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Anatomy, Physiology and Congenital anomalies of the lens

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Case History

- Name: abc
- Age: 60 years
- Sex: male
- Address: Peshawar
- Profession: Farmer
- Date of admission: 25\textsuperscript{th} of Jan, 2005
Chief complaints

• Gradual Dimness and blurring of vision in both eyes more in the left-----------------2 years
Slit lamp examination

- Opacity seen in the cortex of the lens
Embryology of the lens

• Embriologically lens is first seen as a thickening of surface ectoderm – lens placode at 22nd day of gestation
• It overlies the optic vesicle
• The lens placode invaginate and sink to form the lens vesicle
• Single layered cells covered by basal lamina
• The cell forming the posterior wall elongates and are filled with protein – crysatalin
• Responsible for transparency
• These elongates-- primary lens fibers
• The nuclei of the lens fiber move anteriorly-- nuclear bow
• Additional lens fibers formed by mitotic division of the anterior epithelial cells – secondary lens fiber that persist throughout the life
Lens capsule

• The capsule is formed from the mesenchyme that surrounds the lens
• Supplied by the hyaloid artery
• Later on this blood supply regresses and vascular capsule disappear before birth
Continued…

• Lens fibers run completely from anterior to posterior surface and come to apposition at sites – sutures
• The anterior sutures – upright Y
• Posterior suture – inverted Y
• In fetus growing lens is supplied by hyaloid artery
• Its spherical, soft reddish tent
• In infants anteroposterior diameter is nearly that of an adult
• Its equatorial diameter is about 2/3 of that reached in the adult
Gross Anatomy

- Lens is a transparent and biconvex situated in between the iris & vitreous.
- Two surfaces: ant & post
- Ant less curved, post is more curved.
- Two poles: ant & post
- Axis: line joining the ant & post poles
- Equator is the marginal circumference of the lens
Continued…

• Diameter of the Lens  . Adults 10mm
• Thickness of Lens  . 4mm
• Diopteric power  +15
• Aging decreases the dioptic power.
• Lens totally depends on Aq/vit for nourishment.
Structure of the Lens

• Three parts:
  – Capsule
  – Lens epithelium
  – Lens fibers
• Capsule:
  – Capsule is an elastic basement membrane that envelops the entire Lens.
  – It is thickest on the ant & post surfaces close to the equator-- 20 um
  – Thinnest at the posterior pole-- 3um
The inner surface of the ant capsule is in direct contact with the lens epithelium.

Posteriorly it contacts the superficial lens fiber cells.

The thickest basement membrane is formed by the lens epithelium anteriorly and by the superficial lens fiber posteriorly.
Continued…

• Capsule has homogeneous appearance consist of 40 lamellae
• It consist of type iv collagen
• Elastic capsule can be stretched up to about 60% of its circumference.
• In equater capsule attached with zonular fibers and cilliary processes
Continued…

- It serves as diffusion barrier
- Capsule chief function to mold the shape of the lens during accommodation
Lens Epithelium

- Found only on the anterior surface
- Cells are cuboidal and lies beneath the capsule
- At the equator these cells elongate and form columnar cells
- Arranged in meridional rows
- At the equator lens mitotic activity is at maximum
Continued…

• Absence of the dividing epithelium cells on the posterior capsule acts as function of the lens epith.
• Lens epithilium----the transport of substances from aqueous into lens
• Secretion of the capsular material.
Lens fibers

- Main mass of the lens
- Lens epithelial cells differentiate into fibers; this process continues throughout the life of the lens.
- As the cells progressively elongate anteriorly, the nucleus moves anteriorly.
- Lens bow
• Each cell become a fiber
• Central fibers become nucleus of the lens and outer fibers become cortex of the lens
• Sutures are formed by fibers- anterior Y & posterior Inverted Y
Continued…

• Continues production of lens fiber from embryonic stage
• Earliest fiber mass in the center-embryonic nucleus followed by fetal nucleus
• Adult nucleus those fibers that formed after the birth
• The newly formed nucleated fibers- lens cortex
• Fibers are tightly packed
• During development lens fibers lose there nuclei and the cytoplasm organelles – crystallin.

