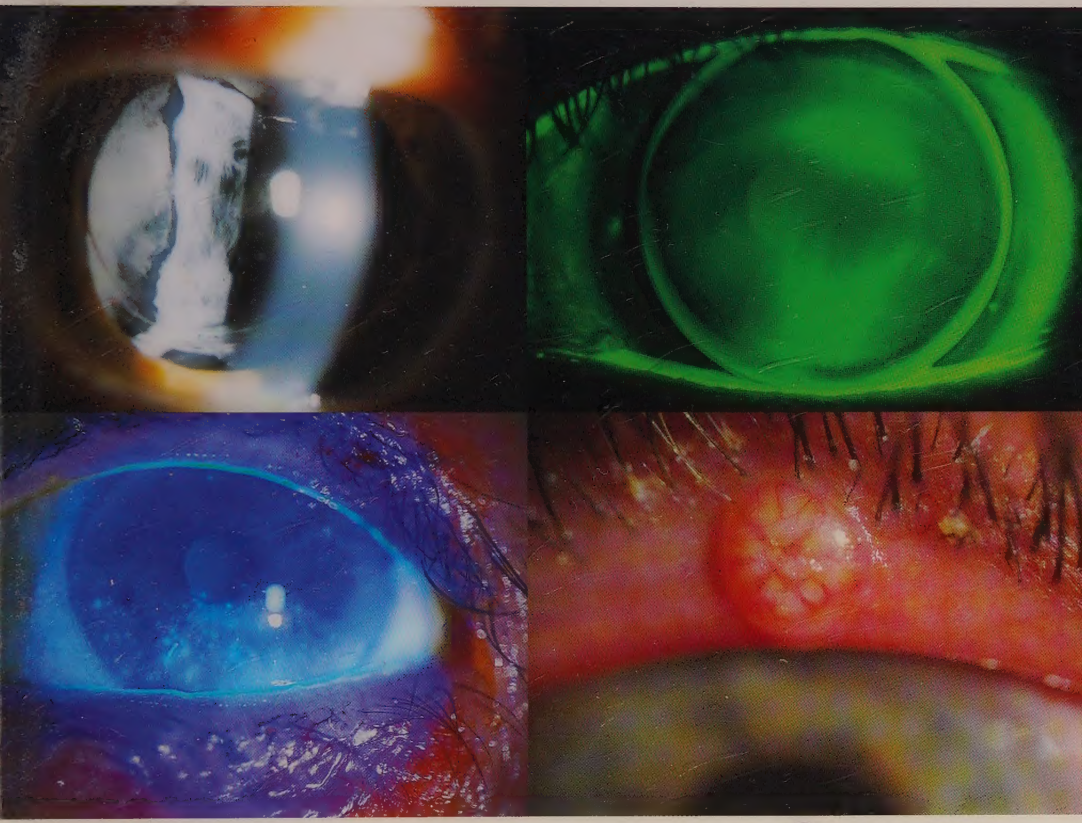
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2nd edition

Anterior Eye Disease and Therapeutics A–Z

Adrian Bruce and Michael Loughnan

CHURCHILL LIVINGSTONE
ELSEVIER

Key to Icons



Advice



Advice – genetic



Bandage and therapeutic contact lenses



Blood tests



Cease contact lens wear



Contact lens refitting



Consulting room procedures



Incisional surgery



Laser surgery



Microbiology



Notifiable condition



Ocular investigations



Oral medication



Pathology



Review



Tear supplements



Therapeutics



Topical treatment



Urgent

✓CAS 8324

Condition groupings

Condition groupings index [diagrams] ____ **CG-2**

Condition groupings index [tabular] ____ **CG-3**

Corneal degenerations, dystrophies, and depositions ____ **CG-4**

Corneal opacifying disorders, non-inflammatory and avascular

Eyelid – hot, tarsal ____ **CG-5**

Inflamed eyelid due to infection, allergy, or autoimmune conditions; conditions affecting the tarsal conjunctiva, particularly superiorly

Eyelid and lacrimal ____ **CG-6**

Degeneration of the eyelid, eyelashes, or lacrimal system, often causing watery eye

Iris and lens ____ **CG-7**

Lens disorders and cataract, iris dysgenesis, and tumors

Keratitis ____ **CG-8**

Corneal inflammation, with ulceration or infiltration

Keratopathy ____ **CG-9**

Non-inflammatory disease of the cornea, with deposits, opacities, or epithelial erosions, due to dessication, mechanical, toxic, or metabolic factors

Red eye – hot ____ **CG-10**

Inflamed red eye, due to infection, allergy, autoimmune, or intraocular pressure rise

Red eye – quiet ____ **CG-11**

Non-inflammatory changes to the bulbar conjunctiva with lumps, bumps, or pigment

Skin – bumps, lumps ____ **CG-12**

Non-inflammatory changes, due to degenerations or neoplasia

Surgical signs and complications ____ **CG-13**

Including operations on the cornea and crystalline lens and filtering procedures

Tear film anomaly ____ **CG-14**

Conditions affecting the aqueous, lipid, or mucous components of the tear film

Trauma ____ **CG-15**

Abrasions, burns, contusions, foreign bodies, sharp or blunt trauma

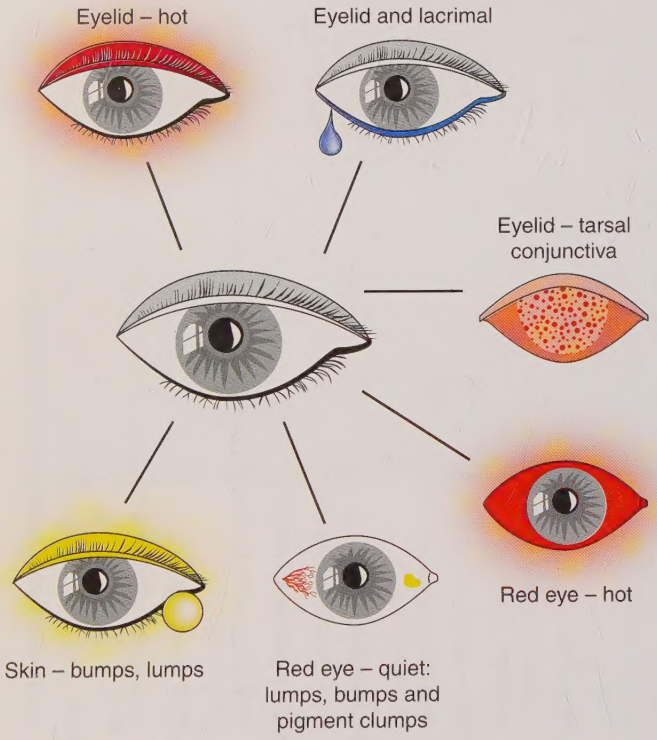
City and Islington College



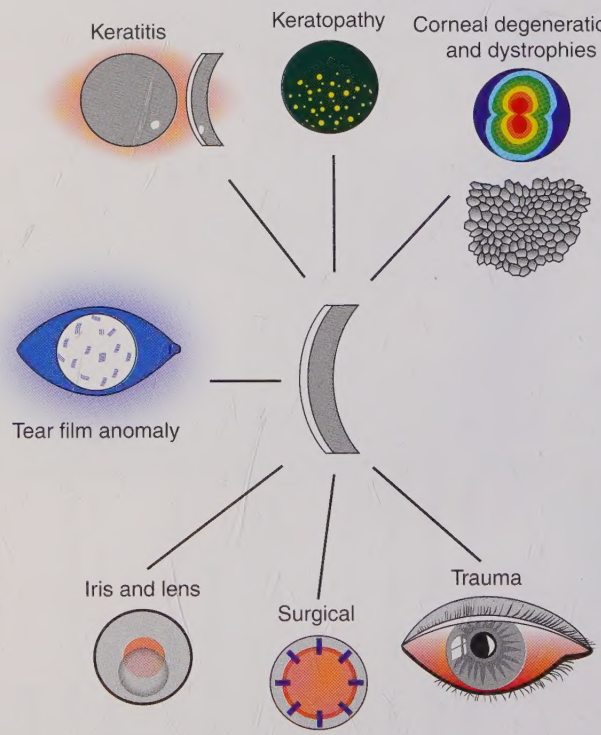
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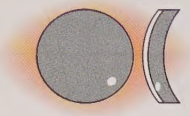
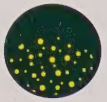
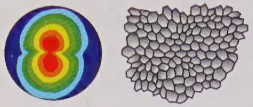
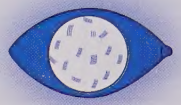
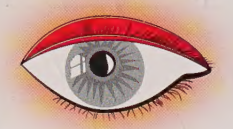
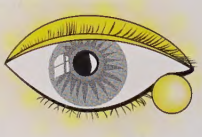
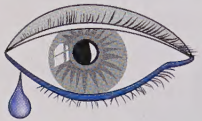
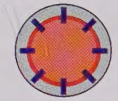
Condition groupings index [diagrams]

Condition grouping diagrams for conjunctiva and eyelid



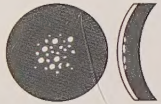
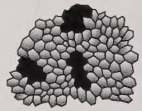
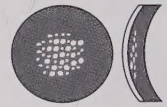
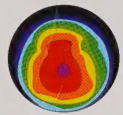
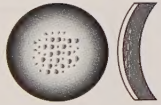
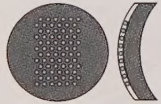
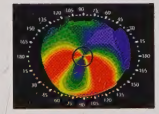
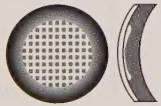
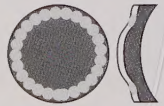
Condition grouping diagrams for cornea, iris, lens and tear film



	Hot eye Infected and or inflamed	Quiet eye Non-inflamed		Dry and watery eye	Surgical and trauma
Cornea	 Keratitis	 Keratopathy	 Corneal degenerations and dystrophies		
Conjunctiva	 Red eye	 Red eye – quiet: lumps, bumps		 Tear film anomaly	 Surgical
Eyelid	 Eyelid – hot	 Eyelid – tarsal conjunctiva	 Skin – bumps, lumps	 Eyelid and lacrimal	 Trauma
Iris and lens	 Red eye	 Iris and lens			 Surgical

Condition groupings index [tabular]

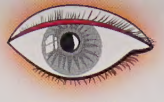
Corneal degenerations, dystrophies, and depositions

 Corneal dystrophies and degenerations overview	 Epithelial basement membrane dystrophy	 Fuchs' endothelial dystrophy	 Granular stromal dystrophy
 Keratoconus	 Keratoconus – forme fruste	 Keratoglobus	 Lattice stromal dystrophy
 Macular stromal dystrophy	 Meesmann's epithelial dystrophy	 Pellucid marginal degeneration	 Posterior polymorphous dystrophy
 Reis–Bückler's (anterior) dystrophy	 Salzmann nodular degeneration	 Terrien's marginal degeneration	**See also** • Keratitis • Keratopathy

Corneal degenerations and dystrophies: corneal opacifying disorders, non-inflammatory and avascular, mostly inherited, progressive, bilateral.



Acute allergic blepharoconjunctivitis



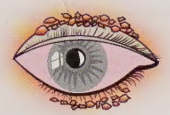
Blepharitis



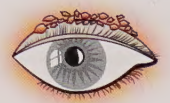
Canaliculitis



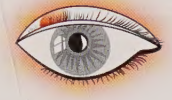
Dacryocystitis



Herpes simplex – primary keratoblepharoconjunctivitis



Herpes zoster ophthalmicus



Hordeolum – internal and external



Lice – phthiriasis palpebrarum



Molluscum contagiousum



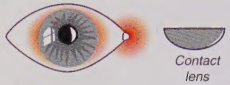
Preseptal cellulitis



Concretions



Conjunctival scarring overview



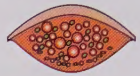
Contact lens complications overview



Floppy eyelid syndrome



Follicular conjunctivitis



Giant papillary conjunctivitis (papillae >1.0 mm)



Stevens–Johnson syndrome



Trachoma



Vernal keratoconjunctivitis

Eyelid – hot; see also

- Keratitis
- Red eye – hot
- Skin bumps, lumps

Eyelid – tarsal; see also

- Red eye – hot
- Eyelid – hot

Eyelid – hot: inflammatory ("hot") red eyelid due to infection, allergy, autoimmune.

Eyelid – tarsal: conditions affecting the tarsal conjunctiva, particularly superiorly.

Eyelid – hot, tarsal

Eyelid and lacrimal



Bell's palsy – facial paralysis



Dermatochalasis



Ectropion



Entropion



Epiblepharon and epicanthic folds



Epiphora overview



Floppy eyelid syndrome



Nasolacrimal duct obstruction – congenital



Ptosis



Punctal stenosis

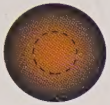


Trichiasis

See also

- Eyelid – hot
- Skin – bumps, lumps and pigment clumps

Degenerative changes to eyelid anatomy or lacrimal system. Often causes watery eye.



Aniridia



Cataract – overview



Ectopia lentis



Iridocorneal dysgenesis and Iridocorneal endotheliopathy (ICE)



Iris cysts and benign tumors



Leukocoria



Melanoma and nevus of the iris



Pseudoexfoliation of the lens capsule



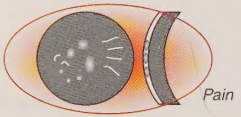
Synechia

See also

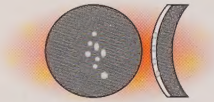
- Surgical
- Red eye – hot

Lens disorders and cataract, iris dysgenesis, and tumors.

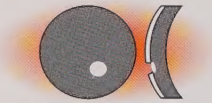
Keratitis



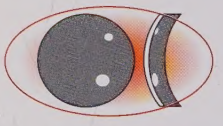
Acanthamoeba keratitis



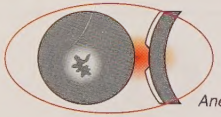
Adenoviral keratoconjunctivitis



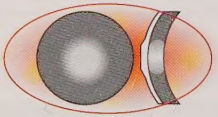
Bacterial keratitis



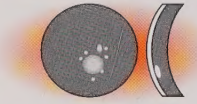
Corneal infiltrates – overview



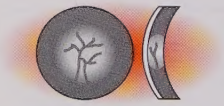
Dendritic ulcer (epithelial disease);
Herpes simplex keratitis (HSK)



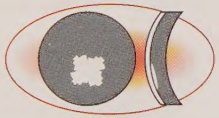
Disciform keratitis
Herpes simplex keratitis (HSK)



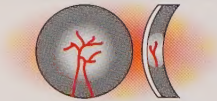
Fungal keratitis



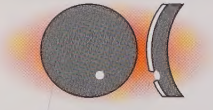
Herpes simplex keratitis – overview



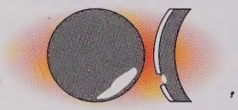
Infectious crystalline keratopathy



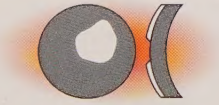
Interstitial keratitis – overview



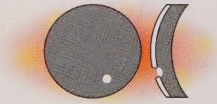
Marginal keratitis



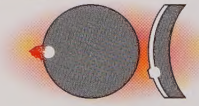
Mooren ulcer – autoimmune



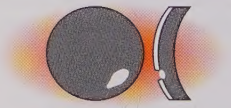
Neurotrophic keratitis, *and*
Persistent epithelial defect – overview



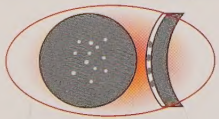
Peripheral ulcerative keratitis – overview



Phlyctenulosis – conjunctival, corneal



Rosacea keratitis



Thygeson's SPK

See also

- Red eye – hot
- Tear film anomaly
- Eyelid – hot
- Trauma (abrasion)
- Keratopathy (recurrent erosion)

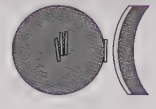
Keratitis – hot eye: corneal inflammation with corneal ulceration or infiltration with/without ocular pain, irritation, or red eye.



Band keratopathy



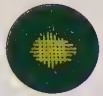
Climatic droplet keratopathy



Corneal edema – overview



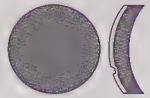
Corneal opacities – peripheral (Arcus, Vogt, Kayser–Fleischer)



Crocodile shagreen



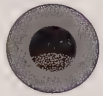
Crystalline keratopathy – overview



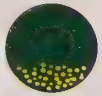
Dellen



Descemet's membrane breaks



Endothelial opacities – overview (Kruckenburg spindle, and others)



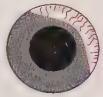
Exposure keratopathy



Filamentary keratopathy



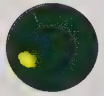
Iron lines



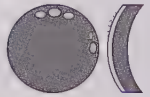
Limbal stem cell deficiency



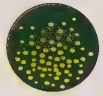
Lipid keratopathy



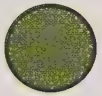
Recurrent corneal erosion syndrome



Salzmann nodular degeneration



Superficial punctate keratopathy



Toxic keratopathy



Vortex keratopathy

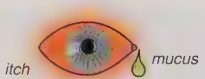
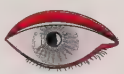
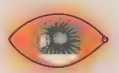
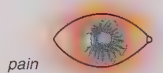
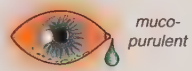
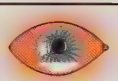
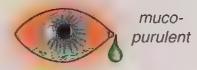
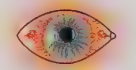
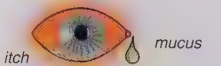
See also

- Corneal degenerations and dystrophies
- Keratitis
- Tear film anomaly
- Trauma
- Red eye – quiet (pterygium)

Keratopathy: non-inflammatory disease of the cornea. Deposits, opacities, epithelial erosions. Causes: desiccation, mechanical, toxic, metabolic.

Keratopathy

Red eye – hot

 Adenoviral keratoconjunctivitis	 Allergic eye disease overview	 Atopic keratoconjunctivitis (AKC)	 Bacterial conjunctivitis
 Chlamydial conjunctivitis	 Conjunctival scarring – overview	 Conjunctivitis – overview	 Contact lens complications – overview
 Endophthalmitis	 Episcleritis – simple and nodular	 Fuchs' heterochromic iridocyclitis	 Glaucoma – acute angle closure
 Gonococcal keratoconjunctivitis	 Hypopyon	 Iritis	 Membranous and pseudomembranous conjunctivitis
 Ocular cicatricial pemphigoid	 Ophthalmia neonatorum	 Scleritis – overview	 Seasonal allergic conjunctivitis (SAC)
 Stevens–Johnson syndrome	 Superior limbic keratoconjunctivitis	 Uveitis–anterior – overview	**See also** • Red eye – quiet • Eyelid – hot • Eyelid – tarsal conjunctiva • Keratitis • Tear film anomaly • Trauma

Red eye (bulbar conjunctiva) – hot: inflamed ("hot") red eye, due to infection, allergy, autoimmune, IOP rise; may be pain, discharge, keratitis, AC reaction.



Caruncular cysts



Conjunctival intraepithelial neoplasia (CIN)



Conjunctival lymphoma



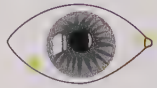
Conjunctival papilloma



Conjunctival pigmentary lesions (nevus, PAM, melanocytosis)



Conjunctivochalasis



Epibulbar choristoma – dermoids, dermolipomas



Melanoma – conjunctival



Pannus and pseudopterygia



Pingueculum



Pterygium



Retention cyst – conjunctival



Sub-conjunctival hemorrhage

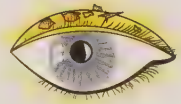
See also

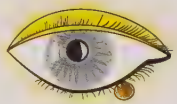
- Red eye – hot
- Eyelid – tarsal conjunctiva
- Surgical
- Tear film anomaly
- Trauma

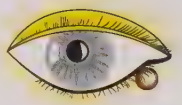
Red eye (bulbar conjunctiva) – quiet: lumps, bumps, and pigment clumps. Non-inflamed eye: may be few symptoms except for cosmetic concern or mild foreign body sensation.

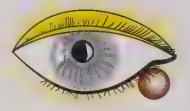
Red eye – quiet

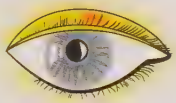
Skin – bumps, lumps

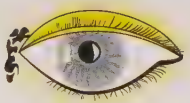

Actinic keratosis

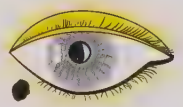

Basal cell carcinoma (BCC)

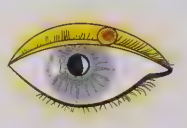

Chalazion

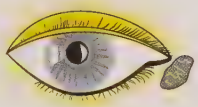

Cysts of the eyelid

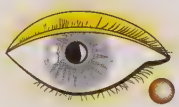

Hemangiomas

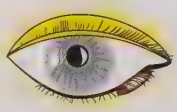

Melanoma – eyelid

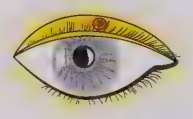

Nevus – eyelid

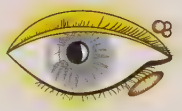

Sebaceous cell carcinoma


Seborrheic keratosis


Skin tumours – benign


Skin tumors – overview


Squamous cell carcinoma (SCC)


Xanthelasma

See also

- Eyelid – hot
- Eyelid and lacrimal
- Surgical
- Trauma

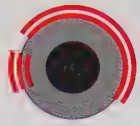
Skin – bumps, lumps: non-inflammatory eyelid skin changes due to neoplasia (benign, malignant, or metastatic) or degenerations.



Bullous keratopathy – pseudophakic and aphakic



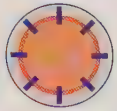
Cataract complications



Cataract incisions



Corneal collagen cross-linking



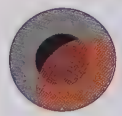
Corneal graft



Corneal graft: lamellar keratoplasty



Corneal graft rejection



Epithelial and fibrous ingrowth



Filtering bleb



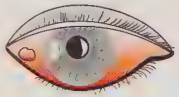
Intraocular lenses



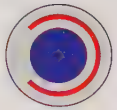
Perforation – corneal



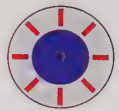
Posterior capsule opacification



Pyogenic granuloma



Refractive surgery – LASIK



Refractive surgery – RK and PRK

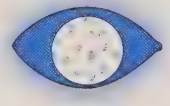
See also

- Trauma
- Red eye – quiet

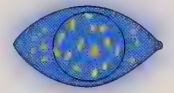
Surgical signs and complications: including operations on the cornea and crystalline lens and filtering procedures.

Surgical signs and complications

Tear film anomaly



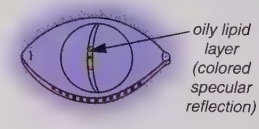
Dry eye syndromes – overview



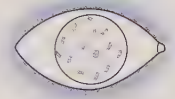
Dry eye syndromes – keratoconjunctivitis sicca (KCS)



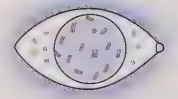
Exposure keratopathy



Meibomian gland dysfunction



Rheumatoid arthritis and the eye



Vitamin A deficiency (xerophthalmia)

See also

- Keratopathy e.g. toxic keratopathy
- Red eye (bulbar conjunctiva) if mild inflammation and irritation

Tear film anomaly: conditions affecting the aqueous, lipid, or mucous components of the tear film.



Abrasion



Chemical burn



Eyelid trauma
(hematoma, laceration, contusion)



Flash burn



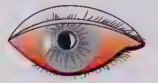
Foreign bodies – corneal
and conjunctival



Hyphema – blunt trauma



Penetrating eye injuries



Sub-conjunctival hemorrhage

See also

- Surgical signs and complications
- Red eye – quiet

Trauma: abrasions, burns, contusions, foreign bodies, sharp or blunt trauma.

Trauma

Anterior Eye Disease and Therapeutics A-Z

2nd edition





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Anterior Eye Disease and Therapeutics A-Z

2nd edition

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To our families:

Cathy, James, and Lachlan
Jacinta, Daniel, Jesse, and Myles

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Preface

In eye care, most therapeutic prescribing is for anterior eye conditions. This book is designed to be the "go to" guide for therapeutic eye care clinicians.

The A–Z format provides fast access and ease of use, essential attributes for a book used by the busy clinician. Furthermore, the coverage for each condition is presented in a double-page spread, giving the key information at a glance.

We have included both common and rare conditions, to make the book as comprehensive as possible. Eye care is somewhat random, in the sense that patients with the more complex or rarer conditions don't always seek out the specialist clinician. The old adage sometimes applies: "It ain't rare, if it's in your chair".

The scope of conditions considered is broadly inclusive of the anterior eye: autoimmune, infectious, and inflammatory conditions; degenerations and dystrophies; developmental anomalies; ocular surface disturbances; systemic manifestations; and tumors. Very rare conditions are not included unless they are well known.

In a conventional handbook, the Index is often one of the most used sections. In this book, the whole text is like an index, with ocular conditions listed in alphabetical order, giving direct access to the information. However, a conventional index has still been included, mainly because some conditions have more than one common name. For a good example, look at "Conjunctival intraepithelial neoplasia"!

There are consistent sub-headings for every condition, including: **Description, Symptoms, Signs, Incidence, Significance, Differential diagnosis, See also (related conditions)**, and **Management**. The contents of the sub-sections and a brief guide to the icons are given in the following pages. Each condition description is enhanced by a representative (rather than a "worst case") image.

Two additional graphical features are present on each page to aid in use of the book. On the right-hand margin is the **Section letter** containing the condition. If you flip through the pages with the thumb of your right hand, the letter will animate and run down the page, making it easier to find the required section. Across the top of the right-hand page for each condition, the **Management icons** highlight the recommended possible managements.

A new "flagship feature" for this book is the **condition grouping** diagrams, appearing as the "Condition groupings" index. The aim is to show "at a glance" as many as possible of a similar type of condition.

If a practitioner has a provisional diagnosis or only knows the condition characteristics, these condition groupings should assist with the short-listing of possible diagnoses or help in considering differential diagnoses. This book is designed to provide the ideal chair-side reference for such situations.

Overall, we have endeavored to meet the needs of both primary and specialist eye care clinicians. With hundreds of possible eye conditions and a similar number of available therapeutic drugs and other treatments, *Anterior Eye Disease and Therapeutics A-Z* aims to make it as easy as possible to arrive at a diagnosis and assist with best treatment.

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Conditions guide

Consistent subheadings have been used for each condition in this book. Below are an overview of the structure of each entry and a brief guide as to the contents of each subsection.

CONDITION NAME

Eponyms and synonyms for the condition

DESCRIPTION

Pathology, etiology, pathogenesis, key issues.

SYMPTOMS

Symptoms are listed and graded as follows:

- foreign body (FB) sensation, sandy, gritty
- cosmetic concern
- itch
- burning
- pain
- blurring of vision
- photophobia/glare sensitivity

(All graded as mild/moderate/severe)

- mass/lesion size, pain, and tenderness
- bilateral/unilateral.

SIGNS

Objective signs characterizing the condition.

INCIDENCE

While not intended to be exact, comments made in this section are intended to give the reader a general idea of the frequency with which the condition may be encountered in clinical practice:

- very common (greater than 1/10)
- common (approximately 1/100)
- uncommon (approximately 1/1000)
- rare (approximately 1/10,000)
- very rare (less than 1/100,000).

SIGNIFICANCE

The significance of the condition is listed with respect to the following issues and gradings:

- malignant versus benign
- cosmetic concern – mild, moderate, marked
- discomfort – mild, moderate, severe
- threat to sight – low, moderate, marked
- sight-threatening – requires prompt investigation and treatment.

DIFFERENTIAL DIAGNOSIS

Refers to related conditions in the book (and occasionally conditions outside the scope of this text) that form possible differential diagnoses. An attempt has been made to cross-reference conditions in an internally self-consistent way, using the condition groupings that are listed below.

The differential diagnoses are usually smaller subsets of the grouping listings found in the condition grouping diagrams in the "Condition groupings" section. The condition grouping diagrams are as follows:

Condition groupings index [diagrams]
Condition groupings index [tabular]
Corneal degenerations, dystrophies, and depositions
Corneal opacifying disorders, non-inflammatory and avascular
Eyelid – hot , tarsal
Inflamed eyelid due to infection, allergy, or autoimmune conditions; conditions affecting the tarsal conjunctiva, particularly superiorly
Eyelid and lacrimal
Degeneration of the eyelid, eyelashes, or lacrimal system, often causing watery eye
Iris and lens
Lens disorders and cataract, iris dysgenesis, and tumors
Keratitis
Corneal inflammation, with ulceration or infiltration
Keratopathy
Non-inflammatory disease of the cornea, with deposits, opacities, or epithelial erosions, due to dessication, mechanical, toxic, or metabolic factors
Red eye – hot
Inflamed red eye, due to infection, allergy, autoimmune, or intraocular pressure rise
Red eye – quiet
Non-inflammatory changes to the bulbar conjunctiva with lumps, bumps, or pigment clumps

Skin – bumps, lumps
Non-inflammatory changes, due to degenerations or neoplasia
Surgical signs and complications
Including operations on the cornea and crystalline lens and filtering procedures
Tear film anomaly (dry eye syndromes)
Conditions affecting the aqueous, lipid, or mucous components of the tear film
Trauma
Abrasions, burns, contusions, foreign bodies, sharp or blunt trauma

SEE ALSO
Other related conditions.

MANAGEMENT
See the explanation of the management categories and icons in the following section.

Management icons guide



Advice: Indicates treatment consists primarily of advice. In this instance, advice may serve to reassure the patient, advocate preventative measures, or to recommend self-monitoring.



Advice – genetic: Indicates an inherited condition, where advice may be provided to the patient with regard to the mode of inheritance and implications for their family's ocular health.



Bandage and therapeutic contact lenses: Indicates use of contact lenses for visual correction of corneal irregularity or for ocular surface protection. A bandage soft lens can promote epithelial stability and healing, reduce corneal stromal edema and reduce pain. For further details, see the following conditions: "Contact lens – gas permeable" and "Contact lens – soft".



Blood tests: Indicates the need to order blood tests, including serological tests, to assess disease activity and to aid in diagnostic and therapeutic decision making.



Cease contact lens wear: Indicates that, if the patient has contact lenses, lens use should be discontinued until the condition has fully resolved. Further lens wear may exacerbate the condition or interfere with treatment. Preservatives present in many topical therapeutic agents can bind to soft lenses and lead to ocular surface toxicity.



Contact lens refitting: Indicates treatment of the condition requires lens refitting to improve oxygen transmissibility, wetting, or the physical fitting of the lens. Modification of the usage and care of the contact lenses may also be required, including compliance with lens care instructions.



Consulting room procedures: Indicates the treatment of an ocular condition using in-office procedures such as foreign body removal, epilation, lacrimal lavage, insertion of punctal plugs, or lancing of a retention cyst.



Incisional surgery: A lesion not amenable to medical therapy may require surgical intervention. This may be in the form of a biopsy to aid diagnosis, curettage of an abscess, or excision of a lesion.



Laser surgery: Indicates a condition requiring removal of diseased tissue in the ocular media or for correction of a refractive error. Examples include phototherapeutic keratectomy (PTK), laser YAG posterior capsulotomy, or laser in situ keratomileusis (LASIK).



Microbiology: Indicates the need to identify the causative organism in an ocular infection. A key requirement is for the specimen to be taken prior to the commencement of antibiotic therapeutics. Laboratory investigations consist of staining and microscopy, culture, and establishment of antibiotic sensitivities.



Notifiable condition: In some countries, certain conditions may need to be reported to public health authorities. These include some sexually transmissible diseases, suspected child abuse, or trauma in an infant.



Ocular investigations: Indicates the need for further investigations to characterize the nature or extent of the condition. Such investigations include computerized videokeratoscopy (corneal topography), specular microscopy to assess the corneal endothelium, B-scan ultrasound, and pachymetry.



Oral medication: Systemic treatment is indicated when adequate drug delivery to the site of action is not achievable with

topical medication. Examples include oral antibiotics, corticosteroids, non-steroidal anti-inflammatory drugs (NSAIDs), and analgesics.



Pathology: Indicates the need to submit tissue for histopathological evaluation. Special stains may need to be ordered to identify specific cell types or pathogenic organisms.



Review: Indicates review is a necessary part of treatment to monitor either the development of the disease itself, response of the disease to treatment, or for possible side effects of drugs. In cases of indeterminate diagnosis, review by another practitioner is a useful alternative.



Tear supplements: Indicates the need for artificial ocular lubricant drops, gels, or ointments for patients with aqueous-deficient dry eye. However, lubricant drops have broader benefits, assisting in reducing antigenic load with ocular allergy or toxic conditions and improving patient comfort if there is significant ocular surface disruption by reducing shear forces associated with blinking. For further details see "Appendix A – Therapeutics", "Section 13 – Ocular lubricants".



Therapeutics: Indicates that application of topical medication is a common treatment option.

The anterior segment of the eye is ideally suited to the delivery of a variety of therapeutic agents by direct topical application. A high concentration of a drug can be readily achieved within the tear film with topical administration. Intraocular penetration is facilitated by the avascular and non-keratinized nature of the cornea. For further details, see "Appendix A – Therapeutics".



Topical treatment: Indicates a number of possible physical treatments of ocular disease. The treatment is conducted by the patient at home and may include lid hygiene, hot compresses, or cold compresses.



Urgent: The ambulance icon indicates that the condition requires urgent treatment. This is usually because the condition is sight- or life-threatening, and there is a risk of rapid progression.

Ocular emergencies covered in this book include bacterial keratitis, chemical (alkali) burns, corneal graft rejection, endophthalmitis, hypopyon, filtering blebs, glaucoma – acute angle closure, penetrating eye injuries, preseptal cellulitis, and uveitis.

A

Abrasion

Corneal abrasion, mechanical trauma

DESCRIPTION

A corneal abrasion is a superficial injury, affecting the epithelium but not involving the underlying stroma. Symptoms usually occur immediately, and the underlying cornea is relatively clear and free from inflammation. While the patient may be aware of the cause of the injury, such as a fingernail, in other cases it may be postoperative or idiopathic.

OTHER CAUSES OF EPITHELIAL DEFECTS

- **Chemical burns:** effects of acid, or particularly alkali, on the cornea can be potentially devastating and represent an ocular emergency.
- **Flash burns** arise due to electromagnetic radiation, either natural or artificial.
- **Foreign body:** organic material embedded in the cornea, e.g. from gardening, carries the risk of infection, and some metals cause persistent inflammation.
- **Contact lens:** corneal abrasion related to contact lens wear carries the risk of bacterial keratitis particularly with *Pseudomonas*, especially with poor patient compliance with lens disposal intervals, solution expiry dates, or contact lens case hygiene.
- **Penetrating eye injuries:** imaging and/or hospitalization may be indicated if the patient has been using power or garden tools that may produce a high speed projectile, or if there is trauma related to a sharp tool or weapon.
- **Recurrent corneal erosion syndrome,** or other idiopathic conditions.
- **Toxic keratitis** with corneal epithelial disruption and mild anterior eye inflammation, related to a toxic substance, medicamentosa, anesthetic

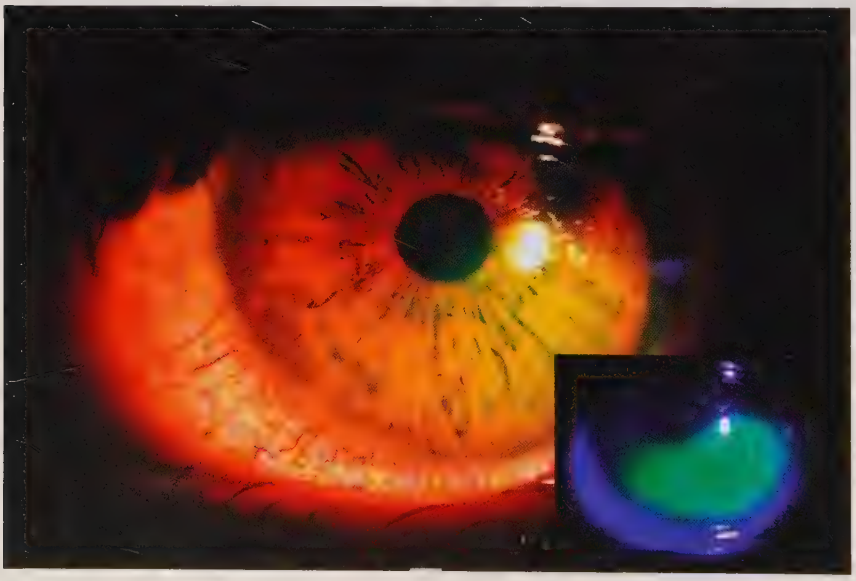


Fig. A-1 Shows staining of an epithelial defect with an underlying clear cornea.



abuse, or foreign matter. An uncommon toxic keratopathy associated with the effects of an airborne insect occurring in southeastern Australia is known as "Christmas eye."

SYMPTOMS

As the cornea is highly innervated, an abrasion usually causes acute pain. The patient will usually clearly recall the nature of the traumatic incident. If the abrasion is over the central cornea, vision may be impaired. The patient also frequently reports blurred vision, photophobia, a red eye, slight ptosis, and increased lacrimation.

SIGNS

An abrasion is a clearly delineated epithelial defect that stains with fluorescein. Abrasions are graded according to size (mm), location (central/peripheral) and effect on visual acuity. The underlying corneal stroma is usually relatively clear and free from inflammation (infiltration), although in larger abrasions there may be underlying corneal stromal edema. The anterior chamber is usually quiet.

DIFFERENTIAL DIAGNOSIS

- Superficial punctate keratopathy (SPK) with partial thickness epithelial lesions.
- Bacterial keratitis or marginal keratitis will show more inflammatory signs including an underlying corneal infiltrate.
- Herpetic keratitis or neurotrophic keratopathy usually causes minimal pain.

SEE ALSO

Trauma; see "Other causes of epithelial defects," above; Eyelid trauma; Chemical burns; Foreign bodies – corneal and conjunctival.

MANAGEMENT

A simple corneal abrasion is self-limiting and will heal in 1–5 days, depending on the extent of the area affected. Management is aimed to improve patient comfort and to ensure there is no secondary infection.

Therapeutics

- Antibiotics: a broad-spectrum topical antibiotic qid is indicated until epithelial healing has occurred. While the ointment form is probably more effective, drops are often adequate.
- Ice packs and oral analgesics may also be indicated for pain management, but patching is rarely used.
- Cycloplegia with e.g. homatropine 2% to prevent ciliary spasm may be indicated for larger abrasions, or if there are signs of an anterior chamber reaction.
- Bandage contact lenses are also sometimes used for larger abrasions to promote comfort and healing. They require prophylactic antibiotics, as above, and regular follow-up.

Advice

Risk factors for infection should be managed. Avoid contact lens wear, and review patient compliance with lens care. If the injury arose from gardening or pets there is a risk of subsequent fungal infection, and in a hot tub the risk of *Acanthamoeba*.

Review

The patient can be reviewed in 48 hours. Normally there is improved patient comfort and signs of healing evident within 24 hours.

A

Acanthamoeba keratitis

DESCRIPTION

Acanthamoebae are ubiquitous free-living protozoans that only rarely infect the cornea. They exist in an active form, a trophozoite, and in a highly resilient dormant cystic form. Infection usually, although not always, occurs in the presence of defined risk factors. Known risk factors include contact lens usage, especially in association with poor disinfection (typically soft contact lenses, although the disease has been reported with rigid lenses), and exposure to pool or spa water (especially in association with an epithelial abrasion). This disease is often missed in its early stages, and a high degree of suspicion should be maintained for any painful epitheliopathy that does not respond to normal treatment, especially in patients with known risk factors.

SYMPTOMS

Symptoms include blurred vision with associated pain that is frequently disproportionate to the degree of clinical signs. Photophobia is also typical of this disease and often initially worsens after commencement of treatment. The infection is bilateral in a significant percentage of patients.

SIGNS

Initially the disease is typified by an epitheliopathy with associated limbitis. At this stage it is commonly mistaken for herpes simplex keratitis. Over a period of weeks

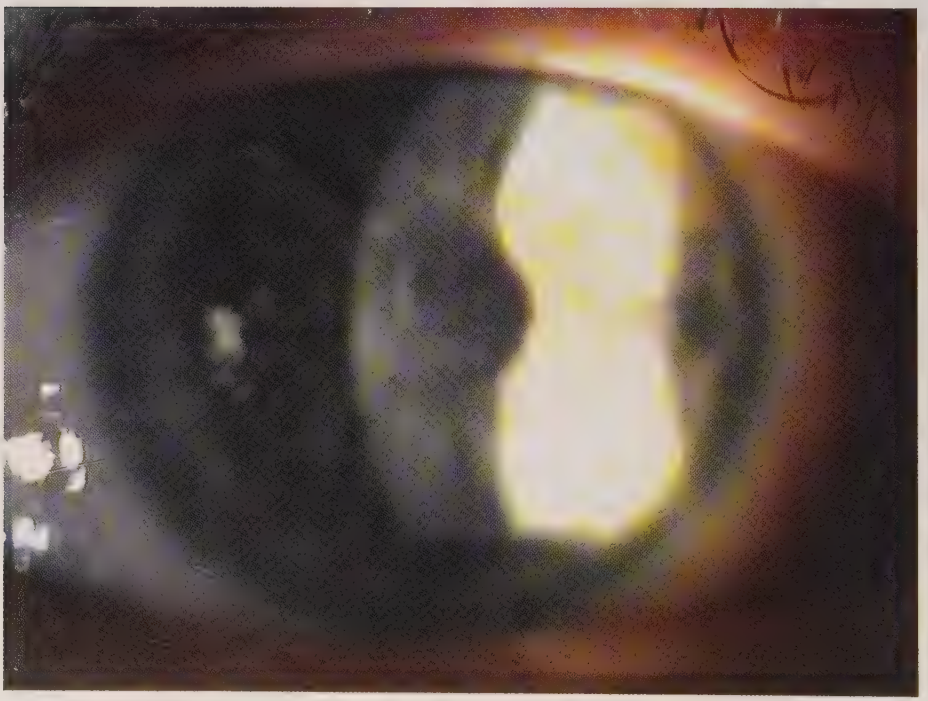
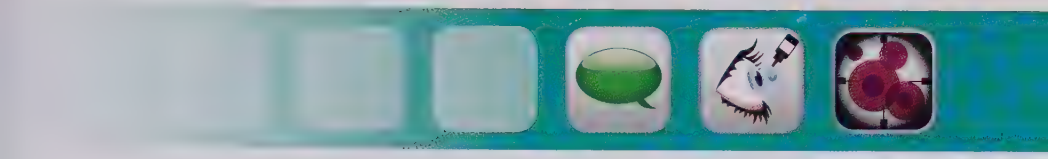


Fig. A-2 Typical "ground glass" epitheliopathy seen in early *Acanthamoeba* keratitis.



stromal signs develop with patchy anterior stromal infiltrates that may be associated with a pathognomonic radial perineuritis, Wessely (inflammatory) ring, and hypopyon. Untreated, it progresses to stromal thinning, epithelial defects, and scleritis.

INCIDENCE
Very rare (less than 1/100,000).

SIGNIFICANCE
Sight threatening, requires prompt investigation and treatment.

DIFFERENTIAL DIAGNOSIS
Herpes simplex keratitis; Contact lens-related conditions; Fungal keratitis.

SEE ALSO
Bacterial keratitis; viral keratitis; Fungal keratitis; Toxic keratopathy.

MANAGEMENT
Contact lens use should be ceased immediately in both eyes. They may be reintroduced when the infection is totally cleared, usually after at least 6 months.

Investigations
Microbiological specimens need to be taken including special stains for amebae and trophozoites and non-nutrient *Escherichia coli*-seeded agar plate for culture. The laboratory identification of the organism can be difficult and needs to be undertaken by a suitably experienced microbiologist. Both cysts and trophozoites may even be observed from staining impression cytology specimens.

Therapeutics
Commence treatment of the disease with specific anti-amebic drugs only after positive microbiological investigations. Treat initially with a combination of anti-amebic and trophozoicidal drugs: propamidine, neomycin, and 0.02% polyhexamethylene-biguanide (PHMB). Some clinicians use topical chlorhexidine instead of PHMB as an alternative cystocidal agent. Weekly review is required until a clinical response is noted. The keratitis may worsen initially with treatment, probably secondary to the liberation of protozoal antigens.

Dosing: Initially G. propamidine (0.15%), and G. neomycin qid and G. PHMB (0.02%) q1h for 1 week. Drop toxicity (toxic keratopathy) is common, and topical neomycin can usually be ceased after a week; propamidine is maintained at qid, and PHMB is slowly decreased to qid. This regimen is then maintained for several months.

Although contraindicated early in the treatment of the disease, topical corticosteroids are frequently used to suppress corneal inflammation once the infection is under good control. They are usually not started until at least several weeks after commencement of treatment with anti-amebics.

Prognosis
The treatment of this disease was greatly improved with the introduction of the first truly cystocidal drug PHMB in the early 1990s. This, coupled with an increasingly rapid diagnosis as awareness of this disease has improved, has led to much better visual outcomes with many patients regaining normal vision.

A

Actinic keratosis

Solar keratosis, keratoses, sun spots

DESCRIPTION
These are slow-growing hyper-keratinized plaques. They are keratinocyte dysplasias, but their potential for malignancy is low. Keratoses occur secondary to ultraviolet light damage and, accordingly, are typically seen in areas of sun-affected skin in elderly fair-skinned individuals. They can give rise to a squamous cell carcinoma.

SYMPTOMS
Patients with keratoses are usually asymptomatic. The affected skin is painless and non-tender. The skin is easily broken and bleeds readily with trauma.

SIGNS
The lesion is flat, or slightly elevated, skin-colored (non-pigmented) and with distinct borders. The surface is scaly and frequently has deep fissures. Occasionally, it is nodular or wart-like (papillomatous), and may even give rise to a cutaneous horn.

INCIDENCE
Uncommon (approximately 1/1000), more common in elderly fair-skinned people.

SIGNIFICANCE
The lesion is commonly of mild cosmetic concern and, although benign, may give rise to a squamous cell carcinoma (SCC). All patients with keratoses should be thoroughly examined to exclude the possibility of a coexisting invasive skin cancer.

DIFFERENTIAL DIAGNOSIS
Basal cell carcinoma (BCC – sclerosing type); Squamous cell carcinoma (SCC) – lid; Seborrheic keratosis.

SEE ALSO
Skin tumors.



Fig. A-3 Actinic keratosis involving the upper eyelid.



MANAGEMENT

Topical treatment

In the early stages, use of an emollient lotion or cream, as well as photoprotection including use of sunscreens, may slow or arrest development. If in any doubt as to the nature of the lesion or the risk of possible transformation, referral is indicated.

Surgery

If there is any concern with regard to malignant transformation into a squamous cell carcinoma, a biopsy should be performed. Chronic erosion at the edges of the lesion, recurrent infection or inflammation may indicate such a change. Cryotherapy is sometimes used, especially when multiple lesions are present.

A

Acute allergic blepharoconjunctivitis

Allergic blepharitis, contact dermatitis, drop allergy, acute allergic edema, contact allergic conjunctivitis (CAC)

DESCRIPTION

Drop allergy, either to the active component of the medication or to some other ingredient (including the preservative), can lead to an acute allergic reaction. As this is an antigen-specific response, prior exposure is required. An immediate (Type I) allergic response occurs with an initial reaction occurring within minutes to hours following exposure, with antigen–antibody (IgE) reaction and histamine release by mast cells. The late-phase response occurs in 6 to 12 hours and is mediated by leukotrienes, prostaglandins, and platelet-activating factor, producing the inflammatory cascade lasting hours to days.

SYMPTOMS

Lid swelling and erythema usually occur within hours of instillation of the allergen. There is a watery discharge, the eye and lid skin may be itchy, and the vision is often blurred. Response may vary from mild to dramatic.

SIGNS

Initial eyelid erythema and edema, tearing, and chemosis. Lid swelling may be unilateral or bilateral depending on exposure to the allergen. The swelling is gravity dependent and as such is often worse on awakening. There is typically an associated allergic conjunctivitis. The swelling is initially quite tense but evolves to folds of redundant skin over several days.

INCIDENCE

Uncommon (approximately 1/1000).

SIGNIFICANCE

Can be distressing to the patient but is not sight-threatening and is self-limiting.

DIFFERENTIAL DIAGNOSIS

Preseptal or orbital cellulitis.

SEE ALSO

Seasonal allergic conjunctivitis (SAC); Allergic eye disease – overview; Toxic keratopathy; Conjunctivochalasis and chemosis.



Fig. A-4 Marked periocular edema secondary to an acute drop allergy.



MANAGEMENT

Withdraw the suspected allergen. If the allergy is to the preservative, unpreserved drops may need to be used in the future.

Topical treatment

The acute episode is usually treated with cold packs applied frequently over the swollen lids.

Therapeutics

If required, a mild topical steroid cream such as hydrocortisone 1% qid may be prescribed for 3 to 7 days.

Oral medication

Oral antihistamines are also sometimes used, especially if there is associated itch and discomfort. It takes several days to a week for the swelling to subside totally.

A

Adenoviral keratoconjunctivitis

Epidemic keratoconjunctivitis (EKC), pharyngoconjunctival fever (PCF), nummular keratitis

DESCRIPTION

This is a disease of varying severity ranging from mild to severe. The incubation period is 4–10 days and, after the onset of conjunctivitis, the virus is shed for up to 12 days. The virus is highly contagious, being spread via ocular or respiratory secretions. The mode of dissemination is by direct contact with infected surfaces, especially towels, tissues, and tonometry prisms. Particular care needs to be taken within an eye clinic to minimize the spread of the virus to both staff and other patients. Thankfully the virus is fairly easily killed with topical alcohol wipes.

Visual disturbance is usually secondary to multiple central subepithelial infiltrates. These typically fade over a few weeks to months; however, occasionally they persist for years following a severe infection.

SYMPTOMS

Typically the patient presents with a watery discharge and a mild-to-moderate burning irritation. Initially worse in one eye, it often becomes bilateral due to auto-inoculation of the other eye. The discharge increases over 1–2 days and, depending on the severity of the infection, the patient develops lid edema, red eye, photophobia, and blur. With pharyngoconjunctival fever (PCF) there are also symptoms of an upper respiratory tract infection (URTI). PCF is characterized by the triad of "follicles, fever, and pharyngitis."

SIGNS

The key signs distinguishing adenoviral keratoconjunctivitis from simple



Fig. A-5 Nummular scars typical of adenoviral keratitis.



bacterial conjunctivitis are a watery discharge, follicular conjunctivitis, palpable preauricular or submandibular lymph nodes, and scattered nummular subepithelial corneal opacities. These have a typical "areolar" appearance with a central dense area surrounded by a less dense periphery. The bulbar hyperemia is purplish/pinkish, with injection usually beginning at the inner canthus then spreading laterally. Keratitis occurs in approximately 80% of cases of epidemic keratoconjunctivitis (EKC) but only approximately 30% of cases of PCF. The opacities are initially inflammatory, presumably a reaction to viral antigen; however, with time they may scar. Once present, a scar normally takes weeks to months to fade but may persist for years.

INCIDENCE

Common (approximately 1/100), usually occurring in epidemics.

SIGNIFICANCE

Usually a self-limiting disease but can result in some long-term increased glare and blurring of vision from persistent corneal opacities.

DIFFERENTIAL DIAGNOSIS

Molluscum contagiosum; Chlamydial conjunctivitis; Granular dystrophy; Herpes simplex keratitis (HSK).

SEE ALSO

Bacterial conjunctivitis; Follicular conjunctivitis.

MANAGEMENT

Topical treatment

HYGIENE

It is important that the patient realizes the highly contagious nature of the infection and is very careful with personal hygiene, especially with regard to food preparation during the infective period. The use of separate towels at home and tissues instead of handkerchiefs should be encouraged.

SUPPORTIVE OR PALLIATIVE MEASURES

Treatment is largely symptomatic and supportive, and spontaneous resolution occurs within 2 weeks. Tear supplements to improve ocular comfort are often useful. Other possible measures are cold packs (every 3–4 hours) and irrigation with saline or cold water (at a similar interval).

POVIDONE IODINE 5%

Betadine (povidone iodine 5%) is being investigated as an antiseptic irrigation treatment for EKC.

Microbiology

If the diagnosis is unsure, polymerase chain reaction (PCR) of a swab from the inferior fornix may be taken to identify viral DNA. Office-based rapid antigen tests are also now available. Alternatively, viral culture can be performed.

Therapeutics

Antiviral agents are ineffective against adenovirus. The use of topical steroids is controversial; they are only used when there is an associated keratitis. They are of most benefit in the early stages of the disease, when the opacities are inflammatory rather than scars; however they can be tried to help fade opacities even several weeks to months after the initial infection. Aspirin or similar may be useful if the ocular discomfort is significant.

Dosing: If indicated, G. fluorometholone acetate (0.1%) qid for 1 week, or longer if corneal infiltrates persist.

Surgery

Excimer laser to remove anterior scars is almost never necessary; even in the most severe cases they tend to fade with time.

Review

Recheck in 7–10 days, or earlier if there is a severe conjunctivitis or keratitis.

A

Allergic eye disease – overview

Seasonal/perennial allergic conjunctivitis (SAC, PAC), giant papillary conjunctivitis (GPC), vernal keratoconjunctivitis (VKC), atopic keratoconjunctivitis (AKC), contact allergic conjunctivitis (CAC), acute allergic blepharoconjunctivitis

DESCRIPTION

Allergic eye disease ranges from the very common and annoying (although non-sight-threatening) conditions, seasonal and perennial allergic conjunctivitis (SAC, PAC), through to rarer, bilaterally sight-threatening conditions, such as vernal keratoconjunctivitis (VKC) and atopic keratoconjunctivitis (AKC). It is often associated with other allergic features such as eczema, asthma, sinusitis, and allergic rhinitis (hay fever). The onset of allergic disease usually occurs in childhood although a significant other group experience onset post-puberty. The hallmark of all these conditions is mast cell degranulation with the release of vasoactive substances, especially histamine causing itch and redness.

Although the different conditions covered by allergic eye disease have a similar etiology, they are sufficiently different so that they can usually be clinically separated. Occasionally, one form will evolve over time into another form of allergic eye disease, typically childhood VKC into adult AKC.

Seasonal and perennial allergic conjunctivitis (SAC and PAC)

SAC and PAC lead to ocular surface itch and irritation and are typified by a velvety, edematous conjunctivitis without scarring.

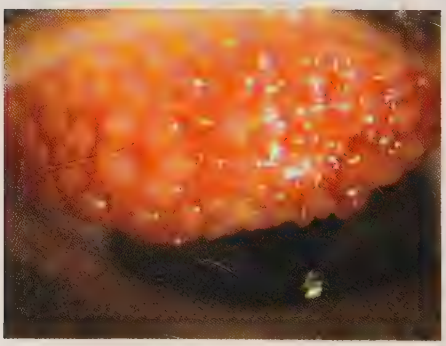


Fig. A-6 Inactive giant papillae with surface scarring in a young patient with vernal keratoconjunctivitis.

Contact allergic conjunctivitis (CAC)

Also called acute allergic blepharo-conjunctivitis, the most common form of contact allergic eye disease is drop allergy; the allergy is usually to either the active ingredient or some other part of the formulation, most commonly the preservative. It can be difficult to distinguish drop allergy from drop toxicity (toxic keratopathy); however, drop allergy usually occurs soon after repeat challenge with a single drop of the agent, whereas toxicity typically builds up over time.

Vernal keratoconjunctivitis (VKC)

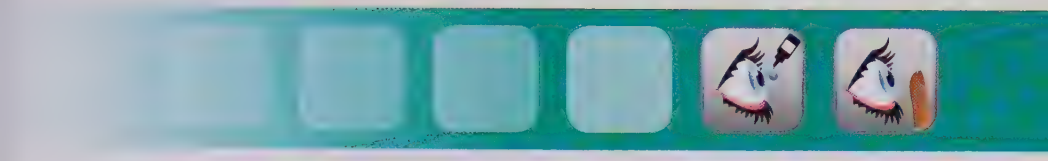
VKC is seen exclusively in children and young adults, most commonly in boys. It is frequently associated with a significant other atopic disease, such as eczema or asthma, and frequently exhibits seasonal exacerbations, especially in spring. There are two main forms: the more common palpebral vernal, typified by giant papillae on the superior tarsal conjunctiva, and the less common limbal vernal, typified by a focal inflammatory reaction at the limbus. When severe, VKC has the potential to lead to significant corneal scarring with resultant long-term visual loss through the development of chronic, non-healing corneal epithelial defects termed shield ulcers.

Atopic keratoconjunctivitis (AKC)

AKC is an ocular surface cicatrizing condition almost exclusively seen in adults with marked atopic disease, usually including facial, peri-ocular eczema. The typical features include marked tarsal subconjunctival scarring and corneal conjunctivalization and scarring.

Giant papillary conjunctivitis (GPC)

GPC affects the superior tarsal conjunctiva, and is most commonly seen with soft contact lens usage, although it can occur less frequently with rigid contact lenses. There appear to be mechanical as well as allergic components to the condition. It may lead to temporary or permanent contact lens intolerance.



SYMPTOMS

The hallmark symptom of ocular allergy is itch. Other symptoms include redness, peri-ocular swelling, irritation, a slight foreign body sensation, and, if marked as in VKC and AKC, photophobia. These conditions are usually bilateral, although frequently the symptoms and signs are asymmetric. Especially if the allergen is a pollen or other plant matter, the disease exhibits periods of exacerbation and remission which may be seasonal in nature.

SIGNS

To varying degrees, all forms of ocular allergy demonstrate signs of chemosis, vascular injection, superficial punctate keratopathy (SPK), lid swelling (even ptosis with severe disease), and conjunctival papillae, usually most marked on the superior tarsal conjunctiva. Pannus formation, corneal vascularization, corneal epithelial ulceration, and corneal scarring may be seen with more severe forms of allergy such as VKC and AKC.

INCIDENCE

SAC and PAC are both very common, affecting up to 35% of the population. VKC and AKC are both very rare. GPC is a rare condition and appears to be becoming less common with modern lenses, probably reflecting improved lens materials, finishes, and coatings. CAC is less common with modern drops and preservatives, and occurs only rarely.

SEE ALSO

Seasonal allergic conjunctivitis; Acute allergic blepharoconjunctivitis; atopic vernal conjunctivitis; Vernal keratoconjunctivitis; Giant papillary conjunctivitis; Toxic keratopathy.

MANAGEMENT

The degree of therapeutic intervention is highly dependent on the severity of symptoms, and the risk to the ocular surface.

Advice/allergen avoidance

Cleaning around the eyes and the flushing of the inferior conjunctival fornix with lubricant drops can be useful in clearing allergens from the ocular surface during an acute attack.

With more marked disease, such as VKC and AKC, the early involvement of an allergist is important to ensure optimal control of the multiple allergic diseases often present in these patients. Drops may need to be altered or ceased with CAC, and contact lenses removed during the early treatment phase for GPC.

Therapeutics

Many different classes of topical medication are used to treat allergic eye disease ranging from over-the-counter (OTC) medications, such as ocular lubricants, antihistamines, decongestants (vasoconstrictors), and mast-cell stabilizers, to the prescription medications, non-steroidal anti-inflammatory agents, corticosteroid agents, and cyclosporin A. The level of therapeutic intervention is matched to the severity of symptoms and risk to the ocular surface and is discussed under the entry for each specific condition. Occasionally, systemic treatment with short-term oral prednisolone is required for acute flare-ups of VKC and AKC.

A

Aniridia

DESCRIPTION

Aniridia is a bilateral, inherited, congenital dysgenesis that occurs secondary to a mutation or deletion in the *PAX6* gene on the short arm of chromosome 11. This is a homeobox gene and, as such, controls the development of a number of structures. This explains the broad nature of ocular defects seen with the disease. Most people with autosomal dominant inheritance have a mutation within the gene and are unlikely to have extraocular manifestations of the disease. People with sporadic or recessive aniridia with no family history of the disease frequently have a deletion within chromosome 11 and commonly have extraocular disease. The presence or absence of extraocular disease appears to depend on the extent of the deletion on chromosome 11. The combination of a Wilms' tumor, aniridia, genitourinary anomalies, and mental retardation is referred to as WAGR syndrome, or occasionally as Miller syndrome, and is due to a deletion on chromosome 11. The combination of aniridia, cerebellar ataxia, and mental retardation is termed Gillespie syndrome. Approximately 85% of all patients with aniridia have no extraocular manifestations.

SYMPTOMS

Ocular symptoms include poor vision, glare, and chronic surface irritation.

SIGNS

Iris dysgenesis is quite variable and ranges from minimal changes with retroillumination defects through to the total lack of iris tissue. Frequently, there is a residual frill of iris tissue in the angle. This is thought to play a role in the development of raised intraocular pressure. Glaucoma occurs in approximately 75% of patients,

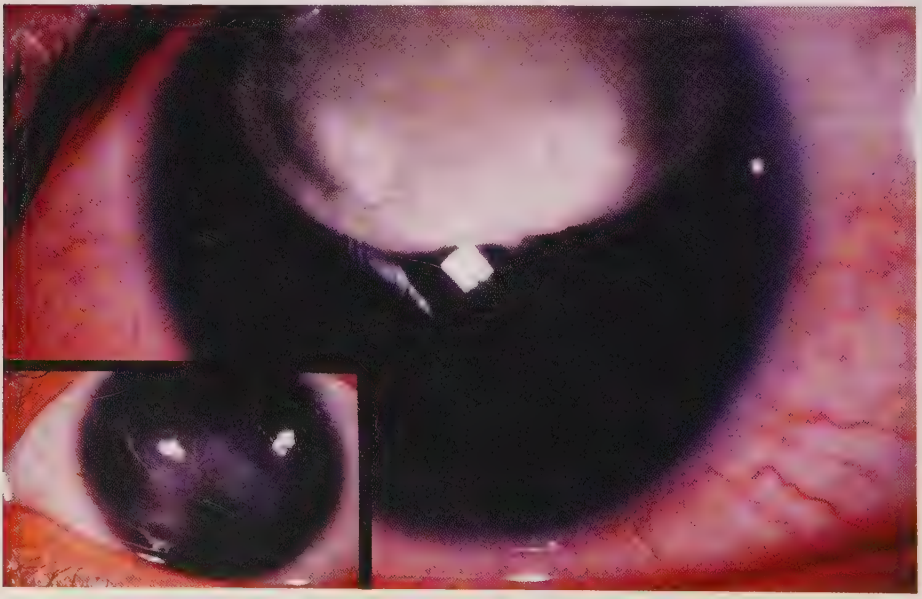


Fig. A-7a Aniridia demonstrating lack of iris, ectopia lentis, cataract, and keratopathy.

Fig. A-7b Inset: Corneal epitheliopathy secondary to primary limbal stem cell dysfunction.



and typically develops in late childhood or early adolescence.

Other commonly seen ocular findings include a cataract, ectopia lentis (usually with superior displacement), nystagmus, ptosis, hypoplastic fovea, atypical colobomata, and disc hypoplasia. Over time patients frequently develop limbal stem cell failure with progressive conjunctivalization (pannus formation) of the cornea.

INCIDENCE

Very rare (less than 1/100,000).

SIGNIFICANCE

Vision is usually poor, being 6/60 or worse, and usually deteriorates with time due to glaucoma, cataract, and epitheliopathy.

DIFFERENTIAL DIAGNOSIS

Iridocorneal dysgenesis; Trauma; Iridocorneal endotheliopathy (ICE).

SEE ALSO

Limbal stem cell deficiency; Ectopia lentis.

MANAGEMENT

Therapeutics

Topical or oral anti-glaucoma medication is frequently required. Patients should have yearly reviews to monitor for the development of glaucoma.

Tear supplements

Tear supplements are frequently used for the associated keratopathy.

Therapeutic contact lens

An opaque tinted soft lens can create an artificial pupil, improve vision, and improve cosmesis.

Surgery

Medical management of the associated glaucoma is often inadequate and filtering surgery, including the use of an artificial filtering shunt, may be necessary. Cataract surgery is also often required, with the preferred approach being a lensectomy/vitrectomy. In planning any surgery, care needs to be taken to minimize trauma to the limbus so as to preserve any residual limbal stem cell function.

Genetic counseling

Aniridia can be inherited as either a recessive or a dominant trait with approximately one-third of all cases being sporadic. The dominant trait has a high degree of penetrance, and therefore approximately 50% of family members are affected.

Prognosis

Visual prognosis is guarded, with visual function frequently deteriorating over time.

A

Atopic keratoconjunctivitis (AKC)

DESCRIPTION

Atopic keratoconjunctivitis (AKC) is a serious sight-threatening bilateral allergic disease that eventually leads to extensive conjunctival scarring. Patients typically have a strong atopic history with eczema, asthma, and hay fever. Onset is usually not until adulthood, although it may evolve from childhood vernal keratoconjunctivitis (VKC).

SYMPTOMS

As with all allergic eye disease, itch is the key symptom. There may be associated blurring of vision and a mucoid discharge with mucous strands.

SIGNS

Ocular

The lids are thickened, macerated, and fissured, and there is frequently associated blepharitis. The conjunctiva is variably affected with anything from a hyperemic, flattened velvety appearance with papillae through to extensive subepithelial scarring with forniceal shortening. Resolved papillae superiorly may lead to typical limbal cystic lesions. There is frequently a significant mucus discharge.

The primary cause of visual loss is corneal scarring. This results from a combination of conjunctival disease and the liberation of pro-inflammatory cellular products that have a directly toxic effect. In less severe cases there is only a superficial punctate epitheliopathy, whereas in severe cases this extends to include persistent epithelial defects, stromal scars, and extensive pannus formation. There may be superior limbal infiltrates and Trantas dots.

In severe cases, an anterior subcapsular cataract may develop.

There may be associated signs of keratoconus that possibly develops secondary to the extensive eye rubbing that characterizes this disease.



Fig. A-8 Fine superficial scarring with loss of the normal tarsal conjunctival architecture resulting from chronic atopic keratoconjunctivitis.



Skin
Atopic dermatitis typically affects the flexures and should be looked for at the wrists, antecubital fossa, neck, and on the eyelids themselves.

INCIDENCE
Rare (approximately 1/10,000).

SIGNIFICANCE
Sight-threatening from corneal surface scarring, and an increased risk of both bacterial and bilateral herpetic keratitis as well as an increased risk of rhegmatogenous retinal detachment.

DIFFERENTIAL DIAGNOSIS
Vernal keratoconjunctivitis (VKC); Trachoma.

SEE ALSO
Keratoconus; Allergic eye disease – overview; Seasonal allergic conjunctivitis (SAC); Cataract.

MANAGEMENT
Allergen avoidance
Cleaning around the eye and the flushing of the inferior conjunctival fornix with lubricant drops can be useful in clearing allergens from the ocular surface during an acute attack.

The early involvement of an allergist, usually via a GP referral, is important to ensure optimal control of the multiple allergic diseases often present in these patients. They can appropriately investigate for any specific allergens and put in place a system of allergen avoidance, such as the removal of carpets from bedrooms and the use of hypoallergenic bedding. The possibility of allergen desensitization can also be explored.

Therapeutics
Many different drugs are used topically to control ocular surface inflammation.

ANTIHISTAMINES (LEVOCABASTINE)
Unfortunately these drugs are of limited usefulness.

COMBINED ANTIHISTAMINE/MAST-CELL STABILIZERS (OLOPATADINE AND KETOTIFEN)
Quite effective, and do not have the side effects seen with steroids. Should be used throughout the year.

NON-STEROIDAL ANTI-INFLAMMATORY AGENTS (KETOROLAC)
Quite useful and often used in addition to mast-cell stabilizers.

CORTICOSTEROIDS
Extremely effective in managing acute exacerbations. Long-term usage of corticosteroids is often associated with side effects and, therefore, they are restricted to the sparing use of drugs of lower efficacy with poor ocular surface penetration (e.g. fluorometholone).

CYCLOSPORINE A
It has been demonstrated to be quite effective, with minimal side effects.

Advice
Advise patients of the importance of avoiding aggressive rubbing of the eye. This can contribute to the development of both ptosis and keratoconus.

B

Bacterial conjunctivitis

Simple bacterial conjunctivitis

DESCRIPTION

Simple bacterial conjunctivitis is a common, usually self-limiting condition most frequently occurring in childhood. There is a wide range of causative organisms, both Gram-positive and Gram-negative. The most common are *Staphylococcus aureus, S. epidermidis, Streptococcus pneumoniae*, and *Haemophilus influenzae*. Recurrent infections may be associated with poor hygiene as well as abnormalities of the lacrimal drainage system, such as nasolacrimal duct blockage or a retained punctal or canalicular plug. Although not as highly contagious as adenoviral conjunctivitis, care should still be taken not to spread the disease. This usually occurs by direct contact with infected mucus and can be avoided by strict personal hygiene.

SYMPTOMS

Typically, patients present with red, gritty eyes with associated mucopurulent discharge. Both eyes are usually involved, although one may be affected before the other. The discharge increases over 1 to 2 days and often becomes a significant problem. Patients often report matting of lashes or that their eyes are stuck together on waking.

SIGNS

The conjunctiva is reddened and inflamed with velvety edematous papillary changes best seen on the superior and inferior tarsi. The mucopurulent discharge leads to crusting along the lids and matting together of the lashes, and there is frequently a long mucous strand in the inferior fornix. The cornea frequently has features of a toxic epitheliopathy with scattered punctate erosions on fluorescein staining.

INCIDENCE

Common (approximately 1/10).

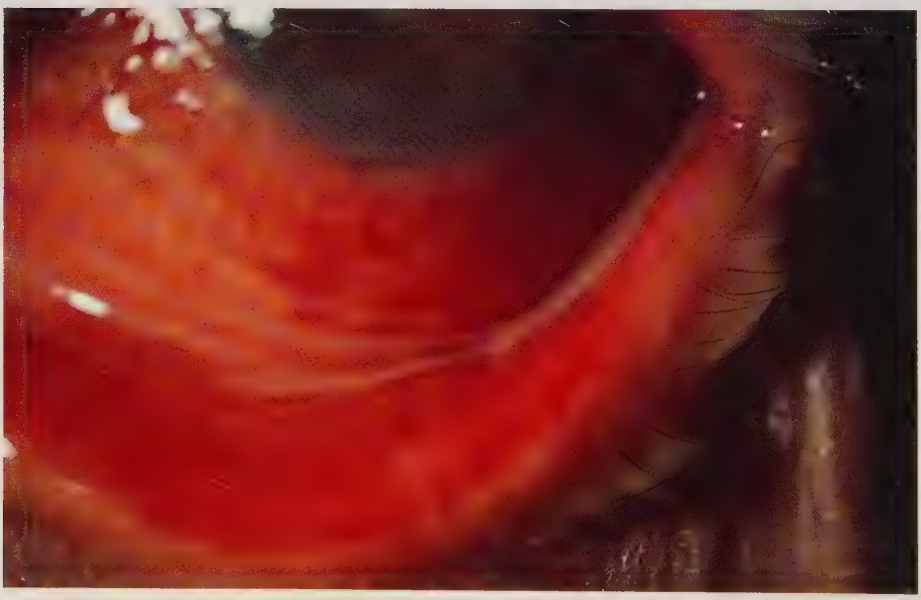


Fig. B-1 Mucopurulent exudate in the inferior fornix with associated conjunctival chemosis and injection in bacterial conjunctivitis.



SIGNIFICANCE

Usually not sight-threatening unless complicated with bacterial keratitis or, unusually, membranous conjunctivitis with symblepharon formation.

DIFFERENTIAL DIAGNOSIS

Viral conjunctivitis.

SEE ALSO

Gonococcal keratoconjunctivitis; Adenoviral keratoconjunctivitis.

MANAGEMENT

Although usually a self-limiting disease resolving in 10–14 days, it is usually treated with topical antibiotics to shorten the course of the disease.

Hygiene

Active cleaning of the lids, lashes, and eye to remove the discharge is important to decrease symptoms, improve penetration of topical drugs, and decrease infectivity. It also helps to protect the ocular surface by removing toxic lytic enzymes present within the discharge.

Microbiology

Microbiological investigation is rarely required; however, bacterial cultures can be performed from a conjunctival swab.

Therapeutics

Dosing: topical antibiotics with a broad spectrum of activity against known Gram-positive and Gram-negative pathogens are usually used 4 times a day for 1 week. Commonly used antibiotics include chloramphenicol, gentamycin, tobramycin, moxifloxacin, and ciprofloxacin. In methicillin-resistant *Staphylococcus aureus* (MRSA) ocular surface infections, consider the use of antimicrobials such as trimethoprim, tobramycin, or chloramphenicol.

B

Bacterial keratitis

Corneal abscess, microbial keratitis, infected ulcer

DESCRIPTION

Bacterial infections of the cornea are acutely sight-threatening and, if left untreated, can rapidly progress to marked stromal loss and even perforation. Bacterial keratitis almost always occurs in the presence of a significant risk factor such as contact lens use, especially with poor lens care or extended wear. Even the higher oxygen transmission lenses worn on an extended wear basis represent an increased risk of developing bacterial keratitis. The most common causative organism in contact lens-related bacterial keratitis is *Pseudomonas aeruginosa*, a bacterium that is notable due to its ability to spread rapidly, and lead to severe inflammation and tissue damage.

Other risk factors include a poor ocular surface, lid disease, prolonged topical steroid usage, lacrimal drainage system dysfunction, corneal abrasion, epithelial defects of any cause, and recurrent corneal erosion syndrome.

SYMPTOMS

Patients usually present with increasing photophobia, lid swelling, blur, and a watery discharge. They also frequently experience a foreign body sensation and increasing pain, although wearing a contact lens may mask symptoms. Occasionally there may be a mucopurulent discharge from an associated bacterial conjunctivitis.

SIGNS

There are several acute inflammatory signs within the cornea itself, with an inflammatory infiltrate, stromal edema, stromal loss, and keratic precipitates. The anterior chamber also displays signs of inflammation with cells and flare, and frequently a hypopyon and synechia. These are not usually due to infection within the anterior chamber but rather are signs of sterile inflammation secondary to the inflammatory mediators and bacterial toxins released within the cornea.

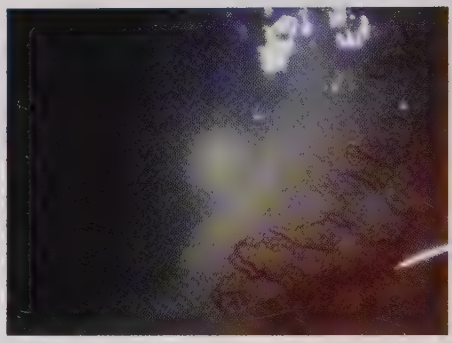


Fig. B-2 Peripheral corneal infiltrate with associated pannus in a patient with bacterial keratitis.

INCIDENCE

Uncommon (approximately 1/1000) outside of defined risk groups.

SIGNIFICANCE

Acutely sight-threatening.

DIFFERENTIAL DIAGNOSIS

Fungal keratitis; Herpes simplex keratitis (HSK); *Acanthamoeba* keratitis; Marginal keratitis.

SEE ALSO

Infectious crystalline keratopathy (ICK); Bacterial conjunctivitis; Gonococcal keratoconjunctivitis.

MANAGEMENT

Prompt investigation and treatment is essential.

Investigations

Whether all microbial keratitis needs to be thoroughly investigated for the causative organism is debatable. Certainly investigation is essential if the infection is not responding appropriately to treatment, to ensure other causative organisms such as herpes simplex virus, Acanthamoebae, fungi, and atypical mycobacteria are not missed.

Routine investigation includes inoculation of slides for Gram and



Blankaphor stains and bacterial culture plates and broth. Special stains for viruses and cultures for viruses, fungi, protozoa, and mycobacteria may also be added if thought appropriate. Even in the presence of a hypopyon there is no advantage in performing an anterior chamber tap; the hypopyon is usually a reactive change to the corneal infection.

Therapeutics

Even prior to bacterial stains and culture results, treatment with broad-spectrum bactericidal antibiotics is commenced. Current practice is either monotherapy with a fluoroquinolone (ciprofloxacin, ofloxacin, or moxifloxacin) or dual therapy with a fortified cephalosporin (cephalothin 5%) and aminoglycoside (tobramycin 1.3%). Drops are commenced hourly, day and night, usually for a minimum of 48 hours. The patient may be admitted to hospital if compliance is thought to be an issue, although most microbial keratitis is treated within the community, not requiring hospitalization. Once a good clinical response is observed the antibiotics can be gradually decreased to 2 hours by day, 4 hourly by night for 2 to 3 days, then qid usually for a week.

Antibiotics are not withdrawn because of the stain results due to the risk of polymicrobial infection, but may be added. If unresponsive to treatment, antibiotics may be changed as a result of sensitivity results although, if a good clinical response is observed, antibiotics are usually continued if the organism is found to be resistant to the antibiotic on sensitivity testing. This is probably explained by a higher drug concentration at the site of infection than is used for sensitivity testing.

The use of topical steroids in the treatment of bacterial keratitis is controversial. They are not used initially but may be added after an appropriate clinical response is noted to reduce inflammation, and thereby the degree of scarring and epithelial defects.

Dosing: G. ciprofloxacin/ofloxacin, or moxifloxacin q1h day, and night for 2 days followed by q2h day, and q4h night for 3 days. In the presence of a good clinical response, then qid for 1 week, or at least until closure of any epithelial defect. If indicated, G. fluorometholone acetate (0.1%) is added, bd for 2 days followed by qid, usually commenced on day 3 or 4 following a good clinical response to treatment.

Review

To follow the response to treatment it is best to follow the size, density, and edge of the corneal infiltrate. With effective treatment the initial response, usually observed over the first 1 to 2 days, is a stabilization of the infiltrate. The infiltrate will then decease in size and density, and the edge will usually become more distinct and rounded rather than infiltrative of the surrounding tissue. The hypopyon may increase in size over the first 1 or 2 days of treatment, probably as a response to the liberation of antigens from dead and dying bacteria. It then usually becomes more distinct and gradually resolves over a few days to a week. The epithelial defect may take days to a week or two to fully heal. Following resolution of the infection, an area of focal stromal loss from tissue digestion is often present at the site of maximal inflammation. This is especially so with Gram-negative organisms such as *Pseudomonas* that release lytic enzymes including collagenases.

B

Band keratopathy

DESCRIPTION

Precipitation of calcium salts from the tear film leading to a subepithelial, and anterior corneal stromal opacity.

There are several causes:

- ocular – longstanding inflammation as seen with chronic iridocyclitis, especially with juvenile rheumatoid arthritis, silicone oil in the anterior chamber, and phthisis bulbi
- metabolic – increased serum calcium and phosphate and chronic renal failure
- hereditary
- age-related – typically seen in patients in their 80s and older.

SYMPTOMS

Blurred vision, glare, surface irritation. May be asymptomatic initially.

SIGNS

The typical appearance is that of a diffuse, peripheral interpalpebral, subepithelial, gray opacity that starts near the limbus. This progresses across the inferior cornea being narrowest centrally. In addition, there are frequently small, anterior, star-shaped, chalky white calcium deposits scattered throughout. A clear, unaffected zone is maintained between the edge of the band and the limbus. The epithelium overlying the deposition is typically quite thin. When the band is very old, or if there is an associated stromal defect, there may be large chips of calcium deposited. These typically have an overlying epithelial defect. Often there is an association with cataract formation.

INCIDENCE

Uncommon (approximately 1/1000).

SIGNIFICANCE

Visual acuity is usually reduced several lines.

