Current Clinical Management Options for Corneal Dystrophies and Degenerations

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Corneal Conditions

- **Map-Dot-Fingerprint** 58
- **Reis-Buckler** 1
- **Granular** 19
- **Lattice** 9
- **Macular** 1
- **Posterior Polymorphous** 1
- **Fuch's** 65
- **Salzmann's Nodular** 4

Classification of Corneal Dystrophies

- The Cornea Society
- The International Committee for Classification of Cornea Dystrophies (IC3D)
- New classification system is based on:
  - Anatomy: the corneal layer(s) affected
  - Genetic information: gene locus of the mutation
  - Onset, signs, symptoms, course and examination techniques
- Each dystrophy is also categorized based on the level of evidence available to support it

Classification of Corneal Dystrophies (Weiss et al, 2008)

- **Category 1**
  - A well defined corneal dystrophy in which a gene has been mapped and identified and specific mutations are known.
- **Category 2**
  - A well defined corneal dystrophy that has been mapped to on one more specific chromosomal loci, but the gene(s) remain to be identified.
- **Category 3**
  - A well-defined corneal dystrophy in which the disorder has not yet been mapped to a chromosomal locus.
- **Category 4**
  - This category is reserved for a suspected new, or previously documented, corneal dystrophy, although the evidence for it, being a distinct entity, is not yet convincing

Our Approach

- **Front to back** - anterior to posterior
- **Typical age of onset**
- **Symmetry of the disease**
- **Mode of inheritance** - AD or AR
- **Affected eye** - OD, OS or OU
- **Type of opacity/deposit**
  - Size, shape and location
- **Management options**

Dystrophies

Characteristics of Corneal Dystrophies

- Primary disorders – no systemic involvement
- Bilateral & symmetrical
- Early onset – usually by 3rd decade
- Involvement of only one layer of the cornea (usually)
- Central corneal location
- Inheritance: autosomal dominant (AD)

Epi- & Subepithelial Dystrophies

1. Epithelial Basement Membrane Dystrophy (EBMD)
2. Meesmann's Dystrophy
3. Reis-Buckler's Dystrophy

EBMD

- **Map-Dot-Fingerprint / Cogan's Microcystic / Anterior Basement Membrane**
- *Most common of all anterior corneal dystrophies*
- Bilateral
- Present in adult life (3rd decade or older)
  - Rarely seen in children
- Equal predilection for both sexes
- ***IC3D:*** *Cat 1 and most cases have no inheritance pattern*

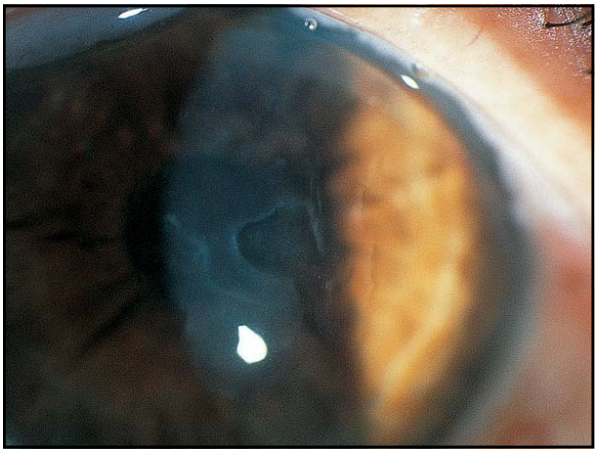
EBMD

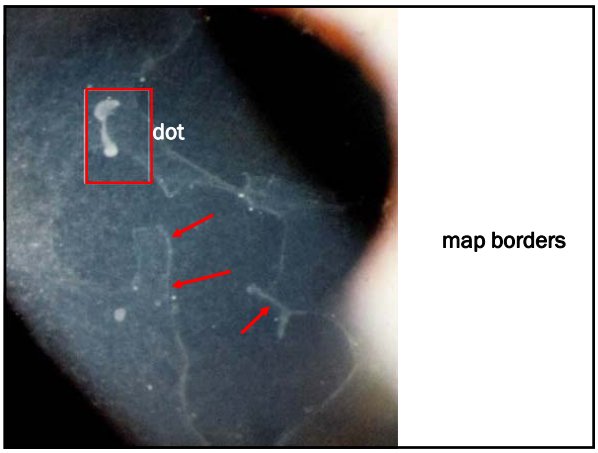
- Corneal signs:
  - Geographic **maps**
    - Most common corneal finding
  - Intraepithelial **dots**/microcysts
  - **Fingerprint** lines
- Corneal signs can appear together or individually

