Cornea and External Disease
Robert Cykiert, M.D.
Revised 2003

I. Basics

papilla

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

follicles

lymphatic response

acute	**chronic**
EKC, pharygoconjunctival fever	adult inclusion conjunctivitis
medicamentosa (epinephrine, neosynephrine)	toxic
Parinaud's oculoglandular,syndrome	r/o sarcoid
HSV primary conjunctivitis	r/o GPC, vernal conjunctivitis
Newcastle's conjunctivitis	

with acute follicles, always check lid margin for HSV vesicles, ulcers

membranes

conjunctivitis	ocular cicatricial pemphigoid	erythema multiforme
Stevens Johnson syndrome	Srogrens syndrome	atopy
Symblepharon	scieroderrna	burns
radiation burns	trachoma	EKC
sarcoid	drugs	

filaments

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

Enlarged Corneal nerves

MEN TIIb	icthyosis	Hanson's
Kconus	Refsums	Fuchs corneal dystrophy
old age	failed PKP	congenital glaucoma

trauma neurofibromatosis

MEN TIIb

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 microbulla
then subcellular with frank bulla

stomal

all extracellular

factors

imbibition pressure = IOP - swelling pressure (nl 50)
fluid into cornea from IOP, gyclosoaminoglycans'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

<u>mild</u>
muro 128, hair dryer, control IOP
<u>moderate</u>
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, violacious, blanches, increased with age
telangiectasia that bleed easily, rarely in retina and look like HTN or DM retinopatby
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
psuedotrachoma 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 corjunctivitis withwhite 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, maculopapultar 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	lues	TB

lymphgranuloma veneream

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 mucopurlent 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 scaning flerbervs pits (lirnbal 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)
doxycyline 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 neomcularization, 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- 1 0 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 If 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, sutuIres
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

erythema, 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
Doxycyline 100mg bid for a month
maybe even chronically bid/qd

Rosacea

F, 30-50, rhinophyma, telangiccwia, pustules, crythema
MGD, marginal keratitis, inferior corneal pannus with subepitlielial infiltrates

Subconjunctival hemorrhage

Anemia	ASHD	conjunc;tivitis	decreased platlets
DM	menses	nephritis	subacute bacterial endocarditis
Trauma	trichinosis	valsalva	vascular anomaly

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, Ehlors 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
Tl- 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, episcieritis
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 nearotrophic 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- 1 00 mg po x 3 days
pain medication
r/o HIV especially if < 45 year old or risk group
if microdissemination, pt may be im. immunecompromised
consider IV steroids, use IV acyclovir

HZO Neurological syndromes

postherpetic neuralgia

pain decreased with time (80% gone in I yr), worse if pt is older
Rx capsiacin, 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

atopy	Wegenees	periarteritis nodosa
dysentery	Crohn's/ UC	influenza
GC arthritis	dengue fever	hookworm
gold poisoning		
porphyria	brucellosis	acute leukemia
rheumatoid arthritis	SLE	scleroderma

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 cuffmg of nerves
risk factor contamin CL, corneal trauma, r/o topical anesthetic abuse
bx , stains faster and better than culture
touch material instead of smearing on slides-
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 CrystaUine 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 bacitmcin

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 mesenchyrnal 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 degencration

unilateral F>M middle age, non-inflamm end years later to old kerititis 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 resection, 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 deposts at edge of pannus, epithelium intact, decreased Va from astigmatism
rare perforation, rx with mild steroids chronically to suppress inflammation
RX lamellar or PKP

Marzinal 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 floureseein staining
debridement 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

TI	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

recurance post PKP common

Macular

AR, periphery involved
decreased 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 assymmetry
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 gutatta 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 scaning, V, VII CN, avitaminosis A, lagopthalmos, 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, tarsorraphy, 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

Exposure

Eyelid Burns

Immediate

often with swelling (<3rd degree)
when resolves, lubricate ql/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 epidermilization, 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 I5O 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 Phemphigoid

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 (oclironosis)

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
quinacrine 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, Tlll

Mucopoysaccharidosis (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 mmn 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 degeration

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 ftom HZO, HSV

Neurogenic

neurosyphillis, lesions of ciliary ganglion

V. Tumors

NonPigmented

Papillomas

viral

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

neoplastic

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

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

b**ulbar 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

conjunctiva

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