CORNEAL DYSTROPHIES AND DEGENERATIONS: DIAGNOSIS AND TREATMENT

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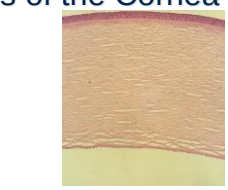
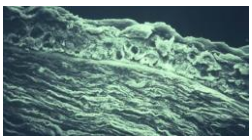
GOALS

- **Differentiate dystrophy vs. degeneration**
- **Review Normal vs. abnormal**
- **Classify the condition by location**
  - **Layers of the cornea**
  - **Central vs. peripheral**
- **Determine appropriate treatment and present in order of complexity**
  - **Similar treatment for various conditions**

Review the Layers of the Cornea

- Tear film 7-11 *u*m
- Epithelium 50 *u*m
- Epithelial BM <128 *n*m
- Bowman 8-14 *u*m
- Stroma 500 *u*m
- "Dua Layer" 15 um
- Descemet 5-10 *u*m
- Endothelium 5 *u*m





SCLAFANI

CORNEAL DYSTROPHY

- Rare conditions
- Slowly progressive, bilateral, central location
- Primary involvement of single corneal layer *
- Variable penetration and severity
- No associated systemic or ocular disease
- No sex predilection.
- Onset by age 20, stabilize by age 40 (except Fuchs)
- Autosomal dominant (50%)

CORNEAL DYSTROPHY

- **Epithelial**
  - Map/dot/fingerprint
  - Meesman's
- **Subepithelial/ Bowman's**
  - Reis-Bücklers Dystrophy (CDB 1)
  - Thiel-Behnke Honeycomb Dystrophy (CDB 2)
  - Subepithelial Mucinous
- **Endothelial**
  - Fuchs' dystrophy
  - CHED—congenital hereditary endothelial dystrophy
  - PPMD—posterior polymorphous dys
- **Stromal**
  - Lattice Dystrophy
  - Granular Dystrophy
  - Avellino Dystrophy
  - Macular Dystrophy
  - Gelatinous Drop-Like Dystrophy
  - Schnyder Crystalline Dystrophy
  - Central Cloudy Dystrophy of Francois
  - Fleck Dystrophy
  - Cornea Farinata
  - Pre-Descemet's Dystrophy
  - Posterior Amorphous Corneal Dystrophy
  - Congenital Hereditary Stromal Dystrophy
  - Primary Band Keratopathy

CORNEAL DEGENERATION

- Non-familial, late onset
- Asymmetric, unilateral, central or peripheral
- Changes to the tissue caused by inflammation, age, or systemic disease.
- Characterized by a deposition of material, a thinning of tissue, or vascularization

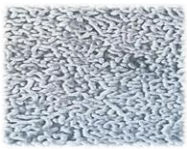
Corneal Degenerations

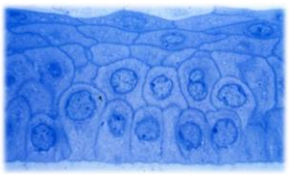
From Periphery to Center (arbitrary division)

- Arcus senilis
- Lipid keratopathy
- White limbal girdle of Vogt
- Senile furrow
- Terrien's marginal degeneration
- Hassall-Henle bodies
- Calcific band keratopathy
- Calcareous degeneration
- Spheroidal degeneration
- Iron deposition
- Coats' white ring
- Crocodile shagreen
- Corneal farinata
- Salzmann's corneal degeneration
- Corneal keloids
- Corneal amyloid degeneration

EPITHELIUM

- 50 *um* non-keratinized stratified squamous epithelium
- 5-10 layers central 8-10 peripheral
- Superficial layers have microvillae that attach tears.
- Exfoliation q 5-7 days
- Deeper layers (Basilar Columnar cells) have hemi-desmosomes
  - connect the epithelium to basement membrane which connects to Bowman's Layer.
- First phase of wound healing occurs with migration of existing cells over the wound





EPITHELIAL BASEMENT MEMBRANE "DISORDER" EBMD

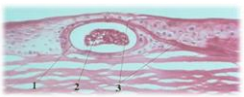
- Most common anterior corneal disorder
- DYS: inherited, single layer, bilateral
- DEG: Prevalent 43%, 25% unilat,> 29y,trauma
- Abnormality of epithelial turnover, maturation, and production of BM and adhesion complexes

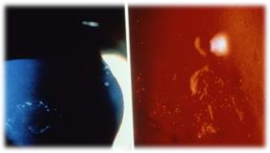
Thickened BM ultimately weakens the epithelium and causes recurrent corneal erosion (RCE).

EBMD

- The basal cells produce abnormal finger-like projections that bend intra-epithelialy and trap cells/debris that form cysts.
- MAPS : multi-lamination of BM and collagen
- DOTS: grey opacities,cysts
- FINGERPRINT: reduplication of BM

AD: TGFBI/BIGH3 gene

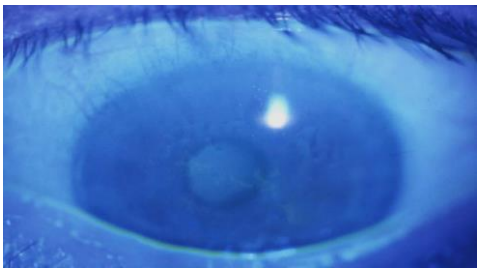


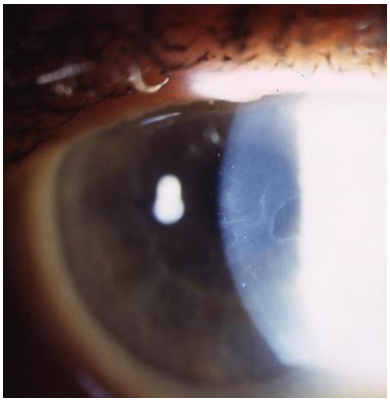


SLX of EBMD

- Negative NaFL pattern and instantaneous TBUT
- No Rose Bengal Stain
- When Microcysts surface and erupt , + NaFL
- Asymptomatic vs. Variable degrees of Blur, diplopia, photophobia, dryness, FBS, or pain.

EBMD





Bandage Contact lens
Focus Night/Day
BC 8.4/13.4
Plano 20/20+
CW 6 pm

TREATMENT FOR EBMD

- Indicated if vision or comfort are compromised.
- Manage co-existing ocular surface disease
- Environment/ diet /bedtime eyeshields
- Lubricants
- Punctal occlusion
- Bandage Contact Lens (BCL)
- Surgical: PTK

AT Containing Hyaluronic Acid HA

- Most common in Europe and Asia
- Higher concentration of Sodium Hyaluronate
- Strong ability to bind water
- Visco-elastic properties to stabilize tear layer
- Predominant glycos-aminoglycan to appear at the wound site and aids in cell proliferation, migration and ultimate healing
- High MW HA = cohesive
- Low MW HA= dispersive

TREATMENT FOR EBMD/ABMD to Prevent Recurrent Erosion

- Avoid preservatives or surfactants
- Electrolytes nourish eye
- DED benefits from Hypo-osmotic drops
  - Counteract the high salt contact of dry eyes
  - Bland ointment help retain fluid on the eyes
- Edematous Corneas or Weak junctions that lead to RCE benefit from Hyper-osmotic agents
  - Sodium Chloride vs. High Oncotic Pressure

  Muro 128: Solution (2-5%) vs. ointment (5%)

  Ung: comfort, > concentration

  Treat 6 weeks Solution /3-6mo ung
- Warm Packs: QID 2-3 weeks/shields

Vs.

