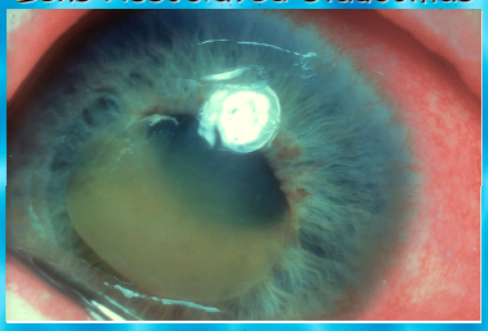
Lens Associated Glaucomas



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Lens Associated Glaucoma

- Lens-induced open angle glaucoma
  - Phacolytic
  - Phacotoxic, phacoanaphylactic
  - Lens particle
- Lens-induced angle closure
  - Phacomorphic
- Lens dislocation

Lens Associated Glaucoma

- Post cataract extraction
  - Retained lens cotex
  - IOL-related
  - Post Nd:Yag posterior capsulotomy

Phacomorphic Glaucoma

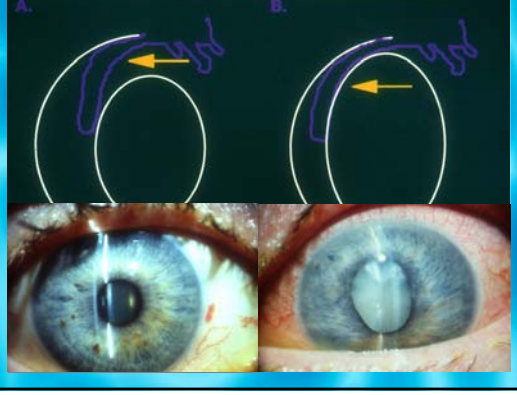
- Secondary angle closure from intumescent lens
- Pupillary block glaucoma
- Increased risk: Hyperopia
  Weak zonules
  (Trauma, Exfoliation, Age)

Phacomorphic Glaucoma (Lens Intumescence)

A. Pupillary Block
   - Central depth deeper than peripheral
   - Angle opens with laser iridotomy
   - Fellow eye usually narrow

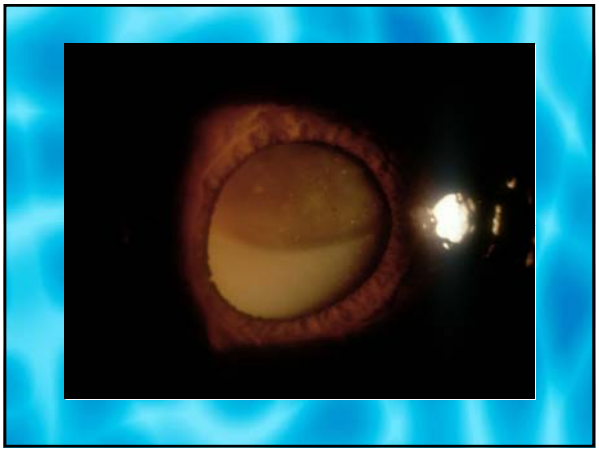
B. Mechanical angle closure from enlarged lens
   - Chamber diffusely shallow
   - Angle remains narrow after iridotomy
   - Depth fellow eye variable

