NORMAL AND ABNORMAL LENS

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METHOD OF OBSERVATION

The examination of the lens is performed by using direct illumination, parallelepiped, optic section and/or retro-illumination at the slit-lamp. Retro-illumination may also be performed using the direct ophthalmoscope. When observing with the parallelepiped and optic section, one should scan from the anterior lens capsule to the posterior lens capsule both medially and laterally. When observing the tissues with retro-illumination, focus at the plane of the iris. Retro-illumination gives an overall view of the cataract, although note that the depth of focus is usually not sufficient to provide an assessment of both the anterior and posterior lens at the same time, particularly with higher magnification, so that separate assessments are required.

NORMAL

The examiner is able to view the various layers of the lens using a parallelepiped and optic section. From anterior to posterior, the layers include the anterior capsule, anterior subcapsular space, anterior cortex, nucleus, posterior cortex, posterior subcapsular space, and posterior capsule. Anterior and posterior portions should appear clear and free from opacities. Zones of discontinuity, a series of zones of clear media in both the anterior and posterior lens cortex delineated by a curved line of scattered light, can be seen in normal adult lenses. These zones are made up of lens fiber layers with different scattering properties, likely due to different refractive indices in the continually growing lens cortex. The anterior erect Y suture and the posterior inverted Y suture are visible within the fetal nucleus. These are caused by the meeting of fibers that arch over the lens equator and join with other fibers to form branching suture lines. The sutures are visible

because of the large amount of light scatter caused by the non-uniform shape and size of the lens fiber ends, which is in contrast to the very minimal amount of light scatter caused by the rest of the uniformly shaped and orderly arranged lens fiber cells. In adolescence and adulthood, star and complex star sutures are formed, but these are much more difficult to see.

By using retro-illumination the examiner can view the red reflex which should be devoid of opacification.

During lens development new fibers from the epithelial layer are produced continuously and migrate centrally towards the cortex and nucleus. Due to this development, the lens increases in sagittal width and the nucleus becomes less flexible with increasing age.

The embryonic blood supply to the lens is the tunica vasculosa lentis and it is replaced during development. Sometimes it does not completely atrophy. In this case there remains an embryological remnant attached to the posterior capsule. It is termed a Mittendorf dot and it can be viewed by using an optic section (when it will appear white in direct illumination) or retro-illumination (where it will appear black against the orange glow from the fundus). It is most commonly located inferior and nasal to the optic axis. More rarely, more extensive remnants of the tunica vasculosa lentis result in small, light brown or tan star-shaped deposits on the anterior lens surface (epicapsular stars). They can be bilateral or unilateral and single or multiple.

ABNORMAL

Cataracts

Cataract refers to the opacification of the lens. Because of the high prevalence of lens opacity with age, the clinical definition of cataract as “an opacification of the lens that causes vision loss” is more commonly used. Note that vision loss could refer to loss of contrast sensitivity or increased disability glare and not just visual acuity loss. Cataracts are classified either as acquired or congenital and result from a variety of aetiologies:

Acquired	Congenital
Age-Related (old term: senile) Traumatic Metabolic/Systemic Disease Genetic Disease Toxic Intraocular Disease	Maternal Infection Birth Trauma Metabolic/Systemic Disease Genetic Disease Ocular Maldevelopment Syndromes

ACQUIRED CATARACTS

AGE-RELATED CATARACTS

Age-related cataracts (ARC) are the most common form of cataract that one will encounter in the clinic. Nuclear sclerotic (NS), cortical spoke (CS) and posterior subcapsular cataract are forms of ARC. Other, less common aetiologies of cataract do exist and may have significant impact on functional vision. Through clinical experience, thorough history taking, and access to academic resources, proper recognition of the aetiology of cataractogenesis should be identified.

S: Symptoms include decreasing vision, glare and difficulties with night driving. Patients may commonly report that “my glasses are dirty, but cleaning them doesn’t help”. Other symptoms may include monocular diplopia that is often associated with cortical cataracts.