Autologous Serum Drops

- Utilizes patient's own blood serum
- Blood is drawn and the serum is spun down and mixed with artificial tears. Devoid of cells and clot factors
- Replaces "personal" growth factors
- Replaces individualized antibodies
- Serum contains growth factors, fibronectin, Vit. A and anti-proteases
- Requires blood donation 2-3 times year $150-$300 Hospital/Lieters
- Consider 5-10% serum albumin drops qid instead

Original Article

Gary N. Foulks, MD, EDITOR

Abdominal Breathing Increases Tear Secretion in Healthy Women

Kokoro Sano, RN, PHN, Motoko Kawashima, MD, PhD, Kazuhiro Ikeura, DDS, Reiko Arita, MD, PhD, and Kazuo Tsubota, MD, PhD

Abstract

PURPOSE: To determine the relationship between abdominal breathing and tear meniscus volume in healthy women, we investigated the change in tear meniscus volume in two groups: normal breathing and abdominal breathing.

METHODS: We used a crossover experimental model and examined 20 healthy women aged 20-54 years (mean ± SD, 32.7 ± 11.1 years). The participants were randomly assigned to one of two groups. During the first visit, the normal breathing group was subjected to normal breathing for 3 min, whereas the abdominal breathing group was subjected to abdominal breathing (4-second inhalation and 6-second exhalation) for 3 min. During the second visit, the protocols were swapped between the two groups. We estimated the R wave to R wave (R-R) interval, tear meniscus volume, salivary amylase activity, pulse, and blood pressure before and immediately after, 15 min after, and 30 min after completion of the breathing activity.

RESULTS: After abdominal breathing, compared to that before breathing, the tear meniscus volume increased significantly 15 min after breathing ($P < .01$). Furthermore, systolic blood pressure showed a significant decrease immediately after abdominal breathing ($P < 0.05$). No significant difference was found in the test parameters in the normal breathing group.

CONCLUSION: Abdominal breathing for 3 minutes increases the tear meniscus volume in healthy women. Consequently, abdominal breathing may be considered in the treatment of dry eye disease.

The Ocular Surface

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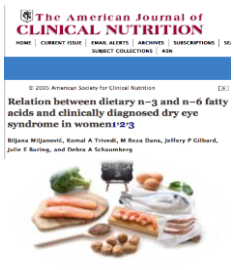
ANTI-INFLAMMATORY EFFECTS OF EFA

- WOMENS HEALTH STUDY
- Published 2005
- N > 32K
- Association of Low Dietary intake of ω3 and DED
- 30% reduction in risk/1g/day
- Elevated Risk DED 15:1
  when ω6>ω3
- When balance is off, it is associated with lipid abnormalities
- Lacrimal gland preventing apoptosis of the secretory epithelial cells



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Relation between dietary n−3 and n−6 fatty acids and clinically diagnosed dry eye syndrome in women1·2·3

Bijana Miljanović, Komal A Trivedi, M Reza Dana, Jeffery P Gilbard, Julie F Buring, and Debra A Schaumberg

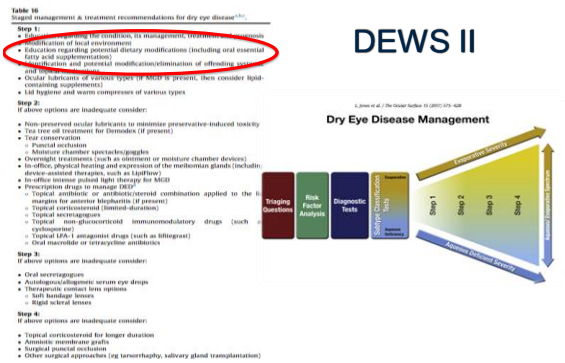


Table 16
Staged management & treatment recommendations for dry eye disease[1,8,9]:

Step 1:
- Education regarding the condition, its management, treatment and prognosis
- Modification of local environment
- Education regarding potential dietary modifications (including oral essential fatty acid supplementation)
- Medications and potential modification/elimination of offending systemic medications
- Ocular lubricants of various types (if MGD is present, then consider lipid-containing supplements)
- Lid hygiene and warm compresses of various types

Step 2:
If above options are inadequate consider:
- Non-preserved ocular lubricants to minimize preservative-induced toxicity
- Tea tree oil treatment for Demodex (if present)
- Tear conservation
  - Punctal occlusion
  - Moisture chamber spectacles/goggles
- Overnight treatments (such as ointment or moisture chamber devices)
- In-office, physical heating and expression of the meibomian glands (including device-assisted therapies, such as LipiFlow)
- In-office intense pulsed light therapy for MGD
- Prescription drugs to manage DED[9]
  - Topical antibiotic or antibiotic/steroid combination applied to the lid margins for anterior blepharitis (if present)
  - Topical corticosteroid (limited-duration)
  - Topical secretagogues
  - Topical non-glucocorticoid immunomodulatory drugs (such as cyclosporine)
  - Topical LFA-1 antagonist drugs (such as lifitegrast)
  - Oral macrolide or tetracycline antibiotics

Step 3:
If above options are inadequate consider:
- Oral secretagogues
- Autologous/allogeneic serum eye drops
- Therapeutic contact lens options
  - Soft bandage lenses
  - Rigid scleral lenses

Step 4:
If above options are inadequate consider:
- Topical corticosteroid for longer duration
- Amniotic membrane grafts
- Surgical punctal occlusion
- Other surgical approaches (eg tarsorrhaphy, salivary gland transplantation)

DEWS II

J. Jones et al. / The Ocular Surface 15 (2017) 575–628

Dry Eye Disease Management

Caffeine is a non-selective competetive adenosine antagonist, that increases the level of acetylocholine, for the parasympatheic path..
Acts on lacrimal gland o increase secretion
Protective against MRSA

ecretion

Kwaku Antwi Osei*, Godwin Ovenseri-Ogbomo†, Samuel Kyei‡, and Michael Ntodie*

ABSTRACT

Purpose. Caffeine, probably the most widely consumed psychoactive substance, is claimed to have conflicting effects on some tear film dynamics. This study sought to investigate the effect of orally ingested caffeine on tear secretion.

Methods. In an examiner-masked, placebo-controlled, crossover experimental model, the effect of caffeine intake on tear secretion was studied in 41 healthy volunteers aged 20 to 26 years (mean, 23.0 ± 2.1 years). Participants were randomly assigned into two groups, A and B, to receive two different treatments in two sessions. Subjects in group A were exposed to 5.0 mg/kg body weight of caffeine dissolved in 200 mL of water on their first visit, whereas those in group B were exposed to 200 mL of water. On the second visit, however, the order of treatment was reversed. Schirmer 1 scores were measured repeatedly at 45, 90, 135, and 180 minutes after treatment. The baseline Schirmer 1 scores were compared with posttreatment scores.