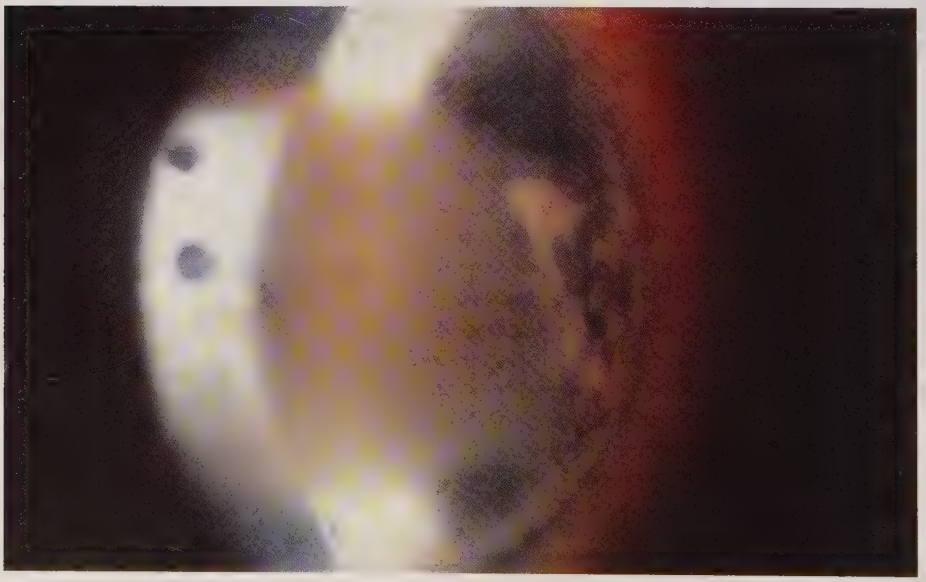


Fig. B-3 Band keratopathy with two large circular clear areas, and denser calcium deposition in the paralimbal area.



DIFFERENTIAL DIAGNOSIS

Rarely confused with other conditions as appearance is so typical. A similar deposition of calcium salts can occur on poorly cared-for contact lenses.

SEE ALSO

Iritis.

MANAGEMENT

Tear supplements

Topical lubrication may be helpful for associated surface irritation. Bandage contact lenses may be of benefit.

Surgery

Treatment is indicated for improvement in vision, or ocular comfort, with intervention often precipitated to maximize visualization during cataract surgery. Large chips of calcium can be manually removed with forceps, and residual calcium removed either by a combination of debridement and chelation or with excimer laser phototherapeutic keratectomy (PTK). Chelation is usually achieved with either sodium versenate or EDTA.

Prognosis

Unfortunately, recurrence of the band is common. This is especially the case with metabolic causes unless the underlying metabolic defect can be corrected. It is less of a problem with age-related band keratopathy where recurrence occurs very slowly over years. Consider referral for hyperparathyroid (hypercalcemic) work-up in an adult presentation.

B

Basal cell carcinoma (BCC)

Rodent ulcer

DESCRIPTION

This slow-growing tumor arises from the basal layer of the epidermis, and accounts for approximately 90% of all malignant eyelid tumors. It most commonly occurs on the lower eyelid, followed by the medial canthus, upper eyelid, and lateral canthus. Although non-metastatic, this tumor is locally invasive and can cause extensive local destruction if left untreated. It can invade the orbit (approximately 1–2%) and paranasal sinuses; this is most common with medial canthal tumors. Rarely, perineural spread may even occur (approximately 3%). Basal cell carcinoma (BCC) almost exclusively occurs in older patients and is typically seen in sun-damaged skin arising secondary to ultraviolet light exposure.

SYMPTOMS

The patient normally notices a slow-growing, often ulcerated painless lesion.

SIGNS

There are three clinical morphological types, all associated with loss of the lashes in the affected area. Posterior extension may lead to localized cranial nerve palsies.

Nodular

This is the most commonly seen lesion, and presents as a shiny, firm, pearly-edged nodule, often covered with small, dilated blood vessels.

Nodulo-ulcerative (rodent ulcer)

Appearance is similar to the nodular type. However, there is a central ulcerated area surrounded by a raised rolled edge.

Sclerosing (morpheaform, fibrosing)

This is the least common and most aggressive form of the disease, and may arise from a nodular BCC. It is characterized

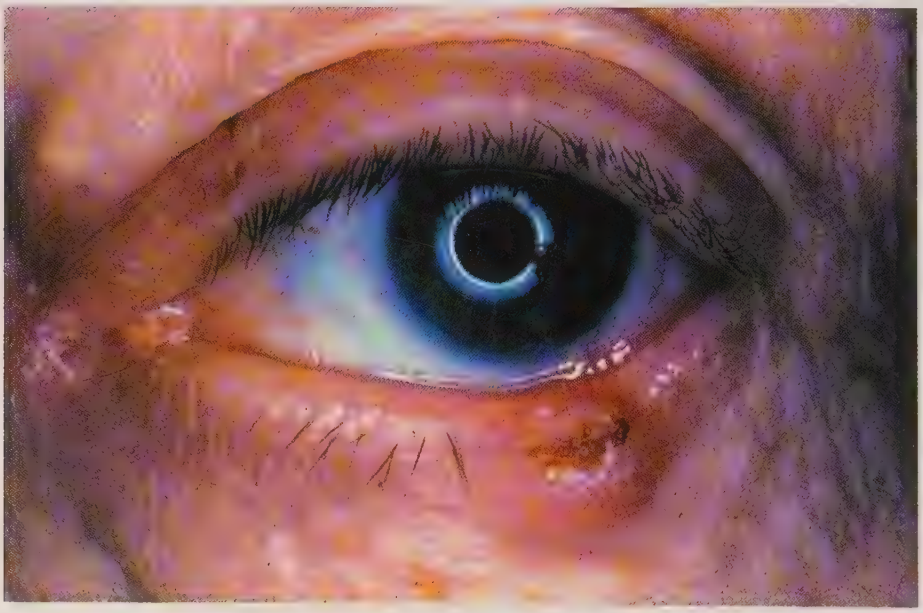
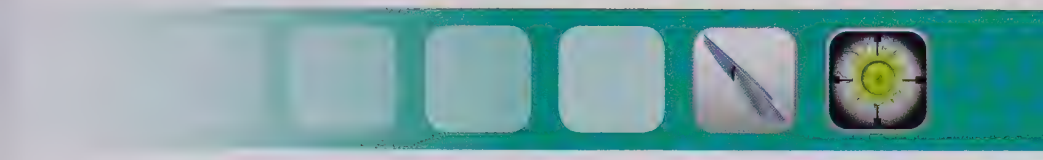


Fig. B-4 A small nodulo-ulcerative BCC involving the lower eyelid.



by a flat, indurated plaque of thickened skin. The extent of the lesion is often difficult to define on clinical inspection.

INCIDENCE

Uncommon (approximately 1/1000), especially more common in the elderly, and patients with a history of extensive UV exposure.

SIGNIFICANCE

Can be both sight- and life-threatening if left untreated for a long period of time.

DIFFERENTIAL DIAGNOSIS

Chalazion; Squamous cell carcinoma (SCC); Blepharitis – anterior; Actinic keratosis; keratoacanthoma.

SEE ALSO

Sebaceous cell carcinoma; Skin tumors.

MANAGEMENT

Surgery is the treatment of choice for all BCCs of the eyelid. Accurate referral including details of size, location, and history are therefore important.

Surgery

Although incisional and excisional biopsies may be appropriate to enable a histological diagnosis to be established, this is an aggressive tumor and can become more rapidly invasive following incomplete excision. For this reason, complete surgical excision is required. Adequate excision of the lesion can lead to the substantial loss of eyelid tissue, so that reconstruction of the defect rather than direct closure is sometimes required.

Cryotherapy

This has been used to treat some small nodular BCCs. It has the advantage of minimizing tissue loss, inconvenience, and discomfort to the patient but has the disadvantage of not allowing histological examination of tissue to establish both the diagnosis and the adequacy of treatment.

B

Bell's palsy – facial paralysis

Seventh cranial nerve palsy, facial nerve palsy

DESCRIPTION

Disruption of the seventh cranial nerve, the facial nerve, results in paralysis of the ipsilateral facial muscles, including the orbicularis oculi responsible for eyelid tone and closure. The facial nerve can be injured by many mechanisms at multiple sites on its long tortuous course from the brain stem. These are traditionally divided into supranuclear, nuclear, and infranuclear lesions.

Bell's palsy is the most common type of facial neuropathy, and refers to idiopathic facial nerve palsy with typical clinical features. The term is also frequently used to refer to all facial nerve palsies.

In addition to Bell's palsy, stroke, infection, trauma, and tumors can damage the nerve. The most common tumor to involve the facial nerve is an acoustic neuroma, either directly or as a result of surgical trauma during its resection. In this case, damage to the fifth cranial nerve, the trigeminal nerve, is also possible, leading to corneal anesthesia, and possible neurotrophic keratitis.

SYMPTOMS

Bell's palsy usually has a rapid onset, and is more common in people with diabetes mellitus and hypertension. It may be associated with decreased tearing and diminished taste. Approximately 75% experience complete recovery. Recovery usually commences within several weeks and is complete in 2 to 3 months. In the remaining 25% of patients a variable degree of weakness persists.

If exposure of the ocular surface occurs, redness, pain, photophobia, blurring of vision, and epiphora are common.

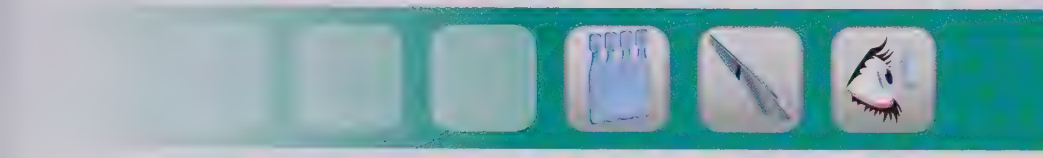
SIGNS

There is facial drooping on the affected side, including the mouth. Often the eye on the affected side cannot be closed and



Fig. B-5a Unilateral facial weakness with associated paralytic ectropion.

Fig. B-5b Inset showing patient with right Bell's palsy in primary gaze.



it may be difficult to move the forehead. There may be associated paralytic ectropion.

The external auditory canal should be inspected for evidence of a herpes zoster vesicular eruption (Ramsey–Hunt syndrome).

With exposure, conjunctival infection, edema, and epithelial disruption are common. Corneal epithelial disturbances are also common, ranging from mild punctate erosions to frank ulceration. Corneal ulcers resulting from exposure are usually inferior, rounded, and have a regular edge.

INCIDENCE

Uncommon (approximately 1/1000).

SIGNIFICANCE

There is usually a marked cosmetic concern and, if exposure keratopathy occurs, moderate discomfort and threat to sight.

DIFFERENTIAL DIAGNOSIS

Ectropion; Entropion.

SEE ALSO

Neurotrophic keratopathy.

B

MANAGEMENT

Tear supplements

Tear supplements are of prime importance in protecting the ocular surface from desiccation. During the acute phase of the disease, this is achieved with the frequent use of usually unpreserved lubricant drops and ointment. Consider also the use of a therapeutic contact lens. During the day ocular protection can be aided with the use of sunglasses, and at night with taping of the eyelid.

Dosing: lubricant ointment bd, and topical drops and/or gel qid continued until good lid function returns.

Surgery

A tarsorraphy, inferior lid shortening procedures, and superior lid lowering procedures are usually only used when the paralysis is permanent. They are particularly important if there is a neurotrophic keratitis from an associated trigeminal nerve palsy.

B

Blepharitis – anterior

Seborrheic blepharitis, Staphylococcal blepharitis

DESCRIPTION

An infection and/or inflammation of the anterior eyelid margin. This region is defined as anterior to and including the lash line, but not including the meibomian glands (see "Blepharitis – posterior"). It affects the glands of Moll, modified sweat glands at the base of the lashes, and the glands of Zeis, oil-secreting glands on the anterior lid margin. The condition is usually bilateral, symmetrical, and chronic in nature.

Staphylococcal overgrowth and seborrhea are thought to be intimately involved in the development of anterior blepharitis. Staphylococci are known to produce markedly inflammatory exotoxins, and it is likely that sensitivity to these toxins plays a major role. Staphylococci overgrowth is probably encouraged by the excess production of lipid associated with seborrhea.

SYMPTOMS

A disparity between the degree of signs and symptoms is a common feature of this disease. Symptoms result from disruption to normal ocular surface function and reduction in tear stability as reflected in a decrease in the tear break-up time (TBUT).

Typical symptoms include burning, grittiness, a foreign-body sensation, and mild photophobia as well as crusting and redness of the lid margins.

Symptoms typically fluctuate and are usually fairly symmetric. There may be exacerbations during winter, especially in cold climates. As with most forms of dry eye, symptoms are also often made worse in drying environments such as with air travel and air-conditioned offices.

SIGNS

There is variable lid thickening, vascular dilatation, and telangiectasia. The lashes



Fig. B-6 Mixed staphylococcal, and seborrheic blepharitis with associated folliculitis centrally. Note seborrheic scurf at base of lashes, and staphylococcal collarettes.



are often reduced in number (madarosis) and non-pigmented (poliosis). They are matted together and encrusted with scales; staphylococcal scales are typically hard and brittle, and form collarettes around the base of the lashes, whereas seborrheic scales are soft and greasy (scurf). Scarring within the lid margin from chronic blepharitis can lead to individual lashes becoming misdirected and touching the ocular surface (trichiasis).

There are frequently associated ocular surface signs with punctate fluorescein and rose bengal staining adjacent to the lower lid margin. Marginal keratitis may occur during acute exacerbations. An external hordeolum (stye) may develop if the glands of Moll or Zeis become infected.

INCIDENCE

Very common (greater than 1/10).

SIGNIFICANCE

Usually associated with low-grade symptoms, and a cosmetic problem. Chronic anterior blepharitis can cause tear-film instability and secondary changes in the conjunctiva and cornea.

DIFFERENTIAL DIAGNOSIS

Blepharitis – posterior; Basal cell carcinoma (BCC); Squamous cell carcinoma (SCC); Sebaceous cell carcinoma; Dry eye syndrome.

SEE ALSO

Rosacea keratitis; Marginal keratitis; Hordeolum – external.

MANAGEMENT

The chronic recurrent nature of the disease should be explained to the patient.

Lid hygiene

This is the mainstay of long-term treatment and is aimed at decreasing seborrhea and staphylococcal antigen load. Scrubbing of the lashes and lid margin is performed with a mild detergent to remove built-up scale and oils. This can be done with a commercially available product or diluted baby shampoo or a solution of sodium bicarbonate. Lid hygiene antiseptic foam solutions are also available and, used regularly, usually several times a week, can significantly reduce the bacterial load.

Tear supplements

Instability of the tear film, due to chronic inflammation, and dysfunction of the lipid component of tears, leads to many of the symptoms seen in this condition. This can be improved with the sparing use of tear supplements.

Therapeutics

Topical weak corticosteroids such as fluorometholone and antibiotics such as chloramphenicol or azithromycin 1% may be useful in acute exacerbations to reduce inflammation and staphylococcal load. They are used 4 times a day for 1–2 weeks. They are very much a short-term measure until other treatment modalities begin working. A topical liposomal tear-stabilizing agent (e.g. Tears Again) may be of particular benefit in the presence of a lipid layer abnormality with an abnormally short TBUT.

B

Blepharitis – posterior

Meibomian gland dysfunction, meibomianitis, meibomitis, meibomian seborrhea

DESCRIPTION

There are 30 to 35 meibomian glands opening onto both the upper and the lower eyelid margins, posterior to the lash line. These glands run for up to a centimeter through the tarsal plate. Lipid produced by the glands mixes with the aqueous tears to help stabilize the tear film. As such, dysfunction of the secretion of lipid (meibome) from these glands can lead to local inflammation and dry eye symptoms. Blockage of the glands can lead to sterile (chalazia) or infected cysts (internal hordeolum).

SYMPTOMS

As with anterior blepharitis there is frequently a disparity between the degree of signs and symptoms. Symptoms result from disruption to normal ocular surface function and reduction in tear stability as reflected in a decrease in the tear break-up time (TBUT).

Typical symptoms include burning, grittiness, a foreign-body sensation, and mild photophobia as well as crusting and redness of the lid margins.

Symptoms typically fluctuate and are usually fairly symmetric. There may be exacerbations during winter, especially in cold climates. As with most forms of dry eye, symptoms are also often made worse in drying environments such as with air travel and air-conditioned offices.

SIGNS

The key sign in posterior blepharitis is an abnormality of the meibomian gland secretions. In meibomian seborrhea excess secretions manifest as oil globules capping the meibomian gland orifices and form an oily or foaming tear film. In meibomianitis there is inflammation or obstruction of the glands. This may manifest as little pustules at the orifice. Expression of the contents of the glands demonstrates many to be blocked; those that do express release yellowy, viscous, even toothpaste-like secretions.

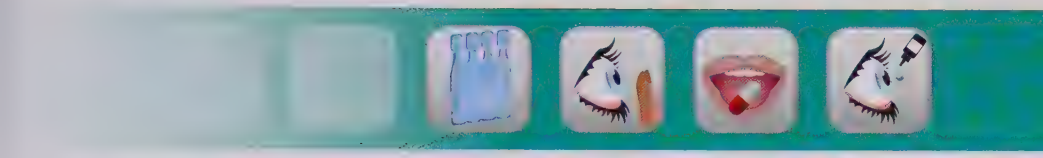
The lid margin is thickened and inflamed with vascular dilatation and telangiectasia. If chronic, the posterior margin is frequently scalloped secondary to scarring. Eversion of the lids may demonstrate small meibomian cysts. There is typically a course of superficial punctate keratopathy involving the inferior third of the cornea and inferior interpalpebral conjunctiva.

Pediatric blepharitis

Special mention is worth making of pediatric blepharitis because of its ability to cause a significant reduction in vision. This



Fig. B-7 Blocked meibomian gland orifices associated with lid thickening, telangiectasias, and lid scarring in chronic posterior blepharitis.



is a relatively uncommon condition usually affecting young boys and girls between 4 and 12 years of age. It is frequently bilateral. Signs include meibomian gland plugging, with or without associated lid inflammation, inferior superficial punctate keratopathy (SPK), and, frequently, inferior corneal neovascularization. The visual acuity may be decreased, and there may be a history of recurrent chalazia or hordeola.

INCIDENCE
Common (approximately 1/100).

SIGNIFICANCE
Usually only ocular surface irritation and cosmetic problem. When associated with marginal keratitis and rosacea keratitis it can be sight-threatening.

DIFFERENTIAL DIAGNOSIS
Blepharitis – anterior; Sebaceous cell carcinoma; Dry eye syndrome; Squamous cell carcinoma (SCC); Basal cell carcinoma (BCC).

SEE ALSO
Rosacea keratitis; Chalazion; Hordeolum – internal; Marginal keratitis.

MANAGEMENT

Advice
The chronic recurrent nature of the disease should be explained to the patient.

Lid hygiene
Although less central to disease control than with anterior blepharitis, this is an important component of the long-term treatment of posterior blepharitis. Scrubbing of the lashes and lid margin is performed with a mild detergent to remove built-up scale and oils. This can be done with either a commercially available product or diluted baby shampoo. Cleaning should be supplemented with warm compresses to melt solidified sebum, and expression of meibomian gland secretions. Lid hygiene antiseptic foam solutions are also available and, used regularly (usually several times a week), can significantly reduce the bacterial load.

Oral medication
Systemic tetracyclines are the mainstay of the treatment of posterior blepharitis. They are broad-spectrum antibiotics and are also thought to have a primary action in altering the nature of the lipid produced by the meibomian glands. They are also useful in the management of any associated facial rosacea. They are usually used initially at a high dose and then reduced to a lower dose for maintenance. A typical regimen is doxycycline 100 mg oral daily for 1 month followed by 50 mg orally for 2 months. Doxycycline is avoided in the treatment of pediatric blepharitis in children less than 8 years of age because of its ability to lead to staining of the teeth. In this group it is replaced with oral erythromycin syrup 30–50 mg/kg per day in divided doses. This is usually maintained for at least several months.

Therapeutics
Topical mild corticosteroids (fluorometholone), and broad-spectrum antibiotics (e.g. chloramphenicol or azithromycin 1%) can be used initially qid for 1 to 2 weeks to reduce inflammation and bacterial load.

Tear supplements
Instability of the tear film due to chronic inflammation and dysfunction of the lipid component of tears leads to many of the symptoms seen in this condition. This can be improved with the sparing use of tear supplements. A topical liposomal tear-stabilizing agent (e.g. Tears Again) may be of particular benefit in the presence of a lipid layer abnormality with an abnormally short TBUT.

B

Bullous keratopathy – pseudophakic and aphakic

Post-surgical corneal edema, ABK, PBK

DESCRIPTION

Anterior segment trauma experienced during intraocular surgery can lead to corneal endothelial cell damage, resulting in corneal edema. Although the edema will frequently clear in the days to weeks following surgery, there is occasionally sufficient damage to the endothelium such that months to years later irreversible edema will develop. Occasionally, surgical damage to the endothelium is so severe that irreversible corneal edema occurs immediately following the operation. Although significant damage to the endothelium can occur with any anterior segment procedure, it is most commonly seen following cataract surgery. Thankfully, irreversible corneal edema rarely occurs following modern small incision, phacoemulsification surgery by an experienced surgeon. However, it was far more common following extracapsular and intracapsular cataract surgery. This is in large part due to the improved control of the anterior chamber during phacoemulsification surgery and the introduction of viscoelastics; these help to protect the endothelium during surgery. Although probably best described as post-surgical corneal edema, the common terms for this condition are pseudophakic (PBK) and aphakic bullous keratopathy (ABK), depending on whether an intraocular lens (IOL) was or was not implanted. These terms are only used to describe permanent irreversible corneal edema.

SYMPTOMS

Poor vision, from both corneal edema and associated problems such as cystoid macular edema and glaucoma, is common. Initially, blurring of vision is most marked in the morning. Glare sensitivity and photophobia are also common. With the development of corneal epithelial blisters (bullae) and microcystic edema, the patient reports a sore gritty eye with occasional episodes of several days of acute pain from rupture of a bullous.

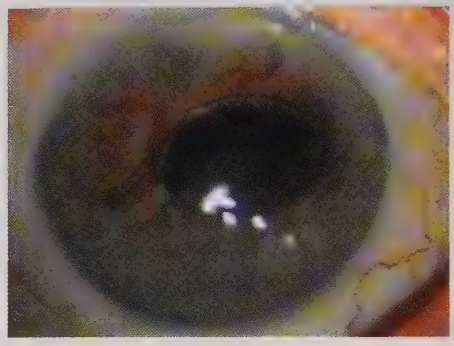


Fig. B-8a Pseudophakic bullous keratopathy. Note the disruption of the iris, and presence of an anterior chamber IOL.

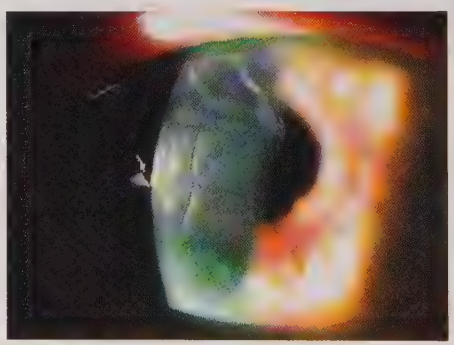
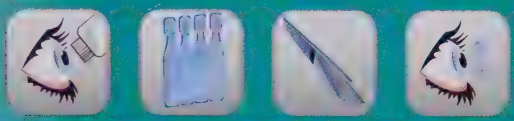


Fig. B-8b Marked corneal edema in bullous keratopathy.

SIGNS

The key sign is the presence of bullae, although these may be transient. They are seen as a small- to medium-sized (1 mm^2) area of raised epithelium with an underlying layer of clear fluid separating the epithelium from the corneal stroma. This is best seen on a slit lamp with high magnification and a narrow oblique slit. Other signs of corneal edema are usually present, with stromal thickening, Descemet's folds, and microcystic epithelial edema, seen as a "ground glass" appearance of the epithelium. Other signs of anterior



segment trauma may be present, including iris disruption, a peripheral iridectomy, vitreous disruption (with vitreous in the anterior chamber and to the wound), corneal scarring, and the presence of lens remnants. The position and style of any intraocular lens should be noted. Glaucoma is common, from both direct damage to the angle and the formation of peripheral anterior synechia. Cystoid macula edema is also common and may significantly reduce the visual potential.

INCIDENCE

Previously relatively common; however, with improved surgical techniques is now uncommon.

SIGNIFICANCE

Frequently leads to discomfort and loss of vision.

DIFFERENTIAL DIAGNOSIS

Fuchs' dystrophy; occasionally postoperative edema occurs because of unrecognized pre-existing endothelial disease.

SEE ALSO

Corneal graft; Corneal graft – lamellar keratoplasty; Corneal edema – overview; Cataract incisions.

MANAGEMENT

Specular microscopy

Residual endothelial cell number can be assessed with specular microscopy. Normal cell density in the elderly is approximately 2000 cells/mm^2; corneal edema typically occurs when the cell count falls below approximately 1000 cells/mm^2.

Tear supplements and therapeutics

Lubricants can reduce the grittiness and foreign-body sensation of microcystic epithelial edema. Hypertonic (5%) saline drops and/or ointment can be used to osmotically desiccate the corneal epithelium and improve vision; they have no action on stromal edema. Occasionally, daily use of a weak topical corticosteroid such as fluorometholone may help reduce ocular surface inflammation and increase comfort.

Therapeutic (bandage) soft contact lens

Application of a therapeutic (bandage) soft contact lens, often worn on a 30-day extended wear basis, can significantly improve ocular comfort.

Surgery

PBK and ABK are one of the most common indications for performing either a full thickness or a lamellar (DSAEK) corneal graft. If the IOL is in a poor position, it may be replaced or repositioned at the time of the corneal graft surgery. If the IOL is an older style lens associated with corneal endothelial damage (such as an iris clip or closed loop anterior chamber lens), it should be replaced at the time of surgery. If the eye is painful and there is very limited visual potential, corneal micropuncture or a total conjunctival (Gunderson) flap may be used to stabilize the ocular surface and reduce pain.

Laser surgery

In a painful eye with poor visual potential, a deep phototherapeutic keratectomy (PTK) may render the eye pain-free and stabilize the ocular surface.

C

Canaliculitis

Lacrimal canaliculitis, dacryostenosis

DESCRIPTION

Canaliculitis is an infection of the lacrimal passage, usually caused by the Gram-positive bacillus *Actinomyces* (*Streptothrix* sp.). This condition usually occurs without any recognizable predisposing factors and frequently relapses.

SYMPTOMS

Presentation is with chronic unilateral epiphora with associated mucopurulent discharge.

SIGNS

The punctum is often pouting with tear retention at its surface. There may be slight tenderness and redness over the course of the canaliculus. Firm indentation and expression of the canaliculus often results in expression of large, chalky white, "sulfur" granules.

Attempted dilatation and syringing reveals signs of canalicular obstruction.

INCIDENCE

Very rare (less than 1/100,000).

SIGNIFICANCE

Epiphora will persist if the disease is not treated.

DIFFERENTIAL DIAGNOSIS

Dacryocystitis.

SEE ALSO

Epiphora.

MANAGEMENT

Referral to the general practitioner is appropriate.

Surgery

Repeat curettage of the canaliculus to remove the granules coupled with several weeks of topical antibiotics may be sufficient to treat the disease successfully. If this is insufficient, incision of the



Fig. C-1 Concretion expressed from the inferior punctum in chronic canaliculitis.



canaliculus with associated curettage is indicated. Unfortunately, the incision itself may sometimes lead to chronic epiphora from canalicular scarring.

Therapeutics

Traditionally the drug of choice for treating the underlying infection has been topical penicillin. As allergy to topical penicillin is so common, other drugs such as topical ciprofloxacin can be used.

C

Caruncular cyst

Onchocytoma, eosinophilic cystadenoma, apocrine cystadenoma, oxyphilic adenoma

DESCRIPTION

This rare cystic tumor is one of only a few pathological lesions affecting the caruncle. Histologically, one or more cystic cavities are lined by proliferating epithelium.

SYMPTOMS

The lesion is usually painless and slow growing. It does not lead to ocular surface symptoms.

SIGNS

The tumor is typically yellowish, tan, or reddish and has a rounded, shiny surface. It has the appearance of a large grain of rice under the epithelium. It most commonly occurs on the caruncle, but may rarely involve the conjunctiva, lacrimal sac, lacrimal gland, or accessory lacrimal glands of Krause.

INCIDENCE

Very rare (less than 1/100,000).

SIGNIFICANCE

Usually benign but may rarely undergo malignant transformation.

DIFFERENTIAL DIAGNOSIS

Normal caruncular tissue can occasionally appear raised and slightly cystic.

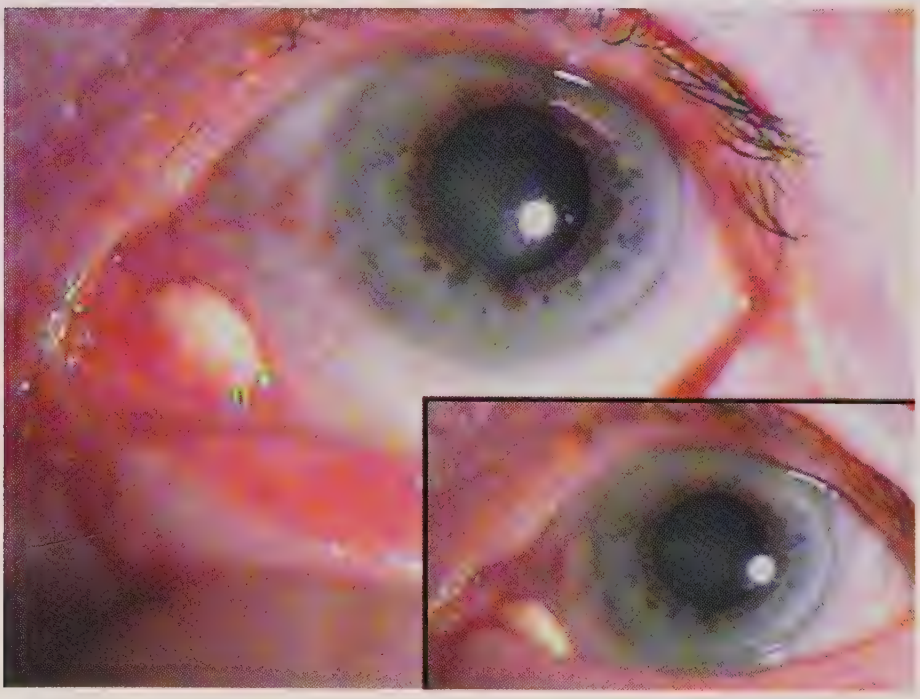


Fig. C-2a Typical caruncular tumor with dome-like raised appearance. The main figure shows the growth from the caruncle tissue.

Fig. C-2b Inset photograph shows the normal appearance of the lesion with the patient's eye in primary gaze.



SEE ALSO
Conjunctival pigmentary lesions; Conjunctival papilloma.

MANAGEMENT

Surgery
Simple excision is indicated for most tumors, although temporization with simple observation is quite reasonable. The lesion is normally easily resected as a single intact tumor. If tissue is left after excision it is said to represent a greater risk for malignant transformation.

Cataract

Nuclear sclerosis, cortical cataract, posterior subcapsular cataract

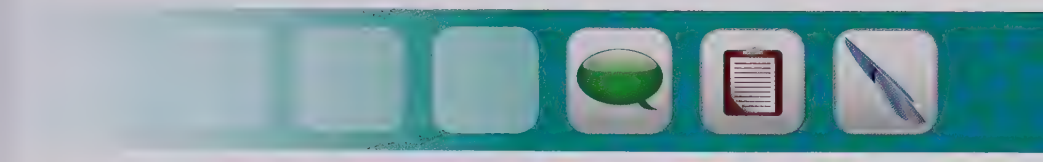
DESCRIPTION

Cataract refers to any opacity within the crystalline lens. Opacities may be acquired or congenital, focal or generalized, large or small, multiple or singular, and may affect any layer of the lens, nucleus, or cortex, anterior or posterior. In a mature cataract the whole lens is opaque, whereas a hypermature cataract is characterized by a loss of fluid from the lens, causing shrinkage with wrinkling of the anterior capsule. Finally, in a Morgagnian cataract, the cortex has liquefied, and the totally sclerotic nucleus sinks within the capsular bag.





Fig. C-3a, 3b Grading of the two most common forms of cataract using the Wilmer Institute Cataract Grading System. a Nuclear sclerosis. b Cortical cataract.



CAUSES

Age-related

Age-related changes within the lens are the result of accumulation and compaction of lens fibers and precipitation of lens proteins, particularly the alpha-crystallins. There are three types of senile cataract:

- nuclear sclerosis – leads to hardening, and brunescence of the nucleus
- cortical cataract – results in typical white, spoke-like opacities, often with associated vacuoles or "water-clefts"
- posterior subcapsular cataract (PSC) – is a plaque just anterior to the posterior capsule and results from posterior migration of equatorial lens epithelial cells.

Trauma

Direct penetrating trauma of the lens usually leads to rapid cortical opacification; rarely, this may remain isolated to the area of trauma. Blunt non-penetrating trauma can result in an anterior subcapsular opacity, typically with a flower-like appearance. Electrical shock, ionizing radiation, and microwaves can also lead to cortical and PSC changes.

Other causes

These include nutritional deprivation and drug–induced (especially with topical or systemic corticosteroids), inflammatory (from chronic uncontrolled anterior uveitis or atopic keratoconjunctivitis), and inherited disorders. Congenital cataracts are typically localized to delineated nuclear layers. Cataract is also more common in tobacco smokers, and occurs earlier in patients with high degrees of ammetropia.

SYMPTOMS

Lenticular opacities lead to a number of symptoms, the most common of which is blurring of vision. Cataract can also lead to glare, particularly with cortical opacities. Patients may also report halos around headlights, especially with night driving. Nuclear sclerotic cataract may lead to a myopic shift and may also alter color perception as it acts as a yellow filter.

INCIDENCE

The incidence is age-related. By 90 years of age everyone has some lens opacity, and in developed countries approximately 25% of 80-year-olds have had a cataract extraction. Cataract is the most common cause of blindness worldwide.

SIGNIFICANCE

Cataract can pose a marked threat to sight if left untreated. This is particularly a problem in developing countries with limited health resources.

SEE ALSO

Cataract incisions; Intraocular lenses (IOLs); Posterior capsular opacification (PCO).

MANAGEMENT

Grading

Grading of the cataract can be performed using the Wilmer Institute Cataract Grading System. The photographs demonstrate grades 1 to 4 nuclear sclerosis on the left, and the consideration of the lens in 16 equal parts to grade cortical cataract on the right. Posterior subcapsular cataract may be graded in terms of area (mm^2).

Advice, and review

Review is appropriate when the cataract is not considered functionally or clinically significant. Consider a 12-month interval for review.

Surgery

The most common indication for cataract surgery is a visual functional deficit attributable to the cataract. The age, infirmity, and visual demands of the individual patient need to be taken into account when making the decision whether to offer surgery. The most common surgical technique for cataract removal is now small-incision phacoemulsification with intraocular lens implantation. This procedure is usually performed as day surgery with local anesthesia and usually results in rapid visual recovery.

C

Cataract incisions

DESCRIPTION

All aspects of cataract surgery have evolved over the past 30 years, including wound construction, wound location, wound closure, lens extraction techniques, intraocular lenses, and perioperative drug administration. As such, many different types of cataract surgery wounds are seen in everyday clinical practice.

SIGNS

There are three main cataract extraction techniques and three main intraocular lens (IOL) styles. The IOL style directly affects the size of the wound. Wounds are usually made with single-use disposable metal blades or reusable diamond blades. The femtosecond YAG laser is also being trialed to perform not only the wound incisions but in addition the capsulorhexis and even nuclear disruption.

Intracapsular cataract extraction (ICCE)

This is now largely of historical interest, as ICCE was phased out in favor of extracapsular cataract extraction (ECCE) techniques in the early 1980s in most developed countries. Extraction of the whole crystalline lens intact requires an 11–12-mm wound. This can be placed at the superior limbus or just slightly more peripheral in the sclera. A superior peripheral iridectomy is routinely performed at the time of surgery to reduce the risk of pupil block.

Extracapsular cataract extraction (ECCE)

Expression of the nucleus with subsequent aspiration of the cortex requires a 10-mm wound, through either the superior limbus or sclera. Although this was the standard method of cataract extraction up to the early 1990s it is now a much less common technique in developed countries. A variety of smaller, often self-sealing, incision ECCE techniques have been developed and are commonly used in developing countries, especially for more advanced cataracts.

Small incision techniques (mainly phacoemulsification)

Modern cataract surgery using one of several phacoemulsification techniques uses a 3.2–2.0-mm incision placed through the sclera, limbus, or clear cornea, superiorly, obliquely, or temporally,

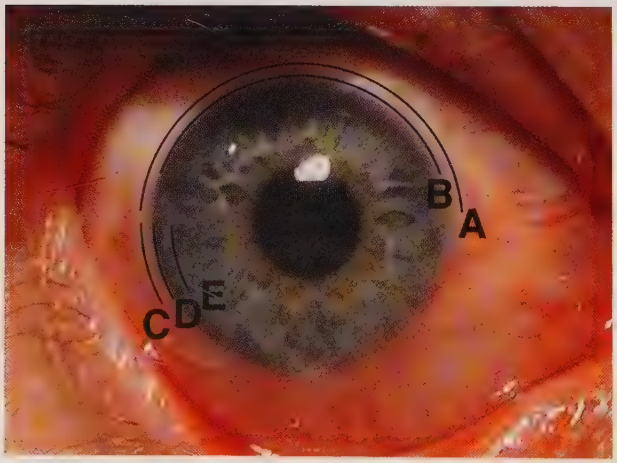


Fig. C-4 External entry site for standard cataract incisions. **A** Superior intracapsular incision. **B** Superior extracapsular incision. **C** Temporal scleral (phacoemulsification) incision. **D** Temporal limbal (phacoemulsification) incision. **E** Temporal clear corneal (phacoemulsification) incision.



for both the cataract extraction and IOL insertion. All these incisions enter the anterior chamber through a small trans-corneal tunnel. In addition, one or two small (typically 1 mm in length) paracentesis wounds through the clear cornea or limbus are made at the time of surgery to facilitate nuclear manipulation and cortex extraction.

The entry into the anterior chamber through Descemet's membrane with both the main wound and the paracentesis wounds can often be seen years later as a white line of faint scarring on the posterior surface of the cornea.

The ICCE and ECCE incisions require sutured closure while only a small percentage of phacoemulsification wounds require suturing.

INCIDENCE

Very common (greater than 1/10); approximately 25% of all 80-year-olds in developed countries have already had a cataract extraction.

SIGNIFICANCE

Cataract surgery can lead to a range of early- and late-postoperative complications. These include suture rupture, endophthalmitis (usually within the first week after surgery), bullous keratopathy, cystoid macular edema (CME), posterior capsular opacification (PCO), and retinal detachment. Most of these complications are more common if the surgery was complicated with posterior capsular rupture and disruption of the anterior vitreous face.

A cataract wound can also lead to astigmatism. This is much more common with large sutured wounds used in extracapsular and intracapsular surgery. Slippage of the wound results in flattening of that axis of the cornea, while tightening, as can occur with sutured wounds, typically results in steepening along that axis. As such, wound slippage with a superior wound results in against-the-rule astigmatism while an overly tight superior wound results in with-the-rule astigmatism.

SEE ALSO

Corneal edema; Posterior capsular opacification (PCO); Endophthalmitis; Cataract; Intraocular lenses (IOLs); Cataract surgery complications.

MANAGEMENT

Therapeutics

Preoperatively, patients are treated with a topical non-steroidal anti-inflammatory drug to reduce the risk of postoperative cystoid macular edema. Standard postoperative drops are a broad-spectrum antibiotic and a moderate strength corticosteroid. More intensive corticosteroid drops are used if there is a predisposition to increased inflammation such as in diabetics or patients with a history of uveitis or in patients with a corneal graft. Tear supplements can also be used in the early postoperative period to reduce ocular irritation.

Dosing: G. ketoralac qid 3 days prior, and for 1 week post-surgery then bd for 2 weeks. G. chloramphenicol or a fluoroquinolone qid 1 week post-surgery. G. prednisolone acetate (or a similar strength topical corticosteroid) qid for 1 week then bd for 2 weeks post-surgery.

Advice

As indicated above, the presence of previous cataract surgery may indicate a predisposition to other pathology.

Sutures are much less commonly used with a modern small incision wound. If present, they are usually removed 1 to 4 weeks after surgery. Dissolvable sutures are not used in the main wound in cataract surgery.

Additional investigations

If there is any doubt as to the patency of the incision, the intraocular pressure should be measured and outflow of aqueous from the incision tested with a Seidel test.

C Cataract surgery complications

DESCRIPTION

Cataract surgery complications, though thankfully uncommon, can be visually devastating. The incidence of complications varies significantly among different studies and depends on which complication is being investigated. One study found an average rate of significant adverse events to be 1/200, although this rate decreased significantly for higher-volume surgeons. Complications occur more frequently in high-risk patients such as those with pseudoexfoliation syndrome (PXF), previous trauma, vitrectomy, diabetic retinopathy, or with dense nuclear sclerosis. It is important to identify risk factors prior to surgery to allow alterations to the surgical plan to reduce the risk. In addition, appropriate prophylactic medical treatment of patients, both pre- and postoperatively, can reduce the risk of complications.

Risk factors

Many pre-existing conditions can raise the risk of complications occurring during and following cataract surgery. An abbreviated list of the main risk factors and associated complication/s is given below.

- PXF: zonular dehiscence, delayed intraocular lens (IOL) dislocation in the capsular bag, posterior capsular rupture
- Dense nuclear sclerosis: increased risk of corneal edema and posterior capsular rupture
- Previous vitrectomy: posterior capsular rupture
- Diabetic retinopathy: macular edema
- Alpha-1A adenoceptor receptor blocker (doxazolin): intraoperative floppy iris syndrome with posterior capsular rupture and iris trauma
- Small pupil: posterior capsular rupture and iris trauma
- Previous trauma with zonular loss: posterior capsular rupture and lens dislocation

Some intraoperative complications can lead to an increased risk of postoperative complications.

- Prolonged surgery time: corneal edema and photic damage to the macula
- Posterior capsular rupture, lens dislocation with vitreous loss: macular edema

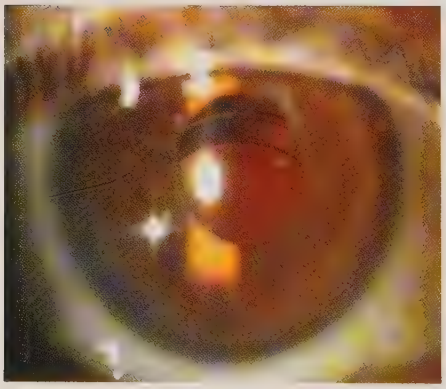


Fig. C-5 Subluxation of a posterior chamber intraocular lens.

Refractive complications

Postoperative refractive problems include:

- Incorrect IOL spherical power – may arise from irregular corneal curvatures, especially following corneal refractive surgery. Soft contact lenses need to be out for 1 week and rigid lenses for 1 month prior to keratometry for IOL power calculation.
- Incorrect IOL toric power – effective power may be altered if placed on incorrect axis.
- Positive dysphotopsias – may arise from edge effects, especially with large irregular pupils.
- Negative dysphotopsias – thought to arise from light reflection anterior to the IOL.
- Induced corneal astigmatism – may arise from a wound burn or corneal sutures.

INCIDENCE

Posterior capsular rupture occurs at a rate of approximately 1/100, or less; endophthalmitis with the use of



intracameral antibiotics occurs at a rate of around 1/8000 cases. With the use of pre-operative topical ketoralac, clinically significant cystoid macular edema occurs at a rate less than 1/200 cases.

PREVENTATIVE MEASURES

The surgical plan should be altered in the presence of certain known preoperative risk factors. A good example of where significant changes to the surgical plan can help prevent complications is with patients known to have used an alpha-1A adrenoceptor receptor blocker. In this case a combination of the following can reduce risk: planned longer wounds to reduce the risk of iris prolapse; the use of cohesive viscoelastic, iris hooks, or a pupil expansion ring; intracameral phenylephrine; and reduced intraocular fluid flows.

Sometimes planning extends to the months prior to surgery. Diabetic patients with pre-existing macular edema are at a high risk of worse edema following surgery and should be treated prior to surgery.

Preoperative treatment with topical ketoralac qid for 3 days followed by several weeks of postoperative treatment appears to significantly reduce the risk of developing postoperative cystoid macular edema. Intraoperative intracameral (into the anterior chamber) antibiotics significantly reduce the risk of developing endophthalmitis. A drop of a topical antihypertensive agent such as alphagan reduces the risk of an early rise in intraocular pressure. Postoperative topical corticosteroids reduce postoperative inflammation.

SEE ALSO

Endophthalmitis; Cataract; Cataract incisions; Intraocular lenses (IOLs).

MANAGEMENT

The specific management of each complication is beyond the scope of this article; many are covered under specific conditions.

Therapeutics

A common perioperative drug regimen is ketoralac qid for 3 days prior to surgery, intracameral cephazolin 1 mg at the end of surgery, 1 drop of brimonidine prior to application of the shield at the end of surgery. Following surgery, a topical antibiotic qid for 1 week; ketoralac and a topical corticosteroid such as prednisolone acetate qid for 1 week then bd for 2 weeks.

C

Chalazion

Meibomian cyst

DESCRIPTION

Within the tarsal plate of both the upper and the lower eyelid are 30–35 modified sebaceous glands (meibomian glands). Blockage of one of these glands can lead to retention and stagnation of the sebaceous secretions and formation of a chronic, sterile, lipogranulomatous inflammatory lesion termed a chalazion. Chalazia are composed of an aggregation of activated macrophages, called epithelioid cells, surrounded by lymphocytes. Epithelioid cells may fuse to be multi-nuclear giant cells. These chalazia occur spontaneously or may follow an acute hordeolum. They occur more commonly in people with chronic blepharitis, facial rosacea, and seborrheic dermatitis. Extremely rarely, a sebaceous cell carcinoma of the tarsal plate can have an appearance clinically indistinguishable from a chalazion.

SYMPTOMS

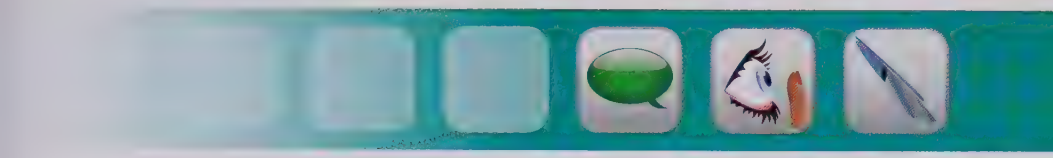
As the lesion is painless and rarely affects vision, patients usually present because of



Fig. C-6a Normal external appearance of a chalazion.



Fig. C-6b Chalazion as a skin lump under the palpebral conjunctiva.



the lump. Chalazia are usually unilateral and develop over a period of weeks. They are commonly multiple or recurrent.

SIGNS

A rounded, firm, non-tender subdermal lump within the tarsal plate is easily palpated. A small chalazion is 2–3 mm in diameter, whereas a large chalazion is 7–8 mm in size. Frequently the chalazion will rupture spontaneously. It usually ruptures posteriorly through the palpebral conjunctiva. If this has occurred there will be a polypoid, granulomatous reaction noticeable at the site of rupture. If it ruptures anteriorly through the skin, an ulcerated skin lesion may be present.

INCIDENCE

Common, especially in patients with the risk factors listed above.

SIGNIFICANCE

Normally only a cosmetic problem.

DIFFERENTIAL DIAGNOSIS

Hordeolum – internal and external; Skin tumors (neoplasms – particularly in elderly patients).

SEE ALSO

Blepharitis – posterior; Rosacea keratitis; Sebaceous cell carcinoma.

MANAGEMENT

Advice

Chalazia do not necessarily require treatment and may be left to spontaneously resolve, if there is no effect on vision and if the patient is happy with the cosmetic appearance. Spontaneous resolution of the lump typically takes several months. Neoplastic lesions should be ruled out.

Lid hygiene

Topical or systemic antibiotics are not effective and not justified due to the sterile nature of the lesion. Local lid therapy can be effective for small-to-medium-size lesions. Hot compresses at least four times daily, combined with gentle cleansing of the lid margins, may be effective. One method of applying heat is with microwaved wheat bags (commonly used for arthritis therapy). If massage of the lump is performed, it should be gentle to avoid gland rupture. With local therapy it may take 2 to 4 weeks for the chalazia to resolve.

Surgery

Surgical incision of the gland with curettage of the contents is the most common form of treatment required and may be used for larger chalazia or if topical therapy is ineffective. Following surgery patients are usually placed on a 1-week course of topical antibiotics and corticosteroids to clear any associated infection and suppress the inflammatory reaction. The material obtained from primary incision and curettage is rarely submitted for histopathological examination, unless there are features suggestive of an underlying sebaceous cell malignancy.

Steroid injection

Injection of the lesion with 0.1–0.2 mL of a corticosteroid such as triamcinolone is a good alternative to surgery. This has an approximately 80% success rate for a single infection.

Prevention

If associated with facial rosacea or posterior blepharitis, ongoing lid hygiene or even oral doxycycline may be needed to prevent recurrences.

C

Chemical burns

Alkali burn, acid burn

DESCRIPTION

Acute chemical burns of the ocular surface are frequently bilateral and may occur accidentally, usually at work, or from intentional assault. The most important noxious chemicals are acids and alkalis. Detergents and hydrocarbons can also disrupt the ocular surface but usually lead to little more than epithelial haze and mucus layer disruption that typically heals rapidly.

Depending upon the pH and degree of exposure, both acids and alkalis can damage ocular surface and intraocular structures. Alkalis are particularly dangerous because of their ability to penetrate the epithelium and damage the limbal stem cell apparatus, thereby leading to ocular surface scarring, epithelial defects, and vascularization.

Sodium hydroxide (NaOH), lye, is one of the most common alkalis and is supplied in both a dry flake and liquid form. It is a common ingredient in industrial cleaning solutions. Other alkalis include calcium hydroxide [$Ca(OH)_2$ or lime] and potassium hydroxide (KOH or caustic potash).

SYMPTOMS

Immediate pain, redness, epiphora, and blurring of vision occur with most burns. Very severe burns may be fairly pain-free because of damage to superficial sensory nerves. In the long term the degree of ocular surface recovery and restoration of vision depends largely on the extent of the burn, the effectiveness of first aid, and initial treatment.

SIGNS

Signs of injury are often worse inferiorly, secondary to both gravity and the protective nature of the upper lid.

Initial presentation

Following first aid the eye is assessed for the extent of the injury. Initial grading of the injury may be difficult, and grading may change when reassessed a day or two later. The extent of the injury can be graded depending on the degree of corneal epithelial loss, limbal ischemia, corneal stromal opacification, and intraocular damage as evidenced by cataract, iris damage, or a raised intraocular pressure.

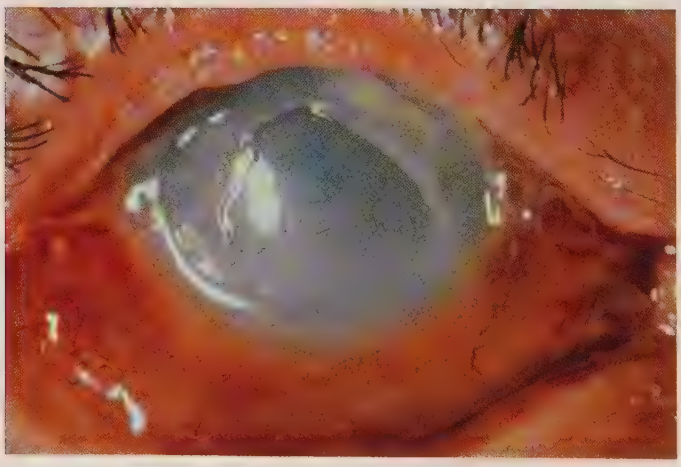


Fig. C-7 Corneal epithelial loss, and limbal ischemia following recent alkali burn.



- Grade 1: clear corneal stroma and no limbal ischemia
- Grade 2: corneal haze but less than 1/3 limbal ischemia
- Grade 3: corneal haze blurring iris detail with 1/3 to 1/2 limbal ischemia
- Grade 4: opaque cornea with more than 1/2 limbal ischemia

Long-term prognosis

In most burns no long-term damage occurs. In severe cases, damage to the various anterior segment and ocular surface structures can lead to many problems. These include limbal stem cell dysfunction with epithelial disturbances, trichiasis, entropion, ectropion, ptosis, conjunctival scarring, dry eye, cataract, and glaucoma. Although epithelial opacification with superficial punctate keratopathy (SPK) and epithelial loss is common, for the purpose of grading the injury the cornea is only assessed for the degree of stromal opacification.

INCIDENCE

With improved work safety, first aid, and improvements in medical management, loss of vision secondary to chemical burns has become less frequent in most developed countries. Unfortunately it is probably becoming more frequent in many industrializing nations.

SIGNIFICANCE

While many chemical injuries are mild and result in no ongoing ocular problems, severe burns can result in bilateral blindness.

SEE ALSO

Limbal stem cell deficiency; Conjunctival scarring – overview.

MANAGEMENT

First aid

The most important, vital aspect of treatment is to rapidly return the ocular surface to as near as possible a normal pH of 6.9. This can be confirmed with litmus paper applied to the inferior fornix. First aid is achieved by immediate, copious irrigation of the eye with tap water for at least 15 minutes. Where available, sterile saline should be used and continued as the patient is sent to the emergency clinic. Any particulate alkali should be removed with forceps. Aside from the ocular injury, it is important to assess and treat any related skin injuries.

Review

All chemical burns should be reviewed the following day because of the risk of a significant change in signs over the first day or two.

Initial treatment

- Grades 1 and 2. Very mild Grade 1 burns can be treated with topical antibiotics and review. If more marked, treat with topical steroids such as fluorometholone acetate and a broad-spectrum antibiotic such as chloramphenicol qid until the epithelium heals.
- Grades 3 and 4. This is a medical emergency: prompt treatment and early management are the keys to early epithelial healing and maximal visual recovery. These patients are frequently admitted to hospital and are treated with hourly topical unpreserved corticosteroids, homatropine 2% bid, chloramphenicol tds, sodium citrate 10% 2-hourly, and sodium ascorbate 10% 2-hourly. This regimen is designed to reduce inflammation and loss of corneal stromal tissue. The drops are tapered as the epithelium begins to heal.

Long-term treatment

Corneal grafts, amniotic membrane transplants, tarsorhaphies, and limbal stem cell transplants have all been tried with varying success in the management of the long-term effects of severe chemical burns. The outcome is commonly disappointing, underscoring the extensive damage to multiple structures that occurs with such injuries.

C

Chlamydial conjunctivitis

Trachomatous inclusion conjunctivitis (TRIC), adult inclusion conjunctivitis, adult chlamydial conjunctivitis

DESCRIPTION

The obligate intracellular organism *Chlamydia trachomatis* causes three different ocular diseases in humans – ophthalmia neonatorum, trachoma, and adult inclusion conjunctivitis. Different serotypes predominate in each type of disease. Adult inclusion conjunctivitis is essentially a sexually-transmitted disease and, unlike most viral and bacterial causes of conjunctivitis, is chronic, with symptoms often persisting for many months. This chronicity is due to the systemic nature of the disease, with repeat inoculation of the ocular surface.

SYMPTOMS

Adult inclusion conjunctivitis can lead to an acute or chronic slight red eye with irritation and a watery or mucopurulent discharge. Symptoms may be uni- or bilateral. The vision may be blurred if there is also an associated keratitis.

SIGNS

Adult inclusion conjunctivitis leads to an acute or chronic follicular conjunctivitis, often with associated mucopurulent discharge. Follicles appear as avascular, whitish, and amorphous nodules, like small grains of rice, located in the tarsal and forniceal conjunctiva. There may be an associated keratitis with scattered nummular subepithelial infiltrates, marginal infiltrates, or even pannus formation. There is often a palpable and slightly tender preauricular or submandibular lymph node. If chronic, a superior micropannus may occur.



Fig. C-8 Follicles involving the superior tarsal conjunctiva in adult chlamydial conjunctivitis.



INCIDENCE

Chlamydial infections are one of the most common causes of chronic conjunctivitis. Young, sexually active adults are most at risk of contracting adult inclusion conjunctivitis. It is an uncommon infection outside this group.

SIGNIFICANCE

Unlike trachoma, adult inclusion conjunctivitis does not lead to scarring of the ocular surface. It is, however, associated with pelvic inflammatory disease, a chronic chlamydial infection of the cervix, uterus, and fallopian tubes. This can lead to infertility through fallopian tube scarring and obstruction and, therefore, the diagnosis and treatment of this disease is especially important.

DIFFERENTIAL DIAGNOSIS

Adenoviral keratoconjunctivitis; Molluscum contagiosum; Herpes simplex keratitis (HSK); Trachoma.

SEE ALSO

Conjunctival scarring; Follicular conjunctivitis; Bacterial conjunctivitis; ophthalmia neonatorum (in neonates).

MANAGEMENT

Adult inclusion conjunctivitis is often a sexually transmitted disease. It is essential that any sexual partners as well as the patient are identified and treated; this is often best done through referral to a sexual health clinic or the patient's general practitioner. This is important both for the health of the sexual partner and to prevent reinfection of the patient. Chlamydial infections are a notifiable disease in many countries.

Microbiological investigation

Chlamydia trachomatis can be detected by both staining and culture. It can also be detected by direct immunofluorescence as well as polymerase chain reaction (PCR) to detect bacterial antigen. PCR has become the investigation of choice in many centers because of its ease of collection, specificity, and sensitivity.

Oral antibiotics

As described above, adult inclusion conjunctivitis is a systemic infection that is commonly associated with pelvic inflammatory disease in women. Treatment is with oral doxycycline 100 mg daily for 10–14 days. Single dose treatment with 1 gram of azithromycin orally has previously been used to treat uncomplicated chlamydial cervicitis successfully. Referral to a gynaecologist or urologist is appropriate in women with urinary symptoms. Standard topical antibiotic or topical steroid treatments are ineffective because they do not treat the systemic infection.

C

Climatic droplet keratopathy (CDK)

Labrador keratopathy, spheroidal degeneration, elastotic degeneration, Bietti's nodular dystrophy

DESCRIPTION

Climatic droplet keratopathy (CDK) is a degenerative disease of unknown cause although UV light exposure, especially extreme sun exposure, appears to be the main etiological factor. Other postulated causative factors include aging, low humidity, microtrauma from wind and sand, UV exposure from welding, and exposure to extremes of temperature. There may also be a familial component to its development.

Interestingly, one of the first reports in the Western ophthalmic literature was by Bietti, who described the disease in Somali fishermen who, no doubt, experienced extreme solar UV exposure.

Histopathologically, the lesions are localized to Bowman's membrane and are difficult to differentiate from calcium. They are considered a form of elastotic (actinic) degeneration.

SYMPTOMS

As this condition only occurs where there has been extensive UV light exposure, there are often associated actinic changes such as pterygia and pingueculae. Consequently, surface irritation and burning are common.

SIGNS

In the initial stages of CDK small, superficial, golden-colored globules are seen in the peripheral interpalpebral cornea. As the

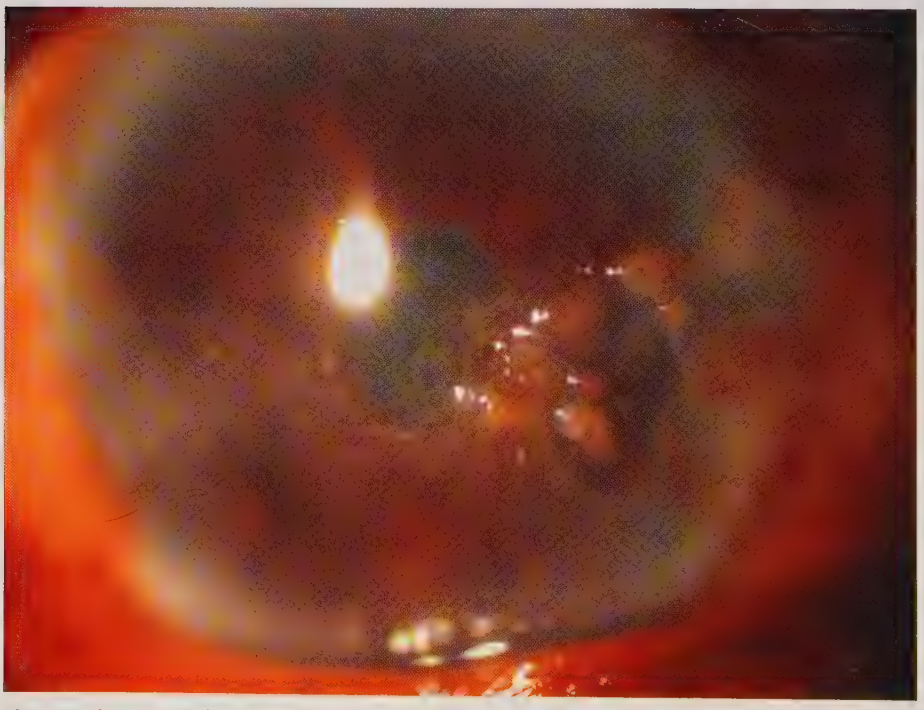
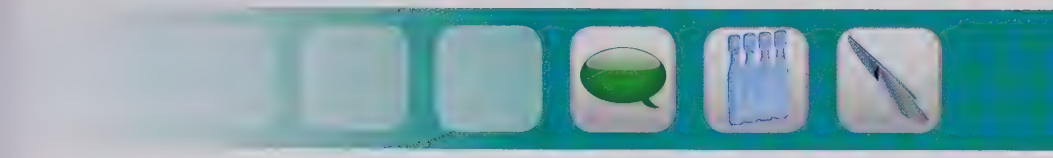


Fig. C-9 Large central golden-colored droplets with an associated, more generalised corneal opacity in an advanced case of CDK.



disease progresses the lesions develop in the more central cornea and coalesce to form raised lesions. The surrounding corneal stroma is frequently slightly opaque. The condition is almost always bilateral.

INCIDENCE
Rare, and isolated to risk groups as described above.

SIGNIFICANCE
The condition is most commonly fairly mild and restricted to the peripheral cornea, and as such is usually not sight-threatening. If it progresses to involve the central cornea, a reduction in vision may occur.

DIFFERENTIAL DIAGNOSIS
Band keratopathy; Granular dystrophy; Salzmann nodular degeneration.

SEE ALSO
Pterygium; Pingueculum.

MANAGEMENT
Advice
Strong encouragement with regard to the wearing of adequate sunglasses with good side-protection is important. This can reduce continued UV damage that not only can lead to progression of this condition but may also lead to other UV-related ocular diseases.

Tear supplements
Ocular lubricants are often helpful in reducing associated ocular surface irritation.

Surgery
If severe, superficial debridement or a superficial keratectomy may be indicated. Occasionally, a penetrating keratoplasty (corneal graft) may be necessary to restore corneal clarity and regularity to the corneal surface.

C

Concretions

Conjunctival lithiasis, calcium concretions

DESCRIPTION

Concretions are small white to yellow crystalline deposits most commonly seen in the tarsal and forniceal conjunctiva. They are more commonly seen in the elderly and following conjunctivitis. Grouped concretions can sometimes appear on the palpebral (tarsal) conjunctiva associated with an old chalazion or internal hordeolum. In this situation they probably form secondary to blockage of the meibomian glands with accumulation of lipid under the conjunctiva. Histopathologically they appear to be epithelial inclusion cysts filled with epithelial and keratin debris, although secondary calcification can occur. They are distinct from follicles, which are focal lymphoid hyperplasias having diffuse edges, and inclusion cysts, which are thin-walled lesions containing fluid.

SYMPTOMS

They are usually asymptomatic although, rarely, may erode and lead to a recurrent foreign-body sensation.

SIGNS

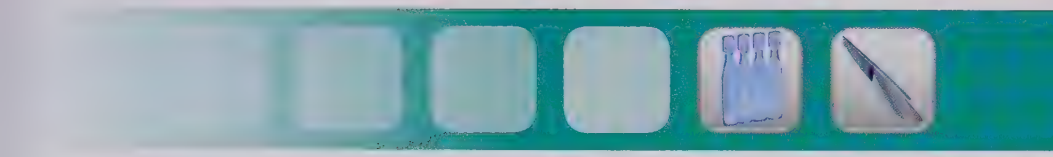
The concretions are chalky white to yellow lesions with distinct edges typically seen in the inferior conjunctival fornix or on the palpebral conjunctiva. They are typically quite small, having a diameter less than 1 mm, although they may have a diameter of up to 3 mm. Smaller concretions are usually flat although larger ones may be raised. Forniceal concretions are often seen within a small clear cyst.

INCIDENCE

Very common (greater than 10%), especially in the elderly.



Fig. C-10 Multiple eroded concretions stained yellow with fluorescein.



SIGNIFICANCE
May rarely lead to slight ocular surface irritation.

DIFFERENTIAL DIAGNOSIS
Abrasion; conjunctival retention cyst.

SEE ALSO
Chalazion; Trachoma.

MANAGEMENT
Tear supplements
Foreign-body sensation can be reduced by the use of ocular lubricants.

Surgery
Eroded concretions leading to ocular surface irritation can be removed under topical anesthetic at the slit lamp with a 25-gauge needle.

C

Conjunctival intraepithelial neoplasia (CIN)

Squamous cell carcinoma, corneal intraepithelial neoplasia, conjunctival squamous cell carcinoma, ocular surface squamous neoplasia, Bowen's disease, conjunctival dysplasia, carcinoma in situ

DESCRIPTION

Squamous tumors are the most common neoplastic tumors of the ocular surface and most frequently arise in the interpalpebral limbus or conjunctiva. There is often a history of excessive sun exposure; consequently, other UV-related ocular surface changes, such as pterygia, pinguecula, or lipoidal degeneration, may be present. Rarely, the tumors may be secondary to contact lens use, when they may arise superiorly, or from a rare inherited skin disease, xeroderma pigmentosa. They are usually slow growing and spread locally by direct extension, only very rarely metastasizing to a distant site.

Tumors that are restricted to the epithelium are usually referred to as intraepithelial dysplasia. Once they extend through the epithelial basement they are referred to as a squamous cell carcinoma. Recurrence following excision is common (25–50%) and is thought to occur both secondary to incomplete excision of the initial lesion and from new lesions occurring in other areas of UV-damaged epithelium.

SYMPTOMS

Patients may present complaining of ocular surface irritation, a mass, or blurred vision if the area of abnormal epithelium extends into the pupil. Up to 30% of tumors are picked up as an incidental finding on routine examination.

SIGNS

The three main morphological types are:

- en plaque – this is the most common appearance; tumor is gelatinous, raised, and has a "stuck-on" appearance, with tufts of superficial blood vessels

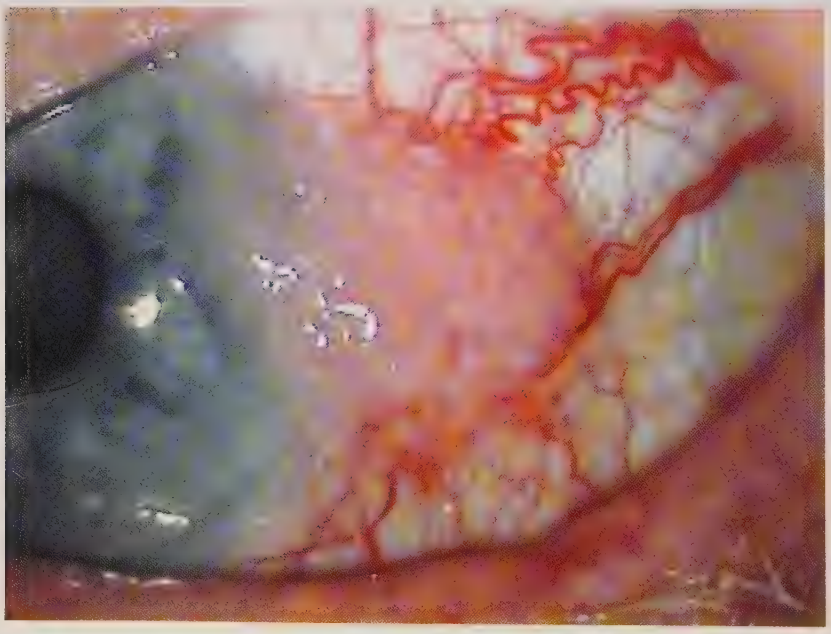
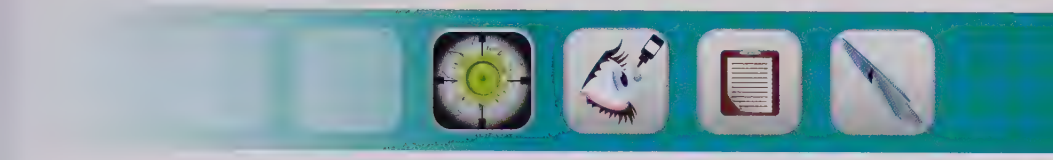


Fig. C-11 Large papillomatous CIN with prominent "feeder" vessels.



- papillomatous – the appearance is wart-like with a raised discrete lesion with surface corkscrew-like blood vessels
- diffuse – this is the least common, with the tumor diffusely involving an area of thickened conjunctiva or limbus. It may resemble an area of chronic conjunctivitis.

All three types are commonly associated with an area of gray-white hyperplastic epithelium that extends centripetally onto the cornea, typically with fimbriated edges. All may be associated with an overlying whitish area of keratinization.

INCIDENCE

In high sun exposure areas still rare (approximately 1/100,000), and usually occurs in the elderly. The incidence is significantly higher in patients with HIV infection.

SIGNIFICANCE

Although usually easily resected, this tumor can very rarely threaten vision through invasion of the globe and life through posterior extension into the middle cranial fossa.

DIFFERENTIAL DIAGNOSIS

Pterygium; Melanoma – conjunctival; Nevus; Limbal stem cell deficiency; Conjunctival lymphoma; Epibulbar choristoma; Episcleritis.

SEE ALSO

Squamous cell carcinoma (SCC) of the eyelid; Conjunctival pigmented lesions.

MANAGEMENT

Impression cytology (although not commonly available) may be useful in making the initial diagnosis. A superficial layer of epithelial cells is harvested from the ocular surface by adherence to a nitrocellulose filter and examined by a pathologist.

Surgery

Simple excision of the tumor and surrounding tissue with subsequent histopathological examination is the treatment of first choice. Adjunctive treatment such as diathermy or cryotherapy is recommended by some clinicians but does lead to more tissue damage.

Therapeutics

Recurrences are increasingly being treated with topical agents. The most commonly used drug is mitomycin C. This is a radiomimetic agent and has the potential to damage the ocular surface, even the limbal stem cells, if used at too high a dose or too many times. Therefore, after two rounds of treatment with mitomycin C many clinicians will change to topical interferon-alpha or, less frequently, vitamin A. Topical mitomycin C frequently leads to a red, slightly sore eye, even occasionally an epithelial defect. If this occurs the drug should be stopped immediately. Inflammation of the inferior lid skin is also common but can be prevented by careful application of the drop followed by active cleaning of the skin with a towel and warm water.

The advantages of topical treatment over repeat surgery are that it limits the degree of tissue damage and treats other unsuspected small areas of dysplasia.

Dosing: G. mitomycin C (0.04%) qid for 1 week, then nil for 1 week then qid for a second week. G. interferon-alpha (1 million units/mL) treatment is qid for several months.

Review

As recurrences can occur years following excision of the initial lesion, indefinite yearly review is recommended.

C

Conjunctival lymphoma

Lymphoid tumors

DESCRIPTION

Lymphomas are potentially malignant neoplasias of the immune system predominantly affecting the lymph glands. They are classified as Hodgkin or non-Hodgkin disease. Hodgkin lymphomas of the conjunctiva are exceedingly rare. Conjunctival lymphomas are usually restricted to the conjunctiva itself but, rarely, may occur in association with more generalized systemic disease. Histologically, the majority of conjunctival lymphomas are composed of monoclonal proliferations of B-cell lymphocytes and are divided into two categories; MALT (mucosal-associated lymphoid tissue, often termed MALTomas), and non-MALT tumors. MALT tumors are most common and are less aggressive with an excellent 5-year survival rate, whereas non-MALT tumors are considered highly malignant and invasive. Lymphoid tumors of the conjunctiva are associated with systemic lymphoma in about 30% of patients. Clinically, conjunctival lymphomas are indistinguishable from non-malignant lymphoid tumors, benign reactive lymphoid hyperplasia, or atypical lymphoid hyperplasia. These tumors contain mature follicles composed of B-cell lymphoblasts.

SYMPTOMS

The lesion is usually asymptomatic except for concern expressed by the patient for the enlarging mass and associated redness.

SIGNS

The tumor is located in the conjunctival stroma, or deep to Tenon's fascia, involving the space between the conjunctival epithelium and the sclera. It is usually unilateral, but may be bilateral in as many as 20% of cases, is salmon pink in color (hence the term "salmon patch"), and is usually relatively fleshy and diffuse. It often originates in the fornix or adjacent to the limbus. Differentiation between malignant



Fig. C-12 Typical salmon-colored subconjunctival thickening seen in conjunctival lymphoma.



and non-malignant lymphoid lesions cannot be reliably made on clinical history and examination.

INCIDENCE
Very rare (less than 1/100,000). Most commonly seen in young to middle-aged adults.

SIGNIFICANCE
May be life-threatening.

DIFFERENTIAL DIAGNOSIS
Benign lymphoid hyperplasia; non-pigmented conjunctival nevus.

SEE ALSO
Conjunctival intraepithelial neoplasia (CIN); Melanoma – conjunctival; Episcleritis.

MANAGEMENT
Surgery
An excisional biopsy is indicated to determine the nature of the lesion. Routine histopathology is supplemented with special monoclonal antibody investigations to establish the nature of the lymphocytes, and whether they are monoclonal or polyclonal. Even with these studies it may be very difficult to establish the true malignant potential of an individual lesion. Monoclonal B lymphocyte proliferation is more suggestive of malignancy.

Radiotherapy
If, following histopathological investigation, malignancy is suspected, local radiation to the area is indicated due to the fairly radiosensitive nature of these lesions.

C

Conjunctival papilloma

Conjunctival warts, human papilloma virus infection

DESCRIPTION

Conjunctival papillomas are caused by infection with the human papilloma virus (HPV) serotypes 6 and 11. HPV is a double-stranded DNA virus transmissible by direct contact. Conjunctival papillomas usually commence as a single lesion and multiply by autoinoculation leading to multiple lesions. They exist in two forms:

- pedunculated papillomas, which most commonly involve the inferior forniceal or palpebral conjunctiva and may extend to involve the puncta and caruncle
- sessile papillomas, which are less common and are broad-based, relatively vascular lesions. They usually occur on the bulbar conjunctiva.

SYMPTOMS

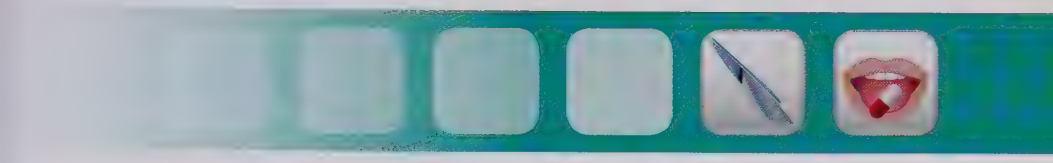
Conjunctival papillomas are often asymptomatic or may lead to epiphora from direct involvement of the punctum or through secondary occlusion. Occasionally, they lead to a foreign-body sensation.

SIGNS

Pedunculated papillomas have a typical uneven cobblestone appearance with dilated tortuous vessels visible below the epithelial surface. They are the same



Fig. C-13 Multiple papillomata involving the caruncle.



color as the normal conjunctiva, or slightly whiter, and are often multiple. Sessile papillomas are invariably single and unilateral. They are fleshy and vascularized. Papillomas vary widely in size and may even grow so large as to interfere with normal lid function.

INCIDENCE

Rare (approximately 1/10,000), and occur usually in children or young adults.

SIGNIFICANCE

Usually only a cosmetic problem, but may lead to epiphora. HPV has also been implicated in the pathogenesis of conjunctival squamous tumors.

DIFFERENTIAL DIAGNOSIS

Papillomatous squamous neoplasia; conjunctival mucoepidermoid carcinoma (clear cell tumor), dermoid.

SEE ALSO

Conjunctival intraepithelial neoplasia (CIN); Caruncular cyst; Skin tumors – benign.

MANAGEMENT

Like skin warts, conjunctival papillomas also usually eventually spontaneously regress with time. If they are large and lead to ocular surface irritation, epiphora, or significant cosmetic concern, intervention may be considered, so referral is advisable.

Intralesional injection

Injection of the papilloma with 0.1–0.2 mL of 106 units/mL interferon-alpha frequently leads to rapid regression of the lesions. This is usually not done in association with surgery, as it is felt that surgery may result in liberation of the virus, and lead to seeding across the conjunctiva. This may be followed with a 4- to 6-week course of topical interferon-alpha, 106 units/mL qid. Bleomycin has also been shown to be an effective treatment for skin papillomata when used as an intralesional injection.

Oral medication

The H_2-receptor antagonist cimetidine has recently been successfully used to treat recalcitrant papillomas. This is taken orally as a 6-week course 400 mg bid.

Surgery

Surgical excision has been used to remove the lesions. Unfortunately, this leads to some conjunctival scarring, and recurrence of the lesions following excision is common. Cryotherapy and carbon dioxide laser vaporization have also been tried with some success.

C

Conjunctival pigmented lesions

Primary acquired melanosis, caruncular nevus, conjunctival melanoma, conjunctival nevus, ocular melanocytosis, freckle

DESCRIPTION

The conjunctiva is anatomically subdivided into bulbar, forniceal, and palpebral areas. In addition, two specialized areas exist medially, the plica semilunaris and caruncle. Pigmented substances such as drugs and foreign bodies can stain any part of the conjunctiva (notably argyrosis with silver compounds). In addition, primary pigmented melanocytic lesions can arise within the conjunctiva. These are divided histopathologically into melanosis, nevus, and melanoma. Melanoma is a malignant highly metastatic tumor of the melanocytes. It is discussed under "Melanoma – conjunctival."

Benign melanosis

Accumulations of normal melanocytes within the epithelium are referred to as melanosis. Benign racial melanosis occurs in many dark-skinned people. It arises during early childhood and affects both eyes asymmetrically. The pigmentary lesions are flat, intraepithelial, and are often most marked around the limbus and perforating branches of the anterior ciliary artery or nerve. Freckles (ephelis) are also flat but often singular and may exist in fair-skinned individuals.

Primary acquired melanosis (PAM)

PAM is a premalignant condition, usually unilateral, and usually occurring in fair-skinned races. It usually affects older people and appears as a flat, light to dark brown pigmentation, most commonly affecting the bulbar conjunctiva. The affected area is often extensive and may evolve over time. There are two types of histological classification – with (high-risk) or without (low-risk) cellular atypia. Lesions with cellular atypia are premalignant and carry an approximately 50% chance of malignant transformation.

Nevi

A conjunctival nevus is a common benign ocular nevus consisting of an accumulation of melanocytes, epithelial cells, and nevus cells (related to melanocytes) within the epithelium and/or the subconjunctival tissue. They are histologically classified into junctional, subepithelial, compound, blue, or congenital melanocytosis. The usual benign nevus is flat or slightly elevated and moves freely in the conjunctiva over the sclera. They are often cystic, paralimbal, and contain a variable degree of pigment, ranging in color from deep-brown to pink. They often become more pigmented around puberty and most commonly occur at the nasal or temporal limbus and, rarely, in the fornix, tarsus, or on the cornea. Blue nevi are rare congenital nevi and are typically deeply pigmented areas appearing blue to slate-gray in color.