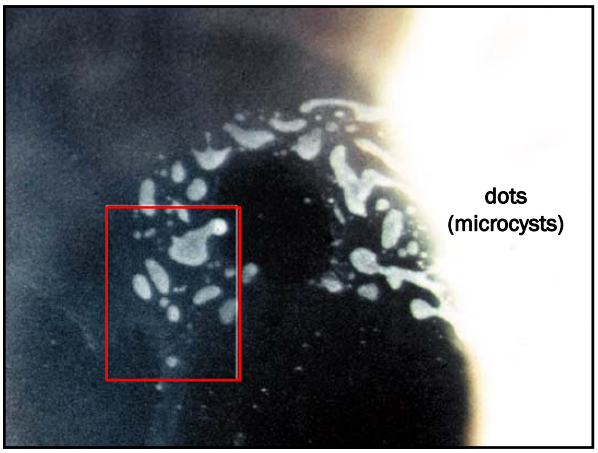
Metabolic Process

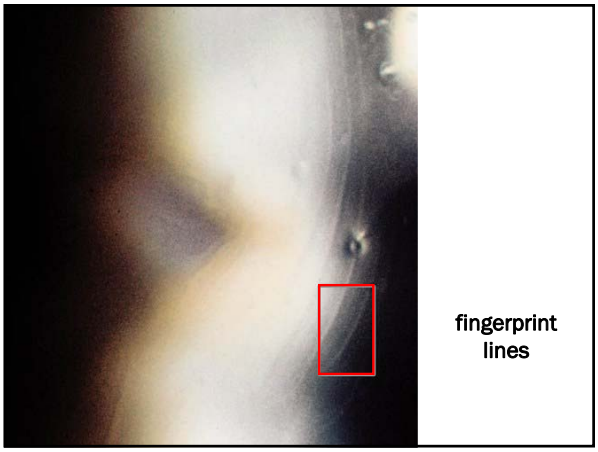
1. Abnormal turnover, maturation and synthesis of the corneal basement membrane
2. Blocks normal migration of epithelial cells towards the surface

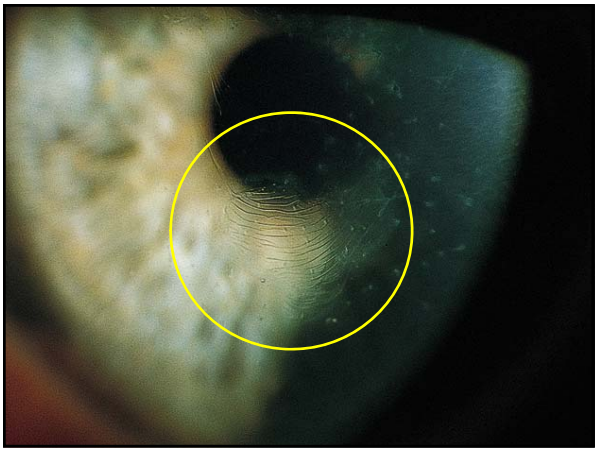
- **"Maps" & "fingerprint lines"** are due to thickening of the basement membrane
- The microcysts produce the **"dot"** appearance which are trapped cellular debris











Symptoms

- Patients are usually asymptomatic with map and fingerprint findings
- When microcysts erupt:
  - Mildly uncomfortable to painful
  - Tearing
  - Photophobia
  - Transient blurring of vision

Symptoms

~10% of all EBMD patients will develop recurrent corneal erosions (RCE)

Treatment & Management

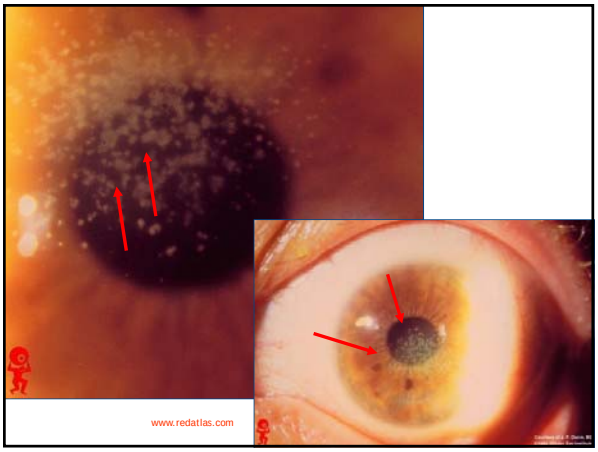
- Mild cases:
  1. Lubricating eye drops
  2. Hypertonic drops (Muro-128)
  3. Bandage soft contact lens
- Mild/moderate cases:
  - Tx is the same as in "mild cases" but add:
    1. Oral tetracycline or doxycycline (50mg x bid)
    2. PF, FML, Lotemax or Alrex
  - Corticalsteroids inhibit proteinases that weaken epithelial binding

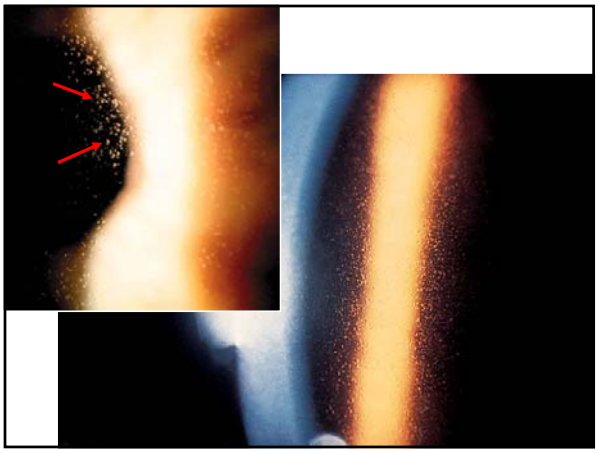
Treatment & Management

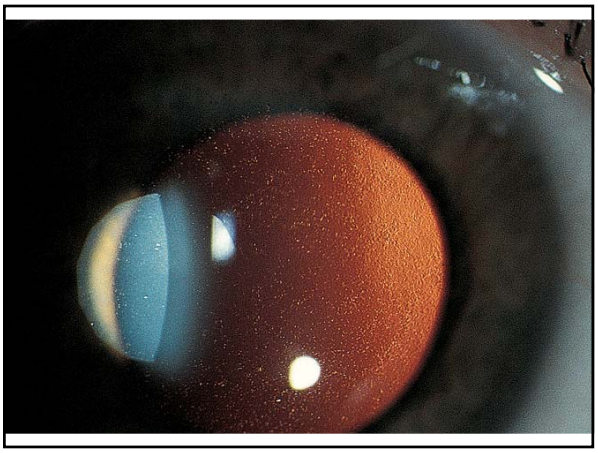
- Severe cases (with frequent RCE) :
  1. Phototherapeutic keratectomy (PTK)
  2. Epithelial debridement
  3. Stromal puncture
     - If not within the visual axis
- SCLs or GP lenses to mask surface irregularities and to improve vision
- Use bandage SCL's for comfort and protection during episodes of corneal erosion