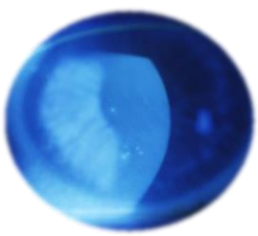
Results. Schirmer 1 scores increased after caffeine intake. The increase was statistically significant at 45 and 90 minutes ($p < 0.05$) after caffeine intake. Age, body mass, and blood pressure had no correlation with Schirmer 1 scores (Spearman correlation test, $p > 0.05$). There was no influence of gender in caffeine's effect on tear secretion ($F = 0.994$, $p = 0.399$).

Conclusions. From our study, orally ingested caffeine appears to stimulate tear secretion in healthy non–dry eye subjects.

(Optom Vis Sci 2014;91:171–177)

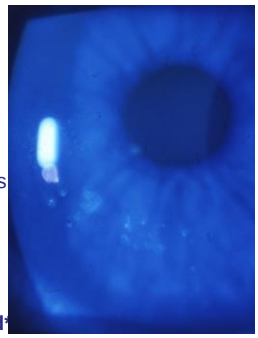
Superficial Punctate Keratitis of Thygeson (SPKT)

- Chronic, usually bilateral disorder characterized by focal epithelial lesions favoring central visual axis
- Mean age 29 (2 to 70)
- Long duration with remissions and exacerbations
- Asymptomatic (esp. later) vs. FBS, epiphora, photophobia
- Corneal sensation not effected although occasional hypo-aesthesia… r/o HSV

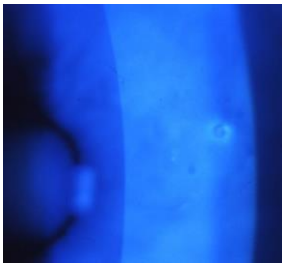


SPKT

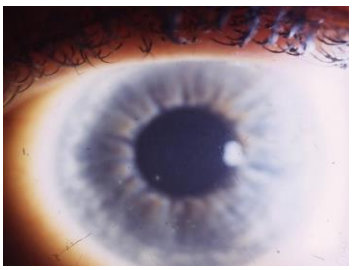
- Average of 15-20 lesions
  - (1 to 50)
- Lesions have +NaFl/RB staining and are elevated during active disease process
- Each lesion comprised of multiple lesions
- Change position over time
- No stromal involvement
- **Conjunctiva is not inflamed***



Lesions are round and show negative staining



Conjunctiva: usually not inflamed unless during the developmental stage:1-2 wks



RPS Adeno Detector™ Rapid Pathogen Screen

RPS RAPID PATHOGEN SCREENING
Point-of-care diagnostic devices

Collecting the Sample

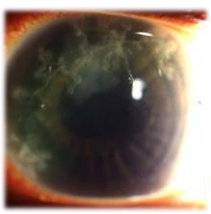
Dab the *sampling pad* inside the lower eyelid (palpebral conjunctiva) 4 – 6 times. Allow the *sampling pad* to rest against the conjunctiva (membrane on inside of the eyelid) for an additional 3 seconds to ensure saturation of the *sampling pad* with eye fluid.

Etiology of SPKT Unknown

- Possibly Viral due to latency, recurrence, lesion appearance, duration
- PCR testing proved that it is NOT HSV 1, HSV 2, VZV or Adenovirus
- Still investigating HPV since both have minimal inflammation
- Prolonged SPKT Associated with Salzmans Nodular Degeneration
- Suggested association with eczema, urticaria, asthma
- HLA-DW3 and DR3 association: gluten sensitive, DM2, Lupus, Graves

Treatment for SPKT

- Lubricants for comfort
- BCL to smooth surface
- Good response to steroids however needs long taper and can prolong the course or worse. Confident DDX…
- Antivirals ?
- Cyclosporine
  Reinhard showed 70% suppression with 2%
- Lack of response to systemic or topical AB, debridement/ cautery of tissue

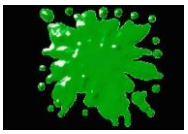


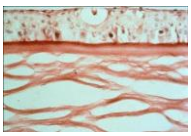
5 CHARACTERISTICS OF THYGESONS

1. Chronic, usually bilateral disorder characterized by central focal epithelial lesions and no stromal involvement
2. Long duration with remissions/exacerbations
3. Eventual Healing without scars
4. Lack or response to AB treatment
5. Striking response to steroid TX

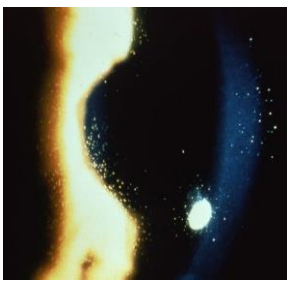
MEESMAN'S DYSTROPHY

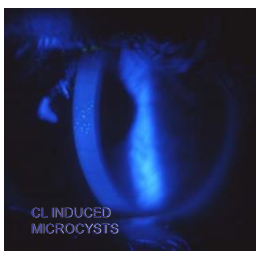
- Diagnosed within first year of life
- A "peculiar" substance is produced which thickens the BM.
- Numerous epithelial vesicles that extend to limbus*
  - Contain debris,cells,GAG
- No scarring. Photophobia. Irritation
- May have slight decrease in VA.
- CLS are not contraindicated and may be therapeutic when rupturing
- LISCH : whorl-shaped clusters





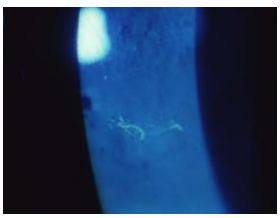
MEESMAN'S DYSTROPHY





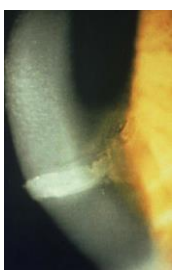
RECURRENT CORNEAL EROSION

- Traumatic erosions due to thickened BM with poor hemi-desmosomal attachments.
- May result from incomplete healing following trauma
- Associated with EBMD (50%) or Stromal Dystrophies esp.Lattice



RECURRENT CORNEAL EROSION

- Onset in the am due to edema or shearing effects
- Symptoms may be more severe than it appears
- Epithelial loss surrounded by pooling and loose ends



Treatment for RCE

- ACTIVE
  - Aggressive corneal abrasion TX = BCL heals slower
- PROPHYLAXIS
  - Lubricants/ Hyperosmotic agents
  - Long-term BCL, Amniotic Tissue, MMP inhibitors
- SURGICAL
  - Debridement
  - Anterior Stromal Puncture
  - PTK with PRK

Prospective, Multicenter, Clinical Evaluation of Point-of-Care Matrix Metalloproteinase-9 Test for Confirming Dry Eye Disease

TREATMENT OF RCE

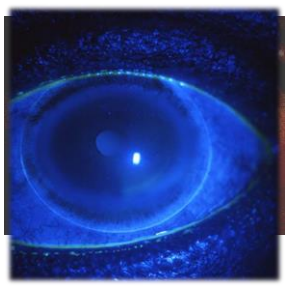
PROPHYLAXIS

- Patients with RCE show a chronic increased level of metallo-proteinase enzymes (MMP 2&9) which may dissolve the basement membrane and fibrils due to inadequate neutralization. TX = inhibit MMP
- Doxycycline: oral, 50mg BID
  - 2 months treatment time. Reduced MMP 70%.
- Topical Steroids
  - Pred Forte, FML, Lotemax, TID,2-3 weeks
- No recurrences in 21 months.