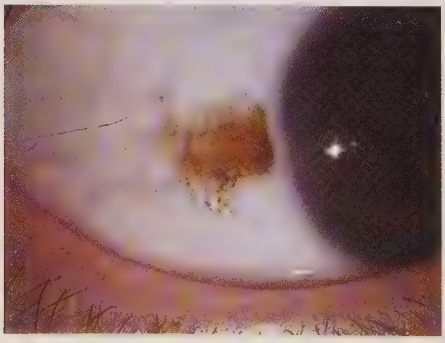


Fig. C-14a Prominent pigmented conjunctival nevus.

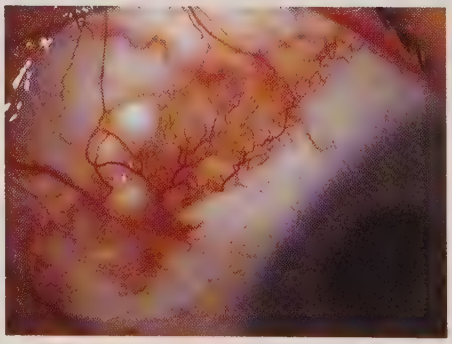


Fig. C-14b Non-pigmented cystic conjunctival nevus.



Congenital melanocytosis

May involve only ocular tissue (ocular melanocytosis) or may also involve skin (oculodermal melanocytosis or nevus of Ota). As with blue nevi, the lesion is usually blue to gray and may be associated with a uveal nevus. They are both associated with malignant transformation, usually leading to uveal or skin rather than conjunctival melanoma.

SYMPTOMS

Patients are asymptomatic except for concern with regard to the appearance and potential for malignant transformation. Children with nevi and freckles often present around puberty, because of a noted change in the color of the lesion.

INCIDENCE

Melanosis is the norm in some dark-skinned races. Nevi are common (approximately 1/100). Melanoma and primary acquired melanosis are both rare (approximately 1/10,000).

SIGNIFICANCE

Most pigmented lesions are benign although some have malignant potential. Of particular malignant potential are PAM and congenital melanocytosis.

DIFFERENTIAL DIAGNOSIS

Conjunctival intraepithelial neoplasia (CIN).

SEE ALSO

Melanoma – conjunctival; Melanoma – eyelid; Melanoma and nevus of the iris; Nevus – eyelid.

MANAGEMENT

Advice

Patients should be advised to re-present for examination if there is any alteration in size, color, or height of the pigmented lesion. Six- to 12-monthly review of an evolving nevus is appropriate. Photographic records of lesions are very useful.

Therapeutics

Topical mitomycin C 0.04% may be used to treat high-risk PAM not amenable to surgical treatment. Care should be taken with respect to the length of treatment; more than 4 weeks of total accumulated treatment time appears to place patients at a heightened risk of developing limbal stem cell deficiency.

Surgery

A thorough history and examination reveals the nature of most conjunctival pigmentary lesions. If there is any concern with regard to malignant transformation, an incisional or excisional biopsy should be performed with submission of the lesion for histopathological evaluation.

Multiple biopsies are needed in all cases of PAM to identify any areas with cellular atypia. If present, such areas can be excised and cryotherapy applied to reduce the risk of transformation into melanoma.

C

Conjunctival scarring – overview

Conjunctival cicatrization, ankyloblepharon, symblepharon

DESCRIPTION

Scarring of the conjunctiva can be a visually devastating end point of several diseases. It can result in combined dry eye syndrome from a reduction in aqueous tears because of scarring of the lacrimal ductules and a loss of the mucin layer from loss of conjunctival goblet cells. It can also lead to trichiasis and disturbance of lid function through forniceal shortening or ankyloblepharon (adhesion of the lid margins to each other) and symblepharon (adhesion of one conjunctival surface to another) formation. The ultimate result of such a severe disturbance of the function of the ocular surface is keratinization and opacification of the cornea.

CAUSES

Conjunctival scarring may be mild to severe and focal or generalized, depending upon its cause. A thorough examination of the ocular surface is essential to ensure that what one is seeing is not the start of a severe conjunctival scarring condition such as ocular cicatricial pemphigoid.

Surgical trauma

Many surgical procedures on the eye require a trans-conjunctival approach, thus resulting in localized conjunctival scarring. This may interfere with subsequent surgery, such as glaucoma filtering surgery.

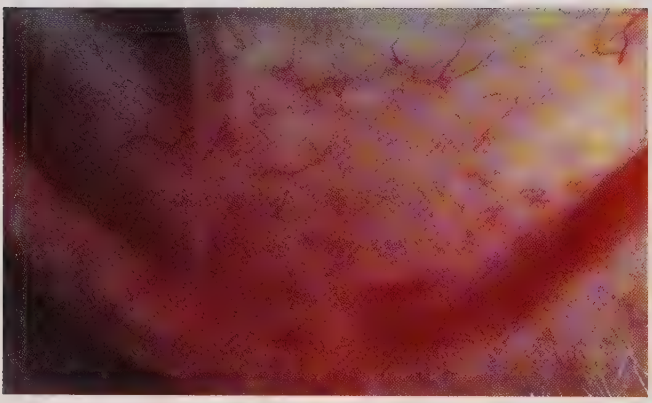


Fig. C-15a Subconjunctival scarring with resultant inferior forniceal shortening in ocular cicatricial pemphigoid.

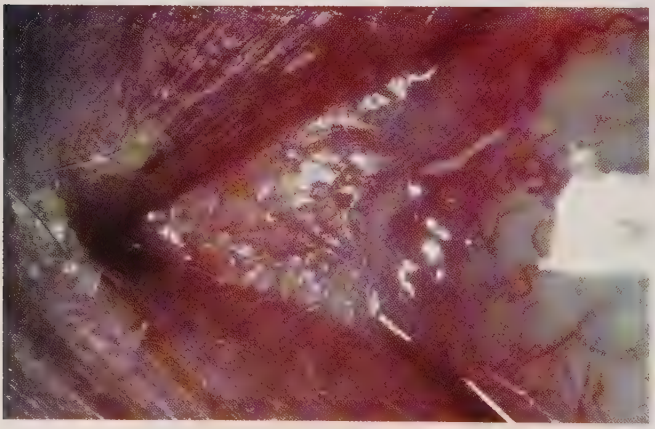


Fig. C-15b Typical caruncular flattening, scarring, and keratinization seen in ocular cicatricial pemphigoid.



Trauma
Chemical, thermal, radiation, and mechanical trauma can all lead to conjunctival scarring. The most destructive of these is chemical injury, especially acid and alkali burns. These can lead to widespread conjunctival epithelial loss and resultant scarring.

Immune mediated
Both ocular cicatricial pemphigoid and "graft versus host" disease can lead to generalized conjunctival scarring and may result in devastating dry eye.

Infection
Most infectious conjunctivitis does not lead to conjunctival scarring. The exception is with membranous conjunctivitis where there is epithelial loss and secondary conjunctival shrinkage and symblepharon, and even ankyloblepharon, formation. Trachoma also results in typical conjunctival scarring with formation of a Von Arlt's line on the superior tarsal conjunctiva and secondary trichiasis.

Allergy
Atopic keratoconjunctivitis typically leads to conjunctival shrinkage, forniceal shortening, and generalized subconjunctival scarring.

SYMPTOMS
If severe enough, disruption of the ocular surface from interference with tear and lid function can result in symptoms of grittiness, burning, and foreign-body sensation. This may lead to blurring of vision from corneal epithelial changes.

SIGNS
Aside from looking for signs helpful in identifying the specific diagnosis, signs of ocular surface abrasion from trichiasis and features of dry eye, such as a reduced tear meniscus and fluorescein and rose bengal staining, should be sought.

SIGNIFICANCE
Although often mild, depending on the etiology and extent of conjunctival scarring, conjunctival cicatrization can be a very significant and sight-threatening condition.

DIFFERENTIAL DIAGNOSIS
See causes listed above.

SEE ALSO
Ocular cicatricial pemphigoid (OCP); Trachoma; Chemical burns; Atopic keratoconjunctivitis.

MANAGEMENT
Tear supplements
Retentive and viscous tear supplements, as both ointments and drops, are helpful in restoring ocular surface function and reducing symptoms.

Surgery
Progressive scarring of unknown cause may require investigation with a conjunctival biopsy to look for typical features of ocular cicatricial pemphigoid with complement and immunoglobulin deposition along the basement membrane zone. Histopathological assessment of atopic keratoconjunctivitis will reveal typical changes with multiple eosinophils.

C

Conjunctivitis – overview

DESCRIPTION

This is an overview of the many different forms of conjunctivitis that are covered in more detail in individual articles in this book. Conjunctivitis is a non-specific term signifying inflammation of the conjunctiva, with dilation of the conjunctival blood vessels giving the characteristic red eye appearance. One of the main features distinguishing between acute bacterial infections and other causes is its chronicity: chronic conjunctivitis (lasting longer than 2 weeks) is uncommon with bacterial and simple viral infections.

CAUSES

Conjunctivitis occurs in a number of pathological conditions.

Infective

- Bacterial – acute, hyperacute, and chronic
- Viral – including adenovirus, herpes simplex, molluscum
- Chlamydial – ophthalmia neonatorum, trachoma, adult inclusion conjunctivitis
- Lice – phthiriasis palpebrarum

Allergic

Seasonal, giant papillary, vernal, and atopic.

Ocular surface disease

- Blepharitis
- Keratoconjunctivitis sicca – Sjögren's syndrome, lacrimal, and lipid layer anomalies

Immune

Ocular cicatricial pemphigoid (OCP), Stevens–Johnson syndrome.

Toxic

Drop sensitivity, chemical injury, photokeratopathy.

Mechanical

Eye rubbing, mucus fishing syndrome, artifactual conjunctivitis, or contact lens use.

Nasolacrimal duct obstruction

Chronic dacryocystitis.

Neoplastic

Some invasive ocular surface neoplasias, such as conjunctival intraepithelial neoplasia (CIN), can mimic conjunctivitis.

SYMPTOMS

The patient presents with red eyes and may also have noticed a purulent or watery discharge. A watery discharge is associated



Fig. C-16 Severe bilateral conjunctivitis due to Stevens-Johnson syndrome.



with a viral conjunctivitis while a purulent discharge is associated with a bacterial infection. Chlamydial conjunctivitis can have a mixed presentation – like bacterial or viral. Both of these conditions tend to be unilateral initially, then bilateral, although herpes simplex disease tends to stay unilateral. A foreign-body sensation is common with most causes of conjunctivitis, although itch will tend to suggest an allergic etiology.

SIGNS

- Follicles consist of lymphoid hyperplasia within the palpebral conjunctival stroma, more pronounced inferiorly. Clinically they appear as multiple small "grains of rice" embedded in the tarsal conjunctiva. There are surrounding accessory blood vessels. Possible causes include viral or chlamydial infections, as well as a hypersensitivity to topical medication.
- Papillae are focal elevations of the conjunctiva, particularly on the superior palpebral, and are best highlighted on lid eversion by using fluorescein. Papillae consist of a central blood vessel, with conjunctival epithelial hyperplasia, and stromal edema. There may also be a diffuse white infiltrate of chronic inflammatory cells. Giant papillae are greater than 1 mm in diameter; conversely, normal papillae can occur on the superior edge of the tarsal plate (inferior edge when everted). Papillae can occur in bacterial conjunctivitis, blepharitis, atopic conjunctivitis, giant papillary conjunctivitis, and vernal conjunctivitis.
- Membranes or pseudomembranes consist of coagulated exudate firmly or loosely adherent to the conjunctiva. Acute bacterial conjunctivitis should be considered, including gonococcal and severe adenoviral conjunctivitis among others.
- Discharge and hyperemia. See above under "Symptoms."
- Chemosis is the term given to sub-conjunctival edema and is seen especially with acute allergic conjunctivitis.
- Sub-conjunctival hemorrhages. Multiple small hemorrhages can occur in viral and bacterial conjunctivitis.
- Scarring. An uncommon sign in developed countries, but consider trachoma, ocular cicatricial pemphigoid, and atopy.
- Lymphadenopathy. Enlarged lymph nodes in the pre-auricular and sub-mandibular regions are often seen with viral or chlamydial infections.
- Keratopathy. Associated corneal infiltrates may be seen with some infective forms of conjunctivitis such as chlamydial and adenovirus.

INCIDENCE

Very common (greater than 1/10).

SIGNIFICANCE

Conjunctivitis tends to be associated with mild to moderate cosmetic concern and discomfort, although some of the less common causes such as ocular cicatricial pemphigoid can be sight-threatening.

DIFFERENTIAL DIAGNOSIS

See causes listed above.

SEE ALSO

Conjunctival lymphoma; Sub-conjunctival hemorrhage (ecchymosis); Scleritis and Episcleritis; Membranous and pseudomembranous conjunctivitis; Concretions; Conjunctival scarring; Follicular conjunctivitis.

MANAGEMENT

Treatment ranges from advice for simple viral conjunctivitis through to biopsy and treatment with systemic immunosuppressive agents for ocular cicatricial pemphigoid.

C

Conjunctivochalasis and chemosis

Conjunctival chalasis, conjunctival edema

DESCRIPTION

Conjunctivochalasis refers to a chronic degenerative loosening and redundancy of the conjunctiva. The conjunctiva remains largely aligned with the ocular surface and so the subconjunctival space is not significantly expanded. Conjunctivochalasis is most commonly seen with aging of the ocular surface. However, it may also result from prolonged conjunctival edema as occurs with thyroid eye disease, ocular allergic conditions, and some orbital tumors.

Chemosis (conjunctival edema) may occur acutely in many different conditions including allergic conjunctivitis, bacterial conjunctivitis, and chemical burns. Chronic chemosis is less common and can occur with orbital tumors, thyroid eye disease, or as an idiopathic finding. Chemosis results from conjunctival inflammation with extravasation of fluid from the conjunctival blood vessels, leading to expansion of the subconjunctival space. The conjunctiva is elevated from the sclera, giving the eye a bulbous appearance.

SYMPTOMS

With conjunctivochalasis, patients may report chronic ocular surface irritation, with foreign-body sensation and increased lacrimation. In contrast, chemosis symptoms reflect the cause – for example, the symptom of itching in allergic conjunctivitis.

SIGNS

Conjunctivochalasis

The redundant inferior conjunctiva is often slightly inflamed, forming a horizontal ridge of tissue that prolapses over the inferior eyelid margin. The ridge of conjunctiva, tarsal surface of the inferior lid margin, and inferior cornea all stain moderately with fluorescein and rose bengal. Changes are usually bilateral. Occasionally sufficient redundancy occurs that the inferior conjunctiva forms a ridge of tissue rubbing on the cornea.

Chemosis

In contrast to conjunctivochalasis, the changes seen with chemosis include an edematous expansion of the subconjunctival space. If the entire conjunctiva is chemotic then the "ballooning" of the conjunctiva for 360° around the cornea gives the eye a "watch glass" appearance. The conjunctiva is frequently inflamed with associated hyperemia. Occasionally, a discharge may be present.

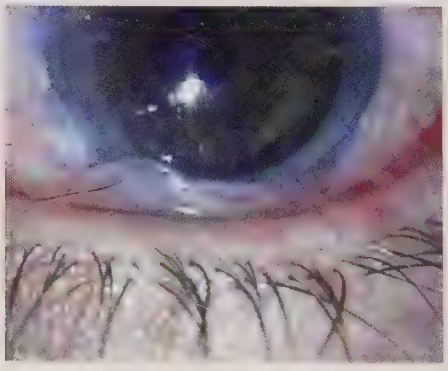


Fig. C-17a Area of conjunctivochalasis rubbing on the inferior cornea in a patient who has previously had a corneal graft.

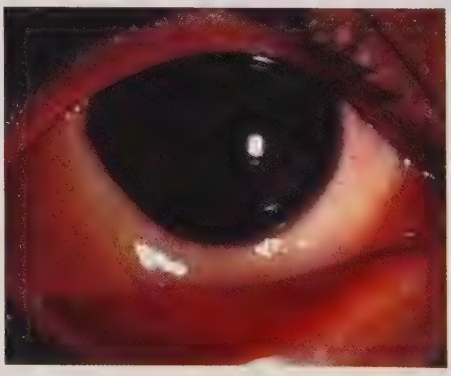


Fig. C-17b Marked conjunctival chemosis in acute allergic conjunctivitis.



INCIDENCE
These conditions can be common (approximately 1/100) in certain subgroups. Conjunctivochalasis is usually seen in older patients. Chemosis is commonly seen secondary to bacterial, viral, or allergic conjunctivitis.

SIGNIFICANCE
Both conditions can cause irritation but in isolation are not sight-threatening.

DIFFERENTIAL DIAGNOSIS
Conjunctivitis; Acute allergic blepharoconjunctivitis; Retention cyst and lymphangectasia; Atopic keratoconjunctivitis (AKC).

SEE ALSO
Superior limbic keratoconjunctivitis (SLK); lid retraction (exophthalmos/proptosis).

MANAGEMENT
Conjunctivochalasis
This condition is a chronic degenerative change within the conjunctiva and as such is largely irreversible.

TEAR SUPPLEMENTS
The frequent use of viscous tear supplements acts to lubricate the ocular surface and provide temporary symptomatic relief.

SURGERY
Surgical excision is frequently required if there are significant ocular surface symptoms. The ridge of conjunctiva and a frill of 1–2 mm of conjunctiva are excised. Usually this can be performed at the slit lamp with topical anesthetic and local infiltration. Postoperatively the patient is placed on topical antibiotics and a corticosteroid qid for 1 week, or until healing of the epithelial defect.

Chemosis
TOPICAL THERAPY
Chemosis due to allergic conjunctivitis responds well to the frequent application of cold compresses, leading to vasoconstriction. The allergen can be flushed out and the eye soothed with tear supplements (preferably unit dose) every 2 hours for 1–2 days.

THERAPEUTICS
If chemosis due to allergic conjunctivitis is moderate to severe, a combination topical antihistamine and vasoconstrictor may be prescribed: for example, naphazoline and pheniramine combination qid for 24–48 hours. An oral antihistamine such as diphenhydramine can also assist. See also comments under "Acute allergic blepharoconjunctivitis" and "Atopic keratoconjunctivitis."

C

Contact lens: gas permeable (GP)

Rigid contact lens, hard contact lens

INDICATIONS

In addition to the correction of common refractive errors and presbyopia, gas permeable (GP) contact lenses have unique benefits for the conditions described below.

Corneal irregularity

GP lenses provide correction of corneal irregularity, including conditions such as keratoconus, keratoglobus, pellucid marginal degeneration, Terrien's marginal degeneration or, after refractive surgery, corneal trauma or grafting. In such cases, rigid gas permeable (GP) lenses offer better vision than a soft lens, since the lens does not flex to the abnormal corneal contour. The tear layer posterior to the lens enables the optical correction of the corneal irregularity.

High prescriptions

High spherical or astigmatic prescriptions may be corrected with GP lenses using high oxygen transmissibility materials, when silicone–hydrogel soft lens parameters may not be available. Tear exchange and the smaller lens diameter associated with many GP lens designs provide additional modes of oxygen delivery to the cornea and also provide a reduced risk of corneal infection compared to soft lenses.



Fig. C-18a Comparison of lens diameters for (left to right) a 24-mm scleral, 10-mm rigid, and 14-mm soft lens.

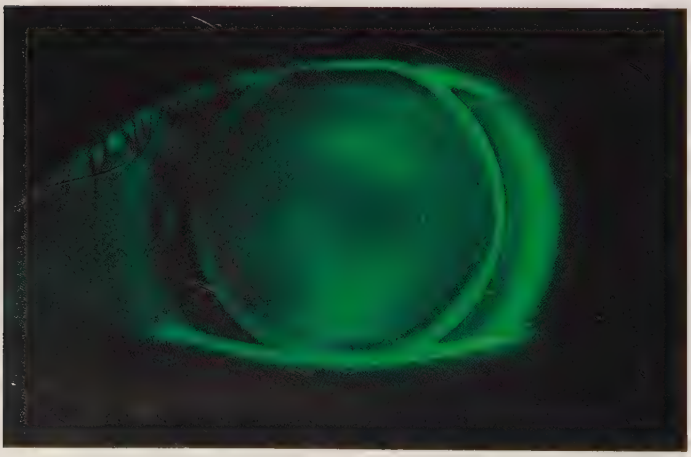


Fig. C-18b Menicon Z corneal lens fitted two-thirds toward the flatter keratometry value, showing a near-alignment fluorescein fitting pattern.



Ocular surface disease

Treatment of ocular surface disease may be assisted with GP lenses, since the lens materials are water-impermeable. Larger diameter GP lenses can offer ocular surface protection in severe keratoconjunctivitis sicca, Stevens–Johnson syndrome, ocular cicatricial pemphigoid, lagophthalmos, advanced atopic keratoconjunctivitis (AKC), and persistent epithelial defect.

Corneal reshaping

Temporary myopia reduction and corneal reshaping is possible using reverse-geometry orthokeratology GP lenses.

LENS DESIGNS AND MATERIAL TYPES

GP lenses are durable, rather than disposable, lasting on average 1–2 years. The lenses are primarily suited to full-time wear, since the initial comfort of GP lenses is less than for soft lenses, and the patient may take 1–2 weeks to adapt to wearing them.

Corneal lenses

Corneal lenses are smaller than the cornea, usually being 9–10 mm in total diameter. Lens diameter is selected to fit within the palpebral aperture or, if possible, to attach to the superior eyelid margin. Lens base curve is typically closely matched to the curvature of the central cornea, a fitting relationship that is termed "alignment." Using fluorescein and the slit lamp cobalt blue filter, there should be a relatively even glow of fluorescence from the tear layer under the lens (Fig. C-18b). Vertical lens movement with blinks is typically at least 2 mm more than occurs with a soft lens. In keratoconus the lens may exhibit "3-point touch" where there is less fluorescence in the fitting pattern centrally and also in the mid-periphery.

Mini-scleral lenses

Mini-scleral lenses are GP lenses with a diameter of 16–18 mm. They are primarily used for therapeutic applications. The lens is fitted to rest on the sclera without blanching or impinging on blood vessels. Scan the slit lamp optic section across the lens to ensure the central fitting exhibits clearance from the cornea. With corneal grafts a mini-scleral lens may approximately align with the central cornea and exhibit clearance peripherally. Mini-scleral lenses do not move vertically with blinking but may seal at the sclera or may "rock" if fitted to align centrally for a corneal graft.

Scleral lenses

Scleral lenses are large (about 25 mm, or 1 inch) diameter GP lenses (Fig. C-18a). They are primarily used for therapeutic applications when other lens types have failed.

GP lens materials

Contemporary lens materials have high oxygen permeability and deposit resistance, e.g. Boston XO (Bausch & Lomb), Menicon Z (Menicon), and HDS100 (Paragon). Oxygen-impermeable plastic (PMMA) may be used for trial lenses, but is contraindicated for normal wear.

LENS CARE

Most patients use their GP lenses for daily wear and remove them at night to store in a case. Extended wear remains relatively uncommon, as there is an increased rate of complications. While a multipurpose GP lens care solution is convenient, the separate cleaning and soaking solutions are more effective. A weekly enzymatic protein removal procedure is warranted for patients with long wearing times or lens deposits. Mini-scleral lens wearers may fill the lens with saline or a non-preserved lubricant drop prior to lens instillation.

C

Contact lens: soft

Disposable contact lenses, silicone–hydrogel contact lenses

INDICATIONS

Soft lenses are widely used for the correction of common refractive errors and presbyopia. In addition, soft contact lenses have unique benefits for the conditions described below.

Pain relief

A soft lens may be used as a bandage for pain relief. Possible causes of corneal disruption include band keratopathy, bullous keratopathy, Meesmann's dystrophy, superior limbic keratoconjunctivitis, and Thygeson's superficial punctate keratopathy. If a filamentary keratopathy is refractory to other measures, a bandage contact lens may be helpful in breaking the cycle of epithelial damage and filament formation.

Ocular surface protection

Bandage lenses may assist in the short term for trichiasis, entropion, tarsal scars, and concretions.

Promote epithelial healing

A bandage lens fitted over an abraded or denuded cornea provides a barrier to tear film flow into the corneal stroma, thereby reducing edema. In addition, a bandage lens reduces the shearing action of the eyelids on the epithelium, allowing the epithelium to slide over, and adhere to the basement membrane, in cases of persistent epithelial defect (PED), neurotrophic keratitis, chemical burns, or recurrent corneal erosion.

Post-surgical

A lens may provide pain relief and promote epithelial healing after refractive surgery or corneal surgery.

Opaque tinted

A conventional soft lens with opaque coloring may be used for prosthetic purposes to mask corneal scarring, ocular disfigurement, or aniridia. Visual indications may include persistent uncorrectable diplopia or polyopia from ocular media anomalies or binocular vision problems.

LENS DESIGNS AND MATERIAL TYPES

Soft lens materials

Modern soft lens materials include silicone, which gives lenses a high oxygen transmissibility (Dk/T > 25). The older hydrogel materials (low to moderate Dk/T) are perhaps more suited to part-time or limited wear. All soft lens materials are characteristically porous and water absorbent, making the lenses comfortable.

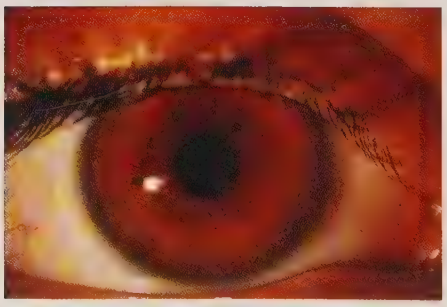
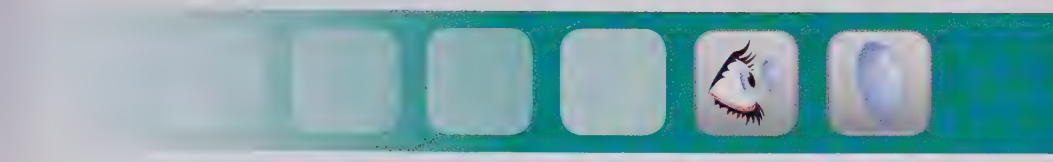


Fig. C-19a Disposable contact lens of 14-mm diameter on eye.



Fig. C-19b Cosmetic opaque contact lens for disfigured eye (inset).



Disposable lenses

Disposable soft lenses are supplied in multiple blister packs. Lenses are usually discarded fortnightly or monthly, depending upon manufacturer recommendation and patient factors, to minimize complications related to lens deposits. On lens removal, they are cleaned and disinfected with a multipurpose solution.

Daily disposable lenses

Daily disposables are single-use lenses, supplied in packs of 30 or 90 lenses. They tend to be of a thin design in high water-content hydrogel material (moderate Dk/T, 12–24). No cleaning or disinfection system is required, as the lenses are not reused.

Conventional soft lenses

Conventional (non-disposable) soft lenses are a largely obsolete modality, due to more complex maintenance requirements and a higher rate of complications.

Extended wear lenses

In extended wear the lenses are left in at night and removed weekly or monthly. While most silicone–hydrogel lenses have adequate oxygen transmissibility for extended wear, there remains an increased rate of infective and inflammatory complications compared to daily wear.

Bandage lenses

A bandage contact lens may be a plano-powered silicone–hydrogel lens. Examples of lenses that may be used include Acuvue Oasys (Johnson & Johnson), AirOptix Night & Day (CibaVision), and Purevision (Bausch & Lomb). Bandage lens treatment is typically used for short periods (e.g. from 1 week to 1 month), since there is a slightly increased risk of microbial keratitis. If there is an epithelial defect, then prophylactic topical antibiotics are usually used concurrently, either as preservative-free drops or instilled when the lens is not in the eye to avoid preservative accumulation in the lens and toxicity reactions. The epithelial defect must be "clean" (i.e. not infected, inflamed, contaminated with foreign material, or related to a prior contact lens condition). Corticosteroid and non-steroidal anti-inflammatory drops are usually contraindicated during bandage contact lens treatment.

Prosthetic opaque lenses

Conventional lenses may be custom colored to match the patient's other eye color, with the option of an opaque black pupil for unsighted eyes. Milder anomalies may be able to be masked with opaque-colored disposable contact lenses. Special care and maintenance may be required for prosthetic contact lenses, often with a hydrogen peroxide care system. Regular aftercare is required, as for all conventional soft lenses.

C

Contact lens-related conditions

Acute red eye, solution hypersensitivity, solution toxicity, vascularization

Modern contact lenses provide safe comfortable visual correction for the vast majority of wearers. However, some inflammatory complications uncommonly occur in contact lens wear.

In addition, contact lens wear may be an additional risk factor for some general eye conditions. Such conditions covered in this book include:

- abrasion
- *Acanthamoeba* keratitis
- bacterial keratitis
- corneal infiltrates
- exposure keratopathy – tear film instability on the surface of the lens, causing desiccation
- fungal keratitis
- giant papillary conjunctivitis (GPC)
- marginal keratitis (sterile peripheral ulcer)
- superficial punctate keratopathy (SPK)
- superior limbic keratoconjunctivitis.

CONTACT LENS ACUTE RED EYE (CLARE)

Contact lens-induced acute red eye (CLARE) is a unilateral ocular inflammatory reaction, generally restricted to those patients wearing soft lenses in extended wear. The infiltrates are typically located within 3 mm of the dilated blood vessels of the limbus. However, the overlying epithelium is usually intact, showing minimal staining. Ocular pain and acute bulbar hyperemia are other hallmarks. In some instances the patient will be awakened at 2 or 3 am due to ocular pain. However, there is no mucopurulent discharge and usually no anterior chamber reaction. Bacteria causing CLARE are adherent to the contact lens, rather than colonizing the ocular surface or eyelids, which is consistent with the observation that symptoms rapidly subside once lenses are removed. Both Gram-negative *Hemophilus influenzae* bacteria and Gram-positive *Streptococcus pneumoniae* have been isolated from the lenses of patients with CLARE reactions in extended wear.

SOLUTION HYPERSENSITIVITY AND TOXICITY

Fine scattered infiltrates within the epithelium are an infrequent response to solution preservatives. Infiltrates are usually accompanied by marked conjunctival hyperemia and epithelial punctate staining.

Small intraepithelial infiltrates occur early in the inflammatory response, with larger subepithelial infiltrates developing if the condition is chronic. The origin of intraepithelial infiltration is attributed to the preservatives in the tear film. The presence of infiltrates is diagnostic of a hypersensitivity reaction, whereas a toxicity

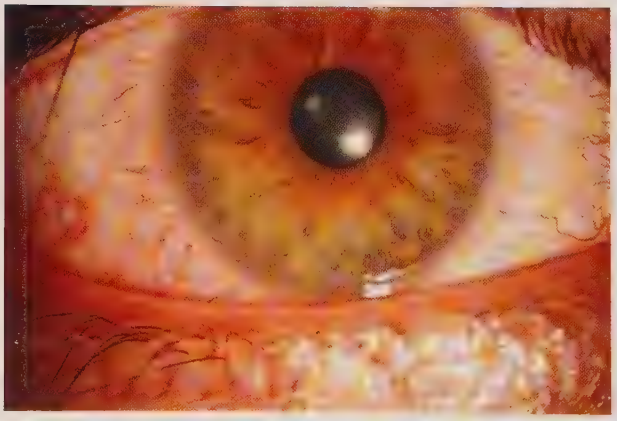


Fig. C-20 Mild infiltrative keratitis related to contact lens solution hypersensitivity.



reaction is a direct chemical effect causing superficial punctate keratopathy.

VASCULARIZATION

Vascularization of the cornea is often defined as blood vessels extending more than 1.0–1.5 mm from the limbus. It is significant because it is indicative of chronic edema (see "Corneal edema") and for the risk of corneal opacification (see "Lipid keratopathy"). In contact lens wear, vascularization is most commonly a sign of chronic hypoxia. It is rare with modern high oxygen transmissibility contact lenses.

SYMPTOMS

Hypoxic effects may be asymptomatic. Allergic and toxic effects may cause stinging or itching, usually bilateral. Infection may cause pain or discharge, usually unilateral.

INCIDENCE

Contact lens complications range from very common (greater than 1/10; e.g. punctate epithelial erosions) to very rare (less than 1/100,000; e.g. *Acanthamoeba* keratitis).

SIGNIFICANCE

Discomfort: mild to moderate; threat to sight: low to moderate.

DIFFERENTIAL DIAGNOSIS

Conjunctivitis; Glaucoma – acute angle closure; Adenoviral keratoconjunctivitis, where the infiltrates are anterior stroma.

MANAGEMENT

Acute inflammatory reactions

In acute red eye (CLARE) or solution hypersensitivity, contact lenses should be immediately removed and disposed of. Most symptoms resolve within 2 days; however, the infiltrates generally remain for at least a week. A mild steroid (e.g. fluorometholone qid) will hasten symptomatic relief. If there is a significant overlying epithelial defect or other suspicious signs, treatment for infectious keratitis should be considered (see appropriate condition).

Prevention

To minimize the risk of inflammation or infection, potential sources of bacterial contamination, such as hands and eyelids, should be kept as clean as possible and lenses, case, and solutions replaced regularly. Contact lens wearers with an upper respiratory tract infection should discontinue wear until it resolves. Depending upon the severity of the reaction, extended wear may need to be curtailed. If the lens care multipurpose solution is suspected, change the care regimen to a hydrogen peroxide-based system.

Vascularization

The lenses should be refitted to increase oxygen availability. If the lens is already optimized, reduce wearing time. Review is an essential part of management as many hypoxic signs are asymptomatic.

C Corneal collagen cross-linking (CXL)

DESCRIPTION

Corneal collagen cross-linking is a chemical method of stiffening the cornea mainly used to halt progression of corneal ectatic diseases such as keratoconus. It was first introduced into clinical practice in the early 2000s after initial experimental development in the late 1990s in Germany. The corneal epithelium is usually removed, the corneal stroma is saturated for approximately 15–30 minutes using riboflavin-5-phosphate (vitamin B2) drops, and the cornea irradiated for 30 minutes with ultraviolet light (UVA). This results in the riboflavin becoming chemically unstable. It becomes stable again upon binding to the corneal collagen, forming chemical cross-links in the anterior one-third to two-thirds of the corneal stroma leading to a multiple-fold increase in corneal stiffness. The process also leads to the generation of oxygen free radicals resulting in the death of keratocytes in these layers. It has been found not only to halt progression of keratoconus but also frequently results in some flattening of the cornea with an increase in best spectacle-corrected visual acuity. Occasionally CXL is combined with implantable corneal ring-segments aiming to further flatten an ectatic cornea.

Faster, less toxic methods of cross-linking the corneal collagen using alternative cross-linking agents are actively being developed and are likely to become clinically available within the next several years.

INDICATIONS

CXL is an evolving treatment with the indications for treatment, treatment protocols, and outcome measures still actively being developed. Only now are randomized controlled trials starting to be published.

CXL is mainly used for treating progressive keratoconus; therefore, it is mainly used in patients in their mid to late teens and into their 20s. It has also been used for treating patients with other forms of progressive corneal ectasia, such as post-LASIK keratoectasia, pellucid corneal marginal degeneration, and keratoglobus, and provides a means of stiffening the host cornea prior to corneal transplantation. It may also rarely be used in an attempt to flatten the cornea to allow a patient to become more contact lens tolerant, thereby avoiding the need



Fig. C-21a Bisected human cornea that has been soaked in riboflavin.



Fig. C-21b Other half of bisected human cornea from Figure C-20a that has been soaked in riboflavin and cross-linked with exposure to UV light. Note that the cornea is significantly stiffened.



for a corneal graft. In addition, it has been used for the treatment of painful bullous keratopathy and non-responsive microbial keratitis, especially fungal keratitis, where the generation of free radicals results in sterilization of the anterior cornea.

The definition of progressive corneal ectasia can be based on a number of grounds: steepening cornea curvature, progressive corneal thinning, increasing refractive astigmatism, decreasing best spectacle-corrected acuity, and progressively steepening rigid contact lens fit. Due to the potential for significant test-to-test variation, a corneal steepening on the maximum or mean keratometry reading of at least 1.2 diopters is usually required to indicate a true progression of the disease.

To be amenable for treatment, the cornea needs to be clear and of adequate thickness, preferably at least 400 microns with the epithelium on. The concern is that cross-linking of a very thin cornea could result in free radical damage to the corneal endothelium.

SYMPTOMS

Following the cross-linking procedure patients usually experience several days of discomfort due to pain secondary to the epithelial defect. This is often reduced if a bandage soft contact lens (BSCL) is applied immediately postoperatively. The BSCL is usually removed once the epithelium is fully healed; this usually takes 3–4 days. It is common for patients to be mildly light-sensitive for up to several weeks. They can usually resume contact lens usage 1–2 weeks after the procedure.

SIGNS

A very mild, non-visually significant central haze is typically seen in the anterior stroma from the time of the treatment, peaking at around 3 months, and often fully resolved by 6–12 months. This is taken to be an indicator of the depth of cross-linking.

THERAPEUTICS

Immediately following the CXL procedure the patient typically uses a topical antibiotic, such as chloramphenicol or ciprofloxacin, and a topical corticosteroid, such as fluorometholone acetate qid. The antibiotic is ceased when the epithelium is healed, and the BSCL removed. The corticosteroid is used qid for 1 week then bd for 3 weeks then ceased.

MONITORING OF PATIENTS

Patients are followed with serial corneal topographic maps both preoperatively to assess for progression and postoperatively to assess the effect of the treatment. Corneal topography is preferably performed with an instrument such as an Orbscan or Pentacam capable of measuring the curvatures of both the anterior and the posterior corneal surfaces as well as topographic pachymetry.

To assess the need for CXL, patients with keratoconus disease are usually followed every 6 to 9 months. If they clearly have stable keratoconus, and are in their late 20s to 30s, it is reasonable to cease regular screening.

Following CXL patients usually have a 3-month, 6-month, and then 6- to 12-monthly maps performed for several years.

SEE ALSO

Keratoconus; Pellucid marginal corneal degeneration; Keratoglobus.

C

Corneal dystrophies, degenerations, and depositions – overview

DESCRIPTION

Corneal degenerations, dystrophies, and depositions are common types of corneal opacifying disorders. Various conditions are covered individually in this text.

Dystrophies

Corneal dystrophies are non-inflammatory, progressive, usually bilateral, and mostly inherited disorders. The genetics of each dystrophy are discussed in the individual article. They tend to affect the central cornea, be avascular, and have an onset early in life. The mode of inheritance is usually autosomal dominant, although inheritance of keratoconus is not so clearly defined.

- Anterior: epithelial basement membrane dystrophy (Cogan's, map-dot fingerprint); Meesmann's epithelial dystrophy; Reis–Bücklers dystrophy
- Stromal: granular stromal dystrophy; lattice stromal dystrophy; macular stromal dystrophy
- Posterior: Fuchs' endothelial dystrophy; posterior polymorphous dystrophy; congenital hereditary endothelial dystrophy (CHED, not covered in this text)
- Ectatic: keratoconus and keratoglobus; pellucid marginal degeneration

Degenerations

Corneal degenerations may be unilateral or bilateral, may be asymmetric, are most often located peripherally, and tend to onset later in life. The degenerations tend to occur in relation to processes such as aging, trauma, or disease. Degenerations related to inflammation can be vascularized.

- Age-related: corneal opacities – peripheral (arcus senilis, Vogt's white limbal girdle); crocodile shagreen and corneal mosaics; Terrien's marginal degeneration
- Inflammation-related: band keratopathy; Salzmann nodular degeneration; lipid keratopathy; pseudogerontoxon in vernal disease (see "Corneal opacities – peripheral")
- UV radiation and exposure-related: climatic droplet keratopathy (CDK; spheroidal degeneration); pterygium,

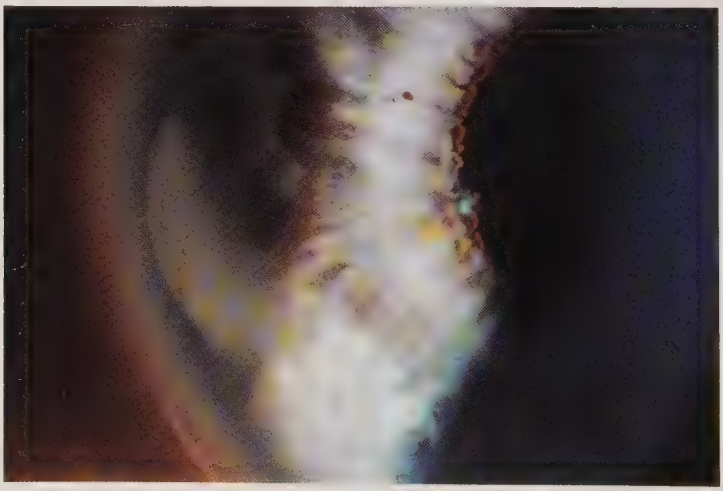
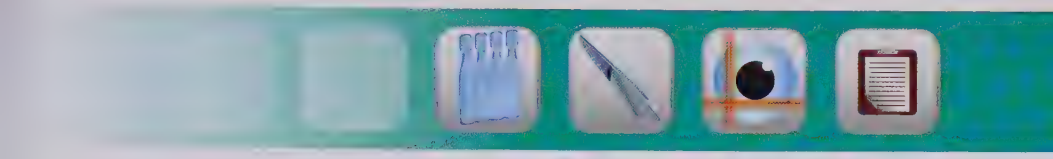


Fig. C-22 Early band keratopathy, a degeneration due to the deposition of calcium salts at the level of Bowman's membrane.



although a conjunctival degeneration is manifest at the corneal periphery

Depositions
- Corneal opacities – peripheral (Kayser–Fleischer ring; Wilson disease; copper deposits)
- Vortex keratopathy (chloroquine, amiodarone, Fabry)

SYMPTOMS
Dystrophies
Some of the corneal dystrophies are associated with acute painful episodes due to recurrent erosions. Fuchs' dystrophy is associated with glare and blurred vision, particularly on awakening.

Degenerations and depositions
Some of the corneal degenerations can be associated with significant symptoms of ocular discomfort and effect on vision. The age-related degenerations such as arcus and shagreen tend to be asymptomatic.

SIGNS
The principal sign of a corneal dystrophy or degeneration is the development of opacity in any layer of the cornea. A dystrophy will tend to affect the central cornea and have an earlier onset in life, whereas degeneration more frequently affects the peripheral cornea and has an onset in middle age or later.

Despite the possible appearance of an opacity that may resemble corneal infiltration, there is an absence of associated signs of infection including conjunctival hyperemia, anterior chamber reaction, and discharge. The epithelial surface is usually intact with no fluorescein staining.

INCIDENCE
Dystrophies
Uncommon to rare (approximately 1/1000–1/10,000) across all age groups.

Degenerations
Very common (greater than 1/10) in the older age groups.

SIGNIFICANCE
Dystrophies and degenerations are more likely to cause ocular discomfort than vision loss. Depositions may have systemic implications.

DIFFERENTIAL DIAGNOSIS
See the conditions under "Description"; corneal scar.

SEE ALSO
Pterygium; Pannus; Corneal opacities – peripheral.

MANAGEMENT
Most of these conditions are chronic, and progressive. While to do nothing and review is an option, management or treatment at some point in time is likely to be required, sooner for some conditions than others. For the conditions related to UV, general advice about the use of sunglasses is helpful.

Tear supplements
Topical lubrication can be helpful for ocular surface irritation.

Surgery
Either incisional, or laser surgery may be indicated. Some of the more significant dystrophies may require corneal grafting.

C

Corneal edema – overview

Endothelial decompensation, microcystic epithelial edema

DESCRIPTION

The corneal stroma is composed of lamellar sheets of collagen interspersed with abundant osmotically active mucopolysaccharides. Without the pumping action of the endothelium, and to a lesser extent the epithelium, the cornea swells, and the normal orientation of the lamellar collagen is lost, leading to decreased corneal clarity. Edema of the epithelium increases forward light-scatter and has a greater effect on vision than stromal edema.

CAUSES

Corneal edema is most often a result of endothelial cell dysfunction although it may also occur from epithelial and stromal disease. In addition, it may result from breaks in Descemet's membrane as occurs in corneal hydrops.

Endothelial cell dysfunction occurs for a number of reasons, as discussed below.

Hereditary

Endothelial dystrophies such as Fuchs' endothelial dystrophy, congenital hereditary endothelial dystrophy (CHED), and posterior polymorphous dystrophy (PPD).

Inflammatory

Herpes simplex and herpes zoster keratouveitis can both lead to significant endothelial dysfunction and corneal edema.

Post-surgical

Most commonly, cataract surgery leading to bullous keratopathy – aphakic (ABK) or pseudophakic (PBK).

Intraocular pressure

A very high intraocular pressure (>50 mmHg) as seen with acute angle closure glaucoma typically leads to microcystic epithelial edema, while a low pressure (<5 mmHg) typically leads to deep stromal edema.

SYMPTOMS

Corneal edema leads to blurring of vision, increased glare, photophobia, and halos around light sources. Microcystic epithelial edema also leads to a gritty foreign-body sensation. Rupture of an epithelial bullous leads to an acute painful episode that typically lasts several days.

SIGNS

Epithelial edema

Accumulation of fluid both within and underneath the epithelium leads to microcystic epithelial edema and bullae. Microcystic edema gives the epithelium a roughened uneven appearance, while bullae appear as raised areas of epithelium with negative fluorescein staining.

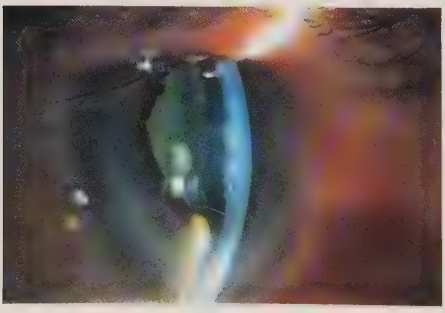


Fig. C-23a Stromal edema with folds in Descemet's membrane.

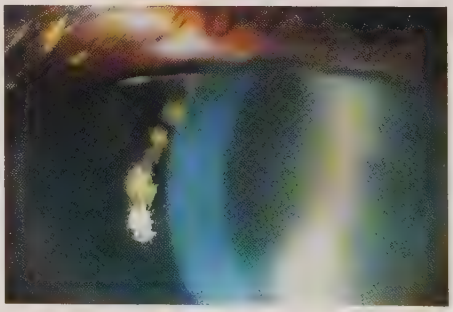


Fig. C-23b Microcystic corneal epithelial edema.



Stromal edema
Accumulation of fluid in the deep stroma leads to characteristic folds in Descemet's membrane and generalized opacification of the stroma.

Associated signs
Signs that can help to identify the cause of the edema are evidence of previous intraocular surgery, endothelial guttae, PPD lesions, intraocular inflammation with keratic precipitates, and an abnormal intraocular pressure.

SIGNIFICANCE
Long-standing corneal edema can have a severe effect on ocular comfort or vision. It is often associated with cystoid macular edema (CMO) and typically leads to a painful red eye. It is not uncommon for a ruptured bullous to lead to microbial keratitis, and as such threaten survival of the eye.

DIFFERENTIAL DIAGNOSIS
See causes listed above.

SEE ALSO
Corneal graft rejection; Descemet's membrane breaks (hydrops); Disciform keratitis; Fuchs' endothelial dystrophy; Glaucoma; Bullous keratopathy – pseudophakic and aphakic.

MANAGEMENT
The underlying cause of the edema should be treated; this is especially the case with disciform keratitis. The most common causes of gross corneal edema are post-surgical or dystrophies. In such cases irreversible endothelial dysfunction may be treated with the measures discussed below.

Tear supplements
Lubricants can reduce the grittiness and foreign-body sensation of microcystic epithelial edema.

Therapeutics
Hypertonic (5%) saline drops and/or ointment can be used to osmotically desiccate the corneal epithelium; they have no action on stromal edema.

Therapeutic contact lens
The temporary use of a bandage contact lens is usually useful in reducing symptoms but does increase the risk of microbial keratitis in these damaged eyes, which are predisposed to epithelial defects.

Surgery
Although a corneal graft is the most common surgical procedure for irreversible edema, corneal micropuncture and a conjunctival flap are also sometimes used to stabilize the ocular surface.

Laser surgery
In a painful eye with poor visual potential a deep phototherapeutic keratectomy (PTK) may render the eye pain-free, and stabilize the ocular surface.

C

Corneal graft

Penetrating keratoplasty (PK, PKP), lamellar grafts

DESCRIPTION

Although the most common form of corneal grafting is replacement of the central cornea with a 7.5–8-mm diameter full thickness allograft, many variations exist. These include larger grafts, partial thickness grafts (also called lamellar grafts; e.g. deep anterior lamellar keratoplasty [DALK] and Descemet's stripping automated endothelial keratoplasty [DSAEK]), peripheral grafts, and grafts of different sizes and shapes (patch grafts). Over the past decade many variations of lamellar grafts have been developed; over this period lamellar grafts have increased from a very small percentage to a significant percentage at present of corneal grafts performed in developed nations. Almost all grafts (except DSAEK) are sutured into position with 10/0 nylon with interrupted sutures (most commonly 16), a continuous-running suture, or with a combination of both. These sutures are usually deep partial corneal thickness, not full thickness.

INDICATIONS

There are three main indications for a corneal graft. These are to:

- improve vision (visual)
- reduce pain (therapeutic)
- repair a hole in the cornea (tectonic).

The two minor (and usually secondary) indications are to improve appearance (cosmetic) and to establish a diagnosis (diagnostic). Overall, the most common indications for a corneal graft in developed countries are keratoconus, pseudophakic bullous keratopathy (PBK), Fuchs' dystrophy, and replacement of a failed graft.

SYMPTOMS

After the initial postoperative period there is usually no irritation or discomfort associated with the presence of a corneal graft. A loose suture can lead to a foreign-body sensation. Both regular and irregular astigmatism are common.

SIGNS

Aside from the size and suturing style, the graft should be assessed for clarity and compactness and any loosening of the sutures. The anterior chamber should be assessed for any sign of rejection, the intraocular pressure recorded, and the refractive error noted.

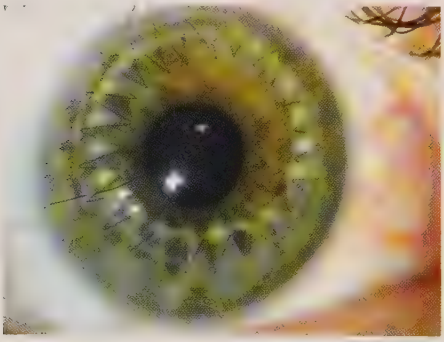


Fig. C-24a A central penetrating corneal graft with a 24-bite running suture in place. This suturing style is commonly used with keratoconus.

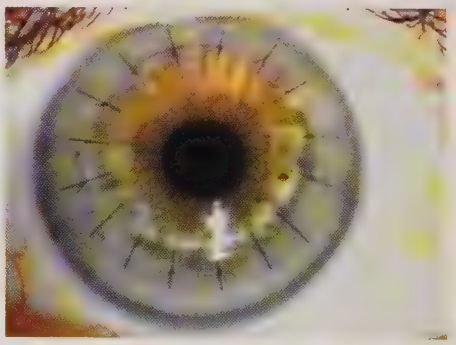


Fig. C-24b A central penetrating corneal graft with 16 interrupted sutures in place. This suturing style is used for edematous or vascularized corneas where the risk of postoperative loosening of sutures is more common.



INCIDENCE

In developed countries, the rate of corneal grafts performed varies widely, but is commonly between 50 and 100 per million people per year. The rate of corneal grafts has fallen steadily in many developed countries over the past 2 decades.

SIGNIFICANCE

The presence of a full-thickness corneal graft or DSAEK indicates there is a risk of endothelial rejection. As such, any symptoms or signs of intraocular inflammation need to be assessed carefully.

SEE ALSO

Corneal graft – lamellar keratoplasty; Corneal edema; Keratoconus; Fuchs' endothelial dystrophy; Corneal graft rejection; Bullous keratopathy – pseudophakic and aphakic; Cataract incisions; Refractive surgery signs.

MANAGEMENT

Advice

An eye with a corneal graft, with the exception of posterior lamellar grafts such as DSAEK, is always at an increased risk of significant damage from ocular trauma. Even a moderate injury can result in rupture of the graft–host junction, especially in the early postoperative period before the graft has healed into position. As such, patients should be advised of the need for protective eyewear in high-risk situations, and the need to avoid heavy contact sports.

Therapeutics

Postoperative topical corticosteroids are essential to decrease the risk of graft rejection. In general, some topical corticosteroids should be used until at least 1 week following removal of all sutures. A topical antibiotic and corticosteroid are used qid for 1 week immediately following suture removal to reduce the risk of rejection and microbial keratitis.

In high-risk grafts, regrafts, or following a rejection episode, one drop of a topical corticosteroid may be used indefinitely.

Surgery

With penetrating grafts the donor tissue is usually 0.25 to 0.5 mm greater in diameter than the host trephination. A common combination is an 8.00-mm diameter graft transplanted into a 7.75-mm trephined host cornea. While both the donor and the host are routinely cut with round surgical blade trephines, several investigators are trialing use of the femtosecond YAG laser to perform the incisions. Sutures are removed when the graft has healed into position. The timing of suture removal is very dependent on the age of the patient. Younger patients in their 20s and 30s usually heal in the graft within 6–12 months while patients in their 70s and 80s may take 2 years or more. As sutures can loosen and break as they degrade with time, they are usually all removed by 2 years following surgery. Sutures are also removed if they break or loosen, or sometimes to control graft astigmatism if interrupted sutures have been used.

Contact lenses

Rigid lenses are sometimes required to achieve good vision following grafting due to postoperative astigmatism. Lenses are not usually fitted until all the sutures are removed.

C

Corneal graft – lamellar keratoplasty

Lamellar grafts, DSAEK, DSEK, DMEK, DALK

DESCRIPTION

Lamellar keratoplasty (LK) involves transplantation of only a partial thickness of the cornea. LKs have undergone a significant renaissance over the past decade due to much improved surgical techniques, although the final best corrected visual acuity gained with lamellar grafts of all forms is often one to two lines worse than that for a traditional penetrating keratoplasty (PKP), full thickness graft. There are two main types of lamellar grafts, anterior and posterior.

Anterior grafts, such as a deep anterior lamellar keratoplasty (DALK), involve transplanting the corneal stroma and epithelium while leaving the host Descemet's membrane plus or minus some deep stroma. The advantage of these techniques over a PKP is the preservation of the host endothelium, thereby preventing the risk of endothelial allograft rejection, although overall survival rates for DALK and PKP appear to be very similar. Some clinicians also feel that the graft–host junction with a DALK is less likely to be ruptured following trauma.

Posterior grafts involve transplanting a lenticule of donor Descemet's membrane alone (Descemet's membrane endotheliokeratoplasy [DMEK]), or Descemet's membrane and deep stroma (Descemet's stripping automated endothelial keratoplasty [DSAEK], and Descemet's stripping endothelial keratoplasty [DSEK]) after first stripping off the host Descemet's membrane, leaving the epithelium and the corneal stroma. The advantage of these techniques over a PKP is the removal of the need for corneal suturing and faster visual rehabilitation and less induced astigmatism, although the rate of primary graft failure appears to be significantly higher than for PKPs.

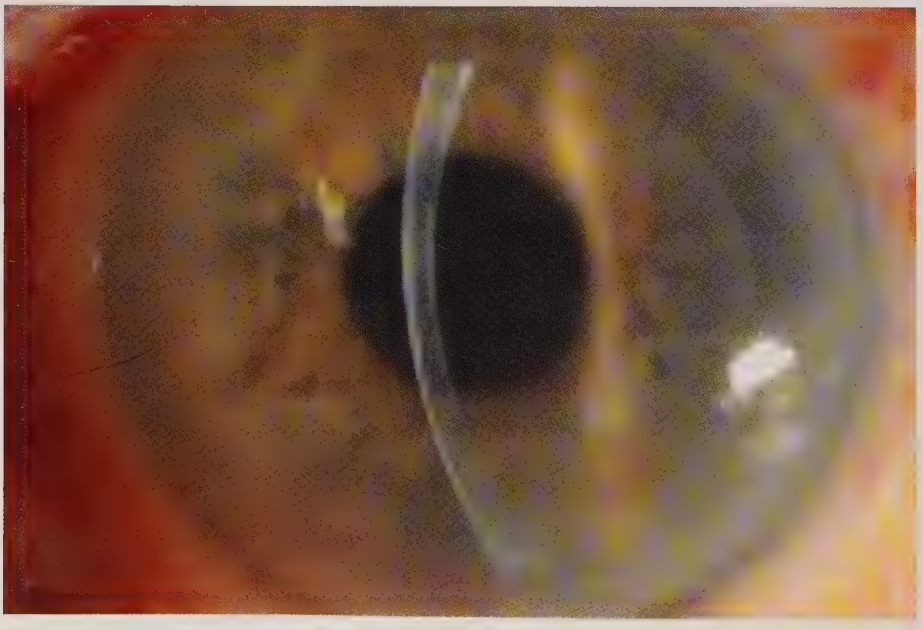
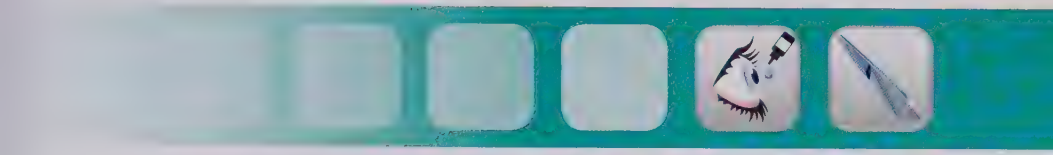


Fig. C-25 DSAEK graft in a patient with Fuchs' endothelial dystrophy.



INDICATIONS

Anterior grafts are most commonly used to treat keratoconus and stromal dystrophies. Posterior grafts are used to treat endothelial failure, most commonly Fuchs' corneal endothelial dystrophy, and pseudophakic bullous keratopathy.

SYMPTOMS

After the initial postoperative period there is usually no irritation or discomfort associated with the presence of a corneal graft. A loose suture can lead to a foreign-body sensation.

SIGNS

An anterior graft has an appearance very similar to a penetrating graft; often a DALK can be almost indistinguishable from a PKP even on careful slit-lamp examination. The suturing of the two grafts is also identical.

A posterior lamellar graft is easily distinguished from a PKP as it is inserted into the anterior segment through a limbal or scleral wound and, therefore, there are no sutures securing it to the host cornea. A DSAEK/DSEK adds another approximately 8-mm diameter lenticule of 100–250 microns thickness of donor stroma to the overall thickness of the cornea. Therefore, they are easily identified; on pachymetry a clear compact cornea may be 750–900 microns thick, the normal cornea being 500–600 microns. This may lead to an artifactual rise in the measured intraocular pressure.

INCIDENCE

Corneal grafts are performed in most developed countries at a rate of around 50–100 grafts per million of population per year. Lamellar grafts are now approaching 30–35% of all grafts performed in many such countries.

SIGNIFICANCE

As with penetrating grafts, posterior lamellar grafts are at risk of endothelial rejection. Similar degrees of astigmatism occur in both PKPs and anterior lamellar grafts. Lamellar grafts frequently have a best correct vision of 1 to 2 lines of acuity less than that of penetrating grafts.

Another relatively common complication of posterior lamellar grafts is spontaneous dislocation of the donor lenticule into the anterior chamber. If this occurs, surgical intervention is required to reattach the graft by injection of an air bubble into the anterior chamber.

SEE ALSO

Corneal graft; Keratoconus; Keratoglobus; Fuchs' endothelial dystrophy; Corneal graft rejection.

MANAGEMENT

Advice

An eye with an anterior lamellar graft is probably at as high a risk of significant damage from ocular trauma as a penetrating graft. Even a moderate injury can result in rupture of the graft–host junction, especially in the early postoperative period before the graft has healed into position. Therefore, patients should be advised of the need for protective eyewear in high-risk situations and the need to avoid heavy contact sports.

Similarly, a posterior lamellar graft is at risk of graft rejection, and any signs of increased intraocular inflammation or corneal edema need to be assessed promptly for the possibility of graft rejection.

Therapeutics

Postoperative topical corticosteroids are essential to decrease the risk of graft rejection. Both anterior and posterior lamellar grafts receive a topical corticosteroid such as prednisolone acetate qid for 2 months post-surgery followed by a slow tapering to once daily by 6–9 months following the graft, and then once daily for at least 1, often 2–3, years.

C

Corneal graft rejection

Allograft rejection

DESCRIPTION

Clarity of a corneal transplant is dependent on the continued function of transplanted endothelial cells to pump fluid from the corneal stroma against the osmotic action of stromal mucopolysaccharides. As the transplanted tissue is foreign to the host, these cells may be attacked by the host immune system. Such an episode is termed "graft rejection." If the damage is to the point where insufficient endothelial cell function remains for the graft to ever be cleared, this is termed "graft failure," or a "rejected graft."

Rejection can occur at any time following surgery but is uncommon in the first 2 months. It is much more common in certain situations, such as when there have been previous corneal grafts that have failed or when there has been previous inflammation or infection.

Aside from rejection being directed against the endothelial cells, it can also be directed against the epithelium, when it is termed "epithelial rejection," or the stromal keratocytes, "stromal rejection." As both these cell populations are self-renewing, these forms of rejection by themselves are not a risk to the continued clarity of the graft.

SYMPTOMS

Frequently, the first symptom is photophobia, followed by a red eye with associated mild ache and possible blurring of the vision. Occasionally, rejection can occur without the patient noticing any symptoms.

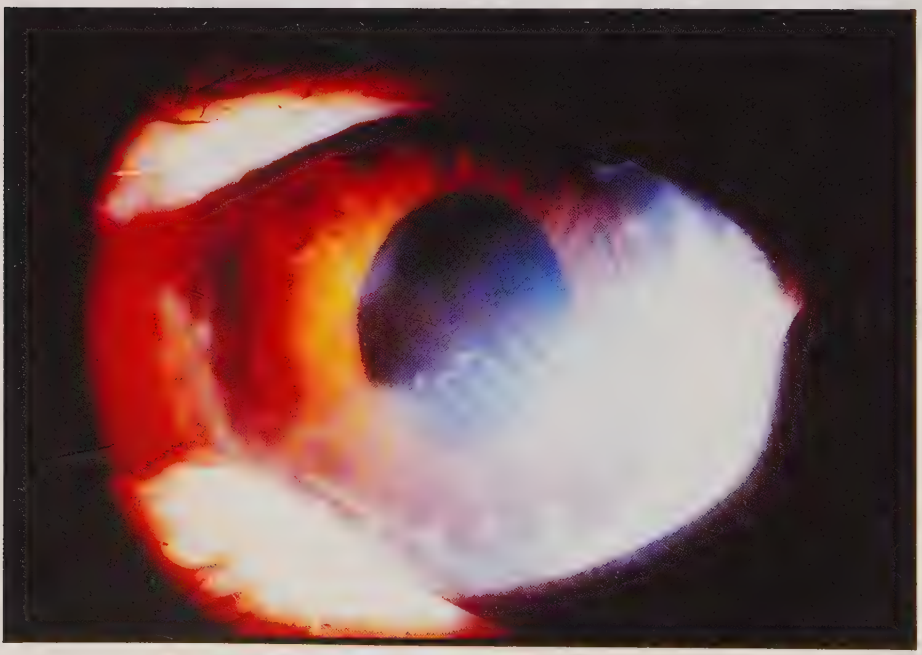
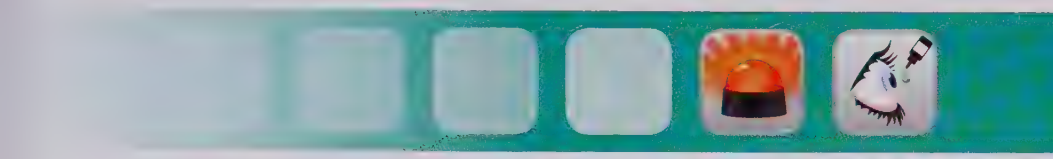


Fig. C-26 Acute corneal graft rejection. Note the loss of graft clarity inferiorly, secondary to corneal stromal edema, whereas the superior cornea is still quite clear. At the junction of the clear and edematous cornea there is a line of keratic precipitates, a Khodadoust rejection line.



SIGNS

Depending on how established and aggressive the rejection episode is, intraocular inflammation as manifested by anterior chamber cell and flare may vary from very mild to marked. Keratic precipitates are almost always present, and may be grouped or generalized. If in a line, this is termed a rejection or Khodadoust line. There is also often a variable degree of corneal edema that may also be sectorial.

INCIDENCE

Depending on the associated risk factors for the graft, as few as 3% (for keratoconus) and as many as 75% (for active infection at the time of graft) fail within the first 5 years following surgery.

SIGNIFICANCE

Graft rejection represents an acute risk to long-term graft survival and as such is an emergency. Prompt treatment is required to limit damage to the graft and to maximize the chance of restoring graft clarity.

DIFFERENTIAL DIAGNOSIS

Disciform keratitis; Iritis.

SEE ALSO

Corneal graft; Corneal edema; Bullous keratopathy; Fuchs' endothelial dystrophy.

MANAGEMENT

Prevention

Topical corticosteroids are used prophylactically to reduce the risk of graft rejection until all sutures are removed. Any manipulation of the graft or increased inflammation in an eye with a graft increases the risk of a rejection episode. Whenever any pro-inflammatory event occurs, such as with suture removal, suture manipulation, or cataract surgery, moderate-strength topical corticosteroids should be used prophylactically, usually qid for 1 week. The survival rate of grafts performed for herpes simplex keratitis is significantly increased with the prophylactic use of long-term low-dose oral acyclovir.

Therapeutics

Initial treatment of an episode of graft rejection is with intensive topical steroids such as prednisolone. Some clinicians supplement this with subconjunctival or oral corticosteroids. After a response is noted with decreased intraocular inflammation, the steroid dosage can be slowly tapered over several weeks to 2 to 3 drops a day. Some topical corticosteroids are then maintained for at least 6–12 months, and in a high-risk graft may even be used indefinitely.

Dosage: initial treatment is with G. prednisolone acetate 1% (or a topical corticosteroid of similar strength such as dexamethasone 0.1%) q1h day and night for several days. After a clinical response is observed with a reduction in anterior chamber activity and KP, the steroid can then be reduced to q2h by day, q4h by night for several days and then qid.

C

Corneal infiltrates – overview

Keratitis, contact lens-associated red eye, microbial keratitis

DESCRIPTION

Corneal infiltrates are focal areas of corneal stromal inflammation composed of accumulations of white blood cells (predominantly neutrophils), cellular debris, and protein with associated edema. If persistent, they may lead to stromal tissue loss (keratolysis). Although they may be seen in some non-infectious diseases, they are of particular concern because they may be the main presenting sign in microbial keratitis.

Infectious

Microbial keratitis, where bacteria, viruses, fungi, or protozoa have infected the cornea, leads to an inflammatory reaction characterized by corneal infiltrates, usually with an overlying epithelial defect. The inflammation often extends to the anterior chamber with focal keratic precipitates around the site of keratitis, aqueous cell and flare, and even hypopyon formation and posterior synechia.

Non-infectious (antigen sensitivity)

A focal sterile inflammatory reaction can occur to either microbial or non-microbial antigens. They are usually peripheral, less dense, and smaller than infiltrates from infectious causes, and the overlying epithelium may be intact. Infiltrates can be associated with soft contact lens wear, especially with poor lens care, or extended wear. People with posterior blepharitis can also develop corneal infiltrates adjacent to the lid margin (marginal keratitis).

SYMPTOMS

Secondary to any significant corneal inflammation, the eye becomes red and sore, photophobic, and often the vision is blurred. There is also increased lacrimation.

SIGNS

A discrete opacity is seen usually within the anterior corneal stroma. The center

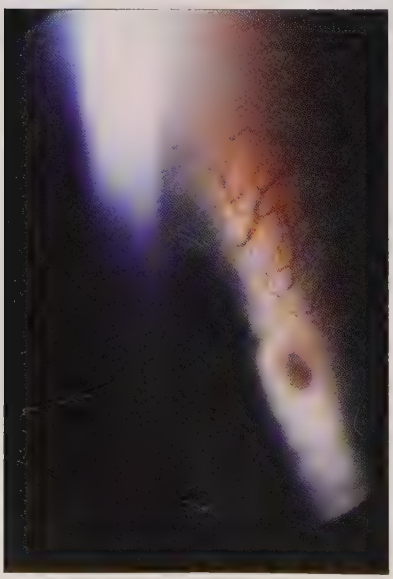


Fig. C-27a Two views of a focal sterile corneal infiltrate in a marginal keratitis. The degree of density of the infiltrate in retro-illumination.



Fig. C-27b Negative NaFl staining where the epithelium is raised not ulcerated.



of the infiltrate is often densely opaque, and white to gray in color. Surrounding this central dense area there is often a halo of less intense stromal infiltration, such that individual inflammatory cells can be seen on slit-lamp examination. There is also usually some degree of limbal or conjunctival hyperemia. The lesion should be assessed for the presence of an associated epithelial defect.

It may be possible to differentiate between infectious and sterile infiltrates by their location and size, as well as the presence of associated signs ("the company they keep"). The **PEDAL** mnemonic can be helpful in distinguishing the two conditions – an infected ulcer is more likely to be associated with **P**ain, **E**pithelial defect, **D**ischarge, **A**nterior chamber reaction, and a more central **L**ocation.

INCIDENCE

Corneal infiltrates are uncommon. However, they are more common in at-risk groups such as contact lens wearers and people with rosacea-associated blepharitis.

SIGNIFICANCE

Infectious infiltrates are very important, requiring immediate treatment because of their sight-threatening nature.

DIFFERENTIAL DIAGNOSIS

Corneal scarring; microbial keratitis; Marginal keratitis; contact lens-associated infiltrates; Dellen; Adenoviral keratoconjunctivitis; Herpes simplex keratitis.

SEE ALSO

Granular dystrophy; Rosacea keratitis; Foreign bodies – corneal and conjunctival; Corneal dystrophies, degenerations, and depositions.

C

MANAGEMENT

Infectious

Microbiological investigation and treatment with the appropriate antimicrobial agents is indicated. See the appropriate condition (e.g. "Bacterial keratitis," "Herpes simplex keratitis"). These require urgent treatment.

Non-infectious

- If sure that the lesion is non-infectious, then a mild topical steroid qid will usually lead to rapid resolution of the lesion. This is often covered with a broad-spectrum topical antibiotic qid.
- If contact lens-associated, withdrawal of contact lens use and increased lens hygiene are indicated. Changing to disposable SCL may also be indicated.
- If associated with blepharitis, treatment of the lid disease with lid hygiene and oral tetracycline may be indicated.

Review

Usually within 1–2 days to ensure resolution and lack of progression that may indicate an infection. If there is doubt about the diagnosis, the infiltrate should be treated as a microbial keratitis.

C

Corneal opacities – peripheral

Arcus senilis, Vogt's white limbal girdle, Kayser–Fleischer ring

DESCRIPTION

There are several conditions that lead to exclusively peripheral opacification of the cornea. The more important conditions affecting the peripheral cornea, such as marginal keratitis, Terrien's marginal degeneration, conjunctival intraepithelial neoplasia (CIN), and Mooren's ulcer, are dealt with in separate articles whereas more benign lesions are dealt with here. The peripheral nature of these conditions probably reflects the main difference that

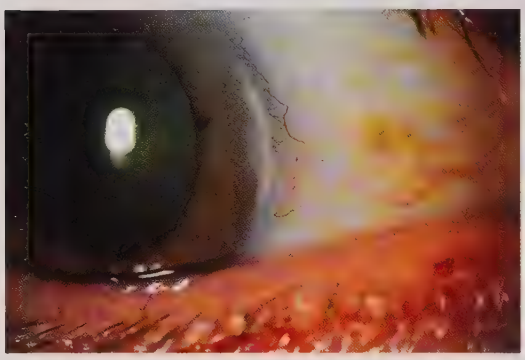


Fig. C-28a Vogt's white limbal girdle.

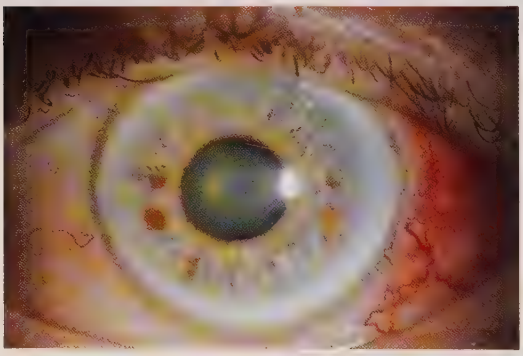


Fig. C-28b Arcus senilis.

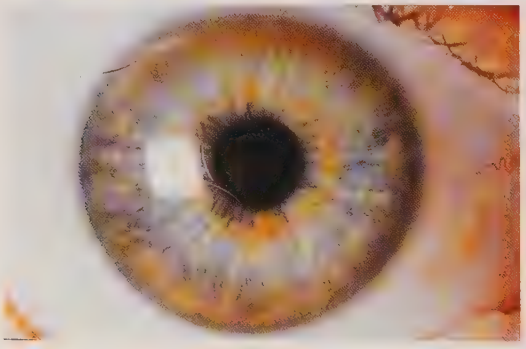
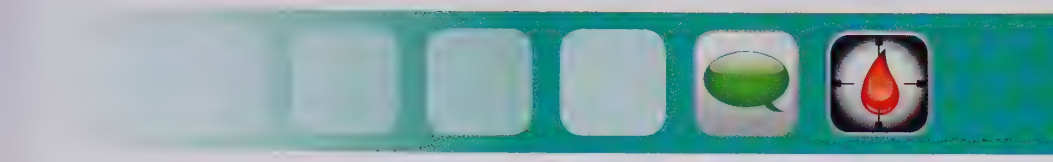


Fig. C-28c Kayser–Fleischer ring.



exists between the peripheral and central cornea, its close proximity to limbal blood vessels.

Vogt's white limbal girdle

This is a very common totally benign, exclusively peripheral corneal lesion. The appearance is that of a chalky white crescent-shaped lesion within the anterior stroma of the interpalpebral cornea. It is usually but not always separate from the limbus. It can affect either the nasal and/or the temporal cornea and is almost exclusively seen in older people. No treatment is required.

Arcus senilis

Arcus senilis, as the name indicates, is a crescent-shaped degenerative change that affects the peripheral cornea. It is extremely common, affecting almost all people over 80 years of age. It is usually not associated with any systemic disease, although it may rarely be associated with abnormal blood lipids. It may also occur in the rare inherited corneal disorder Schnyder's crystalline dystrophy. Blood lipid disturbances are associated with atherosclerotic disease, stroke, and heart attack. Any suspicious presentation of arcus senilis, such as in a young patient or if it is particularly dense or extensive, should be investigated with serum lipids.

The lesions initially occur in the superior and inferior limbal cornea and extend to involve the whole of the peripheral cornea. There is usually a clear zone between the arcus and the limbus. Occasionally the cornea within this peripheral clear zone can become thinned. This usually only occurs in quite elderly people and is referred to as a peripheral furrow.

Histologically, lipid is first deposited in Descemet's membrane and then in the anterior stroma. This leads to the formation of two triangles, apex to apex, visible both clinically and histologically.

Wilson's disease: Kayser–Fleischer ring

Wilson's disease (hepatolenticular degeneration) is a rare inherited metabolic disease affecting normal copper metabolism. It results in both liver and central neurological dysfunction. Excess copper is deposited in the peripheral cornea where it binds to a receptor protein in Descemet's membrane leading to an orange-colored opacity, a Kayser–Fleischer ring. This can also occur with chronic liver failure resulting in elevated serum copper levels. With treatment and restoration of serum copper levels to normal, the ring disappears. There may be an associated "sunflower" lens opacity.

DIFFERENTIAL DIAGNOSIS

Pseudogerontoxon (arcus juvenilis) of vernal keratoconjunctivitis; posterior embryotoxon (see "Iridocorneal dysgenesis").

SEE ALSO

Band keratopathy; Mooren's ulcer; Lipid keratopathy; Terrien's marginal degeneration; Pellucid marginal corneal degeneration; Marginal keratitis; Rosacea keratitis; microbial keratitis; Blepharitis – posterior; Corneal infiltrates – overview; Pannus; Conjunctival intraepithelial neoplasia; Peripheral ulcerative keratitis.

MANAGEMENT

No treatment is required for Vogt's white limbal girdle or the majority of cases of arcus senilis.

INVESTIGATIONS

Serum lipids should be investigated in cases of heavy arcus or arcus occurring in younger patients. Patients with a Kayser–Fleischer ring invariably present with pre-existing liver disease.

C

Crocodile shagreen

Corneal mosaic patterns, François cloudy dystrophy

DESCRIPTION

The term "crocodile shagreen" refers to the resemblance of polygonal opacities within the cornea to crocodile hide (shagreen). The basis for this mosaic appearance is probably the normal arrangement of stromal collagen lamellae.

There are several types of corneal mosaic patterns. Although there is no commonly agreed classification, they can be classified as follows:

- physiological anterior mosaic pattern
- anterior crocodile shagreen (mosaic shagreen of Vogt)
- posterior crocodile shagreen (central cloudy dystrophy of François)
- peripheral crocodile shagreen
- LASIK flap shagreen folds (these are discussed under refractive surgery signs).

SYMPTOMS

All except LASIK flap folds are asymptomatic, with a reduction in visual acuity from the opacities only occurring extremely rarely with posterior crocodile shagreen.

SIGNS

All have typical grayish-white polygonal opacities separated by relatively clear spaces.

Physiological anterior mosaic pattern

This can occur if the normal tension within Bowman's layer is relaxed and may result from trauma, hypotony, and aging. It is sometimes evident in the tear film fluorescein pattern following applanation tonometry and may be seen following rigid contact lens use.

Anterior crocodile shagreen

This can occur as a separate inherited condition or in association with other conditions such as megalocornea, iris malformations, and band keratopathy.

Histology reveals Bowman's membrane to be calcified and ridged with flattened overlying epithelium.

Posterior crocodile shagreen

The mosaic pattern is deep, being in the posterior third of the corneal stroma. It is only present in the central two-thirds of the cornea. Some clinicians distinguish this from François cloudy dystrophy although others consider the two entities to be the same. It occurs as an autosomal dominant trait. The histology is unique with a saw-tooth configuration of the collagen lamellae seen on electron microscopy.

Peripheral crocodile shagreen

A peripheral form of mosaic patterning can occur in the elderly. The lesions are usually spread throughout the cornea.

INCIDENCE

Uncommon. Physiological anterior mosaic pattern may be readily induced with corneal applanation.

SIGNIFICANCE

Very low, almost never causes a reduction in vision.

DIFFERENTIAL DIAGNOSIS

Corneal edema; folds in a LASIK flap (see "Refractive surgery signs – LASIK").

SEE ALSO

Corneal dystrophies, degenerations, and depositions.

MANAGEMENT

No intervention is required.

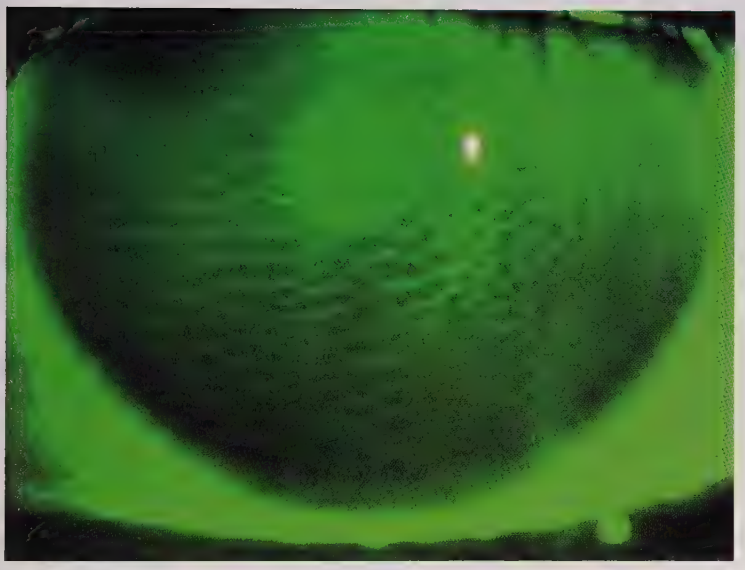


Fig. C-29a Physiological anterior mosaic pattern highlighted with fluorescein: a normal corneal pattern demonstrated in this patient after she gently rubbed her eye through the eyelid.