Meesmann's Dystrophy

- **Juvenile hereditary epithelial dystrophy**
  - Onset is during the first 5 years of life
  - Bilateral/symmetrical
  - AD inheritance
  - ***IC3D:*** *Cat 1*
- Signs:
  - Numerous epithelial vesicles most dense within the inter-palpebral region







Symptoms

- Asymptomatic until middle age
- Mild ocular irritation
- Photophobia
- Blurred vision caused by ruptured vesicles
- Recurrent erosions are rare

Treatment & Management

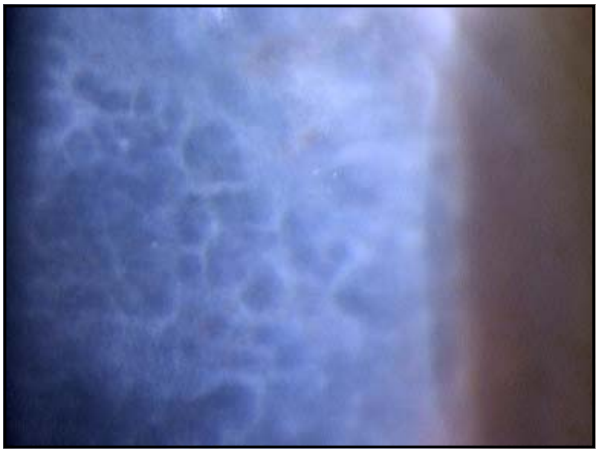
1. Most patients require no treatment!
2. Keep corneal surface lubricated
3. Monitor the status

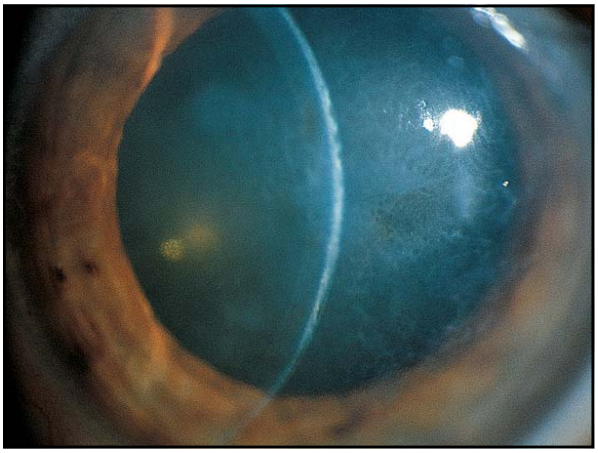
Reis-Buckler's Dystrophy

- Dystrophy of Bowman's Layer
- Bilateral/symmetrical
- AD inheritance
- Onset is in the 1st decade of life, poor vision by age 20
- Phenotypic appearance similar to ***Granular Dystrophy*** of the stroma
- ***IC3D:*** *Cat 1*

Reis-Buckler's Dystrophy

- Signs:
  - Honeycomb shaped opacities within the central cornea
  - Opacities seen in Bowman's layer & anterior stroma
    - Bowman's layer is replaced by a connective tissue layer
  - Epithelial binding to Bowman's is poor
    - Frequent RCE's





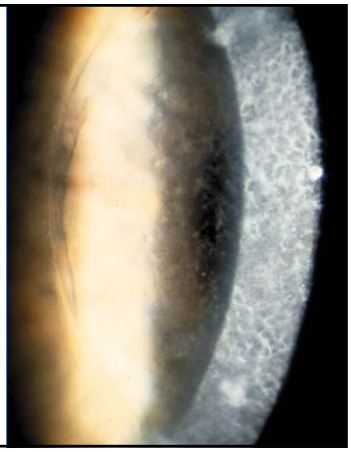
Symptoms

- Decreased vision
  - Clouding of the cornea
  - Irregular astigmatism
- Progresses from central to peripheral cornea
- Pain/discomfort due to RCE
  - Frequency of RCE increases with severity of the dystrophy

Treatment & Management

1. Monitor & keep corneal surface lubricated
2. Manage episodes of RCE
   - Bandage contact lenses
   - Ocular surface lubrication
   - PTK with Mytomycin C
3. Penetrating keratoplasty (PK)
   - High incidence of recurrence in donor graft