The case history may also uncover other risk factors for age-related cataracts. There may be a family history of cataract as age-related nuclear and cortical cataracts have an association with hereditary factors. There may also be a history of cigarette smoking, which is an identified risk factor associated with nuclear cataracts. PSC are the least common form of

ARC and are more often times associated with non-age related risk factors. These risks include diabetes, history of systemic steroid use, and secondary to ocular disease such as retinitis pigmentosa (RP), uveitis, or eye trauma.

O: The three main morphological types of ARC include nuclear sclerotic (NS), cortical spoke (CS) and posterior subscapular (PSC).

Nuclear Sclerotic Cataracts (NS) present as a homogeneous increase in light scatter in the lens nucleus and can be associated with an increased yellowing or brunescence, which is indicative of blue wavelength-dependent light absorption. These changes also occur to a lesser degree with normal ageing. The use of a slit-lamp optic section technique is an accurate way of detecting and assessing nuclear cataracts and it is best performed with a dilated pupil, although is still possible undilated. Unless digital imaging is available, nuclear yellowing and opacification can be graded on a scale of 0 to 4+ with 0 being the absence of opacity and 4+ being the most severe form of opacity. Several standardized systems of grading cataract do exist, we will discuss The Lens Opacification Classification System (LOCS) below.

Cortical Spoke Cataracts (CS) are wedge shaped opacities found in the anterior and/or posterior lens cortex. Significant functional vision symptoms are most commonly reported when the cortical spokes enter the pupillary area having an impact on the optical axis. Opacification in the cortex that remains hidden behind the iris and does not enter the pupillary area may have no associated visual symptoms. Cortical opacities are most often found in the inferior-nasal part of the lens, suggesting the association of ultra-violet B radiation involvement in their aetiology. Opacification is due to the scattering of light when it meets irregular interfaces between regions of different refractive indices.

Cortical opacities are best seen using retro-illumination from the fundus, when the opacities appear black against the red fundal glow. The brightest reflection from the fundus is obtained when the illuminating beam strikes the optic nerve head, so that typically the illumination system is placed on the temporal side of the biomicroscope. The slit-lamp illumination beam size, shape and position can be altered to avoid the cortical opacities on its way to the fundus. Some slit-lamps allow a half-moon shape illumination beam that can be placed inside the edge of the pupil. Retro-illumination gives an overall view of the cataract, although note that the depth of focus is usually not sufficient to provide an assessment of both the anterior and posterior lens at the same time, particularly with higher magnification, so that separate assessments are required.

Cortical cataracts can also be viewed using an optic section and this can indicate whether the cortical opacities are in the anterior or posterior cortex. The cataracts appear white in direct illumination. They are often associated with water clefts, which are optically clear wedges that can be seen with slit-lamp biomicroscopy. Direct illumination is less useful for cortical cataracts as it can show large amounts of light scatter due to backward light scatter and reflections that do not cause vision loss. In addition, the opacity is viewed in many small sections and an overall view of the cataract is only possible by mentally putting together all the different sections.

Unless digital imaging is available, record cortical cataracts by drawing them. The undilated pupil can be recorded as a dashed line on this diagram. If there are cortical opacities in both the anterior and posterior cortex, both should be recorded.

Posterior Subcapsular Cataracts (PSC) presents at the posterior portion of the lens just in front of the posterior capsule. In the age-related vacuolar type of PSC cataract, localized reductions in refractive index and vacuoles are found in the early stages. In the later stages of development there is a posterior migration of epithelial cells from the lens equator. These epithelial cells converge on the posterior pole, forming the balloon or bladder cells of Wedl. A membranous appearance occurs in the later stages of PSC development due in part to the many organelles in the epithelial cells and the membrane formed by the migrating cells. Although an optic section may aid in determining the depth of any opacity of the lens, a PSC is best viewed through a dilated pupil using retro-illumination. Documentation of the extent of a PSC may be completed by using digital imaging or by simply drawing the cataract on your medical record. Recording the dimension of the undilated pupil as a dashed line on the diagram will aid in locating the PSC with respect to the optical axis.

