81 y. o. w. f. c/o chronic fbs ou. “Equate” AT ou qid + w/ minimal relief. Coumadin, Premarin, Antivert, Toprol XL, Diovan, Lanoxin, Tricor, Xanax.
Tx: D/C cheap tears
Freshkote OU bid - tid
Warm comp. qpm x 15 min.
Lid hygiene qod
Pathophysiology

- Morphologic changes in BM of epithelium
  - Inadequate hemidesmosome formation
    - “fingerprint” lines
  - Fibro-granular deposits
    - “map” and “dot” deposits

Basement Membrane Dystrophy

- Most common anterior corneal dystrophy!
- AD
- 2% of population
EBMD

Tx
- Early
  - Lubrication
  - Hypertonic gtts
  - DTS control
- Later
  - Recurrent erosions
    - Debridement or PTK
    - BCL
    - Anti-inflammatory Tx
      - Topical and Oral
      - Antibiotic Tx


Dry Sponge v. Wet Sponge
RCE Treatment

- **Abrasions**
  - Antibiosis
  - BCL
  - Anti-inflammatory
    - Topical and/or Oral Tetracyclines

- **Chronic Treatments**
  - Copious Lubrication
  - Topical Sodium Chloride
  - Topical Anti-inflammatory
  - Oral Tetracyclines

RCE Treatment

- **Chronic Treatments**
  - Surgical
    - PTK
    - PRK
    - Stromal Puncture
    - Epithelial Debridement
Anterior K Dystrophies

- **Reis Buckler’ s Dystrophy**
  - Scarring Bowman’ s membrane, 1st decade

- **Meesman’ s Dystrophy**
  - AKA juvenile epithelial dystrophy
  - Extremely rare, multiple small clear cysts
Anterior K Dystrophies

- Lisch dystrophy
  - ? EBMD
- Thiel-Behnke dystrophy
  - ? Reis Buckler

35 year-old male
Myopia
-5.75 OD; -6.50 OS
Pre-operative ocular exam was normal
  - Scant small white subepithelial flecks in cornea OU
Ladarwave CustomCornea LASIK in 2005
  - Amadeus Microkeratome used
Uncorrected distance vision
- 20/20 (6/6) OU for 1st 6 months

1-year visit

- Vision 20/60 OD; 20/25 OS
- Interface opacities in the flap interface, more prominent OD > OS

Treatment
- Flap Lift and scrape with mitomycin C OD
Vision was 20/80 OD and 20/25 OS

- Penetrating keratoplasty OD
Granular corneal dystrophy Type II (Avellino Corneal Dystrophy - ACD) (Granular-lattice corneal dystrophy)

Excimer laser corneal ablation
- Contraindicated¹-⁴
- Includes PTK with or without mitomycin¹
- PRK, LASEK (Advanced surface ablation)²
- LASIK³⁴

³) Lee WB et al. JRCS 2007;33:133-8
Stromal K Dystrophies

- **Granular Type I (Groenouw)**
  - Sm. granules 2nd to 3rd decade
  - Progressive, coalescent, recurring lesions


- **Granular dystrophy Type II (Avellino)**
  - AD, cross b/t Type I and Lattice
  - Tx: DALK, PK

Stromal K Dystrophies

- Lattice dystrophy Type I
  - Most common stromal dystrophy
  - Lattice or cracked glass appearance, 1st decade
  - PTK, DALK, PK

H&E stain of cornea with lattice. Note pink amorphous deposits in stroma
Congo red stain, highlighting amyloid

Apple-green birefringence of amyloid with cross-polarization.