Recurrence

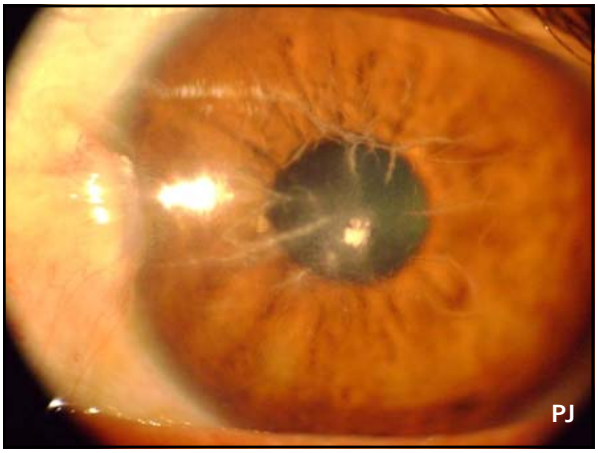


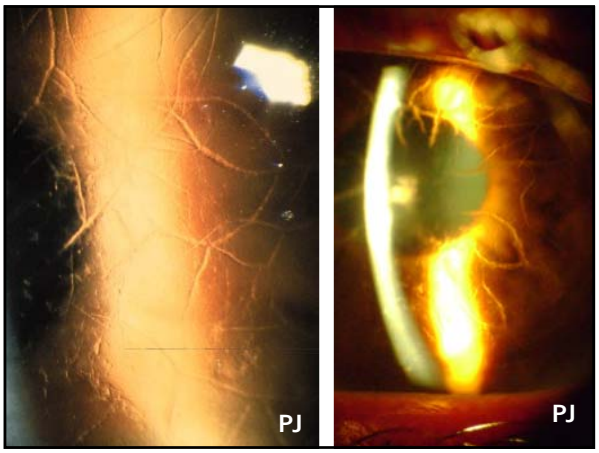
Stromal Dystrophies

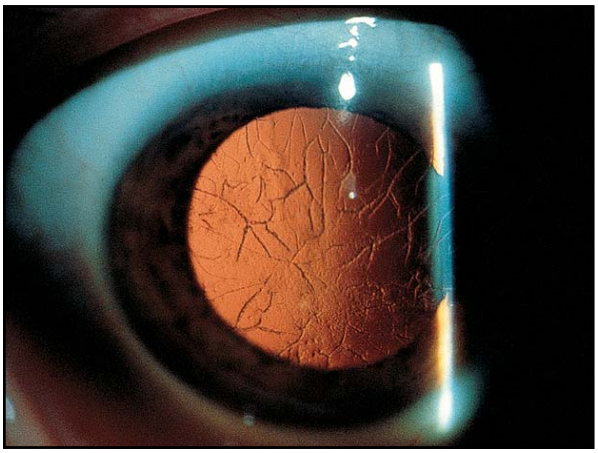
- Lattice Dystrophy
- Granular Dystrophy
- Macular Dystrophy
- *IC3D: All 3 stromal dystrophies fall under Category 1*

Lattice Dystrophy

- Bilateral/symmetrical/AD
- Onset is in the 1st decade of life
- Signs:
  - Branch-like thin lines
  - Mid-stroma
  - Opacities are made of amyloid
- Progression from central to peripheral cornea
  - The peripheral 1mm always remain clear





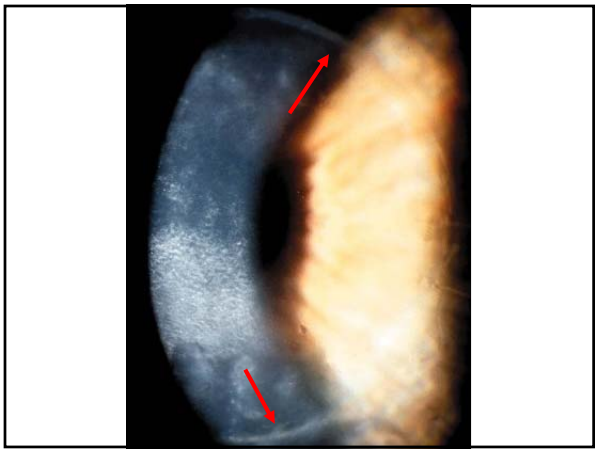


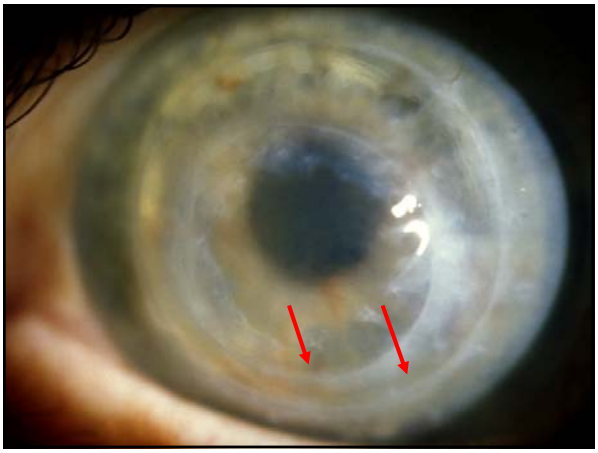
Symptoms

- Decreased visual acuity
  - Distortion the architecture of the corneal lamellae
  - Visual impairment by fourth decade
- Amyloid opacities disrupts Bowman's layer, leading to frequent episodes of RCE

Treatment & Management

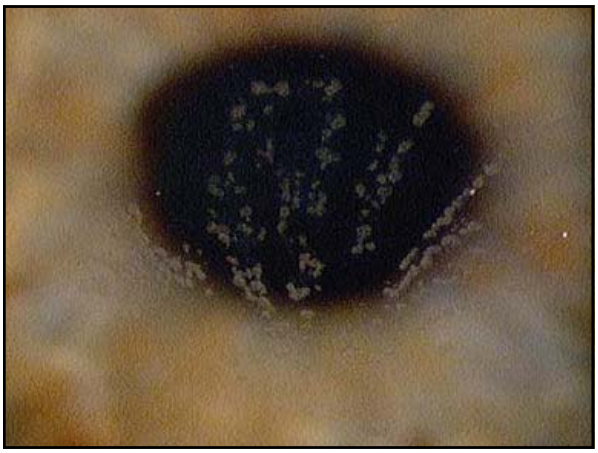
- Manage episodes of RCE
  - Lubrication
  - BCL
  - PTK / epithelial debridement
- Penetrating keratoplasty
  - Preferred treatment
  - Only 5% recurrence rate in the corneal graft
  - Occurs in periphery & migrates centrally

