I. Basics

papilla
  vascular response
  if giant, the differential includes atopy, vernal, GPC, prosthesis, suture

follicles
  lymphatic response

acute
  EKC, pharygoconjunctival fever
  medicamentosa (epinephrine, neosynephrine)
  Parinaud's oculoglandular syndrome
  HSV primary conjunctivitis
  Newcastle's conjunctivitis
  with acute follicles, always check lid margin for HSV vesicles, ulcers

chronic
  adult inclusion conjunctivitis
  toxic
  r/o sarcoid
  r/o GPC, vernal conjunctivitis

membranes
  conjunctivitis
  ocular cicatrical pemphigoid
  erythema multiforme
  Stevens Johnson syndrome
  Srogers syndrome
  atopy
  Symblepharon
  scieroderrna
  burns
  radiation burns
  trachodema
  EKC
  sarcoid
  drugs

filaments
  exposure (keratoconjunctivitis sicca, neurotrophic, patching
  recurrent erosion)
  bullous keratopathy
  HSV
  superior limbic keratoconjunctivitis psoriasis
  med
  aerosol keratitis
  radiation
  Thygeson's SPK
  diabetes mellitus
  retained FB

ptosis

Enlarged Corneal nerves
  MEN TIIb
  ichthyosis
  Hanson's
  Kconus
  Refssums
  Fuchs corneal dystrophy
  old age
  failed PKP
  congenital glaucoma
MEN IIb
AD with thick corneal nerves, medullary thyroid cancer, pheochromocytoma, mucosal neuromas, and marfanoid habitus
thickened lid margin with rostral lashes, thick lips, epibulbar neuromas
cafe au lait spots, periungual, lingual neuromas
often confused with NFI
often die early from amyloid producing thyroid cancer in 10-20 year old with distant mets at dx
thick nerves precede the cancer!

corneal edema
whenever epithelium disrupted, can stimulate iritis via reflex arc

epithelial
intracellular first
intercellular with microbullae
then subcellular with frank bulla

stomal
all extracellular

factors
imbibition pressure = IOP - swelling pressure (nl 50)
fluid into cornea from IOP, glycosaminoglycans's
fluid out of cornea by dehydration, pump
IOP is inverse with swelling pressure
with nl endothelium, high IOP-- epithelial edema
with nl IOP, poor endothelium-- stromal edema

rx
mild
muro 128, hair dryer, control IOP
moderate
soft CL, cycloplegia, PK, conjunctival flap

Brown McClean syndrome
peripheral edema in aphakic, pseudophakic with orange pigment on endothelium

Stains
Flouresecin staining when disruption of cell-cell junctions
Rose Bengal stains with disruption of precorneal tear film
cell death increases permeability to these dyes, but Rose Bengal can still be blocked with tears
II. Lids/conjunctiva

**Congenital**

**Epitarsus**
fold of conjunctiva on palpebral lid

**Osler Weber Rendu**
hereditary hemorrhagic telangiectasia
AD, dilated blood vessels on palpebral conjunctiva in star/sunflower shape 1-3 mm, violaceous, blanches, increased with age
telangiectasia that bleed easily, rarely in retina and look like HTN or DM retinopathy
problems with epistaxis and GI bleeding

**Congenital lymphedema**
XLR, AD, usually massive edema of legs
 dysplasia of lymphatics

**Medicamentosa**

**Anaphylactic**
sulfonamides, bacitracin, anesthetic

**Allergic**
with eczema, SPK, red eye
atropine, homatropine, aminoglycosides, antivirals

**Toxic**
often after 1 wk of use, especially keratoconjunctivitis sicca pts
papilla, redness, SPK, no itch
aminoglycosides, antiviral, preservative

**Follicular**
months to years later
big follicles, pannus, SPK
psuedottrachoma syndrome
atropine, miotics, epinephrine, antivirals
Conjunctivitis

EKC adenovirus 8,9,13; can cause symblepharon
EBV virus with mono can cause conjunctivitis, keratitis
Newcastle disease virus with poultry exposure
measles may have papillary conjunctivitis with white avascular spots ci
caruncle/conjunctiva like Koplik spots in mouth

Parinaud's OGS

cat scratch
children with cats, hx of scratch, sneeze, 2 wk latency, nodule in
superior or inferior conjunctiva
intense chemosis, injection, lymph nodes may appear up to 2 wks later
systemic fever, malaise, maculopapular rash
DX:
Hanger Rose test 90% sensitivity skin test
Warthin Starry stain for bacilli
RX:
Doxycycline 100mg bid x 1 month

tularemia
lymph nodes, fever, chills, vomiting, pneumonia but ocular involved
<5%
necrotizing, ulcerating conjunctivitis, corneal ulcer, optic neuritis,
dacryocystitis, panophthalmitis
rabbit hunters, hx of tick bites with punched out lesion
DX:
with agglutination titers 1: 160 or higher in 2 weeks and peak
in 4-8 wks
RX:
streptomycin, tetracycline

sporotrichosis
spherical elastic movable nodule, pink then purple then black and
necrotic
multiple subcutaneous nodules along lymphatics, multiple yellow
nodules in conjunctiva
sporotrichosis conjunctivitis seen in HI pts
no systemic illness
DX:
culture on Sabouraud's
RX:
KI 1 ml/day

Misc

sarcoid                      leptothrix                      chancroid
glanders          Crohn's
fungi               fungi               TB
lymphgranuloma venereum
culture conjunctiva and scrape, blood clx if febrile, VDRL, FTA,
PPD, viral titers, biopsy

Reiters
bilateral conjunctivitis, iridocyclitis, urethritis, polyarthritis
fever, lymph nodes, pericarditis, pneumonitis, myocarditis
think if chronic nonfollicular mucopurulent conjunctivitis
SPK, corneal infiltrate, corneal neovascularization
steroids, chlamydia/dysentery antibx?