• Alpha and beta – 60% of the lens fibers

• High refractive index is due to crystallin

• It differs in different parts of the lens

• It compensate spherical and chromatic aberrations
Suspensions

• The lens is held in position by radially arranged fibers—suspending ligaments or zonule

• The zonule fibers arise from the epithelium of the ciliary process.

• The larger bundles are straight-ant zonular sheet the smaller fibers curve backward- post zonular sheet
Physiology of lens

• Lens plays a role in the process of accommodation by which light rays are focus on the retina.

• It’s the remarkable transparency of the lens that allows it to play this important role in the normal vision.
Continued...

• Lens is avascular and receives its nutrition from aqueous & vitreous.

• It is important in
  – To maintain its own clarity.
  – To provide ref. power.
  In accommodation.
  – To absorb UV rays.
• Lens epithelial cells have the metabolic capacity to carry out all the normal cell activities. Including DNA, RNA, Protein and Lipid Biosynthesis.

• Glucose metabolism-energy-ATP-lens growth-transparency.

• ATP-transport of

  » Electrolytes

  » Amino acids
Glucose Metabolism

• Glycolytic pathway. Anaerobic - 1mole glucose--2mol ATP
• Kreb cycle. Aerobic. 1mole glucose-36ATP.
• HMPshunt. NADPH 5% of Glucose by this route.
• Sorbitol.
• Glucose metabolism is regulated by hexokinase.
Electrolytes transport

• Maintenances of ionic balance depends upon the communications between epithelial cells and fibers.
• Energy is required for this process
• Internal Na⁺ is maintained about 20mMol & K⁺ 120mMol.
• The respective concentrations in aqueous are 150mMol & 5mMol.
Continued...

- Sodium ions extrusion. Less outward.
- Potassium ions intake. More inward.
- Na+/K+ pump is regulated by Na+, K+, ATPase.
- 1 Mole of ATP pump, 3 Mole of Na outward & 2 Mole of K inward.
- The major site of active cation transport is ant. Epith. Of lens.
- Amino acids are actively transported into the lens.
Continued...

- Active transport of Na, K, amino acid and proteins occurs through the ant. Epith. & diffuse out with the concentration gradient through the back of the lens.
- K+ is more concentrated in the anterior lens & Na+ in posterior.
- Manipulation may inactivate the energy dependent enzyme pumps.
Glutathione

- It is synthesized in lens mostly in reduce form.
- Levels decrease with age.
- It helps to preserve the equilibrium of lens protein.
- To maintain transport pump & mol. Integrity of lens fiber membrane.
- It plays a central role in protecting the lens from the oxidative insult (free radical).
- Oxidative damage — cat. Formation
Continued...

- **Lens mainly consists of**
  - 66% water
  - 33% protein

- **Lens protein**
  - soluble 85% - cortex
    » 15% alpha crystalline
    » 55% beta crystalline
    » 15% gamma crystalline
  - Insoluble 15% - nucleolus
Accommodation

• It is the ability of eye to change its diopteric power.

• Near accommodation
  – Contraction of the ciliary's muscles
    » Choroid & cilary body moves forward
    » Zonules inward towards lens
    » Increases the convexity of lens
    » Diopteric power increases
Continued...