Dursan and Plugfelder. "Treatment of Recalcitrant RCE with inhibitors of matrix metalloproteinase –9." American Journal of Ophthal. 2001,132:8-13

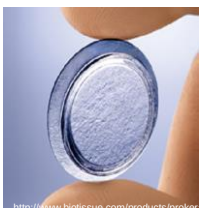
BANDAGE CONTACT LENSES

- To aid in healing by offering protection
- To provide comfort for decompensating corneas with erupting microcysts
- To aid in dehydration
- To produce a more regular refracting surface
- To aid in drug delivery
- To reduce inflammation



Amniotic Membrane Grafts (AMG) "ACTIVE" treatment over BCL

Biotissue-Prokera, Amniograft, & Amnioguard



IOP Ophthalmics-Ambiodisk

AMG Actions

1. Reduces inflammation
2. Inhibits scarring
3. Inhibits angiogenesis
4. Promotes epithelialization
5. Possesses anti-microbial properties
6. Restoration of lost corneal thickness*

TREATMENT OPTIONS

TARSORRHAPHY

- Surgically close the palpebral fisssure by suturing

the superior and inferior lids at the lateral aspect

STAMLER LID SPLINT

- Adhesive on one side with enou rigidity on the other to hold the the closed position

TAPESORRHAPHY

- Tagederm

AMNIOTIC TISSUE GRAFTS

- Allow for use of meds and examination

Treatment for RCE

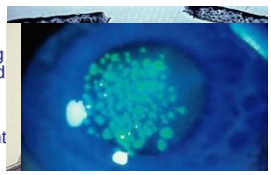
- ACTIVE
  - Aggressive corneal abrasion TX = BCL heals slower
- PROPHYLAXIS
  - Lubricants/ Hyperosmotic agents
  - Long-term BCL, Amniotic Tissue, MMP inhibitors
- **SURGICAL**
  - **Debridement**
  - **Anterior Stromal Puncture**
  - **PTK with PRK**

CORNEAL DEBRIDEMENT

- Soften epithelium
  1-2 gtt topical anesthetic
  q 15-30 seconds for 2-3 minutes
- Use cotton swab, spatula, spud or jewelers forceps
- Remove flaps by pulling edges toward center
- Don't pull directly up or out
- Remove flaps down to tight, firm edges.
- Tx abrasion (>50-100%)
  - Recurrence Rate 18%

ANTERIOR STROMA MICROPUNCTURE

- Disturb Bowman's Layer to promote tighter adhesion and stimulate cornea to produce functional BM complexes
- Topical anesthetic and a 27g cannula: use forceps to bend needle to avoid puncture
- Closely spaced (.5mm) punctures damaged/adjacent
  - Anterior Stroma :100-150 *u*
  - Apply tangential force
  - Avoid Visual axis since minimal scarring can occur
  - RR 40% Yag 80% success

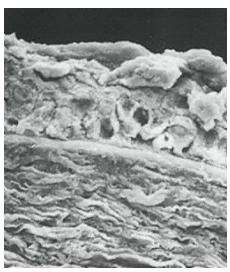


PTK

Phototherapeutic Keratectomy
Combined PRK
Wavefront Technology

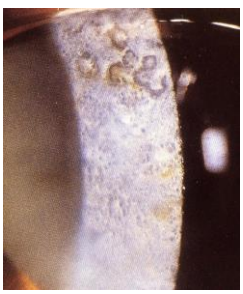
Bowman's Layer

- Acellular modified layer of anterior stroma
- Type 1 collagen fibers randomly arranged
- Pores for corneal nerves to pass
- Fxn? Not found in many species yet good vision and normal epithelial-stroma junctions.
- Not replaced however when damaged, causes significant opacification which effects VA

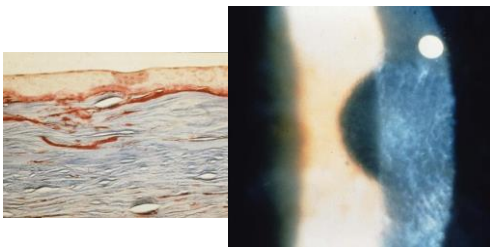


REIS-BUCKLER DYSTROPHY

- Bilateral, symmetric, AD, by age 5
- Bowman's layer is obliterated and replaced with randomly arranged regular collagen that thickens.
- Linear, ring-like or "Honey comb"
- Marked VA loss due to superficial stromal haze or topographical changes from elevation of tissue
- Painful if recurrent erosions occur.
- TX: PKP or LK but may recur

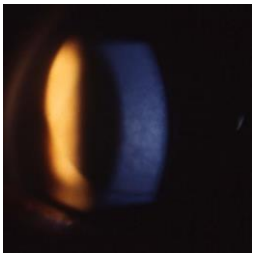


REIS-BUCKLER DYSTROPHY



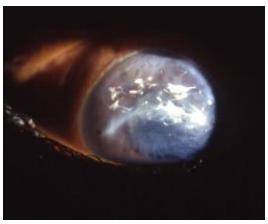
ANTERIOR MOSAIC

- Dystrophy or Degeneration
- AKA: Anterior Crocodile Shagreen
- Breaks in Bowman's that appear like central grey polygonal opacities with clear spaces.
- Blanches with limbal pressure.
- Asymptomatic

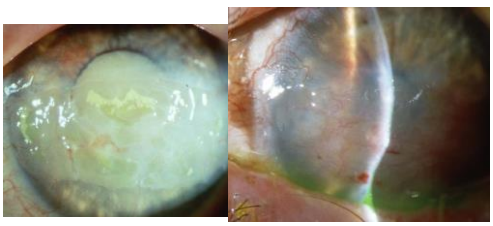


BAND KERATOPATHY DEGENERATION

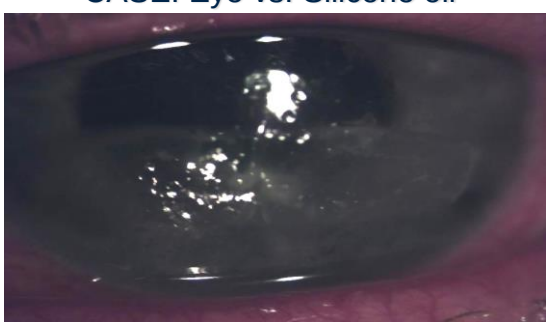
- Deposition of Calcium salts in Bowman's layer
- Located interpalpebral region
- History of uveitis, renal failure, prolonged use of miotics, syphillis, interstitial keratitis,hyperparathyroidism
- TX: Chelation with EDTA 1%
- TX: Therapeutic CL