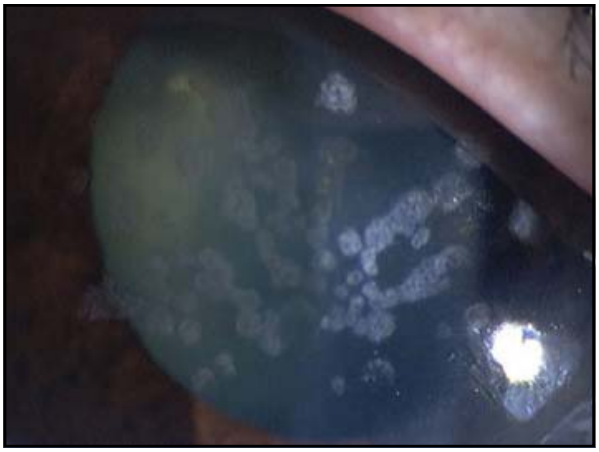


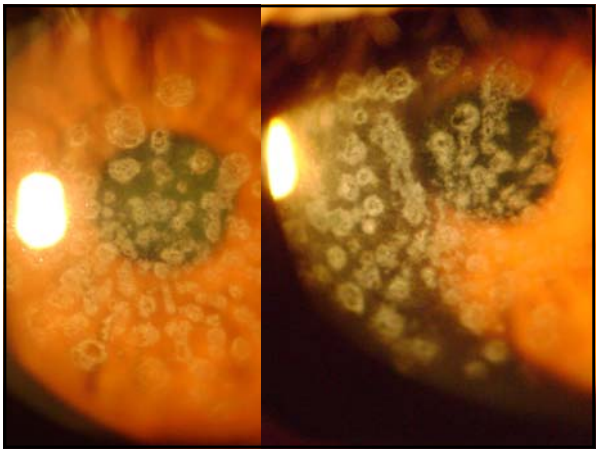


Granular Dystrophy

- *Most common of the stromal dystrophy*
- Bilateral/symmetrical/AD
- Onset in 1st decade of life
- Signs:
  - Snowflake or breadcrumb opacities made of eosinophilic hyaline
  - Opacities located in the anterior stroma
  - Progresses from central to peripheral cornea







Symptoms

- Visual prognosis good in early years
- Glare & photophobia
- Decreased VA's around 5th decade of life
  - As the opacities increase in number/size, they begin to coalesce
- RCE episodes are rare
  - Eosinophilic hyaline doesn't disrupt Bowman's layer

Treatment & Management

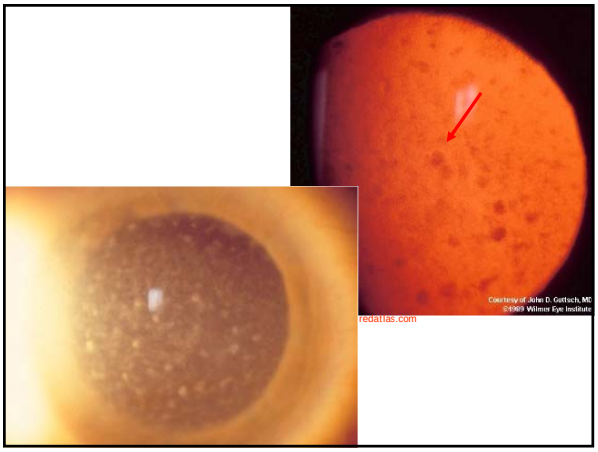
1. Manage episodes of RCE
   - Ocular lubrication
   - Bandage contact lenses
   - PTK
2. Penetrating keratoplasty
   - High recurrence rate
   - Occurs in periphery & migrates centrally

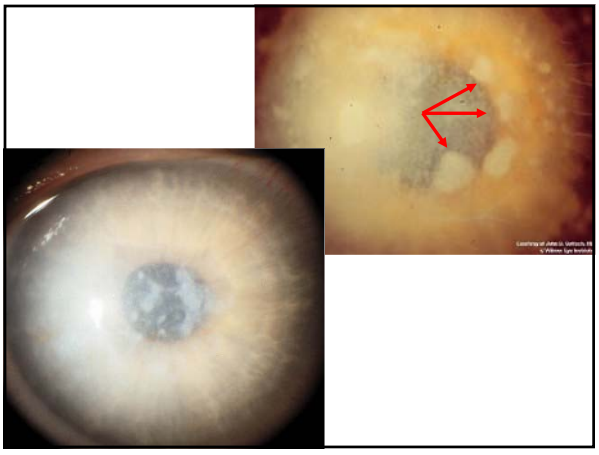
Macular Dystrophy

- Autosomal recessive
  - Only dystrophy with AR inheritance pattern
  - Affects patient more severely than AD dystrophies
- Onset in the 1st decade of life
- Opacities are made of glycosamine-glycan
  - Opacities eventually enlarge and coalesce with progression

