LENS

The lens is a transparent biconvex crystalline mass placed b·w the iris and the vitreous in a saucer shaped space called the patellar fossa.

FUNCTIONS OF LENS

- It transmits and refracts the light
- It absorbs UV light of < 350 nm wavelength
- Contributes to 35% of refractive power of eye
- helps in accomodation

Anterior and Posterior Pole

- The centre of anterior and posterior surface is called the anterior pole and posterior pole respectively
- Anterior pole is 3mm from centre of cornea

Refractive Properties of Lens

- Refractive index = 1.39
- Refractive power : 16-17 D

Anatomy of Lens

- Lens Capsule
- Lens epithelium
- Lens fibres

Lens Capsule

- Thin, transparent hyaline cartilagenous membrane surrounds the lens
- Lens capsule is elastic but doesn't have any elastic fibers
- Produced continuously throughout life
- Produced by basal portion of epithelium anteriorly and posterior lens fibers posteriorly
- Thicker anteriorly than posteriorly
- Thicker at equator than poles
- Thinnest at posterior pole

* Posterior capsular rupture is a complication of cataract surgery bcz. posterior capsule is weakest at posterior pole

Lens Epithelium

- There is no posterior epithelium left after embryonic period (Posterior epithelium of lens vesicle is used up to form the primary lens fibers during embryonic period)

Anterior Lens Epithelium

- Cuboidal nucleated epithelium cells below the lens capsule
- Most metabolically active part of lens
- Epithelium of equatorial region → columnar cells → actively dividing cells

Zones in epithelium

1- Central zone
2- Peripheral zone
3- Germinative / Equatorial zone

Central Zone

- Central cells: cuboidal in shape
- Decrease with age
- stable, no mitosis normally
- can show mitosis in pathologic condition

Glaukomaflecken in Acute Angle Closure Glaucoma → metaplasia of cuboidal cells into myofibroblasts

Intermediate / Peripheral Zone

- smaller and more cylindrical
- located peripheral to central cells
- rarely undergo mitosis

Bow Region

- The newly laid lens fibers elongate
- Nuclei are more anterior to the nuclei of superficial cells

Posterior Capsular Opacification

- Residual epithelial cells migrate posteriorly
  ↓
  Differentiate into a balloon like / wing cell
  ↓
  Wedl cell
  ↓
  Elshnig pearl

- Sometimes donut shape configuration
  ↓
  Soemmering's Rings

Zonal Arrangement of Lens Fibres

Lens
↙ ↘
Nucleus Cortex

Primary Lens Fibers → develop before 3 months of age, from posterior epithelium

Secondary Lens Fibers → develop from equatorial zone / germinative zone

- The central nucleus of lens → oldest cells
- The periphery of cortex → youngest cells

Fibers of lens are split into regions depending on age & origin

- Embryonic Nucleus → 3 months of embryonic life
- Fetal Nucleus → 3-8 months of fetal life
- Infantile Nucleus → last month of intra uterine life till puberty
- Adult Nucleus → corresponding to the lens in adult life
- Cortex consisting of youngest fibers

Suspensory Ligaments / Zonules

- Lens is held in place by suspensory ligament or zonule of Zinn
- Consist of bundles of strands from surface of ciliary body to equatorial capsule where they join with the zonular lamella
- Majority of zonules arise from posterior end of pars plana up to 1.5mm from ora serrata
- Suspensory zonular complex is divided into 4 zones

1. Pars orbicularis → lies on pars plana
2. Zonular plexus → lies b/w ciliary process in region of pars plicata
3. Zonular fork → point of angulation of zonules at midzone of ciliary valleys
4. Zonular Limbs
   - Anterior zonular limb → pass from pars plana to pre-equatorial part of lens
   - Posterior Zonular Limb → pass from pars plicata to post equatorial part of lens
   - Equatorial Zonular Limb → pass from pars plicata to lens equator