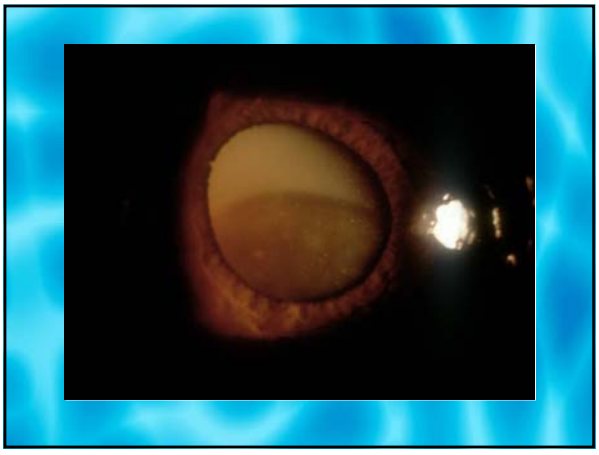
Phacomorphic Glaucoma / Mechanism



Phacolytic Glaucoma: Background

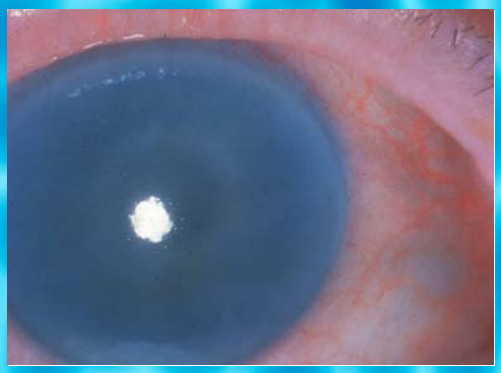
- Zeeman, 1943
- Irvine and Irvine: Blockage of TM by macrophage
- Flocks: "Phacolytic glaucoma"





Phacolytic Glaucoma

- Clinical presentation
  - Rapid onset of pain and redness
  - Marked elevation of IOP
  - Corneal epithelial edema
  - Keratic precipitates



Phacolytic Glaucoma

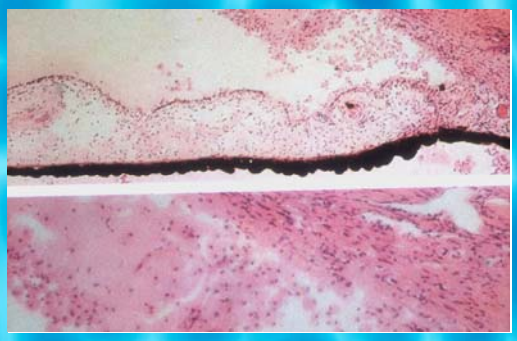
- Clinical Presentation
  - Open angle
  - Minimal-moderate AC reaction, large cells, flare
  - Mature cataract (total opacification) or
  - Hypermature cataract (liquefied cortex)
  - White patches (macs) on ant. lens surface

Phacolytic Glaucoma: Differential Diagnosis

- Primary angle closure
- Angle closure from mature cataract
- Neovascular glaucoma
- Uveitic glaucoma

Phacolytic Glaucoma

- AC Aspirate
  - HMW lens protein (not seen in ordinary cataract)
  - Macrophages



Phacolytic Glaucoma: Diagnosis

- Paracentesis: engorged macrophages
- Phase-contrast microscopy and Millipore filter
- Macrophage # does not correlate with IOP
- May not be present if corticosteroid given

Phacolytic Glaucoma

- Medical Treatment
  - Beta blocker, CAI
  - Apraclonidine
  - CAI
  - Osmotic agent
  - Topical corticosteroid
- Surgical Treatment

Phacolytic Glaucoma: Pathophysiology

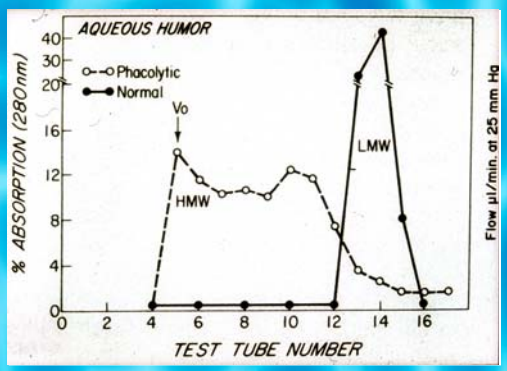
- Microscopic defects in lens capsule
- Soluble lens protein enters aqueous humor
  - (May occur intermittently)
- Obstruction of aqueous outflow

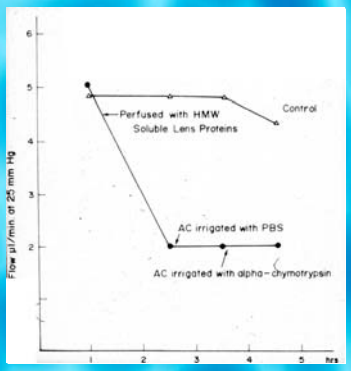
Phacolytic Glaucoma: Pathophysiology

- High molecular weight lens protein
  - > 150 million g/mol
  - Amount increases with age and cataract formation
  - Absent in infant, juvenile patients
- Usually found in nucleus, not cortex
  - Phacolytic: in liquid cortex

Phacolytic Glaucoma

- Epstein:
  - Enucleated human eyes
  - HMW lens protein infused into AC
  - Obstruction of aqueous outflow
  - Decreased outflow with increased protein perfusion
  - Obstruction not relieved by vigorous irrigation