• Accommodation far distance
  
  Relaxation of ciliary muscles
  – Zonules contracts
  – Increases diameter of lens
  – Convexity decreases
  – Diopteric power decreases

• Lens diopteric power is 15 D
• Ref. index is 1.39
• PH is 6.9
Continued…

- Accommodation develops two months of age
- 6—8 months of age acc. Well dev.
- Max in childhood
- Decreases in middle age
- Amplitude depends upon background luminance
  - Dec. lumin.dec.amplitude
Lens transparency

- High content of protein
- Regular arrangement of lens fibers
- Lamellar conformation of protein
- Even distribution of protein
- Short range order of crystalline
- Metabolism of lens
- Electrolytes balance
Purkinje images

• It is 4 in number
  • Ref. from ant. surface--cornea
  • Ref. from post. surface--cornea
  • Ref. from the ant. surface--lens
  • Ref. from the post. surface--lens
Congenital abnormalities

- Shape
- Size
- Position
- Transparency
Shape—Anterior lenticonus, posterior lenticonus
Shape--Coloboma
Coloboma of Iris and Lens
Size--Microphakakia
Size--Microspherophakia
Position--ECTOPIA LENTIS

- It refers to displacement of lens from its normal position.
- The lens may be completely dislocated (luxated) from the pupillary space or partially displaced (subluxated).
ECTOPIA LENTIS

Dislocation or luxation when lies completely outside the lens patellar fossa and may appear in one of the following areas;

1. Incarcerated in the pupil.
2. In the anterior chamber.
3. In the vitreous.
4. Sub retinal space.
5. Remain as wandering lens.
6. Extruded out of the globe partially or completely.
ETIOLOGY

• A. Hereditary.

• B. Traumatic subluxation/ dislocation.

• C. Secondary to other ocular conditions.
Hereditary

a) Isolated forms;
   1. Simple ectopia lentis.
   2. Simple microspherophakia.

b) With associated ocular anomalies;
   1. Ectopia lentis et pupillae.
   2. Aniridia.
MARFAN SYNDROME

• Limbs are long compared to trunk.
• Pubis to sole measurement is in excess to the pubis to vertex measurement.
• Arm span is in excess of the height.
• Arachnodactyly (fingers are long and spider like).
• Pectus excavatum.
• Joint laxity.
HOMOCYSTINURIA

- Tall, slim build.
- Arachnodactyly may occur.
- Vertebral osteoporosis.
- Scoliosis.
- Increased platelet stickiness.
HOMOCYSTINURIA

- Increased risk of thromboembolism, both arterial and venous.
- Partial or complete vascular obstruction in various organs.
- Fine and fair hair, fair complexion, mental retardation (50%), seizures, spastic gait, muscular weakness, chest deformities and erythematous patches on the skin of face (malar flush).
WEILL-MARCHESANI SYNDROME

• Short stature.
• Short, small and stubby fingers.
• Limited joint mobility.
• Well developed muscular appearance.
• Thick skin.
• Brachycephaly.
• Mental retardation.
HYPERLYSINAEMIA

- Lax ligaments.
- Hypotonic muscles.
- Seizures.
- Mental handicap.
- Microspherophakia.
- Occasionally superior subluxation.
SULPHITE OXIDASE DEFICIENCY

Very rare, AR inheritance.
Disorder of sulphur metabolism.
Characterized by ectopia lentis, poor feeding, neurological abnormalities, seizures, myoclonus, decerebrate posturing, mental retardation and death usually before the age of 5 years.
EHLER-DANLOS SYNDROME

- Rare, AD hereditary disease.
- Disorder of collagen caused by deficiency of hydroxylysine.
SIMPLE ECTOPIA LENTIS

May be congenital or spontaneous.

- Congenital type is AD inherited, occurs early in life, bilateral and symmetrical. Displacement is upward and temporally. Defect in fibrillin gene on chromosome 15.

Spontaneous type occurs at any time during adult life (between 20-65 years). Displacement is often downward. Glaucoma is common.
Transparency

• Pathology—Any opacity in the lens or its capsule is called CATARACT.

• Clinically—Any opacity in the lens or its capsule interfering with vision is called CATARACT.
Transparency

LENSES OPACITIES (CATARACTS)

• Congenital
• Acquired
Thanks