Symptoms

- Vision is stable until 2nd decade
- Poor vision by 2nd or 3rd decade
- RCE's are very common





Treatment & Management

- Tinted cosmetic SCL's
  - Improve appearance
  - Reduce photophobia
- Bandage SCL's for episodes of RCE
  - PTK
- PK – Surgery of choice
  - Pts need PK sooner since VA is poor by 2nd-3rd decade
  - Recurrence in graft is rare
    - Occurs in periphery & migrates centrally

	Granular Dystrophy	Macular Dystrophy	Lattice Dystrophy
Onset of Deposits	1st decade	1st decade	1st decade
Onset of Symptoms	3rd decade or asymptomatic	1st decade	2nd decade
Heredity	AD	AR	AD
Reduced vision	4th or 5th decade	1st or 2nd decade	2nd or 3rd decade
Erosions	Uncommon	Very Common	Very Common
Opacities	-Discrete sharp borders -Intervening stroma is clear early but becomes hazy -Not to limbus	-Indistinct margins -Hazy intervening stroma -Extends to limbus -Endothelium affected	-Early refractile lines and dots -Subepithelial spots -Diffuse central haze -Limbal zone clear in mild cases
Corneal Thickness	Normal	Thinned	Normal
Deposit Type	Hyaline	Glycosaminglycan	Amyloid
Clinical Features	Clear limbal zone	Opacities reach limbus Cornea thinned	Lattice lines

Fuch's Dystrophy

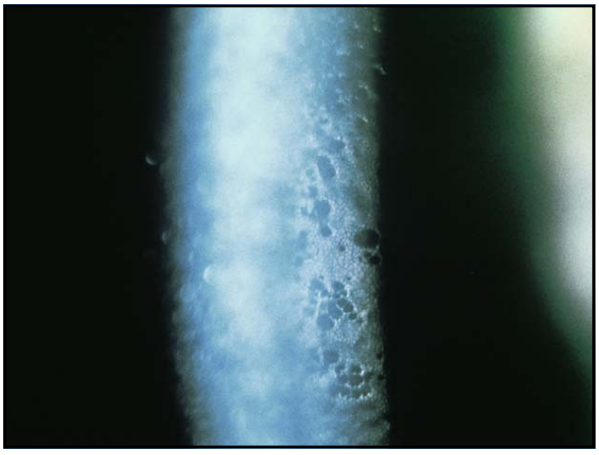
- Bilateral / AD
  - IC3D: Found no inheritance pattern
- Late onset (4th - 5th decade of life)
- More common in females
  - Literature reports equal predilection for both males & females
- Characterized by 3 stages
  1. Guttata
  2. Stromal or epithelial edema
  3. Corneal fibrosis or scarring

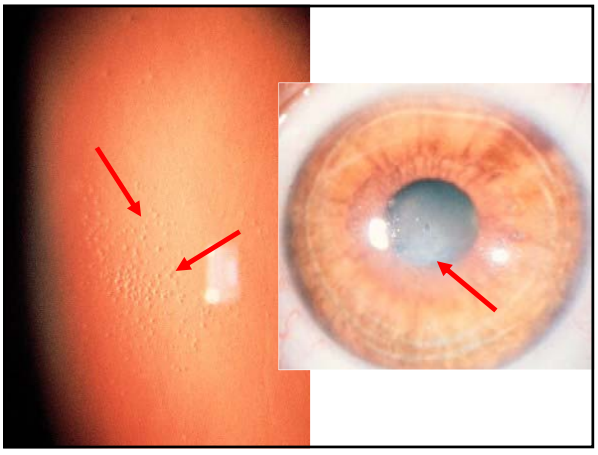
Metabolic Process

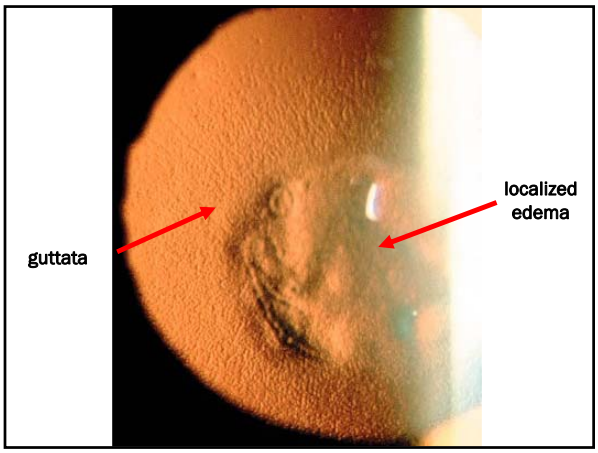
- Excessive production of basement membrane material by the endothelium
- Guttata is the accumulation of this material projected into the anterior chamber, causing endothelial cells to "drop out"

Fuch's Dystrophy

- Signs
  - Corneal guttata
    - "Orange peel" or "beaten metal" appearance
  - Best seen in retro-illumination
  - Stromal edema from endothelial decompensation
    - Worse in the AM
    - Pts report "blurry" vision that improves throughout the day
  - In severe cases
    - Epithelial bullae
    - Bullous keratopathy