EDTA Treatment

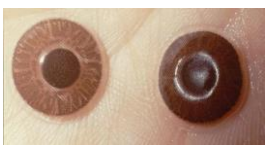


CASE: Eye vs. Silicone oil

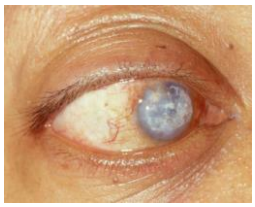


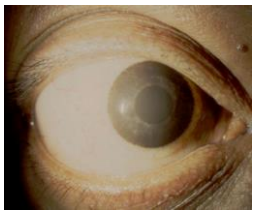
Treatment: Cosmetic Contact Lens

- Black Underprint: color is applied to a dark background to mask and make a scar more uniform This darkens and mutes the overlaid color.
- Store in glass vials
- 53% H20 to maintain dye



Occluder Contact Lens for Band K





SALZMANN'S NODULAR DEGENERATION

- Bluish, superficial nodular elevations
- Inflammatory/Non-inflammatory event that exposes the cornea and results in excess COLLAGEN plaques that replace BOWMANS
- Post-chronic-keratitis
- Asymptomatic to very painful and sight threatening depending on location and severity
- TX: BCL/AB/NSAID, PTK,PKP

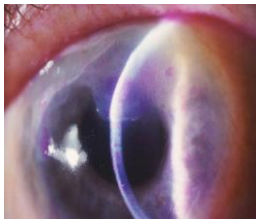
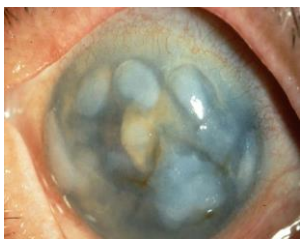
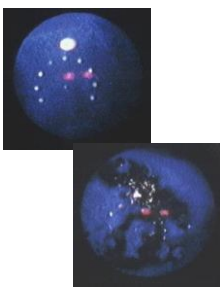


Photo-therapeutic Keratectomy:PTK
Debulking Effect for Salzmann's Nodular Degeneration
PRK with Localized Spot Ablations 3-6 mm diameter





STROMAL DYSTROPHYS

- 90 % of corneal thickness
- 22%: Comprised of TYPE 1 collagenous lamellae interspersed with keratocytes and ground substance(proteoglycans, glycoproteins, serum)
- GAGS: affect hydration, thickness, transparency
- 78%: rest is water.
- Abnormal Substance found within the cells or fibrils that have distinct histological-stains

Name of Dystrophy
Name of Deposition
Pathology Stain

- ***M**arilyn **M**onroe **A**lways*
- ***G**ets **H**er **M**an*
- ***L**os **A**ngeles **C**ounty*
- ***S**outhern **C**alifornia **O**cean*

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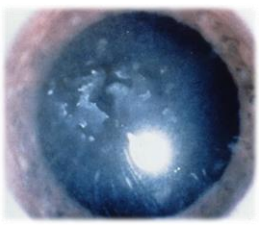
MACULAR DYSTROPHY

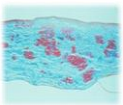
- Clouding due to build-up of muco-polysaccharides
- Begins centrally & superficially then extends limbus to limbus thru all layers
- Thinning, no clear spaces between
- Primary involvement of the endothelium: guttata*
- Begins in 1st decade of life: aggressive causing early & severe VA loss
- Predominant in Virginia area
- Autosomal recessive*
- TX: PKP/DALK

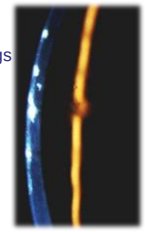


- *M**acular** / **M**ucopolysaccharide / **A**lcian Blue*

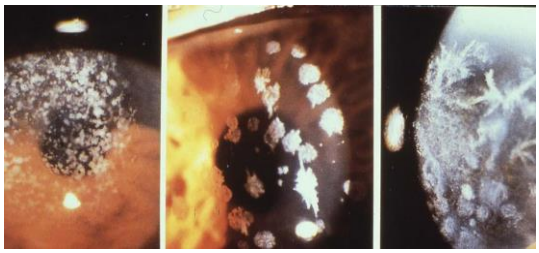
GRANULAR DYSTROPHY

- Central, anterior to mid-stromal deposits of Hyaline
- Discreet white spots (translucent) to powdery rings
- Clear areas between lesions in early years
- Erosions can break thru BM.
- Autosomal dominant w/ complete penetrance*
- *Granular / Hyaline/ Masson Trichrome*





GRANULAR DYSTROPHY



GRANULAR DYSTROPHY TREATMENT

- Pinhole effect may maintain VA (20/20) until the lesions coalesce and reduce VA=20/200.
- PKP was only treatment and recurrences were common
- Present treatment includes PTK/DALK and BCL:
  - Smooth epithelial surface to treat monocular diplopia
  - Pain management following PTK or erosions
  - Induced anisometropia
  - Spectacle distortions of high plus lens

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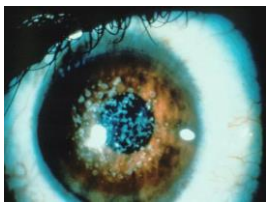
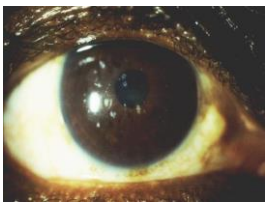
PTK Treatment for GRANULAR





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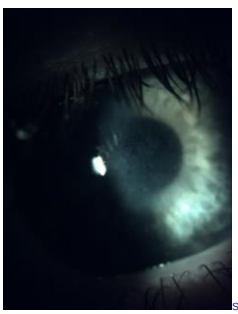
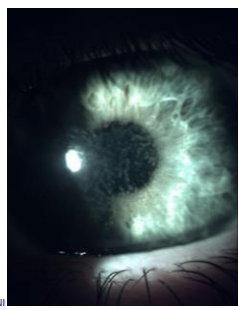
27 yo WF: Granular Dystrophy





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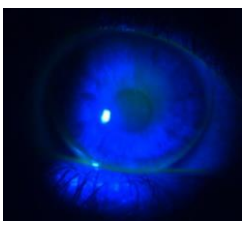
Contact Lens Fit for Granular

- C/O anisometropia/haze
- RE +7.50 – 2.00 x 010 20/30
- LE +.25 – 2.25 x 170 20/30
- CL FIT
  - DIL +3.50 8.08/11.2 20/25 +
  - PV pl -1.75 x 180
- Refit OD at 4 months pg
  - Hydrasoft Options
  - +8.75 -2.00 x 010 20/25 !!!