Stromal K Dystrophies

- Macular dystrophy
  - AR, gray dep. w/ stromal haze, limbus to limbus.
  - 1st decade, PK rather than PTK
**Mucopolysaccharide deposits**

Staining w/ Alcian Blue

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<table>
<thead>
<tr>
<th>Table 1: Corneal Stromal Dystrophies</th>
<th>Table 2: Mnemonic for remembering corneal stromal dystrophies</th>
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<tbody>
<tr>
<td>*Lattice</td>
<td>*Marilyn – Macular Dystrophy</td>
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<tr>
<td>*Granular</td>
<td>*Monroe – Mucopolysaccharide</td>
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<tr>
<td>*Avellino</td>
<td>*Always – Alcian Blue stain</td>
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<td>*Gelatinous Droplike dystrophy</td>
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<tr>
<td>*Congenital Hereditary Stromal Dystrophy</td>
<td>*Angeles – Amyloid</td>
</tr>
<tr>
<td></td>
<td>*California – Congo Red</td>
</tr>
</tbody>
</table>
Posterior K Dystrophies

- Fuchs’ Dystrophy
  - Most common post. dyst
  - AD, 4\textsuperscript{th} - 6\textsuperscript{th} decade, var. exp., polymegathism
  - Dec. Va, k edema late
Posterior K Dystrophies

- Palliative Treatment:
  - Topical osmotic agents
- Definitive Treatment
  - DSAEK
  - DMEK

Congenital Hereditary Endothelial dystrophy
- AR (Most Common)
  - Edema at birth, nystagmus
- AD
  - Opacities dev. Later, no nystagmus
Make it stop!!!

- 34 y. o. w. m. c/o 1 d Hx of severe FBS, photophobia, OD, after waking this morning.
- Fourth occurrence
- Initial occurrence fingernail injury
RCE Healing

- S/P Debridement
  - BCL or AMT
  - Antibiotic
  - Steroid
  - Oral Tetracycline or Azythromycin

RCE Treatment

- Epithelial debridement
  - Weck cell sponge
  - Beaver Blade
  - Foreign body spud / debridement hoe

- Bandage CL / Amniotic Membrane
  - Topical antibiotic
  - Topical anti-inflammatory
  - Oral Tetracycline antibiotic therapy
RCE

- Treatment
  - Surgical
    - PTK / PRK
    - Debridement – Amniotic Membrane CL
C. B.

- 40 y. o. w.m. LEE 2 years prior. Si Hy DW CL wearer. Obvious non-compliant SCL wear.
- Noticed dec. Va, FBS, injection OS 1 d prior. No discharge or pain. Removed CL OS immediately.
- BVa: 20/20 OD, 20/100 OS
- P, EOM, MB, CF = Normal
- $T_A$: 16 mmHg, OU
C. B.

- Fundus = Normal
- Phone consult w/ derm. = prob. Erythema Multiforme
- Dx: Presumed HSK, OS
- Tx: Viroptic OS q2h, P. F. A. T. prn.
  - Start Valtrex 500mg bid, finish Z-Pack as instructed.
- Derm. consult. following day.
C. B.

2 d f/u

- BVa OS: 20/30
- IOP OS: 17
- EM confirmed by bx of skin lesion.

Add: Lotemax qid
Cont. Viroptic qid x 3d.
C. B.

- 8 d f/u
- BVa: 20/20
- IOP: 16 mmHg
- Tx: Lotemax qid x 1 more wk. then stop.

Herpes Simplex Keratitis

- Most common cause of corneal blindness in US, 50,000 new or recurrent cases/yr.¹

- Epithelial Disease
  - Vesicles, Dendrites, or Geographic ulcers.
  - Check K sensitivity
    - Viroptic q2h or 8x/d
    - Zigan gel
    - Debridement?
    - Orals?
      - HEDS II
  - **Absolutely NO STEROIDS w epi. defects.

---
HS Stromal Keratitis

- Recurrent disease
- ISK
  - Retained Viral antigen in stroma.
- Nec. Strom. Keratitis
  - Dense infil., ulceration, and necrosis
- HEDS I
- HEDS II
  - Sig. benefit tx w/ orals.

**H S Endothelialitis**

- KPs, Cells and Flare
- Stromal / Epi. edema
- No neovasc. or infiltr.
- Disciform, Linear, Diffuse
- ? CMV
- Tx: Steroids, Top. Antivirals, and Oral Antivirals (1-2 gm/d)*


**HZO**

72 y. o. 1 wk hx “shingles” c/o dec. Va OS. Acyclovir 800 mg 5x/d.