Phacolytic Glaucoma: Role of Macrophage

- Macs observed in TM of eyes with phacolytic
- Response to lens material
- Engorged macs in aqueous of children after aspiration of cataract (no glaucoma)
- Dueker: rabbit oil-laden macs cause no IOP rise

Phacolytic Glaucoma with Immature Cataract

- Irvine and Irvine, 1952
  - Clinically immature cataract
  - Pathological exam: liquefied posterior cortex
- Chandler 1958
  - Phacolytic glaucoma induced by immature cataract
  - Relieved by lens extraction

Phacolytic Glaucoma with Immature Cataract

- Presumed localized liquefaction of posterior cortex
- Vision may be good
- Anterior uveitis, immature cataract
- Non-responsive to steroids, glaucoma meds
- Diagnostic paracentesis

Posterior Dislocation with Phacolysis

- Mild injection
- Moderate IOP rise
- Moderate AC reaction
- White patches on lens capsule

DDx of Phacolysis with Posterior Dislocation

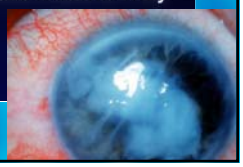
- POAG
- Angle recession glaucoma
- OAG with lens dislocation
  - (do not see white deposits)

Lens Particle Glaucoma

- Retained lens cortex
- Setting
  - Cataract extraction
  - Penetrating lens injury
  - Nd:Yag posterior capsulotomy

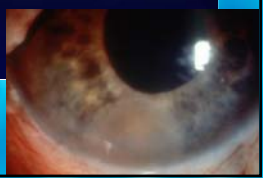
Lens Particle Glaucoma: Clinical

- Cortical lens material in AC
- Corneal edema if IOP highly elevated
- Moderate flare and cell (PMNs and macs)
- Gonioscopy: open angle, lens material may be present



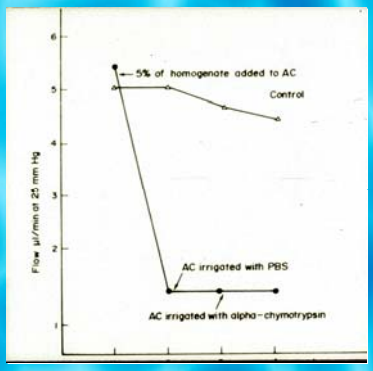
Lens Particle Glaucoma

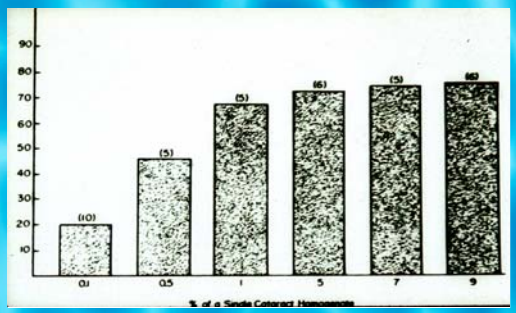
- Cortical and capsular material in TM
- Severity of glaucoma correlates with amount of lens material
- Days to weeks between insult and glaucoma



Lens Particle Glaucoma: Pathophysiology

- Epstein, Grant: Lens material obstructs TM
- Small amount can cause significant obstruction
- Role of inflammatory cells, viscoelastic
- May cause trabecular dysfunction after many years





Lens Particle Glaucoma: Therapy

- CAI, beta blocker, +/- osmotic
- Cycloplegia, corticosteroid
- Removal of lens material/vitreous

IOP Rise Post Capsulotomy

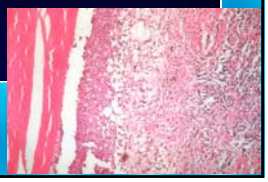
- 59-67%: > 10 mm above baseline
- 38%: > 40
- Peak after 3-4 hr; Baseline after 1 week
- Increased risk: > 200 mj total energy, pre-existing glaucoma

IOP Rise after Nd:Yag Posterior Capsulotom

- Correlates with reduced facility of outflow
- Small lens fragments or proteins in TM
- Shock wave damage to endothelium or blood-eye barrier
- Vitreous molecules pass anteriorly into TM

"Phacoanaphylaxis"