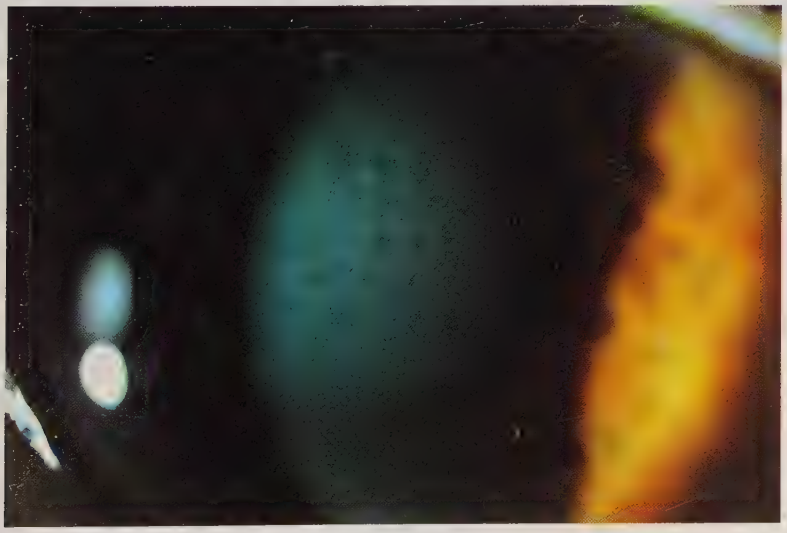


Fig. C-29b Anterior crocodile shagreen.

C

Crystalline keratopathy

Cystinosis, chrysiasis, paraproteinemia

DESCRIPTION

Many different substances can deposit in the corneal stroma in crystalline form. These include lipid, gold (chrysiasis), the amino acid cysteine (in cystinosis), proteins (in paraproteinemia), and urate (in gout).

Corneal stromal crystals are most commonly seen in lipid keratopathy and are composed of cholesterol. Cholesterol is also deposited in infectious crystalline keratopathy and in the rare inherited corneal disorder Schnyder's crystalline dystrophy.

The most serious crystalline deposition occurs with the deposition of protein with life-threatening paraproteinemias such as multiple myeloma, lymphoma, and Waldenstrom macroglobulinemia. It can also rarely be seen with more benign conditions such as benign monoclonal gammopathy.

Chrysiasis affects most patients who have received more than 1 g in total of gold.

SYMPTOMS

Only lipid keratopathy involving the central cornea, infectious crystalline keratopathy, and Schnyder's crystalline dystrophy usually result in decreased vision. Cystinosis is associated with severe photophobia and has obvious systemic features including severe growth retardation and renal failure. Chrysiasis and paraprotein deposition are usually asymptomatic.

SIGNS

Most crystalline depositions are characteristic enough to allow specific identification from their appearance. Lipid keratopathy occurs around an area of vascularization and extends throughout the corneal stroma. Gold deposits occur throughout the cornea as small, glittering granules and are more common in the deep and peripheral stroma. Occasionally, they can also deposit in the crystalline lens. Cysteine deposits are widespread throughout the cornea but most marked in the periphery and anteriorly. They appear as a "snowstorm" of small, fine, white crystals. Paraprotein deposits occur diffusely throughout the cornea. However, they are more marked superficially and in the periphery.

INCIDENCE

Aside from lipid keratopathy, all other forms of crystalline keratopathy are very rare.

SIGNIFICANCE

Cystinosis and paraproteinemias may be life-threatening. Crystalline keratopathy may be the first sign of a paraproteinemia.

DIFFERENTIAL DIAGNOSIS

Verticillata; Corneal scarring; drug depositions such as chlorpromazine.

SEE ALSO

Lipid keratopathy; Infectious crystalline keratopathy; Corneal dystrophies, degenerations, and depositions.

MANAGEMENT

Ocular investigations

Specular microscopy can occasionally definitively identify certain crystals due to their characteristic morphology. Cholesterol crystals have a characteristic notched border with sharp edges.

Blood tests

Patients with scattered crystalline deposits of unknown cause should be investigated for increased serum paraproteins with plasma electrophoresis and a full blood examination.

Specific treatments

Specific treatments exist for several crystalline keratopathies with systemic treatment of any malignancy for paraproteins and topical cystinase for cystinosis. The antibiotic treatment of infectious crystalline keratopathy is discussed in its specific article.

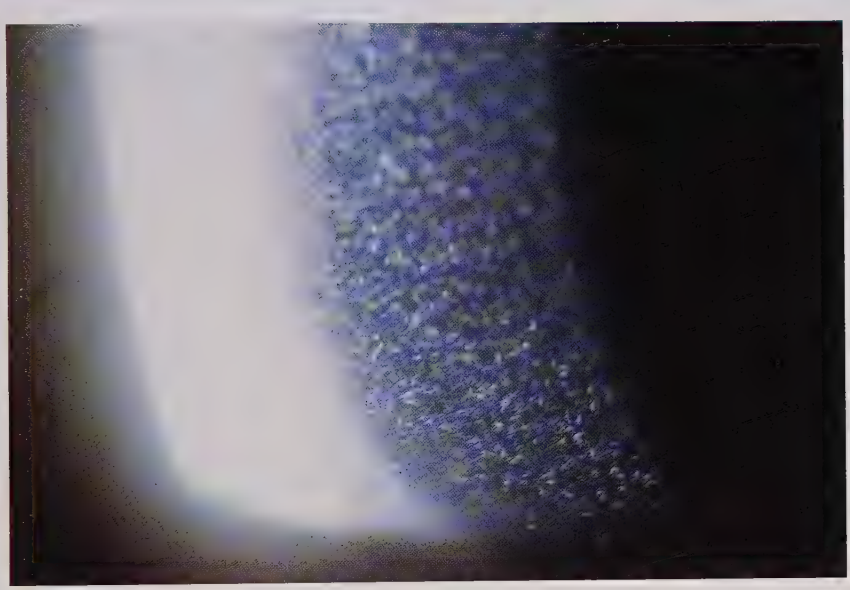


Fig. C-30a A "snowstorm" of small, white, crystals seen in a patient with cystinosis.

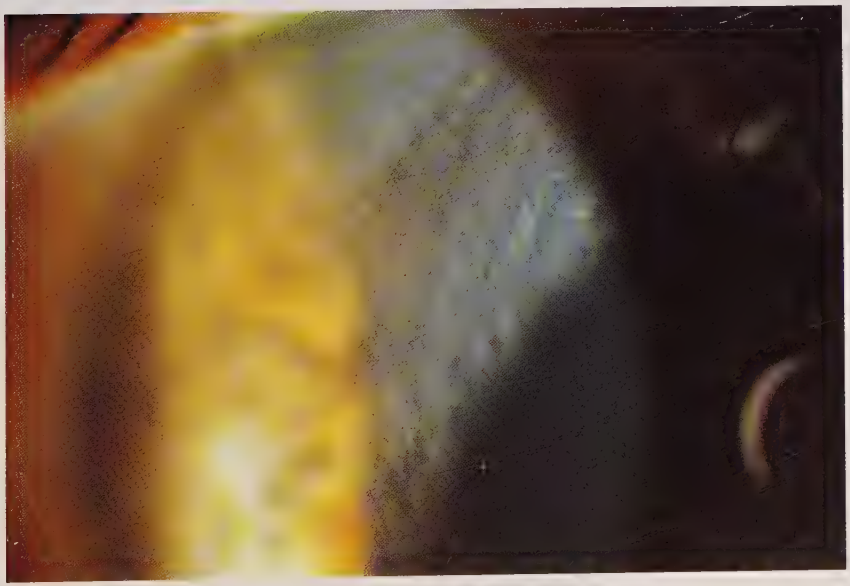


Fig. C-30b Similar crystal effect seen in monoclonal gammopathy.

C

Cysts of the eyelids

Cyst of Moll, eccrine sweat gland hidrocystoma, cyst of Zeis, sebaceous cyst, milia

DESCRIPTION

Cysts can arise in any of the different glands around the eyelid. Aside from the meibomian glands that run within the tarsal plate itself, the eyelid skin has its own eccrine and sebaceous glands. There are also modified sebaceous glands associated with the lash follicles (glands of Zeis) and modified eccrine glands (glands of Moll). All of these glands can become blocked, resulting in eyelid cysts. Chalazia and internal hordeolum affect the meibomian glands. They, along with external hordeolum, are discussed in separate articles.

SYMPTOMS

None of these cysts is painful. Patients normally present because of the cosmetic appearance or because of a concern that the lesion may be malignant.

SIGNS

Cysts of Moll

As they are eccrine glands, cysts arising from the glands of Moll are filled with clear fluid and, as such, are partially translucent. They occur along either the upper or the lower lid margin.

Eccrine sweat gland hidrocystoma

As these arise from skin sweat glands, they have an appearance similar to cysts of Moll. However, they are not confined to the lid margin.

Cysts of Zeis

Like the cysts of Moll these are confined to the lid margin but, as they are derived from sebaceous glands, they are filled with more opaque fluid than cysts of Moll.

Sebaceous cysts

These are less common and are derived from normal skin sebaceous glands. They are often larger than cysts of Zeis and typically have a small central punctum.

Milia

These are tiny white superficial cysts derived from hair follicle-associated glands. They are usually multiple and easily visible under the skin.

INCIDENCE

All of these cysts are common, cysts of Moll being the most common.

SIGNIFICANCE

None of these lesions has malignant potential and, therefore, they are only of cosmetic concern.

DIFFERENTIAL DIAGNOSIS

It is important to exclude the possibility of a malignant skin tumor such as a squamous cell carcinoma or basal cell carcinoma.

SEE ALSO

Chalazion; Hordeolum – internal and external; Skin tumors – benign; Skin tumors – overview; Sebaceous cell carcinoma.

MANAGEMENT

Advice

Patients can be reassured that these lesions are benign. Surgical treatment is only indicated for cosmetic reasons.

Surgery

Simple incision and expression of the contents is usually sufficient to remove these lesions. This can usually be performed at the slit lamp without anesthesia. If they recur, deroofing of the cyst allowing it to heal by secondary intention is usually effective.



Fig. C-31 Cysts affecting different glands around the eyelid. **a** Cyst of Moll (demonstrating characteristic transillumination). **b** Eccrine sweat gland hidrocystoma. **c** Milia of the upper eyelid. **d** Cyst of Zeis.

D

Dacryocystitis

Acute dacryocystitis, chronic dacryocystitis, mucocele

DESCRIPTION

Dacryocystitis is an inflammation within the lacrimal sac and can be acute or chronic. Acute dacryocystitis is due to a bacterial or, less frequently, a fungal infection within the lacrimal sac. This usually occurs due to tear stasis from pre-existing nasolacrimal duct obstruction. Chronic dacryocystitis is also secondary to chronic nasolacrimal duct obstruction but occurs without the dramatic signs of an acute bacterial infection.

While usually due to idiopathic blockage, less common causes of nasolacrimal duct obstruction include tumors, non-infectious granulomatous lesions, facial fractures and trauma, or foreign bodies such as punctal plugs. While dacryocystitis can occur at any age, it most frequently occurs in young children, secondary to congenital nasolacrimal duct obstruction, or in the elderly.

SYMPTOMS

Acute dacryocystitis usually presents with a painful, sometimes massive, enlargement of the lacrimal sac that develops over 1 to 2 days. There is usually an associated mucopurulent discharge and a history of epiphora.

Chronic dacryocystitis presents with epiphora with an associated painless swelling at the medial canthus.



Fig. D-1 Acute dacryocystitis with signs of lacrimal sac distension. Associated inflammation with redness and swelling of the overlying skin and eyelids is also present.



SIGNS
A distended tender lacrimal sac is present inferomedial to the medial canthus. There may be a lighter colored yellowy area within the swelling due to pointing of the pus within the distended sac. There is associated lid swelling and redness, and there may even be an area of cellulitis. Chronic dacryocystitis is associated with a mucocele, a painless swelling over the lacrimal sac.

INCIDENCE
Uncommon to rare (1/1000 to 1/10,000).

SIGNIFICANCE
Usually leads to chronic nasolacrimal duct obstruction with epiphora.

DIFFERENTIAL DIAGNOSIS
Canaliculitis.

SEE ALSO
Epiphora; Nasolacrimal duct obstruction – congenital; Punctal stenosis.

MANAGEMENT
Probing, dilation, and irrigation should not be attempted with an acute dacryocystitis because of the risk of spreading the infection leading to cellulitis.

Systemic antibiotics
Oral broad-spectrum antibiotics should be commenced immediately. A full course of antibiotics, usually for 1 week, is indicated.

Surgery
If very painful, immediate drainage of the sac with either a stab incision or aspiration with a large gauge (19G) needle is appropriate. A dacrycystorhinostomy (DCR) is usually required after an acute dacryocystitis, both to relieve epiphora and to prevent recurrence. A DCR is also appropriate in the management of chronic dacryocystitis with mucocele.

Dellen

Fuchs' dimple

DESCRIPTION

A dellen is a reversible, localized corneal depression. It is due to dehydration, with resultant compaction of the normal corneal stroma. This typically occurs from disruption of the pre-corneal tear film secondary to adjacent elevated structures such as a pterygium or limbal tumor. Contact lenses can also lead to the formation of a dellen; this is most commonly seen with rigid lenses. Dellens can also occur spontaneously for no obvious reason in the elderly.

SYMPTOMS

Foreign-body sensation is common, often with slight photophobia.

SIGNS

Dellens typically occur in the peripheral interpalpebral cornea. The appearance is that of a small, focal, depressed divot. The overlying epithelium is desiccated and demonstrates both pooling of fluorescein within the lesion and delayed staining. The underlying stroma is uninflamed. However,

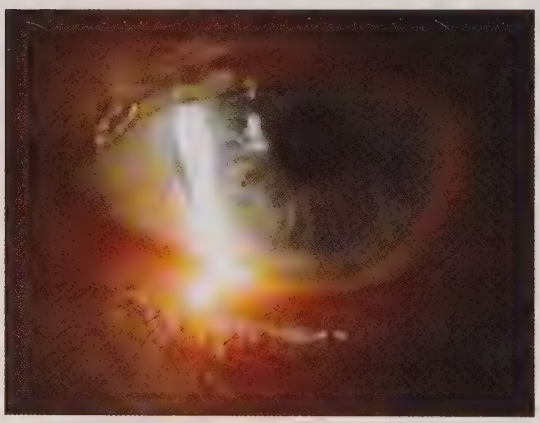


Fig. D-2a Corneal dellen secondary to a raised limbal lesion.

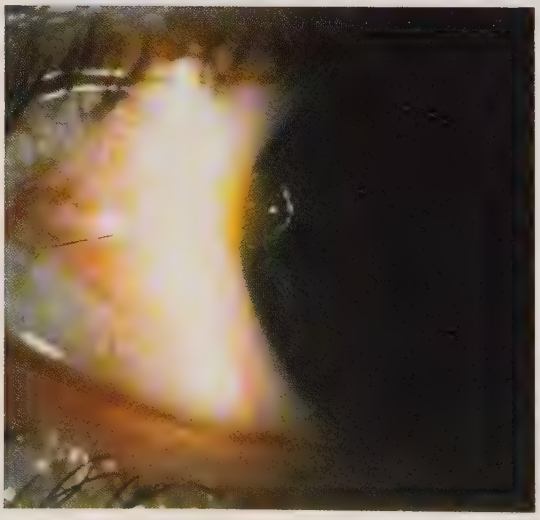


Fig. D-2b Slit-lamp view demonstrating both depressed "scalloped-out" nature of the lesion and fluorescein leakage through the compromised epithelium at the base of the dellen.



it is slightly opaque, and grayish in color. A causative limbal lesion is often seen.

INCIDENCE
Rare.

SIGNIFICANCE
Usually dellen are transient and innocuous. However, the overlying epithelial disruption and compromise of normal stromal structure may predispose to infection. Although a dellen does not lead to stromal loss, if it is long-standing stromal scarring may occur.

DIFFERENTIAL DIAGNOSIS
Corneal infiltrate.

SEE ALSO
Microbial keratitis; Pterygium.

MANAGEMENT
Tear supplements
Improving the precorneal tear film with tear supplements can reduce epithelial disruption and lead to resolution of the dellen.

Cease lens wear
If caused by rigid contact lens use the temporary cessation of lens wear and even refitting with hydrogel lenses may be indicated.

Surgery
Removal of the causative limbal mass may be indicated if problems persist.

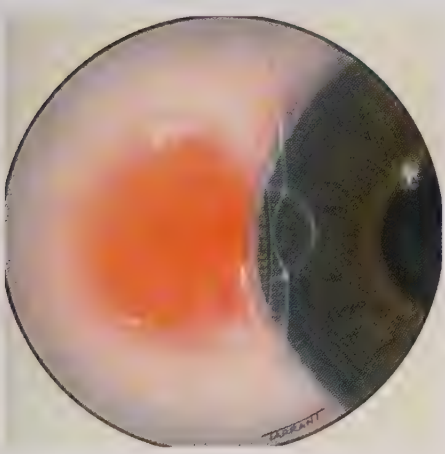


Fig. D-2c A dellen adjacent to a raised limbal lesion.

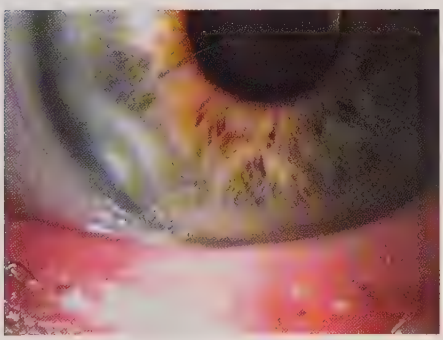


Fig. D-2d Limbal dellen with white crescentic opacity secondary to tissue compaction.

Dendritic ulcer

Herpes simplex keratitis

DESCRIPTION

The herpes simplex virus (HSV) can cause a wide spectrum of corneal disease. One of the most common and recognizable is a dendritic ulcer. It is caused by active viral replication within the corneal epithelium. The term dendritic refers to the branched nature of the lesion. Dendritic ulcers are a secondary manifestation of herpes simplex infection resulting from reactivation of dormant virus that had been residing in the cell bodies of the corneal sensory neurons. Although not supported by a recent investigation, many clinicians feel that reactivation often occurs following a precipitating event or "trigger factor," such as excessive sunlight exposure, an intercurrent illness such as influenza, or a particular life stress.

SYMPTOMS

Patients usually experience a slight foreign-body sensation. Pain is quite uncommon, probably because of the neurotrophic nature of the virus, traveling along sensory axons. Therefore, the corneal signs are often disproportionately marked compared to the symptoms. If a patient does have significant pain, it is more likely that the lesion is a healing epithelial defect. Tearing, slight blurring of vision, and a slightly red eye are also common. There may also be a history of recurrent attacks.

SIGNS

The key feature of this form of herpes simplex keratitis is the characteristic epithelial defect. This commences as an area of punctate erosions and rapidly leads to an irregular linear defect with one or several areas of viral activity usually dichotomously branching off from the main lesion (termed "end bulbs"). The epithelium in these areas stains with both rose bengal and fluorescein as a result of cellular damage secondary to active viral replication. Fluorescein leaks through the epithelium in these areas, due to damage to the normal cell–cell adhesion complexes, and this leads to late staining around the area. There is frequently slight opacification of the underlying corneal stroma. Enlarged, especially topical, corticosteroid modified-ulcers are termed "geographic ulcers." This can evolve into a chronic non-healing defect – a "meta-herpetic ulcer." Corneal sensation is adversely affected, especially in recurrent disease.

INCIDENCE

Affects about 1/1000 people at some time in their life. More common in young adults but can occur at any age.

SIGNIFICANCE

May lead to scarring of the anterior corneal stroma. Recurrence is common and may lead to other forms of herpes simplex keratitis such as stromal necrotic disease or disciform keratitis.

DIFFERENTIAL DIAGNOSIS

Healing epithelial defect; microdendrites of herpes zoster ophthalmicus; *Acanthamoeba*, herpes simplex primary blepharokeratoconjunctivitis.

SEE ALSO

Herpes simplex keratitis – overview; Disciform keratitis; Herpes zoster ophthalmicus; Follicular conjunctivitis; Persistent epithelial defect (PED).

MANAGEMENT

Advice

Two key features to emphasize to the patient are the need to avoid trigger factors where possible and to present promptly if they feel they may have a recurrence of the disease. Trigger factor avoidance largely comes down to good sunglasses with adequate side-protection when in high UV light. Prompt presentation is important as it can limit the degree and extent of corneal scarring.



Additional investigations

If the diagnosis is not certain direct immunofluorescent staining for viral antigens can be performed on an epithelial specimen.

Therapeutics

Gentle debridement of the epithelium along the edge of the dendrite with an anesthetic-soaked cotton bud is a very effective treatment. It is all the more effective when linked with the administration of a topical antiviral agent such as acyclovir ointment or trifluridine drops. With treatment, approximately 90% of dendrites are healed within 1 week. A recent publication from the Herpes Eye Disease Study (HEDS) has demonstrated the effectiveness of long-term prophylactic oral acyclovir in preventing recurrences.

Dosage: Oc. acyclovir 3%, or G. trifluridine 1% × 5 daily for 10 days.

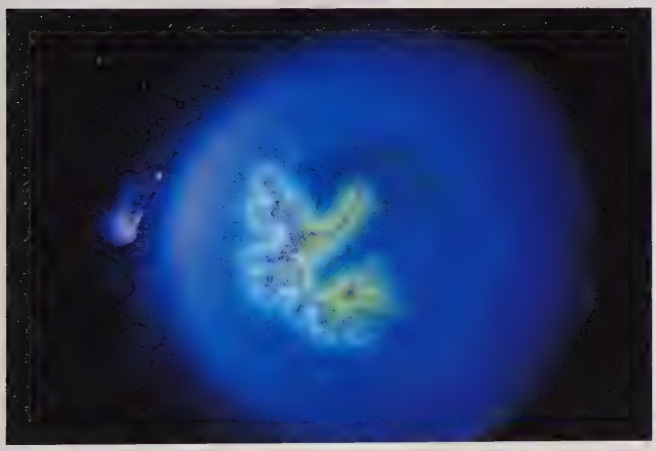


Fig. D-3a Large central dendritic ulcer. Note two areas of active viral replication on the right-hand side of the lesion. This photograph was taken several minutes after the instillation of fluorescein, and demonstrates late fluorescein leakage around these areas of viral activity.

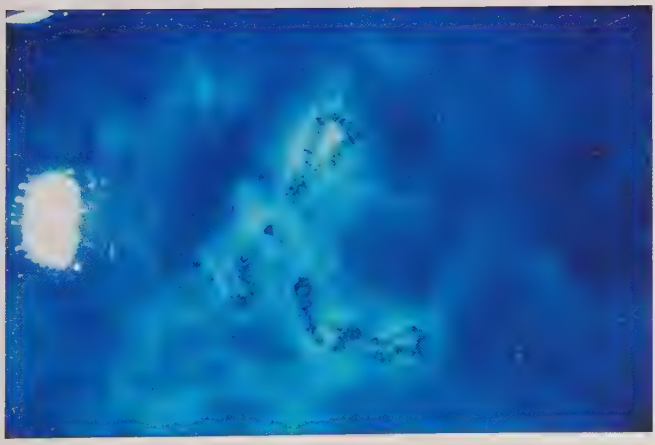


Fig. D-3b Early dendritic epithelial changes.

Dermatochalasis and blepharochalasis

Pseudoptosis

DESCRIPTION

Both of these conditions affect the eyelids leading to redundant skin and a baggy appearance around the eyelids.

Dermatochalasis

This is a very common, usually bilateral, degenerative condition affecting both the upper and lower eyelids and resulting in redundancy of the skin of the eyelids. It typically affects the elderly. Degenerative changes are seen not only within the eyelid skin and deeper tissue but also frequently within the septum orbitale. This leads to orbital fat prolapse into the eyelid.

Blepharochalasis

This is a very rare, bilateral, inflammatory condition occurring in young patients, and affecting the upper eyelids with bouts of acute, painless, non-pitting edema. Attacks usually commence around puberty, each lasting several days, and decrease in frequency over time. Eventually the eyelid skin becomes thinned and redundant resembling dermatochalasis. Both ptosis and anterior herniation of orbital fat may also occur.

SYMPTOMS

Patients with dermatochalasis present either because of concerns about the cosmetic effect or because of reduced vision from pseudoptosis. The redundant skin may even press on the upper lid lashes leading to blurring of vision by obscuring the visual axis. Blepharochalasis leads to typical, episodic lid edema.

SIGNS

The eyelid skin is thinned and redundant. There is often a loss of the normal upper eyelid skin crease, occasionally with eyelid skin draped down over the lid margin. Orbital fat herniation leads to a fullness of the eyelid; this is typically seen centrally, medially, or laterally on the lower eyelid although it can occur medially in the upper lid.

INCIDENCE

Dermatochalasis is very common while blepharochalasis is very rare.

SIGNIFICANCE

Although usually a cosmetic problem either may lead to reversible visual loss from pseudoptosis.

DIFFERENTIAL DIAGNOSIS

Ptosis; Acute allergic blepharo-conjunctivitis.

SEE ALSO

Entropion; Ectropion.

MANAGEMENT

Advice

Usually no treatment is required. The most common cause for intervention is cosmetic concern by the patient. Occasionally, intervention is indicated for visual reasons, with a superior visual field defect or blurring of vision from interference of the redundant skin or lash ptosis with the visual axis.

Surgery

Where indicated a blepharoplasty can be performed either with surgical excision of the redundant skin or with application of laser resulting in shrinkage of the affected skin. If there is significant fat herniation, surgical repair of the degenerate aponeurosis is also indicated.



Fig. D-4a Dermatochalasis. Note the bulge just inferior to the lower lid margin secondary to orbital fat herniation.



Fig. D-4b Dermatochalasis with the upper eyelid skin resting in a fold on the superior eyelashes.

D

Descemet's membrane breaks

Corneal hydrops, forceps injury, Haab striae

DESCRIPTION

An acute break in Descemet's membrane leads to rapid and gross corneal stromal edema, termed corneal hydrops. This most commonly occurs with the ectatic corneal dystrophies (keratoconus, keratoglobus, and pellucid marginal corneal degeneration). Breaks in Descemet's membrane can also occur secondary to birth trauma (usually a forceps injury) or from stretching of the membrane with congenital glaucoma (Haab striae). Thankfully, forceps injury breaks are now very uncommon due to the increasing use of caesarean section for difficult births. Following acute rupture and hydrops, the break is healed by the migration of endothelial cells over the defect and the gradual deposition of a new membrane.

SYMPTOMS

Acute hydrops results in marked photophobia, blurring of vision, discomfort, and a red watery eye. Congenital glaucoma and forceps injury usually result in reduced visual potential due to the effects of glaucoma or from amblyopia. Years later, endothelial cell dysfunction may progress with the development of irreversible corneal edema.

SIGNS

The break itself is spindle-shaped, often with tapered ends. Scrolled Descemet's membrane can sometimes be observed next to the break. If acute, as occurs in keratoconus, there is usually dense overlying corneal edema and microcystic edema of the corneal epithelium. The

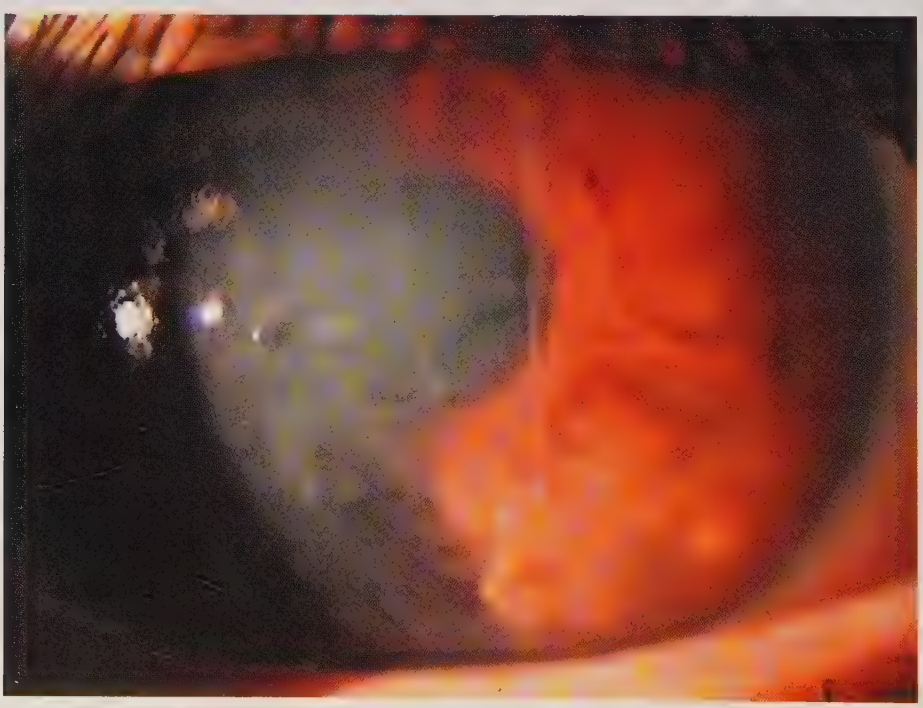


Fig. D-5 Break in Descemet's membrane resulting from a forceps injury. Note the overlying corneal edema secondary to endothelial cell dysfunction.



break is seen as a clearer area deep within the edematous cornea. It is usually in the inferior cornea and orientated vertically. Breaks from birth trauma or congenital glaucoma are usually associated with more generalized edema.

Breaks resulting from birth trauma are typically orientated diagonally with the long axis of the split running from superotemporal to inferonasal. They are more commonly seen in the left eye. Haab striae are more curvilinear and may be oriented in any direction but are most commonly horizontal. Other signs of congenital glaucoma that may be present in addition to Haab striae are an enlarged globe, an increase in corneal diameter (buphthalmos), cataract, and optic nerve head cupping and increase in size. Focal corneal stromal scarring commonly occurs over an area of hydrops and leads to local corneal flattening.

INCIDENCE

Hydrops in keratoconus is uncommon and usually occurs only in patients with advanced disease with significant corneal ectasia. It is most commonly seen in Down syndrome patients in whom corneal grafting surgery is usually avoided, even in advanced cases, due to poorer outcomes and increased risks of complications. Congenital glaucoma and birth trauma-associated breaks are both very rare.

SIGNIFICANCE

May lead to corneal edema with loss of vision and painful bullous keratopathy.

DIFFERENTIAL DIAGNOSIS

Posterior polymorphous dystrophy lesions may resemble Descemet's membrane splits; corneal surgical wounds such as clear corneal cataract incisions.

SEE ALSO

Corneal edema; Keratoconus; Pellucid marginal corneal degeneration.

MANAGEMENT

The management of corneal edema, resulting from a break in Descemet's membrane, with lubrication and hypertonic saline is the same irrespective of the cause. Specific treatments for the different causes of this condition are:

- Corneal hydrops. Usually a self-limiting condition unless the break is particularly extensive, resolving after 6 to 10 weeks. Supportive treatment in the interim is with sunglasses and often a topical corticosteroid such as prednisolone sodium phosphate 0.5% tds; a topical mydriatic can also be used to reduce photophobia. A penetrating keratoplasty (corneal graft) may be indicated if the edema fails to clear or if significant scarring develops. Occasionally, early marked corneal neovascularization develops. This is an indication for early and expedited grafting to avoid excessive corneal vascularization.
- Buphthalmos. Usually associated with very poor visual potential due to the development of both glaucomatous disc damage and amblyopia. As such a corneal graft is rarely indicated.
- Forceps injury. There is often associated amblyopia but, if good visual potential exists, a corneal graft may be performed for both visual improvement and pain relief.

D

Disciform keratitis

Herpetic keratouveitis, herpes zoster ophthalmicus, endotheliitis

DESCRIPTION

Several viral infections can lead to inflammation directed against the corneal endothelium. The corneal involvement is often, although not always, circular, hence the term disciform keratitis. It is seen with several herpes virus infections, most commonly with herpes simplex keratitis (HSK) and herpes zoster ophthalmicus (HZO). More rarely, it can also occur with chicken pox (the primary infection of herpes zoster) and measles. Herpes simplex disciform keratitis may be seen without any past history of HSK or in conjunction with other forms of HSK, such as dendrites and stromal necrotic disease. With HZO, onset of the disciform keratitis is typically several weeks after the initial skin lesions first appear. Rather than being secondary to active viral replication, it is thought that the excessive inflammation seen in keratouveitis is directed at residual non-replicating viral antigen remaining in the tissue after the initial infection.

SYMPTOMS

Patients have minimal pain and discomfort but do experience significant photophobia and blurring of vision. A dull ache around the eye, watery discharge, and red eye are common. There may be a history of previous HSK or recent HZO.

SIGNS

Endotheliitis and deep corneal edema with overlying microcystic edema of the epithelium may involve one area or the whole of the cornea. Underlying the area of edema there are focal keratitic precipitates, and the anterior chamber shows signs of inflammation with mild to moderate cell and flare. A mid-stromal whitish immune (Wessely) ring may be present. This is usually 5–6 mm in diameter and has an indistinct border. There may be an associated rise in intraocular pressure (hypertensive uveitis) due to trabeculitis; this is especially so with herpes simplex. Full recovery of vision is common following

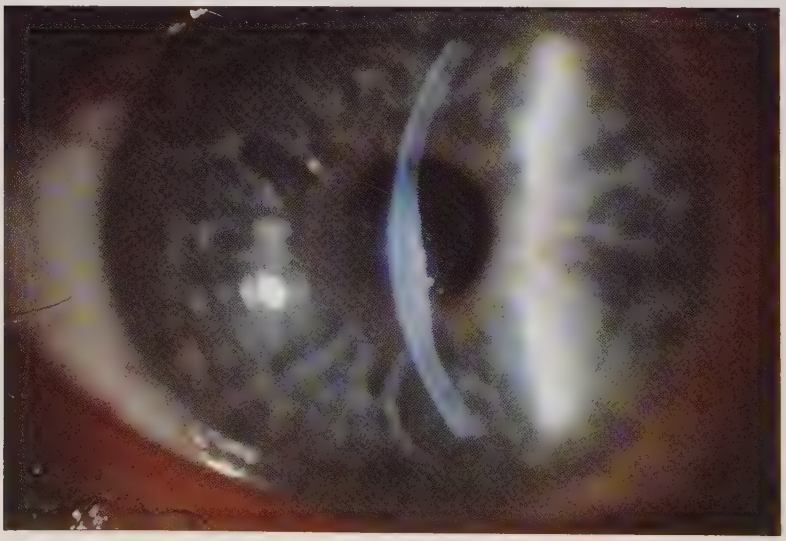
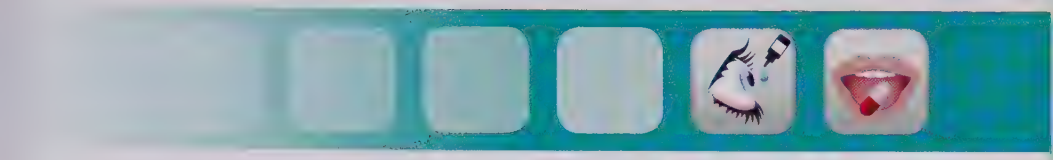


Fig. D-6 Herpes simplex disciform keratitis with central stromal edema and keratitic precipitates.



treatment with clearing of the corneal edema, although some residual stromal opacity due to scarring may persist for months to years following resolution of the active inflammatory episode. The scarring typically has a "ground glass" or reticular appearance.

INCIDENCE
Uncommon to rare.

SIGNIFICANCE
Most cases resolve with appropriate anti-inflammatory treatment, although recurrence is common with herpes simplex. Residual stromal scarring, with consequent loss of vision and photophobia, frequently occurs if the inflammation is long-standing, although it usually fades over months to years. HZO disciform keratitis usually takes years to fully resolve and, if uncontrolled, may lead to permanent corneal edema due to endothelial cell loss. The development of a nuclear sclerotic cataract is also common with HZO disciform keratitis.

DIFFERENTIAL DIAGNOSIS
Iritis; Interstitial keratitis; Corneal edema from other causes.

SEE ALSO
Corneal edema; Dendritic ulcer; Herpes simplex keratitis – overview; Herpes zoster ophthalmicus.

MANAGEMENT
Treatment is directed at resolution of active inflammation to reduce corneal edema and any rise in intraocular pressure. For disciform keratitis caused by herpes simplex virus this needs to be carried out in conjunction with adequate antiviral coverage.

Therapeutics
The patient is treated with intensive topical corticosteroids such as prednisolone acetate until some resolution of inflammation occurs. This is then slowly tapered over several months. Recurrence with cessation of the topical steroid is common and, for this reason, some topical corticosteroids are usually used for 4 to 6 months. With HZO it is common for patients to require several or more years of topical corticosteroids before being able to cease all medication without suffering a flare-up in their condition. Any associated rise in intraocular pressure should be treated with a topical anti-glaucoma medication, most commonly a topical beta-blocker such as timoptol, until the intraocular inflammation is adequately controlled. Topical prostaglandin inhibitors such as latanoprost have a proinflammatory effect, and topical carbonic anhydrase inhibitors such as dorzolamide can lead to worsening of the corneal edema: as such both of these medications are relatively contraindicated. A topical antiviral agent such as acyclovir ointment should be used prophylactically during corticosteroid treatment of herpes simplex disciform keratitis. This is used to suppress any enhancement of viral replication from the use of topical steroids. This is generally not needed for HZO disciform keratitis where it just adds to surface toxicity.

Dosage: Initially G. prednisolone acetate 1% q2h for 2 days then qid for several weeks followed by a slow taper usually tds for 1 month, bd for 1 month, then daily for several months. A topical antiviral such as Oc. acyclovir 3% or G. trifluridine 1% is administered bd until the steroid is down to bd, at which point it can be ceased.

Oral medication
The Herpetic Eye Disease Study (HEDS) has demonstrated the benefit of long-term prophylactic oral acyclovir (400 mg oral bd) in reducing the risk of recurrence of disciform keratitis. Unfortunately, this benefit only persists as long as the medication is taken.

D

Dry eye – overview

DESCRIPTION

The cornea and conjunctiva are unique in the body in that they are an exposed mucous membrane requiring constant wetting to maintain their health and, in the case of the cornea, its clarity. Ocular surface wetting is achieved by the action of the tear film, eyelids, lacrimal glands, and ocular surface. The tears consist of an aqueous component from the lacrimal and accessory lacrimal glands and lipid from the meibomian glands. A layer of mucin produced by both the conjunctival goblet cells and the corneal epithelium facilitates wetting of the ocular surface. A deficiency in any one of these components can result in tear-film instability and lead to drying of the ocular surface. This is referred to as dry eye; a decrease in aqueous production is often referred to as aqueous tear-deficiency dry eye (ATDDE); an abnormal increase in the evaporation of tears from the ocular surface, as occurs with a lipid layer deficiency, is referred to as evaporative dry eye (EDE).

Aqueous deficiency

A dry eye resulting from a decrease in the aqueous component of the tear film is referred to as keratoconjunctivitis sicca (KCS) (see "Dry eye syndrome – KCS"). A minor degree of aqueous tear film deficiency is common, especially in post-menopausal women.

Lipid anomaly

Chronic blepharitis with an alteration in the lipid component of the tear film may lead to a dry eye through instability of the tear film (see "Blepharitis – posterior").

Mucus deficiency

This is a rare cause of dry eye, and usually leads to particularly severe disease. It results from goblet cell destruction secondary to causes of conjunctival scarring and, if marked, may lead to ocular surface keratinization. (See "Chemical burns," "Vitamin A deficiency," and "Ocular cicatricial pemphigoid.")

Dysfunction of tear film distribution

Ocular surface dryness can result from dysfunction of the normal wetting action of the eyelids as occurs with a decreased blink rate (with Alzheimer's disease) or lagophthalmos (see "Exposure keratopathy") and malposition of the lids (see "Ectropion"). It may also occur with ocular surface irregularities (see "Pterygium," "Pingueculum").

Allergy

The presence of inflammatory cells or other inflammatory components in the tear film, as occurs in allergic eye disease, also causes tear instability. Chronic irritation from other forms of dry eye can also cause ocular surface inflammation, exacerbating the underlying condition.

SYMPTOMS AND SIGNS

Symptoms and signs of dry eye may be grouped into seven categories dependent on the method of investigation:

- Symptoms. Patients may report symptoms such as "dryness," burning, sandy or foreign-body sensation, with varying frequency and severity.
- Stability of the tear film. Most notably fluorescein tear break-up time (TBUT) and

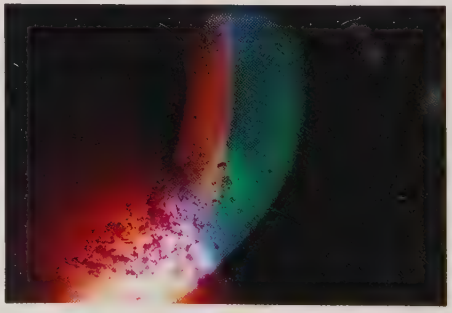


Fig. D-7a Signs common to the various forms of dry eye. Fluorescein and rose bengal staining of the inferior interpalpebral cornea and conjunctival epithelium with associated sparse tear film.



Fig. D-7b Rapid break-up of the tear film.

non-invasive break-up time. A TBUT less than 5–10 s is considered abnormal.
- Ocular surface evaluation. In particular, fluorescein staining, rose bengal staining, and impression cytology.
- Secretion or quantity tests. Such as the Schirmer and cotton thread tests.
- Slit-lamp biomicroscopy. Including tear meniscus height measurement, debris assessment, and lipid layer interferometry.
- Surfacing by the eyelids. Including blink rate, completeness, surface irregularities.
- Solute tests. Such as tear osmolality, tear fern ing, lactoferrin analysis (non-clinical).

INCIDENCE
Common, especially due to meibomian gland dysfunction, and in patients with collagen vascular diseases such as rheumatoid arthritis.

SIGNIFICANCE
Although often only causing minimal problems, severe dry eye can cause epithelial defects, chronic ocular irritation, and increased risk of infection.

DIFFERENTIAL DIAGNOSIS
Toxic keratopathy; Superficial punctate keratopathy; Superior limbic keratoconjunctivitis; conditions listed above in "Description."

SEE ALSO
Dellen; Persistent epithelial defects.

MANAGEMENT
Any underlying cause, such as blepharitis or eyelid disturbance, should be treated.

Advice
Avoidance of drying situations such as being in hot or cold winds, near air-conditioners, or radiant heaters. Protective glasses may be useful.

Tear supplements (lubricants)
These are usually in multi-use bottles for occasional application, ointment form for overnight use, and as unpreserved unit-dose form if required more than several times a day. A topical liposomal tear stabilizing agent (Tears Again) may be of particular benefit in the presence of a lipid layer abnormality with an abnormally short TBUT. Oral omega-3 supplements of 1–2 g daily, flaxseed oil, and fish oil may be of benefit, especially in the presence of a poor diet.

Therapeutics
In any allergic or severe forms of dry eye, a mild anti-inflammatory may be used. Ocular surface keratinization may be reversed with the use of 1% topical vitamin A ointment; care should be taken when used in darker skinned individuals because of its ability to lead to skin depigmentation.

Consulting room procedures
Maximizing both normal tears and lubricant effect, by occluding the puncta, may be appropriate for an aqueous tear deficiency.

Tarsorrhaphy
Reduction of the ocular surface area with a lateral tarsorrhaphy is reserved for severe sight-threatening cases of dry eye refractory to more conservative treatments.

D

Dry eye syndromes – keratoconjunctivitis sicca (KCS)

Sjögren's syndrome

DESCRIPTION

A dry eye resulting from a decrease in the aqueous component of the tear film is referred to as keratoconjunctivitis sicca (KCS). More severe forms are often associated with other diseases such as rheumatoid arthritis, Sjögren's syndrome, graft versus host disease following bone marrow transplantation, or following herpes zoster ophthalmicus. Several drugs can cause or exacerbate KCS, including retinoids, propanolol, and tricyclic antidepressants. A minor degree of aqueous deficiency is common in post-menopausal women; a recent study suggested hormone replacement therapy (HRT) may exacerbate the condition. KCS may be made worse if there are other concurrent causes of dry eye, such as blepharitis or a reduced blink rate. KCS may also occur in some patients following corneal refractive surgery, especially LASIK.

SYMPTOMS

Intermittent burning, a gritty foreign-body sensation, a red eye, and a mild mucous discharge are all common. Paradoxical reflex tearing may occur secondary to ocular surface irritation from the dryness. Patients with severe dry eye are usually quite photophobic. There may be associated blepharitis symptoms or symptoms of a dry mouth with Sjögren's syndrome. Increased evaporation from the ocular surface by hot or dry winds or being in an under-humidified environment also characteristically makes symptoms worse.

SIGNS

KCS will typically show a decreased lid margin tear meniscus height (less than 0.3–0.4 mm). A reduction in both basal and reflex aqueous tear production may be demonstrated with a Schirmer test (less than 6 mm of wetting after 5 min) or in tear volume with the phenol red cotton thread test (less than 10 mm after 15 s). As with all forms of dry eye, signs of an unstable or insufficient tear film include a reduced tear break-up time (TBUT), with less than 5–10 s considered abnormal. Ocular surface damage is observed with punctate fluorescein (an exclusion dye) and rose bengal (a vital stain) staining of both the corneal and conjunctival epithelium. This is particularly seen in the interpalpebral area with a band of staining across the central cornea and triangular wedges of staining on the perilimbal bulbar conjunctiva. The degree of staining reflects the severity of the dry eye. In severe dry eye, epithelial disturbance may be so severe as to cause a corneal ulcer.

Increased mucus production as evidenced by the presence of a mucous strand in the inferior fornix is common.

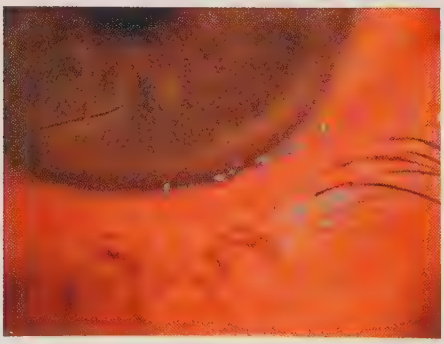


Fig. D-8a Sign of an aqueous tear film deficiency, scant inferior tear film meniscus.



Fig. D-8b Test for reduced aqueous tear film production, phenol red cotton thread test.



Ocular surface inflammation and posterior blepharitis are also common and reflect the debilitated ocular surface.

INCIDENCE
Very common, especially in post-menopausal women and patients with collagen vascular diseases such as rheumatoid arthritis.

SIGNIFICANCE
Severe dry eye can cause epithelial defects and even corneal perforation.

DIFFERENTIAL DIAGNOSIS
Toxic keratopathy; other causes of superficial punctate keratopathy.

SEE ALSO
Blepharitis – anterior; Blepharitis – posterior; Rheumatoid arthritis, and the eye; Herpes zoster ophthalmicus; Superior limbic keratoconjunctivitis (SLK); Persistent epithelial defects (PED).

MANAGEMENT
Advice
Important non-drug treatments include avoidance of drying situations, such as being in hot or cold winds, near air-conditioners, and being too close to radiant heaters such as fires. Protective glasses may be useful in such situations.

Tear supplements
Tear supplements (also termed lubricants) are absorptive polymers. They are available as drops, ointments, and gels and can be preserved or unpreserved. Many of the preservatives, notably benzalkonium chloride and cetrimide, can be toxic to the epithelium and, for this reason, if required more than several times a day unpreserved lubricants are used.

Therapeutics
In severe dry eye the addition of a mild anti-inflammatory agent such as fluorometholone daily may significantly improve comfort. Topical cyclosporin A is used as a long-term treatment for KCS because of its ability to lead to increased lacrimal gland function with increased tear production. A topical liposomal tear stabilizing agent (Tears Again) may be of particular benefit in the presence of a lipid layer abnormality with an abnormally short TBUT. Oral omega-3 supplements of 1–2 g daily, flaxseed oil, and fish oil may be of benefit, especially in the presence of a poor diet. Exact dosage recommendations are often difficult due to considerable variation of component oils in various preparations.

Punctal occlusion
Maximizing both normal tears and lubricant effect by occluding the puncta is usually employed if tear supplements by themselves are insufficient. This can be done permanently with cautery. This is usually preceded by a trial of dissolvable collagen canalicular plugs, or non-dissolvable silicone punctal plugs.

Tarsorrhaphy
Reduction of the ocular surface area with a lateral tarsorrhaphy is reserved for severe sight-threatening cases of dry eye refractory to more conservative treatments.

E

Ectopia lentis

Subluxed lens, dislocated lens, zonular dehiscence

DESCRIPTION

The term "ectopia lentis" refers to displacement of the crystalline lens from its normal position. If it is totally displaced from the pupil, this is referred to as dislocation; if it is still partially within the pupillary space, it is termed subluxation. Dislocation of the lens occurs because of loosening of the zonular adherens (zonules), the normal support structures for the lens.

There are several causes of ectopia lentis, explained below:

- Trauma. This is the most common cause of ectopia lentis and occurs secondary to traumatic rupture of the zonules. Most commonly this is caused by blunt trauma to the eye such as with a punch or from a squash ball. There is often accompanying damage to other intraocular structures, such as the lens (cataract), trabecular meshwork (leading to glaucoma), and iris (leading to hyphema and sphincter ruptures).
- Hereditary ectopia lentis. Both dominant and recessive forms of inherited ectopia lentis exist and are not associated with any other ocular or extraocular abnormalities.
- Marfan's syndrome. This is an autosomal dominant inherited disorder of connective tissues leading to skeletal disorders, muscular underdevelopment, cardiovascular abnormalities, and ectopia lentis. The lens is usually displaced superiorly. Other ocular manifestations include an increased risk of retinal detachment and glaucoma.
- Homocystinuria. This is an autosomal recessive inherited disorder affecting amino acid metabolism. The lens is usually displaced inferiorly. Patients also have a clotting disorder due to vascular endothelial abnormalities; this can lead



Fig. E-1 Superior subluxation of the crystalline lens in a patient with Marfan's syndrome. Note the elongated and stretched zonules still attached to the lens inferiorly.



to sudden death due to intravascular coagulation.
- Other inherited conditions. Ectopia lentis is also occasionally seen in a range of other conditions including Weill–Marchesani syndrome, aniridia, and Ehlers–Danlos syndrome.

SYMPTOMS
Poor vision and refractive instability are the most common presenting problems. Posterior dislocation of the lens can lead to induced myopia while total dislocation leads to effective aphakia with resultant hypermetropia. The lens is often mobile (phacodonesis), due to loosening of the zonules. This leads to an unstable refraction with regard to both the cylinder and the sphere. Best-corrected acuity is also often reduced. Due to a loss of the normal accommodative reflex from dysfunctional zonules the eye is effectively presbyopic.

SIGNS
Phacodonesis, lens displacement, and iris distortion can be observed on the slit lamp; this is best seen with a dilated pupil. Testing demonstrates a loss of accommodation. Refraction often reveals myopic astigmatism with an oblique cylinder; the refractive error is often unstable.

INCIDENCE
Very rare.

SIGNIFICANCE
It may be associated with a life-threatening disease such as homocystinuria or Marfan's syndrome.

DIFFERENTIAL DIAGNOSIS
Keratoconus (can also lead to myopic astigmatism with an unstable refraction with oblique cylinder).

SEE ALSO
Aniridia; Eyelid trauma; Intraocular lenses (IOLs).

E

MANAGEMENT
Therapeutics
If the lens is only slightly displaced, the daily use of a miotic such as 1% or 2% pilocarpine may constrict the pupil sufficiently to remove optical distortions derived from the edge of the lens, and thereby improve vision and reduce glare.

Genetic advice
If an underlying genetic defect is suspected referral to a physician for appropriate investigation is important.

Surgery
Surgery is indicated when the visual disability induced by the mobile and displaced lens is sufficient to adversely affect the patient's quality of life. A lens extraction and intraocular lens implantation is performed. If the lens is markedly displaced, and quite mobile, a pars plana vitrectomy and lensectomy is the procedure of choice. Otherwise, a usual anterior transcorneal approach is employed. Placement of the intraocular lens depends on the integrity of the capsule and the zonules. If the zonules are very weak or almost nonexistent an anterior chamber intraocular lens may have to be used.

E Ectropion

DESCRIPTION

Ectropion refers to eversion of the eyelid whereby it rests away from the eye. This almost exclusively affects the lower eyelid and may affect only a part of the eyelid (e.g. medial ectropion) or the whole length of the lid.

The causes of ectropion are:

- Involutional. With aging, degenerative changes occur within the medial and lateral canthal tendons and the tarsal plate. This can result in overall loosening of the lower eyelid and lead to ectropion.
- Cicatricial. Scarring of the skin and underlying tissue can pull the eyelid margin away from the eye. This is most commonly caused by prolonged sun exposure with actinic skin changes but can also occur with burns, skin tumors, and with some scarring skin conditions.
- Paralytic. Disruption of the facial nerve (the seventh cranial nerve) leads to a lack of tone in the orbicularis oculi muscle. This leads to loosening of the lower eyelid with ectropion and inferior ptosis. This is most commonly seen with Bell's palsy but may also occur from trauma or tumors along the length of the nerve.
- Mechanical. Tumors on or near the lid margin can lead to mechanical eversion and ectropion.
- Inflammatory. Inflammatory eyelid swelling from infection or allergy may lead to ectropion.

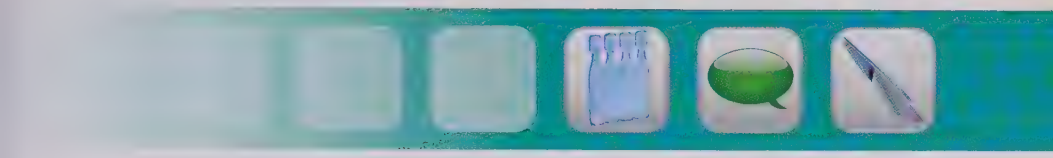
The main concern with ectropion is disruption of normal lid function resulting in a disturbance of the ocular surface. This can cause corneal drying and ulceration predisposing the eye to microbial keratitis.

SYMPTOMS

Epiphora is common, especially with medial ectropion. This results from malposition of



Fig. E-2 Involutional (senile) ectropion with forward displacement of the eyelid margin, and exposure of the inferior tarsal and forniceal conjunctiva.



the inferior punctum. A red sore eye and mucus discharge are also common.

SIGNS

The cause of the ectropion should be sought, looking for lid laxity (involutional), scarring of the skin (cicatricial), facial weakness (paralytic), and eyelid tumors (mechanical). The ocular surface should be closely examined for signs of epithelial disruption such as punctate fluorescein staining or even ulceration.

INCIDENCE

Common, especially in the elderly.

SIGNIFICANCE

Aside from leading to a sore, watery red eye, ectropion may result in corneal epithelial disturbances.

SEE ALSO

Bell's palsy; Entropion; Epiphora; Punctal stenosis.

MANAGEMENT

Advice

Mild ectropion may require no treatment other than advice to minimize rubbing and trauma of the lower lid to limit progression.

Tear supplements

Tear supplements are of prime importance in preventing desiccation of the ocular surface from the eyelid dysfunction seen with ectropion. If the ocular surface is disrupted, lubricant drops and ointment are important. After application of the lubricant, active blinking and even manual support of the lower lid may be important to facilitate distribution of drops or ointment across the ocular surface. During the day ocular protection can be aided with the use of sunglasses with good side protection.

Surgery

Surgical repair with a tarsorrhaphy or inferior lid shortening procedure may be indicated depending on the severity and cause of the ectropion. Indications for surgical intervention may include ocular surface disruption, chronic epiphora, recurrent irritation, recurrent bacterial conjunctivitis, and cosmetic concern.

E Endophthalmitis

DESCRIPTION

Endophthalmitis is one of the most feared complications of intraocular surgery. It is one of the few true ophthalmic emergencies and refers to an infection primarily involving the anterior chamber and posterior segment including the vitreous. Endophthalmitis is usually caused by bacteria, although rarely it can also be caused by fungi. Some bacteria, such as streptococci and *Bacillus cereus*, lead to more severe inflammation and a heightened risk of vision or even globe loss whereas others, such as *Staphylococcus epidermidis*, have a lower risk of significant visual loss. The organism can gain access to the eye via one of two routes:

- Exogenous. Entry to the eye is via a wound in intraocular surgery, following penetrating trauma or corneal perforation, along a suture track or, rarely, from posterior spread of microbial keratitis. Significant risk factors for the development of endophthalmitis following a penetrating eye injury include the presence of an intraocular foreign body, a dirty injury, and delayed time until repair of the wound. The rate of endophthalmitis following a penetrating eye injury in some studies approaches 30%. Late endophthalmitis can occur via a leaking conjunctival bleb following glaucoma drainage surgery. Postoperative endophthalmitis is overwhelmingly caused by Gram-positive organisms, especially staphylococci and streptococci.

  The risk of postoperative endophthalmitis can probably be reduced by the preoperative management of chronic periocular infections such as staphylococcal blepharitis, chronic dacryocystitis, or canaliculitis. The instillation of a few drops of povidine/iodine solution into the inferior fornix immediately preoperative has been demonstrated to reduce the risk of endophthalmitis while performing cataract surgery at the same time as lid procedures have been demonstrated to increase the risk. Other measures used by some clinicians in an attempt to reduce the risk of endophthalmitis include a drop of topical broad-spectrum antibiotic immediately preoperatively and a single subconjunctival injection of a broad-spectrum antibiotic at the end of the operation. Despite this, the only antibiotic

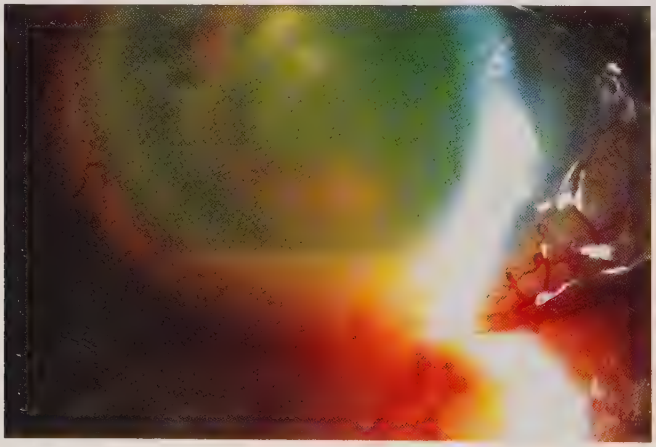


Fig. E-3 Acute bacterial endophthalmitis with corneal edema, hypopyon, and fibrinous exudate.



administration shown conclusively in a randomized controlled study to significantly reduce the risk of developing endophthalmitis is the intracameral (into the anterior chamber) administration of the cephalosporin antibiotic cefuroxime at the end of surgery.

- Endogenous. This mode of infection is quite uncommon and occurs when the microorganism gains access to the eye via the bloodstream. This usually occurs from septicemia complicating a systemic infection such as pyelonephritis or from intravenous drug use (where there is a higher than normal rate of fungal endophthalmitis).

Although usually acute with a rapid onset, endophthalmitis can occasionally present as a chronic infection. This is especially the case with *Proprionibacterium acnes* infections following cataract surgery.

SYMPTOMS

Over several hours to a day the eye becomes red, sore, and photophobic. Vision dims, and the Snellen acuity may be worse than 6/60. The patient is usually significantly distressed by the rapid reduction in vision to urgently seek medical attention. Symptoms typically commence 3–7 days following intraocular surgery.

SIGNS

The eye is inflamed with cells and flare in the anterior chamber and vitreous. There may be a hypopyon, anterior chamber fibrinous exudate, and collections of pus within the vitreous cavity. The cornea is usually edematous with deep Descemet folds, and the iris is inflamed, frequently with posterior synechia.

INCIDENCE

Approximately 1/500 to 1/1000 following cataract surgery, although the incidence is reduced to approximately 1/8000 with the instillation of intracameral cefuroxime or cephazolin at the end of the surgery.

Moxifloxacillin has also been used as a prophylactic intracameral agent and is probably as effective.

SIGNIFICANCE

Approximately 50% of eyes with endophthalmitis end up with severely reduced vision.

DIFFERENTIAL DIAGNOSIS

Uveitis (especially in poorly-controlled diabetes following cataract surgery), retained lens material; Disciform keratitis.

SEE ALSO

Cataract incisions; Hypopyon.

MANAGEMENT

Urgent treatment is essential. Even a few hours delay in treatment can result in significantly increased visual loss. For this reason urgent referral to an appropriate center such as an ophthalmic emergency department is required. Contact should be made with the hospital or ophthalmologist to alert them to the case.

Surgery

Urgent surgery is required to allow both the collection of appropriate microbiological specimens and the intravitreal injection of antibiotics (usually vancomycin and ceftazidine) and corticosteroid (dexamethasone). A vitrectomy may be performed at the same time although a large randomized study has demonstrated that this probably has little effect on the final visual outcome. It is essential the antibiotics are given intravitreally; topical, oral, or intravenous antibiotics are inadequate to treat the infection. Following surgery the patient is commenced on topical mydriatics, corticosteroids, and antibiotics. Occasionally, a repeat injection of intravitreal antibiotics and corticosteroids is required 2 to 3 days following the initial injections if the signs fail to resolve.

E Endothelial opacities – overview

DESCRIPTION

Opacities on the endothelial surface of the cornea can occur in a number of pathological and non-pathological conditions. Causes are discussed below:

- Surgery. Both surgical and non-surgical trauma can affect the endothelium. The most common surgical trauma is with cataract surgery. Modern clear corneal incisions lead to a linear scar parallel to and several millimeters in from the limbus. This may be superior or temporal. In addition there is a scar from the port site for the second instrument used in cataract surgery. This may be temporal, nasal, superior, or inferior.
- Trauma. Non-surgical trauma may occur at the time of birth leading to breaks in Descemet's membrane (see "Descemet's breaks") or from penetrating ocular trauma. Breaks in Descemet's membrane also occur with hydrops complicating keratoconus.
- Degeneration. Although typically seen with Fuchs' endothelial dystrophy, guttae may develop as a normal aging change in elderly patients. Guttae are small, usually central, and sometimes grouped raised areas on the endothelial surface. They are described as excrescences and lead to a stippled appearance of the endothelium. They are often best seen on retroillumination.
- Inflammation. Some of the most commonly seen endothelial opacities are keratitic precipitates. These are collections of inflammatory cells, both lymphocytes and neutrophils, seen with any anterior segment inflammation. This includes iritis, microbial keratitis, and corneal graft rejection. Even when the inflammation has settled the precipitates may persist for several weeks to months. As they age they often become smaller and more pigmented.
- Dystrophies. Two main dystrophies lead to endothelial opacities: Fuchs' endothelial dystrophy and posterior polymorphous dystrophy. Fuchs' results in central guttae (described above), often with associated pigment cell deposition, and deep stromal edema. Posterior polymorphous dystrophy leads to typical grouped vesicular lesions sometimes described as resembling frogspawn.
- Deposits. Both pseudoexfoliative material from pseudoexfoliation (PXF) and pigment granules can deposit on the endothelial surface. Neither leads to endothelial cell dysfunction. Pigment deposits occur more commonly with pigment dispersion syndrome (a Krukenburg spindle and associated with open angle glaucoma), Fuchs' endothelial dystrophy, in heavily pigmented people, and following intraocular surgery with iris manipulation.
- Downgrowth. Cellular downgrowth on the endothelium can occur following a penetrating wound, either surgical or nonsurgical. It especially occurs when there is a history of wound leakage and when the wound is through the peripheral cornea or limbus. Two types of downgrowth occur – fibrous or epithelial. The area of downgrowth usually has a distinct border, arises from the wound site, and spreads over the cornea and often into the angle and onto the iris. It is a disease with a high risk of sight-threatening complications.
- Depositions. Some systemic drugs, such as chlorpromazine, can precipitate on the endothelium leading to scattered deposits.

SYMPTOMS

Only a few of the endothelial opacities lead to symptoms themselves, most symptoms being related to the underlying disease. Disruption to normal endothelial pumping function, as seen with Fuchs' endothelial dystrophy, marked posterior polymorphous dystrophy, and downgrowth, leads to blurring of vision, often worse in the morning. There may also be some grittiness with foreign-body sensation from microcystic edema.

SIGNS

Close examination of the opacity, the cornea for stromal edema, and the rest of the anterior segment should allow accurate assessment of the nature of the opacity.



INCIDENCE
Endothelial opacities are uncommon in the general population.

SIGNIFICANCE
While some opacities pose no threat to vision, many are associated with sight-threatening disease.

DIFFERENTIAL DIAGNOSIS
Cataract incisions; Corneal graft rejection; Corneal edema; Descemet's membrane breaks; Epithelial and fibrous ingrowth; Fuchs' endothelial dystrophy; Posterior polymorphous dystrophy; Uveitis anterior – overview.

SEE ALSO
Disciform keratitis; Fuchs' heterochromic iridocyclitis; Hypopyon; Iritis; Pseudoexfoliation of the lens capsule.

MANAGEMENT
Management of the underlying cause of the opacity is indicated. This is covered in the article on each specific condition.

Ocular investigations
Specular microscopy, although rarely needed to make the diagnosis of the opacity, may be helpful in quantifying residual endothelial cell number in several conditions such as Fuchs' endothelial dystrophy and following penetrating trauma.



Fig. E-4a Large active (mutton fat) keratic precipitates in acute granulomatous uveitis.

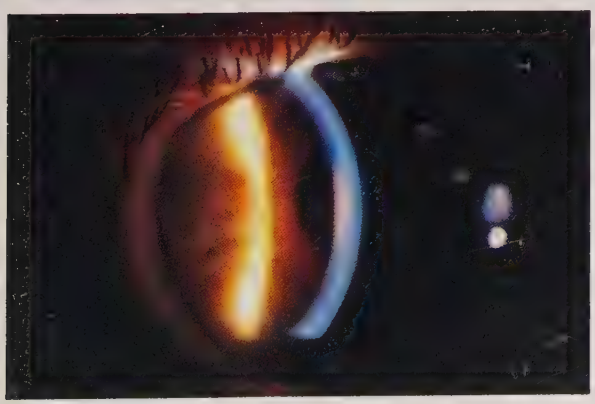


Fig. E-4b Krukenburg's spindle in pigment dispersion syndrome.

E Entropion

DESCRIPTION

Entropion refers to inversion of the eyelid margin; it is less common than ectropion. Entropion may lead to trichiasis and corneal damage from inturned lashes abrading the ocular surface. The causes of entropion are:

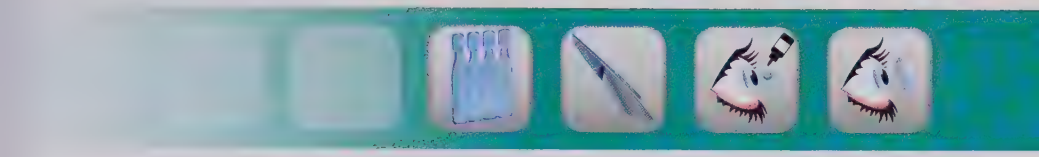
- Involutional (senile). This is the most common cause of entropion and most commonly affects the lower eyelid. It typically occurs in the elderly and results from degenerative changes within the eyelid. These include horizontal lid laxity, laxity of the inferior lid retractors, and overriding of the preseptal (inferior) over the pretarsal (eyelid) orbicularis oculi muscle during lid closure, leading to inwards tilting of the tarsal plate.
- Cicatricial. Scarring of the palpebral and forniceal conjunctiva can pull the lid margin inwards. This is usually only seen with conditions that lead to severe scarring such as chemical burns, Stevens–Johnson syndrome, trachoma, and ocular cicatricial pemphigoid.
- Spastic. This typically occurs in patients with a predisposition to involutional entropion and results from spastic contraction of the orbicularis muscle. This is usually the result of ocular surface irritation and as such may occur after ophthalmic procedures such as cataract extraction.
- Congenital. This very rare cause of lower lid entropion is secondary to maldevelopment of the lower lid retractors. This abnormality also results in the absence of the lower lid crease.

SYMPTOMS

Rubbing of the lashes on the ocular surface leads to irritation, increased lacrimation with occasional epiphora, and blurring of vision.



Fig. E-5 Involutional entropion of the lower eyelid with misdirection of the eyelashes.



SIGNS
The cause of the entropion should be sought by assessing lid laxity, tarsal plate stability, conjunctival scarring, and the presence or absence of a lower lid skin crease. The ocular surface should also be assessed for signs of trichiasis with resultant epithelial disturbance, spanning from a superficial punctate keratopathy through to frank ulceration.

INCIDENCE
Involutional entropion is uncommon to rare although more common in the elderly. The other forms of entropion are all rare.

SIGNIFICANCE
Aside from chronic irritation, entropion may lead to permanent visual loss through corneal scarring and vascularization.

DIFFERENTIAL DIAGNOSIS
Dermatochalasis with misdirection of the lashes of the upper eyelid can give the appearance of entropion. Epiblepharon may be confused with congenital entropion.

SEE ALSO
Ectropion; Ocular cicatricial pemphigoid; Stevens–Johnson syndrome; Chemical burns; Trachoma; Conjunctival scarring; Trichiasis.

MANAGEMENT
Tear supplements
It is important to treat any dry eye that may be associated with cicatricial causes of entropion.

Consulting room procedure
Especially with involutional entropion, some relief may be provided prior to surgery by taping the lower lid. If the area of trichiasis is localized, epilation of selected lashes may also help protect the ocular surface until a more definitive procedure can be performed. Other, usually short-term, measures that can be very effective, especially with spastic entropion, are the insertion of everting (Quickert) sutures or the injection of botulinum toxin.

Therapeutic contact lens
A silicone–hydrogel soft lens may be used to protect the ocular surface from the trichiasis associated with entropion. The lens treatment may be only a short-term measure prior to surgery, if epilation alone cannot control the lash growth.

Surgery
Surgical correction of the underlying cause of the entropion is important to protect the ocular surface and prevent ulceration and microbial keratitis. This may entail tightening of the inferior retractors, horizontal shortening of the eyelid, or inferior conjunctival mucous membrane grafting.

E

Epiblepharon and epicanthic folds

DESCRIPTION

Epicanthic folds and epiblepharon are both common, usually benign, eyelid conditions. They both occur in infants and often resolve with normal childhood development.

- Epicanthic folds. Epicanthic folds are bilateral vertical skin folds adjacent to the medial canthus. They are classified into four categories depending on the location and orientation of the folds:
  - —Superciliaris. The fold runs from the brow down to the lateral aspect of the nose.
  - —Palpebralis. The fold runs symmetrically from the upper to lower lid medially.
  - —Tarsalis. The fold runs from the lateral aspect of the upper lid and extends medially into the medial canthus.
  - —Inversus. The fold runs from the lower lid up to the medial canthus.

  There are racial differences in the occurrence of these folds, with epicanthus palpebralis being common in European babies but usually resolving with age and epicanthus tarsalis being common in Asians and often persisting.
- Epiblepharon. Epiblepharon are an extra horizontal skin fold present adjacent to the lower eyelid margin and are particularly common in Asian infants. The skin fold may press on the lashes leading to vertical misdirection and even trichiasis.

SYMPTOMS

As the children are so young, they do not complain of any symptoms. The parents may present concerned about esotropia in children with epicanthic folds.

SIGNS

With epicanthic folds, it is important to exclude a true esotropia with a thorough examination of the eye and eye movements. With epiblepharon, the ocular surface



Fig. E-6a Epicanthic fold (epicanthus palpebralis).



Fig. E-6b Epiblepharon inferiorly.

should be examined for any evidence of trauma from rubbing of the lashes.

INCIDENCE

Both are very common, especially in Asian infants, while epicanthal folds are relatively common in European infants.

SIGNIFICANCE

Usually clinically insignificant although, rarely, epiblepharon can lead to such marked misdirection of the lashes that it can lead to trichiasis with corneal epithelial abrasion.

DIFFERENTIAL DIAGNOSIS

Epicanthic folds can lead to a misdiagnosis of esotropia, and epiblepharon can be misdiagnosed as the rare condition congenital entropion.

SEE ALSO

Entropion; Trichiasis.

MANAGEMENT

Review

Review in several months is indicated to ensure that the child does not have a true esotropia in the case of epicanthic folds and that the cornea is not traumatized in epiblepharon.

Surgery

Rarely, surgery is performed to correct these skin folds. This is usually for cosmetic purposes with epicanthic folds and as such is performed in teenagers or adults. Surgery for epiblepharon is usually performed in children in the uncommon situation where it leads to trichiasis and corneal epithelial trauma.

Advice

Epiblepharon may resolve spontaneously as the face matures.

E

Epibulbar choristomas

Dermoids, lipodermoids, dermolipomas

DESCRIPTION

A choristoma is a congenital overgrowth of histologically normal tissue occurring at an abnormal site. This is in contrast to a hamartoma, an abnormal overgrowth of normal tissue seen at a site where it normally occurs. Both are benign and arise during fetal development, although they may exhibit a limited degree of growth in early childhood.

Two main types of choristoma occur around the eye: dermoids, comprised mainly of skin elements, and lipodermoids, where there is also significant overgrowth of fat cells. Both may be associated with a rare inherited condition, Goldenhar syndrome. In this syndrome, there are usually pre-auricular skin tags, and there may be associated cardiac and hearing defects.

SYMPTOMS

Lipodermoids are almost always asymptomatic. In contrast, dermoids can lead to several problems other than cosmetic concern. They can interfere with normal lid function, obscure the visual axis, and are frequently associated with induced hypermetropic astigmatism. Occasionally, hairs can grow from their surface and lead to ocular irritation.

SIGNS

Lipodermoids are usually located at the outer canthus and have a gelatinous appearance with a distinct medial border. The lateral border is usually quite indistinct, with the lesion extending superiorly into the orbit.

Dermoids are smooth rounded, usually white- to yellow-colored well-demarcated lesions; occasionally, fine hair may arise from the surface of the dermoid. They most commonly occur at the limbus and are covered with conjunctiva. The conjunctiva is usually immobile over the lesion, and the dermoid is attached to the deep tissue, sclera, or cornea. There is usually a white sclerotic crescent present in the cornea adjacent to limbal dermoids.

Associated signs of Goldenhar syndrome should also be sought, with examination of the external ear and adjacent skin.



Fig. E-7a Limbal dermoid leading to distortion of the lid margin in a child.



INCIDENCE
Rare.

SIGNIFICANCE
Lipodermoids usually have no clinical significance whereas dermoids may be associated with significant loss of vision from obstruction of the visual axis, corneal opacification, or induced (usually hyperopic) irregular astigmatism.

DIFFERENTIAL DIAGNOSIS
The palpebral lobe of the lacrimal gland can, rarely, be mistaken for a lipodermoid; conjunctival mucoepidermoid carcinoma (clear cell tumor).

SEE ALSO
Conjunctival lymphoma; Conjunctival papilloma; Conjunctival intraepithelial neoplasia.

MANAGEMENT
Advice
Patients should be reassured about the benign nature of the lesions and that they generally do not increase in size.

Surgery
Surgery is usually not indicated for lipodermoids because of the risk of damage to lacrimal ductules leading to an aqueous tear deficiency. If surgery is performed, only the exposed portion of the lesion is excised.

Dermoids can be surgically removed if indicated due to lid dysfunction, vision loss, or for cosmetic purposes. With limbal dermoids, this is usually achieved with a shave excision leaving deeper tissue untouched. Dermoids affecting the central cornea can also occasionally be removed with a shave excision, although a corneal graft is sometimes required if there is deep extension of the lesion.



Fig. E-7b Lipodermoid seen laterally.

E Epiphora

DESCRIPTION

Epiphora refers to overflow of tears from the ocular surface. They usually run down the cheek from the medial canthus. However, especially in the elderly with lateral canthal tendon laxity, the tears may overflow from the lateral canthus.

The main causes of epiphora are detailed below:

- Increased lacrimation. A marked increase in tear secretion occurs with reflex tearing. This is triggered by emotional or environmental factors or ocular irritation, including underlying dry eye (paradoxical tearing).
- Decreased outflow. Physical or functional obstruction of the lacrimal outflow system can occur anywhere from the punctum, through the canaliculi, common canaliculus, lacrimal sac, nasolacrimal duct to the outlet of the nasolacrimal duct in the inferior nasal meatus on the lateral wall of the nose. The most common sites of physical obstruction are at the punctum with punctal stenosis, the lacrimal sac, and the nasolacrimal duct. In infants, failure of the lower end of the nasolacrimal duct to canalize is the most frequent cause of epiphora. This can rarely lead to a congenital dacryocele, a cystic dilation of the lacrimal sac.
- Lacrimal pump failure. The lids act to pump the tears medially across the eye toward the puncta and along the canaliculi into the lacrimal sac. Failure of this mechanism, as occurs with medial ectropion and facial nerve paralysis, can also lead to epiphora.

SYMPTOMS

Patients usually complain of a watery eye, slight ocular discomfort, and, frequently, blurring of vision due to an excess tear film. The frequency and extent of epiphora should be sought. Epiphora made worse by cold winds is usually due to anatomical or functional obstruction, whereas epiphora that becomes worse in hot, dry conditions or after prolonged concentration is often due to dry eye.

SIGNS

An increase in the marginal tear strip is observed on slit-lamp examination; this is more obvious with the application of fluorescein. Signs of the cause of epiphora, such as punctal stenosis, ectropion, entropion, and dry eye, should also be sought. Skin excoriation may occur from the overflow of tears.

Clinical tests of lacrimal drainage are detailed in the section "Management."

INCIDENCE

Common, especially in the elderly and infants.

SIGNIFICANCE

Epiphora is usually only a cause of ocular irritation and frustration at the need to constantly dry the eye. It can, however, become a more significant problem if skin excoriation or dacryocystitis occurs.

SEE ALSO

Dacryocystitis; Dry eye syndromes; Bell's palsy; Punctal stenosis; Ectropion; Canaliculitis; Nasolacrimal duct obstruction.

MANAGEMENT

Consulting room procedures

If obstruction of the lacrimal drainage system is suspected, the following tests may be useful in defining the cause.

DILATION AND IRRIGATION (SYRINGING)

After instillation of a drop of local anesthetic, the punctum is dilated with a punctal dilator, and a lacrimal cannula inserted. Sterile saline or water is then injected. If the canaliculus is blocked, fluid will reflux through the same punctum. Blockage to the common canaliculus or more distal structures will lead to efflux of fluid through the other punctum. If the drainage system is patent, fluid will pass through to the back of the throat.

JONES DYE TEST

This is a test for functional blockage. Fluorescein is instilled into the tear film.



After 5 minutes, an anesthetic-soaked cotton bud is placed under the inferior turbinate. If fluorescein is collected, the drainage system is functionally patent.

Ocular investigations

DACRYOCYSTOGRAM

Radio-opaque dye is directly injected into the canaliculi, and X-rays taken immediately and 5 minutes post-injection. This reveals any physical obstruction to the passage of dye.

Lacrimal scintillogram

A radionuclide (usually technetium-99) is instilled into the tear film, and its passage through the lacrimal drainage system followed radiologically. This provides a more physiological test of drainage function and can reveal a functional blockage.

Therapeutics

Any cause of increased tear production, such as ocular surface inflammation or dry eye, should be treated.

Surgery

Surgical intervention with a dacryocystorhinostomy to establish a fistula from the lacrimal sac to the nasopharynx for nasolacrimal duct obstruction, probing for congenital nasolacrimal duct obstruction, or Jones' tube for a blockage to the canaliculi is often indicated. Failure of lid function from an ectropion is also surgically repaired.