GRADING ARC: OBJECTIVE CLASSIFICATION

Several objective classification systems have been created for the purpose of formally grading the extent of cataract development. We will discuss The Lens Opacification Classification System III (LOCS III). LOCS is in its third generation and is a series of standardized color photographs.

THE LENS OPACIFICATION CLASSIFICATION SYSTEM III (LOCS III)

The LOCS provides a simple and accurate means of grading the type and severity of age-related cataracts by comparing a patient's cataract to a set of standard photographs of cataracts. The presence or absence of opacification is assessed in **3 major anatomical zones**: the cortex, the nucleus, and the posterior subcapsular zone. The standard set of colored photographs of optic sections and retroillumination of the lens are used to grade different degrees of Nuclear Opalescence (NO), Nuclear Color (NC), Cortical Cataract (C), and Posterior Subcapsular Cataract (P) in the lens.

The severity is denoted by numbers given by the standard photos and the classification can be modified for clinical purposes. Rather than noting a decimal grading for severity in between standards, one simply uses a (+) to denote that the severity lies somewhere between two standards. For example, if an unknown fell between standards 1 and 2 its severity would be noted as 1+. The decimal grading for severity appears below.

Cataract Type & Nuclear Color	Severity
NO = Nuclear Opalescence	0.1-6.9
NC = Nuclear Color	0.1-6.9
C = Cortex	0.1-5.9
P = Post. Subcapsular Zone	0.1-5.9

Slit-Lamp Technique:

- the patient is seated at the slit-lamp
- dilation to a 6mm pupil diameter is recommended
- LOCS III standards are placed over the patient's shoulder on transparencies or photos
- room lights are dimmed
- the examiner can use **any** slit beam intensities, heights, widths, angles, etc. to discover location of cataract components in the lens.
- for grading and classifying cataract components, the examiner **must** use a:

  45° SL angle for **nuclear assessment**
  0° SL angle for **cortical and subcapsular assessment**

Grading Nuclear Opalescence (NO)

NO is graded by comparing a colored slit image to nuclear standards NO1 to NO6. The average opalescence of the entire nucleus is compared to the standards and assigned a grading of severity. Average opalescence is graded by examining the nuclear area, which is comprised of 2 anatomical zones of increased scatter. They are the **figure** which includes the embryonal nucleus and its outer shells, and the **ground,** comprised of the background regions of the nucleus. In **early nuclear opacification** the background is clear and the figure becomes brighter. **Intermediate nuclear opacification** appearance differs by the increased haze of background and this reduces the contrast between the figure and ground.

Grading Nuclear Color (NC)

NC is graded by comparing color of the unknown lens to standards NC1 to NC6. This is done in 2 regions of the nucleus: the entire cross-sectional view and the posterior subcapsular reflex. The color should be compared to colors in NC1- NC6 reference standards. Use a plus to indicate severity between standards.

Grading Cortical Cataracts (C)

Grading is achieved by comparing the opacity to the standard photographs using retroillumination. The images are focused anteriorly (at iris plane) or posteriorly (at posterior capsule plane). Isolated cortical changes such as vacuoles and dot opacities are disregarded. Dots, lamellar separations, water clefts and vacuoles that are clustered with well-defined edges are graded as cortical opacification. Opacities ≥ size of opacity at 6:00 in C1 standard are considered significant in the grading system. Compare aggregate area of opacity to C1 to C5 standards and select an appropriate interval by using a (+) to reflect severity between intervals. Opacities seen only in a posteriorly focused view are graded as cortical if they are closer to the periphery than to the center of the lens. If they are closer to center, then they are graded as P for posterior subcapsular cataracts.

Grading the Posterior Subcapsular Zone (P)

They are considered to be present only if they are visible against red reflex. Only posteriorly-focused retroillumination image is used for grading. The examiner must compare the area of opacity in lens to P1 to P5 standards and assign a (+) to reflect severity between intervals. A posterior opacity is considered large enough to grade if ≥ size of the opacity at 6:00 in the C1 standard photograph.

