approximately 14% of lens glucose is metabolized via the pentose shunt, a scheme where the direct oxidation of glucose-1-phosphate occurs.⁹ The pentose shunt does not generate ATP. It does however, form pentoses required for ribonucleic acid synthesis. Also, NADPH generated is utilized to maintain lens glutathione in the reduced state.

The utilization of glucose in the lens by the sorbitol pathway is a mystery. The mechanism of utilization is understood, however, the function of the pathway remains an enigma.

Each pathways contribution in the metabolism of lens glucose is controlled through the inhibition of key enzymes in the pathway. The hexokinase activity appears to be the limiting factor in the lens through its sluggish activity. Therefore, as the concentration of glucose is increased in the lens, the hexokinase enzyme becomes saturated, limiting further production of glucose-6-phosphate or lactate. Any excess glucose is diverted to the sorbitol pathway where it may either accumulate or re-enter the glycolytic pathway.

TYPES OF CATARACTS

For descriptive purposes in this paper, classification of cataracts will be by the age at which they occur. The major categories will be congenital, juvenile/developmental, and senile. Other less common but relevant cataracts that will be described are traumatic, radiation, and toxic.

Congenital Cataracts

Congenital cataracts are generally considered to be lens opacities which are present at birth or within three months after birth. Interference with vision is dependent the size and location of the opacity. Usually, congenital cataracts do not interfere with vision unless located directly in the visual axis. The majority of these cataracts are bilateral and stationary, and may involve the entire lens or only portions of the lens. The most commonly involved area is the cortex adjacent to the fetal nucleus. The most practical method for gauging the period of onset of an opacity is to use the Y-sutures as landmarks during slit lamp examination. Congenital cataracts will usually be located in deep embryonal fetal nuclear fibers due to the developmental period they begin in. The majority of these cataracts which interfere with vision are noted and managed at the time of birth by the attending physician. Therefore, from a practical out look, the practicing optometrist will see very few of these lenticular anomolies.

Anterior axial embryonal cataracts are the most frequent of the congenital cataracts, occurring in approximately 25% of the population.¹⁰ They are usually fine, white, multiple, irregularly formed opacities located near the anterior suture, which cause no interference with vision. Uniquely, they are the only form of cataract that does not occur symmetrically in anterior and posterior halves of the lens.

Stellate, or sutural, cataracts are much rarer and can occur at either suture, but most frequently the posterior. To the examiner, they appear as a blue-green thickening of the otherwise white sutural line. These cataracts probably develop subcapsularly during intra-uterine life in the end of the third trimester. Like most congenital cataracts, they are stationary but larger ones may interfere with vision.

Congenital punctate cataracts appear as disc shaped regions of small gray to light blue opacity surrounding the embryonal nucleus. The punctate opacities represent late forming fibers which have degenerated and broken down to granular debris, being surrounded by healthy fibers. These opacities are extremely rare but may be found in conjunction with stellate cataracts.

Anterior polar or pyramidal (central punctiform) cataracts are sharply circumscribed opacities of the anterior lens capsule. Often laminated in appearance, they can project conically outward from the lens or into the lens itself. Fine thread-like projections may radiate from the opacity, possibly connecting it to the cornea. The size of these cataracts vary with related visual loss being size dependent. Although they are normally stationary, progression has been noted. A similar cataract may occur following corneal perforation due to trauma.

Capsular flakes (Vogtkapselstar flecken) are noted as white, round, sharply circumscribed, dense opacities up

to 1mm in size. They appear as multiple, flat prominences on the capsular surface resulting in loss of the characteristic lens shagreen. Usually associated with pigment threads and star shaped pigment debris, they are rarely axial and therefore are not connected with visual loss normally.

Spurious posterior capsular cataracts, more commonly known as Mittendorf dots, are relatively common. Generally located inferior and nasal to the posterior pole, they will appear as a black dot with the ophthalmoscope and as a white dot with the slit lamp. They are simply the remnant of the hyloid artery attachment in the formation of the tunica vasculosa.

Posterior lenticonus is a rare anomaly presenting as a conical or globular protuberance on the posterior lens surface. According to Vogt, 80% of the cases report with posterior lenticular opacity complications, resulting in a dimness of vision and an increase in minus power of the central lens.¹¹ They can also occur on the anterior surface.