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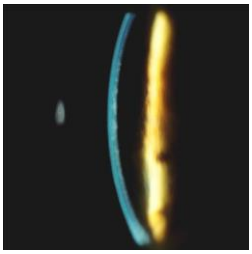
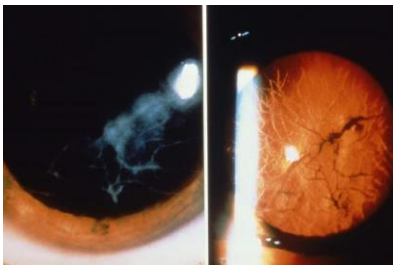
LATTICE DYSTROPHY

- Branching refractile filaments of AMYLOID
- Symptoms occur around 20 yo
- RCE are common
- Resultant scars and late intervening haze can blur VA
- Lines thicken with age & penetrate deeper layers
- Autosomal Dom/ Recessive
- TX: DALK, PKP
- *Lattice / Amyloid / Congo Red*



LATTICE DYSTROPHY

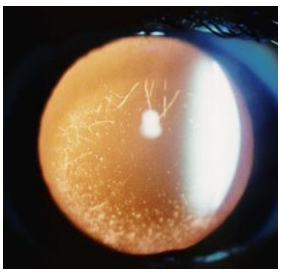


TYPES OF LATTICE DYSTROPHY

- **T**YPE 1
  Poor VA by age 40-60
- TYPE 2
  Merotoja syndrome
  Bilateral Facial palsy, skin thickens,
  Depressed brows, prominent
  VA loss >65y
- TYPE 3
  Floppy ears, protruding lips, Auto-R
  Larger deposits, mid stroma, no RCE
  VA loss > 60 y

AVELLINO DYSTROPHY

- Avellino, Italy
- Typical granular dystrophy with axial anterior stromal haze and mid-stroma discreet linear opacities.
- Congo red



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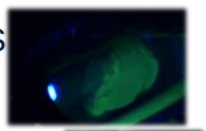
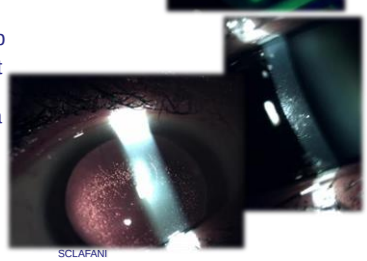
CASE EXAM FINDINGS

- Presents to ER with abrasion
- At 3 day follow-up
- SLX: fine discreet lines in the mid-peripheral stroma with translucent white spots. VA 20/20





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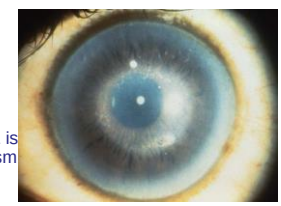
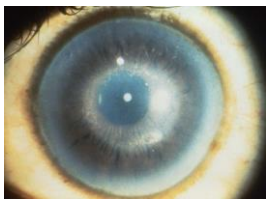
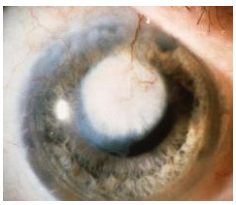
CENTRAL CRYSTALLINE DYSTROPHY OF SNYDER

- Deposits of cholesterol crystals in anterior stroma
- Premature peripheral arcus
- Vision is generally good
- Usually normal serum lipid profile: +/- hyperlipidemia
- Expressivity is variable
- B120 gene on chromosome 1 is responsible for lipid metabolism and transport
- ***S**nyder / **C**holesterol /**O**il*



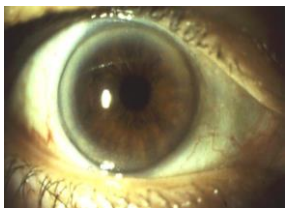
CENTRAL CRYSTALLINE DYSTROPHY OF SNYDER

- Vs. Secondary Lipid Keratopathy: Cholesterol and Lipid deposition as a result of long-standing deep NEO 2• to HSV/HSK.
- TX of NEO via topical steroids, photocoag. of vessel, LKP/PKP

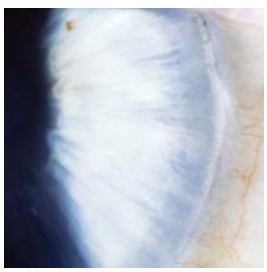
ARCUS SENILIS

- Effects >60% population between 40-60 years
- Peripheral lipid deposition
- Located anterior to Descemet's layer and in Bowman's layer
- Juevenile form usually represents hyperlipidemia
- Be suspicious of carotid disease if this is present to a greater degree in one eye.



WHITE LIMBAL GIRDLE OF VOGT

- Effects > 50% population between 40-60
- With/Without clear zone
- Represents subepithelial degeneration and sometimes calcium deposition
- Does not affect visual acuity
- Located in the horizontal meridian

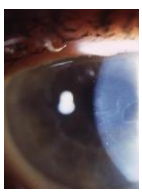


POSTERIOR EMBRYOTOXIN

- Extremely prominent ring of Schwalbe
- Eye is normal but may be associated with corectopia, aniridia, or corneal conditions that are part of systemic syndromes

Restasis® Allergan

(Cyclosporine Ophthalmic Emulsion 0.05%)



- **Indicated for patients who do not get relief with compresses and lubricants**
- **Restores tear production**
- **Increases goblet cells**
- **Excellent safety profile**
- **BID dosing**
- **Mild stinging**

OFF LABEL USES

- EBMD
- Thygesons Dx
- Blepharitis
- Refractive Surgery
- Graft vs.Host in BMT
- Viral Conjunctivitis
- Chronic Uveitis

Shire LIFITEGRAST= XiiDRA

- Preservative-free topical eye solution made up of a small-molecule integrin antagonist
- First-in-class molecule that inhibits T-cell inflammation by blocking the binding of 2 key cellular surface proteins (LFA-1 and ICAM-1) that mediate the chronic inflammatory cascade
- Apr 15. FDA accepts lifitegrast for priority review reducing assessment time from the standard 12 months to just eight months. [10]
- ADHD and Hunter Syndrome (GAGS)

Normal Changes to the Endothelium

- Descemet's layer thickens from 3-17*u*
- There is a decrease in the # of endothelial cells
  - from 3500 cells/mm^2 to 1200
  - this single layer spreads out: lacks mitosis
- High density mitochondria : 90% pump
- Lenses produce reversible polymegathism

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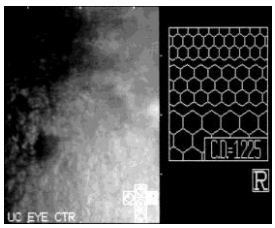
Abnormal Changes to the Endothelium

- Endothelial cells become more irregular = Polymorphism
- Cells secrete collagen towards Descemet's causing multilamination = guttata
- This breaks down the barrier function and results in stromal and epithelial edema



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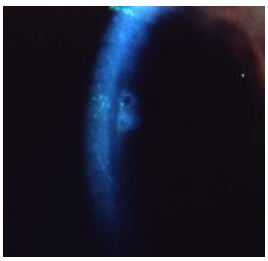
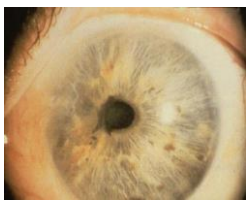
POSTERIOR POLYMORPHOUS DYSTROPHY- PPMD

- Isolated to coalescent vesicles that intervene between normal endothelial cells.
- Areas of normal or thickened Descemet's membrane representing a collagenase material
- These vesicles can lead to stromal edema.
- Association with keratoconus