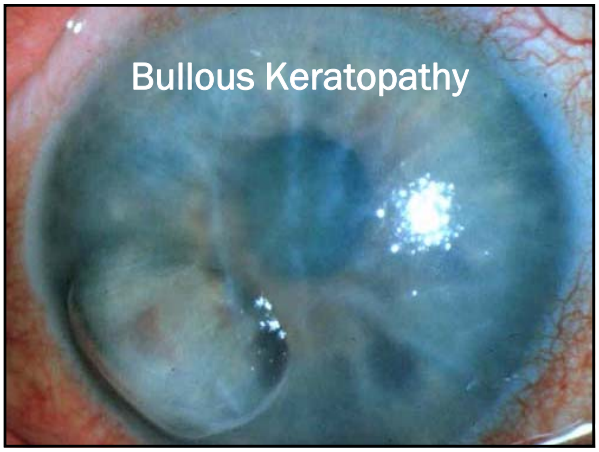


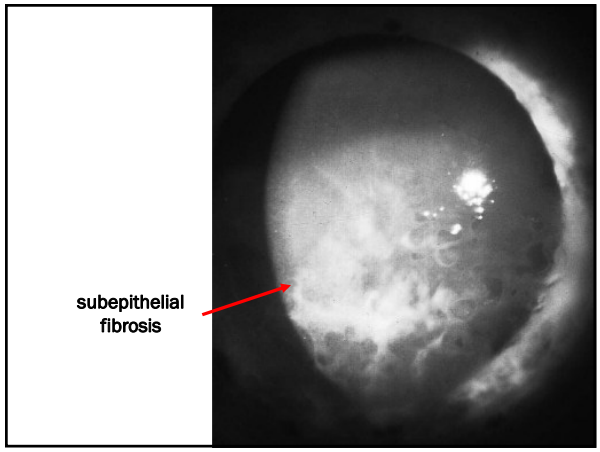




Symptoms

- Initial stages, blurry vision in the AM that improves throughout the day
- In later stages, epithelium also takes on water, may lead to bullous keratopathy
- Decreased VA's secondary to:
  - Corneal surface changes
  - Subepithelial fibrotic scarring
- Pain, discomfort, foreign body sensations
- Scarring from chronic edema





Treatment & Management

- Monitor early cases
- Hyperosmotics for edema
  - (Muro 128, 2-5%)
- Bandage SCL's
  - For discomfort
    - A high Dk SCL with full coverage
  - Can use hyperosmotics over a high water content disposable SCL

Treatment & Management

- Penetrating keratoplasty
  - Most common indication for PK
    - Clear grafts reported in 80% of pts 2 yrs post-surgery
  - Double procedure:
    - Cataract extraction performed at the time of transplant

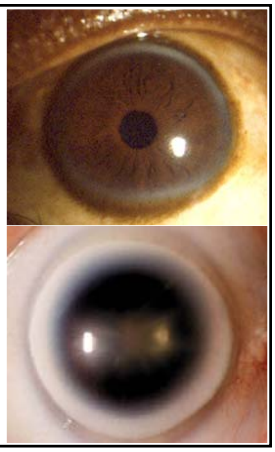
Degenerations

Corneal Degenerations

- Secondary changes
- Associated with **normal aging**
  - Presents later in life
  - Occurs in peripheral cornea
  - Little or no effect on vision
- Associated with **ocular insult**
  - Acute: bacterial keratitis
  - Chronic: recurrent uveitis
  - Can occur centrally
  - Affects vision

Corneal Arcus

- Most common peripheral opacity
- Bilateral & symmetrical
- Gray-white or yellow annular rings
- Clear zone between ring and limbus
- 10% more common in males than females
- Vision is not affected



Arcus Senilis

- Theory: lipids leak from limbal blood vessels
- Seen in 75% of those over 70 and 100% of those over 80
- Occurrence in men under 50 is associated with ***systemic hyperlipidemia***
  - Refer for blood work

Corneal Arcus

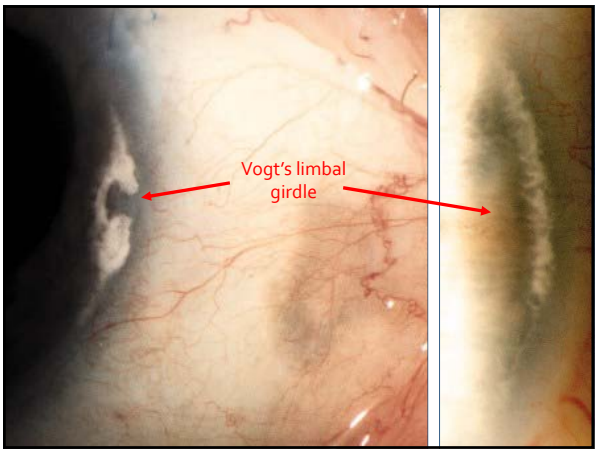
- Recent studies indicate arcus to be an indicator of elevated cholesterol *regardless* of age
- No specific hereditary characteristics
- ***What about UNILATERAL arcus???***
  - Watch for carotid occlusive disease
  - The side with arcus is normal
    - The side with faint or less arcus is occluded
  - Refer for doppler ultrasound

Treatment & Management

- No treatment necessary
- If patient is under 40 y.o., refer for serum cholesterol measurement
- *When questioned, patients will admit to having elevated cholesterol levels*

Limbal Girdle of Vogt

- White arcuate opacity in the inter- palpebral zone at 3 & 9 o'clock
- Symmetrical
- Two types
  - Type I
  - Type II