Zonular (Lamellar) cataracts are one of the most frequent types of congenital cataracts. They present as a uniform gray zone of lamellar opacity surrounding a clear zone, like an orange peel around the fruit. This opacified zone is then surrounded by clear cortex. The size of the opacity varies with the lenticular development at the time of lens damage; the earlier the disturbance the smaller

the opacity and the more centrally located it will be. In a lenticular cross section, they would appear as thick, gray, radially u-shaped opacities with anterior and posterior limbs which saddle the lens equator. Generally tending to increase in density with age, visual disturbance can result. They are usually bilateral.

Embryonal nuclear cataract (*cataracta centralis pulverulenta*) appear as a dense gray-white opacity located between the sutures in the region of the embryonic nucleus. It is probably related to a developmental disturbance during the first trimester. On inspection, the examiner will see a 1-2mm gray globe in the center of the lens surrounded by otherwise clear lens fibers. The lens is of normal size and thickness, however the embryonic nucleus did not develop properly. This cataract is generally bilateral, stationary, and shows hereditary tendencies. Surprisingly, considering its location and density, it shows little if any visual interference.

Axial fusiform cataracts (Spindle cataract) are anterior-posterior cortical cataracts connected by thread-like opacities running axially through the lens. Normally bilateral with equal involvement, the vision can be disturbed depending on the amount and density of the opacification. This lens anomaly is due to a disturbance in early fetal life effecting axial areas of the lens interfering with growth of new lens fibers in the region.

The total congenital cataract is relatively rare and usually zonular. Bilateral involvement is the rule with a shrunken appearance to the lens. The lens may degenerate to liquification resembling a hypermature morgagnian cataract. The probable cause is a metabolic change occurring in the third trimester.

Congenital cataracts following viral infection of the mother during pregnancy are less common now than in the past due to better public education of expecting mothers. Numerous viral infections can cause lenticular anomalies such as rubella, encephalitis, influenza, hepatitis, mononucleosis, mumps, measles, and many more.

The most common of the viral congenital cataracts is rubella induced. It is the result of the mother being infected during the first trimester and it involves all but the outermost layers of the lens. Normally bilateral, it appears in two forms: 1) a pearly white central opacity with smaller clearer peripheral zone, 2) the entire lens being opaque.

Persistent hyperplastic primary vitreous is a failure of the primary vitreous and the blood vessels to regress during development. It occurs in full term infants and presents at birth, normally being unilateral. It appears as a dense opaque tissue containing blood vessels located at the posterior lens surface. The opacity decreases in density towards the periphery allowing long, thin, rudimentary ciliary

processes to be seen by the examiner. These processes extend and attach to the posterior lens surface. The lens itself is characteristically smaller than normal but clear. If opacification does occur, it will begin at the anterior pole. The lens could also absorb leaving only capsular remnants over the retrolental tissue. Upon presentation at birth, a differential diagnosis from retinoblastoma and pseudoglioma is necessary.

Juvenile/Developmental Cataracts

Several types of cataracts form during childhood or early adulthood. These are called juvenile or developmental cataracts. Most cataracts developing during this time period affect the lens periphery, progress slowly, and rarely interfere with vision unless occurring concurrently with congenital or progressive senile lens changes. The incidence of these cataracts is relatively low at .03% for the general population.¹²

Galactosemia is a relatively rare disease which can result in cataracts in the young child. The cataracts are formed due to the systems inability to metabolize galactose normally. The earliest lens change is an increase in the refractive power about the fetal nucleus, which appears to the examiner like a drop of oil in the lens, followed by a zonular or nuclear cataract formation. These cortical changes appear as faint striate opacities in the anterior cortex. With diagnosis and treatment instituted prior to the beginning of the second month, regressive

prognosis is good, otherwise the cataract progresses rapidly and matures without treatment.