Floppy Lid
SLK
burning, no itch or discharge, symp worse than signs
corridor hyperemia, velvety papilla upper tarsus
+Rose Bengal, micropannus, fine SPK, filaments in 1/2
50% with mild thyroid dysfunction
soft contact lens can also cause similar picture
RX:
- scrape conjunctiva, pressure patch, soft contact lens, resect conjunctiva

Thygeson's keratitis
can often mimic SEI
recurrent hx with quiet white eye
bilateral raised heaped up epithelium with microcysts seen in retroillumination
can mimic HSV
coarse grey white lesions slightly elevated without flourescein stain
dramatically responds to topical steroids often after 2 doses, taper in one wk
some need chronic therapy due to rebound if steroids stopped

molluscum
SPK, pannus, follicles, pseudotrachoma

Graft vs Host
S1                conjunctiva hyperemia
S2                chemosis, exudate
S3                pseudomembranous
S4                corneal epithelial slough
higher stage correlates with increased severity of disease and mortality
keratoconjunctivitis sicca most commonly, cicatticial lag, ectropion, persistent
epithelial defects, iritis
Chlamydia

**Trachoma**
- superior pannus, SPK, corneal infiltrates,
- lid destruction and exposure are key elements
- tetracycline, erythromycin, or sulfonamides x 3 months
- S1 conjunctival follicles, cytoplasmic inclusion bodies
- S2 inflammation, increased corneal pannus
- S3 scanning flesherでは pits (limbal depressed necrotic follicles), Arlt's line
- S4 end stage

**Adult TRIC**
- 5% with urethritis, 1-2 wk latency, meibomianitis, lid edema and redness
- follicular conjunctivitis, EKC-like SEI
- NO membranes
- superior pannus (not seen in EKC)
- doxycycline 100mg bid 10 days or erythromycin 250mg qid 3 wks, treat partner

**Newborn TRIC**
- no follicles, more discharge, + pseudomembrane
- 4-12 days post partum, r/o GC
- Giemsa incl bodies 40%, 90% with + clamydiazime
- otitis, pneumonitis in 15%, recurrence 20%
- Erythromycin syrup 50mg/kg qid x 2 wks, treat mom

**LGV**
- Parinaud's OGS, follicular conjunctivitis, conjunctival granuloma
- can have keratitis, corneal neomacularization, anterior uveitis

**Atopy**

**RX**
- allergy testing and environmental control
- compresses, pressure patch, air conditioning, pulse steroids
- vasoconstrictors, Acular, livostin, mast cell stabilizers
- immunosuppressives

**Hayfever**
- rapid, lid swelling, chemosis (pale palpebral conjunctiva), itching, mucus, dellen
- pressure, rhinitis/asthma, episodic, may have no signs
- T1 hypersensitivity, elevated tear IgE, eosinophils in scraping in chronic cases
**Vernal (VKC)**

- bilateral seasonal young (3-25 year old) in warmer climates, M>F
- FHx of atopic allergies
- self limited average 4-10 years
- ITCH (worsens in evening, dust, lights, wind, rubbing), clear tears
- ropy discharge but lids don't get crusted or stick together unless bacterial superinfection
- GPC (may see a membrane form at the slit lamp), limbal involvement more in blacks (can be 360 degrees)
- Homer Trantas dots (clumps of degenerated eosinophils), clear elevated cysts
- SPK, flour dust of epithelium, intraepithelial cysts, shield ulcers usually upper cornea
- pseudoarcus, myopic astigmatism, associated with keratoconus, rare corneal neovascularization
- >2 eosinophils/hpf pathognomonic, increased tear histamine
- topical cyclosporine 2% qid can be used as alternative to steroids

**Atopic (AKC)**

- M>F, teens to 40's, burns out by 40-50 year old, small papilla, milky edema, corneal neovascularization
- fix of atopic eczema (3% of pop), similar to venial findings but no seasonal changes
- symblepharon, foreshortening of inferior fornix, usually lower palpebral conjunctiva affected
- in severe cases bilateral cataracts (anterior subcapsular, or posterior polar)
- 10% of all atopic dermatitis associated with keratoconus, iritis, cataract
- RD from pars plana tears or ora dialysis (can have photoreceptor outer segments in anterior chamber which look like cells)

- increased serum IgE, few eosinophils in scraping and rarely free granules
- systemic antihistamines, nasal cromolyn, topical mast cell stabilizers, steroids and cream, doxycycline
- path: increased T helper, macrophages, increased class II HLA similar to OCP and rosacea.
  - more complex than simple mast cell allergic rx
  - No BM deposition

**GPCofCL**

- usually develops within first year of lens wear, but RGP can develop after years
- also seen in art eyes, sutures
- r/o VKC (no tear histamine, no free eosinophil granules, only 1/4 have eosinophils in scrapings
early mucous discharge, mild itch with increased in severity, then pain with CL, blurred Va
papilla may stain (sign of activity), whitish material on lens
conjunctival injection or chemosis when severe, thick shem of mucus, soft <
hard CL
switch brands/ types of lenses, increased use of enzyme, topical steroids,
mast cell stabilizers, stop lenses