POSTERIOR POLYMORPHOUS DYSTROPHY- PPMD

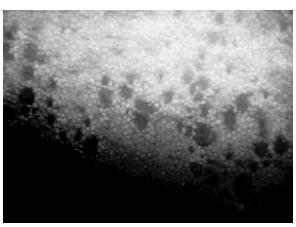
- Can be present at birth
- Wide variety of expression
  - Non-symptomatic
  - Grouped vesicles cause blur
  - Stromal edema
  - Correctopia and irido-corneal adhesions resulting in glaucoma if they enter TM

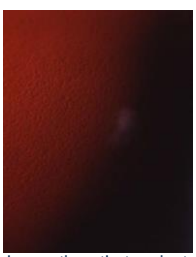


FUCH'S DYSTROPHY

- Bilateral, asymmetric, begins in 5th or 6th decade
- More predominant in women (3x)
- Initially pigment dusting
- Non-symptomatic

FUCH'S DYSTROPHY STAGE 1

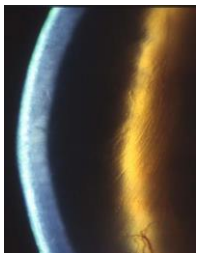




Guttata represent clear, vesicular endothelial secretions that project into the potential space between the endothelium and Descemet's

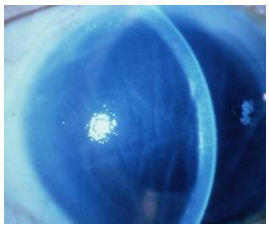
FUCH'S DYSTROPHY STAGE 2

- Guttata interrupt the normal pumping mechanism = edema
- Edema begins around Descemet's and Bowman's layers and then spreads the entire thickness.
- Pt. experience glare/hazy VA
- Bullae appear: they reduce vision and cause pain when they rupture, especially in am



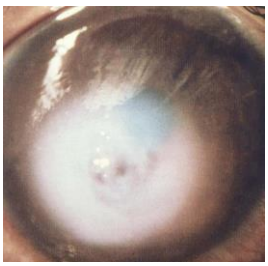
FUCH'S DYSTROPHY STAGE 2

- Guttata interrupt the normal pumping mechanism = edema
- Edema begins around Descemet's and Bowman's layers and then spreads the entire thickness.
- Pts experience glare/hazy VA
- Bullae appear: they reduce vision and cause pain when they rupture, especially in am



FUCH'S DYSTROPHY STAGE 3

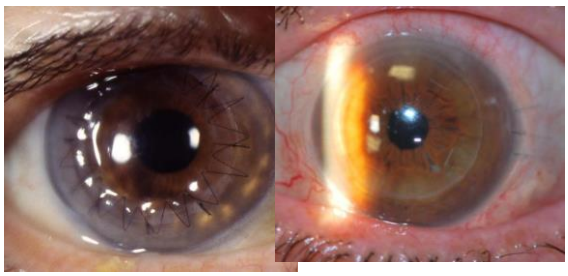
- Edema is reduced but sub-epithelial connective tissue grows and causes reduced vision.
- Patient is comfortable due to reduced corneal sensitivity.
- Elevated IOP, peripheral neovascularization, and corneal erosions.



FUCH'S DYSTROPHY TREATMENT

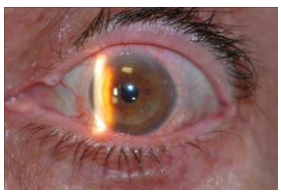
- Hypertonic solutions to draw fluid out 4-6 x
  - Sodium Chloride
  - Muro 128 (2% or 5%) solution, 5% ointment-PF
  - Fresh Kote
- BCL to aid in comfort for ruptured bullae
- Hair Dryer in the am
- Lubricants for comfort
- Lower IOP
- Conjunctival flap
- Corneal transplant to restore vision: PKP vs. DSEK

PKP vs. DSEK

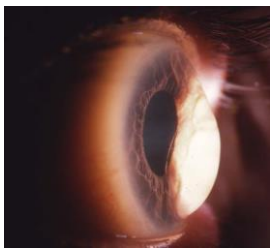


DSEK: Descemet Stripping Endothelial Keratoplasty

Faster visual recovery
Less astigmatism created since there are no sutures
Eye is much stronger and more resistant to injury since only the diseased tissue rather than the entire cornea
Surgery time is quicker
Chance of rejection is reduced
Procedure can be combined with cataract surgery
VA 20/30-20/40
1-2 D Hyperopic Shift, thicker

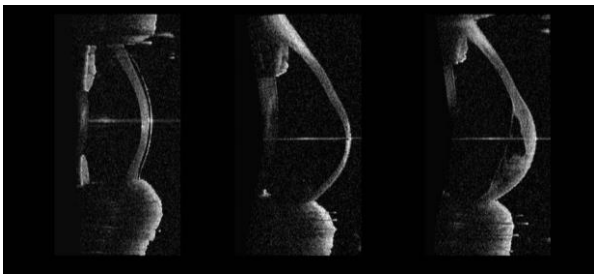


KERATOCONUS



"Keratoconus is a clinical term to describe a condition in which the cornea assumes a conical shape because of thinning and protrusion"

Evolution of KCN: Ectasia to Hydrops

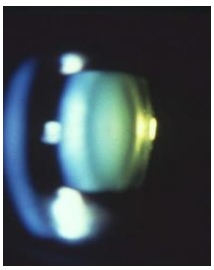


Keratoconus- Keratometry

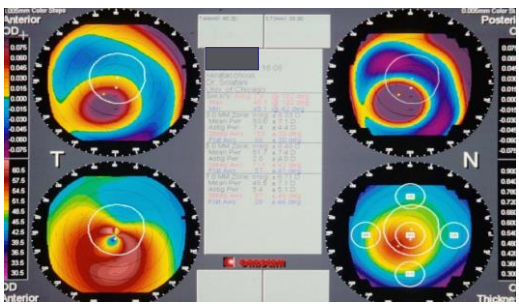
- Initially, mires get small and then there is a lack of parallelism
- Expand perimeters by use of +1.25 SPH and add 7 D to your reading
- Steepening begins infero-temporally and progresses clockwise
- TOPOGRAPHY- more sensitive
- PLACIDO RINGS- get closer

RETINOSCOPY

- Scissors Reflex
- Against motion that breaks apart
- Represents multiple refractive powers within the optic zone



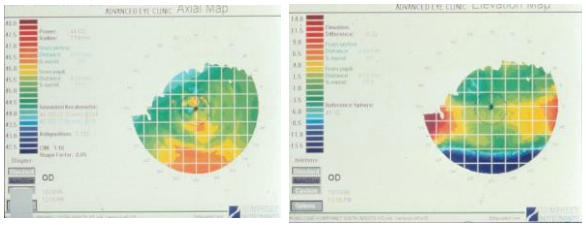
Topography of Keratoconus



PSEUDOKERATOCONUS

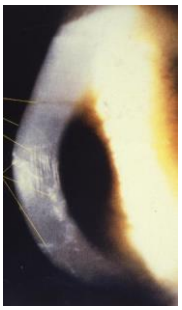
- Corneal warpage topography can mimic KC
- Repeat topography must be performed and a measurable change would indicate pseudo-KC
- Evaluation of elevation maps at steep zone:
- Predicts the elevation or depression of the cornea if the best fit sphere was on cornea