Surgical Anatomy of Lens

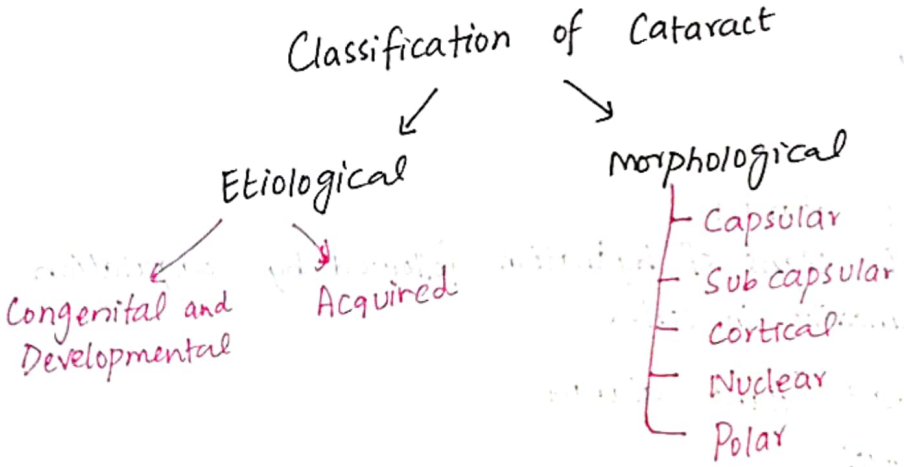
- Central hard nucleus
- Epinuclear plate of variable thickness
- A layer of cortex
- Capsule

Hardness of Lens based on its color

- Grade I: Grey
- Grade II: Amber, greenish yellow
- Grade 3: Yellow
- Grade 4: Brown / Brunescent cataract
- Grade 5: Black

CATARACT

- Opacification of lens is known as cataract



* Congenital and Developmental cataract → due to disturbance in normal lens development i.e. formation of lens fibres
  - Developmental opacities are partial and stationary

* Acquired Cataracts → due to degeneration of already formed lens fibres
  - Acquired opacities progress until entire lens is involved

Senile Cataract

- It is related to aging
- caused by lifelong exposure to sunlight or uv light
- Rare in persons younger than 50 yrs unless associated with some metabolic disturbance such as diabetes

Risk Factors

- Age > 50 yrs
  If cataract develops before 45 yrs → Pre senile cataract
- ~~Gender male~~
- uv radiation
- Dietary factors → deficient in proteins, amino acids, vitamins (Riboflavin, Vit C, E)
- Dehydration Crisis → prior episodes of severe dehydration (diarrhea, cholera)
- Smoking
- Cyanates

Pre-senile Cataract Risk Factors

- Hereditary
- Diabetes mellitis
- Atopic Dermatitis
- Myotonic Dystrophy

TYPES OF SENILE CATARACT

1. Cortical Cataract (Soft Cataract)
   wherein the classical signs of hydration followed by coagulation of proteins appear primarily in cortex
2. Nuclear / Sclerotic Cataract (Hard Cataract)
   - slow sclerosis in nucleus

STAGES OF MATURATION OF CORTICAL CATARACT

1- Lamellar Separation

- Demarcation of cortical fibres owing to their separation by fluid
- Lamellar separation can be seen only with a slit lamp and is invisible ophthalmoscopically
- A grey appearance to pupil seen
- General increase in refractive index of cortex in old people
- Increase in reflection of scattering of light

2. Incipient Cataract (cuneiform vs cupuliform)

- Early detectable opacities with clear areas b.w them appear in the periphery of the lens
- Sectorial alterations in the refractive indices of lens fibres, thus producing irregularities in refraction, some visual deterioration and polyopia

Cuneiform Cataract:

- wedge shaped opacities with clear areas in between
- extend from equator towards the centre
- can be demonstrated on dilatation of pupil
- seen in lower nasal quadrant first

- With oblique illumination, the opacities appear grey whitish color
- In retro illumination, they are black against the red background of fundus