**Contact dermatitis**
earythema, itching, scaling of lids, papilla
chronic meds (Neomycin most commonly), metals, cosmetics, false lashes
cement, fingernail polish

**Blepharitis**
all get hot compresses and lid scrubs bid
can try antibiotic ungt qhs to qid, if severe may use blephamide
chronic doxycycline antibx associated with vaginitis, allergy,
photosensitivity, take on empty stomach, no breastfeeding

**Staph**
collarettes, ulcers at base of lashes, papilla, purulent discharge, marginal
ulcers
absent, thin, broken, misdirected, or white lashes
younger, F (80%), short duration, dry eyes

**Seborrheic**
older, more chronic, oily margins, crusting, papilla, follicles, dermatitis, dry
eyes

**Meibomian Gland Dysfunction (MGD)**
acne rosacea, conjunctival injection, SPK, Rose Bengal stain, more burning,
bulbar injection
Doxycycline 100mg bid for a month
maybe even chronically bid/qd

**Rosacea**
F, 30-50, rhinophyma, telangiectasia, pustules, erythema
MGD, marginal keratitis, inferior corneal pannus with subepitelielial
infiltrates

**Subconjunctival hemorrhage**

<table>
<thead>
<tr>
<th>Anemia</th>
<th>ASHD</th>
<th>conjunctivitis</th>
<th>decreased platelets</th>
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<tbody>
<tr>
<td>DM</td>
<td>menses</td>
<td>nephritis</td>
<td>subacute bacterial endocarditis</td>
</tr>
<tr>
<td>Trauma</td>
<td>trichinosis</td>
<td>valsala</td>
<td>vascular anomaly</td>
</tr>
</tbody>
</table>
Superglue in eye
    toxic to endothelium
    cold water compresses to loosen adherance
    wait 3-4 hours, can cut lashes

Kaposi's sarcoma
    Grade I, II patchy and flat and <4 mon duration, Gr III > 3mm thick
    on bulbar conjunctiva or eyelid
    standard rx with radiation
    consider double freeze thaw cryo for Gr I, II on eyelids
    simple excision of conjunctiva for bulbar conjunctiva leaving bare sclera
    excision s/p vessel diueation by flourescein angiograrn for Gr III on
    conjunctiva

III. Cornea

Congenital Anomalies

megalocornea
    >13mm, bilateral, most commonly XLR, usually isolated, nonprog
    r/o glaucoma, increased risk factor for ectopia, cataract, glaucoma
    rarely associated with renal cell cancer, r/o congenital glaucoma
    nl endothelial density
    F carriers may have slightly larger cornea

microcornea
    <10mm, hyperopes, AD>AR
    20% with angle closure glaucoma, usually eye nl
    r/o nanophthalmos, microphthalmos, trisomy 13, Ehlers Danlos, dwarfism

Anterior seg dysgenesis AD

Posterior embryotoxin

Axenfelds
    hyperteloric, small shoulders, facial asymmetry, 1/2 with glaucoma
    (syndrome)

Rieger's
    craniofacial anomalies, mental retardation, Marfans, 1/2 with
    glaucoma
    syndrome with maxillary hypoplasia, telecanthus, microdontia,
    skeletal and spine deformities

Peter's
    80% bilateral, clearing of leukoma with waiting, glaucoma
    T1- nl lens and no systemic involvement
    T2- lens involved, bilateral, often glaucoma
    r/o von Hippel's internal corneal ulcer (no lens abnormal)
local posterior keratoconus (endothelium/descemet present)

**Haab's striae**

**Congenital opacities**
- facets, nebula, macula, leukoma
- corneal keloid probably from intrauterine trauma

**Sclerocornea**
- bilateral, often with systemic and other ocular problems

### Infections

#### Viral

**HSV Primary**
- conjunctivitis in children, bilateral, fever, preauricular adenopathy, URI, 3-9 days incubation
- varied present (mild follicular conjunctivitis to pseudomemb)
- lid vesicles, ulcerative bleph, 1/2 get small fleeting corneal dendrites
- SPK, conjunctival dendrites

**HSV Recurrent**
- most common cause of central infectious corneal ulcer
- sunlight, fever, stress, memses, steroids
- 1 yr 25%, 2 yrs 50% chance of recur
- lids (psuedozoster)
- rare follicular conjunctivitis
- epithelial keratitis
  - SPK, dendritic, geographic, marginal
  - shaggy borders, ghost scars of prior dendrites, decreased corneal sense
  - often with mild stromat edema
  - 1/4 recur in 1 yr, 1/2 recur in 2 yrs
  - metaherpetic lesion from poorly healing epithelium, gray thickened heaped up edges

**RX:**
- debride, viroptic 9x/day taper off 2 wks
- see in weekly, don't overtreat
- limbal lesions resist antivirals, slower healing

**diff dx of dendrite**

- CL filaments
- HZO Mygeson's
- tyrosinemia TII healing corneal abrasion

**Disciform**
- immune rx at endothelium
- central corneal edema with fuie KPs under edema, mild iridocyclitis, increased IOP, Wessley ring
diff dx HZO, local bullous keratopathy
self limited 2-6 months with variable scar
cycloplegic, may use steroids, Muro 128, antiviral cover, later PKP