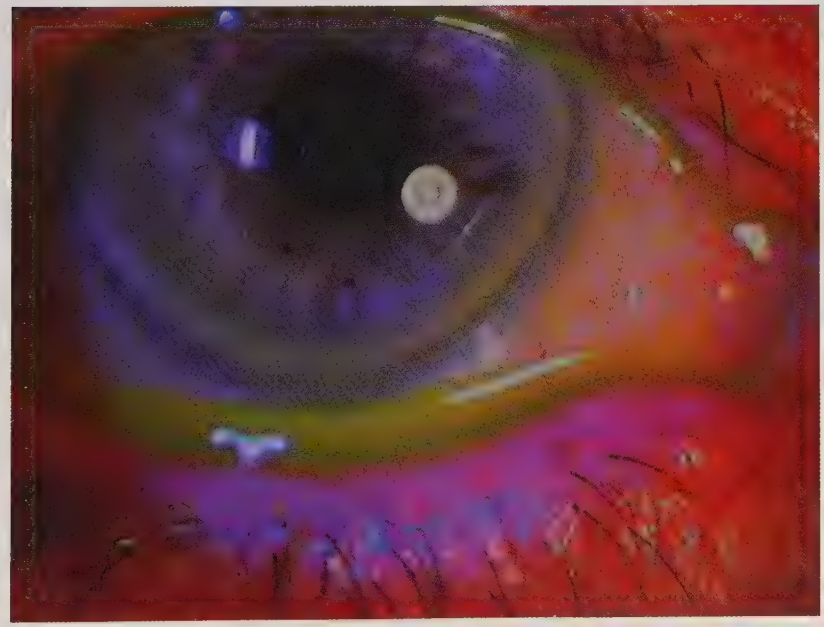


Fig. E-8 Increased marginal tear film stained with fluorescein.

E

Episcleritis

DESCRIPTION

Episcleritis refers to inflammation in the space between the conjunctiva and the sclera involving both the episclera and Tenon's capsule. The key point with respect to episcleritis is its totally non-destructive nature. It does not erode the sclera or lead to corneal changes and, as such, is not sight-threatening. It typically affects young women and is both recurrent and self-limiting. Although most episcleritis is idiopathic it is uncommonly associated with an underlying collagen vascular disease (especially rheumatoid arthritis), viral infection (such as herpes zoster ophthalmicus), or gout.

Three different patterns of episcleritis occur – simple sectorial, simple diffuse, and nodular. Simple sectorial is the most common, followed by simple diffuse, and then nodular.

SYMPTOMS

Episcleritis typically looks much worse than it feels. Despite the redness of the eye, there is normally no pain. However, a slight

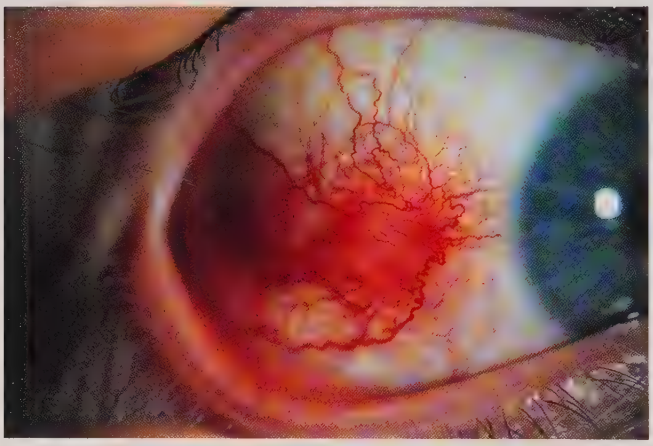


Fig. E-9a Simple sectorial (focal) episcleritis. Note the inflamed vessels are superficial; the deeper scleral vessels visible superior and inferior to the area of episcleritis are uninflamed.

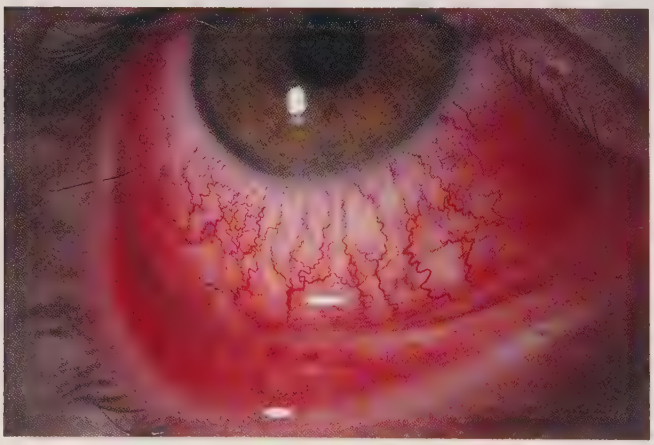


Fig. E-9b Simple diffuse episcleritis.



dull ache is common. Minimal tenderness over the area and some slight lacrimation are also common. Untreated attacks usually last for weeks to a month or two. Nodular episcleritis usually lasts longer than the other forms of the disease.

SIGNS

Simple sectorial and simple diffuse are characterized by an area of redness, dilated episcleral vessels, and slight edema. Nodular episcleritis is focal, with significant edema leading to formation of an inflammatory nodule. Unlike with conjunctivitis, the signs are restricted to the globe, completely sparing the tarsal and forniceal areas.

Features distinguishing episcleritis from scleritis include the level of the edema, the level of the dilated vessels (both best assessed with a slit lamp), and the response to topical vasoconstrictors. With application of dilute phenylephrine drops, episcleral vessels tend to blanch, whereas scleral vessels stay dilated.

INCIDENCE

Relatively common with a strong predisposition for women between the ages of 20 and 50 years; it is uncommon outside this group.

SIGNIFICANCE

Although episcleritis causes irritation, and is frequently a cosmetic concern, it is not sight-threatening. It is only rarely associated with an underlying systemic disease.

DIFFERENTIAL DIAGNOSIS

Scleritis; limbal vernal (vernal keratoconjunctivitis); Foreign body; Pterygium; Pingueculitis. Diffuse episcleritis can look like conjunctivitis.

SEE ALSO

Limbal lesions; Rheumatoid arthritis and the eye; Conjunctival intraepithelial neoplasia.

MANAGEMENT

If mild, no treatment is needed, the attack often resolving spontaneously after several weeks. If the patient is disturbed by the cosmetic effect or ache, treatment may be instituted.

Advice

It is important to reassure the patient that the disease will not damage the eye and as such is not a threat to sight. As this is a chronic relapsing disease, it is also important to stress the risks of long-term corticosteroid usage.

Therapeutics

A short course of a topical corticosteroid, for example prednisolone sodium phosphate qid for 2 weeks, almost always leads to resolution of the current attack. Topical non-steroidal anti-inflammatory agents (such as ketorolac) have also been used to suppress attacks. They have the advantage of avoiding the possible side effects of topical corticosteroids.

Tear supplements

Topical lubrication may be helpful for associated surface irritation.

Oral medication

If attacks are frequent, a 6-month course of an oral non-steroidal anti-inflammatory agent (NSAID) may be prescribed to reduce both the frequency and the severity of attacks. A commonly used regimen is piroxicam 10 mg orally daily.

E

Epithelial and fibrous ingrowth

Epithelial and fibrous downgrowth

DESCRIPTION

Growth of epithelial or fibrous tissue into the eye frequently results in the progressive destruction of intraocular structures. Both are destructive to the eye but epithelial ingrowth is usually a much more aggressive disease. Epithelial and fibrous tissue usually gains access to the anterior segment through a fistulous tract, resulting from either surgical or penetrating trauma. It may even extend along a suture tract. With the development of smaller, improved-profile wounds for cataract surgery and the introduction of better sutures and suturing techniques, the incidence of ingrowth resulting from either surgical or accidental wounds has markedly decreased over the past several decades. Epithelial stem cell theory predicts that cells capable of continuous self-renewal and division only exist in the peripheral cornea and limbus. This is supported by the observation that epithelial ingrowth only occurs from peripheral and limbal wounds.

Once inside the eye, epithelial or fibrous tissue proliferates and spreads first across the anterior segment involving the cornea, angle, iris, and lens. It can then spread into the posterior segment to involve the ciliary processes and retina.

SYMPTOMS

Symptoms depend on the degree and extent of ocular involvement and include a red, photophobic eye and blurred vision.

SIGNS

The key sign of epithelial ingrowth is the recognition of a spreading membrane across the cornea or iris. There is also a history of a surgical or accidental penetrating wound, and there may be a history of a fistulous tract or prolonged reduction in intraocular pressure indicating aqueous leakage. The membrane has a rounded smooth edge; on the cornea, the overlying stroma is usually slightly edematous, and there is microcystic epithelial edema. On the iris, the membrane leads to loss of normal iris architecture and may contract, leading to polycoria and corectopia. Epithelial ingrowth also often results in a few circulating epithelial cells visible in the anterior chamber and resembling inflammatory cells.

The extent of iris involvement can be identified with the use of the argon laser. On hitting normal iris, low power (500 microns × 100 mW × 0.1 s) leads to slight contraction. On an epithelial membrane this leads to a characteristic fluffy gray area. On invading the posterior segment, ingrowth may involve the ciliary processes and retina, leading to a retinal detachment and phthisis bulbi.

The distinction between fibrous and epithelial ingrowth may be difficult to make, especially early on in the course of the disease. In general, fibrous ingrowth does not result in as obvious a membrane as epithelial ingrowth, but does lead to the formation of peripheral anterior synechia and shallowing of the anterior chamber. Over time, the more aggressive and progressive nature of epithelial ingrowth allows the diagnosis to be made with more confidence.

INCIDENCE

Extremely rare following modern intraocular surgery. Most commonly occurs following a penetrating injury involving the limbus or peripheral cornea.

SIGNIFICANCE

Epithelial ingrowth usually leads to eventual loss of the eye. Fibrous ingrowth is less destructive but may lead to glaucoma and cataract formation.

DIFFERENTIAL DIAGNOSIS

Iridocorneal endotheliopathy; Iridocorneal dysgenesis; Aniridia; Trauma.

SEE ALSO

Perforation – corneal; corneal/conjunctival lacerations; Cataract incisions.



MANAGEMENT

Medical treatment is usually inadequate for managing epithelial ingrowth, with early aggressive surgical treatment being necessary unless the disease is very limited. Fibrous ingrowth is much less aggressive and may only lead to glaucoma due to the formation of extensive peripheral anterior synechia with the preservation of other anterior segment structures.

Therapeutics

Anti-glaucoma medication is frequently required to control both fibrous and epithelial ingrowth, although in the long-term medical treatment is usually insufficient.

Surgery

Early, often quite destructive, surgery was popularised in the mid-1970s as a way of preserving affected eyes. This often involved a broad iridectomy, a corneal graft, lens extraction, and anterior segment cryotherapy. Even with such aggressive surgery the risk of losing the eye is quite high. Glaucoma filtering surgery frequently fails due to epithelial invasion of the trabeculectomy site; for this reason a filtering seton may be required.

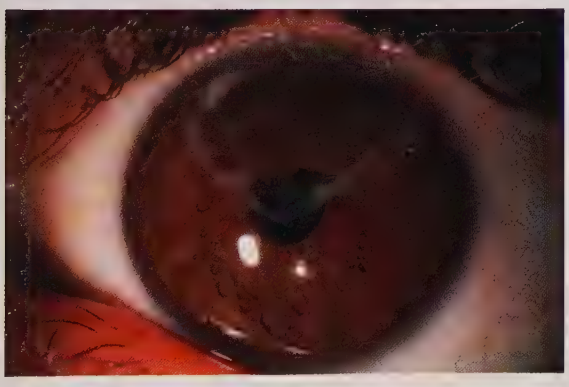


Fig. E-10a Epithelial ingrowth following cataract surgery with a superior wound; there is a membrane on the endothelial surface of the cornea extending from the superior limbus with the inferior edge of the membrane visible through the pupil.

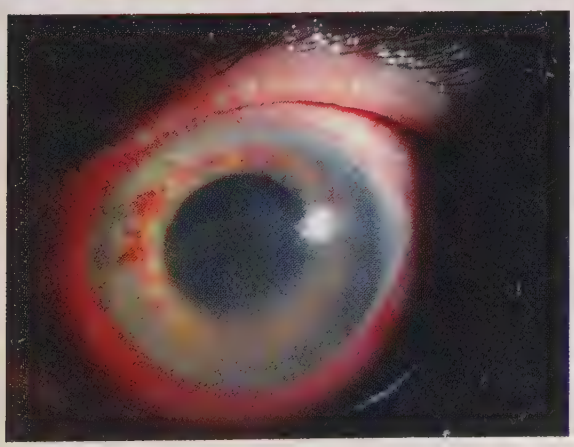


Fig. E-10b Epithelial ingrowth with associated focal inferior corneal edema. The edge of the membrane is also visible through the pupil.

E

Epithelial basement membrane dystrophy (EBMD)

Map–dot–fingerprint dystrophy, Cogan's microcystic corneal dystrophy

DESCRIPTION

This common corneal dystrophy affects the corneal epithelium leading to characteristic cystic and whorl-like opacities. It shows no particular gender preference, and inheritance is very variable. Histology reveals several unique epithelial changes that occur in this condition:

- Microcysts are due to small cystoid spaces in the epithelium into which otherwise normal superficial corneal epithelial cells desquamate.
- Dots (putty spots) are areas of desquamated cells under an area of basement membrane reduplication.
- Fingerprints are due to folds of reduplicated basement membrane within the corneal epithelium.
- Maps are due to changes not in the epithelium but in the anterior stroma (Bowman's membrane) with the formation of both new basement membrane and collagen.

SYMPTOMS

Most patients are symptom-free throughout life, with the condition only being found on routine examination. About 10% of patients develop problems with recurrent corneal erosions. This is uncommon in patients less than 30 to 40 years of age. Typical symptoms include ocular irritation with acute exacerbations that may last minutes to hours. The pain experienced with these exacerbations is usually quite

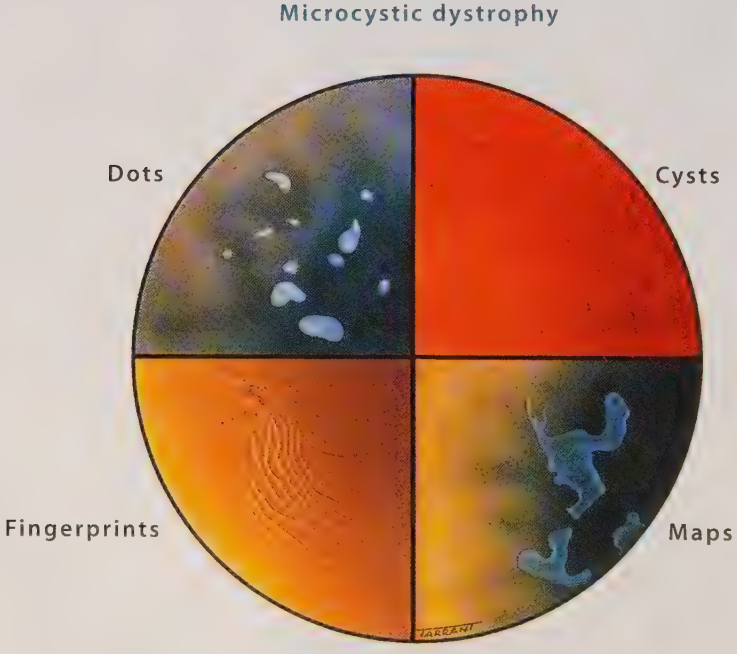


Fig. E-11 Four types of epithelial changes seen in epithelial basement membrane dystrophy.



severe and comes on either in the early hours of the morning or on waking. Vision is usually blurred for days to weeks following a significant attack. Rarely, the maps and fingerprints are themselves so marked that they slightly decrease vision.

SIGNS

There are four epithelial changes that occur in this condition – microcysts, dots, maps, and fingerprints. Microcysts are much more commonly seen than dots, maps, and fingerprints and are commonly the only sign present. The signs can change with time, are bilateral, and may be very variable, ranging from difficult-to-see grouped microcysts to easily identified widespread maps and fingerprints. While all these signs may be subtle, the microcysts are particularly easy to miss. They occur most commonly inferiorly and are best seen with high-magnification and on retroillumination with a dilated pupil. Once observed it may be useful to ask the patient to blink to ensure the observed disturbances are not just present in the tear film.

INCIDENCE

This is probably the most common of any corneal dystrophy.

SIGNIFICANCE

Although EBMD may be asymptomatic, and only noted as an incidental finding, it may be associated with recurrent corneal erosion syndrome, ocular irritation, or contact lens intolerance.

DIFFERENTIAL DIAGNOSIS

Meesmann's dystrophy; Reis–Bücklers dystrophy; corneal epithelial microcystic edema; corneal scarring.

SEE ALSO

Recurrent corneal erosion syndrome; Corneal edema; Vortex keratopathy.

MANAGEMENT

Tear supplements

Ocular irritation is treated with topical lubricants. Recurrent corneal erosions are initially treated with node lubricant ointment or gel. This needs to be continued for several months after the last recurrent erosion attack.

Therapeutic contact lens

A therapeutic (bandage) contact lens can be fitted at the time of an epithelial erosion to both provide pain relief and facilitate healing. It is used on an extended-wear basis for up to a week. As the lens is worn when there is a pre-existing epithelial defect, it does heighten the risk of microbial keratitis and as such is usually accompanied by prophylactic topical antibiotics. A bandage lens probably has no long-term benefit with regard to preventing subsequent erosions.

Surgery

Several surgical interventions have been employed to stabilize the epithelium and prevent further attacks. The easiest technique is simple debridement of the affected area under topical anesthesia. Superficial micropuncture with a bent needle has also been tried and appears to be relatively effective. Several studies have also reported success with debridement coupled with alcohol delamination. There is a similar success rate of around 30–50% for debridement alone and micropuncture, slightly higher for debridement coupled with alcohol delamination.

Several studies have also demonstrated a superficial phototherapeutic keratectomy (PTK) (8 to 10 microns deep) to be about 80% successful. If successful, late recurrences are uncommon. Treatment failures tend to occur quickly, within the first few weeks to months following surgery and can be treated with a repeat PTK with a high rate of success.

E

Exposure keratopathy

Lagophthalmos

DESCRIPTION

The eyelids are critically important in maintaining the tear film and, consequently, the health of the ocular surface. They act both to contain the tears within the palpebral fissure and to spread the tears across the cornea and bulbar conjunctiva. Interruption to the function of the eyelids interferes with this mechanism and leads to local drying and typical ocular surface changes collectively known as exposure keratopathy.

There are three main disturbances in eyelid function:

- Decreased blink rate. This occurs in several degenerative brain diseases such as dementia, Alzheimer's disease, and Parkinson's disease.
- Incomplete blink (lagophthalmos). This occurs with facial nerve palsies, orbital diseases with proptosis, and lid diseases such as ectropion. There may also be associated inferior eyelid ptosis. Nocturnal lagophthalmos refers to the eyes being partially open while sleeping. Exposure may also occur from inadequate eyelid closure during a prolonged general anesthetic.
- Lid retraction. This occurs with proptosis and dysthyroid ophthalmopathy. It results in an unstable tear film, especially inferiorly.

With nerve palsies, the ocular surface sensation may also be decreased, leading to a neurotrophic keratopathy. The coexistence of exposure and a neurotrophic keratopathy represents a marked risk to the ocular surface.

SYMPTOMS

Disturbance to the tear film and ocular surface from inadequate lid function leads to discomfort with grittiness and burning and often increased lacrimation. Blurring of vision can result from tear film disturbances, epithelial changes, and increased lacrimation.

SIGNS

The hallmark of exposure keratopathy is inferior epithelial disturbance. There is an unstable tear film with a decreased tear break-up time (TBUT) and associated epitheliopathy. Initially, there are superficial punctate erosions affecting both the corneal and the conjunctival epithelium. These may coalesce into an area of epithelial disturbance. Ulceration of the epithelium may occur, with the typical ulcer being inferior and oval; the long axis of the



Fig. E-12a Upper and lower lid retraction with associated lid swelling and conjunctival chemosis in a patient with dysthyroid ophthalmopathy, one of the causes of exposure keratopathy.

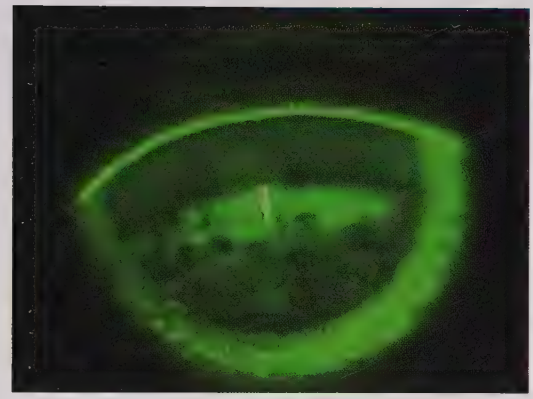


Fig. E-12b Moderate corneal fluorescein staining typical of lagophthalmos, with the characteristic superior sparing of epithelial disturbance.

oval defect is usually horizontal. The edge of the defect is usually smooth and slightly heaped up. If chronic, stromal loss with thinning of the cornea may occur. There is also an increased risk of microbial keratitis, usually with a Gram-negative bacterium.

INCIDENCE
Uncommon, although more commonly seen in the elderly.

SIGNIFICANCE
Usually results in some blurring of vision and may lead to permanent vision loss through scarring and infection.

DIFFERENTIAL DIAGNOSIS
Neurotrophic keratopathy; Dry eye syndromes.

SEE ALSO
Bell's palsy; Ectropion; Superficial punctate keratopathy; Atopic keratoconjunctivitis; Persistent epithelial defects.

MANAGEMENT
Tear supplements
These can help to stabilize the tear film; ointments and gels are especially useful at night to prevent drying from nocturnal lagophthalmos.

Temporary corneal protection
If recovery of lid function is anticipated, as can occur with facial nerve damage during neurosurgery, temporary corneal protection is required. Therapeutic (bandage) contact lenses are not usually used to protect the epithelial surface in exposure keratopathy. Desiccation of the lens and inadequate lid function to move the lens can result in increased epithelial damage and may even predispose to microbial keratitis. Taping of the eyelid closed at night or even during the day may be necessary.

Surgery
If permanent, surgical correction may be indicated. Correction of any specific cause of the exposure should be carried out at first. This includes ectropion repair, upper lid gold implants for lid retraction, and orbital decompression for dysthyroid ophthalmopathy proptosis.

If exposure persists, a lateral tarsorrhaphy can be extremely effective in protecting the ocular surface.

E

Eyelid trauma

Laceration, hematoma, canalicular laceration, lid contusion

DESCRIPTION

Eyelid trauma is typically caused by either a blunt or a sharp injury. Blunt injuries result in a hematoma (black eye) while sharp injuries lead to an eyelid laceration. Both may be associated with significant ocular or periocular tissue involvement. Sharp injuries may lead to ocular penetration, while blunt trauma may result in non-penetrating damage to the globe and orbit. If severe, as occurs in motor vehicle accidents, assault, and some sporting injuries, a blunt injury may lead to a fracture of an orbital wall (blow out) or of the skull.

Sharp injuries characteristically lead to a skin laceration; this may be superficial or full thickness and may damage the proximal lacrimal drainage system, punctum, and canaliculus.

SYMPTOMS

Damage to the lacrimal drainage system may lead to epiphora whereas an orbital fracture can result in diplopia. Diplopia may also result from an orbital hemorrhage or bruising of an extraocular muscle. Blurring of vision may be due to lid trauma with increased lacrimation and ptosis or may result from damage to intraocular structures. A skull fracture may lead to an altered conscious state.

SIGNS

The affected area should be examined for skin lacerations, areas of swelling, and, in particular, for any damage to the canalicular system.

A thorough examination of both the globe and periocular tissue is also indicated. This includes measurement of the intraocular pressure, slit-lamp examination to assess the presence of



Fig. E-13 Lower eyelid hematoma with swelling, skin discoloration, and associated subconjunctival hemorrhage.



a hyphema, or lens subluxation and gonioscopy for angle recession. Assess pupil reactions and facial sensation for integrity of the cranial nerves. Dilated fundal examination should be performed to assess retinal or optic nerve damage. Ocular movements should be assessed, specifically looking for signs of an orbital wall fracture with restriction of movement or diplopia, especially with upward or lateral gaze.

A base of skull fracture usually leads to characteristic bilateral eyelid hematomas, termed a "panda" or "raccoon" sign.

INCIDENCE

Common, especially seen with assault and from sporting injuries.

SIGNIFICANCE

A full thickness laceration may lead to chronic epiphora or ocular surface irritation secondary to trichiasis from a lid margin defect.

DIFFERENTIAL DIAGNOSIS

Orbital floor fracture; basal skull fracture; Preseptal cellulitis.

SEE ALSO

Sub-conjunctival hemorrhage; Hyphema (blunt trauma); Epiphora; corneal and conjunctival lacerations.

MANAGEMENT

Additional investigations

If there is any suggestion of a possible retained foreign body or orbital wall or base of skull fracture, radiological investigations with a plain X-ray of the skull and CT scan should be performed. An MRI may then be indicated to further define the injury.

Advice

An eyelid hematoma resolves spontaneously but may benefit from application of a cold compress. The patient should be advised that, as the eyelid hematoma resolves, the area of skin discoloration usually migrates inferiorly with a sharp inferior arcuate border, following the insertion of the septum orbitale.

Review

Review at 1, 6, and 12 months and even on a yearly basis is indicated following significant trauma, especially with angle recession, to ensure that there is no late rise in intraocular pressure from damage to angle structures.

Surgery

A full-thickness skin laceration, canalicular laceration, or orbital wall fracture usually requires surgical repair.

F

Filamentary keratopathy

Corneal filaments, mucous filaments

DESCRIPTION

Damage to the corneal epithelium coupled with tear film dysfunction can lead to the presence of mucous filaments. These filaments are composed of both degenerate epithelial cells and mucus.

The most common causes of a filamentary keratopathy are severe dry eye and post intraocular surgery with filaments attached to a wound or sutures. This is most commonly seen in the first few weeks following a corneal graft. Other causes include neurotrophic keratitis, superior limbic keratoconjunctivitis, drop toxicity (toxic keratopathy), and recurrent corneal erosion syndrome.

SYMPTOMS

Corneal filaments lead to marked ocular surface irritation with foreign body sensation. The marked discomfort associated with filamentary keratopathy is due to the tugging on the corneal epithelia that occurs from movement of the filament during normal blinking and eye movement. There is often associated photophobia and blurring of vision. There may also be epiphora from an increase in reflex tearing.

SIGNS

The filaments appear as grayish comma-shaped lesions that move in the tear film with blinking. They are anchored to the epithelium by a narrow strand and usually thickest and globular at the distal end; they are typically from 1 mm to several mm in length. Due to their mucous composition, they stain freely with rose bengal but poorly with fluorescein. Their position should be noted as it can help identify the cause. Inferior filaments are more common with dry eye; postoperative strands are usually over sutures; filaments arising from superior limbic keratitis usually involve the superior cornea.

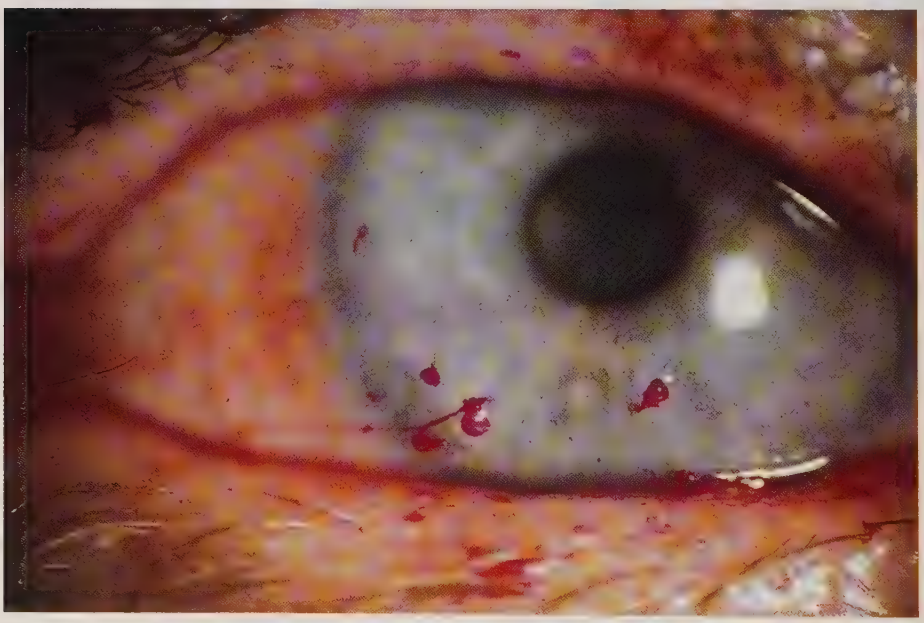


Fig. F-1 Several corneal filaments stained with rose bengal inferiorly in a patient with severe dry eye.



There is often some reactive ptosis and, unless the filament is secondary to dry eye, an increase in the tear film. Signs of the underlying cause should also be sought.

INCIDENCE
Uncommon.

SIGNIFICANCE
Leads to significant ocular surface discomfort, photophobia, and tearing.

SEE ALSO
Dry eye syndromes; Neurotrophic keratopathy; Toxic keratopathy; Superior limbic keratoconjunctivitis.

MANAGEMENT
In addition to treating the filaments, it is important to treat the underlying cause to prevent recurrence.

Removal
The filaments can be removed with either forceps or a cotton bud soaked in anesthetic. This usually leads to a focal epithelial defect at the site of removal. Consequently, topical antibiotics are usually used for several days following removal. If the filament is attached to a loose corneal suture this should also be removed.

Tear supplements
As most filaments are associated with a tear film disturbance, tear supplements are indicated. Punctal plugs can also be of significant benefit in restoring normal tear film function.

Therapeutics
Topical corticosteroids may be useful in reducing ocular surface inflammation and, consequently, filament formation. Topical acetylcysteine, a mucolytic agent, has also been used to dissolve filaments, although it is rarely used long-term due to its significant stinging on instillation, and rapid degradation in the bottle.

Dosing: G. fluorometholone 0.1% bd, G. acetylcysteine 10% qid.

Therapeutic contact lens
If the filamentary keratopathy is refractory to other measures, a bandage contact lens may be very helpful in breaking the cycle of epithelial damage and filament formation.

F

Filtering bleb

Trabeculectomy, drainage bleb, glaucoma procedure

DESCRIPTION

The intraocular pressure (IOP) may be lowered via the establishment of an outflow channel (fistula) between the anterior chamber and the sub-Tenon's space (a trabeculectomy). This leads to the development of an elevated filtering or drainage bleb adjacent to the limbus. The procedure is usually performed superiorly; at the same time a surgical peripheral iridectomy is performed to prevent the iris from blocking the outflow channel. The aqueous humor drained via the bleb either is removed via absorption through conjunctival veins or lymphatics or, if the overlying conjunctiva is thin, may pass directly through the conjunctiva into the tear film. As the outflow channel can heal over with postoperative scarring, adjunctive treatment with antimetabolites such as 5-fluorouracil (5-FU) or mitomycin C (MMC) is frequently used. This leads to a reduction in fibrotic activity (scarring) and increases the success rate of the procedure. These antimetabolites, especially mitomycin C, also typically lead to thinner-walled blebs more prone to infection and rupture.

Postoperative complications are relatively common, and fall into two main categories.

Early postoperative complications

- Shallow anterior chamber: this is common following surgery and usually rectifies itself. If persistent, it can lead to peripheral anterior synechia, cataract, endothelial dysfunction from lenticulo-corneal touch, choroidal effusions, and hypotonous maculopathy. Causes of a shallow anterior chamber include excessive filtering, wound leak, pupil block, and malignant glaucoma (aqueous misdirection syndrome).
- Endophthalmitis
- Persistent corneal epithelial defects: this is more common when adjunctive antimetabolites have been used.

Late postoperative complications

- Bleb failure: failure of filtration can occur if the drainage channel is scarred over or blocked with vitreous or iris. In addition the bleb can become encysted (encapsulated) or fibrosed.
- Cataract formation: early nuclear sclerosis commonly develops.
- Blebitis/endophthalmitis: bacterial infection within the filtering bleb is termed blebitis. This is more common in patients with a thin-walled, cystic bleb associated with adjunctive antimetabolite usage. Blebitis may rapidly evolve to endophthalmitis.

SYMPTOMS

Patients with a bleb may be aware of a slight foreign-body sensation. This

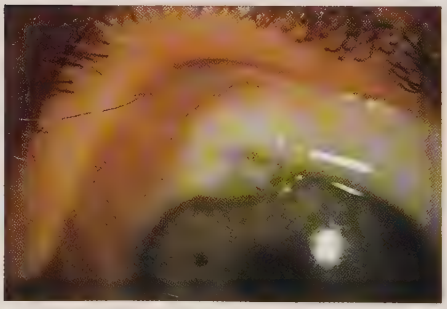


Fig. F-2a Thin-walled, polycystic bleb with whitening of the underlying sclera, typically seen with adjunctive mitomycin C treatment.

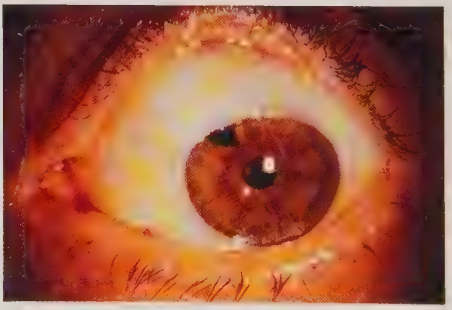


Fig. F-2b Thin, flat, diffuse filtering bleb associated with good IOP control.



is especially the case with a cystic or encapsulated bleb. They may also develop symptoms associated with the above complications with blur, ache, increased lacrimation, and photophobia.

SIGNS

There are several different morphological types of filtering blebs; some are associated with better control of the IOP. Thin-walled polycystic blebs are usually effective in lowering IOP but are associated with an increased risk of blebitis. Flat, thin, diffuse, relatively avascular blebs are also associated with good IOP control. Flat, vascular, fibrosed blebs without an underlying subconjunctival space are usually non-filtering and as such ineffective with regard to IOP control. An encysted bleb is characterized by an elevated cyst-like bleb and occurs secondary to hypertrophy of Tenon's capsule; this is also non-filtering. Blebitis is characterized by an inflamed conjunctiva, whitened bleb often containing inflammatory debris, and, frequently, slight anterior chamber cell and flare. In addition to evaluating the bleb, the IOP should be recorded, and the anterior chamber assessed for shallowing or inflammation. Seidel's test may be performed if leakage of aqueous from a thin-walled bleb is suspected.

INCIDENCE

Rare. Although glaucoma has an incidence of approximately 5%, more effective medical management with newer anti-glaucoma topical medication has recently led to a decrease in the number of filtering surgeries being performed in many countries.

SIGNIFICANCE

A patient with a filtering bleb is always at risk of bleb failure and an increase in IOP. A thin-walled bleb is always at risk of developing blebitis.

SEE ALSO

Endophthalmitis; Hypopyon; Glaucoma – acute angle closure; Cataract incisions.

MANAGEMENT

Advice

The patient should be advised to re-present promptly if there is any reduction of vision or if the eye becomes inflamed.

Review

Six-monthly review for glaucoma assessment is appropriate.

Therapeutics

Topical antibiotics and corticosteroids are used immediately postoperatively. Topical anti-glaucoma medication may also need to be used to maintain adequate pressure control.

Surgery

Occasionally, additional surgery is needed to maintain bleb function with 5-FU injections, YAG-laser, or even surgical revision of the bleb.

F

Flash burns

Photokeratopathy, snow blindness, actinic keratitis

DESCRIPTION

Ultraviolet light (UV) can acutely damage the corneal epithelium. The most damaging wavelengths are between 260 and 290 nm, although with prolonged exposure longer wavelengths can also lead to corneal damage. As the ozone layer allows essentially no UV light below 290 nm to enter the Earth's atmosphere, the major sources of damaging, low-wavelength UV are man-made. An exception to this is snow blindness, where altitude, reflection from the snow, and prolonged exposure combine to lead to a photokeratopathy.

The most common man-made sources are arc welders, UV suntanning lamps, and from shorting of an electrical board.

SYMPTOMS

The eyes feel gritty and sore with a sandy foreign-body sensation. There is usually marked lacrimation, photophobia, and lid swelling. Pain may be intense, and there is blurring of vision from both the increased lacrimation and epitheliopathy. Symptoms typically begin 8–12 hours following exposure. Therefore, patients often present to an emergency department in the middle of the night having been woken by the pain.

SIGNS

Involvement is usually bilateral with marked lid swelling and tearing. There may also be signs of skin damage from the UV exposure with redness and swelling. Both the conjunctival and corneal epithelia display a widespread punctate keratopathy, staining freely with fluorescein. The area of staining is restricted to the exposed interpalpebral area. As such, the forniceal conjunctiva and superior cornea is



Fig. F-3 Conjunctival hyperemia and chemosis in a patient with a recent welding flash exposure.



uninvolved. Rarely, with very severe UV exposure, stromal swelling and even an anterior uveitis can occur.

INCIDENCE
Common, especially among welders, electricians, and beginner skiers.

SIGNIFICANCE
Heals within 2 to 5 days without scarring and has no long-term effects on vision.

DIFFERENTIAL DIAGNOSIS
Toxic epitheliopathy.

SEE ALSO
Toxic keratopathy; Superficial punctate keratopathy.

MANAGEMENT
Advice
It is important to reassure the patient that the vision will recover fully and that there will not be any adverse long-term effects. An explanation of the nature of the injury is important to reduce the possibility of a repeat injury. Advice should also be given with regard to the importance of suitable protective eyewear.

Tear supplements
Antibiotics are not needed as the epithelial damage is usually not full thickness and recovers quickly. Tear supplements, ice packs, padding, and sunglasses may help with pain relief.

F

Floppy eyelid syndrome

DESCRIPTION

Floppy eyelid syndrome is a rare and frequently misdiagnosed condition capable of leading to significant upper eyelid laxity and conjunctival and corneal damage. It is thought to arise from spontaneous eversion and trauma to the upper eyelid during sleep. It is highly associated with sleep apnea and typically affects obese middle-aged men. The condition may be unilateral or bilateral and is usually chronic. It may also affect patients with injuries requiring them to always sleep on one side. In this case the condition is unilateral, involving the same side as that on which the patient sleeps.

The association with sleep apnea is of particular importance because of its association with sudden unexplained death. Patients with sleep apnea usually report a history of snoring and poor quality sleep, waking without feeling refreshed.

SYMPTOMS

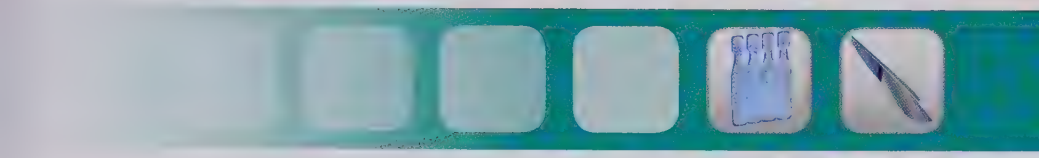
The affected eye is gritty and sore; there may be some blurring of vision and associated increased lacrimation. Symptoms are typically chronic and fluctuate over time.

SIGNS

The upper lid lashes typically point downwards (lash ptosis) and lose their normal regular orientation, becoming crossed and overlapping. The upper lid usually displays marked horizontal laxity especially affecting the medial canthal tendon. The upper lid is extremely easy to evert; eversion may occur simply by pulling on the supraorbital skin. The superior tarsal conjunctiva is markedly inflamed with a beefy red papillary reaction. Corneal complications are common with a superior superficial punctate keratopathy, filamentary keratopathy, and peripheral



Fig. F-4 Demonstration of upper eyelid laxity with associated papillary conjunctivitis with floppy eyelid syndrome.



pannus formation having all been reported. Several cases of floppy eyelid syndrome being associated with keratoconus have also been reported.

INCIDENCE
Rare.

SIGNIFICANCE
Can lead to sight-threatening corneal complications with scarring and epithelial defects. Floppy eyelid syndrome and sleep apnea have been strongly associated with the development of keratoconus.

DIFFERENTIAL DIAGNOSIS
Rosacea keratitis; Dry eye syndrome; chronic Canaliculitis; chronic Dacryocystitis; Toxic epitheliopathy; eyelid imbrication syndrome (the upper eyelid overriding the lower eyelid).

SEE ALSO
Giant papillary conjunctivitis; Filamentary keratopathy.

MANAGEMENT
The floppy eyelid syndrome should be brought under good control prior to corneal graft surgery if there is associated keratoconus.

Sleep laboratory investigations
All patients with floppy eyelid syndrome should be investigated for sleep apnea at a sleep laboratory. These have become more common over the past decade as the importance of sleep apnea and other sleep disturbances has been realized.

Non-drug treatments
Treatment of sleep apnea with nasal continuous positive airway pressure (CPAP) usually results in significant improvement in the floppy eyelid symptoms and signs. If the patient is obese, weight loss may also help. Other treatments include the use of a shield at night to prevent rubbing and eversion of the lid, lubricant ointments to reduce friction between the lid and cornea, and taping of the lids at night.

Surgery
If treatment of the sleep apnea is insufficient, a horizontal lid-shortening procedure prevents spontaneous eyelid eversion. The effect is usually only temporary unless the underlying sleep apnea is also treated.

F Follicular conjunctivitis

DESCRIPTION

Conjunctival follicles result from hyperplasia of local lymphoid tissue. They are the conjunctival equivalent of an inflamed lymph node and often occur in association with inflammation of the draining preauricular and submandibular lymph nodes. They are important clinically as they only occur in a limited number of pathological conditions. However, a few scattered follicles, especially in the inferior fornix, are common and of no clinical importance. Children especially may have inferior forniceal follicles in the absence of any disease.

Pathological causes of a follicular conjunctivitis can be grouped into four categories.

Viral infections

Primary herpes simplex conjunctivitis (keratoblepharoconjunctivitis), molluscum contagiosum infections around the eyelid and brow, adenoviral conjunctivitis (EKC), chicken pox lesions around the eye.

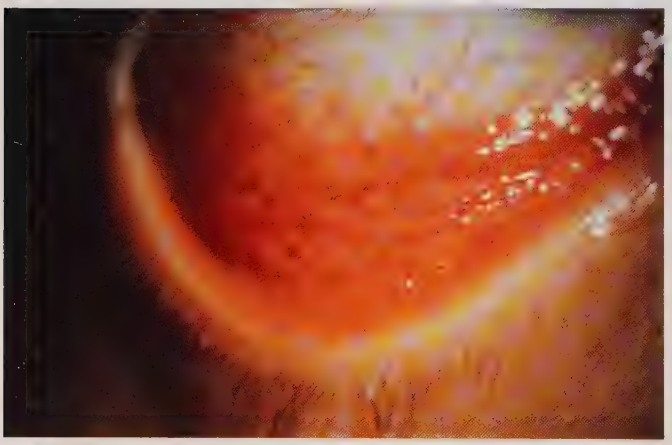


Fig. F-5a Conjunctival follicles: rows of multiple, discrete, smooth, pink-to-yellow elevated lesions.



Fig. F-5b In contrast, an example of a marked papillary reaction on the superior tarsal conjunctiva.



Chlamydial infection
The adult and childhood forms of chlamydial infection around the eye, adult inclusion conjunctivitis and trachoma, both lead to a follicular conjunctivitis. The neonatal chlamydial infection, neonatal chlamydial conjunctivitis (a cause of ophthalmia neonatorum), does not lead to follicles, as newborns are unable to form follicles until several months of age.

Therapeutics
Follicular conjunctivitis may occur from a chronic low-grade allergic response to topical medication; this is most commonly seen with anti-glaucoma medication.

Parinaud's oculoglandular syndrome
This is an uncommon type of conjunctivitis with associated gross lymphadenopathy. It occurs secondary to a number of granulomatous infections and inflammations, most commonly sarcoid and cat scratch fever.

SYMPTOMS
Most causes of follicular conjunctivitis are associated with a slightly red eye, watery discharge, and slight irritation and discomfort.

SIGNS
Follicles are best seen in the inferior temporal fornix, although they are usually also present on the superior and inferior tarsal conjunctiva. They are uncommon on the bulbar conjunctiva although they do occur at the limbus in trachoma. They appear as rows of multiple discrete smooth pink-to-yellow elevated lesions. They are largest in the fornix and smaller on the tarsal conjunctiva. Each follicle is surrounded by displaced blood vessels.

Follicles are most commonly confused with papillae, although the two are readily distinguished. Papillae result from epithelial hyperplasia and edema and contain a vascular core; they are usually smaller and much redder than follicles. A papillary reaction is much less specific and therefore of less diagnostic importance than is a follicular reaction.

Other features that should be looked for with a follicular conjunctivitis are local lymphadenopathy, keratitis, molluscum lesions around the eye, and herpetic vesicles on the skin.

INCIDENCE
Follicular conjunctivitis is common and, due to the infectious nature of many of its causes, may occur in epidemics.

SIGNIFICANCE
There is usually a specific underlying cause for follicular conjunctivitis, including several that are chronic and require systemic treatment.

DIFFERENTIAL DIAGNOSIS
Papillary conjunctivitis; conjunctival granuloma (in sarcoidosis).

SEE ALSO
Herpes simplex primary keratoblepharoconjunctivitis; Molluscum contagiosum; Chlamydial conjunctivitis; Adenoviral keratoconjunctivitis; Trachoma; Conjunctivitis – overview.

MANAGEMENT
Management of the individual causes of follicular conjunctivitis are listed under the specific diagnosis.

Microbiology
Unless the cause is apparent it is important to perform microbiological investigations to exclude chlamydial conjunctivitis.

F Foreign bodies – corneal and conjunctival

DESCRIPTION

The eye is relatively protected from contact with foreign bodies by the brow, cheek, eyelashes, and eyelid. It is also protected by the action of the lids and Bell's phenomenon, acting together to prevent anything but fast moving objects from hitting the unprotected cornea. As such, foreign bodies most commonly hit the eye when travelling at high speed, as occurs with drilling or grinding. Small windborne particles may also escape these protective mechanisms and become embedded in the ocular surface.

Foreign bodies are commonly classified into several groups: metallic or non-metallic, embedded or superficial, and penetrating or non-penetrating.

SYMPTOMS

A foreign body on the ocular surface leads to lacrimation, redness, and irritation and may cause blurring of vision. The degree of these symptoms depends on the size, depth, location, and composition of the foreign body.

SIGNS

Slit-lamp examination is performed to determine the depth of the foreign body. The inferior and superior fornices and tarsal conjunctiva should also be examined. Fast-moving metallic foreign bodies usually become embedded in the cornea and lead to a focal scalloped out area of stromal loss. If the foreign body contains iron, and is left for hours or days, an associated rust ring may develop.

While most are fairly superficial, full-thickness penetration of the cornea can occur, especially with high-speed injuries (e.g. drilling and grinding). Signs of intraocular penetration include shallowing of the anterior chamber, a lowered intraocular pressure, anterior chamber inflammation, cataract, pupil distortion, and egress of aqueous humor from the wound site. With ocular penetration, aqueous humour passing out through the wound results in an obvious dilution of fluorescein and is easily seen on a slit lamp with a cobalt blue filter in place. This is termed a positive Seidel's sign.

INCIDENCE

Very common.

SIGNIFICANCE

Usually no long-term effect on sight. Deep central foreign bodies may lead to some permanent loss of vision and glare problems from scarring. This results from induced focal irregular astigmatism and opacity.

SEE ALSO

Abrasion; Recurrent corneal erosion syndrome; Perforation – corneal.

MANAGEMENT

Advice

Patients should be advised of the need for adequate protective eyewear to prevent further, possibly more severe, injuries.

Therapeutics

If there is an epithelial defect after removal of the foreign body, a broad-spectrum topical antibiotic is usually administered qid for several days.

Removal of the foreign body

Superficial, non-embedded corneal and conjunctival foreign bodies can be irrigated away or manually wiped off with a cotton bud after the instillation of a drop of topical anesthetic into both eyes.

Embedded corneal foreign bodies can be removed either by wiping with a cotton bud or if necessary carefully lifted off the cornea with the tip of a 25-gauge needle. The needle can be mounted on a cotton bud to allow easier handling and, under slit-lamp control, is advanced to the foreign body parallel to the iris plane and used to lift out the foreign body. The tip of the needle may be partially bent over to facilitate hooking out the foreign body. This can be achieved by pressing the tip of the needle against its protective plastic sheath.

If there is an associated rust ring, this can be removed either with the needle, if very superficial, or with a dental burr if the rust has penetrated the superficial stroma.

Laser surgery

An excimer laser phototherapeutic keratectomy (PTK) is rarely indicated to treat post-traumatic corneal scars. They are usually too deep and removal can lead to both corneal haze and a significant hyperopic shift.

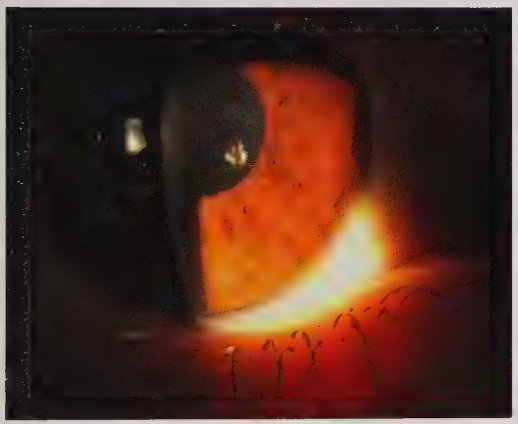


Fig. F-6a Central corneal foreign body scar with central rust staining.



Fig. F-6b Paracentral imbedded non-metallic corneal foreign body.

Fuchs' endothelial dystrophy

Guttae

DESCRIPTION

Fuchs' endothelial dystrophy is a hereditary, often autosomal dominant, disorder. It has variable penetrance and affects women more than men at a proportion of approximately 5:1. It primarily affects the corneal endothelial cells with a progressive reduction in endothelial cell number and pumping capacity, eventually resulting in persistent corneal edema.

Symptoms normally begin in the sixth or seventh decade of life and progress over several years. Progression is often somewhat asymmetric, with one cornea frequently decompensating a year or two before the other. Rapid progression of corneal edema can occur following routine uncomplicated cataract surgery, underlying the minimal endothelial reserve frequently present in patients with this disease.

SYMPTOMS

The first symptom is usually early morning blurring of vision and halos. Typically, this clears with time, initially lasting only half an hour or so. However, with progression of corneal endothelial dysfunction this period extends. Eventually, usually after several years, the cornea fails to clear at all over the entire day. Blurring of vision is often accompanied by ocular discomfort and grittiness due to corneal endothelial edema. If left untreated bullous keratopathy with associated episodic severe pain can occur.

SIGNS

Aside from the signs of corneal edema, the typical finding is that of central endothelial guttae. These are small stippled raised lesions on Descemet's membrane mainly seen in the central cornea. Often there are numerous pigment granules scattered around the guttae.

A few scattered peripheral excrescences in the peripheral Descemet's membrane are common, especially in the elderly. These are termed Hassall–Henle bodies, or Descemet's warts, and are not indicative of Fuchs' dystrophy.

There is also frequently an associated nuclear sclerotic cataract.

INCIDENCE

Uncommon, although the third most common corneal dystrophy overall after epithelial basement membrane dystrophy and keratoconus. Most commonly seen in women over the age of 50.

SIGNIFICANCE

Slowly leads to significant bilateral visual loss unless treated.

DIFFERENTIAL DIAGNOSIS

Keratouveitis; Uveitis.

SEE ALSO

Corneal edema; Corneal graft; Corneal graft – lamellar keratoplasty; Disciform keratitis; Corneal dystrophies, degenerations, and deposits – overview; Bullous keratopathy.

MANAGEMENT

Review

Guttae noted on routine examination with no or only minimal corneal edema require no active intervention. Six-monthly to yearly review may be carried out to assess the progression of the disease.

Genetic advice

Inheritance is autosomal dominant with a high degree of penetrance, being more commonly seen in women. Patients should be advised that, although it is possible their children will also develop the disease, this is not particularly common. Missense mutations in several genes have been linked to the disease, with a recent study suggesting Fuchs' dystrophy and posterior polymorphous dystrophy are allelic variants of the same disease continuum. Another study found an association with a mutation in the gene for collagen VII, a major component of Descemet's membrane.

Ocular investigations

Corneal pachymetry and specular microscopy may be useful in defining the

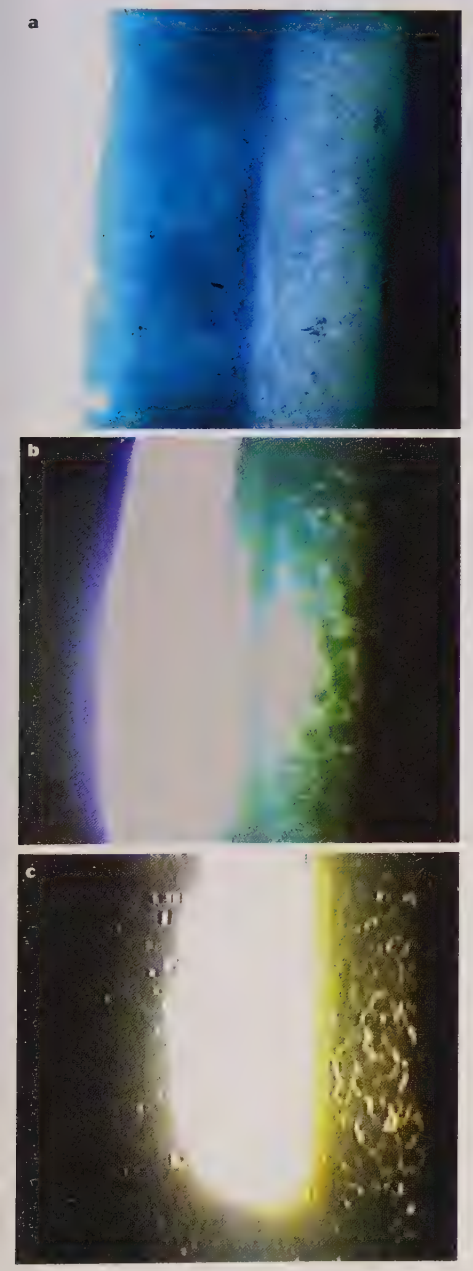


Fig. F-7 Endothelial guttae in Fuchs' dystrophy: **a** normal endothelium, grade 0; **b** mild guttae, grade 2; **c** moderate guttae, grade 3.

degree of corneal edema and extent of endothelial cell loss. Pachymetry readings may demonstrate diurnal variation, the cornea being thicker in the morning.

Therapeutics

Initially, ocular discomfort can be reduced by the use of tear supplements. In the early stages of the disease, vision can be improved by treating the corneal epithelial microcystic edema with hypertonic saline (5%) drops or ointment. This acts to osmotically dry out the epithelium; it does not reduce stromal edema. Drying of the corneal epithelium can also be achieved with warm dry air from a hair dryer. The hair dryer should be set on low and held at arm's length with the airflow directed above the head.

Surgery

A corneal graft (penetrating or lamellar, such as DSAEK) is currently the only option to restore corneal clarity. This is often performed earlier for the first eye to enable visual recovery before the second eye loses too much vision. Surgery in the first eye is indicated when vision is reduced below 6/12 to 6/18 and fails to clear by midday to early afternoon. The number of corneal grafts performed for Fuchs' endothelial dystrophy has been steadily rising over the years, probably secondary to an aging population. A cataract extraction and intraocular lens implantation is often performed at the time of the graft (a triple procedure).

F

Fuchs' heterochromic iridocyclitis

Fuchs' uveitis syndrome

DESCRIPTION

Although the features of Fuchs' heterochromic iridocyclitis are unique, it is often misdiagnosed. It is a chronic, recurrent, non-granulomatous anterior uveitis of unknown cause. It differs significantly from other forms of anterior uveitis with regard to the symptoms, signs, prognosis, and treatment. Fuchs' heterochromic iridocyclitis typically affects young adults, although it may present in patients of any age.

Key differences from other forms of anterior uveitis include:

- Iris changes with heterochromia are common.
- Glaucoma is very common.
- Corticosteroids have little to no effect on the intraocular inflammation.
- Both anterior peripheral and posterior synechia do not occur; therefore, there is no need to treat with mydriatics.

The cause of Fuchs' heterochromic iridocyclitis is unknown although two mechanisms postulated to be involved are infection with the rubella virus and *Toxoplasma gondii*, the cause of ocular toxoplasmosis.

SYMPTOMS

Early in the course of the disease there are few if any symptoms. Frequently, the presenting symptom is blurring of vision from cataract or, less commonly, from advanced glaucoma. Patients do not develop a red or sore eye although they may notice floaters from the vitritis.

SIGNS

Signs are almost always unilateral (approximately 10% are bilateral), with typical scattered small stellate keratic precipitates (KP). These are non-pigmented, gray to white in color, and come and go with time. There may be an associated low-grade anterior chamber reaction with minimal cell and flare. There is also frequently a low-grade anterior vitreous reaction with cells and opacities being present.

The iris heterochromia characteristic of this condition is present in most cases and results from progressive atrophy of both the iris stroma and the posterior pigment layer. This results in a flattened, somewhat featureless iris with prominent blood vessels; this is often most marked in the peripupillary region. Retroillumination defects are common, especially around the pupil margin, and the resting pupil is often somewhat enlarged due to atrophy of the sphincter muscle. If iris heterochromia is present, the affected eye is usually, although not always, lighter in color.

The most common vision-threatening feature is a cataract, although the most important complication to look for is raised intraocular pressure and signs of glaucomatous disc damage. On gonioscopic examination, the angle may appear normal or may show signs of trabecular meshwork scarring and neovascularization.

INCIDENCE

Rare, accounts for approximately 4% of all cases of uveitis.

SIGNIFICANCE

Visual loss is common from both cataract and glaucoma. The glaucoma is frequently difficult to treat either medically or surgically, and advanced nerve damage is common.

DIFFERENTIAL DIAGNOSIS

Anterior uveitis; Posner–Schlossman syndrome (glaucomatocyclitic crisis).

SEE ALSO

Iritis; Disciform keratitis.

MANAGEMENT

Review

Six-monthly reviews with pressure checks and optic nerve-head examination are indicated because of the frequency and insidious nature of glaucoma.



Fig. F-8a Typical small, stellate, scattered keratic precipitates in Fuchs' heterochromic iridocyclitis.

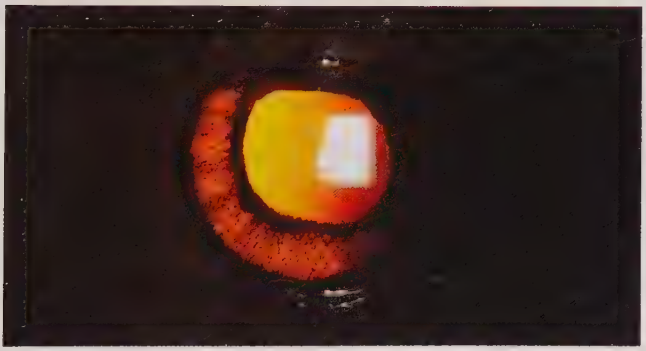


Fig. F-8b Extensive peripupillary iris transillumination defects.

Therapeutics

As mentioned above, unlike other forms of anterior uveitis, topical corticosteroids are ineffective and mydriatics unnecessary. Topical anti-glaucoma medication is indicated if the pressure is raised. However, unfortunately they are frequently not wholly effective. Proinflammatory drugs such as latanoprost are contraindicated as they may exacerbate the underlying disease.

Surgery

Cataract surgery is usually eventually required with Fuchs' heterochromic iridocyclitis. The surgery is sometimes made slightly more difficult as the iris often dilates poorly. However, there is no increased risk of corneal endothelial cell decompensation leading to corneal edema.

Glaucoma-filtering surgery is also commonly required, although unfortunately the failure rate for trabeculectomy is quite high. As such, most practitioners recommend that adjunctive antimetabolites such as 5-FU or mitomycin C are used during surgery.

F

Fungal keratitis

Keratomycosis

DESCRIPTION

Of the four classes of organism that can infect the cornea – bacteria, viruses, fungi, and protozoa – fungi are one of the most destructive and most difficult to treat. Two types of fungi infect the cornea: septate-filamentous fungi and yeasts. The common filamentous infective organisms are *Fusarium* and *Aspergillus*, while the common infective yeast is *Candida albicans*. Fungal keratitis usually occurs in an already-compromised eye with a history of ocular surface disease and long-term topical corticosteroid use. The infection takes a slower and, unfortunately, more relentless course than bacterial keratitis.

SYMPTOMS

Fungal keratitis even in the early stages leads to marked symptoms of pain, photophobia, and increased lacrimation. The development of symptoms is usually slower than with a bacterial keratitis. Patients typically present within several weeks rather than several days of a sore red eye. There is also sometimes a history of an agriculture injury with exposure of the cornea to a dirty foreign body or of long-term topical corticosteroid use.

SIGNS

The infected eye usually demonstrates marked conjunctival injection. The corneal infiltrate is usually quite dense with an indistinct (feathered) border. Occasionally, there are typical surrounding infiltrates, termed "satellite lesions." Corneal edema, anterior chamber cell and flare, and hypopyon are common.

INCIDENCE

Very rare in most developed countries, although relatively common in some tropical and subtropical developing



Fig. F-9 Fungal keratitis with large, dense, central infiltrate with overlying epithelial defect, showing the typical surrounding infiltrates (satellite lesions), and hypopyon.



nations. Incidence is related to climate; fungal keratitis (especially with *Fusarium*) has been shown to be more common in the southern USA than in northern states.

SIGNIFICANCE

Sight-threatening, with about 50% of eyes ending up with "count fingers" or less vision. Early diagnosis and treatment is of paramount importance.

DIFFERENTIAL DIAGNOSIS

Bacterial keratitis; *Acanthamoeba* keratitis; Herpes simplex keratitis; Herpes zoster ophthalmicus.

SEE ALSO

Herpes simplex keratitis – overview; Endophthalmitis; Corneal infiltrates; Hypopyon; Infectious crystalline keratopathy.

MANAGEMENT

Although the cornea and anterior chamber are typically quite inflamed, topical corticosteroids are contraindicated because of their ability to potentiate fungal activity. Topical antibiotics are inactive against fungi.

Microbiological investigations

If suspected, a corneal scrape with submission of the specimens for special fungal stains and culture is indicated. If scrapes fail to identify an organism, a corneal biopsy may be necessary. Several stains have been used to identify fungi; one of the most sensitive and widely used is a Blancophor preparation. Fungi are usually cultured on a Sabouraud agar slab, a special fungal culture medium. Antifungal sensitivities are usually not performed, as they take a long time to complete and are notoriously unreliable.

Antifungal medication

As the clinical differentiation of a fungal keratitis from other corneal infections is so unreliable, and the antifungal agents used are often somewhat toxic to the cornea, treatment with antifungal agents should only be commenced when there is a positive microbiological diagnosis.

Often a combination of both topical and oral antifungals is used. Occasionally, corneal intrastromal injection of voriconazole may be used to treat non-responsive infections. No single drug is universally active against all fungi, and drug treatment failures are unfortunately common. Four classes of antifungals are commonly used for treating fungal keratitis: polyenes (natamycin and amphotericin B), imidazoles (ketoconazole and fluconazole), triazoles (voriconazole), and fluorinated pyrimidines (flucytosine). Some drugs are particularly indicated for specific species of fungus, such as amphotericin B for candidal infections and voriconazole with *Scedosporium* species. Unfortunately, many of these drugs have poor penetration into the cornea, and many clinicians actively remove healing epithelium over the infiltrate to enhance penetration into the stroma.

With successful treatment the pain subsides, the edge of the corneal opacity becomes more distinct (rounds up), and the satellite lesions fade.

Surgery

Successful treatment often requires surgical intervention with a penetrating keratoplasty or sclerectomy to limit the spread of the disease. If contemplated, it is usually best performed early, before spread of the organism into surrounding tissue.

G

Giant papillary conjunctivitis (GPC)

Contact lens-induced/associated papillary conjunctivitis (CLPC)

DESCRIPTION

Giant papillary conjunctivitis (GPC) affects the upper tarsal conjunctiva and is usually seen with soft contact lens use, although it is also less commonly seen with rigid lenses. GPC has also been reported to occur with an ocular prosthesis (glass eye) or following cataract surgery as a response to corneal sutures.

Giant papillae are classically described as being 1 mm in diameter, although there is clearly a continuum, and they may be quite a bit smaller in less severe forms of the disease. For this reason, the term "giant" is sometimes omitted, giving the term "papillary conjunctivitis," or in contact lens wear, CLPC. The condition has an insidious onset over weeks and may even occur in people who have successfully worn contact lenses for years.

GPC is thought to be caused by a combination of an allergic reaction to deposits on the lens material and mechanical irritation related to passage of the upper lid over the surface of the lens. GPC appears to occur more commonly with poor lens hygiene and in extended wear. It is less common with more frequent lens replacement.

Histological studies of the conjunctiva have demonstrated an increase in neutrophils, plasma cells, and eosinophils underlying the allergic and inflammatory nature of the disease.

SYMPTOMS

Ocular discomfort, increased awareness of the lens and lens movement, itch, increased mucus production, and blurred vision are all common. Symptoms are made worse with lens use but frequently do not totally resolve for hours to weeks following discontinuing lens usage.

SIGNS

Examination of the contact lenses may reveal increased protein deposition. The following grading scale for GPC is widely used, with the papillary reaction being graded in the central upper tarsal conjunctiva. The appearance of the papillae is accentuated with fluorescein:

- Grade 1 (preclinical) – slight conjunctival injection with fine papillae and no symptoms
- Grade 2 (mild) – mild injection, 0.3–0.5-mm papillae, and mild symptoms
- Grade 3 (moderate) – moderate injection with some early subconjunctival scarring and 0.5-mm or larger papillae, moderate symptoms with lens awareness increased
- Grade 4 (severe) – moderate to severe injection with papillae greater than 0.75 mm and significant lens intolerance.

As with the symptoms, signs may persist for months after ceasing lens use.

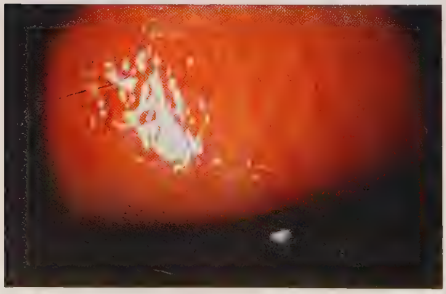


Fig. G-1a Mild papillary conjunctivitis, highlighted in specular reflection.

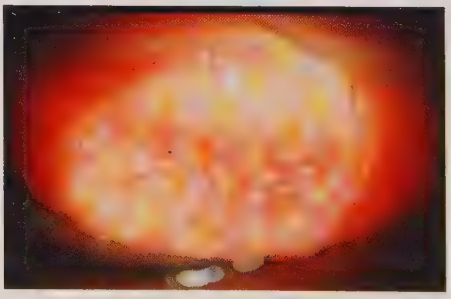


Fig. G-1b Severe papillary conjunctivitis with giant papillae.



INCIDENCE
Preclinical and mild grades are commonly seen in long-term soft contact lens users whereas moderate to severe grades are uncommon.

SIGNIFICANCE
May result in long-term contact lens intolerance.

DIFFERENTIAL DIAGNOSIS
Superior limbic keratoconjunctivitis (SLK); Seasonal allergic conjunctivitis (SAC); Vernal keratoconjunctivitis (VKC).

SEE ALSO
Follicular conjunctivitis; Contact lens-related conditions; Infectious crystalline keratopathy.

MANAGEMENT
The degree of intervention is highly dependent on the grade of GPC.

Advice
Successful treatment requires the patient to have a clear understanding of the nature of the condition to improve compliance with both the modification of lens usage and topical medication.

Change or refit contact lens
The use of biweekly/monthly or daily disposable lenses may be appropriate with all grades of GPC; occasionally, refitting with a rigid gas permeable (RGP) lens may be necessary.

Modification of lens use
Contact lens modification focuses on changing to daily disposables, reducing wear time and frequency, ceasing extended wear, and re-emphasizing the importance of proper lens care, including the "rub" step. In grades 1 and 2, patients should be advised to modify lens use to reduce wear time to less than 8–10 hours a day for a maximum of 6 days a week. Patients should be advised lenses should not be used on an extended-wear basis, even if they are using a silicone hydrogel lens approved for such use. In grades 3 and 4, cessation of lens use for 1–2 months while topical medication takes effect may be required. Although now rarely worn, non-disposable conventional contact lens use may be assisted by changing to hydrogen peroxide solution and more frequent enzymatic cleaning.

Before recommencing lens wear, the patient should be examined to ensure tarsal conjunctival inflammatory changes have significantly resolved. The lens is then reintroduced, gradually increasing lens wear times.

Therapeutics
Topical corticosteroids are commonly used in the short term (qid for 2 weeks) to settle grade 3 and 4 disease. A topical combined antihistamine/mast cell stabilizer such as olopatadine or ketotifen fumarate is the mainstay of topical treatment. This may be continued for several months, even after reintroduction of contact lens use.

Dosing: G. fluorometholone 0.1% qid for 2 weeks. G. olopatadine 0.1% or ketotifen fumarate bd for several months.

G

Glaucoma – acute angle closure

Acute pupil-block glaucoma

DESCRIPTION

Acute angle closure glaucoma is usually a dramatic, rapid-onset glaucoma leading to a very high intraocular pressure (IOP); it presents a marked and immediate threat to vision. The mechanism of angle closure is usually blockage of aqueous flow through the pupil from the posterior to anterior segments, with subsequent bowing forward of the peripheral iris (iris bombe), leading to occlusion of the angle. A shallow anterior chamber (less than 2-mm deep centrally) markedly increases the risk of angle closure. This is more common in Asian peoples, the elderly, women, and people with small corneas, large lenses (i.e. with a nuclear sclerotic cataract), short axial length (also associated with hypermetropia), and a positive family history.

Occasionally, angle closure can be subacute, with the patient giving a history of intermittent episodes of ache, blur, and halos around light. Rarely, pupil block can also occur with intraocular lenses and surgical gas or silicone oil in the anterior chamber. Aside from pupil block, acute angle closure can also occur in the rare condition of plateau iris. Acute angle closure can also be precipitated by pharmacological dilation of the pupil in a patient with a shallow anterior chamber, and narrow drainage angle. Pupil block has become much less common in developed countries, probably due to the increasing incidence of cataract surgery.

SYMPTOMS

The patient often presents in the evening or overnight with a severe, dull, ocular ache, reduced vision, halos around lights, and even nausea and vomiting. Despite the usually dramatic nature of the presentation, some patients have remarkably few symptoms.

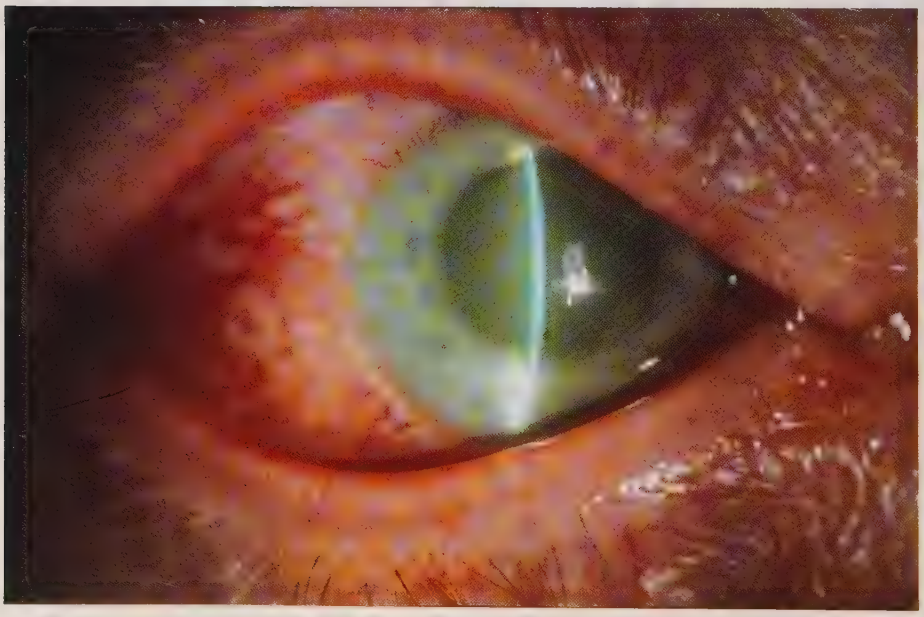


Fig. G-2 Acute angle closure glaucoma with conjunctival injection, corneal edema, mid-dilated pupil, and nuclear sclerotic cataract.



SIGNS

The eye is injected, the cornea edematous, and the IOP raised, usually to between 50–80 mmHg. The anterior chamber is shallow, and the iris bowed forward; the pupil is totally non-reactive, often oval, and in mid-dilation. Gonioscopy is difficult due to corneal edema but, if possible, reveals the angle to be fully occluded. If the optic nerve head is visible, it is usually swollen, and vascular flow nonexistent.

INCIDENCE

Rare in most developed countries but remains a significant problem in some developing countries, especially in parts of Asia.

SIGNIFICANCE

Untreated, it may lead to total visual loss from optic nerve damage within a matter of 24–36 hours.

DIFFERENTIAL DIAGNOSIS

Neovascular glaucoma; glaucomatocyclitic crisis (Posner–Schlossman syndrome) – see associated text, *Posterior Eye Disease and Glaucoma A–Z*; Disciform keratitis.

SEE ALSO

Corneal edema; Pseudoexfoliation of the lens capsule (PXF); pigment dispersion (see "Endothelial opacities").

MANAGEMENT

Urgent ophthalmic intervention is required to protect vision, with the majority of cases being treated at hospital emergency departments.

Therapeutics

Topical pilocarpine (1–2% every 15 minutes to 1 hour), to constrict the pupil and relieve the pupil block, may be effective early on in the disease. It is usually ineffective with IOP above 50 mmHg due to the induced iris ischemia. It should also be instilled in the other eye to prevent the development of pupil block. Exercise caution if retinal detachment risk factors are present. Topical anti-glaucoma medication should also be instilled; apraclonidine 0.5%, timoptol 0.5%, or dorzolamide 2%, all 1 drop stat, may be used. Commonly, both apraclonidine and dorzolamide are used. Latanoprost is not used because of its slow onset of action and pro-inflammatory effect.

Oral and intravenous medication

Indicated if IOP over 50 mmHg, or if significant vision loss. Established pupil block is initially treated with oral or intravenous acetazolamide, usually 500 mg stat, to reduce aqueous production. Hyperosmotic agents such as oral glycerin or intravenous mannitol may be used to reduce vitreous volume if acetazolamide is insufficient.

Laser iridotomy

The definitive treatment is with a peripheral iridotomy. Timing of a peripheral Nd:YAG laser iridotomy is somewhat controversial and may be difficult to perform early on due to corneal edema. The aim is to re-establish a communication between the posterior and anterior chambers. This will relieve the pupil block and can lead to instantaneous resolution of angle closure. It is frequently performed acutely to rapidly lower the IOP. It is important to perform the same procedure in the other eye within a few days to protect it from a similar attack.

Surgery

Cataract extraction is an effective measure to prevent the development of pupil block in the other eye.

G Gonococcal keratoconjunctivitis

DESCRIPTION

Gonococcal conjunctivitis is an acute bilaterally sight-threatening disease caused by infection with the Gram-negative diplococcus, *Neisseria gonorrheae*, the same organism that causes gonorrheal urethritis. Gonococcal conjunctivitis is usually contracted as a sexually transmitted disease, in which case there is an associated infection of the genitourinary tract, leading to a purulent urethritis in men and urethritis and vaginitis in women. Women especially may have genitourinary infection without symptoms. Neonates can also contract the disease during passage through the birth canal, leading to ophthalmia neonatorum.

Epidemics of gonococcal conjunctivitis, mainly affecting children, have also been reported in Aboriginal communities in outback Australia. In these cases, direct hand-to-eye contamination and flies have been the suggested mode of transmission.

SYMPTOMS

The patient presents with hyperacute, usually bilateral, conjunctivitis. There is usually a characteristic marked mucopurulent discharge. They may also have symptoms of urethritis with burning and stinging on urination.

SIGNS

Aside from the discharge, intense conjunctival hyperemia, chemosis, and pseudomembrane formation are common. Other common signs are marked lymphadenopathy and eyelid edema. In severe cases ulcerative keratitis may develop, initially involving the marginal cornea but extending to involve the whole of the cornea, leading to keratolysis and even perforation.



Fig. G-3 Marked purulent discharge and conjunctival inflammation in adult gonococcal conjunctivitis.



INCIDENCE
Rare, although more common in some communities. Gonococcal urethritis is more common than gonococcal conjunctivitis.

SIGNIFICANCE
Serious sight-threatening disease due to corneal ulceration, scarring, and the risk of perforation.

DIFFERENTIAL DIAGNOSIS
Other causes of acute mucopurulent conjunctivitis, including Stevens–Johnson syndrome.

SEE ALSO
Bacterial conjunctivitis; Conjunctival scarring – overview; Ophthalmia neonatorum; Perforation – corneal; Membranous and pseudomembranous conjunctivitis; Conjunctivitis – overview.

MANAGEMENT
Advice
As the infection is sexually transmitted, identification and treatment of sexual partners is important, both to halt the spread of the disease and to prevent re-infection.

Microbiology
Specific microbiological identification of *N. gonorrheae* is important, because of both the social implications of identifying a sexually-transmitted disease and the need to treat sexual partners. Treatment can proceed on the basis of the Gram stain, especially in ophthalmia neonatorum where rapid intervention is required.

Lid hygiene
Much of the ocular surface damage is from the active lytic enzymes in the mucopurulent discharge. As such, frequent lid hygiene and eye flushing with sterile saline to remove any discharge is important. Initially, this may need to be performed every hour.

Therapeutics
As this is a systemic infection, systemic antibiotics are required. While penicillin has traditionally been the treatment of choice, many strains are now penicillin-resistant. Consequently, ceftriaxone 1 g intramuscularly daily for 5 days is now commonly used. Topical antibiotics such as gentamycin 1.3% hourly are also usually used. In children, cefotaxime 25 mg/kg bid for 7 days is usually the treatment of choice.

Notifiable
In some countries this infection may need to be reported to public health authorities.

G

Granular dystrophy

Groenouw I dystrophy

DESCRIPTION

The corneal dystrophies are a group of progressive, usually bilateral, inherited (although non-congenital) corneal disorders with a typical age of onset. The stromal dystrophies result in characteristic stromal opacities. There are many different stromal dystrophies; the three main types are granular, macular, and lattice. Granular dystrophy was first described in the Western literature by Groenouw in 1898 and is occasionally referred to as Groenouw I dystrophy. It is now known that granular dystrophy (along with Avellino, lattice, and Reis–Bücklers dystrophies) occurs secondary to a mutation in the kerato-epithelin gene *BIGH3* on the long arm of chromosome 5. The typical "bread crumb"-like deposits seen in granular dystrophy are from deposition of an amorphous hyaline material within the corneal stroma. This is best seen histopathologically with Masson's trichrome, where it stains bright red. There are three subtypes of granular dystrophy:

- type I with an early onset usually in the first decade of life

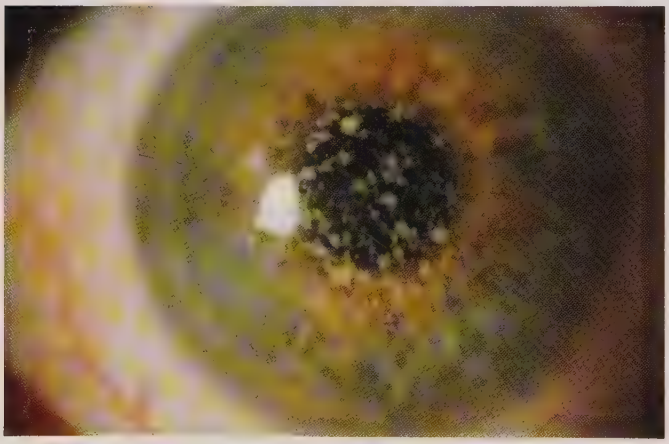


Fig. G-4a Moderately advanced type 1 granular corneal dystrophy.