CLINICAL ASSESSMENT OF ARC

In addition to slit-lamp or direct ophthalmoscopic assessment of the cataract, objective assessments should include refraction (as nuclear and cortical cataracts can cause significant changes in refractive error). Note that retinoscopy is often difficult with cataracts due to minimal light returning to the retinoscopist. This can sometimes be overcome by using the larger sight hole (in those retinoscopes in which the sight hole size can be changed), using as few trial lenses as possible (to reduce light loss due to reflections from the retinoscope lenses) and working closer to the patient than the traditional 50 or 67cm. Assessment of visual function should include contrast sensitivity and disability glare testing in addition to visual acuity (preferably using a logMAR chart). The former two tests provide a better measure of the patient's functional vision compared to the high contrast, low luminance test provided by clinical visual acuity assessment.

A: It is important to differentially diagnose the morphological type(s) of cataracts as they have different risk factors and management plans (see below). A significant myopic shift in refractive error in an older patient strongly suggests a nuclear cataract and should be investigated. A significant shift in the power or axis of astigmatism in an older patient can also suggest cortical cataract and should similarly be investigated.

A PSC may be obstructed by the corneal and lens reflexes when viewed through an undilated pupil. Care should be given to conduct a thorough dilated examination of the lens to identify the extent of cataract development.

P: Cataracts are monitored until the decrease in functional visual acuity affects daily activity. Ultimately, surgery is required to remove the cataract and replace it with an artificial lens, known as an intraocular lens (IOL). Surgical referral is dependent on individual patient symptoms.

Prior to referral, there are several options that should be considered to improve the patient's visual function.

i) Provide an updated refractive correction, particularly in nuclear cataracts (myopic shifts) and cortical cataracts (astigmatic changes).
ii) Suggest an anti-reflection (AR) coating on their glasses. This is particularly useful if you suggest that they should read with their back to a window (see below), limiting reflections from the back surface of their spectacle lenses.
iii) Suggest an ultra-violet (UV) coating for glasses. Some cataracts contain fluorescent pigments that can scatter light and reduce vision further.
iv) Suggest wearing a broad brimmed hat. This can be better than suggesting tinted spectacles as a tint reduces the light from the object that you are looking at as well as the sun's light.
v) Suggest a tint for glasses. A dilated pupil may allow for light to pass around a PSC allowing for reduced light scatter and less distorted visual acuity. Providing a tint for any patient that complains of discomfort due to glare may be beneficial. Use caution in cases of CS cataracts as tints can dilate the pupil slightly and expose more of the CS, potentially reducing vision.
vi) Suggest that the patients read or do their near activities with the window/sun behind their back and/or with a lamp shining over their shoulder. Light should fall on the near task and not in their eyes. This can be further improved

using a typoscope (a black card with a wide slit in it). The black card stops light reflecting from the white pages of the book/newspaper into their eyes and the patients must move the slit in the card as they read the page.

TRAUMATIC CATARACTS

S: Patients may report a history of penetrating or concussive injuries, exposure to radiation (environmental or therapeutic), or electric shock.

O: Penetrating injuries display opacification local to the area of involvement. Concussion may lead to a Vossius ring or what is viewed as an imprinting of the iris pigment onto the anterior lens capsule. A more diffuse opacification of the lens may be seen in cases of cataracts resulting from electric shock or exposure to radiation. Exfoliation of the anterior lens capsule may result from intense UV exposure.

A: Penetrating, Concussion, Radiation, Glassblower's, Electric shock

P: In cases of cataract resulting from trauma, damage to the lens may be threatening to the health of the eye resulting in a possible medically necessary lens extraction. In cases of a ruptured lens capsule, lens proteins may "leak" into the aqueous of the anterior and posterior chambers and cause an anaphylactic response. These cases are medical emergencies and need to be assessed by an ophthalmologist. As in the case of any type of opacification of the lens that does not affect the integrity of the eye health, traumatic cataracts may be monitored until the decrease in visual function affects daily activity.

METABOLIC/SYSTEMIC DISEASE

S: Patients may report a history of diabetes mellitus (DM), hypertension (HTN), galactosaemia, mannosidosis, Wilson's disease, hypocalcemia, galactokinase deficiency, Fabry's disease and others.