- Uncommon, granulomatous uveitis
- Response to lens material
- Rarely seen after ICCE
- Glaucoma rare; hypotony more common

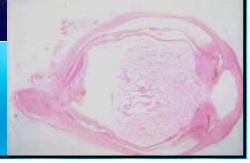


Sensitization to previously isolated lens protein

- Following extracapsular cataract extraction
- Cataract removal in one eye; subsequent cataract surgery or hypermature lens in fellow
- Traumatic rupture of lens capsule

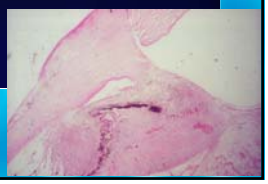
"Phacoanaphylaxis" Clinical Picture

- Hours to months after insult
- Uveitis mild to severe
- KPs on cornea and intra-ocular lens
- Marked AC reaction +/- hypopyon
- Residual lens material
- Vitritis, inflammatory membranes



Phacogenic Uveitis: DDx

- Bacterial endophthalmitis (P. acnes)
- Sympathetic ophthalmia (may co-exist)
- Other granulomatous uveitides

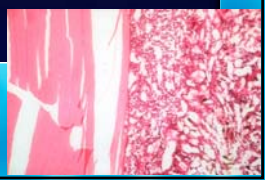


Phacogenic Uveitis: Pathophysiology

- Autoimmunity to lens proteins
- Zonal, granulomatous reaction
- PMNs, epithelioid and giant cells, monocytes
- No circulating lens proteins in AC
- Glaucoma: lens particle, inflammatory cells, PAS
- Experimental model: auto-antibodies;? role of cell-mediated immunity

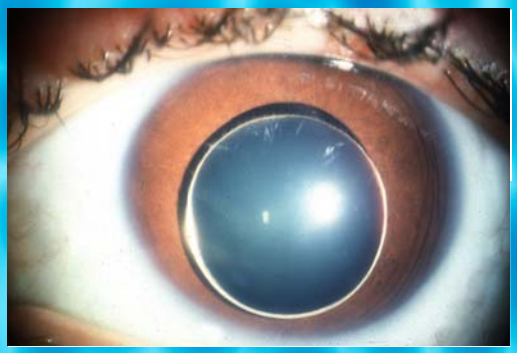
"Phacoanaphylaxis" Management

- Refractory to corticosteroids
- Surgical removal of residual lens material/vitrectomy
- Approach to cataract removal in fellow eye



Ectopia Lentis

- Displacement of lens from central position in PC
- Dislocation: No remaining zonular attachment; lens in AC, PC or vitreous
- Subluxation: Loosening of zonules, decentered in pupillary axis



Ectopia Lentis: Backgound

- 1749 Berryat: first report
- 1846 Sichel: traumatic vs spontaneous
- 1849 Arlt: congenital

Conditions Associated with Ectopia Lentis[a]

Trauma	Alport's syndrome
Simple ectopia lentis	Mandibulofacial dysostosis
Ectopia lentis et pupillae	Klinefelter's syndrome
Marfan's syndrome	Retinitis pigmentosa
Homocystinuria	Persistent pupillary membrane
Weill-Marchesani syndrome	Axenfeld-Rieger syndrome
High myopia	Dominantly inherited blepharoptosis and high myopia
Uveitis	Marfan-like syndrome with hyaloretinal degeneration
Buphthalmos	Sturge-Weber syndrome
Megalocornea	Syphilis
Ehlers-Danlos syndrome	Crouzon's disease
Hyperlysemia	Refsum's syndrome
Sulfite oxidase deficiency	
Aniridia	
Scleroderma	

Ectopia Lentis: signs

- Iridodonesis
- Phakodonesis
- Asymmetry in AC depth (between eyes or quadrants)
- Peripheral iris concave, central iris conical)
- Iris bombe if pup. block present
- Zonules attached to lens capsule or retracted