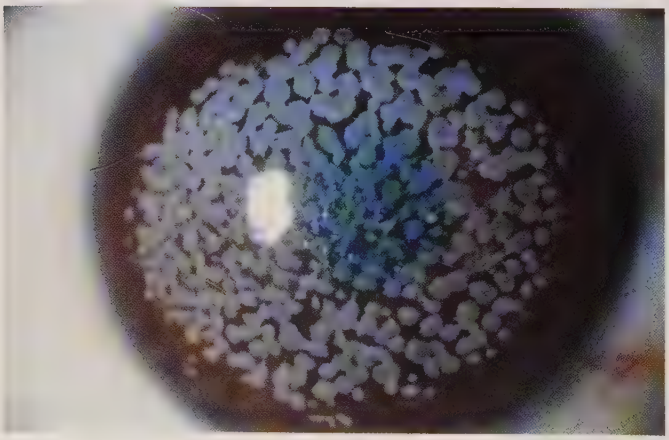
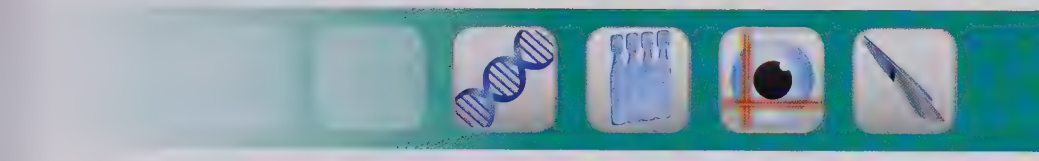


Fig. G-4b More advanced disease with extensive opacities.



- type II with an onset in the fourth or fifth decades
- type III or Avellino dystrophy with an onset late in life and lattice lines as well as granular deposits.

SYMPTOMS
Recurrent corneal erosion symptoms, ocular discomfort, and blurring of vision from the developing opacities all increase over time.

SIGNS
The deposits of granular dystrophy evolve from small "crumb-like" white opacities to almost confluent white lesions. They initially involve the superficial stroma, but spread deeper to involve the middle stroma. One of the characteristic features is that the intervening cornea between lesions remains clear. The lesions of granular dystrophy type 2 remain fairly superficial and are smaller in size. The lesions are most marked centrally, but also spread peripherally, although they do not involve the limbus. The overlying epithelium is frequently affected, being raised and exhibiting areas of negative fluorescein staining. There may also be areas of epithelial instability associated with recurrent corneal epithelial erosions.

INCIDENCE
Rare to very rare (less than 1/10,000).

SIGNIFICANCE
May lead to a marked reduction in visual acuity secondary to corneal opacification.

DIFFERENTIAL DIAGNOSIS
Avellino dystrophy; contact lens deposits; ciprofloxacin deposits; Microbial keratitis; Band keratopathy; Reis–Bücklers dystrophy; Corneal grafts – lamellar keratoplasty.

SEE ALSO
Macular stromal dystrophy; Lattice stromal dystrophy; Recurrent corneal erosion syndrome; Corneal dystrophies, degenerations, and deposits.

MANAGEMENT
Genetic advice
Granular stromal dystrophy is usually inherited as an autosomal dominant trait with a high degree of penetrance.

Tear supplements
These are commonly used to treat ocular surface discomfort associated with an unstable epithelium and recurrent corneal erosion syndrome.

Phototherapeutic keratectomy
Phototherapeutic keratectomy with the excimer laser has been used to treat superficial granular deposits both to improve vision by removing opacities and to stabilize the epithelium. Unfortunately, adequate treatment of opacities usually requires a fairly deep ablation and results in a significant hypermetropic shift. It can also lead to significant scarring and thinning of the cornea.

Surgery
Occasionally, in advanced disease with significant visual loss, a lamellar (deep anterior lamellar keratoplasty) or penetrating keratoplasty is performed. Superficial recurrences within the graft are common and may lead to renewed visual disturbances. Thankfully, this normally takes several years to occur.

Hemangioma

Strawberry nevus, nevus flammeus (port wine stain), cavernous hemangioma

DESCRIPTION

A hemangioma is a benign neoplasm (tumor) of superficial blood vessels. The lesion is not encapsulated and is composed of anastomosing blood vessels with an infiltrative growth pattern. Capillary hemangiomas are among the most common tumors in infants, arising as congenital lesions at or soon after birth. They may regress or fade by adolescence.

Strawberry nevus is a capillary hemangioma, most commonly occurring on the upper lid. The angioma can less frequently involve other skin areas, including elsewhere on the face or on the body.

Port wine stain (nevus flammeus) is a hemangioma where the lesion is subcutaneous, usually segmental, and most commonly on the face. Larger lesions may be associated with multi-system disorders. For example, Sturge–Weber syndrome has an association of port wine stain with intracranial or ocular hemangiomas and glaucoma.

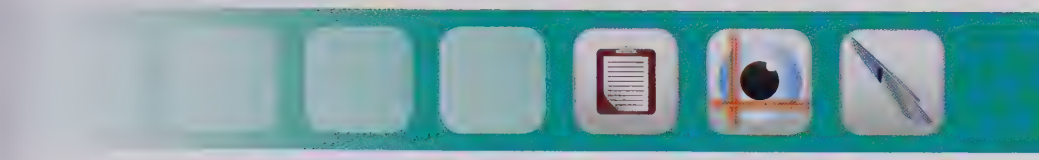
Cavernous hemangioma is a benign tumor that can occur within the orbit. When it occurs posterior to the globe, it



Fig. H-1a Capillary hemangioma (strawberry nevus) on the upper lid.



Fig. H-1b Right proptosis due to a cavernous hemangioma.



may lead to proptosis, compression of the optic nerve, or cause papilledema. Presentation is most commonly in females in the fourth or fifth decade. Unlike the capillary hemangiomas, these tend to be encapsulated.

SYMPTOMS

The lesions are painless but may be a cosmetic concern. They are usually unilateral. The parent may present with an infant with a cosmetic or visual concern.

SIGNS

A skin hemangioma appears as a flat or slightly elevated, circumscribed, colored lesion on the skin surface. The color of the strawberry nevus is more red to blue, whereas the port wine stain appears dark blue to purple through the overlying skin. Superficial lesions may blanch on pressure (unlike a nevus or melanoma).

Visual and oculomotor function in the infant should be evaluated, if there is eyelid or orbital involvement. A glaucoma work-up is required for patients with ocular involvement with a port wine stain.

INCIDENCE

Common (approximately 1/100) in infants under 6 months of age.

SIGNIFICANCE

Although these conditions are benign, there can be mass effects. If a strawberry nevus is located on the upper lid, there is potential for ptosis and amblyopia in the growing infant eye. If located elsewhere within the orbit, there is a potential for displacement of the globe, strabismus, or development of a refractive anomaly.

When the superior lid is involved in a port wine stain, there is an increased risk of glaucoma in that eye. There may be an association with rare systemic syndromes, such as Sturge–Weber syndrome.

DIFFERENTIAL DIAGNOSIS

Seborrheic keratosis; Actinic keratosis; Pyogenic granuloma; Nevus – eyelid.

MANAGEMENT

Review, and advice

Small lesions may simply be documented and monitored. A photograph can provide more information than a drawing. Review is indicated usually at an interval of 6 to 12 months if invading orbit or eyelid.

Surgery

Excision is rarely indicated, as the lesions are benign. Most hemangiomas are not encapsulated and have a propensity to bleed if excised. If the hemangioma is life- or sight-threatening, treatment may be via an intralesional injection of corticosteroids (triamcinolone), or systemic interferon-alpha may be tried for a capillary hemangioma.

Laser surgery

Erbium laser treatment may be used for a port wine stain to try to reduce the amount of skin coloration.

H Herpes simplex keratitis (HSK) – overview

DESCRIPTION

The herpes simplex virus is a double-stranded DNA virus and is the most ubiquitous viral infection in humans. Humans are its only known host. Ocular infection is common. It is estimated that approximately 500,000 people in the USA have had herpes simplex keratitis (HSK) and that approximately 10% of these have at least one recurrence each year. The extremely protean nature of its corneal involvement is one of the distinctive features of HSK and has led to it being labeled a "great masquerader." The initial or primary infection typically occurs in childhood and leads to eyelid skin vesicles, follicular conjunctivitis, and, frequently, corneal microdendrites, a blepharokeratoconjunctivitis. Following the primary infection, the virus remains in the body, resident in the trigeminal ganglion. This establishment of dormant virus within the neuronal cell bodies is termed latency. Secondary disease occurs when the virus becomes reactivated and travels down the sensory neuronal axon to infect the cornea. Several manifestations of secondary disease occur as a result of an inflammatory response to residual, often non-replicatory, viral antigen rather than from florid viral replication.

CLASSIFICATION

The different types of corneal involvement with secondary HSK can be classified on an anatomical basis or according to the degree of viral replication and inflammation. The latter is more useful from a clinical perspective as it helps to direct the usage of topical antivirals and corticosteroids. There are three anatomical classifications:

- Epithelial
  —Dendritic ulcer

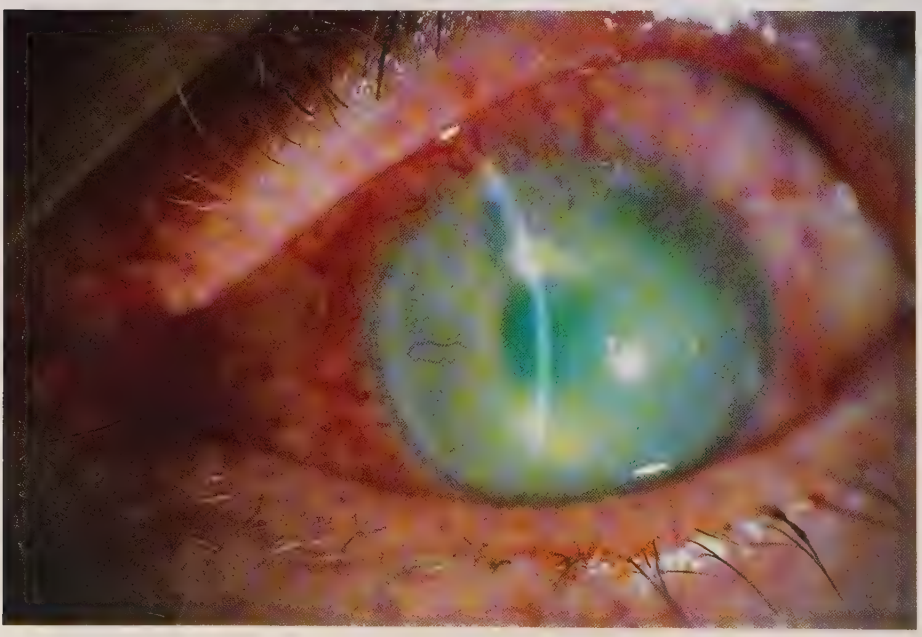


Fig. H-2 Inflamed vascularised, edematous cornea with focal opacities in stromal necrotic herpes simplex keratitis.



—Geographic ulcer
—Metaherpetic (indolent) ulcer
- Stromal
  —Stromal necrotic keratitis
  —Disciform (herpetic keratouveitis) keratitis
  —Interstitial keratitis
- Endothelial
  —Disciform (herpetic keratouveitis) keratitis
  —Linear endotheliitis

While some viral replication within the cornea may occur at a low level in all these conditions, dendritic and geographic ulcers are the conditions primarily caused by viral replication.

SYMPTOMS

Blurring of vision, increased lacrimation, photophobia, and ocular discomfort are common with all manifestations of HSK. Significant pain is rarely a symptom of HSK, probably due to the neurotrophic nature of the virus leading to reduced corneal sensation. This is especially the case with recurrent disease.

SIGNS

Signs of ocular inflammation and viral activity should be sought. Ocular inflammation usually leads to corneal edema, keratic precipitates, an anterior chamber reaction, and an increased intraocular pressure. Viral replication is most obviously manifest by an epithelial dendritic staining pattern. The condition is usually unilateral.

INCIDENCE

Affects approximately 1/5000 people every year. Metaherpetic ulcers, interstitial keratitis, limbal vasculitis, and linear endotheliitis are all very rare.

SIGNIFICANCE

HSK is the most common cause of corneal scarring in most developed countries.

DIFFERENTIAL DIAGNOSIS

Herpes zoster ophthalmicus; chicken pox; healing epithelial defect.

SEE ALSO

Dendritic ulcer; Disciform keratitis; Herpes simplex primary blepharo-keratoconjunctivitis; Interstitial keratitis; Conjunctivitis – overview; Perforation – corneal.

MANAGEMENT

The management of each condition is given in full in their individual articles. The following general principles apply.

Advice

Patients with recurrent disease should be advised to avoid known trigger factors, such as UV light exposure and ocular trauma, with the use of tight-fitting wrap-around sunglasses. They should also be especially attuned to the possibility of a recurrence around the time of a significant intercurrent illness, such as influenza. They should be advised of the need for prompt presentation if they feel they may have a recurrence of HSK.

Microbiology

Culture, or (more commonly) direct immunofluorescent staining, PCR, or viral DNA can be undertaken from tissue swabs to identify specific viral antigen.

Therapeutics

Topical antiviral agents are used either in a full-treatment dose (i.e. acyclovir ointment 5 times daily for 7 days to treat active viral replication) or prophylactically (i.e. acyclovir bd to prevent viral reactivation during concurrent topical steroid usage). Topical steroids are used to suppress inflammation and are usually tapered over several months to prevent rebound inflammation from occurring. Specific treatment regimens for the different manifestations of HSK are given under the specific condition articles.

Oral medication

Long-term, low-dose oral acyclovir has been shown to be effective in reducing the number of recurrences, especially in patients with recurrent disease.

Herpes simplex primary blepharokeratoconjunctivitis

Primary herpes simplex infection

DESCRIPTION

The herpes simplex virus is a double-stranded DNA virus and is the most ubiquitous viral infection in humans. Humans are its only known host. The initial or primary infection usually occurs in children and involves the mucocutaneous junction of the lips. Occasionally, the initial lesion involves the eye, leading to a blepharokeratoconjunctivitis. The source of infection is most commonly from direct contact with a family member or friend.

The primary ocular infection is usually mild, self-limiting, and often subclinical. Its importance lies more in the establishment of viral latency within the trigeminal ganglion, predisposing to secondary herpes simplex keratitis at a later date.

A primary ocular infection can also occur from exposure to the virus during passage of a baby through the birth canal; this is one of the causes of ophthalmia neonatorum. Extremely rarely, the infection may progress to a meningitis or encephalitis, requiring urgent admission to hospital and intravenous antiviral therapy.

SYMPTOMS

There is often a low-grade fever and symptoms of a mild generalized viral illness. The skin lesions may be unilateral or bilateral. The affected area is irritable, and the child often has photophobia and increased lacrimation. In the very rare event of an associated encephalitis, the child's conscious state is affected.

SIGNS

The skin lesions typically appear first and crop over several days. The lesions have the typical vesicular appearance of a cold sore; in contrast to shingles, they do not respect a normal dermatomal distribution. An associated conjunctivitis is common with follicles, a watery discharge, and preauricular lymphadenopathy. Occasionally, a keratitis also occurs; this is usually quite benign and non-scarring, with fine, grouped, superficial punctate erosions or even microdendrites.

INCIDENCE

Common; most common in children.

SIGNIFICANCE

Usually benign, but predisposes the child to secondary herpes simplex keratitis and may, rarely, lead to herpetic meningitis or even encephalitis.

DIFFERENTIAL DIAGNOSIS

Other causes of childhood vesicular rashes such as chicken pox (varicella), rubella, and measles.

SEE ALSO

Dendritic ulcer; Disciform keratitis; Herpes simplex virus – overview; Interstitial keratitis; Herpes simplex ophthalmicus; Ophthalmia neonatorum.



Fig. H-3 Typical vesicular eruption affecting the skin around both eyes in a child with primary herpetic blepharokeratoconjunctivitis.



MANAGEMENT

Often no treatment is necessary. Some clinicians do treat the disease with oral acyclovir in an effort to reduce the risk of the virus establishing latency within the nerve ganglion. The skin lesions should be kept clean to avoid bacterial superinfection.

Review

Review every several days until the lesions stop cropping and the conjunctivitis begins to resolve is common practice.

Microbiology

Culture or, more commonly, direct immunofluorescent staining can be undertaken from tissue swabs to identify specific viral antigen. Polymerase chain reaction (PCR) to establish the presence of viral DNA is now the initial investigation in many laboratories.

Therapeutics

Topically administered antiviral agents have little or no effect on skin lesions and as such are not indicated. Topical antiviral therapy with acyclovir ointment or trifluridine is often given if there is a keratitis. This is probably not necessary as the keratitis is almost always self-limiting and non-scarring.

Dosing: Oc. acyclovir or G. trifluridine 1% × 5 daily for 10 days.

Herpes zoster ophthalmicus (HZO)

Shingles

DESCRIPTION

Herpes zoster ophthalmicus (HZO) is caused by infection of the first division of the trigeminal (fifth) nerve (V1) with the herpes varicella zoster virus (VZV). The same virus causes chicken pox (varicella), usually in children. After the original chicken pox infection the virus lies dormant in the sensory neuronal cell body, in the dorsal root ganglion, or trigeminal ganglion. Reactivation with development of the typical vesicular lesions of shingles usually occurs in adult life, often in people over 60 years of age.

For unknown reasons a disproportionate percentage of shingles (approximately 15%) involves the first (ophthalmic) division of the trigeminal nerve. About 50% of these infections involve the nasociliary branch of the ophthalmic nerve and lead to ocular involvement, occasionally directly infecting the retina with acute retinal necrosis. A significant percentage of patients with HZO also show signs of involvement of the maxillary (second) division of the trigeminal nerve. Thankfully, only a small percentage of HZO leads to ongoing eye disease.

The major ophthalmic importance of HZO is its ability to cause widespread and irreversible damage to ocular and periocular tissues. The infection can be divided into three main phases: an acute phase, where there is massive viral replication; a post-acute phase, with tissue inflammation largely due to an inflammatory reaction to residual viral antigen; and a chronic phase, secondary to tissue damage from the first two phases.

The long-term complications of HZO fall into four main groups:

- nerve damage – nerve palsies, optic neuropathy, neurotrophic keratitis
- scarring – trichiasis, canalicular obstruction, ectropion, raised IOP from scarring of the trabecular meshwork
- persistent inflammation – disciform keratitis, scleritis, uveitis, cataract
- dry eye – ductule damage, chronic blepharitis.

SYMPTOMS

The onset of the rash is often preceded by a viral prodrome and a burning or tingling sensation affecting the skin around the eye and forehead. Photophobia, blurring of vision, and increased lacrimation may occur to a varying degree depending on the extent of ocular involvement. Following resolution of the rash, post-herpetic neuralgia frequently occurs with burning and paresthesia over the affected area. This is uncommon in patients less than 40 years of age.

SIGNS

The characteristic non-confluent vesicular skin rash is restricted to the dermatome of the ophthalmic nerve (V1) with the rash extending from the occiput to the upper eyelid; it does not cross the midline. The exception is with concurrent involvement of the maxillary nerve (V2) where the rash extends to the cheek. Vesicles on the tip of the nose (Hutchinson's sign, "herpes on the hooter") indicate involvement of the external nasal branch of the nasociliary nerve and an increased risk of ocular involvement. Preauricular lymphadenopathy is common. Corneal sensation may be reduced; test the unaffected eye first. Early signs of ocular involvement include aqueous cell and flare, corneal microdendrites, and conjunctivitis. Many longer term complications (such as disciform keratitis and mucous plaques) may take weeks or months to develop.

INCIDENCE

Uncommon.

SIGNIFICANCE

May lead to chronic sight-threatening ocular disease.

DIFFERENTIAL DIAGNOSIS

Herpes simplex keratitis.

SEE ALSO

Herpes simplex primary blepharokeratoconjunctivitis; Neurotrophic keratitis; Dry eye syndromes; Lipid keratopathy; Disciform keratitis.



Fig. H-4 Typical vesicular skin lesions of HZO.

MANAGEMENT

Advice

HZO is contagious during the vesicular stage. Patients should be warned about contact with others who have not had chicken pox and who have not been immunized. Particular concern should be taken with regard to contact with pregnant women because of the teratogenic effects of VZV.

Microbiology

Although not commonly performed due to the characteristic nature of the lesions, culture, or more commonly direct immunofluorescent staining, or PCR for the viral DNA can be undertaken from tissue swabs to identify specific viral antigen.

Acute management

Administration of oral antiviral agents is indicated if the patient presents within 72 hours of the onset of the rash. While oral acyclovir was previously the drug of choice, valaciclovir (1 g tds) or famciclovir (250 mg tds) are now favored due to their longer serum half-lives. Both are used for 1 week. They are thought to limit the extent of the rash, possibly decrease the incidence of post-herpetic neuralgia, and probably decrease the incidence and severity of some ocular complications. There is little role for topical antivirals, either early on, when systemic antivirals are more appropriate, or in the longer term management, when viral replication has already ceased.

Dosing: famciclovir 250 mg or valaciclovir 1g PO tds for 7 days commenced within 72 hours of appearance of the rash.

Management of long-term complications

Topical corticosteroids, tear supplements, and anti-glaucoma medication are all used in the management of long-term complications. These are each discussed in their specific articles.

H

Hordeolum – internal and external

Stye

DESCRIPTION

A hordeolum is also known as a stye and is essentially an abscess of an eyelid sebaceous gland. The term is derived from the Latin word for "barleycorn" in reference to the appearance of the lesions. An internal hordeolum results from an acute staphylococcal infection of a meibomian gland. An external hordeolum is an acute staphylococcal infection of a lash follicle and its associated gland of Zeis or Moll. Both occur more commonly in individuals with chronic blepharitis, especially seborrheic, and staphylococcal blepharitis. Both resolve spontaneously with time.

A hordeolum is distinct from (though similar to) a chalazion. An internal hordeolum results from an acute bacterial infection of a meibomian gland while a chalazion results from sterile low-grade inflammation within the gland.

SYMPTOMS

Both internal and external hordeola may be slightly painful. The lesion is often inflamed,



Fig. H-5a Large inferior internal hordeolum.



Fig. H-5b Superior external hordeolum.



and tender to touch. Patients usually present concerned with the possible diagnosis and cosmetic appearance.

SIGNS

Both present as a well-defined subcutaneous nodule and may be multiple. With both, there may be an associated localized surrounding area of cellulitis. An internal hordeolum is larger than an external hordeolum and located deep within the tarsal plate. An external hordeolum affects the lid margin at the base of the lashes. Both contain purulent material and may rupture, discharging their contents either anteriorly or posteriorly. An internal hordeolum also leads to lid swelling and distortion of the lid margin. Unlike with a chalazion, hordeola are tender, and the overlying skin erythematous.

INCIDENCE

Common, especially in patients with risk factors as listed above.

SIGNIFICANCE

Normally only a cosmetic problem.

DIFFERENTIAL DIAGNOSIS

Chalazion; Eyelid hematoma; acute Dacryocystitis; Preseptal cellulitis.

SEE ALSO

Blepharitis – posterior; Blepharitis – anterior; Rosacea keratitis; Eyelid trauma (hematoma).

MANAGEMENT

Lid hygiene

Warm compresses and lid hygiene may aid resolution of the acute infection and prevent recurrences.

Therapeutics

A week-long course of a broad-spectrum topical antibiotic drop qid is often useful in resolving the infection, especially with an external hordeolum. The best response to topical antibiotics is seen if they are used within several days to a week of the onset of the hordeolum.

Oral antibiotics

A 1-week course of oral antibiotics is indicated if there is significant associated cellulitis. If recurrent and associated with posterior blepharitis, a 3-month course of oral doxycycline may be helpful in reducing recurrences.

Surgery

Epilation may be useful in hastening drainage of pus from around an external hordeolum, while incision and curettage may be useful in draining an internal hordeolum. Following drainage, topical antibiotics are commonly used.

H Hyphema – blunt trauma

DESCRIPTION

Hyphema refers to the presence of blood within the anterior chamber. This usually occurs secondary to blunt trauma or following recent iris surgery but may also occur spontaneously in patients with juvenile xanthogranuloma and rubeosis iridis. Traumatic hyphemas are fairly common, especially in younger patients, following blunt trauma and result from damage to an iris or angle vessel. Of themselves, they are fairly innocuous and transient. Rarely, a secondary – usually more severe – hyphema (rebleed) can occur several days to 2 weeks following the original bleed. This probably occurs due to normal thrombolysis of the original clot prior to repair of the damaged blood vessel and can totally fill the anterior chamber (an "eight-ball" hyphema).

The importance of the presence of a hyphema is its association with acute glaucoma, damage to other intraocular and periocular structures, and the risk of corneal staining. A rebleed is frequently associated with a dramatic rise in IOP.

SYMPTOMS

The hyphema itself leads to blurred vision. This may be worse on waking as the hyphema can settle across the pupil while the patient is sleeping. If there is an associated rise in the intraocular pressure there may also be a dull ache in and around the eye. Damage to other intraocular structures can also lead to blur. An associated orbital fracture can lead to diplopia.

SIGNS

The anterior chamber blood is usually both circulating within the aqueous humour and settled. Red blood cells circulating within the anterior chamber, with no settled inferior layer, is termed a microhyphema. With time it all becomes settled and starts to contract as it dissipates; it may even separate with a whitish surface layer. Associated iris damage is common, with an irregular or dilated pupil, and signs of sphincter rupture with a tear at the pupil margin. Angle recession is also common and may predispose to glaucoma at a later stage. Damage that may occur to other intraocular structures includes anterior damage with corneal edema and zonular dehiscence (with phacodonesis or subluxation of the lens) and posterior damage with retinal dialysis, retinal detachment, vitreous detachment, choroidal rupture, and commotio retinae.

A combination of an extensive hyphema with a high intraocular pressure can lead to significant corneal opacification from staining with hemoglobin. This can occur very rapidly and may be irreversible or take years to resolve.

INCIDENCE

Uncommon overall except when following blunt ocular trauma.

SIGNIFICANCE

May be associated with damage to other intraocular structures and may lead to a rise in intraocular pressure.

SEE ALSO

Corneal and conjunctival lacerations; Eyelid trauma (hematoma); Hypopyon.

MANAGEMENT

Advice

Patients with a hyphema involving less than one-half the anterior chamber should be reviewed in several days to check the intraocular pressure. Patients with a hyphema involving greater than one-half the anterior chamber need to be reviewed daily until the hyphema has substantially resolved. Patients should be advised to present immediately if they have symptoms of a rebleed with decreased vision or ocular ache. Patients should also be advised to avoid vigorous physical activity for several weeks following a hyphema as this may predispose to a rebleed. Aspirin and other non-steroidal anti-inflammatory drugs (NSAIDs) should be avoided because of their anti-thrombotic affect.

Review

All patients should be checked at 1 month and 6 months following the injury to check for a late pressure rise. The angle should

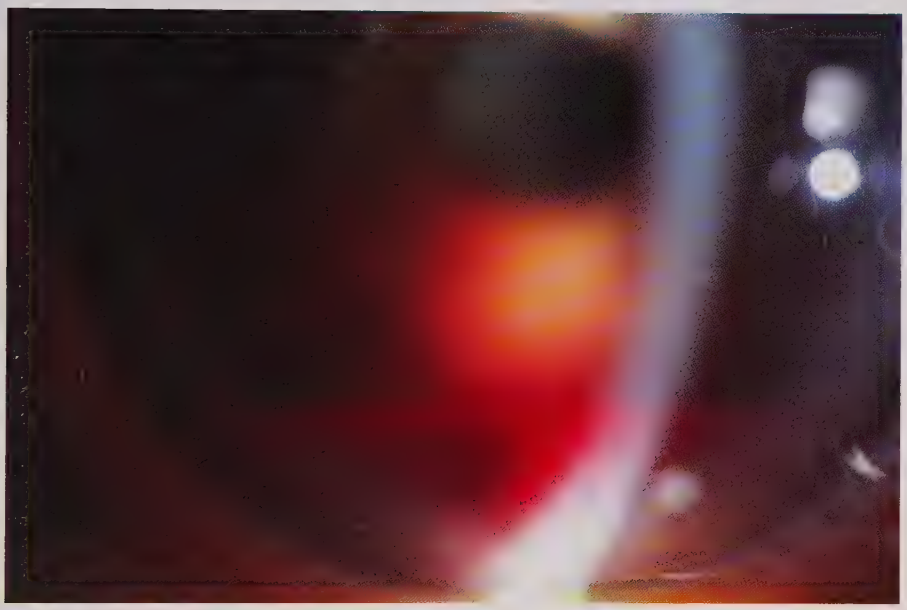


Fig. H-6 Approximately 2-mm high settled traumatic hyphema.

also be examined for evidence of angle recession. If present, longer term pressure checks are appropriate.

Blood tests

Patients of African ancestry who experience a hyphema are investigated with a blood smear to look for sickle cell hemoglobinopathy. Patients with this disease are at an increased risk of acute glaucomatous nerve damage and require aggressive management of any rise in the IOP.

Therapeutics

Most patients are not dilated acutely to check for posterior damage unless the trauma was severe or there is some other indication of posterior segment damage. If they are dilated, it is usually thought best to keep the patient dilated with regular homatropine (tds) or with atropine, to prevent excessive iris movement, as this may predispose to a rebleed.

Any pressure rise is treated on its merits with topical anti-glaucoma medication, oral acetazolamide, or even hyperosmotic agents such as mannitol.

Some clinicians use systemic or (more recently) topical aminocaproic acid, an anti-thrombolytic agent, to stabilize the clot and thereby reduce the risk of a secondary hemorrhage. This is usually only used with a complete hyphema ("eight-ball" hyphema) or where there has already been one rebleed. If corneal blood staining occurs, a trial of topical treatment with the chelating agent desferroxamine may be tried.

Surgery

If a rebleed occurs, urgent assessment is required. Surgical evacuation of the anterior chamber may be required to control the intraocular pressure and to prevent corneal bloodstaining.

H Hypopyon

DESCRIPTION

A hypopyon is a collection of white blood cells (lymphocytes and neutrophils) that has settled within the anterior chamber. It is part of an overall inflammatory reaction and associated with other signs of anterior chamber inflammation.

The inflammation may result from either infectious or non-infectious causes. With endophthalmitis, the hypopyon results from direct intraocular infection whereas, with microbial keratitis, it is due to the effect of bacterial toxins released within the cornea. Severe anterior uveitis, anterior segment ischemia, reaction to an intraocular foreign body and, rarely, fulminant corneal graft rejection or following intraocular surgery (typically a cataract extraction) in a diabetic with extensive retinopathy can lead to a sterile hypopyon. It has also been reported with tight-fitting bandage contact lenses. However, this may be due to bacterial toxins.

SYMPTOMS

Any cause of intraocular inflammation significant enough to cause a hypopyon also leads to blurring of vision, photophobia, and increased lacrimation. There may also be symptoms related to the underlying cause.

SIGNS

Other signs of anterior segment inflammation that should be looked for are aqueous cell and flare, a fibrinous exudate, iris nodules, posterior synechia formation, and keratic precipitates. Anterior chamber cell density can be graded from trace to +4 by counting the number of cells present in a 1-mm by 3-mm oblique slit beam under high magnification:

<1 cell = Trace
5–10 cells = +1
11–20 cells = +2
21–50 cells = +3
>50 cells = +4

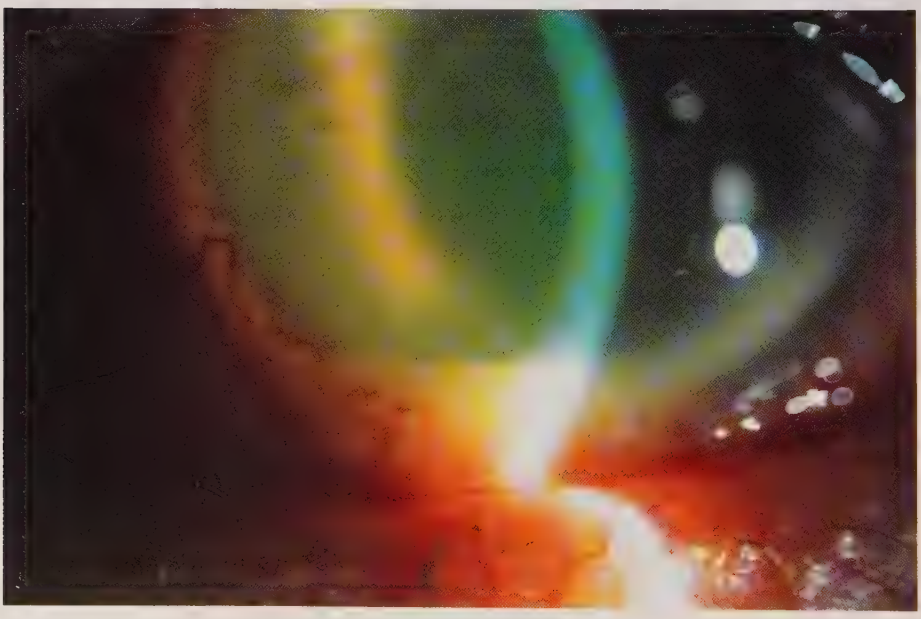


Fig. H-7 Hypopyon with associated corneal edema and anterior chamber cell and flare.



If the inflammation is chronic, there may be an associated anterior cataract. The intraocular pressure should be assessed for an acute rise from a trabeculitis, slight decrease from anterior uveitis, or chronic rise from scarring of the trabecular meshwork. Posterior segment signs include vitreous cells and flare, especially with endophthalmitis and vasculitis, or retinitis with posterior uveitis syndromes such as Behçet's disease.

INCIDENCE
Rare.

SIGNIFICANCE
Always associated with significant sight-threatening disease.

DIFFERENTIAL DIAGNOSIS
Ghost cell glaucoma; phacolytic glaucoma; pseudohypopyon of retinoblastoma (see "Leukocoria").

SEE ALSO
Endophthalmitis; Iritis; Uveitis anterior – overview; Corneal graft rejection; Hyphema; Bacterial keratitis.

MANAGEMENT
Emergency
Because of its ability to cause rapid and permanent visual loss, endophthalmitis needs to be excluded. A hypopyon occurring in the first days to weeks following any intraocular surgery is very suggestive of endophthalmitis and needs to be assessed immediately. Endogenous endophthalmitis can also occur. As such, the lack of recent intraocular surgery is of itself insufficient to exclude the diagnosis of endophthalmitis. The other causes of hypopyon are also sight-threatening and require immediate treatment.

Therapeutics
The anterior segment inflammation is treated with a combination of topical corticosteroids to reduce inflammation, a cycloplegic to prevent posterior synechia formation, and any treatment necessary for the underlying condition.

Surgery
Although surgery is not indicated in the treatment of most of the conditions that cause a hypopyon, endophthalmitis requires prompt injection of intravitreal antibiotics and corticosteroids with a microbiological specimen taken before injection (see "Endophthalmitis").

Infectious crystalline keratopathy (ICK)

Arborescent bacterial keratopathy

DESCRIPTION

Infectious crystalline keratopathy (ICK) is a rare corneal stromal infection usually associated with chronic topical corticosteroid usage. It typically involves a corneal graft, focused at the graft–host junction, although it also occasionally occurs in other (usually scarred and vascularized) corneas with reduced resistance to infection. ICK is typically caused by beta-haemolytic streptococci (*Streptococcus viridans* species organisms), which are slow growing, low virulence bacteria. It may also be caused by more aggressive organisms, such as atypical mycobacteria, and fungi, typically yeast.

Other forms of corneal crystalline deposits differ significantly from ICK, and are discussed under "Crystalline keratopathy."

SYMPTOMS

The patient is usually asymptomatic early on in the disease, the characteristic lesion being picked up on routine examination. As the disease progresses blurring of vision and ocular irritation occur.

SIGNS

The typical lesion is a branching, feathery, chalky-white crystalline lesion involving the anterior to mid-stroma. Occasionally these crystalline changes are seen at the edge of a more usual infective corneal infiltrate. The overlying epithelium is usually intact. In a corneal graft, the affected area arises from the graft–host junction and extends into the graft. Even in a corneal graft, the surrounding cornea is clear and compact, and there are no keratic precipitates or hypopyon, although a mild anterior chamber inflammatory reaction with trace cells and flare may be present. With time, the affected tissue becomes thinned and ectatic. Throughout the course of the disease, the conjunctiva usually remains relatively uninflamed.

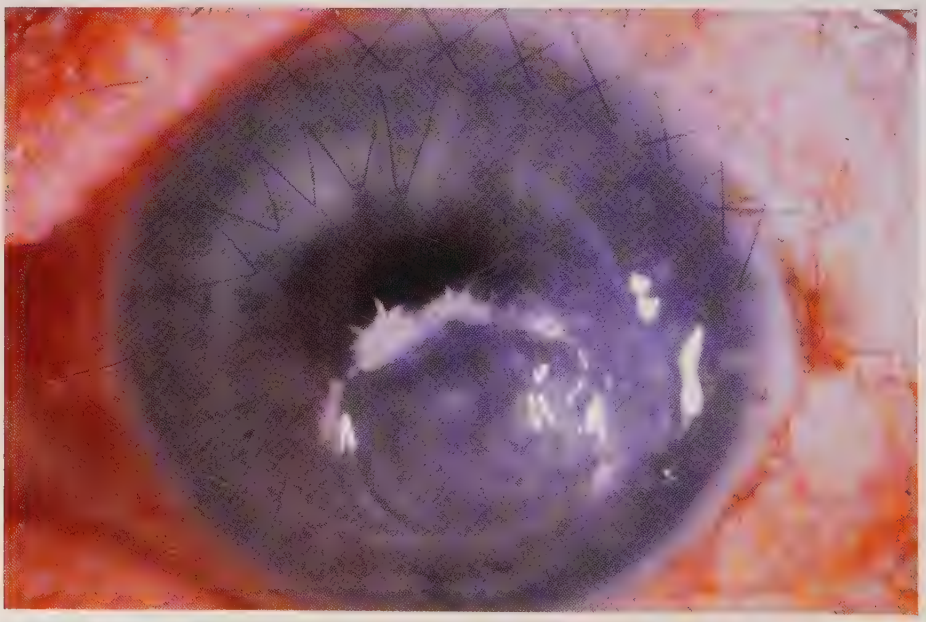
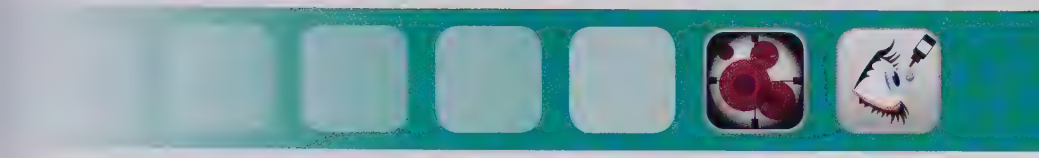


Fig. I-1 Extensive area of infectious crystalline keratopathy involving the graft–host junction in a patient with a previous penetrating keratoplasty.



INCIDENCE
Rare in corneal grafts, very rare in debilitated corneas, and almost never occurs in normal healthy corneas.

SIGNIFICANCE
Although it initially appears quite innocuous and indolent, it does progress with time and is a significant threat to the survival of a corneal graft.

DIFFERENTIAL DIAGNOSIS
Lipid keratopathy; paraproteinemia; Schnyder's crystalline dystrophy.

SEE ALSO
Bacterial keratitis; Corneal graft; Crystalline keratopathy; Fungal keratitis.

MANAGEMENT
Microbiology
Although clinical observation is usually sufficient to make the diagnosis, the collection of microbiological specimens is important to identify the specific infective organism. The causative organism is usually quite slow-growing and fastidious and, as such, may be difficult to identify in the laboratory.

Therapeutics
Treatment is with topical antibiotics. Traditionally, topical 2.5% vancomycin is the treatment of choice. With the introduction of topical fluoroquinolones, ciprofloxacin, ofloxacin, or moxifloxacin are now commonly used. Usually, treatment is initially the administration of topical antibiotics 2-hourly. This is tapered down to qid and maintained for months. During this stage, topical steroids are kept to a minimum or ceased if possible. Some clinicians believe surgical intervention is necessary early in the disease, although medical cures are quite common. If caused by a fungal infection treatment with antifungal medication is required.

Dosing: G. ciprofloxacin/ofloxacin/moxifloxacin q2h day and q4h night for 3 days followed by q2h for 1 week then qid for several months.

Interstitial keratitis (IK)

DESCRIPTION

Interstitial keratitis (IK) refers to an active inflammatory process within the corneal stroma. The key to the definition is that the inflammation is primarily stromal, not arising from an epithelial or endothelial lesion. There are several different causes of IK.

Infectious causes

- Bacteria:
  - —Mycobacteria: tuberculosis (*M. tuberculae*); leprosy (*M. leprae*)
  - —Spirochetes: syphilis (*Treponema pallidum*); Lyme disease (*Borrelia burgdorferi*)
- Viruses: herpes viruses (HSV, HZV)
- Parasites: onchocerciasis

Autoimmune causes

- Cogan's syndrome

Many of these causes can lead to life-threatening systemic disease. Syphilitic IK occurs with both congenital and acquired syphilis. Although the onset of keratitis with congenital syphilis is usually between the ages of 5 and 15 (termed syphilitic rosacea), acquired syphilitic IK may occur at any age. Cogan's syndrome is very rare and may be associated with a life-threatening vasculitis.

SYMPTOMS

Most causes of IK lead to photophobia, blurring of vision, ocular ache, and increased lacrimation. Most are bilateral, although HSK and HZO are usually unilateral. A thorough history should be taken to elicit any non-ocular symptoms of a systemic infection such as fever, weight loss, chronic cough, or skin rashes. A history of an HZO rash should also be sought as should a recent history of hearing loss as occurs with Cogan's syndrome.

SIGNS

During the active stage of the disease, IK results in stromal inflammation with edema, corneal opacities and frequently stromal vascularization. Resolved, inactive IK leads to typical corneal signs with opacification, thinning, and "ghost vessels" (old regressed vessels devoid of blood). The pattern of vascularization and opacification often gives a clue to the underlying cause.

Both acquired and congenital syphilis lead to an acute episode of usually bilateral IK. This results in deep stromal vascularization, scattered corneal opacities, and stromal thinning. There is associated iris atrophy with a typical flattened featureless iris, characteristic Argyll–Robinson pupil signs, and pupillary changes with miosis, pupillary scarring, and irregularity. Other non-ocular signs of congenital syphilis include dental abnormalities, sabre shins, hearing abnormalities, and a depressed bridge of the nose.

Cogan's syndrome leads to a much less marked IK with faint scattered opacities and usually no vascularization. It can also rapidly lead to marked irreversible hearing loss.

INCIDENCE

Very rare.

SIGNIFICANCE

May lead to bilateral reduction in vision. May also be associated with life-threatening systemic infection or autoimmune disease.

DIFFERENTIAL DIAGNOSIS

Old corneal injury; Chemical burn; old stromal necrotic Herpes simplex keratitis; Trachoma.

SEE ALSO

Herpes simplex keratitis – overview; Herpes zoster ophthalmicus.

MANAGEMENT

Urgent

Cogan's syndrome can rapidly lead to permanent hearing loss and even death from large vessel vascular disease. Therefore, if considered, the patient should be investigated immediately by both an ear, nose, and throat specialist and a physician.

Additional investigations

Patients with active IK should be investigated extensively for an underlying cause of the disease. This should include a Venereal Disease Research Laboratory slide test (VDRL),

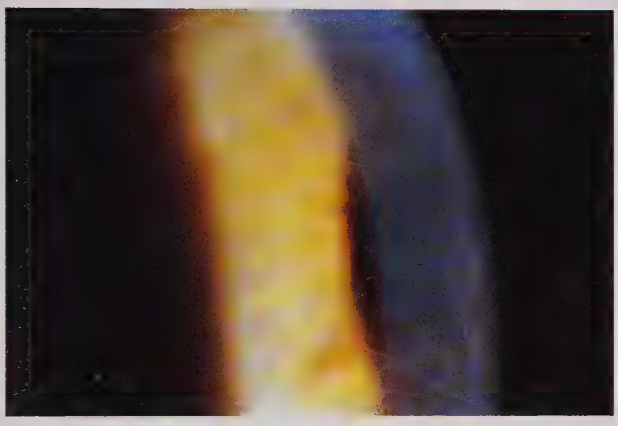


Fig. I-2a Fine "ghost vessels" deep within the corneal stroma in interstitial keratitis.

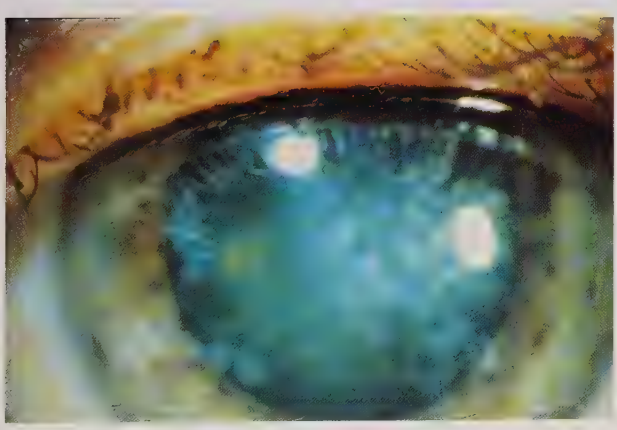


Fig. I-2b Typical scarring of resolved IK.

fluorescent treponemal antibody absorption test (FTA-ABS), erythrocyte sedimentation rate (ESR), C-reactive protein, Lyme disease antibodies (if the patient has been to an endemic area), and a full blood examination. A chest X-ray, Mantoux test, and skin biopsy may be considered for tuberculosis and leprosy. Audiology can be useful to identify hearing loss in Cogan's syndrome. Patients with old IK scarring should also be investigated with syphilis serology.

Therapeutics

Regardless of the cause, the corneal inflammation seen during active IK is treated with topical corticosteroids (such as prednisolone acetate 1% 2-hourly).

Systemic medication

Syphilis, TB, leprosy, and onchocerciasis all require systemic treatment with appropriate anti-microbial agents. Cogan's syndrome is treated with oral corticosteroids.

Notifiable

In some countries syphilitic infection may need to be reported to public health authorities.

Intraocular lenses (IOLs)

DESCRIPTION

Sir Harold Ridley was the first to insert intraocular lenses in the late 1940s. This followed his observation of the inert nature of polymethyl methacrylate (PMMA) windscreen fragments in the eyes of heroic fighter pilots shot down during the Battle of Britain. Since then, intraocular lenses (IOLs) have evolved enormously. There are three components to a modern IOL: the central refracting element (the optic) and two supporting structures (haptics). Most haptics are loops, usually a modified "C" or "J" shape, although some silicone IOLs have "plate haptics," flat haptics extending from the optic. So-called "in the bag" IOLs, those designed to sit within the capsule of the lenses, usually have a relatively sharp edge to the optic to decrease the risk of developing postoperative posterior capsular opacity. All IOLs contain a UV filter while some also have a blue-light filter, making them a soft yellow color. This was introduced because of some experimental laboratory results suggesting a role for blue light in retinal damage, although there is no clinical evidence to support this finding. Current lenses can be grouped into three classifications.

Phakic or pseudophakic IOLs

Most IOLs are inserted to correct aphakia after cataract extraction. This is usually done at the time of the cataract extraction; if performed later, this is referred to as a secondary IOL. IOLs are also increasingly being placed in phakic eyes to correct refractive errors.

Rigid (PMMA) or foldable IOLs

The original IOLs were rigid and were made of PMMA. They required a wound of 6.0–6.5 mm to be inserted. Since the early 1990s surgeons have sought IOLs to complement small-incision phacoemulsification surgery. This has resulted in the popularity of foldable IOLs capable of being inserted through wound sizes as small as 2.0 mm. Foldable IOLs now account for the vast majority of all IOLs inserted in developed countries. Foldable IOLs are most commonly made from hydrophobic acrylic, hydrophilic acrylic, or silicone.

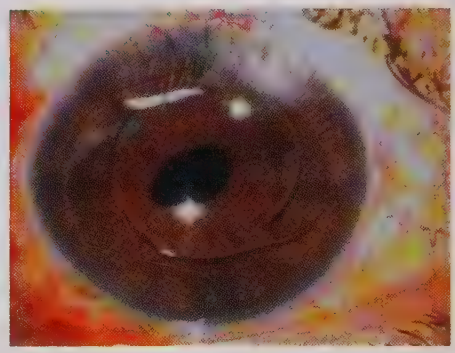


Fig. I-3a Rigid, PMMA anterior chamber IOL.

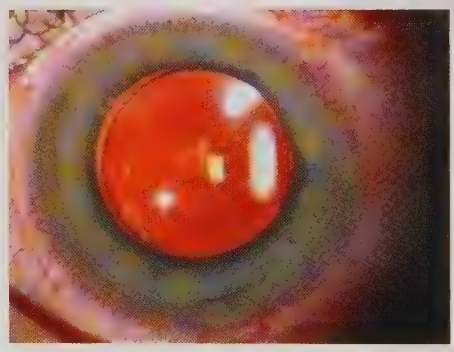
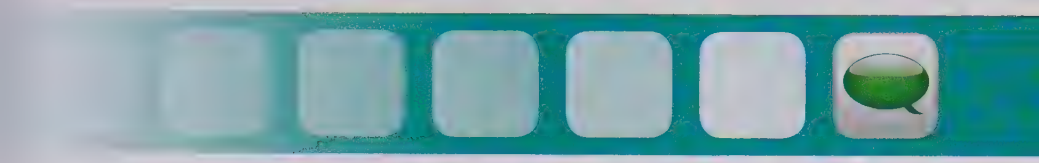


Fig. I-3b Foldable, acrylic posterior chamber IOL in the capsular bag.

Anterior or posterior chamber IOLs

Lens placement depends largely on the availability of adequate support. In cataract surgery, the preferred site for IOL placement is in the capsular bag (so-called "in the bag fixation"). If rupture of the posterior capsule or zonular dehiscence occurs, the IOL may have to be placed with the haptics in the sulcus or angle. Rarely, if there is insufficient capsular or angle (iris) support, an IOL may be sutured into the



sulcus (scleral fixation). Most "in the bag" IOLs with loop haptics can also be placed in the sulcus.

An anterior chamber IOL is supported in the angle, resting in the trabecular meshwork and requiring iris support. The haptics of an anterior chamber IOL are quite different from those of a posterior chamber IOL. Both anterior chamber and posterior chamber IOLs have open loop haptics. However, anterior chamber IOLs provide four-point support in the angle and the overall length of an anterior chamber IOL is longer (usually 13–14 mm versus 12.5 mm for a posterior chamber IOL). Placement of an anterior chamber IOL requires a peripheral iridectomy to be made to prevent the development of pupil block glaucoma.

Other refractive modification

While most IOLs are spherical, several refractive modifications have been introduced over the past decade. Toric IOLs have become very popular because of their ability to reduce postoperative astigmatism in patients with pre-existing astigmatism. In addition, many foldable IOLs are aspheric, designed to address higher order aberrations and increase visual performance, especially under low light conditions. Many IOL companies also make so-called "premium" multifocal (refractive or diffractive) or accommodating IOLs. These are designed to provide both clear distance and reading vision.

SYMPTOMS

The presence of an IOL is usually totally asymptomatic. Occasionally, patients can experience glinting or even halos around lights from surface or edge reflections. This is particularly the case if the IOL has subluxed (sunset syndrome).

SIGNS

Aside from assessing the position and centration of the IOL, signs of any complications of intraocular surgery should be sought. Any intraocular surgery, including placement of an IOL, can lead to a range of early and late postoperative complications. These include endophthalmitis (usually within the first week after surgery), bullous keratopathy, cystoid macular edema (CME), posterior capsular opacification (PCO), and retinal detachment. Most of these complications are more common if the surgery was complicated with posterior capsular rupture and disruption of the anterior vitreous face. The wound can also lead to the induction of astigmatism.

INCIDENCE

Very common. Approximately 25% of all 80-year-olds in developed countries have already had a cataract extraction and IOL insertion. The placement of phakic IOLs to correct refractive errors is also increasing in frequency.

SIGNIFICANCE

May lead to intraocular complications as listed above.

SEE ALSO

Cataract incisions; Endophthalmitis; Bullous keratopathy; Posterior capsular opacification.

MANAGEMENT

Advice

The patient can be reassured that the IOL will last forever (a commonly asked question) and does not require any ongoing care. They should be counselled to re-present if they notice any blurring of vision. Posterior capsular opacification (PCO) is less common with most foldable than with rigid lenses. However, it still occurs at a rate of between 5% and 15%.

Iridocorneal dysgenesis

Anterior segment cleavage syndromes, anterior segment dysgenesis, iridogoniodysgenesis, Axenfeld–Rieger syndrome (ARS)

DESCRIPTION

The iridocorneal dysgenesis syndromes are a group of heterogeneous, very rare, congenital, usually bilateral conditions resulting from abnormal embryological development of the anterior segment. The main syndromes in this group of disorders are Axenfeld syndrome, Rieger's syndrome, and Peters' anomaly, and they appear to be a continuum of the same developmental defect. For this reason they are frequently referred to as Axenfeld–Rieger syndrome (ARS). Being developmental, the underlying defect is non-progressive, although associated pathology, such as glaucoma and cataract, may progress. They are frequently associated with other developmental abnormalities affecting branchial arch (facial) structures as well as, more rarely, cardiac and renal abnormalities, hydrocephalus, and congenital hip dislocation.

The spectrum of disease represents a continuum and, as such, specific classification into one distinct entity may not always be possible. The main structure affected throughout the spectrum of disease is the anterior trabecular meshwork, with a prominent and anteriorly displaced Schwalbe's line. This is termed posterior embryotoxon and is of itself a normal variant, not associated with cataract and glaucoma. With progressively more severe disease, abnormalities include iris defects, glaucoma, cataract, posterior corneal irregularities, corneal opacification, corneal edema, lack of separation of the lens from the cornea, and persistent hyperplastic primary vitreous (PHPV). Many of the mutations responsible for this syndrome appear to occur in either the *PITX2* (pituitary homeobox transcription factor 2) or *FOXC1* (forkhead box transcription factor C1) genes.

SYMPTOMS

Glare and blur are common with all these conditions except posterior embryotoxon and Axenfeld's.

SIGNS

As discussed above, the signs of these conditions fall into a continuum. A constant feature is the presence of posterior embryotoxon. Ocular involvement is usually bilateral but often asymmetric with, for example, one eye having features of Axenfeld's and the other Rieger's.

Axenfeld's anomaly

There is prominent posterior embryotoxon with multiple iris strands (anterior synechia). This condition is rarely associated with glaucoma and is frequently an incidental finding.

Rieger's anomaly

This is more severe than Axenfeld's, with significant iris disruption, and frequently glaucoma. However, the cornea remains clear.

Peters' anomaly

This is similar to Rieger's, but there is the added abnormality of maldevelopment of the cornea with corneal opacification, adherent iris (adherent leukoma), and partial absence of Descemet's membrane. In very advanced cases, there may also be failure of separation of the lens from the cornea as well as PHPV. Other (now rarely used) terms applied to various forms of Peters' include posterior keratoconus and Von Hippel's internal corneal ulcer. Both Peters' and Rieger's anomalies may be part of a wider syndrome with facial and particularly dental abnormalities.

INCIDENCE

All are extremely rare.

SIGNIFICANCE

Rieger's and Peters' are associated with glaucoma and, in the more severe forms, cataract, corneal edema, and opacification.

DIFFERENTIAL DIAGNOSIS

Aniridia; iridoschisis (not included in this text); Iridocorneal endotheliopathy; Epithelial and fibrous ingrowth.

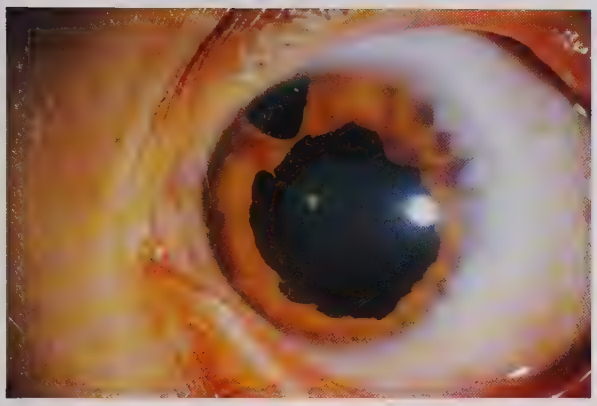


Fig. I-4a Rieger's anomaly with ectropion uveae.



Fig. I-4b Posterior embryotoxon (most marked temporally).

SEE ALSO
Corneal edema.

MANAGEMENT

Genetic advice
These are all inherited diseases; an affected child may have less or more severe disease than their parents. ARS is often inherited as an autosomal dominant trait with a high degree of penetrance, whereas Peters' anomaly typically occurs sporadically.

Therapeutics
Topical anti-glaucoma medication is required if the IOP is raised and the optic nerve head threatened.

Surgery
Glaucoma-filtering surgery may be required to control the IOP if topical medication is insufficient. A corneal graft may by indicated in Peters' anomaly with significant bilateral corneal opacification. Unfortunately, both surgeries may need to be performed in an infant or child where a successful outcome to surgery is less likely.

Iridocorneal endotheliopathy (ICE)

Iridocorneal endothelial syndrome

DESCRIPTION

The iridocorneal endotheliopathies (ICE syndromes) are a rare group of acquired, non-inherited, progressive anterior segment diseases. They primarily affect the iris and cornea, but progress to involve the trabecular meshwork and crystalline lens.

The cause of the ICE syndromes is unknown, but is presumed by many clinicians to be secondary to herpes simplex virus infection. There are three somewhat overlapping ICE syndromes:

- iris nevus syndrome
- Chandler's syndrome
- essential (progressive) iris atrophy.

The first letter from each of these syndromes forms the mnemonic "ICE." The common link between the three syndromes is epithelialization of the endothelial mosaic. The abnormal cells then migrate across the angle and onto the iris. This membrane then contracts and leads to the characteristic iris changes.

SYMPTOMS

Photophobia, blurred vision, and ocular irritation are all common from corneal edema, glaucoma, and cataract.

SIGNS

The disease is almost always unilateral. The iris changes are present in all three syndromes, being most dramatic in essential iris atrophy with polycoria and corectopia. In Chandler's syndrome, the iris appears almost normal, with variable involvement, flattening of the iris ridges, and slight corectopia. In iris nevus syndrome, there are either multiple pedunculated nevi or a diffuse nevus. In all three, there is corneal involvement with stromal edema, microcystic epithelial edema, and an abnormal "hammered-silver" reflex from the corneal endothelium. The corneal edema is very pressure-sensitive and may deteriorate significantly with only a slight rise in intraocular pressure. Cataract



Fig. I-5 Gross iris disruption in essential iris atrophy with pigmentary defects, full-thickness iris holes (polycoria), and displacement of the pupil (corectopia).



is common, and gonioscopy reveals broad peripheral anterior synechia. Glaucoma is thought to occur from trabecular meshwork blockage, from invasion by both the epithelial membrane and peripheral anterior synechia.

INCIDENCE

All syndromes are very rare. However, Chandler's syndrome is the most common of the three. They are more common in women, and onset is usually in the fourth decade of life.

SIGNIFICANCE

All lead to corneal decompensation and often intractable glaucoma. It is common for the affected eye to be blinded over a period of years despite active treatment and multiple operations.

DIFFERENTIAL DIAGNOSIS

Trauma (surgical and non-surgical); Iridocorneal dysgenesis; iridoschisis (not included in this text); Aniridia; Epithelial and fibrous ingrowth; Posterior polymorphous dystrophy.

SEE ALSO

Corneal edema.

MANAGEMENT

No treatment is required in the absence of glaucoma and corneal edema.

Review

Patients should be reviewed 6-monthly because of the high risk of glaucoma.

Ocular investigations

Specular microscopy, if possible through the corneal edema, may reveal the abnormal epithelial-like cells present on the endothelial surface.

Therapeutics

Topical anti-glaucoma medications are commonly required. Topical carbonic anhydrase inhibitors are not appropriate, as they may lead to a reduction in residual endothelial pump function, exacerbating the corneal edema. Topical hypertonic saline drops and ointment are occasionally used to clear microcystic corneal epithelial edema.

Surgery

A trabeculectomy or drainage seton is often required but prone to failure. In addition, a corneal graft is often required but is also prone to failure.

Iris cysts and benign tumors

Lisch nodule, leiomyoma, adenoma

DESCRIPTION

The iris may give rise to a number of benign tumors arising from the pigment epithelium or stroma. The main importance of these tumors is as a differential diagnosis of iris melanoma, although iris melanoma itself is usually a fairly benign lesion (see "Melanoma and nevus – iris").

Primary nonmelanotic iris tumors can be classified on an anatomical basis as arising from the following cell layers:

- Pigment epithelium
  - —Primary epithelial cysts – usually deeply pigmented and globular with a thin wall and fluid-filled.
  - —Adenoma – commonly occurs in the ciliary body and rarely on the iris. Usually deeply pigmented and occurs adjacent to the angle.
- Stroma
  - —Primary stromal cysts – usually single, and moderate sized (1–3 mm). They are less pigmented and less common than primary epithelial cysts.
  - —Brushfield spots – multiple, small white spots usually seen in the peripheral iris giving it a speckled appearance; they are most commonly seen in Down syndrome. Histopathologically, they are areas of iris hypoplasia.
  - —Leiomyoma – appearance closely resembles an amelanotic melanoma with a rounded border and uneven surface.
  - —Lisch nodules – small raised neurofibromata seen in neurofibromatosis; they are bilateral and usually multiple and lightly pigmented.

Secondary epithelial cysts (termed an inclusion or implantation cyst) may arise from ocular surface epithelium that has been implanted into the anterior chamber. They may occur following a penetrating eye injury or intraocular surgery, typically a corneal graft or cataract surgery. They are thin-walled, fluid-filled, and non-pigmented. Occasionally, they have an internal pseudohypopyon, due to the settling of epithelial squames. Rarely, they may even be free-floating within the anterior chamber.

SYMPTOMS

Symptoms usually only arise if the tumor enlarges sufficiently to obstruct the pupil or angle, leading to glaucoma.

SIGNS

Although secondary ocular pathology is uncommon, all but secondary (implantation) cysts should be accompanied by a thorough examination of the angle with gonioscopy.

INCIDENCE

All of these tumors are rare; the exceptions are Brushfield spots (common in Down syndrome) and Lisch nodules (common in neurofibromatosis).

SIGNIFICANCE

Although benign, both primary and secondary cysts may enlarge and lead to obstruction of the pupil or the angle leading to secondary glaucoma.

DIFFERENTIAL DIAGNOSIS

Epithelial and fibrous downgrowth; Busacca (on pupil margin) and Koeppe (on iris surface) nodules in granulomatous Uveitis.

SEE ALSO

Melanoma and nevus of the iris.

MANAGEMENT

A thorough examination and history will allow a diagnosis to be made (in most cases) of an iris cyst or tumor. Even if there is concern that the lesion may be a melanoma, the usual treatment is documentation and observation, unless the lesion is rapidly progressive or causing other problems.

Advice

Patients should be advised that, if they note any change in the size or color of the lesion, they should re-present for repeat examination.

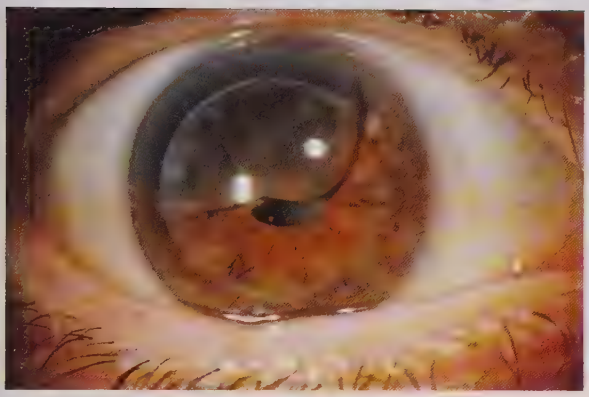


Fig. I-6a Secondary cyst following corneal graft surgery.



Fig. I-6b Lisch nodules in a patient with neurofibromatosis.

Review

While growth is uncommon (in all but secondary cysts) after initial examination, patients should be reviewed annually, preferably with photographic documentation. Any increase in size should lead to more frequent follow-up.

Surgery

If any tumor continues to grow and threatens to occlude the visual axis or significantly invade the angle, surgical excision may be required. This is occasionally preceded by a fine needle aspirate to make a histological diagnosis.

Many different techniques have been used to treat progressively enlarging primary and secondary cysts. These include surgical excision, argon and Nd:YAG laser, cryodestruction, and the injection of either mitomycin C or trichloroacetic acid. All of these carry risk to other structures in the eye, especially the injection of toxic substances capable of damaging the lens, corneal endothelium, and angle structures.

Laser ablation of secondary (implantation) cysts may lead to the liberation of epithelial cells throughout the anterior segment and the development of "sheet-like" epithelial downgrowth. It appears that, currently, the best approach is surgical excision.

Iritis

Anterior acute uveitis (AAU)

DESCRIPTION

Uveitis is classified anatomically, depending on the site of inflammation, as being anterior (including iritis and iridocyclitis), intermediate (pars planitis), posterior, and panuveitis. Iritis is a specific disease entity and, although frequently unassociated with any underlying disease, a significant proportion of patients possess the *HLA-B27* genotype. This is also associated with ankylosing spondylitis and inflammatory bowel disease.

Iritis may also be associated with juvenile rheumatoid arthritis, Reiter's syndrome, and psoriatic arthritis. Iritis is usually unilateral; if bilateral, it is more likely there is an identifiable underlying cause. The most common causes of a bilateral, granulomatous uveitis are tuberculosis, sarcoid, and syphilis. Iritis is also classified according to the severity and chronicity of inflammation as acute, recurrent acute, and chronic.

SYMPTOMS

During an attack, the affected eye is usually red, photophobic, and has a dull ache. There is also often decreased vision and increased lacrimation. Some patients may remain relatively asymptomatic, despite significant inflammation. Symptoms of arthritis, back pain, skin rashes, and urethritis should also be sought.

SIGNS

The classic findings of iritis are circumlimbal injection, anterior chamber cell and flare, keratic precipitates, anterior vitreous cells, and posterior synechia. These signs are described in more detail under the entry "Uveitis."

The pupil is often slightly miotic and slow to react, and the intraocular pressure is often slightly low relative to the other eye. The cell and flare is often quite marked in the anterior chamber but minimal in the anterior vitreous. In granulomatous

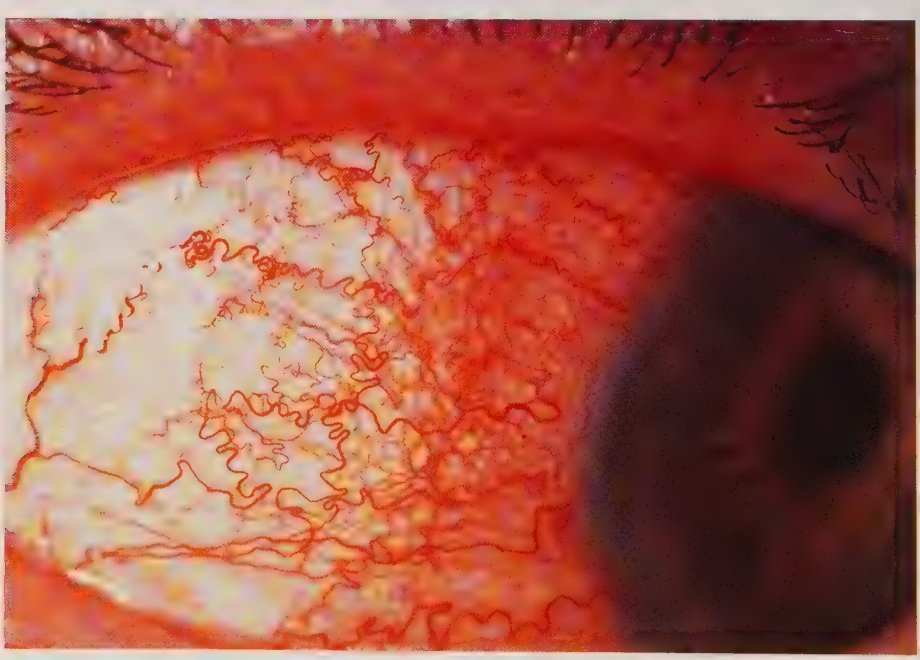
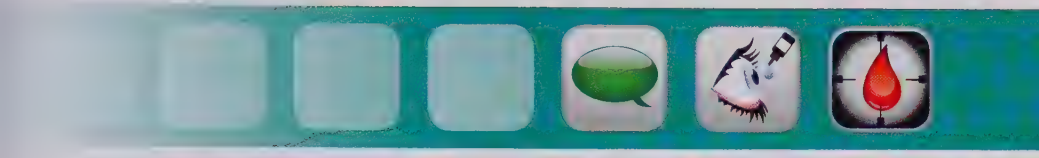


Fig. I-7 Characteristic circumlimbal "ciliary flush" in acute anterior uveitis.



uveitis, the keratic precipitates are typically large, termed "mutton fat" precipitates, and there may be inflammatory nodules on the iris surface (Busacca nodules) or at the pupil margin (Koeppe nodules). Chronic recurrent iritis can lead to a band keratopathy or cataract formation.

INCIDENCE

Rare.

SIGNIFICANCE

May lead to permanently impaired vision from pupil block, open-angle glaucoma, and cataract.

DIFFERENTIAL DIAGNOSIS

Herpes simplex and herpes zoster keratouveitis (Disciform keratitis); Acute angle closure glaucoma.

SEE ALSO

Keratic precipitates; Fuchs' heterochromic iridocyclitis; Endophthalmitis; Hypopyon; Synechia; Glaucoma – acute angle closure; Band keratopathy; Uveitis anterior – overview.

MANAGEMENT

Advice

As iritis is frequently recurrent, patients should be advised to re-present promptly if symptoms recur.

Additional investigations

No investigations are necessary for the first attack of iritis if it is unilateral. Bilateral or recurrent disease is usually investigated for *HLA-B27* positivity. Other tests to identify an underlying cause include angiotensin-converting enzyme (ACE, for sarcoidosis), erythrocyte sedimentation rate (ESR), full blood examination (FBE), antinuclear antibody (ANA, for juvenile arthritis), and a chest X-ray (CXR) and Mantoux test for TB.

Therapeutics

Thankfully, iritis responds well to medical therapy. Initial treatment is with intensive topical steroids such as hourly prednisolone acetate 1%, and a mydriatic such as homatropine 2% tds to prevent posterior synechia formation. If there are already synechia formed, on presentation a drop of homatropine should be instilled and the patient checked 30 minutes later to ensure that they have broken. If they are still present a combination of mydriatics should be instilled, such as phenylephrine 10%, cyclopentolate 1%, and tropicamide 1%, in an attempt to break up the synechia. If this is still ineffective, a subconjunctival injection of mydriatics can be tried.

Following initial treatment, the drops are normally tapered, ceasing the dilating drop first. Mydriasis is not required once the anterior chamber reaction is under control and would add to the photophobia as well as blurring vision. The corticosteroids are usually tapered over 4–6 weeks, unless there is a history of recurrent iritis, when the tapering should be slower.

Iron lines

Fleischer ring, Stocker line, Hudson–Stahli line

DESCRIPTION

Iron can be deposited in the corneal epithelium in several conditions. It is probably derived from the tear film and occurs at the junction of abnormalities in the tear film distribution. Histopathologically, the iron is located predominantly in the basal epithelial layers, although it may also be present in superficial epithelial wing cells.

Some of the commonly observed iron deposition lines have eponymous names. They are:

- Fleischer ring – a circular deposit seen around the base of the cone in keratoconus
- Hudson–Stahli line – a horizontal line seen in normal eyes just inferior to the center of the interpalpebral fissure with a typical "cat's whiskers" appearance at the medial and lateral edges; this may reflect tear film convection flow or the meeting of the upper and lower eyelids on blinking
- Stocker line – seen at the leading (corneal) edge of a pterygium
- Ferry line – seen next to an overhanging filtering bleb.

Iron deposition lines are also occasionally seen following disruption of normal corneal curvature following corneal surgery, such as radial keratotomy and corneal grafts.

SYMPTOMS

The lines themselves cause no symptoms.

SIGNS

All iron lines are best seen with direct observation through the slit-lamp biomicroscope, with maximum illumination



Fig. I-8 A Fleischer ring seen in keratoconus.

and a cobalt blue filter in place. They appear as a dark area in the reflex.

INCIDENCE

A Hudson–Stahli line is very common, especially in older people, and is not associated with any ocular pathology. A Fleischer ring is commonly seen even in mild keratoconus. A Stocker line is common, especially with old, quiescent pterygia. Ferry lines are uncommon.

SIGNIFICANCE

May indicate the presence of another pathology, such as keratoconus with a Fleischer ring.

DIFFERENTIAL DIAGNOSIS

Vortex keratopathy; Krukenburg spindle (see "Endothelial opacities").

SEE ALSO

Keratoconus; Pterygium; Filtering blebs; Corneal opacities – peripheral; Corneal dystrophies, degenerations, and deposits – overview.

MANAGEMENT

No active management is required for the iron deposition itself. Pterygia and keratoconus are treated on their merits.

Keratoconus

Conical cornea

DESCRIPTION

There are three ectatic corneal dystrophies – keratoconus, keratoglobus, and pellucid marginal corneal degeneration. Of these, keratoconus is by far the most common. Keratoconus starts with anterior elevation of the posterior cornea progressing to stromal thinning and bowing forward of the affected cornea, resulting in irregular myopic astigmatism. Onset is usually around puberty; the condition then progresses throughout the teens and into the mid-twenties, usually becoming stable by the late twenties to early thirties. A major recent advance has been the introduction of corneal collagen cross-linking with the ability to prevent progression of the disease.

SYMPTOMS

Aside from induced irregular myopic astigmatism with blurring of vision, glare is a significant problem for people with keratoconus.

SIGNS

Keratoconus is usually bilateral, but may be quite asymmetric. The corneal thinning and ectasia occurs in three main patterns: a nipple cone, where the cone is small, and centered on or near the center of the cornea; an oval cone, where the affected area is larger and usually centered inferotemporally; and a globus cone, where the area of involvement covers most of the central cornea.

Several named signs occur in keratoconus:

- Vogt's striae – deep stromal, usually vertical, stress lines at the apex of the cone
- Munson's sign – bulging forward of the inferior eyelid on downgaze
- Fleischer ring – epithelial iron deposition in a ring around the cone
- Rizzutti's sign – conical reflection on the nasal cornea with shining of a pen light from the temporal side.

The irregular shape of the anterior corneal surface is observed by a scissor or swirling reflex on retinoscopy, an "oil droplet" reflex on slit-lamp retroillumination, irregular and high keratometry readings, and an abnormal corneal map (computer-assisted videokeratoscopy). In advanced keratoconus, scarring may develop in the anterior stroma at the apex of the cone. Spontaneous splits in Descemet's membrane may occur, leading to hydrops (see "Descemet's membrane breaks").

INCIDENCE

Clinical keratoconus is uncommon (about 1/2000). Keratoconus is more common in people with a family history of keratoconus, Down syndrome (approximately 10%), floppy eyelid syndrome, and sleep apnea, and people with ocular allergies, where eye rubbing is considered a significant etiological factor. Many other conditions have been reported to be associated with keratoconus; often, whether these are real or just chance associations is unclear.

SIGNIFICANCE

Keratoconus leads to significant induced myopic astigmatism and can substantially decrease both uncorrected and spectacle-corrected vision. While the cornea may thin, instances of perforation are exceedingly rare.

DIFFERENTIAL DIAGNOSIS

Pellucid marginal corneal degeneration; Keratoglobus.

SEE ALSO

Corneal grafts; Corneal grafts – lamellar keratoplasty; Descemet's membrane breaks; Iron lines; Keratoconus forme fruste; Corneal collagen cross-linking.

MANAGEMENT

Genetic advice

The genetics of keratoconus remain quite obscure. Inheritance is variable. Rarely, it appears to be inherited as an autosomal dominant trait with high penetrance. Most cases are sporadic. If there is no family

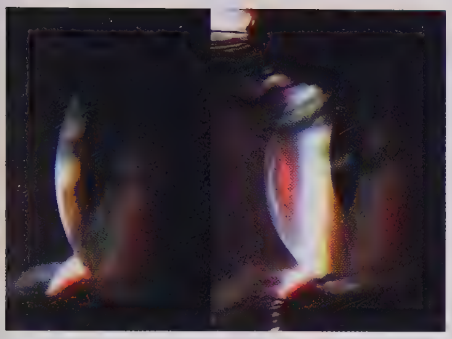


Fig. K-1a Corneal profile of normal and keratoconic eye.

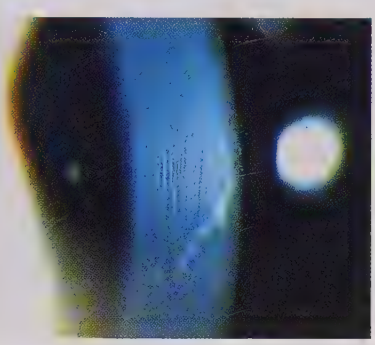


Fig. K-1b Vogt's stria in keratoconus.

history of the disease, patients can be reassured that it is unlikely their children will inherit the disease, the risk being about 1%, although inheritance of slight corneal abnormalities able to be identified on corneal mapping are common.

Advice

Patients with keratoconus should be advised not to have incisional refractive surgery, surface photorefractive keratectomy (PRK), or LASIK, as this can lead to destabilization of the cornea, leading to marked progression of the disease. Patients should also be advised that eye rubbing may lead to progression of the disease.

Contact lenses

Most patients with clinical keratoconus require a rigid contact lens for clear vision. Several options are now available with standard small- or large-diameter RGP lenses, piggyback, hybrid, or reverse geometry lenses, and even preformed mini-scleral lenses for more marked cases.

Corneal collagen cross-linking

Chemical cross-linking of the corneal collagen with riboflavin-5-phosphate activated by UV light has been demonstrated to not only prevent progression of the disease but also frequently results in some flattening of the affected cornea and an increase in best spectacle-corrected visual acuity. Unfortunately, it is frequently not able to be used if the cornea is too thin or scarred.

Surgery

If the patient has poor vision that is unable to be corrected with spectacles or contact lenses, a penetrating keratoplasty (corneal graft) or deep anterior lamellar keratoplasty (DALK) may be appropriate. Other factors such as vision in the other eye, ocular allergies, whether the patient has Down syndrome, the patient's visual requirements, and the age of the patient all impact on the decision of whether or not the patient is suitable for surgery. With advances in rigid contact lens technology, only between 10%, and 20% of patients with clinical keratoconus require surgery. In some cases implantable corneal ring segments are an alternative to corneal grafting and can lead to a significant increase in both uncorrected and spectacle-corrected visual acuity.

K

Keratoconus forme fruste

Sub-clinical keratoconus

DESCRIPTION

With the introduction of corneal surface mapping, a group of patients has been identified with topographical changes consistent with mild keratoconus and steepening of the inferior cornea. Pachymetry often reveals these patients to have a slightly thinner than normal cornea. Despite this they usually retain good spectacle-corrected acuity. This condition has been termed "forme fruste keratoconus." The widespread use of corneal mapping and pachymetry for preoperative refractive surgery screening, especially with LASIK, has revealed that as many as 10% of patients presenting for refractive surgery have evidence of forme fruste keratoconus.