The so-called diabetic cataract is an established anomaly, with two clinical types being recognized. Typical senile cataracts occur at an earlier age in diabetics, and tend to progress more rapidly to maturity. The morphology of the diabetic cataract does not differ in any way from the senile lens changes except in its rate of progression. The duration of the diabetes and the age of the individual are important in cataract development. It also seems to be more common in women than men, with poor diabetic control being a predisposing factor. True diabetic cataracts appear as bilateral white punctate or "snow flake" anterior and posterior subcapsular opacities. Predominantly seen in young people, age 3 to 33, they tend to progress rapidly to a mature opacity. A true diabetic cataract is rare, where senile cataracts in diabetics are common. A true diabetic cataract is usually preceded by a sudden progressive myopia. Senile cataracts tend to occur more frequently in a diabetic population and at a younger age than in a normal population.

Coronary cataracts present as a wreath of club shaped opacities in the periphery of the cortex near the equator of the lens. Appearing about the age of puberty, they are usually bilateral and show a dominantly inherited tendency. Increasing in density with age, the color of these opacities change from blue to white. These opacities can generally be

seen by the examiner only through a widely dilated pupil.

Other less frequently encountered juvenile cataracts include those developing in children having Down's Syndrome or tetany resulting from hypocalcemia. These cataracts may vary in appearance and affect different layers of the lens.

Senile Cataracts

Senile cataracts characteristically first appear in patients beginning in the third to fourth decade of life. They progress at various rates and ultimately result in severe visual impairment. Classification of senile cataracts is usually done in two ways: according to the location within the lens and according to stage of development. The major division is made between nuclear and cortical with the subcapsular opacities being a subdivision of the cortical types. Senile cataracts have a variety of stages, each with its own special name such as: early or incipient, immature or intumescent, mature, hypermature, and nuclear sclerotic.

For practical management of cataract patients, a 1⁺ to 4⁺ grading system has been proposed. This system is based on two clinical data points: The opaqueness or coloration of the cataract on slit lamp evaluation (objective) and the visual acuity reported through the opacity (subjective). Although this system was originally proposed to grade nuclear sclerotic cataracts, it is a very practical clinical tool which can be utilized to uniformly manage any cataract.

It will be explained more fully in the summary of this paper.

It is generally accepted that the prevalence of senile cataracts increases with age; some studies indicating as high as 91% of the population over 80 years old demonstrated some form of senile cataractous change.¹³ However, incidence studies are so variable due to the semantics of the term cataracts, each interpreting it slightly different. All of the various studies indicated that the incidence of senile cataracts is higher in females than males.

Cystoid spaces in the lens appear just beneath the anterior and posterior surface of the adult nucleus in 10% of the patients over 40 years of age.¹⁴ With distal ophthalmoscopy they are seen as small, oval, grayish opacities arranged in a somewhat symmetric fashion. To the examiner with a slit lamp, the lesions seem to bulge forward and give the appearance of an optically empty space. Commonly mistaken for vacuoles which appear with dark borders and light interiors, cystoid spaces have opaque centers with light borders. On a management basis, vacuoles and cystoid spaces are essentially the same.

True exfoliation of the lens capsule is a rare condition largely confined to the lenses of glass blowers, blast furnace operators, and other occupations where exposure to severe heat is common. The superficial zonular lamella of the capsule becomes loosened and peels off in curls. Exfoliation is unusual even in persons exposed to great amount of infra red radiation; more commonly found is a form of posterior

subcapsular cataract.¹⁵

Pseudoexfoliation of the lens capsule occurs much more frequently than true exfoliation, with some studies showing as high as 50% of the patients over 80 years old presenting with it.¹⁶ In this anomaly, an amorphous material is deposited on the lens capsule. The movements of the iris pupillary margin with dilation and contraction rubbing the surface of the lens peel the deposited material off the capsule. The edges of the remaining deposited material are curled and irregular. At the pupillary border of the iris, flecks of grayish-white fluffy material can be seen. On pupillary dilation, a homogenous grayish disc can be seen on the lens capsule where the iris has rubbed the material off. This anomaly is often associated with cataracts, fluid vitreous, and especially glaucoma; therefore when pseudoexfoliation is diagnosed, care must be taken to rule out possible related anomalies.

Epicapsular stars are pigmented remnants of the pupillary membrane, with no visible attachments to the iris, and are frequently found on the anterior lens capsule. They are usually star shaped and peppered, apparently without any orderly arrangement on the capsule. They vary in size and number and rarely affect vision even though they may be present in sufficient number to present a lace-like appearance. Although generally a congenital anomaly, there is an acquired type.