Peripheral Stromal Keratitis
atypical, r/o marginal ulcer, ?association with HIV, corneal neovascularization, interstitial keratitis

Necrotizing
live virus but even - on bx
dense deep stromal infiltrate
no pain, mild iridocyclitis, increased IOP, corneal neovascularization, indolent
self limited 2-12 months
antivirals with steroids and slow taper (even yrs)

Endothelitis
progressive corneal edema with line of KP (looks like PKP rejection), uveitis, increased IOP
acyclovir

Uveitis
iritis with diffuse iris atrophy
multifocal choroiditis

HZO Primary
chickenpox
rare disciform keratitis
SPK
Limbal pustules
lid lesions
usually benign, may treat if corneal involvement

HZO Secondary
2% of adults > 60 year old, is not harbinger of cancer in healthy people
pathology: nerve damage, ischemic vasculitis, inflammatory granulomatous rx
lid vesicles clear in 3 wks and can result in ptosis
conjunctival hyperemia, vesicles, episcleritis
SPK, microdendrites, corneal neovascularization, uveitis, glaucoma, sectoral iris atrophy
stromal nummular keratitis, scleritis (nod>diff, limbal can spread to cornea)
1% with optic neuritis, Homees, EOM palsy (25%)
decreased corneal sensitivity leads to neurotrophic ulcers
RX:
cycloplegic, Acyclovir 800mg 5x/day x 7 days try to start within 72 hrs
topical steroids for iritis
Zovirax or warm Borow's compresses qid
consider prednisone 60-100 mg po x 3 days
pain medication
r/o HIV especially if < 45 year old or risk group
if microdissemination, pt may be im. immunocompromised
consider IV steroids, use IV acyclovir

**HZO Neurological syndromes**

**postherpetic neuralgia**
- pain decreased with time (80% gone in 1 yr), worse if pt is older
- Rx capsaicin, stellate ganglion block, cimetidine, carbamazepine, TCA

**necrotizing angiitis**
- with contralateral hemiplegia and death in 15% wks to months later

**PML-like syndrome**
- with seizures, decreased function, and death

**Bacterial keratitis**

- risk factor CL, trauma, atopy, prior HSV, dry eyes/exposure, bullous keratopathy, OCP/SJ, abnormal lid position
- Staph/Pseudomonas are more than 50% of all cultured cases
- Worst are Pseudomonas, Strep (B-hemolytic, pneumoniae)
- direct epithelial penetration by GC, H egypticus, Diptheroids, Listeria
- Pseudomonas often with hypopyon, diffuse inflammation, epithelial edema
- Staph often with distinct borders, may satellite
- consider calcium alginate swab soaked in trypticase soy broth
- blood, chocolate, tliio, sabourouds for fungus

**Phlyetenular ulcer**
- pinkish white limbal elevation with gray crater, can be on conjunctiva
- children 10-20, allergic rx to staph, TB, Candida, Chlamydia, nematodes
- early Teri4xen's can look similar

**Marginal ulcer**
- gray limbal ulcer usually with clear cornea all around, early corneal neovascularization
- allergic rx usually to staph at 8/10 and 2/4 oclock
- can progress to ring ulcer, usually spread toward lhbus not centrally
- can get superinfected
Acanthameoba

mimics HSV early, later ring infiltrate, often misdiagnosed as HSV
symptoms wax and wane with photophobia, FBS, severe pain
first abnormal epithelium (SPK, persist defects, SEI, edema, whorl like patterns, dendritiform lesions)
then central/paracentral stromal infiltrate with satellites, turns into ring
neurokeratitis with cuffing of nerves
risk factor contamin CL, corneal trauma, r/o topical anesthetic abuse
bx, stains faster and better than culture
use spray fixative instead of air drying
IFA, calcofluor white, conjugated lec-tin fluorescein stain
culture on non-nutrient agar with E. Coli overlay
RX with neosporin, Brolene 0.1%, neomycin, and clotrimazole 1% ql5-qlh, +/-
ketoconazole 200 mg po bid, no steroids
once infection in peripheral cornea, poor candidate for graft

Fungal

trauma, south, mostly filamental, Fusarium, Aspergillus >50 %
Candida 10% in older, keratoconjunctivitis sicca, looks Re bacterial
feathery edges, endothelium plaque, satellite lesion
can penetrate Descemet’s, culture Sabourauds and blood agar
RX:
first line natamycin 5% susp (50mg/ml) q1hr,
Ampho 0. 15% (1.5 mg/ml) especially for candida
flucytosine for yeasts

interstitial keratitis

90% congenital lues with eventual bilateral involvement in 10-20 year old, can be recurrent
acute stage- salmon patch of Hutchinson, KP, stromal infiltrate, corneal edema., pain, photophobia, discharge
often with Hutchinson's teeth, deafness, salt/pepper retina, ON atrophy
usually present endstage with ghost vessels, guttata like bumps, retrocorneal hyaline ridges, fibrous strands into anterior chamber
10% acquired, unilateral, often sectoral, 10 yrs after infection
rare focal avascular interstitial keratitis with vessels
CL induced corneal neovascularization looks very similar
get serology, PPD, and rx for latent lues if present

| TB       | leprosy       | HSV
|----------|---------------|----|
| Mumps    | LGV           | sarcoid
| Kaposi's sarcoma | Hodgkins | Incontinentia pigmenti
| mycosis fungoides | HZO       | protozoan/heiminths