Cupuliform Cataract
- saucer shaped opacity develops
- just below the capsule in central part of posterior cortex
- Posterior Sub capsular cataract
- It gradually extends outwards
- Lies in pupillary axis so vision is affected early
- Near vision is affected more than distant vision

3. Immature Cataract
- opacification further continues but is not complete
- Lens appears greyish white in color but clear cortex is still present
- This is called stage of immature senile cataract

* Intumescent Cataract
- In some patients at the stage of immaturity
- Lens gets excessively hydrated and swollen. This is called intumescence
- Anterior chamber becomes shallow.
- ↑ osmotic pressure inside the lens
- Lens continue to absorb an increasing amount of aqueous and become swollen with stretched glistening capsule

[Shallow Anterior Chamber → pupillary block Risk of Phacomorphic Glaucoma]

4. Mature Senile Cataract
- opacification is complete
- whole cortex is involved
- no iris shadow seen
- intumescence continues
- pearly white color
- Also called 'ripe cataract'

5. Hypermature Senile Cataract → when mature cataract is left in situ

- Morgagnian Hypermature Cataract
- Sclerotic Hypermature cataract

* Morgagnian Hypermature Cataract
  - The whole cortex liquefies
  - Small brownish nucleus may sink to the bottom of lens
  - The liquefied cortex is milky, and the nucleus is seen as a brown mass limited above by a semicircular line, altering its position with changes in position of head
  - Sometimes calcium deposits can be seen on lens capsule

* Sclerotic Hypermature Cataract
  - The cortex becomes disintegrated and is transformed into a pultaceous mass
  - shrunken and inspissated lens
  - Thickened anterior capsule due to proliferation of anterior cubical cells, so that a dense white capsular cataract is formed at the anterior pole in pupillary area
  - The lens and iris become tremulous (phacodonosis and iridodonosis)
  - The anterior chamber deep, and finally, degeneration of suspensory ligament may lead to luxation of lens

PEDIATRIC CATARACT

Pediatric Cataract occur due to some disturbance in ~~gro~~ normal growth of lens

- Congenital Cataract
- Developmental Cataract

CONGENITAL CATARACT

- when disturbance in lens growth occurs before birth
- opacities are present at birth but usually diagnosed within FIRST year of life
- limited to either embryonic or fetal nucleus

DEVELOPMENTAL CATARACT

- when disturbance in lens growth occurs from infancy to adolescence
- Opacities involve the infantile or adult nucleus, deeper parts of cortex or capsule

UNILATERAL PEDIATRIC CATARACTS

- Ocular Anomalies / Local Dysgenesis
  - Persistent Fetal Vasculature
  - Posterior Lenticonus
  - Posterior Segment Tumor
  - Posterior Segment Pathology
  - Retinal Detachments of any cause
  - Uveitis
- Radiation Exposure
- Trauma to eye (Rosette Cataract)
- Idiopathic

- NOT inherited
- mostly not related to systemic disorders
- NOT related to metabolic disorders

METABOLIC CATARACT

BILATERAL CATARACTS

- Genetic mutation
- Metabolic Syndromes
- Chromosomal Anomalies
- TORCH Infection
- Systemic Syndromes

GENETIC MUTATION

Isolated inherited congenital cataracts carry a better visual prognosis than those with co existing ocular and systemic abnormality

Galactosemic Cataract

- This is an autosomal recessive, inherited congenital disease characterized by an inborn inability of the infant to metabolize galactose
- Oil Drop Cataract

Wilson Disease

- Hepatolenticular Degeneration
- Inherited disorder of copper metabolism due to mutation of ATP7B gene
- Affects eye, liver and basal ganglion
  - ↑ copper levels
  - ↓ ceruloplasmin Levels
- Kayser Fleisher Ring
- Sunflower Cataract

Fabry Disease

- X linked lysosomal storage disorder
- Deficiency of enzyme α-galactosidase A
- This leads to abnormal tissue accumulation of a glycolipid
- Angiokeratomas
- Verticellata / Vortex Keratopathy
- Corkscrew vessels