Presenile and senile punctate opacities are found frequently, increasing in number and size with age, and sometimes go to complete opacification. They are normally located in the deep layers of the peripheral cortex and therefore do not normally interfere with vision.

Typical cortical cataract formation is different from that in the lens nucleus. Instead of sclerosing and shrinking, the cortex imbibes water and swells. The water forces lens fibers apart, forming clear spaces; the breakdown products of degenerating lens fibers dissolves in the water, forming opacities; the remaining fibers become opaque resulting in a soft cataract.¹⁷ Therefore, the most common form of early cortical changes result from lens fiber separation by clear water first, and later by cloudy opaque liquid. The lens fibers may separate either radially or between lamella.

A clear water cleft is formed if the fibers separate radially. Clefts are transparent, elongated spaces which appear optically empty. Clefts are most commonly found in the anterior cortex, or in both anterior and posterior cortex. Rarely are they seen in the posterior cortex only. An opacified cleft is termed a spoke.

If the separation of lens fibers is between lamellas, it tends to be less complete with a typical slit lamp appearance of numerous, fine, fold-like, parallel lines running as complete chords across a sector of the lens.

These lamellar separations are most commonly seen in the inferior nasal anterior cortex.

Opacification of the space between separated lamellas results in a cunieforn cataract; the most common type of cataract according to Duane. These cataracts characteristically begin in the periphery of the lens, consisting of flat, wedge shaped, dense white opacities which form a wreath around the lens periphery and gradually encroach on the axial portion. These wedges are often preceded by the fine lines of the lamellar separations. The wedges may fuse together, forming a larger flat opacity which occupies a whole sector of the lens.

An early or incipient cortical cataract is one in which cataractous or precataractous changes cause minimal visual impairment. The lens has not taken in enough water to become swollen at this point.¹⁸

An immature or intumescent cataract is the next stage of cortical cataract. The lens imbibes more water, growing noticeably larger, which results in a decrease depth of the anterior chamber. The duration and degree of this stage is quite variable. The characteristic slit lamp picture of an immature cortical cataract includes water clefts, opaque spokes, lamellar separations with cunieforn opacities intermixed with clear cortex. Vision may remain good for a time but form vision is ultimately lost.

When the entire lens substance has become opaque except for the capsule, the cataract is said to be mature. At this stage, most of the water taken on during the immature stage has been lost and the lens and anterior chamber has returned to normal size. The color of this stage of cataract is extremely variable including, bright white, yellow, gray, brown, and black depending on the softness of the lens material; the lighter the color, the softer the lens material.

The next stage of a cortical cataract is the hypermature cataract which is the result of further lens dehydration and partial calcification. The lens becomes small and irregular in thickness with the capsule appearing shriveled and wrinkled, especially with a marked or complete disappearance of the cortex. Occasionally the cortex is completely liquified allowing the sclerosed brown nucleus to float about in the milky cortical fluid. This is known as a Morgagnian cataract.

Morgagnian cataracts appear to the examiner as if the nucleus is floating in the lens, which it actually is. The upper border of the brown nucleus is visible in the inferior capsule giving a sunken half moon appearance. A tremulous iris may be present due to the deepening of the anterior chamber from lack of lenticular support to the iris.

Posterior subcapsular cataracts are relatively common and can occur at any age but seems to afflict middle-aged

persons more frequently than any other type. It is relatively rare to find an anterior subcapsular cataract unless it is an isolated finding which resulted from inflammation or injury. The early stages of a posterior subcapsular cataract are best seen with a slit lamp using retroillumination, appearing as a slight optical imperfection. Next, a tiny white granular opacity forms which enlarges to form a plaque; this plaque is an arrangement of irregular granules interspersed with vacuoles and crystals. In refracted light, the crystals may give off glints of red, green, and blue. Only a thin layer of subcapsular cortex is involved with the remainder of the cortex remaining clear. In an advanced subcapsular cataract, nuclear sclerosis normally is present to varying degrees. This type of cataract has a particularly devastating affect on vision due to its location in the lens being near the nodal points of the eye. Early symptoms include decreased night vision decreased vision in bright sunlight, and decreased ability to read. Visual impairment usually progresses rapidly to total incapacity over a period of months.