SYMPTOMS

The patient is generally asymptomatic. While there may be irregular myopic astigmatism, spectacle correction or soft contact lens wear is sufficient to meet the

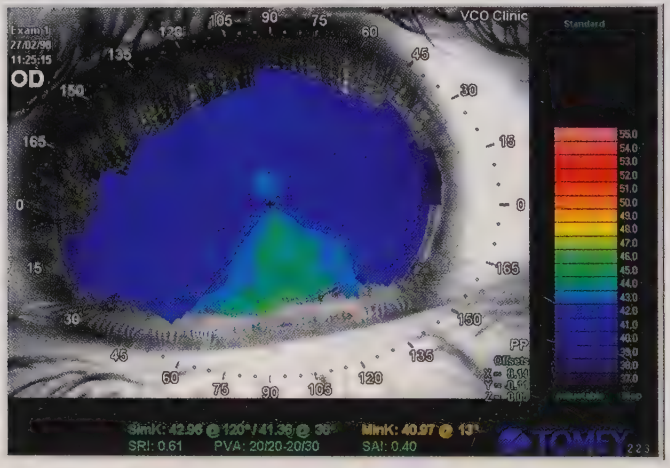


Fig. K-2a Corneal power map of forme fruste keratoconus with inferior steepening with an apical power of 46 D. The patient saw 6/4 (20/12) with a myopic spherical correction.

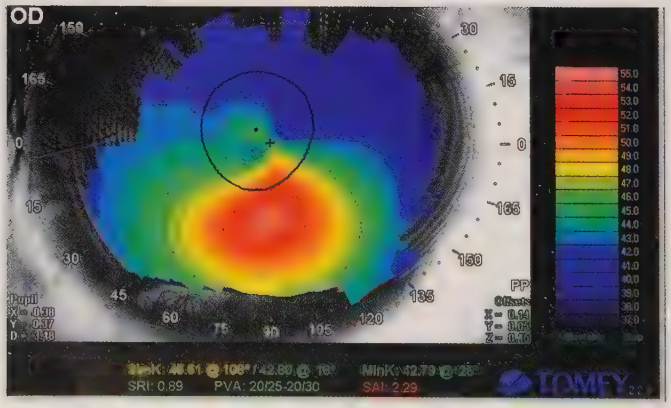


Fig. K-2b Corneal power map of keratoconus with marked inferior steepening. Note that the power scale is the same as for Fig. K-3a.



patient's visual requirements, although there may be occasional mild symptoms such as slight ghosting of lights observed at night.

SIGNS

Keratoconus forme fruste is usually bilateral but may be quite asymmetric. The named signs, such as Vogt's stria, Munson's sign, or Fleischer ring, are not present.

The irregular shape of the anterior corneal surface may be suggested by visual, or refractive signs:

- a scissor or swirling reflex on retinoscopy
- visual acuity not correctable to better than 6/6 (20/20) by spectacles
- irregular keratometry readings
- incongruity between keratometric and refractive astigmatism
- unusual changes in refractive astigmatism over time.

Computer-assisted videokeratoscopy usually reveals mild inferior corneal steepening, although indicative indices fail to meet the criteria for keratoconus.

Suggested quantitative criteria for keratoconus are as follows:

- a mean central corneal power ≥47.00 D and/or
- a difference in central corneal power between the two eyes of ≥1.00 D
- a difference in corneal power of ≥1.50 D at locations 3 mm above and below the visual axis.

The area of focal steepening seen in forme fruste keratoconus fails to meet these criteria.

INCIDENCE

Forme fruste keratoconus probably affects as many as 1/1,000 people

SIGNIFICANCE

Forme fruste keratoconus may develop into full keratoconus and also presents a risk for corneal or refractive surgery. Especially if the patient is young when corneal ectasis is likely to progress, 6-monthly corneal mapping should be performed. If progression is observed, corneal collagen cross-linking should be considered.

DIFFERENTIAL DIAGNOSIS

Keratoconus.

SEE ALSO

Pellucid marginal corneal degeneration; Keratoglobus; Corneal collagen cross-linking.

MANAGEMENT

In general terms, no management is required. Forme fruste keratoconus is an asymptomatic condition, although with potential for development into clinical keratoconus. The clinical course is variable, although the older the patient is at the time of diagnosis, the more likely it is for any change to be slow and that the condition will remain sub-clinical.

Advice

Patients with forme fruste keratoconus should be strongly warned against LASIK, although in some cases they may be suitable for PRK. Patients should also be advised that eye rubbing may lead to progression of the disease.

Contact lenses

Patients considering contact lenses may need to consider rigid contact lenses (as opposed to soft lenses), since rigid lens correction may become essential in future and will often provide better vision.

Keratoglobus

Globular cornea

DESCRIPTION

There are three ectatic corneal dystrophies: keratoconus, keratoglobus, and pellucid marginal corneal degeneration. Of these, keratoglobus is the least common. Unlike the other ectatic dystrophies, keratoglobus is frequently present at birth with subsequent slow progression, although occasionally it may develop in late childhood. Unlike keratoconus, the thinning extends throughout the entire cornea all the way to the limbus. This leads to gross protrusion of the central and inferior cornea.

SYMPTOMS

Vision is characteristically quite poor from the induced myopic astigmatism. As with all irregular astigmatism, glare is a significant problem.

SIGNS

The corneal diameter may be increased, and the cornea is thinned throughout. Unlike keratoconus, maximal thinning is usually in the mid-periphery. There may be scattered deep stromal opacities and, unlike keratoconus, there are no iron deposition lines (Fleischer ring) present. The anterior chamber is characteristically very deep, occasionally being up to 5 mm deep centrally. As with the other ectatic dystrophies, hydrops can occur.

INCIDENCE

Extremely rare.

SIGNIFICANCE

Visual prognosis is very guarded due to the difficulty in treating such steep corneas.

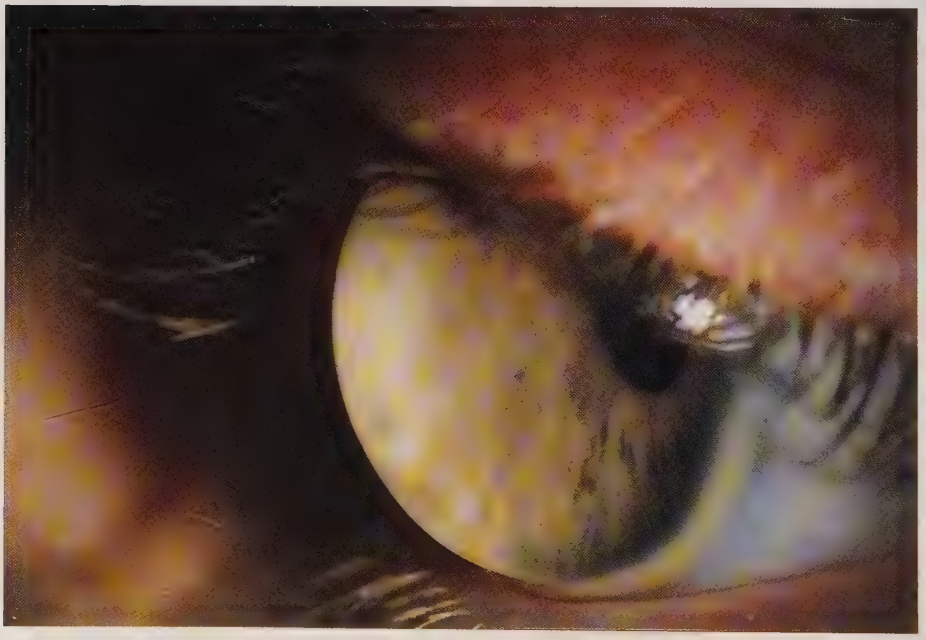
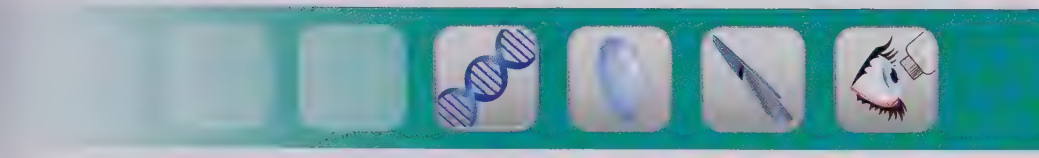


Fig. K-3 Advanced keratoglobus with corneal steepening extending to the limbus.



DIFFERENTIAL DIAGNOSIS

Keratoconus; Pellucid marginal corneal degeneration; megalocornea (not included in this text).

SEE ALSO

Pellucid marginal corneal degeneration; Keratoconus; Corneal collagen cross-linking; Corneal grafts; Descemet's membrane breaks.

MANAGEMENT

Genetic advice

Although it may occur sporadically, keratoglobus frequently appears to have an inheritance pattern consistent with an autosomal dominant trait with a high degree of penetrance.

Contact lenses

Large-diameter rigid lenses with a reverse geometry or aspheric design are usually required. Unfortunately, due to the whole of the cornea being affected, the fitting of even scleral lenses is more difficult.

Corneal collagen cross-linking

Chemical cross-linking of the corneal collagen with riboflavin-5-phosphate activated by UV light may be useful in preventing progression of the disease if the cornea is not too thin or scarred.

Surgery

Standard penetrating keratoplasty has very poor results, probably largely due to the mismatch in corneal thickness between the host and the donor. Onlay lamellar corneoscleral grafts have been attempted, often with a secondary central penetrating keratoplasty. Clear lens extraction and implantation of an intraocular lens of very low or even negative power may be useful in reducing the high degree of induced myopia from the increased central corneal curvature. However, this does not address the induced corneal irregular astigmatism.

Lattice stromal dystrophy

DESCRIPTION
The corneal dystrophies are a group of progressive, usually bilateral, inherited (although non-congenital) corneal disorders with a typical age of onset. The stromal dystrophies result in characteristic stromal opacities. There are many different stromal dystrophies; the three main types are granular, macular, and lattice. Lattice stromal dystrophy (along with Avellino, granular, and Reis–Bücklers dystrophies) occurs secondary to a mutation in the kerato-epithelin gene *BIGH3* on the long arm of chromosome 5.

The typical linear deposits seen in lattice dystrophy are from deposition of amyloid. This is best seen histopathologically with Congo red when it displays birefringence under crossed polarized light. There are three subtypes of lattice dystrophy: type 1 has an onset in the first decade of life; type 2 is associated with a familial form of systemic amyloidosis and has an onset in middle age; type 3 usually has an onset in the fourth decade of life. Lattice lines are also present in granular type 3 (Avellino) dystrophy.

SYMPTOMS
Recurrent corneal erosion symptoms, ocular discomfort, and blurring of vision from the developing opacities all increase over time. Type 2 is associated with typical facial nerve palsies.

SIGNS
The major feature of lattice stromal dystrophy is the presence of fine, relatively translucent criss-cross lines in the anterior stroma. These usually commence as fine dots and coalesce into lines over time. The lines are most marked centrally, but do progress with time both toward the periphery and to involve the deeper stroma. The intervening stroma remains clear. The epithelium overlying anterior lines is often disturbed.

INCIDENCE
Very rare (less than 1/100,000).

SIGNIFICANCE
May lead to symptoms of recurrent corneal erosion syndrome as well as a marked reduction in visual acuity secondary to corneal opacification.

DIFFERENTIAL DIAGNOSIS
Reis–Bücklers dystrophy; Granular (type 3 – Avellino) dystrophy; corneal nerves (these differ from lattice lines in that they are more marked in the periphery and progressively branch as they run centrally; they are also slightly deeper than lattice lines, typically running at the junction of the anterior and middle third of the stroma).

SEE ALSO
Macular stromal dystrophy; Granular stromal dystrophy; Recurrent corneal erosion syndrome; Corneal graft; Corneal graft – lamellar keratoplasty; Corneal dystrophies, degenerations, and depositions.

MANAGEMENT
Genetic advice
All three types of lattice stromal dystrophy are usually inherited as an autosomal dominant trait with a high degree of penetrance. Evaluate the patient for systemic amyloid conditions.

Tear supplements
Tear supplements are commonly used to treat ocular surface discomfort associated with an unstable epithelium and recurrent corneal erosion syndrome.

Laser surgery
Phototherapeutic keratectomy with the excimer laser has been used to treat superficial amyloid deposits both to improve vision by removing opacities and to stabilize the epithelium. Unfortunately, adequate treatment of opacities usually requires a fairly deep ablation and results in a significant hypermetropic shift. It can also lead to significant scarring and thinning of the cornea.

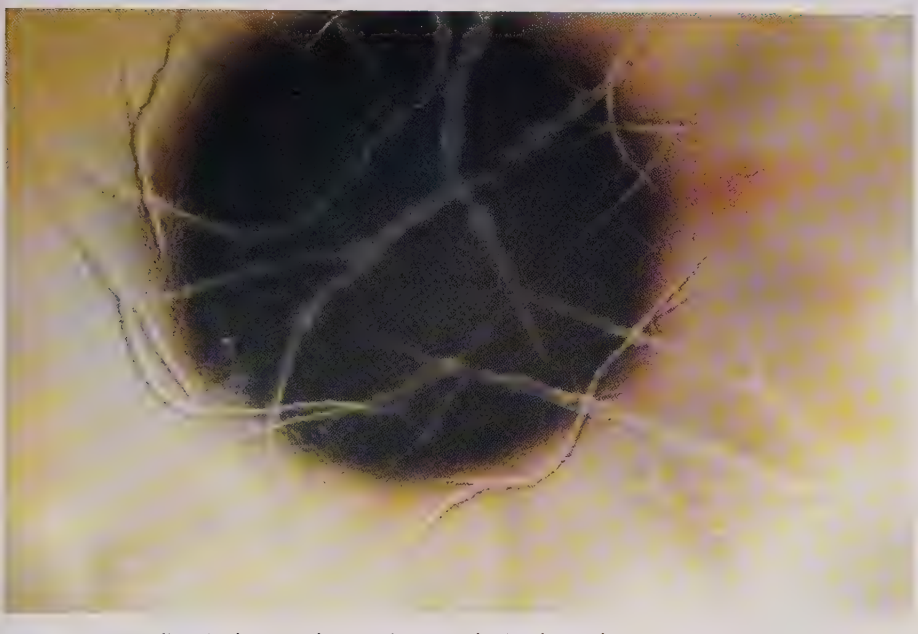


Fig. L-1 Lattice lines in the central cornea in type III lattice dystrophy.

Surgery

Occasionally, in advanced disease with significant visual loss, a lamellar or penetrating keratoplasty is performed. It is uncommon for surgery to be required before middle age. Superficial recurrences within the graft are common and may lead to renewed visual disturbances. Thankfully, this normally takes several years to occur and predominantly affects the peripheral cornea.

Leukocoria

Retinoblastoma, cataract

DESCRIPTION

Leukocoria refers to a white pupillary reflex. An opacity in the lens, vitreous cavity, or at the retina causes the abnormal appearance. While leukocoria may occur for a variety of reasons, perhaps the most notable is retinoblastoma since this condition, while extremely rare, is also potentially fatal.

Retinoblastoma is a malignant tumor of the retina that may be unilateral, bilateral, or multifocal. The tumor arises from primitive retinal cells before their final differentiation and, consequently, the onset is usually before 1 or 2 years of age. Retinal detachment, pseudohypopyon, iris neovascularization, or vitreous hemorrhage may be present. There may be a family history.

Congenital cataract is an opacity of the lens present at birth, which may be unilateral or bilateral, sometimes idiopathic, with a family history or with a systemic basis.

Other possible causes include: toxocariasis, Coats' disease, persistent hyperplastic primary vitreous, retinal astrocytoma, retinopathy of prematurity, coloboma, and retinal detachment.

SYMPTOMS

Since leukocoria predominantly affects infants, the condition is detected when a parent or carer sees the white pupil. However, in some cases the underlying condition causes a secondary strabismus, and the cause is found during dilated fundus examination.



Fig. L-2a Leukocoria in the right eye caused by extensive retinoblastoma.



Fig. L-2b More advanced presentation of retinoblastoma showing orbital inflammation.



SIGNS

Initially a retinoblastoma is a flat or elevated white lesion. With growth, several forms occur:

- Endophytic tumor is a white nodular mass extending into the vitreous, with prominent feeder vessels.
- Exophytic tumor is a mass lesion underlying a retinal detachment.
- Diffuse infiltrating tumor is a rarer form resembling a chronic uveitis.

Congenital cataract, if bilateral, can affect eye movements (nystagmus) or, if asymmetric or unilateral, causes a strabismus. There is a range of forms and densities of cataract that may develop.

Due to the range of conditions that may be present, a fundus examination with mydriasis is a necessary step in assessment of pediatric strabismus.

INCIDENCE

Leukocoria is a rare condition (approximately 1/10,000), primarily affecting young children before the age of 2 years. Retinoblastoma affects about one in 20,000 live births.

SIGNIFICANCE

Sight- and potentially life-threatening, requires urgent investigation and treatment.

DIFFERENTIAL DIAGNOSIS

Cataract; Posterior capsule opacification; Endophthalmitis; Hypopyon; Uveitis.

MANAGEMENT

Urgent hospital evaluation and treatment if retinoblastoma is suspected.

Special investigations

B-scan ultrasound, computed tomography (CT) scan, IV fluorescein angiogram may aid in diagnosis. CT can detect calcification associated with retinoblastoma.

Retinoblastoma treatment

There is a range of possible therapies, dependent upon the tumor size and location within the eye. Therapies include transpupillary thermotherapy with a diode laser, laser photocoagulation, cryotherapy, plaque radiotherapy, chemotherapy, and external radiotherapy. Enucleation may be required for large tumors.

Cataract surgery

Cataract extraction and intraocular lens (IOL) implantation in children with congenital cataract is indicated if the cataract is visually significant. With the foldable IOL and small incision surgery, an IOL can be implanted in children as young as 2 months of age. Lens implantation is often preferable to contact lens wear due to the significant issues of compliance in infants.

Genetics

Congenital cataract is often inherited as an autosomal dominant trait with a high degree of penetrance. Retinoblastoma has a genetic heredity in 40% of cases. In such cases there is a significant risk of siblings being affected. DNA analysis may assist in management.

Lice

Phthiriasis palpebrarum

DESCRIPTION

The eyelashes and eyebrows may become infested with the six-legged louse, *Phthirus pubis*. This is the same louse that causes pubic lice and is distinct from *Pediculus humanus*, the common head louse. Lice may also contaminate bedding, towels, and even clothing; contamination usually occurs when sanitary conditions are poor.

Phthiriasis infestation of the lashes commonly occurs in young adults as a sexually transmitted disease in conjunction with pubic lice. The lice lay eggs, termed nits, which adhere to the base of the lashes. When these hatch, the outer casing of the egg remains as a cast. Infestation often leads to conjunctivitis. This is said to occur secondary to the feces of the lice falling into the eye, a point of which patients are usually happy to remain ignorant.

SYMPTOMS

Infestation results in skin irritation and often a red, watery, slightly irritated eye. Infestation may be unilateral or bilateral.

SIGNS

The lice, nits, and casts have a typical appearance, with the lice being approximately 1 mm in length and the nits and casts being characteristically fusiform (spindle-shaped). There is often a toxic keratoconjunctivitis with papillae and a superficial punctate keratopathy. There may also be red inflamed areas on the eyelid margin from bites by the lice.

INCIDENCE

Rare.



Fig. L-3 Lice and nits at the base of upper lid lashes.



SIGNIFICANCE
Lice cause chronic ocular irritation and are moderately contagious.

DIFFERENTIAL DIAGNOSIS
Blepharitis – anterior.

SEE ALSO
Conjunctivitis – overview.

MANAGEMENT
Removal
Multiple treatments have been tried, including anticholinesterases to kill the lice, ointment to suffocate them, and even Nd:YAG laser. Simple removal of the lice, nits, and casts with forceps is just as effective and quite simple given the limited area of infestation. The eyebrow should also be inspected for signs of infestation.

Advice
Patients should be advised that this may be acquired as a sexually transmitted disease, and symptoms of pubic involvement sought. Treatment of pubic lice and any affected sexual partners is indicated to prevent re-infection. There are multiple over-the-counter lotions available for treatment of pubic hair. All clothes and linen need to be washed and dried.

Therapeutics
Patients may also be treated with the application of an ophthalmic ointment (e.g. lubricant) to the eyelids to suffocate the lice. Application of a topical insecticidal agent such as permethrin 1% may be used if there is also infestation of the pubic hair.

Limbal stem cell deficiency

DESCRIPTION

Following the seminal work of Thoft and others it was recognized that the corneal epithelium is derived from epithelial stem cells situated at the limbus. These stem cells are a self-renewing population that also give rise to transient amplifying cells (TACs). TACs are rapidly mitotic and give rise to corneal epithelial cells that migrate both centrally and superficially, producing the normal corneal epithelium.

An abnormality in this process leads to a breakdown of normal corneal epithelial function, producing an unstable epithelium prone to epithelial defects and slow to heal. The cornea eventually becomes covered in a conjunctival phenotype epithelium with an uneven surface and superficial vascularization. Histopathologically, the central corneal epithelium becomes "conjunctivalized" with the presence of conjunctival goblet cells. In addition, the epithelium stains with conjunctival cytokeratin markers.

Although usually affecting the whole of the cornea, localized limbal stem cell deficiency/failure can lead to localized corneal epithelial changes. Some authors believe that pterygia are a form of localized stem cell deficiency.

The causes of limbal stem cell deficiency can be classified as follows:

- primary
  - —aniridia
  - —KIDS (keratitis/icthyosis/deafness syndrome)
- secondary
  - —trauma
  - —chemical (acids, alkali, antimetabolites, i.e. mitomycin C, and thimerosol)
  - —mechanical (especially following multiple surgeries for conjunctival intraepithelial neoplasia [CIN])
  - —thermal
  - —autoimmune (ocular cicatricial pemphigoid [OCP], and Stevens–Johnson syndrome)

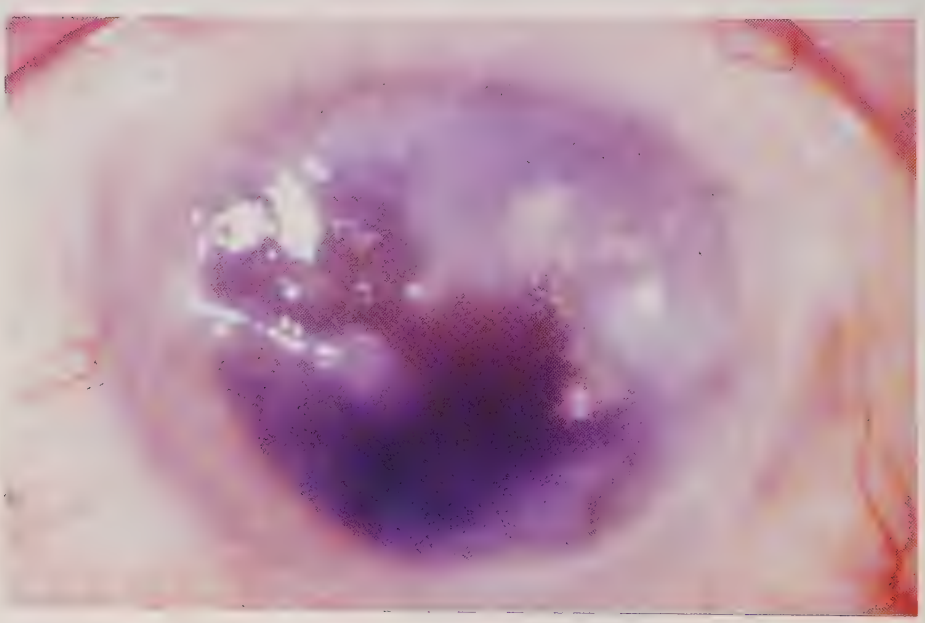
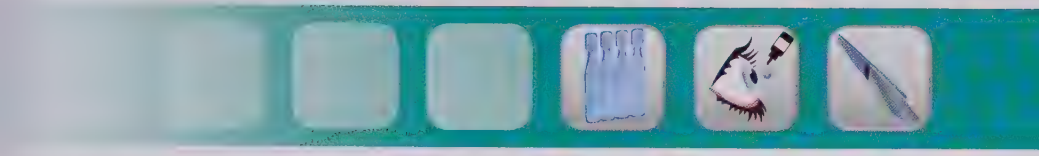


Fig. L-4 Limbal stem cell failure thought to be secondary to thimerosal toxicity.



—allergic (severe atopic keratoconjunctivitis)
—inflammatory (chronic rosacea keratitis idiopathic).

SYMPTOMS
Blurred vision, a red eye, photophobia, increased lacrimation, and a foreign-body sensation are all common with this condition.

SIGNS
The key sign of limbal stem cell deficiency is conjunctivalization of the corneal surface as described above. The limbal architecture is also disturbed with loss of normal vascular and epithelial features. The tear film is unstable over areas of conjunctivalized epithelium with deficiencies in the tear film and a decreased tear break-up time.

INCIDENCE
Rare.

SIGNIFICANCE
Leads to a significant reduction in sight.

DIFFERENTIAL DIAGNOSIS
Toxic epitheliopathy; Rosacea keratitis; Pannus.

SEE ALSO
Chemical burns; Aniridia; Ocular cicatricial pemphigoid; Stevens–Johnson syndrome; Atopic keratoconjunctivitis; Pannus and pseudopterygia.

MANAGEMENT
Tear supplements are useful to stabilize the tear film. Due to the sensitivity of the abnormal epithelium to any chemical insult, only unpreserved lubricants should be used.

Therapeutics
Chronic low-grade inflammation of the ocular surface is common; low-dose topical corticosteroids may be helpful. As with tear supplements, only unpreserved drops should be used such as minims of prednisolone.

Surgery
Limbal stem-cell grafting has been investigated extensively since the early 1990s. A peripheral corneoscleral inlay or onlay autograft from the other eye or allograft (either cadaveric or from a living relative) is performed. This may be performed in conjunction with an amniotic membrane graft. Unfortunately, the results have been quite variable, and many investigators feel that, postoperatively, patients need to be on long-term systemic immunosuppression. Results have been best when the rest of the ocular surface including the tear film is healthy. They have been particularly disappointing in aniridia, possibly because the underlying stromal mechanism for supporting limbal stem cells is deficient. Recent investigations have centered on the expansion of limbal stem cells in vitro, with transplantation of sheets of cultured cells on contact lenses or amniotic membranes.

Lipid keratopathy

DESCRIPTION

Lipid deposition within the corneal stroma usually occurs secondary to incompetent corneal vessels. The cornea is usually avascular due to the presence of soluble vascular endothelial growth factor (VEGF) receptors. Corneal vascularization most commonly results from an infective process such as a bacterial or viral keratitis, typically herpes simplex keratitis. These vessels leak serum lipid resulting in the typical appearance of lipid keratopathy.

Lipid deposition can also occur spontaneously in the peripheral cornea as an arcus senilis. This is especially common in the elderly and people with hyperlipidemia.

SYMPTOMS

Patients frequently notice a slowly enlarging whitish corneal opacity; the eye may be totally comfortable or slightly irritated. If the lipid is deposited centrally, blurring of vision usually occurs.

SIGNS

The intracorneal lipid deposition has a typical appearance, being yellowish to white in color with even, stellate crystalline edges with a distinct edge. There are associated feeder vessels arising from the limbus and extending into the cornea. The depth and extent of the lipid depends largely on the extent and depth of the vessels.

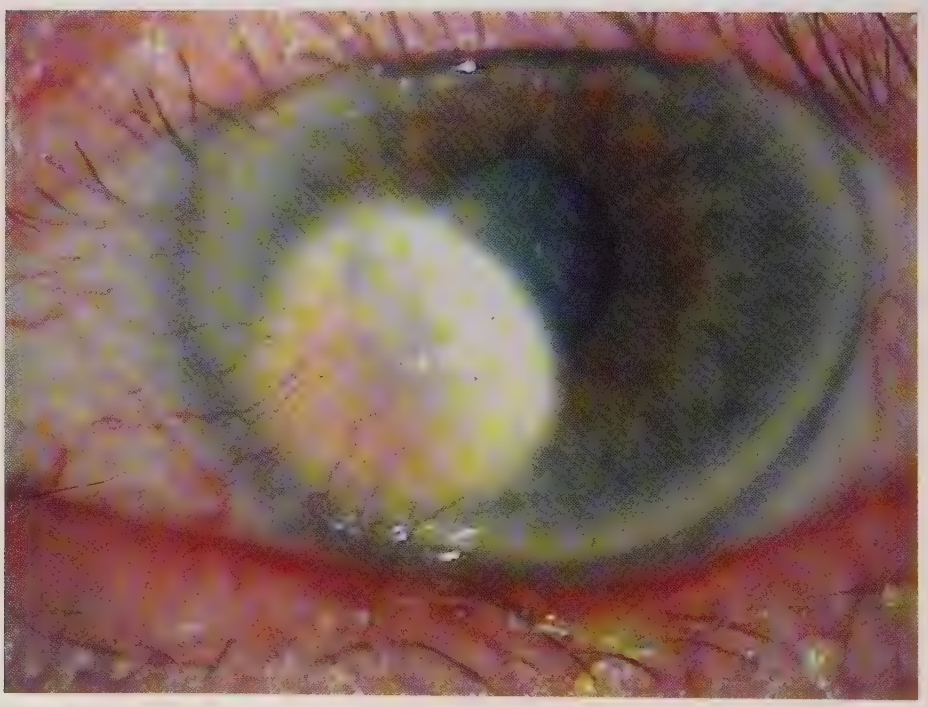


Fig. L-5 Extensive intracorneal vascularization and lipid deposition with formation of a leukoma threatening the visual axis. Corneal vascularization occurred as a result of a peripheral bacterial keratitis.



INCIDENCE
Rare.

SIGNIFICANCE
May become sight-threatening.

DIFFERENTIAL DIAGNOSIS
Fungal keratitis; Infectious crystalline keratopathy; Schnyder's crystalline dystrophy; Bacterial keratitis.

SEE ALSO
Crystalline keratopathy; Herpes simplex keratitis; Herpes zoster ophthalmicus; Corneal opacities – peripheral.

MANAGEMENT
Therapeutics
Any underlying cause of ongoing vascularization, such as active inflammation or viral infection, should be treated. Active inflammation may add to vascular incompetency and, therefore, topical steroids can be tried. It is often very difficult to assess any effect on vascular leakage as the progression or regression of lipid keratopathy is a very slow process.

There are several reports of topical anti-VEGF agents such as bevacizumab being used either topically or as a sub-conjunctival injection to induce regression of corneal vessels.

Laser surgery
Laser disruption and occlusion of the feeder vessels can be useful if the vessels are fairly small and not too numerous. This is performed with the diode laser and usually takes several treatments to occlude all vessels. Commonly used settings are a 200-micron spot size, laser duration of 0.2 seconds, and power of 500 mW. The laser is directed at occluding the arterioles, not the venules, and a string of faint white laser spots are applied along a length of each arteriole. With successful occlusions of the arterioles, the blood flow through the venules gradually diminishes. Care should be taken, as sometimes the vascularized corneal tissue is metabolically dependent on the vessels for oxygen and nutrition. Occlusion of the vessels can lead to sterile ulceration, presumably due to ischemic necrosis.

Surgery
Lamellar excision of the feeder vessels and application of an inlay lamellar graft can be tried if the visual axis is threatened and laser occlusion is unsuccessful.

Macular stromal dystrophy

Groenouw II dystrophy

DESCRIPTION

The corneal dystrophies are a group of progressive, usually bilateral, inherited (although non-congenital) corneal disorders with a typical age of onset. The stromal dystrophies result in characteristic stromal opacities. There are many different stromal dystrophies; the three main types are granular, macular, and lattice.

The characteristic lesions of macular stromal dystrophy result from accumulations of mucopolysaccharide within the endoplasmic reticulum, secondary to an error in normal keratan sulfate metabolism. In type 1 macular dystrophy, an error in a keratan-specific sulfotransferase results in undetectable levels of keratan sulfate in the serum. In type 2 macular dystrophy, serum keratan sulfate levels are low to normal. Histopathologically, the abnormal stromal mucopolysaccharide stains blue with alcian blue.

The result of this metabolic dysfunction is a loss of proteoglycans and an accumulation of dermatan and chondroitin sulfate. This loss of corneal ground substance between corneal collagen lamellae leads to corneal thinning and opacification.

SYMPTOMS

With time, the vision becomes blurred, such that by the fifth to sixth decade of life it has usually decreased to less than 6/60. Recurrent erosions are rare and usually only occur late in the disease.

SIGNS

The characteristic lesions of macular stromal dystrophy first appear in the first decade of life as grayish white dots affecting the central superficial stroma. Over years, these lesions spread to involve both the deep and the peripheral stroma. They develop a characteristic ring-shaped appearance and are approximately 0.5 mm in diameter. In contrast to the other two main stromal dystrophies, granular and lattice, the intervening stroma also becomes progressively opaque. In advanced disease, the whole cornea becomes somewhat opaque, with denser central ring-shaped opacities.

The cornea thins with time due to a decrease in interlamellar spacing, the central corneal often being decreased from

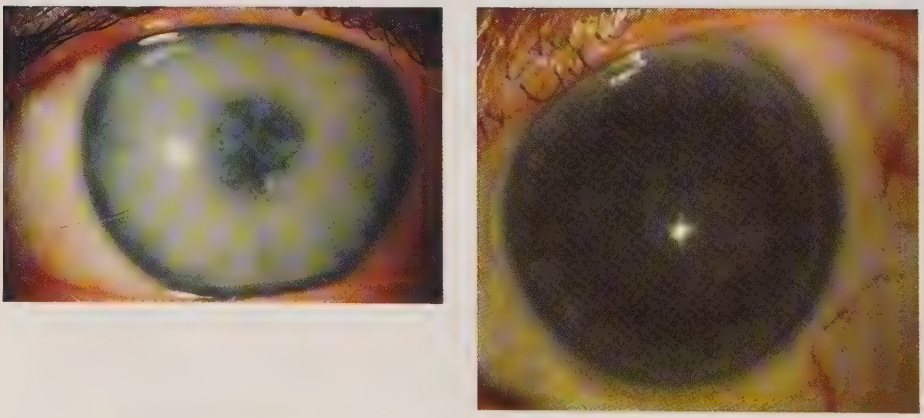
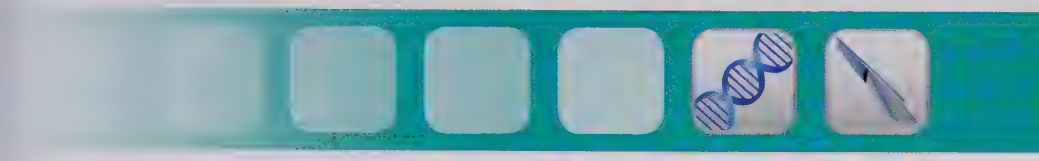


Fig. M-1 Corneal macular dystrophy with generalized haze involving the whole of the cornea with focal denser opacities.



a normal thickness of around 540 microns to only around 400 microns. Endothelial guttae may also develop, and corneal sensation decreases.

INCIDENCE

Extremely rare in most communities. Much more common in some parts of the world, particularly in the Middle East.

SIGNIFICANCE

Usually leads to a marked reduction of vision in both eyes by the fifth or sixth decade of life. Unlike other stromal dystrophies, macular dystrophy does not lead to recurrent corneal erosion syndrome.

DIFFERENTIAL DIAGNOSIS

Interstitial keratitis; bilateral Herpes simplex keratitis; Band keratopathy; Trachoma.

SEE ALSO

Lattice stromal dystrophy; Granular stromal dystrophy; Corneal graft; Corneal graft – lamellar keratoplasty; Corneal dystrophies, degenerations, and depositions.

MANAGEMENT

Genetic advice

Inheritance is autosomal recessive.

Surgery

By the fifth or sixth decade of life, a penetrating keratoplasty or deep anterior lamellar keratoplasty (DALK) is usually required to restore vision. As with other sight-threatening bilateral progressive corneal disease, grafting is usually performed in the first eye before the vision deteriorates markedly to 6/12 (20/40). This is done to allow adequate time for visual rehabilitation in the grafted eye before the vision in the second eye deteriorates significantly. The survival rate for corneal grafts for macular stromal dystrophy is quite high, and recurrences of disease in the graft relatively uncommon.

Marginal keratitis

Staphylococcal hypersensitivity

DESCRIPTION

This is a common peripheral corneal condition thought to occur as an inflammatory reaction to staphylococcal antigens. It is commonly associated with both staphylococcal blepharitis and facial rosacea. It most commonly affects middle-aged people, but may rarely affect children. While the lesion is non-infectious and self-limiting, its main importance lies in a substantial overlap with the symptoms and signs of early microbial keratitis.

SYMPTOMS

During an acute attack, the patient usually complains of photophobia, slight ocular irritation, increased lacrimation, red eye, and slight blurring of vision. Recurrences are common. During any given attack involvement is usually unilateral.

SIGNS

The lesion is usually unilateral, although there may be signs of resolved marginal keratitis in both the affected and the other eye with typical "shovel-shaped" superficial scars. Typically, marginal keratitis affects the inferior paralimbal cornea with a 1–2-mm clear area of uninvolved cornea between the lesion and the limbus (interval of Vogt). Lesions may be multiple and gray to white in color. They are typically rounded, approximately 1 mm in diameter, and involve the superficial stroma.

Occasionally, a lesion can extend around the limbus for a few clock hours as a crescent of infiltrate. There is usually a small overlying epithelial defect. The adjacent conjunctiva is usually slightly injected and inflamed. The anterior chamber is usually quiet or with only a trace of cell and flare. The "PEDAL" mnemonic helps identify lesions more likely to be infectious in nature:

- **P**ain
- **E**pithelial defect
- **D**ischarge
- **A**nterior chamber reaction
- **L**ocated more central.

If untreated, the lesion usually resolves spontaneously over a week or two. Occasionally resolution is accompanied by the ingrowth of small superficial blood vessels, leading to the formation of a localized area of pannus. Signs of facial rosacea with a typical malar flush and, occasionally, pustule formation may be present, as may signs of blepharitis.

INCIDENCE

Common.

SIGNIFICANCE

Usually resolves spontaneously without loss of vision.

DIFFERENTIAL DIAGNOSIS

Microbial (especially Bacterial) keratitis; Phylectenulosis; Corneal (sterile) infiltrate (especially related to contact lens use).

SEE ALSO

Rosacea keratitis; Blepharitis – anterior; Blepharitis – posterior; Pannus and pseudopterygia; Corneal opacities – peripheral; Salzmann's.

MANAGEMENT

If there is associated blepharitis or rosacea, refer to the individual articles in this text for details of treatment. If unsure whether an individual lesion is infective or sterile, it is prudent to err on the side of being overly cautious with the use of a topical broad spectrum antibiotic (see "Bacterial keratitis").

Therapeutics

A combination of an anti-staphylococcal antibiotic, such as chloramphenicol or ciprofloxacin, and a topical corticosteroid usually leads to rapid resolution of the acute disease. Associated posterior blepharitis or rosacea may require treatment with a course of oral antibiotics.

Dosing: G. chloramphenicol or ciprofloxacin qid for 1 week, and G. fluorometholone acetate 0.1% qid for 1 week.



Lid hygiene

If there is associated meibomian gland dysfunction or scaling of the lashes, ongoing lid hygiene may be useful in preventing subsequent attacks.

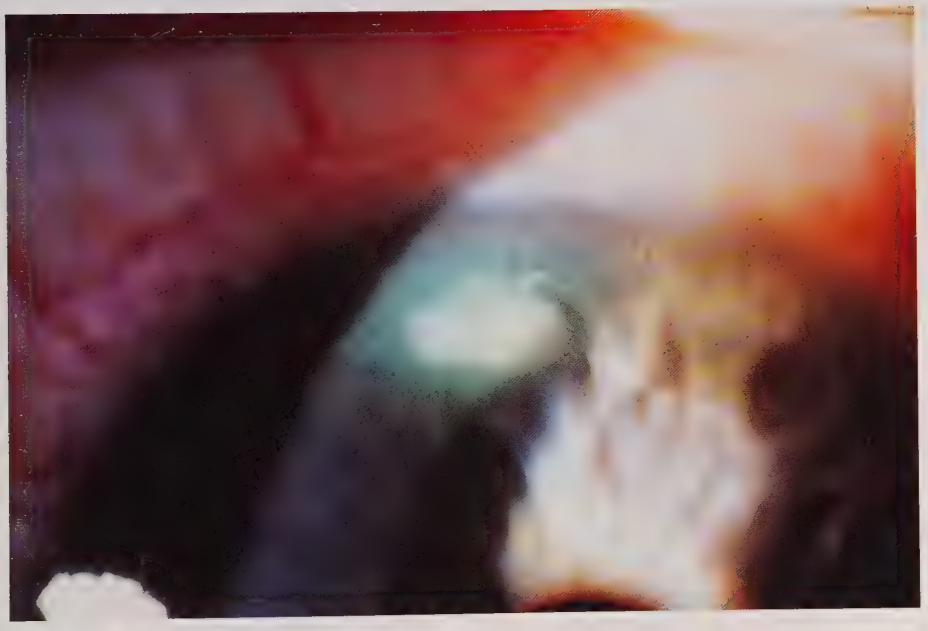


Fig. M-2 Typical paralimbal lesion of marginal keratitis.

Meesmann's dystrophy

Juvenile hereditary epithelial dystrophy

DESCRIPTION

Meesmann's dystrophy is easily the rarest of the three epithelial dystrophies, the other two being epithelial basement membrane dystrophy and Reis–Bückler's dystrophy. This autosomal dominant dystrophy was first described comprehensively by Meesmann in 1938. Onset is usually in the first decade of life with the appearance of intraepithelial cyst-like lesions. These are actually pseudocysts, probably representing areas where cells have died, leaving a space within the epithelium. These pseudocysts have been examined with electron microscopy and contain a characteristic degenerate material dubbed "peculiar substance," probably resulting from cell death and degeneration.

SYMPTOMS

Usual onset of symptoms is not until the fourth or fifth decade of life, with transient blurring of vision, surface irritation, increased lacrimation, and slight photophobia.

SIGNS

As with all dystrophies, the signs are usually bilateral and often relatively symmetric. The characteristic changes are of tiny rounded intraepithelial microcysts. These can occur in a swirling or wedge-like pattern. Initially, these lesions are restricted to the interpalpebral epithelium but with time can involve the entire corneal epithelium, although sparing of the peripheral corneal epithelium is common. Over time, anterior stromal and epithelial basement membrane grayish lesions can occur.

INCIDENCE

Extremely rare (less than 1/100,000).

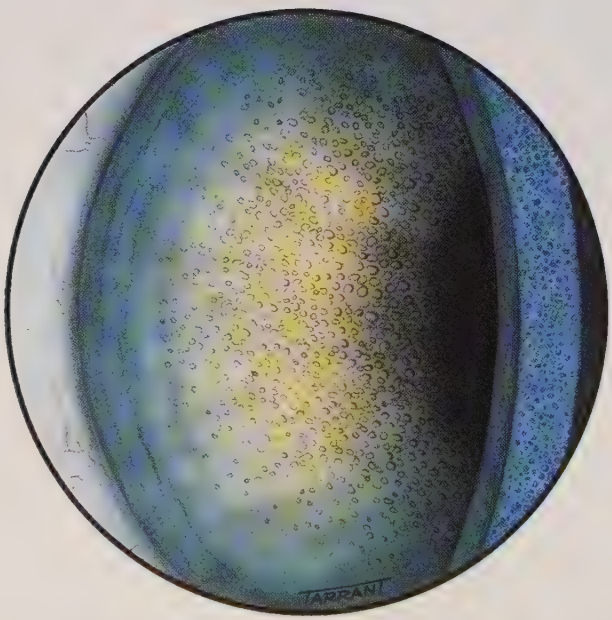
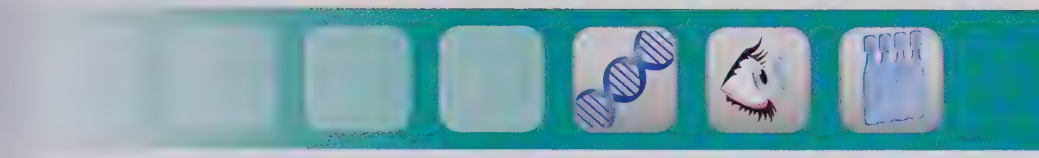


Fig. M-3 Drawing of a slit-lamp view of Meesmann's dystrophy.



SIGNIFICANCE
Usually does not affect vision. However, may lead to slight transient blur, frequently resulting in slight ocular irritation.

DIFFERENTIAL DIAGNOSIS
Recently healed epithelium with microcysts; Epithelial basement membrane dystrophy.

SEE ALSO
Corneal dystrophies, degenerations, and depositions; Reis–Bückler's dystrophy.

MANAGEMENT
Genetic advice
The inheritance pattern is autosomal dominant with a relatively low degree of penetrance.

Tear supplements
Tear supplements are the mainstay of treatment for ocular surface symptoms such as irritation and photophobia.

Therapeutic contact lens
Occasionally, application of a therapeutic contact lens is required to relieve ocular surface symptoms.

Melanoma – conjunctival

Malignant melanoma (MM)

DESCRIPTION

Malignant melanoma (MM) of the conjunctiva mainly affects the elderly. It arises most commonly in a pre-existing nevus or in an area of primary acquired melanosis, and least commonly arises de novo as a primary lesion. It is a potentially sight- and life-threatening disease, with reported death rates varying between different series, but routinely being around 30%. Fair-skinned, blue-eyed individuals appear to be at an increased risk of developing the disease, although a link with increased UV light exposure has not been demonstrated.

SYMPTOMS

Patients usually present having noticed a slow-growing lesion, frequently in a pre-existing area of pigmentation. The eye is usually comfortable.

SIGNS

MM most commonly occurs on the bulbar conjunctiva or at the limbus, although it can rarely arise on the palpebral



Fig. M-4a Primary conjunctival melanoma. Note feeder vessels and the variation in the extent of pigmentation throughout the lesion.

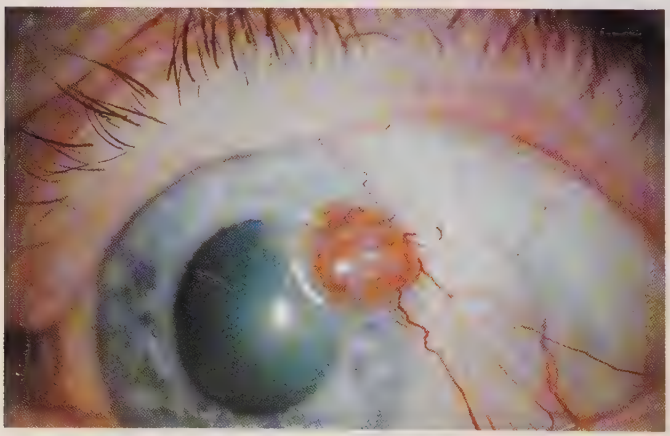
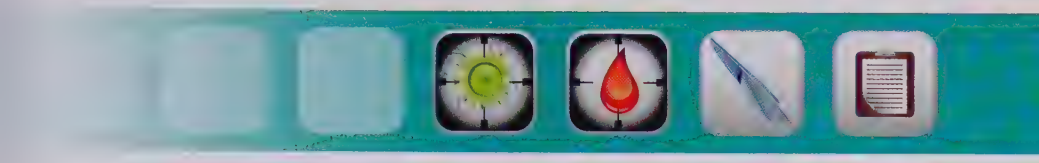


Fig. M-4b Amelanotic melanoma.



conjunctiva. The degree of pigmentation is quite variable, with a reasonable percentage being largely non-pigmented, termed "amelanotic melanoma." The melanoma usually grows in a nodular manner and may even be papillomatous. It is typically quite heavily vascularized with prominent feeder vessels. An associated nevus or area of primary acquired melanosis may be present. The area should be examined under topical anesthesia to define the degree of tethering to the underlying sclera.

A dilated fundal examination should be performed to exclude a ciliary body melanoma that has eroded through the sclera. Local and distant lymph nodes should also be examined for possible lymphatic spread.

INCIDENCE

Very rare (less than 1/100,000).

SIGNIFICANCE

Both sight- and life-threatening.

DIFFERENTIAL DIAGNOSIS

Conjunctival nevus; primary acquired melanosis; melanocytoma (all three listed under "Conjunctival pigmented lesions"); ciliary body melanoma with extension through the sclera. If amelanotic, may be confused with conjunctival intraepithelial neoplasia or a pyogenic granuloma.

SEE ALSO

Melanoma – eyelid; Melanoma and nevus of the iris.

MANAGEMENT

Blood tests/pathology

Referral to an oncologist for investigation of distant spread of the tumor is indicated. Frequently used tests include a full blood examination (FBE) and liver enzymes, chest X-ray (CXR), and liver scan.

Therapeutics

Topical mitomycin C 0.04% may be used to treat disease not amenable to surgical treatment. Care should be taken with respect to the length of treatment; more than 4 weeks of total accumulated treatment time appears to place patients at a heightened risk of developing limbal stem cell deficiency.

Surgery

An initial incisional biopsy is usually performed to confirm the diagnosis. If small, and in the absence of intraocular or orbital spread, treatment is with local excision with resection of the underlying sclera and application of cryotherapy. Cryotherapy seems to be of particular use in reducing the risk of recurrences in MM. Occasionally, conjunctival melanoma, as is the case with choroidal melanoma, may be amenable to local radioactive plaque treatment. If there is extensive local spread, an enucleation or even exenteration may be indicated.

Review

Six-monthly review with annual or biannual systemic investigation is indicated for the rest of the patient's life.

Melanoma – eyelid

Maligant melanoma (MM), amelanotic melanoma

DESCRIPTION

Melanoma is a potentially fatal skin cancer that arises from epidermal melanocytes. It is an invasive neoplastic proliferation of malignant melanocytes in the basal epidermis. In some cases, the melanoma originates from a malignant degeneration of congenital or dysplastic nevi. Melanoma usually occurs after the age of 35.

Several forms of malignant melanoma (MM) are recognized:

- Superficial spreading melanoma. This is the most common form. The plaque has an irregular outline and variable pigmentation. About one-half of the melanomas on the eyelids are non-pigmented.
- Nodular form. An elevated and usually markedly pigmented tumor displaying significant growth and that may be ulcerated at the center.
- Lentigo maligna-related melanoma. These pigmented patches with irregular borders evolve slowly. The precursor lentigo maligna may have been static for many years.

SYMPTOMS

Initially, the patient may have a cosmetic concern about a pigmented lesion on their skin. They may have noticed a change in its mass or size. Symptoms such as pain, tenderness, inflammation, and bleeding may be present, but these usually occur in later stage. Melanoma is usually unilateral.

SIGNS

The challenge in diagnosis is to distinguish between the atypical nevus and a MM. However, there are subtle aspects of pigmented lesions that may allow differentiation. The "**ABCDE**" method is a useful mnemonic for assessing pigmented lesions for the possibility of MM (McCormick and DeLuca 1996). Characteristics are:

A = asymmetrical shape and elevation
B = border is irregular or scalloped
C = color is mottled and variable
D = diameter is usually large (greater than 5 mm)
E = evolutionary change (change in the size).

Rapid growth and altered pigmentation may be important clinical features.

If the melanoma is non-pigmented (amelanotic), the diagnosis can be more difficult. The lesion may have significant vascularity and uneven elevation. Occasionally, distant micro-satellite lesions may form.

INCIDENCE

Melanoma is rare on the eyelids (perhaps 1/1000–1/10,000) and more common on the back of the body and on the legs. Unlike the non-melanoma skin cancers, the risk is less directly related to chronic sun exposure. Epidemiological risk factors include a history of dysplastic nevi, or numerous nevi, a racial predisposition in Caucasians, a relevant family history, the older age group (50 to 70 years), and a history of sunburn (especially when young). In Australia melanoma is the most common form of cancer in people aged 15–44 years. Each year it kills about 1000 Australians, and about 7000 cases are diagnosed (prevalence of about 1 in 3000).

SIGNIFICANCE

The grading system for MM consists of stage 1 – localized primary disease; stage 2 – metastasis to regional nodes; and stage 3 – metastasis to distant nodes or viscera. The prognosis is grim both in stage 2 with multiple node involvement and in stage 3. Tumor thickness is also of prognostic significance, with a lesion of 0.75-mm invasion depth or less being unlikely to have metastasized.

DIFFERENTIAL DIAGNOSIS

Nevus; Basal cell carcinoma (BCC) with secondary melanosis.

SEE ALSO

Skin tumors – overview.



Fig. M-5a Superficial spreading melanoma.

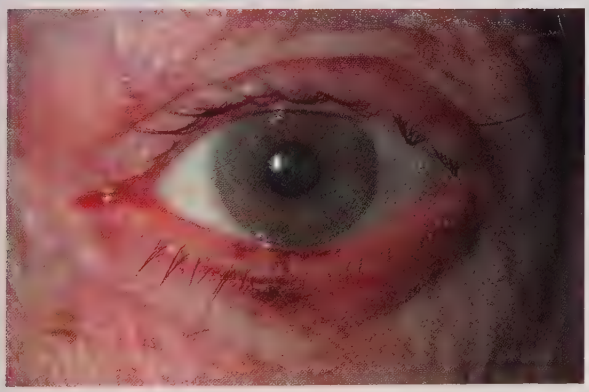


Fig. M-5b Lentigo maligna, also known as melanoma in situ, intraepidermal melanoma, or Hutchinson freckle.

MANAGEMENT

Surgery

Early diagnosis and complete removal are needed for MM. Excisional biopsy with 1-cm skin margins is recommended for melanoma of the eyelid. This requires eyelid reconstruction. In addition, regional lymph node dissection is suggested for melanomas with an invasion depth of 1.5 mm or more. In contrast, acquired nevi should be monitored clinically, but rarely is removal necessary.

Review

For patients with multiple skin lesions, each individual lesion has to be inspected carefully for sign of malignant changes. Melanoma less than 1 mm thick should be reviewed after surgery for 2 years at 3- or 6-month intervals, then annually thereafter. Thicker tumors require at least twice-annual review for the first 5 years.

REFERENCE

McCormick SA, DeLuca RL. Tumors of melanocytic origin. In: Mannis MJ, Macsai MS, Huntley AC (eds). Eye and Skin Disease. Philadelphia: Lippincott-Raven, 1996, pp 381–393.

Melanoma and nevus of the iris

Melanocytic nevus, melanocytoma

DESCRIPTION

Iris nevi and melanomas are largely benign conditions, with neoplastic transformation of a nevus being extremely rare, and iris melanoma, in stark contrast to conjunctival, ciliary body, or choroidal melanoma, also being a largely benign disease. Clinical differentiation between these two conditions may be impossible. Distant metastasis from an iris melanoma is almost unknown. An uncommon subtype of iris nevi is the melanocytoma, also referred to as a magnocellular nevus due to its histological appearance of plump, maximally pigmented, nevus cells. Unlike with skin melanoma, iris lesions are not associated with UV light exposure.

SYMPTOMS

Neither nevi nor melanomas usually cause any symptoms, other than concern at the presence of a colored lesion on the iris and pupil irregularity.

SIGNS

Both iris nevi and melanomas tend to be pigmented, although they may occasionally be amelanotic. Iris nevi may be flat or slightly elevated and are often multiple and bilateral. They do not usually lead to distortion of the pupil margin. Melanomas, by contrast, tend to exhibit several of the following features:

- pupil distortion with ectropion uveae
- growth in both thickness and extent of iris involvement
- prominent vascularity
- secondary glaucoma
- invasion of the angle.

Occasionally, melanomas grow in a more diffuse manner, covering the iris and even leading to heterochromia. As indicated above, clinical differentiation between the two conditions is not always possible. Thankfully, due to the usually benign nature of iris melanomas, this is not a problem.

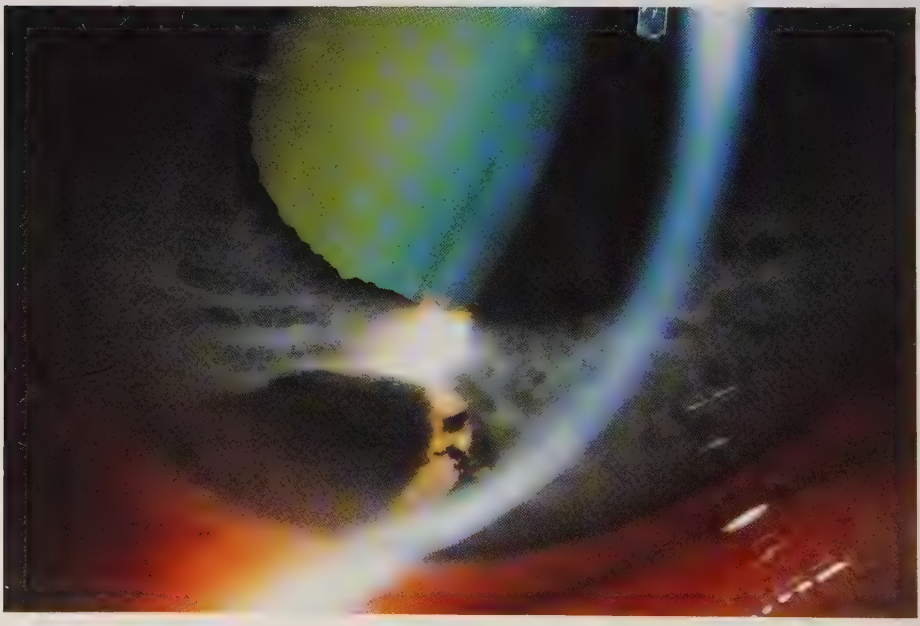


Fig. M-6a Heavily pigmented iris melanoma.



Fig. M-6b Gonioscopic view demonstrating invasion of the angle.

INCIDENCE

Nevi are very common, while melanomas are extremely rare, especially in darkly pigmented people.

SIGNIFICANCE

Both rarely lead to problems although an iris melanoma can (rarely) grow significantly enough to affect iris, angle, or even corneal function leading to glaucoma or corneal edema. Metastasis of an iris melanoma to a distant site is almost unknown.

DIFFERENTIAL DIAGNOSIS

Other iris tumors such as juvenile xanthogranuloma, iris adenoma, iris cysts, leiomyoma, and secondary tumors that have metastasized to the iris; ciliary body melanoma that has eroded through the iris; Lisch nodules in neurofibromatosis; Fuchs' heterochromic iridocyclitis. Nevi may also be seen in the iridocorneal epithelial syndrome subgroup iris nevus syndrome.

SEE ALSO

Melanoma – conjunctival; Melanoma – eyelid; Iridocorneal endothelial syndrome (ICE).

MANAGEMENT

Review

Nevi do not require review. However, melanomas are usually initially reviewed 6-monthly and then annually for life. Slit-lamp photography of the lesion is a very useful means of comparing tumor size and extent over time.

Surgery

Very rarely, an iris melanoma can become so diffuse or increase in size so rapidly that surgical treatment is required. A broad iridectomy, iridotrabeculectomy, iridocyclectomy, or, extremely rarely, enucleation may be indicated.

Membranous and pseudomembranous conjunctivitis

DESCRIPTION

Conjunctival membranes consist of an exudate of loose fibrin-cellular debris. With pseudomembranes, this membrane does not adhere to the underlying epithelium whereas, with true membranes, the conjunctival epithelium is intimately involved within the membrane. Any marked exudative conjunctivitis can lead to membrane formation.

Two bacterial causes of conjunctivitis that typically cause both true and pseudomembrane formation are *Corynebacterium diphtheriae* and *Neisseria gonorrheae*. Other causes of membranous conjunctivitis include severe epidemic keratoconjunctivitis (EKC), Stevens–Johnson syndrome, chemical burns, ligneous conjunctivitis, and vernal conjunctivitis.

SYMPTOMS

The patient usually presents with a severe conjunctivitis, with exudate, tearing, swollen lids, and blurring of vision. The lid swelling may be so marked that the patient is unable to open their eyes. Ocular discomfort is usually quite marked, and a foreign-body sensation from the membranes may occur.

SIGNS

Aside from the signs of the underlying cause of the conjunctivitis, the membranes themselves have a typical appearance. The membrane is a yellow-brown color and is usually seen in the fornices, being most commonly in the inferior fornix. It has an elastic property but does pull apart if stretched too far. Pseudomembranes can be peeled away without the removal of conjunctival epithelium, whereas true membranes result in denuding of the conjunctiva with subsequent conjunctival bleeding. It is not uncommon for a membrane to be predominantly a pseudomembrane, with a small area of true membrane formation. There is



Fig. M-7 A conjunctival pseudomembrane involving the inferior tarsal conjunctiva.



often an associated superficial punctate keratopathy.

INCIDENCE
Rare.

SIGNIFICANCE
May lead to conjunctival scarring.

DIFFERENTIAL DIAGNOSIS
Ligneous conjunctivitis (not included in this text).

SEE ALSO
Gonococcal keratoconjunctivitis; Adenoviral conjunctivitis; Conjunctivitis – overview; Conjunctival scarring – overview, Stevens–Johnson syndrome.

MANAGEMENT
Lid hygiene
Frequent eye hygiene and irrigation with sterile saline to remove any accumulated exudate can reduce ocular discomfort, inflammation, and membrane formation. Membranes can be very irritating to the eye, and removal with forceps and a cotton bud under topical anesthesia can significantly aid ocular comfort.

Glass rodding
The traditional treatment for true membranes of sweeping a glass rod through the inferior and superior fornix to prevent symblepharon formation is now uncommon. It was always of questionable benefit, and aggressive treatment of the underlying cause of the disease is more important.

Therapeutics
As well as supportive treatment with lid hygiene and eye washing, aggressive treatment of the underlying cause with topical antibiotic or corticosteroids as appropriate is important.

Molluscum contagiosum

DESCRIPTION

Molluscum contagiosum is a localized skin infection caused by a pox virus. It is spread by direct skin-to-skin contact and is common in children. Children attending a childcare center are at particular risk of contracting the disease. Adults can also contract the disease, and it can occasionally be transmitted sexually.

The lesions are self-limiting and non-scarring and, as such, are usually only of cosmetic concern. The exception is when they occur around the eye, where they lead to a follicular conjunctivitis.

SYMPTOMS

The skin lesion is normally asymptomatic and grows over 1–2 weeks. As it is usually totally uninflamed, it is frequently not noticed by the patient. The eye is mildly photophobic, watery, and the vision may be slightly blurred. One or both eyes may be affected depending on the extent of the skin lesions. There is no lymphadenopathy.

SIGNS

Molluscum may be solitary or multiple. To cause ocular problems they need to involve the lid margin or skin around the eye or on the brow. It is important to look through the eyebrows, as the lesions may not be readily visible. The molluscum has a typical appearance being about 2–3 mm in diameter, quite raised, and with a lumpy edge. There is a central pit or umbilicus. When expressed the contents are avascular, white, and express separately from the overlying skin.

The eye is red and often slightly watery, and there is usually a marked follicular conjunctivitis. There may also be corneal changes with a superficial punctate keratopathy.



Fig. M-8 Solitary molluscum on the upper eyelid margin with typical central pit.



INCIDENCE
Uncommon.

SIGNIFICANCE
Does not cause any long-term changes and is a self-limiting disease, although it may take months to years to resolve.

DIFFERENTIAL DIAGNOSIS
Other causes of follicular conjunctivitis; skin lesions such as lid cysts, warts, and tumors.

SEE ALSO
Follicular conjunctivitis; Skin tumors.

MANAGEMENT
Advice
The infective nature of the lesion should be explained to the patient and, particularly with children, the need for adequate hygiene to prevent both reinfection and the infection of others should be emphasized.

Histopathology
If the diagnosis is in doubt, routine histopathological examination of the lesion following excision reveals the typical cytological changes of pox virus infection with large cytoplasmic inclusion bodies.

Surgery
Although this is a viral infection, it is not treated with antiviral agents. Molluscum contagiosum is normally quite easy to treat with minor surgery; any insult to the lesion usually results in its rapid regression. Excision, cautery, and expression have all been used successfully. Once the lesion has been removed, the follicular conjunctivitis usually resolves within days to a few weeks.

Mooren's ulcer

DESCRIPTION
Mooren's ulcer is a very rare form of peripheral ulcerative keratitis (PUK) thought to occur from a limbal vasculitis and possible autoimmune reaction directed against corneal collagen. Histopathological examination of the conjunctiva adjacent to the area of ulceration demonstrates an extensive plasma cell infiltrate, suggestive of the autoimmune nature of the disease. There are two forms of the disease. One, mainly affecting older white males, is unilateral and less inflamed; the other is more aggressive, usually bilateral, and affects young males especially of African descent.

SYMPTOMS
Significant ocular surface inflammation with increased lacrimation, pain, and photophobia are typical. This is more marked in the bilateral form of the disease.

SIGNS
The key finding is an area of peripheral corneal ulceration with associated infiltrate and tissue loss typically extending to the limbus. This may occur at any position around the cornea. The area of ulceration is usually crescentic, extending for several clock hours. The junction of the normal and thinned cornea has a characteristic appearance with an "overhanging edge." Unlike with some other forms of peripheral ulcerative keratitis, the sclera is uninvolved.

With time, the area of thinning extends around the cornea and then spreads centrally. Eventually, this can lead to a uniformly thinned cornea with superficial vascularization, termed a "contact lens cornea." Spontaneous perforation is rare but may occur secondary to minor trauma. Cataract formation is common. The whole ocular surface is usually inflamed, particularly the adjacent conjunctiva.

INCIDENCE
This is an extremely rare disease in most communities, although there is a particularly high incidence of the disease in parts of West Africa.

SIGNIFICANCE
Untreated, this condition can lead to bilateral blindness.

DIFFERENTIAL DIAGNOSIS
Other forms of Peripheral ulcerative keratitis (including Rheumatoid arthritis and Wegener's granulomatosis); Bacterial keratitis; Herpes zoster ophthalmicus (serpiginous ulcer); Herpes simplex keratitis (limbal).

SEE ALSO
Terrien's marginal degeneration; Corneal opacities – peripheral; Corneal perforation.

MANAGEMENT
Blood tests/microbiology
No specific serological test exists for Mooren's ulcer, although non-specific markers of systemic inflammation, such as the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level, are occasionally slightly raised. Investigations should be performed to exclude other forms of PUK with anti-neutrophil cytoplasmic autoantibodies (ANCA), angiotensin-converting enzyme (ACE), rapid plasma reagin (RPR), chest X-ray (CXR), hepatitis C antigen (HepC), full blood examination (FBE), and renal function tests. A biopsy of the adjacent conjunctiva may reveal a significant plasma cell infiltrate.

Systemic medication
Systemic immunosuppression is the mainstay of initial treatment, commencing with oral corticosteroids. Long term, the disease is managed with steroid-sparing immunosuppressive agents such as cyclophosphamide, with or without low-dose oral prednisolone. Patients are usually co-managed with a rheumatologist or clinical immunologist.

Therapeutics
In addition to systemic treatment, topical corticosteroids, prophylactic antibiotics,

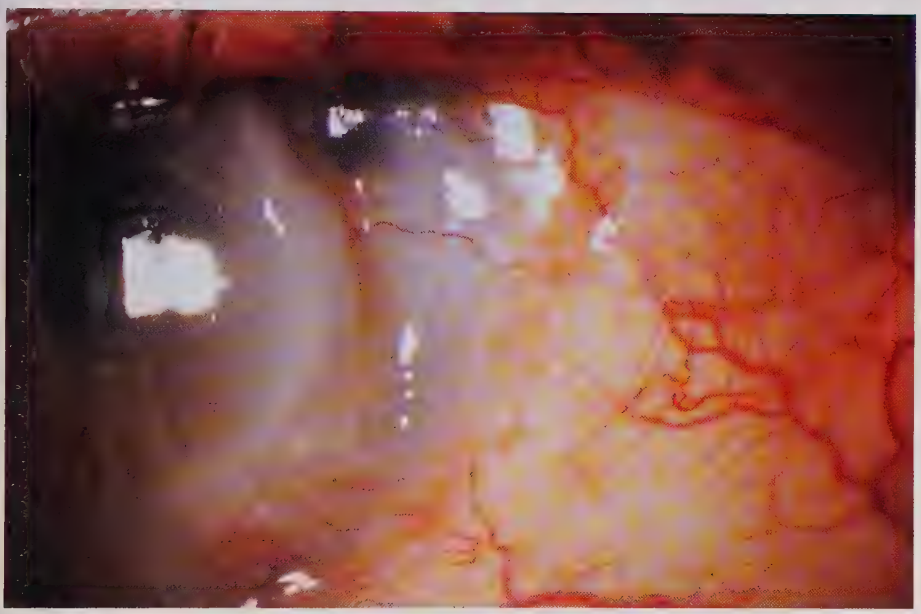


Fig. M-9 Extensive peripheral corneal stromal loss in Mooren's ulcer.

and, occasionally, mydriatics are commonly used.

Surgery

The role of surgery is still somewhat controversial. Many clinicians believe that an early conjunctival resection and recession can lead to a significant reduction in the disease process, and may even lead to total resolution. Surgery in conjunction with systemic immunosuppression is a common approach. Once the disease is brought under control, reactivation is uncommon. Surgery has the added advantage of providing tissue for histopathological examination.

Nasolacrimal duct obstruction – congenital

Dacryostenosis, epiphora – congenital, dacryocele

DESCRIPTION

At birth, the nasolacrimal duct may be delayed in opening, leading to congenital epiphora (tearing). The most common cause of the obstruction is an imperforate valve of Hasner, the mucosal fold covering the opening of the nasolacrimal duct into the nose.

If the lacrimal sac becomes filled with amniotic fluid or mucus due to the absence of lacrimal drainage, the condition is known as a dacryocele.

SYMPTOMS

The parents of the infant may notice epiphora, chronic mucopurulent discharge, or matting of the eyelids.

SIGNS

The main signs are tearing, which may be constant or intermittent. There may be some discharge from the punctum when the lacrimal sac is palpated; such a sign will confirm the diagnosis. The condition is more commonly unilateral but may be bilateral.

INCIDENCE

A delayed opening of the nasolacrimal duct is common (approximately 1/100) although, fortunately, associated infection is less common.

SIGNIFICANCE

The condition may be uncomfortable for the infant and of concern to the parent. There is potential for secondary infection and secondary dacryocystitis.

DIFFERENTIAL DIAGNOSIS

Punctal stenosis (atresia); Glaucoma – acute angle closure; Chlamydial conjunctivitis (neonatal); Ophthalmia neonatorum.

SEE ALSO

Dacryocystitis; Epiphora; Canaliculitis.



Fig. N-1 Congenital nasolacrimal duct obstruction. Bilateral epiphora due to delayed canalization of the nasolacrimal duct.



MANAGEMENT

Advice

In most situations the valve opens spontaneously within weeks to months.

Topical treatment

Massage of the nasolacrimal duct is the most common form of treatment. It consists of applying gentle pressure with a finger over the canaliculi and stroking downward firmly to increase the pressure within the lacrimal sac. Massage should be repeated 5 times qid. The objective is to hasten rupture of the valve obstruction.

Therapeutics

Use a broad-spectrum antibiotic ointment bd if mucopurulent discharge is present.

Surgery

Nasolacrimal duct probing is generally not performed prior to 12–18 months of age, in order to allow spontaneous canalization.

Neurotrophic keratopathy

Corneal anesthetic keratopathy

DESCRIPTION

Corneal sensation is via the nasociliary branch of the trigeminal (fifth cranial) nerve. Aside from providing corneal epithelial sensation, these nerves also appear to provide trophic factors important for normal corneal epithelial function. Interruption of the corneal nerve supply leads to typical corneal changes known as neurotrophic keratopathy.

The corneal nerve supply may be interrupted anywhere along its course, from the trigeminal nucleus in the brainstem through the trigeminal ganglion in the middle cranial fossa and along the course of the trigeminal nerve to the cornea itself.

Causes of damage to corneal innervation include:

- acquired infections – herpes simplex and herpes zoster keratitis
- tumors – acoustic neuromas, other orbital tumors
- surgery – especially surgical removal of an acoustic neuroma, LASIK
- congenital – Riley–Day syndrome (familial dysautonomia), anhidrotic ectodermal dysplasia
- systemic disease – end-stage diabetic disease.

One of the most common causes of damage to the trigeminal nerve is surgical removal of an acoustic neuroma. In this situation, nerve dysfunction is frequently due to neuropraxia and recovers over a period of weeks to months.

The exact cause of epithelial disruption in neurotrophic keratopathy is not known. However, reduced aqueous tear production and the removal of neurotrophic growth factor (NGF) are thought to be important in reduced epithelial metabolism and healing.

SYMPTOMS

Photophobia, redness, and blurring of vision may occur. However, pain is obviously not a significant feature.

SIGNS

Inferior corneal epithelial disturbances are the hallmark of this disease; they range from punctate erosions to frank ulceration. Neurotrophic ulcers typically have a smooth, regular distinct edge and are oval, being longest in the horizontal axis; they are very slow to heal. There is frequently associated ocular surface and even mild intraocular inflammation. Corneal sensation is reduced. Neurotrophic corneas are at an increased risk of microbial keratitis, especially from Gram-negative organisms.

INCIDENCE

Rare (Riley–Day syndrome is extremely rare and mainly seen in Jews of European descent).

SIGNIFICANCE

May lead to permanent visual loss from corneal stromal scarring, secondary to epithelial disease and microbial keratitis.

DIFFERENTIAL DIAGNOSIS

Herpes simplex keratitis; Dry eye syndromes; Toxic epitheliopathy; Exposure keratopathy.

SEE ALSO

Bell's palsy; Superficial punctate keratopathy (SPK); Persistent epithelial defects.

MANAGEMENT

The aim of treatment is to heal any epithelial defects, if present, and to maintain the integrity of the ocular surface, especially the corneal epithelium.

Tear supplements

The mainstay of treatment is to maximize the health of the ocular surface with unpreserved tear supplements. These are usually required on an hourly or 2-hourly basis.

Bandage contact lenses

The use of a bandage contact lens to treat a persistent epithelial defect in neurotrophic keratopathy is particularly problematic because of the increased risk of microbial keratitis. As such, they are rarely used.



Fig. N-2 Grouped superficial punctate epitheliopathy stained with rose bengal in mild neurotrophic keratopathy.

Therapeutics

Low-dose topical corticosteroids can help to settle any ocular surface inflammation. Usually, unpreserved unit dose prednisolone is used to minimize toxic damage to the epithelium. Topical nerve growth factor has also been reported to be of benefit in promoting healing of established epithelial defects; however, it is not commercially available.

Surgery

When there is a permanent reduction of corneal sensation, a lateral tarsorrhaphy is frequently required and is usually successful in protecting the ocular surface. It is almost universally necessary if there is associated facial nerve dysfunction. If the reduction in nerve function is likely to be temporary, as often occurs following intracranial surgery, a botulinum toxin-induced ptosis can be used to protect the ocular surface.

Nevus – eyelid

Pigment spot, mole, melanocytic nevi

DESCRIPTION

A nevus (mole) is an extremely common benign neoplasm. Nevi are composed of clusters of melanocytes that, although normally present at that site, have developed anomalous migration and development. If they are located deeper in the dermis, the cells can be amelanotic.

One classification is according to anatomical location:

- Junctional nevi are in the basal region of the epidermis and are the more common type in childhood.
- Intradermal nevi are located within the dermis and are more common in late adulthood. These may show fatty infiltration and fibrosis, giving the characteristic tumescence.
- Transitional (or compound) nevi are present in both locations and could be considered an intermediate stage between junctional and dermal types.

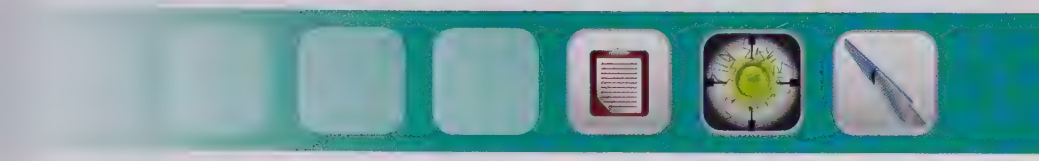
Sun exposure plays a role in the development of some nevi, as they tend to develop in areas of sun-exposed skin. There may also be hormonal influences, as nevi may increase in number or enlarge around puberty or during pregnancy. The age of onset is before 35 years of age, mostly before 25 years. However, the natural course for nevi is usually to gradually fade and flatten with age.



Fig. N-3a Non-pigmented intradermal nevus.



Fig. N-3b Pigmented junctional nevus.



SYMPTOMS
There are usually few symptoms associated with nevi. Nevi may be bilateral or unilateral, but are usually asymmetrical. Unlike a malignant lesion, nevi are not associated with symptoms such as itch, bleeding, inflammation, or discharge.

SIGNS
- Junctional nevi tend to be discrete, flat lesions with moderate to deep pigmentation, usually range from 1–3 mm in diameter, and often present in childhood or adolescence.
- Intradermal nevi are usually dome-shaped, and may be pigmented or non-pigmented. When located on the eyelid margin, lashes may grow through the lesion, but the lash line is not disrupted.
- A dysplastic nevus is defined clinically as greater than 5 mm in diameter, with indistinct margins and irregular pigmentation. Unlike acquired nevi, they tend not to regress with age. They can mimic superficial spreading melanoma.
- Congenital melanocytic nevi are usually large (greater than 15 mm diameter), slightly mounded, and often hairy. Giant congenital nevi are more than 20 mm in diameter.
- Oculodermal melanocytosis (nevus of Ota) is a melanocytic hyperpigmentation, slate gray in color, affecting both dermis and episclera.

Nevi should be distinguished from a malignant melanoma. Nevi are usually even in color, with a regular shape and border. The "**ABCDE**" mnemonic is useful for assessing for the possibility of malignant melanoma:
A = asymmetrical shape and elevation
B = border is irregular or scalloped
C = color is mottled and variable; melanoma may be vascularised
D = diameter is large (usually greater than 5 mm)
E = evolutionary change (in size or pigmentation).

INCIDENCE
Almost everyone has some nevi. They occur in all races, but occur most commonly in Caucasians, particularly those with light pigmentation of the skin and hair.

SIGNIFICANCE
Nevi are a benign condition and usually pose a low risk of conversion to malignancy. If there are large numbers of nevi (e.g. over 100), dysplastic or congenital nevi, there is a slightly higher risk of conversion to malignancy. Nevus of Ota is sometimes associated with melanoma of the choroid, other ocular structures, or central nervous system.

DIFFERENTIAL DIAGNOSIS
Melanoma – eyelid; Basal cell carcinoma (pigmented); Seborrheic keratosis.

SEE ALSO
Skin tumors – overview; Conjunctival pigmented lesions (nevus).

MANAGEMENT
Review
Documentation and review is the management mainstay of nevi. The advent of digital photography has made it easier to document a lesion and can also facilitate discussion with the patient.

Incisional surgery
In general, pigmented lesions considered at risk of malignancy (as defined above) should be excised with narrow margins for biopsy.

O

Ocular cicatricial pemphigoid (OCP)

Neo-natal inclusion conjunctivitis

DESCRIPTION

Ocular cicatricial pemphigoid (OCP) is a bilateral (although frequently asymmetric) progressive cicatrizing (scarring) condition of unknown etiology, but thought to be due to an autoimmune assault on the conjunctival basement membrane. The disease typically progresses slowly over a number of years. In rare cases, it may progress quite rapidly. About 20% of people with OCP have associated skin lesions with vesicles and bullae. Other mucous membranes (especially the oral mucosa) may be affected.

A drug-related form of the disease exists, with ocular features similar to true OCP. The drugs most commonly implicated are chronically used topical medications such as ocular anti-glaucoma medication.