**Cogan's Syndrome**
- tinnitus, periarteritis nodosa, 30-40, decreased hearing, syst vasculitis
- remember congenital lues also with decreased hearing
  - serology

**Infectious Crystalline keratopathy**
- no epithelial defect, feathery "crystalline" edaes, associated with chronic steroids eg PKP
- poor response to topical therapy, bx usually for dx
- Alpha strep viridens, rx with conc topical bacitracin

**Degenerations**

**Pinguecula**

**Pterygia**
- risk factor UV exp especially < 5 year old, no glasses, no ha@ equatorial living
- excision, radiation, conjunctival transplant or flap, mitomycin
- surgery for astigmatism, VF changes, cosmesis, restricted EOMS, inflammation

**Amyloidosis**
- subepithelial, salmon color, avascular
- primary and mycloma tend toward mesenchymal deposit
- secondary to organ deposit
- metachromasia crystal violet, flourescent thiaflavine T, biref@@gence/di@hroism
- Congo Red, +with Siruis Red
  - **primary localized**
    - most common form with palpebral conjunctival asymmetry
      - (brown/yellow waxy firm subconj nodules)
    - lattice is special form
    - can make cornea into pudding
    - does NOT affect lids, but can be orbital presenting as VI n palsy
  - **primary systemic**
    - bilateral symmetric yellow or ecchymotic lid papules, light near dissoc
    - vitreous opacities, EOM palsies, proptosis, glaucoma
secondary local
after chronic inflammation, trichiasis, keratoconus, granular dystrophy
salmon to yellow, fleshy, waxy nodular lesion on cornea

secondary systemic
most common in general medicine and doesn't usually affect eye
lids may be purpuric

Involutional
arcus, hassal-henle bodies, Vogt’s limbal girdle
farinata (lipofuscin, dots, commas in deep stroma)
crocodile shagreen

Deposits

Band Keratopathy
in Bowman's chronic disease, system increased Ca, Phos, mercury, hereditary gout with urates, renal failure uveitis, long standing glaucoma, interstitial keratitis, phthisis, pilocarpine, dry eyes rx with 3% EDTA, scrape and sponge until it clears

Spheroidal degeneration
bilateral M>F, golden brown spheres in anterior stroma/Bowmans in palpebral zone risk factor age, exposure, probably elostatic degeneration of colilgen usually not elevated, small lesions, can be in conjunctiva, can have decreased Va

Salzmann's nodular degeneration
unilateral F>M middle age, non-inflamm end years later to old keratitis usually by hx gray white elevated subepithelial nodules at end of old pannus paracentral cornea asymmetry, tearing, photophobia, or decreased Va rx with simple excision

Coat's white ring FB remnant

Lipid Keratopathy
unilateral or bilateral, areas of -vascularized scars especially surgical often elevated, nodular, yellow between Bowmans and epithelium

Farinata
flour like dusting anterior to Descemet either central or entire cornea, usually bilateral nl Va, products of cellular degeneration

Marginal thinning

Mooren's
idiopathic unilat painful inflamed eye often inferiorly
trauma hx, rare perf, circumferential spread early
central edge undermined in stroma, blunt edge peripherally
two types, one benign, unilateral, in older, responds to surgery
the other, relentlessly progressive and bilateral in 25%, young
RX steroid (top, sys), lamellar keratoplasty with conjunctival
reseption, immunosuppressives
glasses and eye shield
check for Hepatitis C Ag

Terrien's
(NOT an ulcer)
quiet thinning superiorly with fine micropannus can spread 360 degrees
astigmatism in 20-40 and 60-70 year old M>F with steep central wall,
mild inflammation
later, lipid deposits at edge of pannus, epithelium intact, decreased Va
from astigmatism
rare perforation, rx with mild steroids chronically to suppress
inflammation
RX lamellar or PKP

Marzial keratolysis
autoimmune disease, most commonly rheumatoid arthritis
unilateral, inferiorly, may have infiltrate
can have rapid progression
stops if epithelium heals

Age related furrow
lucid areas of arcus, no inflammation, vessels, or perforation

Dystrophies

Anterior

MDF
>30 y.o. 10% have corneal abrasions
50% of pts with recurrent comeal abrasions have MDF
fingerprint best seen with retroillumination, maps with oblique,
usually negative flourescence staining
debrightment with cotton swab
lubricants, soft contact lens, ? excimer

anterior stromal puncture
topical anesthetic, debride area
use 23g needle and penetrate anterior stroma up to 1/3
depth
may perform even in visual axis but space punctures
further apart
antibiotic ointment and pressure patch
warn pts about extreme pain afterwards

Meesman's
childhood, with irritation, small decreased Va
thick BM, intraepithelial microcysts with peculiar PAS+ substance
no rx, retroilluminate, punctate staining

diff dx cystinosis

**Reis-Blacker's**
childhood, progressive gray white at Bowman's layer
reticulated pattern of scarring
painful recurrent erosions, by 50's marked corneal opacities
recur post PKP

**Stromal**
Marilyn Monroe Gets Hers in LA

**Granular**
onset in childhood with decreased Va later >201200
white deposits in anterior stroma
hyaline, + trichrome
periphery may be + for amyloid (ancestry to Avellino, Italy)
may recur yrs after PKP