Ectopia Lentis: Signs

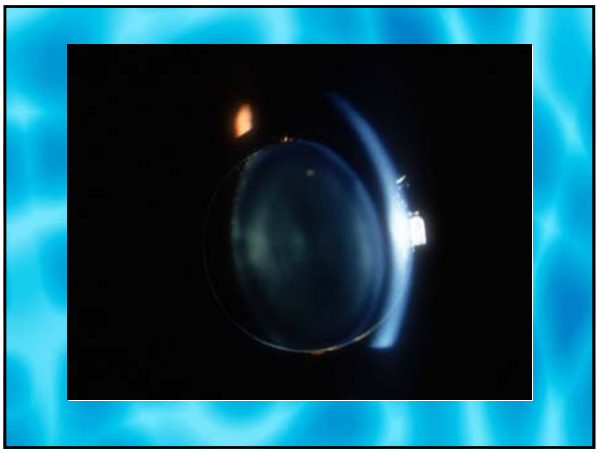
- Lenticular myopia (increased AP length, ant. displacement)
- Lenticular astigmatism (tilting, decentration)
- Variable astigmatism
- Loss of accomodation
- Monocular diplopia, quadriplopia
- Amblyopia

CAUSES OF GLAUCOMA IN THE PRESENCE OF ECTOPIA LENTIS

I. Lens related
   A. Pupillary block by lens
   B. Pupillary block by lens and vitreous
   C. Pupillary block by vitreous
   D. Lens in anterior chamber
   E. Phacolytic glaucoma
   F. Secondary open-angle glaucoma caused by repeated attacks of angle-closure
   G. Peripheral anterior synechiae caused by chronic angle-closure
II. Lens unrelated
   A. Angle recession
   B. Chamber angle anomaly
   C. Coincident primary open-angle glaucoma
   D. Other forms of glaucoma related to an underlying disease process (e.g., ghost cell glaucoma, neovascular glaucoma)

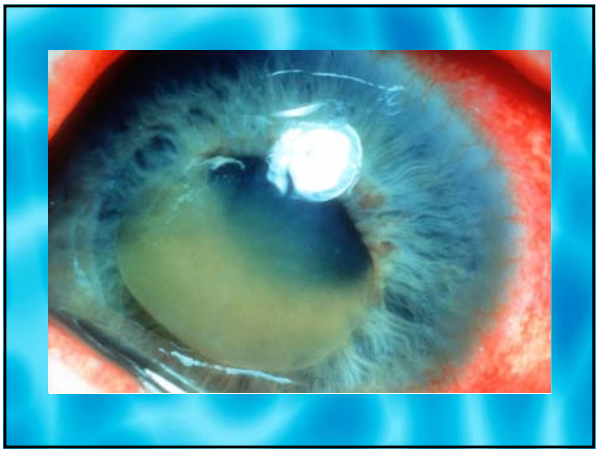
Anterior Dislocation of the Lens

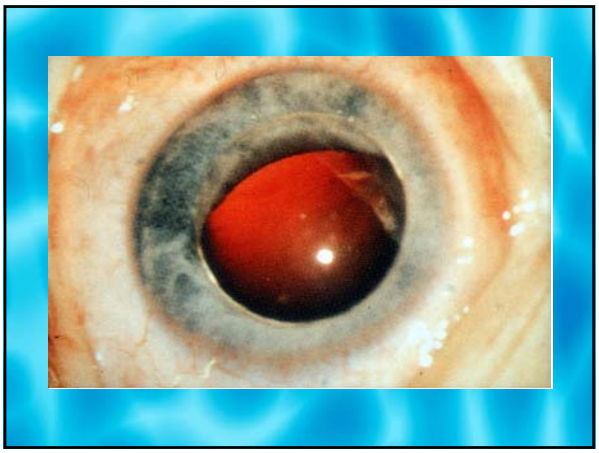
- Marfan's syndrome
- Homocystinuria
- Spherophakia