ELEVATION MAP DIFFERENTIATES KERATOCONUS vs WARPAGE

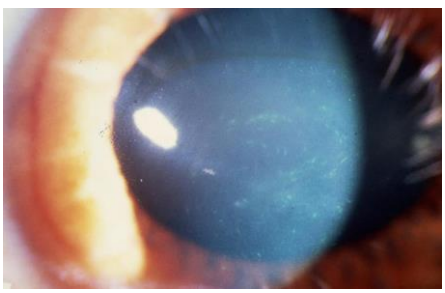


KERATOCONUS-SLIT LAMP FINDINGS

- FLEISCHER RING
- VOGT' S STRIAE
- STROMAL THINNING
- STROMAL SCARS
- SWIRL-LIKE PATTERN
- ENLARGED CORNEAL NERVES
- ACUTE HYDROPS

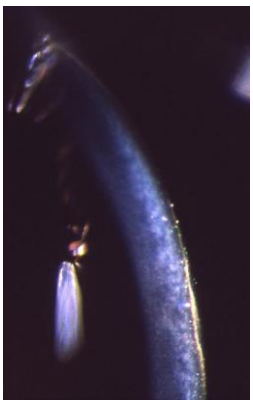


WHORL-KERATOPATHY

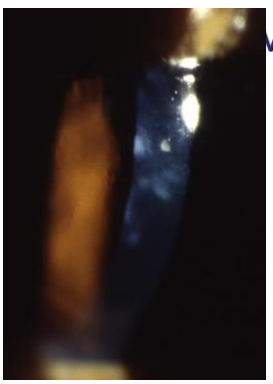


FLEISCHER RING

abrupt change
in curvature
50%



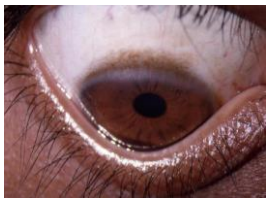
VOGTS'S STRIA (1st Sign)



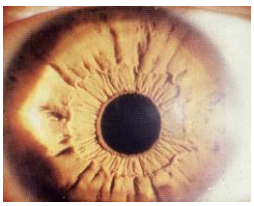
MAL SCAR

CLEK showed
Overall 13% had scars
K > 52 38% scarred
43% of flat fits scarred
26% of steep fits scarred
8% ↑ with each hour WT
↑ scar w/ stain, ring, age, CL (2 fold), ↑FDACL

EXTERNAL FINDINGS

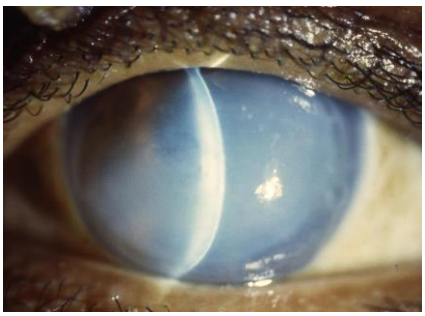


MUNSONS SIGN



RIZZUTIS SIGN

CORNEAL HYDROPS

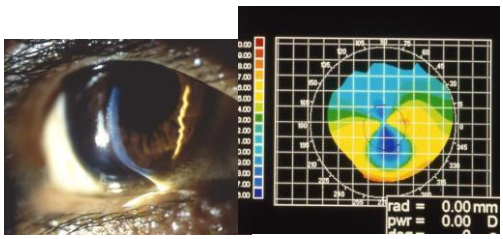


5%

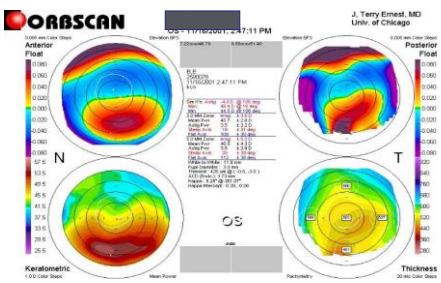
PELLUCID MARGINAL DEGENERATION

- 20-40yo, no gender preference, slow progression
- Thinning occurs below the steep curvature
- Stromal thinning is concentric to the lower limbus and runs from 4-8:00, 1-2mm wide
- Clear, epithelialized, and non-vascularized.
- Absence of lipid: ddx from Moorens or Terriens
- Vertical stress lines and hydrops can occur
- BEER- BELLY CORNEA-

PELLUCID MARGINAL DEGENERATION



PMD vs. KCN

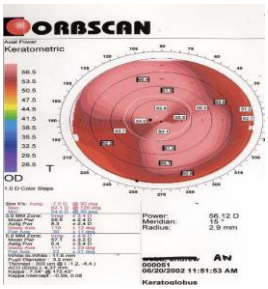


PROGNOSIS FOR PELLUCID

- **Lens fitting is difficult due to inferior apex**
- Central rings show AR/ Inferior rings show WR
- Fitting flat causes bearing and on K (steep) causes too much seal off
- Larger lenses needed due to low positioning/ glare
- **CAREFUL MONITORING-**
- **Poor SX Candidate**

KERATOGLOBUS

- A diffuse thinning of the cornea to 1/3-1/5 the normal thickness
- It is noted early in life and progression is minimal
- Associated with Ehlers-Danlos Syndrome and Leber's Congenital Amaurosis
- Acute hydrops



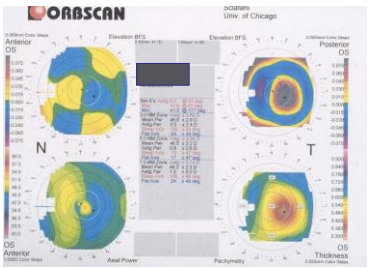
POSTERIOR KERATOCONUS

- Rare developmental defect
- Focal indentations of the posterior cornea with overlying stromal scarring
- Anterior curve not effected
- Descemets' s membrane and endothelium are always present but may be abnormal in the area of thinning

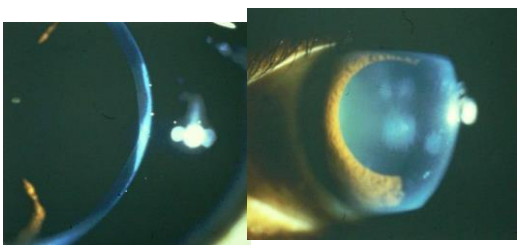
POSTERIOR KERATOCONUS

- Associated Ocular Disease
  - Lens abnormalities, choroidal or retinal sclerosis, PPMD, retinal coloboma, optic nerve hypoplasia, ptosis, iron rings, and posterior synechia,
- Systemic Associations
  - Mental retardation, webbed neck, hypertelerism, short stature, superior placed lateral canthi, genitourinary abnormalilites

POSTERIOR KERATOCONUS



POSTERIOR KERATOCONUS



Thank you for your attention