**Lattice (Amyloid)**
childhood, central lines, dots, haze (ground glass)
recurrent corneal abrasion, decreased Va by 40 years old
T1 AD, can be deep, spares limbus, retroillumination, starts as dots
T2 AD, Meretoja- secondary amyloiddosis with progressive cranial neuropathy and skin changes
T3 AR, thicker lines across entire cornea easily seen, later onset
T3a AD, frequent corneal erosions

recurrence post PKP common

**Macular**
*AR, periphery involved*
dereduced Va in childhood with dimse stromal clouding
GAGs, inability to breakdown KS
gray white opacities with indefinate margins
may be even full thickness, guttata
alcian blue, colloidal iron stain
can recur after PKP

**Central crystallin (Schnyder)**
infant, progressive polychromatic crystals in stroma and haze arcus and limbal girdle
doughnut Re crystals are cholesterol and fats
Oil red 0 stain, frozen section
Va OK, associated genu valgum, hyperlipidemia

**Fleck**
nl Va, no symptoms
rare childhood with assymetry
grey white dandruff to periphery, increased GAGS, lipids
associated limbal dermoid, keratoconus, CCD, PXE, decreased corneal sense
atopy, cortical lens changes

**CCD (central cloudy dystrophy)**
bilateral symm nonprog dense with clear zones
nebulous gray areas with nl vision

**polymorphic stromal dystrophy**
bilateral sym progressive late in life
nl Va, associated with systemic amyloidosis

**PreDescemet farinata**
>30 year old, 0-red-O+, Sudan black+, and PAS+, deep stromal punctate above Descemet’s
nl Va

**Posterior amorphous stromal dystrophy**
rare bilateral child with good Va
extends to limbus, iris processes, endothelium disrupt focally
peripheral varient with clear center

**CHSD**
flaky feathery anterior central opacity at birth
can have decreased Va

**Posterior**

**guttata**
abnormal coliagen, orange peel look, if 2-3+, probably asympt Fuchs

**Fuchs's dystrophy**
guttata are focal retractile clumps of colilgen posterior to Descemet's AD, F>M, post menopause, bilateral and assymetric, rare in Orientals central guttata rust, pigment in endothelium (can have borders)
1 st degree relatives 40% with guttata, incomplete penetrance worse Va in mornings, humid days, increased IOP rarely pigmented guttata can decreased Va (20/60 range) in long-standing cases, subepithelial fibrosis, grayish Descemet thickening posterior collagenous layer can obscure all the guttata
diff dx of corneal edema- other endothelial problems, PPMI), disciform keratitis

**RX:**
only if symptomatic, treat as in other causes of corneal edema
if considering phacoemulsification check corneal thickness
if corneal thickness is > 0.6 mm then do triple procedure pachymetry is optional

**PPMD, posterior polymorphous dystrophy**
AD bilateral childhood, progressive, asymmetry
endothelial cells act like epithelium, may look like ICE
posterior surface with ridges, lines and circles with scalloped edges,
stromal edema, iridocorneal adhesions
best seen in retroillumination

**CHED**
bilateral congenital corneal edema, Descemet’s thick
due to degen of endothelial cells at 5 months gestation
T1 AR, most common, no pain or tearing
stationary, diffuse,
TII AD, 1-2 year old, painful tearing, progressive
no nystagmus, cornea blue ground glass

**Ectatic**

**keratoconus**
sporadic bilateral asymmetric with early astigmatism, F, 10-30 year old
associated with Down's, Marfan's, eye rubbing, vernal, HLA B27, MVP, RP, PMMA CL
Vogt’s striae, Fleischer ring, scarring can be familial (<10% of occur in blood relative)

**keratoglobus**
thinning greater in periphery, mild sear, not genetic, connective tissue disorders
associated Ehlors-Danlos

**Pellucid marginal degeneration**
bilateral clear inferior thinning 2 mm from limbus, 2mm in width
no iron ring, no cone but cornea protrudes above thinned area, no scar, striae
20-40 year old with high astigmatism

**IV. Misc**

dry eyes
80% show excess mucus, thinned tear film with debris, SPK, corneal mucus plaques and filaments
Rose Bengal (>3/9 score), TBT unreliable meniscus height variable
bothered by wind, reading, smoke, steep, @; of skin diseases, tearing/ mucus check conjunctival scanning, V, VII CN, avitaminosis A, lagophthalmos, sarcoid medications implicated include antihistamines, HCTZ, antibx, Inderal, Valium, Pyridium, Timoptic (damages mucus layer, decreased goblet ceffi)
up to 30% of pts with blepharitis have dry eyes due to lipid layer instability

**RX:**
tears, tarsorrathy, goggles, conjunctival flap
treat mucous with mucomyst 10% acetylcysteine (in reality, difficult to obtain, smelly to use)

**punctal plugs**
Collagen plugs leak
if great improvement perform permanent punctal closure

**cautery**
- local anesthetic
- insert into canaliculus
- apply while pulling back and reapply at opening

**Argon laser**
- 400 mW, paint puncta with skin marker
- 200 uM spot in ring 500 uM in center

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### Exposure

**Eyelid Burns**

**Immediate**
- often with swelling (<3rd degree)
- when resolves, lubricate q/2-1 hr
- moisten skin with antibx ungt/frequent saline soaks