O: Diabetic cataracts present in 2 forms. The first, termed "true" diabetic, is due to osmotic over-hydration of the lens & appears as bilateral white punctate or "snowflake" or "rosette" opacities that are located either anteriorly or posteriorly. The second, termed "senile" diabetic cataract, is similar to the common senile changes discussed previously but occurs earlier in the diabetic.

Patients with a history of HTN often develop cataracts due to the medications used to treat their disease, not from the disease itself. The cataracts present similar to ARC.

Alpha Galactosidase A or Fabry's disease cataract has 2 different presentations. The first, sometimes referred to as an "oil droplet" appears as a fine anterior subcapsular cataract that is cream colored & feathery. The other, located in the posterior lens is shaped like a star or cross.

Wilson's syndrome cataract, called green "sunflower", consists of central pigmented opacities with tapering extensions. Because copper is deposited anteriorly & posteriorly within the lens, it takes on a yellow-green appearance

Hypocalcemia cataract consists of multicolored crystals or flecks. They are small white or polychromatic crystals located in the anterior and posterior cortex just beneath the capsule. A clear zone separates this zonular cortical cataract from the nucleus.

Mannosidosis cataract varies in presentation. Galactokinase deficiency is an autosomal recessive disease that presents with lamellar cataract.

A: DM, HTN, alpha galactosidase A, Wilson's disease, hypocalcemia, mannosidosis, galactokinase deficiency, Fabry's disease

P: As any type of opacification, the cataract is monitored until the decrease in visual acuity affects daily function. Referral is based on an individual patient basis.

GENETIC DISEASE

S: Patients may report a history of Myotonic dystrophy or Down's syndrome or other genetic syndromes.

O: Myotonic dystrophy causes a "Christmas tree" cataract, which is a stellate figure in the posterior subcapsular area with or without multicolored dust-like opacities present in the cortex.

Down's syndrome cataracts vary in appearance but generally have the potential to decrease VA.
The more common presentations consist of cortical flake opacities or arcuate opacities.

A: Myotonic dystrophy, Down's syndrome

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

TOXIC DISEASE

S: Use of steroids, phenothiazines, miotics, anti-neoplastics, anti-arrhythmics, gold, and phototherapeutic medications.

O: Steroids have an association with posterior subcapsular cataracts following long term use of either topical or systemic forms.

Phenothiazines – fine yellow-brown crystal deposits that progress to a star shaped opacity of the anterior lens capsule surface. Although these opacities occur in the pupillary area, they rarely affect vision.

Miotics – present as anterior subcapsular vacuoles.

Anti-neoplastics, anti-arrhythmics, gold, and phototherapeutic medications may present in various forms of cataracts.

Phosphospholine iodide, an anticholinesterase, causes cataracts in the form of anterior subcapsular vacuoles.

Amiodarone causes anterior subcapsular lens opacities.

A: Steroids, phenothiazines, miotics, anti-neoplastics, anti-arrhythmics, gold, and phototherapeutic medications.

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

INTRAOCULAR DISEASE

S: Patients may have a history of chronic intraocular inflammation or infection, retinal disease, or vitreo-retinal disease.

O: Uveitic cataracts appear as a colorful luster at the posterior pole of the lens as well as anterior and posterior subcapsular opacities. In cases of recurrent or persistent inflammation, a fibrovascular membrane may develop on the anterior lens surface affecting the optical axis.
Retinal disease such as retinitis pigmentosa (RP), Leber's hereditary optic neuropathy, gyrate atrophy or vitreo-retinal diseases such as Stickler's syndrome or Wagner's syndrome produce posterior subcapsular lens opacities.

Glaukomflecken present as fine whitish-gray anterior subcapsular opacities in the area of the pupil and result from an acute angle-closure attack.

A: Uveitis, retinal disease, vitreo-retinal disease.

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

CONGENITAL CATARACTS

MATERNAL INFECTION/DRUG INGESTION

S: Patients report that their biological mother had a history of gestational viral infection, Rubella(German measles) or ingested medications such as steroids or thalidomide during pregnancy.