The normal lens nucleus hardens and becomes more pigmented with age. The nucleus loses its lamellar structure and becomes increasingly more amber in color. No easily observable changes mark the transition from a normally aging nucleus to that of a nuclear sclerotic cataract. The normal indicators for the 1⁺ to 4⁺ grading system are visual acuity associated with the coloration changes. As the nucleus

hardens, its refractive index increases resulting in an increase in myopia, better known as "second sight." In this period the patient can normally read without the normal reading spectacles. Later, vision gradually becomes blurred, first at distance than at near also. The center of the nucleus and increased in refractive error more so than the peripheral nucleus resulting in an exaggerated spherical aberration. At this point, no change in spectacle prescription will increase vision.

Monocular diplopia or poor hue discrimination can also occur in a patient with a nuclear cataract. Monocular diplopia is usually a result of a small nuclear cataract acting like a prism. Poor hue discrimination occurs in a heavily pigmented lens in which the normal differential filtration of light according to wave length has been exaggerated.¹⁹

There is little difference between a normal aging nucleus and a nuclear sclerotic cataract in its early stages when examined with a biomicroscope. As the process advances, the nucleus loses the lamellar arrangement and deepens in its amber coloration, becoming increasingly distinct in outline against the pale textured cortex. As the nucleus increases in pigmentation, it slowly becomes opaque. The pigmentation may be so extensive that it becomes a cataracta brunescens (brown cataract) or, less frequently, cataracta nigra (black cataract). Nuclear cataracts generally develop slowly spanning many years before the interference

with vision is enough to warrant extraction.

Traumatic Cataracts

Traumatic cataracts are the most commonly seen lens opacity in the young. They are generally unilateral. Care must be taken with an ocular contusion due to the strong possibility of induced glaucoma from a dislocated lens. Besides possible dislocation, injury may result in three types of cataracts: those occurring without rupture of the lens capsule, those following rupture of the lens capsule, and those following perforation of the lens capsule by a foreign body.

Contusion without capsular rupture may present numerous clinical pictures but classically results in two commonly seen entities; one extra capsular (Vossius Ring) and the other intracapsular (rosette cataract). A Vossius Ring can occur following a relatively direct blow to the eye, resulting in the posterior pigmented surface of the iris being forced back into contact with the anterior lens capsule. A ring of pigment is deposited on the capsule after the iris returns to its normal position. There is a tendency for the pigment to absorb with time and vision is rarely impaired. Following ocular contusion a rosette cataract may form which presents as feather-like rays radiating peripherally from the suture lines. These cataracts are characteristically found in anterior and posterior subcapsular regions and, depending on severity and opacification may impair vision.

Traumatic cataracts following rupture of the lens capsule present the examiner with an array of anomalies to contend with. With capsular rupture the lens fibers will quickly swell and opacity, possibly resulting in an acute glaucoma attack by decreasing the anterior chamber depth thus closing off the angle. Lens material may escape with capsular rupture possibly causing a phacoanaphylactic reaction, again leading to glaucoma. Therefore intraocular pressures should be monitored. Later, as absorption of the free lenticular material occurs, the entire lens may become opacified and atrophied, resulting in a shrunken or shiveled appearance. The degree of change is age dependent, with the younger person having an advantage in maintaining vision stability following capsular rupture.

Metallic intraocular foreign bodies may cause cataracts with or without mechanical insult to the lens. Iron and copper alloys compromise the majority of intraocular foreign bodies with each producing quite spectacular lenticular changes. Iron induced (Siderosis) cataracts begin with subcapsular brown dots over the anterior expanse of the lens with eventual yellowing and opacification of the entire lens. Copper induced (Chalosis) cataracts present with a greenish to red-brown discoloration following the sutural pattern of the lens giving a sunflower appearance.²⁰

Dislocation of the lens as a result of contusion occurs in two forms; subluxation and luxation. A subluxated, or partially displaced, lens is the least serious of the

dislocation phenomena. Some but not all of the zonular attachments are broken by the force of the contusion. These usually result from a non-perforating blow but can occur with a perforating one. Luxation is the total displacement of the lens with a loss of all zonular attachments essentially. It is more dangerous than a subluxated lens as the lens may reposition itself in the anterior chamber and induce glaucoma by obstructing the angle. Posterior dislocation into the vitreous usually has a less severe prognosis, but may also induce glaucoma by continuously irritating the ciliary body and creating a partial iris bombe.