**Intermediate**
- healing takes place 3-28 days later with corneas exposure developing rapidly
- surgical rx if significant exposure (chronic injection, flourescein staining, dulling of light reflex)

**Erythema multiforme**
- F>M, all ages but usually young, recurrent vesiculo bullous skin lesions on extremities
- sparing trunk, took for target lesion
- angiitis in dermis and conjunctiva
- **minor** lasting only 2-3 wks
- **major** (Steven's Johnson) M>F 6wks, usually self limited with fever, URI, HA, malaise
- TEN if extensive denudation, mild purulent conjunctivitis, corneal erosions, and loss of lashes
- the eyes are rarely involved with recurrence
- associated with bacteria (Mycoplasma), viral (HSV) infections, drugs (sulfonamides, penicillin, aspirin, dilantin), allergy, connective tissue disorders, vaccines
- reported after topical sulfonamides, scopolamine, tropicamide, proparacaine

**RX:**
- topical steroids, glass rods ?, lubrication, wet dressings, surgery for trichiasis and dry eyes

**OCP**
- F >60 with vesiculobullous skin disorder usually extremeties, inguinal without scar, but localized form on head with atrophic scars
- ocular and mucous membrane involvement
- active disease with small gay conjunctival mound, conjunctival hyperemia/edema
- bilateral dry eyes with epidermiliation, progressive subconj scarring, thin flaccid conjunctival bullae
decreased motility, trichiasis, eosinophils in scraping, Ig in BM, increased pathogens in lids

Drugs associated with (?induced) OCP are IDU, PI, pilo, T1/2, epinephrine

HLA DR4/DQW3, incidence 1:20,000

**RX:**
- steroids (topical and systemic)
- Cytoxan in severe cases
- Dapsone
  - for mild involvement
  - check for G6PD prior to therapy
  - 25 mg bid with gradual increase, stop at 50 mg/day
  - check LFT, CBC, and Met HgB

**Pemphigus vulgaris**
- acantholysis of intraepithelial vesicles, flaccid easily broken blisters, middle age
- no scarring, conjunctiva rarely involved
- catarrhal or purulent conjunctivitis, rarely trichiasis

**Bullous Pemphigoid**
- benign, >60 y.o., tense tough, subepidermal bullae
- may cause shrinking, but rare

**Epidennolysis bullosa**
- blisters after mild trauma
- ocular problems with dystrophic varient with symblepharons
- junctional have recurrent corneal erosions

**Recurrent Erosions**
- painful erosions usually in early morning
- pts often aware of something wrong before they open eyes
- pain similar to prior episode
- often with MDFor trauma from nail, paper

**RX:**
- pressure patch, muro 128, soft contact lens, mild steroids (decreased Bowman's inflammation)
- debride epithelium for MDF
- anterior stromal puncture is better in traumatic cases (see MDF)

**Metabolic disorders**

**Alkaptonuria (ochronosis)**
- AR, no homogentisic oxidase, so homogentisic acid sent to kidneys and with alkali urine turns brown
- pigment of eyes, ears, nose, dura, arthritis, sclerotic heart valves, early ASHD
- brown dots near limbus at Bowman's layer, triangular patches pointing to MR, LR, pigmented piguecula, and coloring of tarsal plates/lids
- quinaacrine and hydroquinone can case ochronosis
- no ocular or medical rx
Cystinosis
AR, fine polychromatic needle like crystals under epithelium that migrate deeper (more peripheral than central) so by 7 y.o. full thickness cornea, in conjunctiva
decreased corneal sensation, glistening dots on iris
photophobia may be incapacitating, can have blepharospasm
dx with conjunctival bx. crystals are water soluble so ask for frozen sections
**infantile**
previously fatal from CRF but now have kidney transplants,
salt/pepper retina
**adolescent**
with less nephropathy, no retinopathy
**adult form**
nl kidney, benign and usually dx with slit lamp
transport enzyme out of lysosome is missing
**RX:**
oral and topical cysteamine forms mixed disulfide which transports cystine out
difficulty is in early diagnosis

Fabry's
XLR, but F carriers have corneal lesions, decreased ceramide trihexosidase
pinhead hyperkeratotic vascular eruptions on breast-, buttocks, and extrem,
parathesias hands/feet with hot weather/exercise, ASHD, CRF with lipid buildup
corneal verticillata, periob edema 1/4, PSC cataract 1/2, conjunctival
aneurysms ½ associated with ON edema, ON atrophy, CME

**diff dx**
chloroquine indomethicin amiodorone
chloropromazine striate melanokeratosis

Familial LCAT deficiency
lecithin:cholesterol Acyl Transferase deficiency, AR
small gray stromal dots in early childhood, peripheral arcus
no ocular therapy

Gauchers
AR sphingolipidosis, excess glucocerebroside
TI-adult onset nonneuropathic, most common, anemia and thrombocytopenia
TII-infant, retroflexion, TIII-adolescent milder neuro
Ocular-pinguecula like lesion with Gaucher cells, can bx

Hyperlipoproteinemia
5 types based on class of lipoprotein
xanthelasma/arcus (most with type II)

Mucolipids
cornea cloudy in GM I type 1, T1, TIII
**Mucopolysaccharidosis (MPS)**
- entire cornea cloudy, colloidal iron, 7 types
- all AR except Hunters (XLR) with gargoyle faces, heart/liver/skeletal/mental retardation/deafness
- RPE changes, ON atrophy, photophobia, papilledema due to hydrocephalus
- gargoyle cells are large storage vacoles in histiocytes
- dx by conjunctival bx of upper bulbar conjunctiva without sedation