O: Rubella infection during pregnancy produces congenital cataracts, which are present unilaterally or bilaterally. They appear as either a diffuse opacity throughout the lens or a dense pearly nuclear cataract that is surrounded by a less dense cortical opacity. Certain cases of rubella may exhibit microspherophakia.

Ingestion of medications during pregnancy produces cataracts, which vary in appearance according to the medication taken. In particular, long-term use of steroids may yield posterior subcapsular cataracts.

A: Rubella, Steroids, Thalidomide

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

BIRTH TRAUMA

S: Patients report a history of trauma at birth

O: Cataract appearance secondary to birth trauma depends on the type of trauma.

A: Trauma

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

METABOLIC/SYSTEMIC DISEASE

S: Patients report a history of Alpha Galactosidase A or no known related history of disease.

O: Alpha Galactosidase A cataract (refer to previous section on acquired cataracts related to metabolic/systemic disease).

A: Alpha Galactosidase A

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

GENETIC

S: Patients report a history of Lowe's oculocerebrorenal syndrome, or Down's syndrome or no known systemic disease.

O: Lowe's syndrome is associated with congenital cataracts. The lens appears thin and small and is referred to as microphakic. Opacities may be present in the capsule, cortex or nucleus.

Down's syndrome is associated with various forms of cataracts. (Refer to previous section on acquired cataracts related to genetic etiologies). The cataracts usually have the potential of causing decreased vision.

Congenital nuclear cataracts without systemic association may present in two forms. The first is termed the embryonal or nuclear cataract (cataracta centralis pulverulenta) which contains small star-shaped opacities that are located in the embryonal nucleus. The fetal nucleus is not affected. This type of cataract has a dominant inheritance pattern, is usually bilateral and does not affect vision adversely. The second is termed the total nuclear cataract. This type of cataract affects both the embryonal and fetal nucleus and therefore may affect vision adversely.

A: Lowe's syndrome, Down's syndrome

P: As any type of opacification, the cataract is monitored until the decrease in visual function affects daily activity. Referral for surgery is based on an individual patient basis.

OCULAR MALDEVELOPMENT

S: Patients report a history of Peter's anomaly or no awareness of a related anomaly.

O: A cataract associated with Peter's anomaly appears in conjunction with a central corneal defect which is in apposition to the lens. The anomaly is referred to as keratolenticular strands.

A: Peter's Anomaly

P: As any type of opacification, the cataract is monitored until the decrease in visual acuity affects daily function. Referral is based on an individual patient basis.

ABNORMALITIES IN LENS SHAPE & POSITION

S: Patients may complain of blurry and/or monocular double vision. There may be a history of Lowe's syndrome, Weill-Marchesani syndrome, Marfan's syndrome, Alport's syndrome, homocystinuria, hyperlysinaemia, or Ehlers-Danlos and others. Also, incomplete closure of the choroidal fissure may result in coloboma of the lens as well as other ocular structures (optic nerve, retina, iris, eyelid).

O: Lowe's - may display posterior lenticonus in which the posterior pole of the lens is conical. Spherophakia may be present which is defined as an abnormally short lens radius of curvature.

Weill-Marchesani - may display spherophakia, subluxation of the lens due to defective lens zonules. After subluxation, the lens may dislocate anteriorly.

Marfan's - may present with microspherophakia. Due to defective lens zonules, patients may present with lens subluxation. The lens is usually displaced upward and this is referred to as ectopia lentis.

Alport's - may present with an anterior lenticonus where the anterior pole of the lens is conical. Anterior lenticonus produces irregular astigmatism.

Homocystinuria - may present with subluxation of the lens due to defective zonules. Lens is usually displaced downward.

Hyperlysinaemia - may present with microspherophakia.

Ehlers-Danlos - may present with subluxation of the lens.

Anomalous, incomplete closure of the choroidal fissure may result in a lens coloboma. This variation presents as a notch at the equator of the lens with associated defect of the local zonules.

A: Lowe's, Weill-Marchesani, Marfan's, Alports, Homocystinuria, Hyperlysaemia, Ehlers-Danlos

P: Treatment is dependent on anomaly and presentation.