Although somewhat rare, certain systemically induced cataracts are worth mentioning since they are pathognomonic of particular diseases. In hepatolenticular degeneration there may be a sunflower cataract. Cortical opacities are seen in patients with Fabry's disease and a characteristic pattern of opacities may be indicative of myotonic dystrophy. A tetany cataract can develop rapidly in patients shortly after parathyroid removal. It has also been documented that cataracts are prevalent in patients with various skin conditions; atopic dermatitis, Kyrle disease, and generalized scleroderma. Diabetic cataracts have been previously discussed in the juvenile cataract section. Current literature suggests that the existence of diabetic cataract as a specific entity, aside from that seen in juveniles, is presumptuous.

Radiation Cataracts

Glass blowers cataract result from prolonged exposure to infra red radiation. This cataract usually begins in the posterior cortex, taking on a small blot-like, flaky appearance. It is normally associated with pseudoexfoliation of the anterior lens capsule and can be prevented by wearing protective goggles.²¹

Small repeated doses of x-ray or radium radiation can result in typical cataract formation. The opacity does not form immediately but can present months to years after the exposure. The normal appearance of this cataract is a thick, dense, disc-like opacity located in the posterior subcapsular region. It progresses to a mixture of fine crumb-like opacities intermixed with vacuole structures with sharply demarcated structures. A neutron cataract is quite similar in appearance but usually progresses faster with more initial damage occurring following the original exposure.²²

Toxic and Nutritional Cataracts

Toxic substances and therapeutic drugs have also been shown to produce cataracts. Of these the most important are the corticosteroids and miotics which are widely used therapeutically. A posterior subcapsular opacity is the typical cataractous change following prolonged use (generally more than one year) of corticosteroids either systemically or topically. Long term use of long-acting miotics such as DFP and

echothiophate may also cause vacuoles initially, with nuclear and posterior subcapsular changes later. Even short-acting miotics such as pilocarpine have been associated with lens opacities. Cessation of the drug will reverse, stop or retard their progress.

Vitamin deficiencies have caused cataracts experimentally. Protein, carbohydrate, fat and certain amino acid deficiencies also have been determined to cause cataracts. But once normal metabolic balance is restored, these cataracts usually stabilize.

Summary

The list of cataract types presented may have seemed extensive but in actuality was quite brief. The point to be taken is that with the numerous types of cataracts that can occur, the practitioner can not be expected to place a label on each anomaly that is seen in the office. Therefore, a grading system that is suitable for uniformly grading of all cataracts based on visual acuity and lenticular opaqueness has been proposed.

The system is quite simple with all cataracts being graded on a 1⁺ to 4⁺ scale; 1⁺ being the least cataractous changes that result in loss of visual function and 4⁺ being the most. Coloration changes, as seen with a slit lamp will vary from practitioner to practitioner, therefore each individual must establish his own scale, combining nuclear sclerotic coloration with cortical opaque changes. Guide lines have been proposed for grading on visual acuity reduction. These can be adapted to fit the individual's scale:

1⁺ 20/25 - 20/30
2⁺ 20/40 - 20/70
3⁺ 20/80 - 20/150
4⁺ 20/200

The benefits of having a uniform grading system is not so two practitioners can discuss a cataract patient, but so the individual practitioner can more easily and reliably manage cataract patients in the office. Unless these concepts

are universally used, discussion of a 2⁺ versus 3⁺ cataract between practitioners is ridiculous due to individual bias in grading systems. But when a practitioner reads on a file card, 2⁺ posterior subcapsular cataract OD, it is readily apparrent to him what was present at the previous examination and he is prepared to accurately assess if any progression has occurred.

Time will be involved in establishing your criterion level for each grade but once established, evaluation becomes increasingly easier and more practical. This is another device to insure consistent quality patient care.

The cataract patient is not difficult to manage if care and patience is taken to assess and document the changes occurring at each examination. Determine first if the lenticular changes have affected vision. Try to identify the type of opacities and their location in the various layers of the lens so that their development and progression can be monitored. Lastly, rule out other causes of poor vision and base the recommendation for surgery, if necessary, on both the function of the patient and your professional opinion.

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