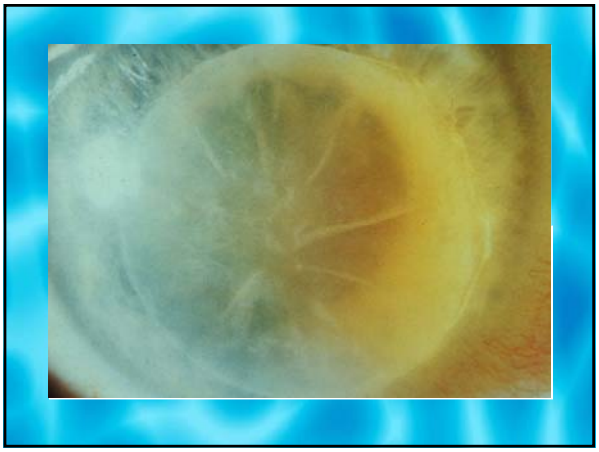


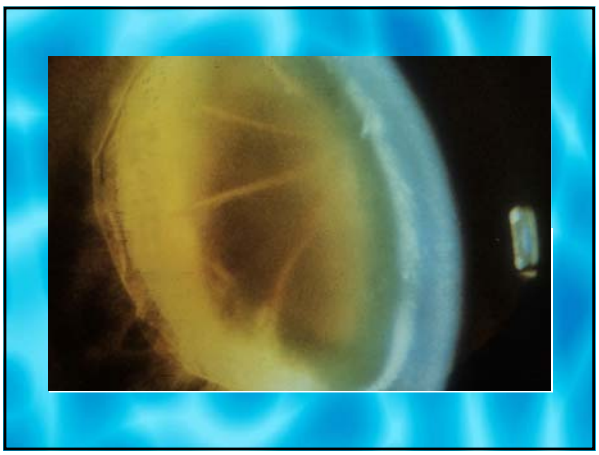
Anterior Lens Dislocation

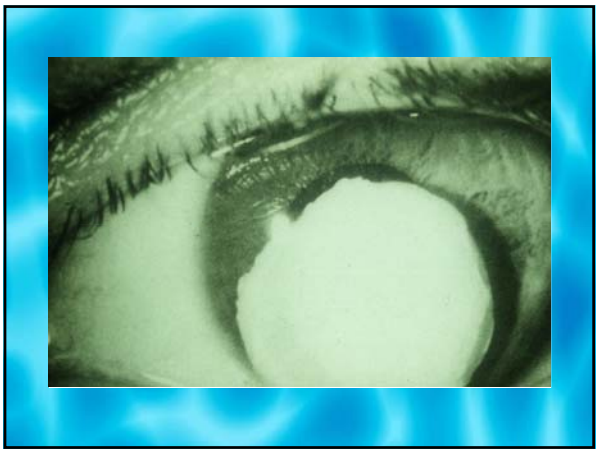
- Pupillary block glaucoma
- Cataract
- Corneal decompensation











Glaucoma Associated with Lens Dislocation

- Spontaneous Dislocation
- Traumatic Dislocation

Anterior Dislocation of the Lens

- Treatment of glaucoma
  - Peripheral iridectomy
  - Prophylactic PI in contralateral eye
  - Miotic

Spontaneous Dislocation into Vitreous Cavity

- Marfan's syndrome
- Marchesani's syndrome
- High myopia

Glaucoma associated with Posterior Dislocation

- Open angle glaucoma
- Pupillary block from vitreous herniation
- Cells and lens material in TM

Posterior Lens Dislocation

- Treatment of glaucoma
  - Peripheral iridectomy if pupillary block
  - Lens removal if phacolysis

POSTERIOR LENS DISLOCATION

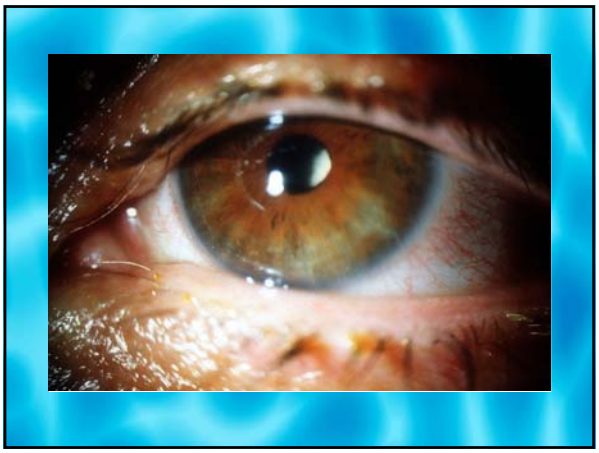
Indications for Surgical Removal

- Inflammation
- Glaucoma
- Visual loss
- Intermittent anterior chamber dislocation with glaucoma

POSTERIOR LENS DISLOCATION

Indications for Surgical Removal

- Hypermature lens
- Capsular rupture/free cortex



Marfan's Syndrome

- Most common inherited disorder with ectopia lentis
- Autosomal dominant, high penetrance
- Muskuloskeletal, cardiovascular abnormalities