Va: 20/30 OD, 20/100 OS
20/100
(-) K staining

Cont. Valtrex 800mg 5x/d
Tobradex oph ung bid / Lotemax qid

3wk: Cont. Lotemax bid

"Pseudo-dendrites" v. "Dendrites"

Pseudodendrites: Tree branches w/o terminal end bulbs.
Dendrites: Tree branches with terminal end bulbs.
“I Can See Clearly Now”

- 40 year old female
- 30 years of blindness in left eye following children’s Motrin at 10 yo
- VA 20/30 OD; LP OS
Diagnosis

- Toxic Epidermal Necrolysis (Lyell syndrome)
- Total Stem cell deficiency and keratinization of the cornea
- Diffuse Symblephara and/or ankyloblephara

Treatment Options

- Total Keratolimbal allograft and penetrating keratoplasty
- Artificial cornea transplant
PRE-OP
Light Perception

POST-OP; 20/40

(6 Months Postop)
Kpro and Amniotic Membrane
Reconstruction

20/30 Distance Acuity
Boston KPro Indications

- Two prior failed grafts
- Poor prognosis for cadaveric grafts
- Vision less than 20/400
- Suboptimal vision in other eye
- No end-stage glaucoma or retinal detachment

KPro Components

- Type 1 Device (PMMA)
- Type 2 Device
- Collar Button
- Back Plate
- Inert Titanium Fixation Screw
Summary

- Keratoprosthetics present a viable treatment option for eyes with previously inoperable corneal blindness
- Visual acuity potential remains excellent
- High retention rates are possible
- Keratoprosthetics have a favorable comparison with high risk corneal grafting
Anterior Segment Predictors of Death

- Xerophthalmia
- Toxic Epidermal Necrolysis
  - (Stevens-Johnson Syndrome)
- Mooren’s Ulcer
- Fabry’s Disease

Mooren’s Ulcer

- Pain
- Unilateral > Bilateral
- Males > Females
- Limbus to central cornea
- Autoimmune
- Prognosis
  - Depends on underlying autoimmune disorder
  - Can lead to death if not treated properly
Fabry Disease

- AKA: Alpha Galactosidase-A Deficiency
- X-Linked
- Acroparesthesias, angiokeratomas, hypohidrosis, tinnitus
- Ocular signs
  - conj. bv tortuosity, radiating psc, K verticillata
- Kidney damage, cardiac damage and stroke potential = life threatening cond.

FABRY DISEASE

MOOREN’S ULCER

National Fabry Disease Foundation, 2009

Klaus D. Teichmann, MD, FRCS, FRACO; Michael D. Wagoner, MD.
The Masquerade: A Case for the FDA

- 17 year old female
- Contact lens wearer
- Redness, pain, and decreased vision for 4 days
- Medical History benign except for “fever blisters”

Initial Examination
Treatment Before Referral
- Contact lens cessation
- Dendrite Debridement
- Viroptic 1 drop every 2 hours
- Pred Forte qid / Valtrex 1 g bid added 4 days before prior (suspected stromal spread)

Referring Diagnosis
- HSV Keratitis

Ring Infiltrate & Radial Perineuritis

Two Weeks later after Steroids
Differential for Dendritic Lesions

- Herpes simplex/zoster
- Epstein Barr
- Tyrosinemia
- Healing epithelial defect
- **Rosacea**  *Lee, Mannis, Schwab. Cornea 2005*
- **Superficial hypertrophic dendritic epitheliopathy (SHDE)**  *Lee et Mannis, Cornea 2006*
- Acanthamoeba

Diagnostics

- **Cornea Culture**
  - Chocolate and blood agar
    - No growth
  - Sabouraud agar
    - No growth
  - Nonnutrient agar
    (E. Coli overlay)
    - Positive

- **Cornea Scraping**
  - Positive for ameobic cysts
Confocal Microscopy
(Confoscan 4)

40 X View

Diagnosis & Treatment

- Acanthamoeba Keratitis
  - 1) Stop Pred Forte & Antivirals
  - 2) Start antiamoebic agents
    - Polyhexamethalene biguanide (PHMB) .02%
    - Chlorhexidine .02%
    - Brolene (Propanidine)
    - Desmadine (Hemxamidine)
  - 3) Cycloplegia
QUESTIONS?

THANK YOU!!!!

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