**Tyrosinemia Type II**
- palm/sole painful erosive/hyperkeratotic skin lesions with mental retardation
- pediatric bilateral painful dendriticlike lesions with purulent debris as a result of crystal rupture of cells
- ddx of HSV (but no pseudopods, min staining, stellate, and no response with antivirals)
- NOTE eye lesions may be first
- tyrosine load test, serum levels, diet may prevent mental retardation
- decreased tyro aminotranferase, or no parahydroxy phenylpyruvate hydroxylase

**Wilson**
- AR, chromosome 13q,
- Kaiser Fleischer ring is golden, red, or green, 1-3 mm wide, at Descemet’s, and starts superiorly
- rare sunflower cataract
- check copper level and ceruplasmin

**DDX:**
- primary biliary cirrhosis, progressive intrahepatic cholestasis of childhood, chronic active hepatitis
- multiple myeloma

**RX:**
- penicillamine

**Avitaminosis A**
- night blindness, keratoconjunctivitis sicca, limbal Bitot’s spots
- pts have decreased mucus, GI, GU, and pulmonary problems
- corneal ulcer, keratomalacia, decreased corneal luster
- malabsorption syndrome: cystic fibrosis, pancreatic disease

**Iris Degenerations**

**Senile degeneration**
- senile miosis with rigidity
- may notice increasing blue color
- moth eaten pupil ruff

**Iridoschisis**
- age related, trauma, angle closure, and miotics
- bilateral over 65 year old, M=F, not familial
- often with shredded wheat appearance

**Ischemic**
- sicke cell, quinine, angle closure, vasculitis from HZO, HSV
Neurogenic
neurosyphilis, lesions of ciliary ganglion

V. Tumors

NonPigmented

Papillomas

viral
younger, pedunculated, bilateral, multiple
fornix, palpebral, caruncle, toxic keratitis
RX observe, cryo, beta radiation
incomplete or partial excision can multiply them

neoplastic
older, unilateral, single, bulbar/perilimbal
sessile, severe conjunctivitis
Rx excise with specimen

CIN (intraepithelial neoplasia)
unilateral, 95% males, light exposed areas, papilloma virus
gelatinous, thickening with leukoplakia, or papilliform (use Rose bengal)
histo-epithelium disarray, disturbed maturation, dysplasia to carcinoma in situ
limbal, excise and cryo (double freeze thaw), scrape Bowman's, recurrence up to 50%

Lymphoma
diff dx benign hyperplasia, dermoid, orbital fat lacrimal gland
salmon color, few vessels, no symptoms, flat smooth and soft, fornix
most conjunctival are localized without systemic spread
prognosis same for unilateral or bilateral (stage I-E), 10% eventually develop systemic later as non Hodgkins and large B-cell types
small cells better prognosis
systemic lymphoma in 2/3 of lid, 1/3 of orbital

Benign hereditary intraepithelial dysplasia
AD, North Carolina ancestry, corneal neovascularization, corneal plaques
bilateral gray horseshoe plaques in bulbar conjunctiva near Iambus
often vascularized, buccal leukoplakia, recur after excision

Mucoepidermoid
like SCC but more aggressive, >60

Oncocytoma
apocrine usually benign tumor of caruncle, elderly F

Misc
inclusion cysts
pyogenic granuloma
Dermoid
Epibulbar Osseus Choristoma
rhabdomysarcoma (embryonal)
Pigmented

**racial melanosis**
nl, bilateral, fades toward fornices, in 95% of blacks, 5% of whites

**congenital melanosis oculi**
unilateral uveal, scleral, episcleral pigment W>B
Nevus of Ota with lid involved, B>W
suspected increased uveal melanoma especially in Caucasians
no increased in conjunctival melanoma

**nevus**
bulbar conjunctival
can grow, often discrete light tan
moveable without extension onto cornea
bx if on palpebral or fornix conjunctiva
often cystic and can be very pigmented

**path**
rare to have junctional nevus of conjunctiva over 25 year old,
probably PAM
usually compound or subepithelial nevi

**iris**
no growth, <3mm width <1mm height minimal vessels, no
glaucoma or cataract

**PAM**
flat, golden brown to chocolate, mobile, indistinct margins
unilat, middle age, grows, does not fade toward fornix, no cysts
PAM without atypia suggest low malignant potential
with atypia has >50% chance of malignancy with subdivision in low and
high risk lesions
multiple small bx, excise smaller lesions, cryo more diffuse lesions,
especially with modularity

**melanoma**
bulbar, vessels, nodular, mobile, NO CYSTS
most from nevi, PAM, but de novo as well
prognosis worse if > 0.8mm, pagetoid PAM, melanoma in situ
form of
PAM, or if lid, caruncle, or fornix is involved
25% mortality
local excision with cryo, metastasis first to regional lymph
nodes

**iris**
no surgery or trab due to increased metastasis
key is to r/o ciliary body melanoma with transillumination,
scleral
depression, U/S
iridic cyts should be transilluminated, gonioscopy and U/S
**Kaposi's sarcoma**
elevated patches of hemorrhage that do not resolve
arise from lymphatics so none in orbit or choroid