Pg. 30

FABRY'S DISEASE

(Mnemonic FABRYC)

- Foam cells / Febrile episodes
- Alpha Galactosidase A Deficiency / Angiokeratomas
- Burning pain in hands and feet "Peripheral Neuropathy / Boys (X linked)
- Renal Failure
- XY genotype (male, X linked Recessive)
- Ceramide trihexoside accumulation / Cardiovascular disease

Lowe Disease

- Lowe Syndrome (oculocerebrorenal) syndrome
- X-linked recessive (gene OCRL1) inborn error of amino acid metabolism
- Neuromuscular, renal and other manifestations
- Posterior Lenticonus

True Diabetic Cataract

- Glucose → Aldose Reductase
- Sorbitol : retained within lens → ↑ osmotic gradient
- water entry / hydration of lens → Vacuole formation and swelling
- Opacification and cataract
- Diabetic Snowflake Cataract

Hypocalcemia / Parathyroid Tetany

- Atrophy
- Removal of parathyroids
- Hypocalcemia

This effects the membrane of lens basically

SYNDROMIC CATARACTS

- Musculoskeletal Syndromes
- Craniofacial Anomalies
- Chromosomal Disorders
- Dental Disorders
- Renal Syndromes
- Skin Disorders

MUSCULOSKELETAL SYNDROMES

Myotonic Dystrophy

- Christmas tree cataract
- ~~Bo~~ (stellate morphology)

Smith Lemli Opitz Syndrome

- microcephaly with bitemporal narrowing
- A short upturned nose with anteverted nares
- Long philtrum
- Unilateral or bilateral ptosis, epicanthus
- Retrognathia
- Polydactyly
- Syndactyly
- Short Stature

Conradi Hunermann Syndrome

- Short stature
- patchy alopecia
- scoliosis
- Asymmetric limb shortening
- microphthalmos microcornea cataracts

Weil Marchesani Syndrome

- Spherophakia - brachymorphia syndrome
- Stocky build and have small, stubby fingers
- Brady dactyly
- microspherophakia
- Anterior dislocation of lens

CRANIOFACIAL ANOMALIES

Hallerman Strief Francoid Syndrome

- Abnormal facial appearance
- Bird like facies
- Dental abnormalities
- Hypotrichiosis
- Skin Atrophy
- Proportionate short stature
- Ophthalmic Features including microphthalmia and congenital bilateral cataracts

Rubinstein Taybi Syndrome

- Broad First toe
- Broad Thumb
- Clinodactyly
- Microcephaly

Bardet Beidel Syndrome

- Retinal degeneration
- Truncal obesity
- Cognitive impairment
- Post axial polydactyly
- Hypogonadism / genito urinary anomalies
- Renal abnormalities

RENAL SYNDROMES

1. ALPORT Syndrome

2. Lowe Syndrome

- Oculocerebrorenal syndrome of Lowe
- male children
- bilateral congenital cataracts
- Associated with Lenticonus
- severe hypotonia
- proteinuria may be most sensitive marker for renal involvement of LS
- Fanconi Syndrome

TORCH

T Toxoplasma
O Other pathogens (syphilis) · varicella
R Rubella
C Cytomegalo virus
H Herpes simplex Virus

Rubella Cataract

- pearly nuclear or more diffuse unilateral or bilateral cataract occur in around 15%.
- Salt and pepper retinopathy

CHROMOSOMAL DISORDERS

Trisomy 21 / Down's Syndrome

- wide gap b.w first and second toe
- clinodactyly, single palmar creare
- epicanthus, upward slant, flat naral bridge
- Blue Dot Cataract
- Symmetrical opacities and often develop in late childhood
- Other features include
  - Brush Field Spots
  - Keratoconus
  - Glaucoma

Trisomy 18 / Edwards Syndrome

- Cataract include ptosis
- Microphthalmos
- Corneal opacity
- Uveal and disc coloboma
- Vitreoretinal dysplasia