Marfan's Syndrome: Ocular FIndings

- Enlarged globe, high myopia, flat cornea
- Iris stromal hypoplasia, correctopia
- Gonio: Iris processes, mounds of iris tissue, vascular anomalies
- Lens dislocation80% (superior, superotemporal); 1st decade

Marfan's Syndrome: Ocular Findings

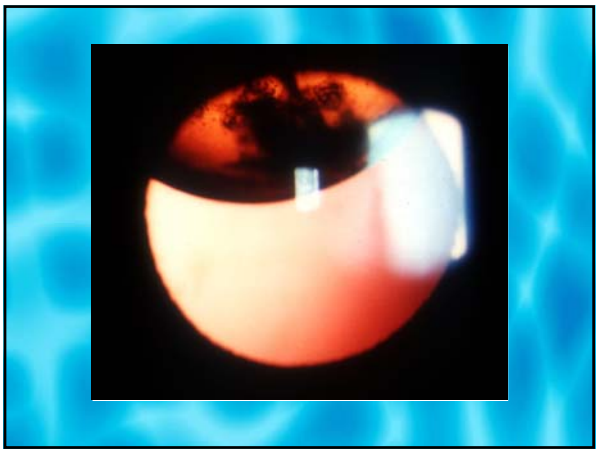
- Lens irregular border, zonules torn and retracted (abnl SEM)
- Lattice, retinal hole, RD
- Heterochromia, keratoconus, strabismus, coloboma of retina/ON

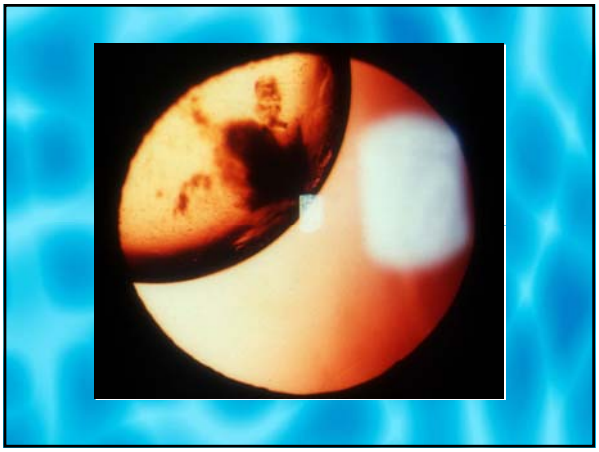
Marfan's Syndrome: Glaucoma

- 8% with ectopia lentis
- Ciliary processes long and narrow
- Longitudinal muscle inserts onto TM
- Circular muscle hypoplastic
- Iris root inserted posteriorly with iris mounds
- Schlemm's canal focally absent

Simple Ectopia Lentis

- Autosomal dominant (rarely recessive)
- Normal pupils
- Onset in childhood (some older pedigrees)
- Bilateral and symmetric
- Dislocated upward and laterally
- Comp: Lens in AC, glaucoma, RD





Ectopia Lentis et pupillae

- Autosomal recessive, bilateral
- Lens and pupil displaced oppositely
- Microphakia
- Pupil distortion
- Consanguinity

Weill-Marchesani Syndrome

- Consanguinity
- Usually recessive
- Brachydactyly, microspherophakia indep. in pedigree
- Lens decrease 20-25% wt and diameter
- Lens zonules weak (strain vs primary disorder)

Weill-Marchesani: Glaucoma

- Anterior lens displacement, pup. block
- Repeated attacks, PAS
- Angle closure worsened by miotic (increase block)
- Contraction of ciliary muscle, lens forward
- Cycloplegia, post. lens movement
- Fully dislocated lens, no effect from miotic

Weill-Marchesani Syndrome Bradymorphia-brachydactyly

- Short stature, stubby hands and feet, brachycephaly, microspherophakia
- Immobility of fingers and wrists
- Normal size globe, small lens
- Narrow angle, forward lens displacement
- Secondary myopia
- Lens dislocation common, early

Homocystinuria

- 3 distinct AR disorders
- Increased homocystinuria in blood
- Cystathionine beta synthetase deficiency
- Skeletal, cardiac, ocular abnormalities;
- MR 50%

Homocystinuria: Ocular

- Lens dislocation 90%
- Inferior, inferotemporal
- 1/3 in AC or vitreous
- No zonules, absent accomodation
- Normal angle structures
- Pupillary block glaucoma
- Optic atrophy (platelet dysfunction)