Limbal Girdle of Vogt

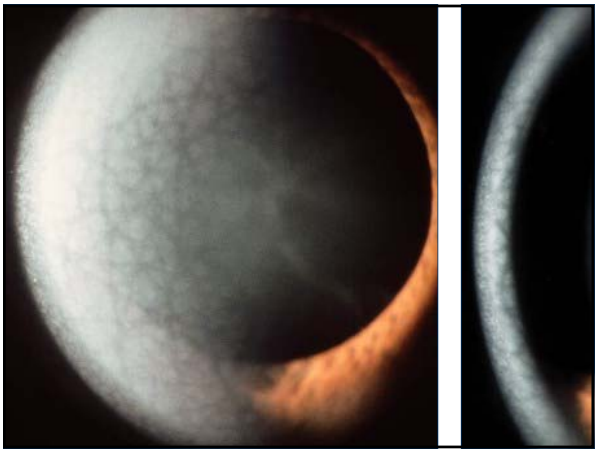
- Type I
  - Early form of calcific band keratopathy
  - Less common
  - Separated from the limbus by a clear zone
  - No extension towards the cornea
  - No treatment necessary

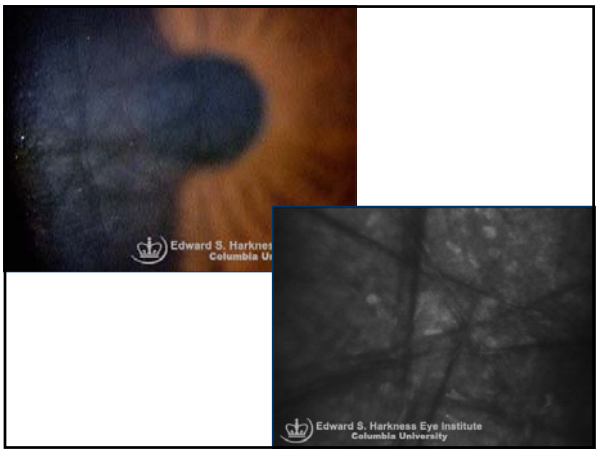
Limbal Girdle of Vogt

- Type II
  - White chalky arcuate band
  - Contiguous with the conjunctiva
  - Degeneration of elastic collagen at the level of Bowman's layer
  - VA's not affected
  - No pathological significance
  - No treatment necessary

Crocodile Shagreen

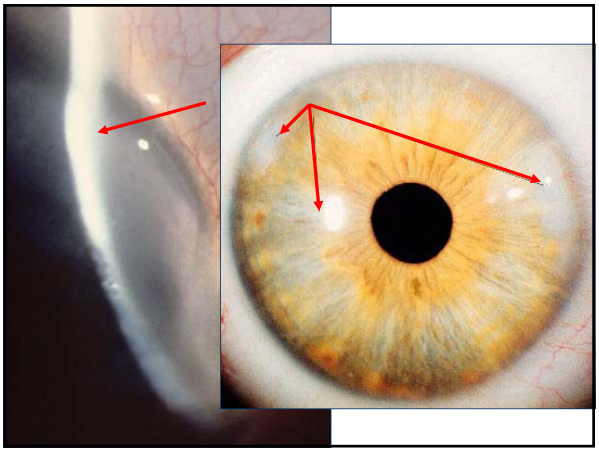
- Mosaic keratopathy
- Polygonal white patches separated by clear intervals
- Breakdown of Bowman's membrane
- Resembles a "mosaic"
- Appearance of cracked ice
- Very rarely symptomatic





Salzmann's Nodular Degeneration

- A non-specific corneal response to chronic insult:
  - Phlyctenulosis, trachoma, keratitis, or corneal inflammation
- Nodules are of hyaline plaques formed in between the epithelium and Bowman's
  - Oval, discrete, avascular, gray-white, elevated & opaque
- Peripheral or mid-peripheral
- More common in females



Symptoms

- Visual acuity is usually OK
  - Irregular astigmatism is common
- Treatment
  - Sugical peel
  - PTK - for superficial nodules
  - Penetrating keratoplasty
    - For lesions extending into mid-stroma
    - Recurrence is possible

Bandage Contact Lenses

Bandage Contact Lenses

- <u>Definition:</u> *Hydrophilic or rigid corneal lenses that are used for the treatment of acute or chronic corneal pathology*
- *What is the rationale in prescribing a bandage contact lens?*
  1. Pain relief
  2. Corneal protection
  3. Wound healing
  4. Enhance vision
  5. Corneal hydration
  6. Drug delivery

Billing & Coding

- *Disclaimer: I am not a billing expert*
- Bandage Contact Lens formerly 92070 is now
  - **92071**
  - **92072**
- This procedure is an ophthalmological procedure where a disposable, soft or extended wear soft, or a hard contact lens is fitted by the physician to treat a diseased or injured eye. Documentation should include indication for use, all lens parameters, use instructions and all follow-up.

Billing & Coding

- **92071**
  - *Fitting of a contact lens for treatment of ocular surface disease*
  - *DOES NOT include supply of lenses*
  - *Must use **99070** or other appropriate supply code*
- *92072*
  - *Fitting of a contact lens for management of keratoconus, initial fitting.*
  - *Submit subsequent visits under appropriate E/M code*

Billing & Coding 92071/92072

- These codes are not part of the patient's original office visit, it is in addition to it
- These codes are **UNILATERAL** codes
  - Bill twice if fitting bilaterally
  - Use a modifier for right (-RT) & left (-LT) eyes

Billing & Coding 92071/92072

- Never use 9231X code when prescribing a bandage contact lens
- The office visit to remove bandage lens is not included
  - Bill under an appropriate E/M code

Thank you for your time and attention.