Cri-Du-Chat Syndrome

- Downward slant
- widely set eyes (hypertelorism)
- Low set ears
- Small jaw, and a rounded face

SKIN DISORDERS WITH CATARACTS

- Cockayne Syndrome
- Rothmund-Thomson
- Atopic dermatitis
- Incontinentia pigmenti
- Progeria
- Ichthyosis

Atopic Dermatitis Cataract

- Bilateral and may mature quickly
- Shield-like dense anterior subcapsular plaque that wrinkles the anterior capsule is characteristic
- Posterior subcapsular opacities may also occur

DRUGS USED IN GLAUCOMA

Glaucoma: ↑ IOP (>21 mm Hg)

Normal IOP = 10-21 mm Hg

↑ IOP

↑ production of Aqueous Humor

Less Drainage

Drugs

↓ production of Aqueous Humor

- β-blockers
- α_2 Agonists
- Carbonic Anhydrase Inhibitor

↑ drainage

↑ Uveoscleral Outflow

- Prostaglandins
  - Latanoprost
  - Bimatoprost

↑ Trabecular Outflow

- Miotics
  - Pilocarpine
  - Physostigmine
- α_2-Agonists
  - Dipivefrine

β-Blockers

Cardioselective
- Betaxolol
- Levo-Betaxolol

Non-selective
- Timolol
- Levobunolol
- Carteolol

α_2-Agonists
- Brimonidine
- Apraclonidine

Carbonic Anhydrase Inhibitors
- Acetazolamide
- Dorzolamide
- Brinzolamide

DRUGS USED IN PRIMARY OPEN ANGLE GLAUCOMA

Ciliary process Receptors

β_2 Receptors (vasodilation) → Amount of blood reaching ciliary body increases → ↑ secretion of Aqueous humor

α_2 Receptors (vasoconstriction) → ↓ secretion of Aqueous humor

- So β-blockers and α_2 Agonists will decrease aqueous humor production

* β-Blockers
  - First line Drugs
  - Approved for use in Glaucoma
    - Timolol
    - Betaxolol → cardioselective (less efficacious but safe in asthmatics)
    - Levobetaxolol
    - Carteolol
    - Metipranolol
    - Levobunolol → Longest Acting

* Prostaglandin Analogues → Drugs of choice for POAG

  PGF2α → ↑ Uveoscleral outflow

  PGF2α Derivatives
  - Latanoprost
  - Bimatoprust
  - Unoprostone

* Non Selective α-Agonists → Act by ↑ trabecular outflow
  - Dipivefrine (prodrug of adrenaline)
  - Adrenaline

  - Dipivefrine can cause cystoid macular edema

* Selective α_2 Agonists
  - Apraclonidine
  - Brimonidine

  Act by ↓ aqueous secretion

- Apraclonidine can cause lid retraction
- Brimonidine associated with anterior uveitis

* Carbonic Anhydrase Inhibitors
  - Acetazolamide (oral) → ↓ Aqueous humor secretion
  - Brinzolamide (Topical)
  - Dorzolamide (Topical)

* Miotics → ↑ aqueous outflow by causing miosis
  - Pilocarpine (Directly acting cholinomimetic) → short acting
  - Physostigmine (Indirectly Acting Cholinomimetic)

- Long Acting Cholinomimetics
  - Demacarium → Rarely used bc they accelerate cataract formation
  - Ecothiophate

DRUGS USED IN ANGLE CLOSURE GLAUCOMA

In Angle closure glaucoma, the iris is abnormally positioned so as to block aqueous outflow through the anterior chamber (iridocorneal angle

Primary Treatment: Surgery → (Laser Peripheral Iridotomy) → opening of iris
↳ Surgical peripheral Iridectomy

Before Surgery, IOP should be reduced

- Cholinomimetics (miotics)
- Acetazolamide
- Osmotic Diuretics (mannitol)

Acute Cases: IV Acetazolamide

All patients with primary acute angle-closure glaucoma should undergo prophylactic laser peripheral iridotomy to the unaffected eye.