SYMPTOMS

Early symptoms are those of chronic ocular irritation similar to dry eye. With progression of the disease, ocular irritation increases; there may be paradoxical tearing from punctal stenosis. In advanced disease, cicatricial entropion may occur with trichiasis. Vision becomes progressively blurred, both from ocular surface disturbances with dry eye and trichiasis and from increasing surface keratinization.

SIGNS

The hallmark of this disease is subepithelial fibrosis with progressive conjunctival shrinkage. In the initial stages, the fornices are blunted and shortened. The conjunctiva is drawn taut by subepithelial fibrosis. An early sign may be flattening of the caruncle with associated medial canthal keratinization. With progression, adhesions develop between the tarsal and bulbar conjunctiva (symblepharon). As the disease progresses further, the fornices are totally obliterated, the cornea becomes opacified as the epithelium becomes



Fig. O-1 Forniceal shortening, subepithelial scarring, and symblepharon formation in early OCP.



heavily keratinized, and adhesions develop between the upper and lower eyelids at the lateral canthus (ankyloblepharon). Associated ocular surface cicatricial changes include punctal stenosis, entropion, and glaucoma, probably partly from obliteration of the subconjunctival space.

In the early stages of the disease, conjunctival inflammation occurs; assessment of this inflammation is important in determining disease activity.

INCIDENCE

Very rare (approximately 1/100,000); it is more common in women by a factor of about 2:1 and usually occurs in the elderly.

SIGNIFICANCE

Bilateral, significantly sight-threatening.

DIFFERENTIAL DIAGNOSIS

Chemical burns; Stevens–Johnson syndrome; Trachoma; Toxic keratopathy.

SEE ALSO

Conjunctival scarring; Dry eye syndromes.

MANAGEMENT

Patients with OCP are usually best managed in a multi-discipline clinic where clinical immunologists and ophthalmic sub-specialists are available to manage individual manifestations of the disease.

Pathology

A conjunctival biopsy is frequently taken and submitted for histopathological examination. In addition to the typical signs of subepithelial fibrosis, immunohistochemistry is used to look for the deposition of complement and immunoglobulin deposition along the basement membrane zone. Identification of such changes is pathognomonic of OCP. However, only approximately 70% of clinical cases of OCP demonstrate such staining characteristics.

Tear supplements

From early on in the disease unpreserved tear supplements are important to maximize the health of the ocular surface.

Oral medication

The mainstay of long-term treatment is systemic immunosuppression to reduce scarring. Dapsone, sulfasalazine and methotrexate are commonly used first-line drugs, whereas azathioprine and cyclophosphamide are used for refractory disease.

Therapeutics

Topical corticosteroids are sometimes used for acute flare-ups of ocular surface inflammation, while anti-glaucoma medications such as beta-blockers (but not pro-inflammatory prostaglandin analogues) are also commonly required.

Surgery

Surgery is usually avoided, as it may lead to a significant increase in disease activity, although occasionally lid procedures for entropion with associated trichiasis are required. OCP has also traditionally been one of the indications for implantation of a keratoprosthesis.

O

Ophthalmia neonatorum

Neo-natal inclusion conjunctivitis

DESCRIPTION

Conjunctivitis occurring in the first 4 weeks of life is referred to as ophthalmia neonatorum. Ophthalmia neonatorum is usually secondary to an infectious cause and is acquired during delivery with passage of the baby through the birth canal. As such, it is unlikely to affect a child born by caesarean section; the causative organisms are also causes of sexually transmitted disease. The four typical causes of infectious ophthalmia neonatorum are:

- *Chlamydia trachomatis* (neonatal inclusion blennorrhea)
- gonococcal conjunctivitis
- other bacteria
- herpes simplex virus (rarely).

The time from birth until the onset of signs is variable, but typically 5 days for chlamydia, 1–3 days for gonococcal disease, 7 days for other bacteria, and over 10 days for herpes simplex virus. Previously (as was often required by law), many babies were treated with a topical 1% or 2% silver nitrate solution to prevent gonococcal disease. This could lead to a low-grade chemically-induced toxic conjunctivitis. Prophylactic treatment has now largely changed to topical erythromycin ointment, because of its enhanced anti-chlamydial activity.

SYMPTOMS

The parent reports a discharge, lid swelling, and red eyes in the infant. The parent may also have a personal history of genital chlamydia, bacterial vaginitis, genital herpes simplex, or gonorrhea.

SIGNS

The typical features of conjunctivitis are seen, with an edematous inflamed papillary change, a discharge, and lid swelling. Most cases are bilateral. No follicles are seen, even in chlamydial conjunctivitis in the newborn. A marked mucopurulent



Fig. O-2 Mucopurulent discharge in ophthalmia neonatorum caused by *Chlamydia trachomatis*.



discharge is typical of gonococcal disease, whereas a watery discharge is more typical of herpes simplex infection. With gonococcal conjunctivitis, corneal stromal lysis with perforation can occur rapidly.

INCIDENCE
Rare in developed countries.

SIGNIFICANCE
Gonococcal conjunctivitis can lead to rapid corneal perforation, whereas chlamydia can lead to pneumonia, and herpes simplex virus can cause encephalitis.

DIFFERENTIAL DIAGNOSIS
Congenital blockage of the nasolacrimal duct.

SEE ALSO
Chlamydial conjunctivitis; Gonococcal conjunctivitis; Herpes simplex keratitis – overview; Conjunctivitis – overview.

MANAGEMENT
Aside from the affected child, treatment of the mother and her sexual partner/s is also required.

Lid hygiene
Frequent eye hygiene and irrigation with sterile saline (to remove any purulent discharge) is required in acute gonococcal disease. This is very important to limit the adverse effect of the mucopurulent discharge on the ocular surface.

Microbiology
Swabs and microbiological investigations are essential to establish the cause of the conjunctivitis. If the cause is thought to be gonococcal, an urgent Gram stain needs to be performed. Identification of the characteristic Gram-negative diplococcus is a medical emergency.

Therapeutics
Only bacterial and toxic conjunctivitis is treated primarily with topical therapy. Other causes of the disease are treated primarily with systemic medication and ocular hygiene. Topical treatment is also often added to topical antibiotics in the treatment of chlamydial and gonococcal disease.

Oral medication
The diagnosis of neonatal gonococcal conjunctivitis is an ophthalmic emergency. Treatment is with systemic penicillin for the sensitive organism or, alternatively, with a cephalosporin. As chlamydial conjunctivitis is only one manifestation of a systemic disease, treatment with oral erythromycin (25 mg/kg of body weight bd for 14 days) is required to clear the infection. Tetracyclines are not appropriate because of their ability to stain the unerupted teeth. Active herpes simplex virus infection in the neonate is frequently treated with systemic antivirals such as acyclovir.

Notifiable
In some countries this infection may need to be reported to public health authorities.

Pannus and pseudopterygia

Superficial corneal vascularization

DESCRIPTION

There are three main types of superficial corneal vascularization, detailed below.

Pannus

Pannus is a fibrovascular infiltration of the superficial subepithelial peripheral cornea, due to centripetal extension of the normal limbal vessels. It is relatively common, may resolve spontaneously, and is usually secondary to sustained paralimbal inflammation, although it may be a sign of localized limbal stem cell dysfunction. The most common cause of pannus is chronic hypoxia from soft contact lens use. Other causes of pannus include trachoma, adult inclusion keratoconjunctivitis (chlamydia), marginal keratitis, chemical burns, ocular cicatricial pemphigoid, rosacea keratitis, and superior limbic keratoconjunctivitis.

Pseudopterygia

While true pterygia do not extend through Bowman's layer (largely sitting on the surface of the cornea), pseudopterygia involve scarring and vascularization of the

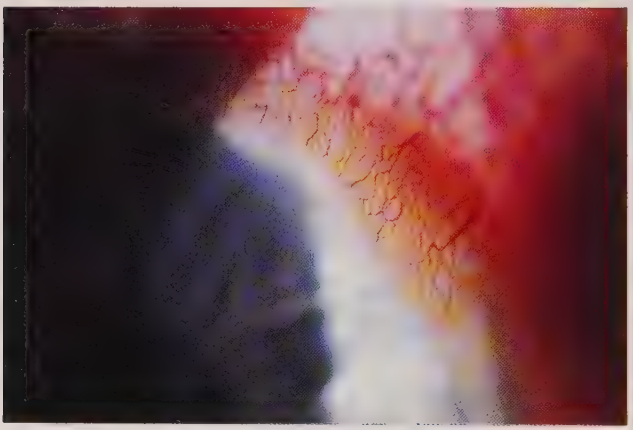


Fig. P-1a Mild-to-moderate superior pannus secondary to marginal keratitis.

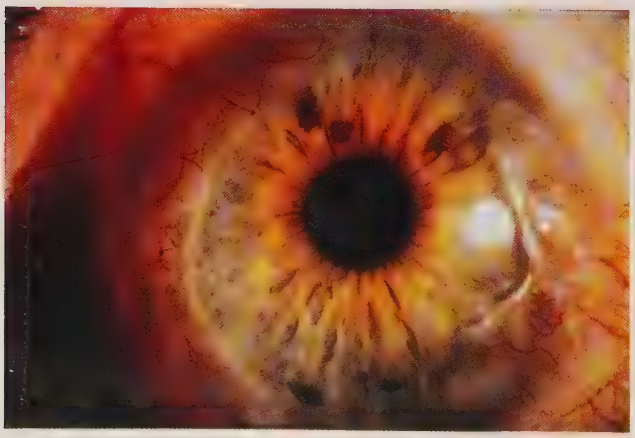


Fig. P-1b Pseudopterygium formation in Terrien's marginal degeneration.



corneal stroma. They are most commonly caused by previous limbal corneal trauma or infection but also occur with some peripheral corneal disease, such as Terrien's marginal degeneration, Mooren's ulcer, and ocular cicatricial pemphigoid.

Fascicular keratitis

A single leash of vessels may extend from the limbus to an old area of keratitis. This is especially common following herpes simplex keratitis and bacterial keratitis.

SYMPTOMS

Unless it involves the central cornea, vascularization by itself causes few if any symptoms, although some slight ocular irritation can occur. Any condition that causes corneal vascularization also typically causes photophobia, blurring of vision, and ocular irritation.

SIGNS

Pannus is usually superior with contact lenses, adult inclusion conjunctivitis, and trachoma. It is usually more inferior with chemical burns, marginal keratitis, and rosacea keratitis. The pannus appears as a superficial fibrovascular opacity extending 1 mm or more from the limbus. The overlying epithelium is usually slightly thickened and raised. Pannus may regress with time if the stimulus for its formation is withdrawn.

Pseudopterygia are thickened and involve distortion of the adjacent limbus and conjunctiva. Frequently, there is some degree of loss of stromal tissue and lipid deposition. In contrast to true pterygia, they are usually more broad-based extending several clock-hours around the limbus. The orientation is often less radial than with true pterygia.

Fascicular lesions are usually focal, and extend directly to an old area of stromal loss with scarring. There may also be associated lipid deposition.

INCIDENCE

Common.

SIGNIFICANCE

Rarely causes vision loss or irritation by itself.

DIFFERENTIAL DIAGNOSIS

Limbic tumors, including: Corneal intraepithelial neoplasia; Melanoma; stromal vascularization; Terrien's marginal corneal degeneration; Pterygium.

SEE ALSO

Limbal stem cell deficiency; Contact lens-related conditions; Lipid keratopathy.

MANAGEMENT

No specific treatment is usually required for the vascularization itself. However, the underlying cause should be treated.

Tear supplements

If causing ocular discomfort, tear supplements are usually of benefit.

Modify lens use

Contact lens-associated pannus may require modification of lens use, with both improved lens cleaning and refitting with lenses that are more oxygen-permeable, such as silicone–hydrogel lenses.

Pellucid marginal degeneration (PMD)

DESCRIPTION

Pellucid marginal degeneration (PMD) is one of the three corneal ectatic dystrophies; as such, the term degeneration is a misnomer. It typically has a relatively late onset, with the first symptoms of corneal astigmatism appearing in the fourth decade of life. The characteristic feature is inferior corneal thinning in a crescent about 1 mm central and concentric to the limbus. The word "pellucid" means clear and refers to the lack of scarring, although some slight superficial vascularization may occur. Although the condition progresses over the years, it is similar to keratoconus in that the degree of involvement is very variable. As with the other ectatic dystrophies, hydrops can occur in extreme cases.



Fig. P-2a PMD with inferior thinning.

SYMPTOMS

Patients with PMD usually do not report any ocular irritation but do have significant and often increasing visual symptoms, experiencing both blurring of vision and glare sensitivity. The blurred vision usually commences in the 30s and results from induced against-the-rule astigmatism. Initially, this is correctable with spectacles. Over time, patients require rigid contact lenses due to the loss of best spectacle-corrected vision due to the irregular nature of the astigmatism. If hydrops occurs, the patient experiences a sudden marked deterioration in vision and significant discomfort, photophobia, and increased lacrimation.

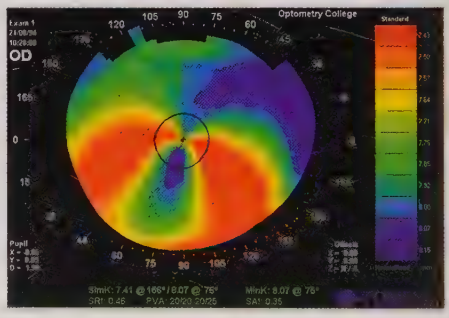


Fig. P-2b Corneal topographical changes typical of PMD.

SIGNS

PMD is almost always bilateral, although it may be asymmetric. There is a clear area of thinning in the inferior cornea approximately 1 mm from the limbus, which typically extends for about 90°, from 4 to 8 o' clock. Unlike keratoconus, where the thinned area is more steeply curved, in PMD the cornea is drawn out and flattened at the thinned area and more steeply curved superior to the thinning.

INCIDENCE

Very rare.

SIGNIFICANCE

Leads to significant blurring of vision due to induced irregular against-the-rule astigmatism.

DIFFERENTIAL DIAGNOSIS

Keratoconus; Terrien's marginal degeneration.



SEE ALSO

Keratoglobus; Corneal graft; Corneal collagen cross-linking; hydrops; Corneal dystrophies, degenerations, and deposits.

MANAGEMENT

Genetic advice

The mode of inheritance of PMD is unclear; most cases occur sporadically. There are reports of PMD and keratoconus both occurring in members of a single family.

Ocular investigations

Computerized videokeratoscopy (corneal topography) can be very useful in confirming the diagnosis, especially in mild cases. There is a characteristic "pinch" sign with a "teardrop" of flattened cornea inferior to the visual axis surrounded by areas of steepening.

Contact lenses

Rigid gas-permeable lenses can be fitted to correct the induced corneal irregularities. Inferior edge lift is a significant problem in this condition. Gas-permeable scleral lenses may be necessary in some cases.

Corneal collagen cross-linking

Chemical cross-linking of the corneal collagen with riboflavin-5-phosphate activated by UV light may be useful in preventing progression of the disease if the cornea is not too thin or scarred.

Surgery

Rarely, the induced corneal irregularity becomes so marked that patients are unable to be successfully fitted with standard rigid contact lenses or even scleral lenses. In this situation, corneal surgery may be indicated. Several different surgical procedures have been tried in PMD. These include a wedge excision of the affected area, thermokeratoplasty, and a large eccentric corneal graft. The most effective method is probably a corneoscleral lamellar inlay graft. This is placed inferiorly, involving the area of corneal thinning. This is usually crescentic in shape and does not involve the central 7–8 mm of cornea. It is used to reduce the central corneal irregular astigmatism.

Penetrating eye injuries

Lacerations, intraocular foreign body

DESCRIPTION

Trauma resulting in a penetrating eye injury commonly lacerates not only the cornea and/or the conjunctiva but can also damage periocular structures such as the eyelids and canaliculi. Common sources of trauma include metal from grinding or drilling, shattered spectacle glass, windscreen glass from an automobile accident, nails from construction, and foliage from gardening. They are commonly work-related but also occur secondary to home and garden maintenance, road accidents, and assault. Of particular concern is the high rate of endophthalmitis and the occurrence of a retained intraocular foreign body (IOFB). In contrast to post-cataract surgery endophthalmitis, where the causative organism is usually a Gram-positive bacterium, post-traumatic endophthalmitis is associated with a high rate of Gram-negative organisms.

SYMPTOMS

There is usually a history of ocular trauma. Corneal lacerations are typically much more painful than conjunctival lesions. There is usually associated tearing, leading to blurring of vision, a red eye, photophobia, and lid edema. A thorough history should be taken to identify any risk of a retained intraocular foreign body. A particularly suggestive feature is a high speed injury, such as with a drill or grinding accident.

SIGNS

A thorough examination of the eye and periocular tissue is required to assess accurately the extent of the damage. Observation of the depth of any ocular injury is particularly important to identify a penetrating injury as well as any damage to intraocular structures. With ocular penetration, aqueous humor passing out through the wound results in an obvious dilution of fluorescein and is easily seen on a slit lamp with a cobalt blue filter in place. This is termed a positive Seidel's sign. Other signs of penetration include shallowing of the anterior chamber, a low intraocular pressure, pupil distortion, and even cataract if the lens has been injured. A thorough fundal examination is necessary, if there is ocular penetration, to exclude posterior segment damage including retinal tears, retinal detachment, and endophthalmitis. Factors identified on multivariate analysis as being associated with a higher risk of developing post-traumatic endophthalmitis are a delay in primary repair of the laceration greater than 12 hours, rupture of the lens capsule, and a dirty wound.

INCIDENCE

Penetrating injuries are uncommon (approximately 1/1000) in developed countries, especially with the increased use of seat belts in cars and safety (protective) glasses in industry. They occur more commonly in the young and in males.

SIGNIFICANCE

Scarring and infection can occur with any ocular surface injury. Therefore, rapid re-epithelialization and protection with topical prophylactic antibiotics is important. Corneal epithelial injuries can predispose to microbial keratitis or recurrent corneal erosion syndrome.

DIFFERENTIAL DIAGNOSIS

Perforation – corneal.

SEE ALSO

Recurrent corneal erosion syndrome; Foreign bodies – corneal and conjunctival; Eyelid trauma (hematoma); Endophthalmitis; Abrasion.

MANAGEMENT

Therapeutics

The initial instillation of a long-acting cycloplegic drug such as atropine to prevent ciliary spasm may be indicated if there are signs of an anterior chamber reaction. Following repair of the wound, patients are normally placed on a topical fluoroquinolone qid and a topical corticosteroid such as prednisolone acetate qid.

Radiological investigation

A plain X-ray or CT-scan is indicated if there is any feature of the history suggestive of a retained intraocular foreign body.

Surgery

Small self-sealing penetrating corneal wounds may not require suturing. A small leak with a well-formed anterior chamber and near normal intraocular pressure may be managed with topical Alphagan bd to reduce the risk of fluid flow through the wound, a bandage contact lens, and eye shield. If the wound has an active leak, or if there is associated intraocular trauma requiring surgical attention such as an acute cataract, the wound should be sutured. Intraocular structures are often not repaired at the time of the initial surgery, allowing time first for the eye to be quietened with primary repair and topical corticosteroids. If cataract extraction is performed at the time of the initial repair, an IOL is frequently not placed but is rather implanted at a later date when keratometry (and therefore calculated IOL power) can be more accurately measured. If required, an iris laceration can be repaired with a McCannel or Siepser suture. Intracameral or even intravitreal antibiotics can be injected at the time of the primary repair to reduce the risk of endophthalmitis.

Review

Even with small self-sealing wounds, the patient should be examined regularly on a daily to twice daily basis following the injury to identify any signs of endophthalmitis or microbial keratitis. In the event of significant damage to the iris or angle, long-term follow-up should be performed initially on a several monthly and later on a yearly basis to monitor for the development of a raised intraocular pressure and glaucoma.

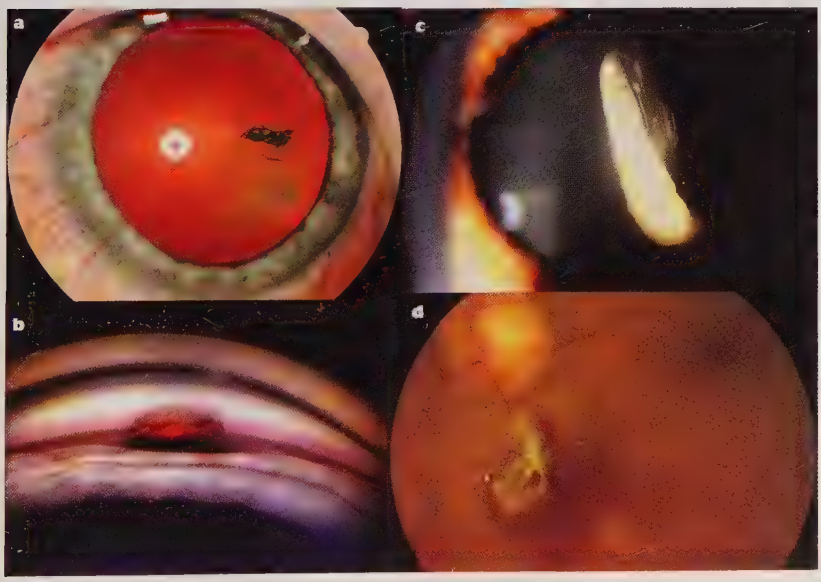


Fig. P-3 Metallic intralenticular (a) and (c), in the angle (b), and intravitreal with associated hemorrhage (d) intraocular foreign bodies (IOFBs).

P Perforation – corneal

DESCRIPTION

Perforation of the cornea occurs from full thickness stromal loss. The causes of perforation include:

- infections – bacterial, fungal, and viral (especially herpes simplex virus) keratitis
- ocular surface disease – dry eye, rosacea keratitis, persistent epithelial defects
- collagen-vascular disease – rheumatoid disease, polyarteritis nodosa (PAN), Wegener's granulomatosis, systemic lupus erythematosus (SLE)
- vitamin deficiency – hypovitaminosis A
- trauma – direct mechanical trauma or chemical burn
- idiopathic – Mooren's ulcer (usually with associated trauma).

Perforation occurs following epithelial ulceration, stromal loss, and rupture of a descemetocoele (area of Descemet's membrane without any overlying stroma or epithelium).

This article deals with perforation and its treatment; details of the specific disease entities listed above are contained in their individual articles.

SYMPTOMS

Patients often complain of the underlying disease and, at the time of perforation, report blurring of vision, increased and fairly constant tearing, and mild ocular discomfort.

SIGNS

The perforation usually occurs within a larger area of stromal loss and epithelial ulceration. The perforation is identifiable as a clear area within the wider area of involvement. A clear area may be a perforation or a bare Descemet's membrane (descemetocoele). Signs that the lesion is leaking aqueous humor are a shallow anterior chamber, lowered intraocular pressure (IOP), and a positive Seidel test, when dilution of concentrated fluorescein by leaking aqueous humor as viewed with a cobalt blue filter in place leads to the appearance of a green plume. The depth of the anterior chamber should

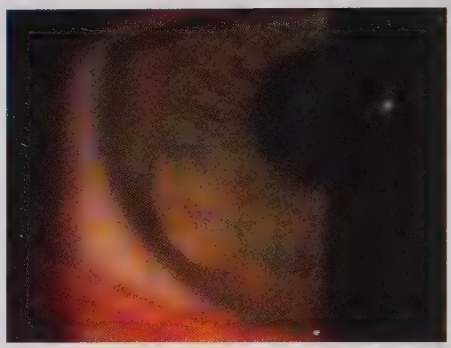


Fig. P-4a Corneal perforation in a patient with keratoconus and hydrops.

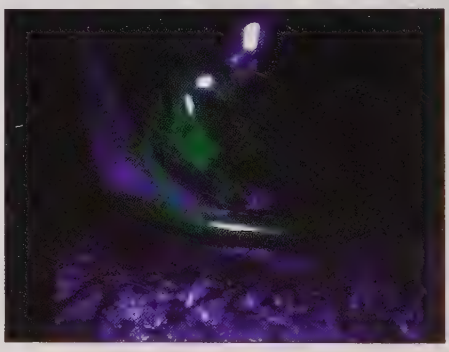


Fig. P-4b Same patient seen after instillation of fluorescein under cobalt blue light illumination, demonstrating positive Seidel's sign.

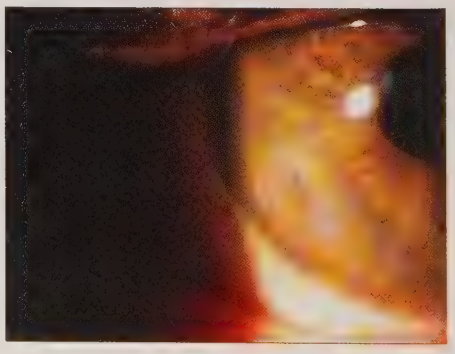


Fig. P-4c The lesion after application of cyanoacrylate glue.



be observed, including any corneal touch by the lens and whether any iris is plugging the defect.

INCIDENCE
Very rare.

SIGNIFICANCE
May lead to a low IOP and flat anterior chamber with hypotonous maculopathy, cataract, and peripheral anterior synechia formation. Rarely, a perforation can also lead to the development of endophthalmitis and expulsion of intraocular contents.

DIFFERENTIAL DIAGNOSIS
Descemetocoele.

SEE ALSO
Rheumatoid arthritis and the eye; corneal/conjunctival lacerations; microbial keratitis; Corneal graft; Mooren's ulcer; Chemical burn; Peripheral ulcerative keratitis; Persistent epithelial defect.

MANAGEMENT
Therapeutics
As necessary, topical medication to control the underlying disease process (such as a microbial keratitis) should be commenced. A topical broad-spectrum antibiotic should be commenced qid to decrease the chance of microbial keratitis or endophthalmitis developing.

Oral medication
Oral acetazolamide (Diamox) is frequently helpful to reduce the flow of aqueous humor through the site of perforation, thereby aiding in the healing of the lesion. If the patient is feeling nauseous, antiemetics are indicated to reduce the risk of the expulsion of intraocular contents that can occur from the rise in IOP with vomiting. If the perforation is caused by a systemic collagen vascular disease, this needs to be brought under control, usually with systemic immunosuppressive agents.

Surgery
Several surgical options exist to seal the perforation. Most commonly, cyanoacrylate glue is applied to the site and a bandage contact lens applied. Application of the glue can be difficult; it is important that just enough glue to seal the perforation is applied. Excess glue sits proud of the site of perforation and is relatively unstable due to the mechanical action of the eyelids. Several methods of applying only a thin film of glue have been developed.

Other surgical options include a lamellar or penetrating keratoplasty and advancement of a thick conjunctival flap.

Peripheral ulcerative keratitis (PUK)

DESCRIPTION

Peripheral ulcerative keratitis (PUK) is not a specific disease entity, but rather a clinical presentation with a typical constellation of peripheral corneal inflammatory signs. The recognition that a patient has PUK is important, as it identifies a limited number of predominantly systemic diseases that cause the condition. The term PUK does not include all peripheral inflammatory (or even ulcerative) conditions, only those with the following signs.

The key features are extensive, usually crescentic, peripheral corneal ulceration (usually greater than 5 mm in length), corneal inflammation, and stromal tissue loss. In some diseases, there may be similar changes in the adjacent sclera. Many of these conditions are extremely rare, and a high level of suspicion and directed investigations are required.

Causes of PUK

- Collagen/vascular diseases
  - —Rheumatoid factor (RhF)-positive disease, rheumatoid arthritis
  - —Anti-neutrophil cytoplasmic antibody (ANCA)-positive disease
    - – Wegener's granulomatosis
    - – Polyarteritis nodosa
    - – Churg–Strauss disease
  - —ANCA- and RhF-negative disease
    - – Relapsing polychondritis
    - – Systemic lupus erythematosus
- Infective
  - —Acute
    - – Bacterial
    - – Viral: HSV, VZV
    - – Fungal
  - —Chronic
    - – Bacterial: syphilis
    - – Viral: hepatitis C
- Autoimmune
  - —Mooren's ulcer
- Idiopathic
  - —Rosacea keratitis

SYMPTOMS

PUK usually has a subacute onset with increasing blur, ocular irritation, photophobia, and redness. The disease is usually unilateral, although may rarely be bilateral, especially with Mooren's ulcer.

SIGNS

PUK is an acute inflammatory process with peripheral corneal ulceration, infiltration, and tissue loss. The area of involvement is usually paralimbal, but may breach the limbus to involve the adjacent sclera. This is especially the case with Wegener's granulomatosis. There may be extensive tissue loss and possibly corneal perforation. The anterior chamber may be secondarily slightly inflamed. Other signs of the causative disease should be sought, such as arthritis with rheumatoid, a vasculitic skin rash with polyarteritis nodosa, nasal granulomata with Wegener's, and auricular cartilage loss with relapsing polychondritis.

INCIDENCE

PUK is extremely rare as are many of the underlying causes.

SIGNIFICANCE

Depending on the specific causative disease, may lead to both loss of sight and, in the case of ANCA-positive diseases, loss of life.

DIFFERENTIAL DIAGNOSIS

Marginal keratitis; Terrien's marginal degeneration.

SEE ALSO

Rheumatoid arthritis and the eye; Perforation – corneal; Mooren's ulcer; Bacterial keratitis; Herpes zoster ophthalmicus; Herpes simplex keratitis – overview.

MANAGEMENT

With recognition of a PUK, the initial management is focused on identifying any underlying cause. Therapy can then be directed, according to the specific diagnosis, as given in each disease article.

Investigations

The following series of investigations should be ordered to aid with making the diagnosis.

- Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP): as non-specific markers of systemic inflammation
- ANCA: especially to identify patients with Wegener's, also a surrogate marker of disease activity
- Rh factor: the patient almost always has known rheumatoid disease; however, the level helps identify systemic disease activity
- Venereal Disease Research Laboratory (VDRL) and fluorescent treponemal antibody absorbed (FTA-ABS): for syphilis
- Hepatitis C antibodies: for hepatitis C
- Chest X-ray: for signs of Wegener's
- Renal function tests: for renal involvement from a systemic vasculitis

Histopathology

A conjunctival biopsy may be useful in making the diagnosis, demonstrating either an extensive plasma cell infiltrate in Mooren's or vasculitis with ANCA- and rheumatoid factor-positive disease.



Fig. P-5 PUK involving the medial paralimbal in a patient with Wegener's granulomatosis.

Persistent epithelial defect (PED)

Persistent corneal ulcer

DESCRIPTION

A persistent epithelial defect (PED), or a chronic ulceration of the corneal epithelium, is one of the more difficult-to-treat ocular surface conditions. Once the defect has occurred, lack of healing may result from many different causes and often from more than one factor. For example, a non-healing defect in herpes simplex keratitis (termed a metaherpetic ulcer) may occur from neurotrophic changes due to chronic viral infection, stromal disruption, ongoing viral replication, and toxicity from topical antivirals.

Factors contributing to lack of healing of the epithelium

- Tear film disturbances: dry eye, dellen, lagophthalmos
- Eyelid disorders: lagophthalmos, exposure keratitis, trichiasis
- Stromal disorders: with significant stromal inflammation and following digestion by lytic enzymes as occurs with a Gram-negative keratitis, the stroma may not support the epithelium
- Infection: ongoing stromal or epithelial infection may result in a PED
- Allergy: chronic ocular surface allergy such as vernal keratoconjunctivitis or atopic keratoconjunctivitis is a potent cause of PEDs
- Corneal anesthesia: leading to neurotrophic keratitis
- Toxicity: especially from multiple and frequent preserved drops and ointments, and with anesthetic abuse keratopathy
- Limbal stem cell deficiency: may lead to both ulceration and PEDs
- Systemic disorders: uncontrolled diabetes mellitus

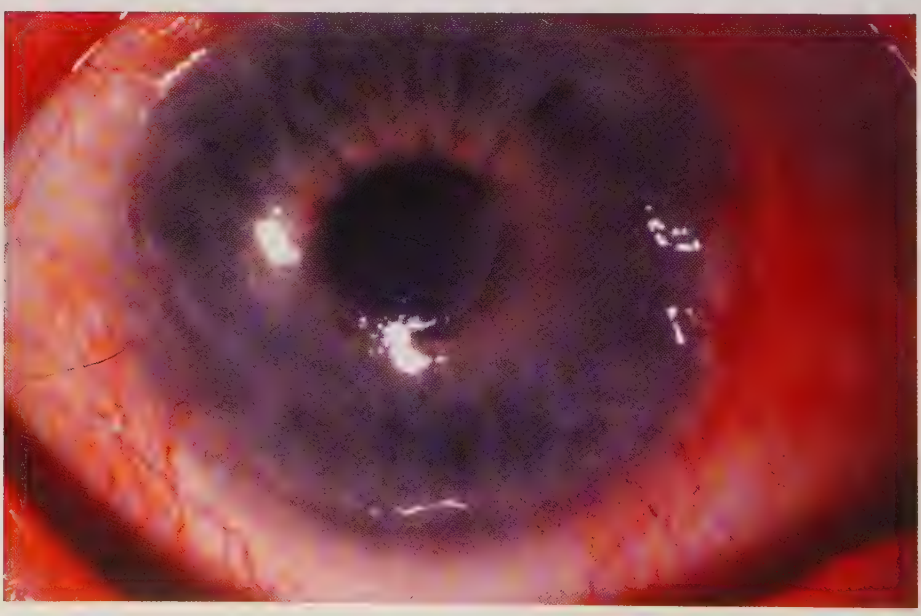


Fig. P-6 PED (metaherpetic ulcer) in a patient with chronic recurrent herpes simplex keratitis.



SYMPTOMS
Most PEDs lead to irritation, increased lacrimation, and a foreign-body sensation but not marked pain. This is probably secondary to localized reduced sensation.

SIGNS
A PED has a characteristic appearance, being round or oval, often with the long axis horizontal. The edges are raised and smooth and are whitish. The exposed stroma is also often slightly whitish, and there may be associated stromal loss with thinning. A thorough examination of the ocular surface and eyelids is indicated to identify any predisposing factors as listed above.

INCIDENCE
Rare.

SIGNIFICANCE
May lead to stromal loss, perforation, infection, and scarring of the cornea.

SEE ALSO
Toxic keratopathy; Limbal stem cell deficiency; Herpes simplex keratitis; Exposure keratopathy; Superficial punctate epitheliopathy; Trichiasis; Perforation – corneal; Neurotrophic keratitis; Dry eye – overview; Recurrent corneal erosion.

MANAGEMENT
The mainstay of treatment is to optimize the health of the ocular surface, and thereby promote epithelial healing. Any underlying cause such as dry eye, infection, trichiasis, and lagophthalmos should be treated. Of prime importance is the reduction of toxicity from topical medication. All topical medications should be kept to a minimum and, wherever possible, should be preservative-free. When outside, sunglasses with good side-protection should be worn to protect the ocular surface from the drying action of hot or cold winds.

Tear supplements
Adequate lubrication is essential, to stabilize the tear film and minimize ocular surface trauma. Only unpreserved drops, gel, or ointment should be used, usually 1- to 2-hourly drops, and ointment at night.

Therapeutics
A topical antibiotic is used prophylactically until the ulcer is healed. If there is stromal or ocular surface inflammation, a mild topical corticosteroid can also be used 2 to 3 times a day. Autologous serum drops have also been used to promote epithelial healing and may be tried in ulcers refractory to other measures.

Therapeutic contact lens
A therapeutic contact lens worn on an extended-wear basis may be applied to protect the ocular surface and promote healing. This is usually only used if more conservative measures have failed. Applying a contact lens when there is an established ulcer and ocular surface disease probably increases the risk of microbial keratitis. Therefore, a topical antibiotic is used 2 to 3 times a day.

Surgery
Protection of the ocular surface can also be achieved with a botulinum toxin tarsorrhaphy or surgical lateral tarsorrhaphy. While a botulinum toxin tarsorrhaphy only lasts several months, a surgical tarsorrhaphy is more permanent and, as such, is usually reserved for cases where ongoing protection is required. Occasionally, other intervention such as a bipedicled conjunctival advancement flap, application of an amniotic membrane graft, or even a corneal graft may be required. If there is significant stromal loss with threatened perforation, application of cyanoacrylate glue and a therapeutic contact lens may be necessary. A superficial keratectomy may also be tried if stromal factors are thought to play a significant role in preventing epithelial closure. Debridement of the edge of the ulcer can also be used to promote healing of a refractory defect.

P

Phlyctenulosis

Migrating phlycten

DESCRIPTION

Phlyctenulosis is a conjunctival and corneal inflammatory disease that predominantly affects children. Similarly to marginal keratitis, the etiology is thought to be a hypersensitivity reaction to bacterial antigens. The two main conditions associated with phlyctenulosis are staphylococcal blepharitis and tuberculosis. Rarer causes include coccidioidomycosis. Although previously more common, phlyctenulosis is now a rare condition. Histopathologically, a phlycten is an inflammatory mass primarily composed of lymphocytes and plasma cells.

SYMPTOMS

Marked photophobia and increased lacrimation are typical of both conjunctival and corneal phlyctens. The vision is often slightly blurred from the increased tearing, and secondary blepharospasm may occur.

SIGNS

Most phlyctens develop at the limbus or on the paralimbal conjunctiva. A conjunctival phlycten is a small, raised, pink-white nodule surrounded by hyperemic conjunctiva. Limbal phlyctens are also raised and whitish, and over time may resolve or migrate centrally onto the cornea. Like limbal and conjunctival phlyctens, corneal phlyctens are raised; this distinguishes them from marginal keratitis in which the affected cornea is ulcerated and depressed. A corneal phlycten often has associated underlying superficial vascularization and stromal loss; these changes follow the migration of the phlycten across the cornea over weeks to months. Rarely, the phlycten may ulcerate or even lead to perforation of the cornea. Associated blepharitis or signs of tuberculosis should be sought.

INCIDENCE

Very rare, although more common in developing countries in people with poor sanitation.

SIGNIFICANCE

Usually self-limiting but may, rarely, lead to permanent visual loss from corneal stroma loss and scarring.

DIFFERENTIAL DIAGNOSIS

Marginal keratitis; microbial keratitis; Pyogenic granuloma (corneal); Pingueculum.

SEE ALSO

Blepharitis – anterior; Interstitial keratitis.

MANAGEMENT

If there is any concern that the phlycten is caused by tuberculosis, referral for investigation with a chest X-ray, Mantoux test, and sputum cultures is indicated.

Therapeutics

Topical corticosteroids such as fluorometholone acetate 1% and a broad-spectrum antibiotic with good staphylococcal coverage (such as ciprofloxacin) are the appropriate initial treatment. These are administered qid for 2–4 weeks. It is reported that tubercular phlyctens are very responsive to topical corticosteroids, whereas staphylococcal phlyctens are relatively resistant. If persistent, staphylococcal phlyctens can also be treated with a several month course of an oral tetracycline such as doxycycline, combined with regular lid scrubs.

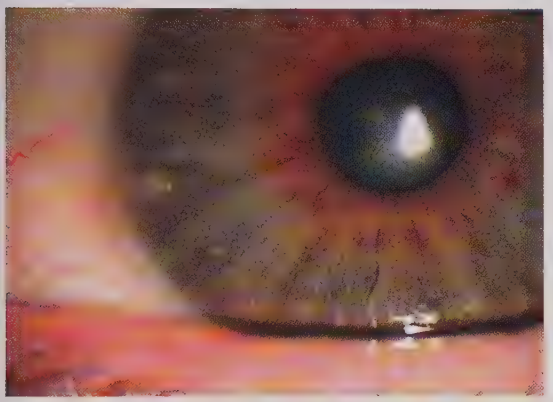


Fig. P-7a Peripheral staphylococcal corneal phlycten.

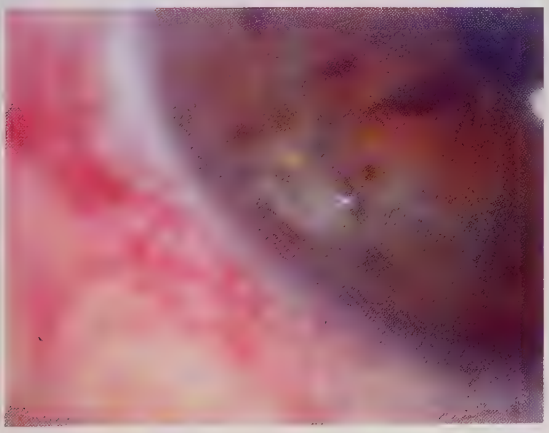


Fig. P-7b Higher-powered view demonstrating associated corneal vascularization.

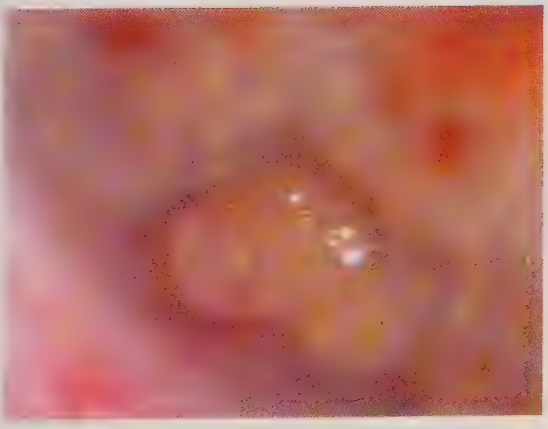


Fig. P-7c Higher-powered view demonstrating raised "stuck-on" appearance.

Pingueculum

DESCRIPTION

A pingueculum is a degeneration of the exposed interpalpebral conjunctiva. It is associated with chronic ultraviolet exposure and chronic irritation from wind and dust.

The pathogenesis of pingueculum is a degeneration of the conjunctival stroma, with degeneration of the collagen fibers and some thinning of the overlying epithelium. Instead of the normal thin and transparent nature of the conjunctiva, there is focal thickening and opaque deposition.

SYMPTOMS

Most patients are asymptomatic, although the pingueculum may at times become inflamed or irritated. The condition is usually bilateral and reasonably symmetrical.

SIGNS

Pingueculum appears as gelatinous deposits in the conjunctiva, adjacent to the limbus at the 3 and 9 o'clock positions, without corneal involvement. They are more common medially. The lesion may

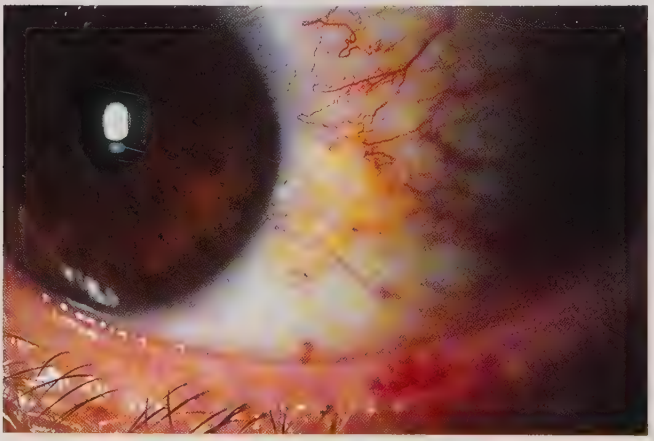


Fig. P-8a A small pingueculum.

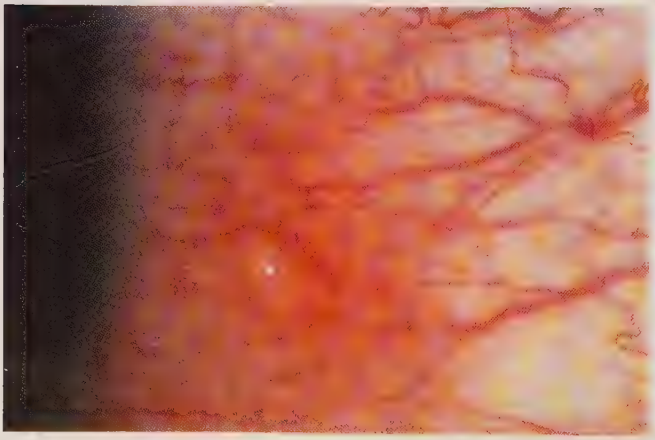


Fig. P-8b Mild inflammation of a pingueculum.



be transparent, white, or yellow, with calcification sometimes present. The surface of the deposit is usually raised, and it may be round or triangular (base toward limbus) in shape.

While a pingueculum may be hyperemic, it is not usually excessively vascularized; the vascularity is similar to or slightly greater than that of the adjacent uninvolved conjunctiva. If the apex of the lesion stains with fluorescein, this is indicative that the pingueculum is creating an ocular tear film-surfacing problem. A corneal dellen may occur if the pingueculum is sufficiently raised to disrupt the adjacent pre-corneal tear film. Following acute UV exposure, the pingueculum may become inflamed (pingueculitis).

INCIDENCE

Very common (greater than 1/10) in older age groups or in particular occupational or recreational groups of people with above-average levels of UV exposure.

SIGNIFICANCE

Pinguecula are a benign degenerative change. There may be mild cosmetic concern or mild discomfort; however, there is no threat to sight.

DIFFERENTIAL DIAGNOSIS

Pterygium; Conjunctival intraepithelial neoplasia; dermoids.

SEE ALSO

Pannus; Dellen.

MANAGEMENT

Advice

The relationship of the condition with UV radiation and environmental exposure should be explained, and the use of sunglasses advocated. Wrap-around sunglasses or sunglasses with side-shields are beneficial in very high UV environments such as on the water or snow.

Tear supplements

Tear supplements can be used to reduce desiccation of the lesion and reduce symptoms, if present. They may be used qid; if more frequent dosing is necessary, unit dose drops will avoid possible toxicity effects from preservatives. Try to avoid the older formulation of tear supplements preserved with benzalkonium chloride (BAK), unless for occasional use only.

Therapeutics, and review

If the lesion is hyperemic or inflamed, it can be decongested with short-term palliative measures such as cold compresses, mild vasoconstrictors, or topical non-steroidal anti-inflammatory drugs (NSAIDs). If there is any uncertainty in diagnosis or nature of the lesion, review may be appropriate.

Surgery

Excision, though rarely required, is usually perfomed for cosmetic reasons or if the pingueculum is interfering with soft contact lens wear. The best cosmetic result is achieved if, similarly to pterygium surgery, the defect is covered with an autograft taken from the superior bulbar conjunctiva.

Posterior capsular opacification (PCO)

Capsular fibrosis, Elschnig pearls

DESCRIPTION

Posterior capsular opacification (PCO) refers to fibrosis and opacification of the posterior capsule of the crystalline lens following cataract extraction. This occurs due to the posterior migration and proliferation of equatorial lens epithelial cells. This remains a significant cause of post-cataract surgery visual loss.

PCO does not normally develop for at least several months and may develop at any time following surgery, although it rarely develops after 5 years following surgery. The degree of opacification slowly increases over months to years. Occasionally, especially with posterior subcapsular and congenital cataract, the capsule is already opaque prior to cataract extraction.

There are two main categories of factors demonstrated to decrease the risk of PCO:

- Surgical factors
  - —A capsulorrhexis at the time of surgery that is both intact and smaller than the diameter of the optic of the IOL
  - —Placement of the IOL with both haptics in the capsular bag
  - —Complete removal of the lens cortex
- IOL factors
  - —IOL material, with hydrophobic acrylic lenses probably having the lowest PCO rate
  - —IOL optic edge design, with a square edge having the lowest PCO rate

SYMPTOMS

PCO leads to blurring of vision and glare sensitivity. Some patients describe "streaking of lights," especially at night. They may describe it as being like the cataract returning.

SIGNS

With PCO, the capsule becomes progressively opaque, with bands or a whitish plaque. There may be associated wrinkling of the capsule, and cystic groups of epithelial cells may be present (Elschnig pearls). Particularly when an anterior capsulorrhexis has been performed, as with phacoemulsification surgery, anterior capsular opacification and contraction (capsulophimosis) may also be present.

INCIDENCE

Common, depends on lens style and type of surgery performed; however, overall between 10% and 50% following cataract surgery. The incidence of visually significant PCO has fallen from approximately 50% with extracapsular cataract extraction and implantation of a rigid PMMA intraocular lens (IOL) to approximately 5–10% with phacoemulsification and the use of newer foldable intraocular lenses.

SIGNIFICANCE

May lead to significant loss of vision.

DIFFERENTIAL DIAGNOSIS

Propionibacterium acnes endophthalmitis; retained cortex.

SEE ALSO

Intraocular lenses.

MANAGEMENT

The main indication for a posterior capsulotomy is a reduction in visual acuity attributable to the PCO; other indications include glare and impaired visualization of the fundus, especially in a diabetic patient requiring retinal laser.

Laser surgery

A Nd:YAG laser posterior capsulotomy is routinely performed for visually significant PCO. The exact style of capsulotomy depends upon both surgeon choice and the specific IOL. In general, a central cruciate capsulotomy is performed with PMMA IOLs, and a central horseshoe-shaped capsulotomy is performed with foldable IOLs. In both, the aim is to achieve at least a 3-mm central clear area.

Complications of this procedure are rare, but include cystoid macular edema, retinal detachment, an early postoperative pressure spike, and posterior dislocation of the IOL (especially with anterior

capsulophimosis and plate haptic IOLs). All these complications are thought to be more common if the capsulotomy is performed early following surgery. As such, most clinicians prefer to wait 3 to 6 months post-surgery before performing a capsulotomy. Pitting of the IOL from individual laser shots is very common, but this is extremely rarely visually significant. Since the introduction of the Nd:YAG laser, surgical excision of a fibrosed posterior capsule is almost never performed.

Therapeutics

Many clinicians treat the patient undergoing a laser capsulotomy with a drop of an antihypertensive agent such as brimonidine (Alphagan) immediately prior and a topical corticosteroid qid for several days following the procedure.

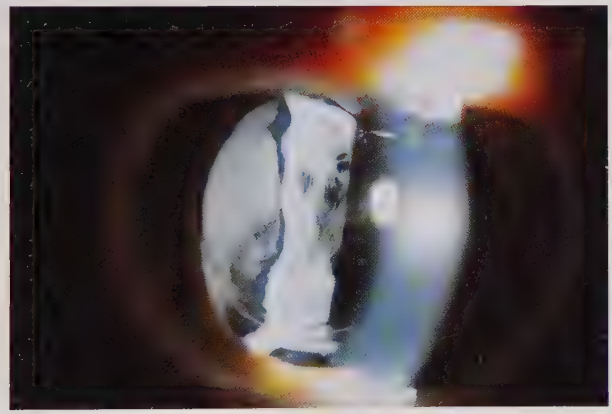


Fig. P-9a Marked PCO as seen with a dilated pupil.

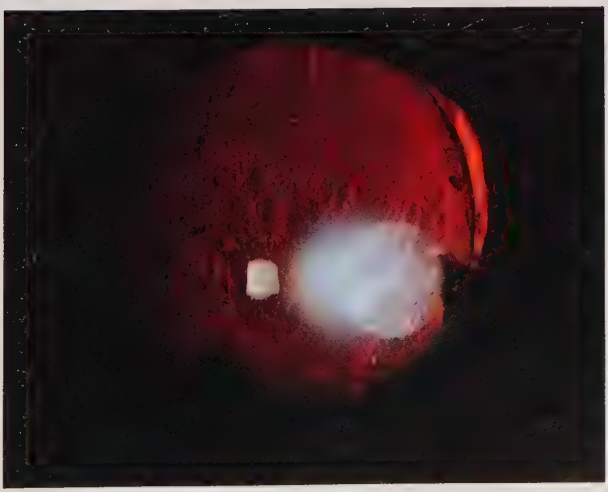


Fig. P-9b Elschnig pearls.

Posterior polymorphous dystrophy (PPD)

DESCRIPTION

Posterior polymorphous dystrophy (PPD) is one of three endothelial dystrophies, the others being congenital hereditary endothelial dystrophy (CHED) and Fuchs' endothelial dystrophy. It is frequently an incidental finding during routine slit-lamp examination, although it may rarely lead to corneal decompensation with increasing corneal edema. PPD severe enough to lead to corneal edema usually does not occur until middle age, and both eyes are usually affected. The lesions are typically bilateral although they may be quite asymmetric. They are present at birth and are usually static or only very slowly progressive.

Histopathologically, the normal endothelial cell morphology is altered, with the cells adopting a more epithelial phenotype and migrating to involve the angle. PPD is inherited as an autosomal dominant trait with a high degree of penetrance.

SYMPTOMS

It is uncommon for patients to have any symptoms associated with PPD unless there is associated glaucoma or corneal edema. The symptoms of corneal edema depend on the degree of edema and include photophobia, blurring of vision (this may show diurnal variation), and ocular discomfort, ranging from a mild foreign-body sensation to acute pain with a ruptured corneal bullae.

SIGNS

The characteristic lesions of PPD are discrete, grouped vesicular or bubble-like lesions (sometimes described as resembling frogs' spawn) involving the corneal endothelium. Lesions may be present anywhere across the endothelial surface, are usually bilateral, and may be asymmetric. Less commonly, band-like lesions that run for several millimeters across the endothelium are present. These lesions usually have linear discontinuous borders and resemble breaks in Descemet's membrane. If severe, corneal edema may also be present with either of these lesions.

Peripheral anterior synechia (PAS) may also occur in PPD and may rarely lead to glaucoma. Gonioscopy is usually required to view these lesions.

Occasionally, examination of other family members may be helpful in making the diagnosis, as they will also show signs of the disease.

INCIDENCE

Rare.

SIGNIFICANCE

Rarely, may lead to corneal decompensation and/or glaucoma.

DIFFERENTIAL DIAGNOSIS

Epithelial ingrowth; Descemet's membrane tear; Iridocorneal epitheliopathy; Fuchs' dystrophy; congenital hereditary endothelial dystrophy.

SEE ALSO

Corneal edema; Corneal graft; Corneal dystrophies, degenerations, and depositions.

MANAGEMENT

The lesions of PPD are normally an incidental finding with no treatment being required. If the lesions are severe, patients should be reviewed annually, checking for the development of both corneal edema and glaucoma.

Genetic advice

Inheritance is autosomal dominant with a high degree of penetrance.

Tear supplements

Rarely, in severe cases, tear supplements are indicated to treat ocular surface discomfort related to corneal epithelial microcystic edema.

Therapeutics

If glaucoma is present, anti-glaucoma medication is indicated. Topical carbonic

anhydrase inhibitors are best avoided as they can potentiate corneal edema. Topical hypertonic saline (5%) may be helpful in alleviating some symptoms related to corneal epithelial microcystic edema.

Surgery

Very rarely, a penetrating keratoplasty (corneal graft) is necessary if irreversible corneal edema occurs.

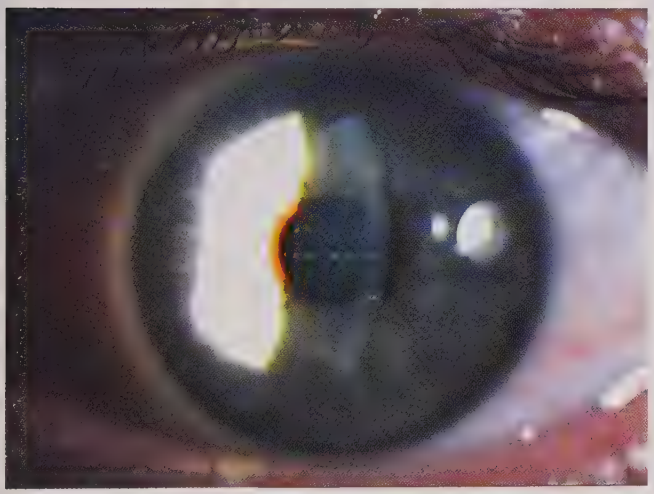


Fig. P-10a Typical vesicular endothelial lesions seen in a mild case of PPD.



Fig. P-10b Severe case of PPD.

Preseptal cellulitis

Eyelid infection

DESCRIPTION

Preseptal cellulitis is an infection of the subcutaneous eyelid tissue, anterior to the orbital septum. It presents as an acute inflammation of the eyelids and is most common in children under 5 years of age. Preseptal cellulitis is a serious condition requiring urgent systemic treatment. The infection is commonly secondary to:

- local infection of the eyelids, including hordeolum, dacryocystitis, impetigo (uncommon superficial skin infection), herpetic blepharitis, or severe conjunctivitis
- trauma or injury of the eyelids or facial area, which has become infected; this may be from an insect bite or laceration
- upper respiratory tract infection, particularly *Haemophilus influenzae* or *Streptococcus pneumoniae*.

There is a need to exclude the more serious condition of orbital cellulitis (see below, under "Signs"), which is potentially vision- or even life-threatening.

SYMPTOMS

The patient may present having noticed swelling and hyperemia of the eyelid, such



Fig. P-11a Right preseptal cellulitis.



Fig. P-11b Axial CT scan of preseptal cellulitis, inferior view, showing tissue swelling anterior to the orbital septum.



that the eyelid is swollen shut and painful. There may be associated fever, headache, and sinusitis, and there may be a history of recent upper respiratory tract infection. The condition is usually unilateral.

SIGNS

There is conjunctival injection, pain, redness, and swelling of the eyelid, particularly the superior eyelid. If the patient is a child under 5 years of age and has no sign of injury or trauma, frequently the cause is an infection with *H. influenzae* or *S. pneumoniae*. *H. influenzae* is classically associated with a sharply-demarcated deep purple discoloration of the eyelid and periorbital area.

An important differential diagnosis is from the much more serious orbital cellulitis. Preseptal cellulitis does not show proptosis, and there is no effect on eye movements or pupillary reactions. Visual acuity maybe slightly affected. Orbital cellulitis does present with these features and a greater effect on visual acuity.

INCIDENCE

Uncommon to rare (approximately 1/1000 to 1/10,000).

SIGNIFICANCE

Preseptal cellulitis is not a superficial or localized infection, but rather a wider spread infection of deeper tissues. Furthermore, it achieves additional significance because of the potential for the infection to spread further via the blood and potentially for it to become a secondary meningitis in children.

DIFFERENTIAL DIAGNOSIS

Other causes of acute eyelid inflammation: Hordeolum; Acute allergic blepharoconjunctivitis; Glaucoma – acute angle closure; Eyelid trauma (hematoma). Other conditions not covered in this series: orbital cellulitis; insect bite; eyelid abscess.

MANAGEMENT

Oral medication

Oral antibiotics are required to treat the tissue infection. The selection of antibiotic is somewhat empirical because the causative organism is often not known when treatment is commenced. Mild to moderate infections usually respond to a penicillinase-resistant penicillin, such as dicloxacillin, or a cephalosporin, such as cephalexin, continued for 10 days.

Topical therapeutics

If there is any sign of external infection or conjunctivitis, the oral antibiotics can be complemented with topical antibiotic ointment tid (e.g. ciprofloxacin).

Urgent

Hospitalization is not usually required; however, in some instances it may be indicated. These may include if the patient is a child under 5 years or if it appears the patient may be non-compliant with home-based treatment or if there is not a positive response to treatment within a few days. The suspicion of *H. influenzae* is also significant because of the risk of rapid progression of secondary meningitis.

In such patients, the intravenous administration of an antibiotic may be indicated, such as a third-generation cephalosporin (e.g. ceftriaxone).

Microbiological cultures

Microbiological cultures are indicated in high-risk patients, as described above. Cultures from the conjunctiva of both eyes, as well as blood cultures, may assist in diagnosis.

CT scan

If there is any concern that the child may have orbital cellulitis, an emergency CT scan of the orbit should be organized.

Review

Review should be daily until there are clear signs of improvement, then every few days until full resolution has occurred.

Pseudoexfoliation of the lens capsule (PXF)

Glaucoma

DESCRIPTION

Pseudoexfoliation (PXF) refers to a gray-white, flaky material observed on the anterior surface of the crystalline lens. Because it appears as if there has been shedding of epithelial cells from the lens capsule, it is termed "pseudoexfoliation." However, the material is believed to be similar to amyloid and may be synthesized at the lens capsule, iris, and ciliary body.

The pseudoexfoliative material is deposited on all surfaces throughout the anterior chamber, including the anterior lens capsule. The characteristic appearance on the anterior lens capsule arises because of movement of the pupil (constriction and dilation), which scrapes the material off the mid-zone of the lens, leaving a clear zone.

PXF is of concern because the material is also deposited in the trabecular meshwork in the anterior angle. In some patients, this leads to a secondary trabecular block with raised intraocular pressure (IOP) and the development of glaucoma. PXF also leads to weakening of the zonules and capsule and can result in lens instability (phacodonesis, pseudophacodonesis) and subluxation or dislocation of the lens or an IOL, and predispose to capsular instability at the time of cataract surgery.

SYMPTOMS

Usually asymptomatic.

SIGNS

PXF can only be readily observed with pupillary mydriasis and slit-lamp examination. White, flaky material is seen on the lens capsule and pupillary border. The pseudoexfoliative material is located in a central disc and in a peripheral circular band, with a clear zone in between. It may be present bilaterally or unilaterally. There may be an iris transillumination defect, due to loss of iris pigment. Pigment dispersion may also be present with signs such as a Krukenburg spindle, fine pigment granules on the iris, and on the anterior lens surface. Gonioscopy commonly shows trabecular hyperpigmentation and a flaky white appearance on the trabeculum.

There may be signs of glaucoma, such as optic nerve cupping, glaucomatous visual field loss, and an increased IOP. There is an increased likelihood of an IOP spike after mydriasis due to the compromised trabecular meshwork. The pupil often dilates poorly.

INCIDENCE

Uncommon to rare (approximately 1/1000 to 1/10,000). More common in Scandinavia, although the reason why is unclear.

SIGNIFICANCE

PXF is of concern because of the risk of a secondary, trabecular block, open-angle glaucoma. There is a risk of approximately 40% of developing glaucoma within 10 years if a patient has PXF. It may also be associated with weakening of the zonules and capsule, which are of significance to cataract surgery.

DIFFERENTIAL DIAGNOSIS

Glaucoma – acute angle closure; Endophthalmitis; Cataract; Iritis.

MANAGEMENT

Ocular investigations

Visual field testing and gonioscopy, to determine other risk factors and whether glaucomatous field defects are present.

Therapeutics

PXF of the lens capsule does not in itself require therapy; however, therapy is required if there are also signs of glaucoma. Medical treatment is the same as for primary open-angle glaucoma. Should pressure be initially high, then some immediate treatment guidelines are given under "Glaucoma – acute angle closure."

Laser surgery

Argon laser trabeculoplasty can be initially effective, more so than in primary open-angle glaucoma. However, the effectiveness of the treatment may diminish after a number of years. Occasionally, surgical intervention with a trabeculectomy is required to control raised IOP.



Review

Many patients with PXF do not have glaucomatous signs. Such patients should be reviewed every 6 to 12 months, depending upon the presence of other risk factors such as a family history of glaucoma.



Fig. P-12a Central disc of pseudoexfoliative material.



Fig. P-12b Pseudoexfoliative material at the pupil margin.

Pterygium

DESCRIPTION

Pterygia are a fibrovascular growth involving both the bulbar conjunctiva (the tail) and cornea (the head of the pterygium). Available epidemiological evidence suggests that they develop from excessive ultraviolet light exposure, with damage to the conjunctival and limbal epithelium. Sidelight (albedo light), internally reflected through the cornea and focused at the limbus, appears to be of particular importance in this process. Pterygia are usually bilateral, although they may be quite asymmetric, are interpalpebral and usually medial. They may (rarely) only be present laterally, or may be present medially and laterally. If present both medially and laterally and touching across the central cornea, they are termed "kissing pterygia." Recurrence of the pterygia following surgical removal is common, especially in darkly pigmented races. Recurrent pterygia often appear slightly different to primary pterygia, being less well centered at the limbus and more distorted.

SYMPTOMS

Depending on the size and height of the lesion, ocular surface irritation with dryness and a foreign-body sensation may occur. If large enough or fibrotic enough, blurring of vision can occur from the induction of irregular astigmatism. Rarely, and usually with recurrent pterygia, positional diplopia may occur.

SIGNS

The pterygium commences with conjunctival changes, with scarring, thickening, and distortion, followed by extension of this process onto the limbus, with subsequent further extension onto the cornea. The leading edge of the pterygium may be preceded by small, round, epithelial lesions resembling Salzmann degeneration; these are termed Fuchs' spots and are seen as a sign of active growth. When in an active growing phase the pterygium is also fairly thickened, obscuring the underlying sclera and cornea. An avascular, slightly opaque area of epithelium (termed a "cap") also frequently precedes the head of an active pterygium. A corneal epithelial iron line (termed a Stocker line) may occur just central to the leading edge of the lesion. Keratometry or corneal mapping may reveal central corneal distortion.

INCIDENCE

Incidence may be as high as 20–30%, depending on UV light exposure.

SIGNIFICANCE

Usually only causes irritation, redness, and cosmetic concern, although the condition can lead to reduced vision or even blindness in very advanced cases. Pterygium has also been associated with an increased risk of developing age-related macular degeneration.

DIFFERENTIAL DIAGNOSIS

Pannus and pseudopterygia; Conjunctival intraepithelial neoplasia (CIN).

SEE ALSO

Pingueculum; Scleritis – thinning and necrosis; Iron lines.

MANAGEMENT

Patients with pterygia should be advised to wear sunglasses with good side-protection when outside in high UV conditions, both to limit further development of the lesion and to prevent it from becoming sore and inflamed.

Tear supplements

Ocular surface lubrication may be useful in alleviating some symptoms of ocular surface irritation.

Therapeutics

Acute inflammation of the pterygium can be treated with topical corticosteroids such as fluorometholone or topical non-steroidal anti-inflammatory agents such as ketorolac. Topical thiotepa is also occasionally used as an adjunctive treatment to reduce the rate of recurrence.



Surgery

Usual indications for offering surgical excision are:

- induced corneal astigmatism leading to decreased vision
- induced corneal astigmatism prior to cataract surgery (intraocular lens power calculations may be significantly compromised secondary to induced astigmatism and flattening of the overall keratometry)
- chronic ocular surface irritation unrelieved by the moderate use of tear supplements
- extensive pterygium threatening the central cornea
- positional diplopia attributable to the tethering action of the pterygium (this is rare)
- cosmetic concern.

Simple excision of the pterygium leaving the exposed sclera uncovered (bare scleral resection) leads to a high rate of recurrence of up to 70%. As such, adjunctive treatments to reduce the rate of recurrence have been developed. Previously the application of beta-radiation postoperatively (often in three fractionated doses) was the most commonly used adjunctive treatment. Mitomycin C, applied either at the time of surgery or postoperatively, was also previously used as an adjunctive measure. While both of these agents were effective, both occasionally lead to significant postoperative complications, including scleral necrosis and delayed epithelial healing. They have largely been replaced with surgical measures aimed at preventing recurrences. The myriad of surgical techniques employed to reduce recurrences include sliding conjunctival grafts, amniotic membrane transplants, and lamellar keratoplasty; an autoconjunctival graft is now the most popular. With this technique, a free conjunctival graft is taken from the superior bulbar conjunctiva of the same eye and sutured, or more rarely glued, in place over the conjunctival defect resulting from pterygium excision. The degree of subconjunctival fibrous tissue excision and the size of the autograft have both gradually increased as surgeons have realized their importance in reducing the risk of recurrence. Using this method, recurrence rates of as low as 0.5% for primary and 1% for recurrent pterygia have been reported.

Following removal, the corneal curvature gradually improves, often returning to normal over several months. For this reason, either a spectacle update or keratometry for IOL calculation for cataract surgery is deferred for 3 months following removal of the pterygium.

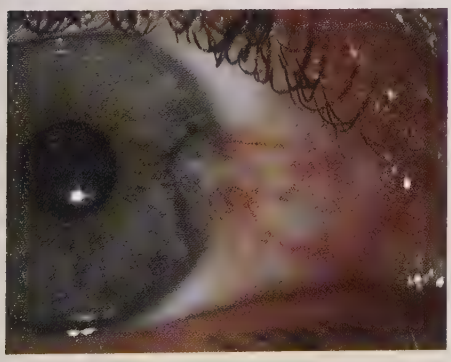


Fig. P-13a Moderate-sized medial pterygium.

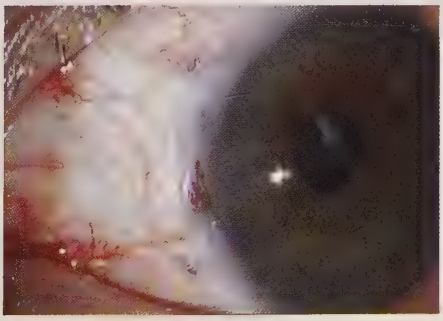


Fig. P-13b Healing conjunctival autograft with two limbal sutures still in place 1 month following pterygium surgery.

P Ptosis and pseudoptosis

DESCRIPTION

Ptosis is an abnormally low position of the upper eyelid, usually such that part of the pupil is covered. A moderate ptosis has the lid margin covering the superior pupil margin, and a severe ptosis has at least half of the pupillary area covered.

Possible causes of ptosis are:

- congenital ptosis – a developmental dystrophy of the levator muscle
- age-related (involutional) ptosis – from degeneration in the levator aponeurosis, the superior extension of the levator muscle
- post-surgical or traumatic ptosis – from a disinsertion of the levator aponeurosis
- paralytic – Horner's syndrome, third nerve palsy due to an intracranial mass, botulinum toxin
- myogenic, e.g. myasthenia gravis (MG), congenital progressive eyelid ophthalmoplegia (CPEO)
- rigid contact lens wear, long-term.

Pseudoptosis is caused by factors other than eyelid levator function such as:

- artificial eye or microphthalmos
- excessive skin on the brow or eyelids (dermatochalasis)
- large eyelid tumor, such as a neurofibroma, or eyelid edema.

SYMPTOMS

Patients will usually be aware of their droopy eyelids. In moderate to severe cases, they may have brow-ache from continual use of their frontalis (brow) muscle to keep their eyes open or complain of a restriction in their superior visual field. An involutional ptosis tends to worsen during the day.

SIGNS

Eyelid margin to pupil reflex distance is measured by shining a pen torch at the patient's eye.

- Normal margin to reflex distance is 4–4.5 mm.
- Mild ptosis is 4 mm.
- Moderate is 3 mm.
- Severe ptosis is 2 mm or less.

Levator muscle action is measured from the change in lid position between extreme upgaze and extreme downgaze. Negate any contribution of the frontalis muscle by pressing against the patient's brow.

- Normal upper eyelid excursion is 15 mm or more.
- Good is 12–15 mm.
- Fair is 5–11 mm.
- Poor excursion is 4 mm or less.

Congenital ptosis frequently has an absence of the upper eyelid crease whereas, in aponeurotic ptosis, the upper lid crease may be raised. A compensatory elevation of the chin is often present in bilateral congenital ptosis, whereas acquired ptosis may show compensatory elevation of the brow.

INCIDENCE

Some degree of unilateral or bilateral ptosis is common (approximately 1/100) in older age groups. Other causes are less common.

SIGNIFICANCE

Mild ptosis may be just a cosmetic concern. However, moderate to severe ptosis causes a substantial reduction in the superior visual field. The patient may be vulnerable to head trauma, since low positioned objects or doorways may not be seen.

DIFFERENTIAL DIAGNOSIS

Floppy eyelid syndrome (blepharospasm); Preseptal cellulitis; Dermatochalasis.

MANAGEMENT

Ocular investigations

If Horner's syndrome is suspected, a test with bilateral instillation of 2.5% phenylephrine should show the affected lid elevating due to denervation hypersensitivity. Testing of ocular excursions enables diagnosis of a third nerve palsy. In myasthenia gravis, fatigue may be increased, as evidenced by lid drooping during a 30-second test period of looking in superior gaze. A high index of



Fig. P-14a Left ptosis.

suspicion is needed to detect a patient with this potentially fatal disease.

Review

In mild-to-moderate cases of ptosis, where surgery is not yet required, review may be appropriate at 6- to 12-month intervals.

Therapeutic contact lens

A scleral lens with a partial shelf may be used as a "crutch" for the eyelid. In this way, the patient can still blink as required, but the lens provides some support between blinks. A spectacle ptosis prop is also possible but does not provide the protection of the ocular surface against desiccation.

Surgery

A levator resection is the most common form of treatment for moderate to severe ptosis. This is appropriate as long as some residual levator function has been demonstrated (see "Signs," above). A shortening of the Muller muscle may assist a mild ptosis. The superior eyelid may be attached to the frontalis muscle if there is no residual levator function. The caution with surgery is in avoiding lagophthalmos or exposure keratitis, should the eyelid be lifted too far.

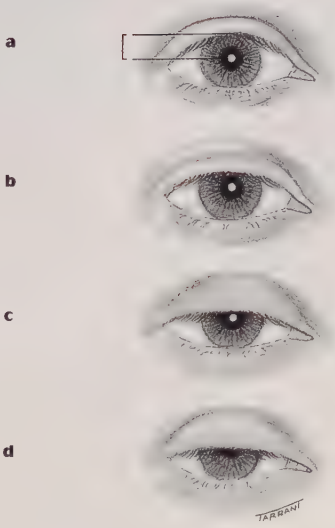


Fig. P-14b Grading of ptosis using the margin to reflex distance: (a) normal, (b) mild, (c) moderate, and (d) severe.

Punctal stenosis

Lacrimal punctal atresia, punctal agenesis

DESCRIPTION

A single lacrimal punctum is located medially in both the superior and the inferior eyelid margins. Progressive constriction of the puncta is called stenosis, whereas congenital absence is termed atresia or agenesis. Punctal stenosis prevents normal lacrimal drainage, leading to epiphora.

The many possible causes of punctal occlusion include:

- idiopathic, primarily age related – the most common cause
- post-infective scarring – herpes simplex infection, trachoma
- cicatricial scarring – ocular cicatricial pemphigoid, Stevens–Johnson syndrome, chemical burn
- medicamentosa – e.g. patients on glaucoma therapy for a long period can also develop conjunctival cicatrization
- iatrogenic – the puncta may have been sealed via cautery or laser as a treatment for aqueous-deficiency dry eye
- secondary stenosis – e.g. from ectropion or punctal eversion
- congenital (agenesis, atresia, imperforate), which may be unilateral or bilateral
- mechanical – obstruction by foreign matter, lashes, or dacryoliths (calcium salts).

SYMPTOMS

Epiphora, the severity of which may be gauged by the frequency of occurrence. The condition may be irritating, because of the frequent need to dry the eye, or lead to blurring of vision due to an excessive tear meniscus. Symptoms may be bilateral or unilateral, and exacerbated by hot or cold winds.

SIGNS

Examination of the puncta is the primary assessment. Normally the punctal orifices are 0.5–2.0 mm in diameter and clearly visible to the examiner on eyelid eversion, either with a focal light or with the slit lamp. Punctal stenosis is present if the puncta are smaller than this, and secondary stenosis present if the puncta are not communicating with the tear meniscus.

The tear film drainage function may be tested using fluorescein dye. The fluorescein disappearance test shows abnormal lacrimal drainage if a significant amount of fluorescein dye is still present in the marginal tear strip 2 minutes after installation. A simplified form of the Jones test is to look for the appearance of fluorescein in the nose, by getting the patient to blow their nose with a tissue after 5 minutes, then examining the tissue under cobalt blue light.

A lacrimal lavage may also be used to confirm patency of the lacrimal drainage system. Other possible specialized tests include dacryocystography or lacrimal scintillography.

INCIDENCE

Common (approximately 1/100) in older age groups.

SIGNIFICANCE

The condition is significant because epiphora will occur without normal lacrimal drainage, causing intermittent or continual irritation to the patient.

DIFFERENTIAL DIAGNOSIS

Canaliculitis; Dacryocystitis; Nasolacrimal duct obstruction.

SEE ALSO

Epiphora; Ocular cicatricial pemphigoid; Stevens–Johnson syndrome; Chemical burns.

MANAGEMENT

Consulting room procedures

Initial treatment is dilatation of the punctum with a fine-tipped punctal dilator (e.g. Reudemann) and using punctal dilators of progressive diameters. Anesthesia of the punctum prior to the procedure can be achieved using a cotton-tipped applicator soaked with

proparacaine. If successful, a lacrimal lavage can then be used to confirm patency of the canalicular system.

Incisional surgery

A simple incision of the posterior wall of the vertical part of the canaliculi is appropriate if testing suggests that the canaliculus is patent beyond the stenosis at the eyelid surface. If the canaliculus is compromised, more invasive surgery may be required, such as insertion of a Lester–Jones tube.

Laser surgery

Laser punctoplasty is suited to more superficial stenosis, such as in medicamentosa, where there is conjunctival overgrowth or superficial scarring.

Review

The punctal dilation procedure may only be a temporary cure, so the patient should be asked to re-present after a few weeks if symptoms persist. Periodic repetition of the procedure may be beneficial if the stenosis is only mild. For moderate-to-severe stenosis, surgery may be the only permanent solution; alternatively, a review may be indicated at 6 to 12 months.

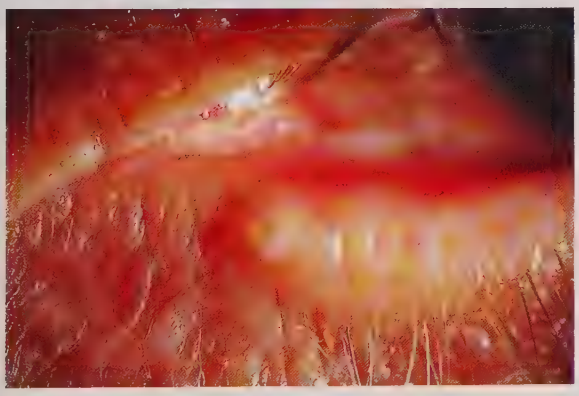


Fig. P-15a Punctal stenosis associated with lid eversion.



Fig. P-15b Lacrimal lavage.

Pyogenic granuloma

Blood blister

DESCRIPTION

A pyogenic granuloma is a proliferative fibrovascular tissue reaction to a break in the epithelial surface, with exposure of deeper tissue. It typically occurs on the conjunctiva following superficial trauma, with exposure of episclera and Tenon's capsule, but may also occur on the tarsal conjunctiva following incision of a chalazion. Other common causes include conjunctival lacerations and following conjunctival surgery, such as pterygium excision with application of a conjunctival autograft. A pyogenic granuloma may also very rarely occur on the cornea, either spontaneously or following trauma. Histopathologically, it is composed of granulation tissue: fibroblasts, capillaries, lymphocytes, and scattered plasma cells. The term "pyogenic granuloma" is a misnomer, as the lesion is neither pyogenic (in that it does not contain pus) nor a granuloma.

SYMPTOMS

Pyogenic granulomas lead to significant photophobia, increased lacrimation, an ocular surface foreign-body sensation, and, frequently, upper lid swelling with a slight reactive ptosis.

SIGNS

The lesion is often pedunculated and appears as an elevated, mobile, amelanotic, fleshy, pink-red mass. It has an uneven velvety surface and extensive superficial fine vascularization. Typically, they are between 2 mm and several millimeters in diameter.

INCIDENCE

Conjunctival pyogenic granulomas may uncommonly occur following trauma, but rarely occur spontaneously. Corneal pyogenic granulomas are very rare.

SIGNIFICANCE

Conjunctival lesions lead to surface symptoms but do not lead to a loss of vision. Corneal lesions may rarely involve the central cornea and lead to corneal scarring and loss of vision.

DIFFERENTIAL DIAGNOSIS

Phlycten; Conjunctival intraepithelial neoplasia; amelanotic melanoma.

SEE ALSO

Pterygium; corneal and conjunctival lacerations; Skin tumors – overview.

MANAGEMENT

Therapeutics

Although the lesion is usually described as being resistant to topical corticosteroids, these can frequently lead to some resolution of the lesion if used frequently and for long enough. A strong steroid such as prednisolone acetate qid for 2 weeks will usually lead to some resolution.

Surgery

Simple excision of the lesion is the most common form of treatment. This can usually be performed on the slit lamp with topical anesthesia, as the lesion itself lacks a nerve supply and as such is relatively insensitive. Due to the extensive blood supply, they do bleed following excision; therefore, application of an antibiotic ointment and double padding is appropriate.

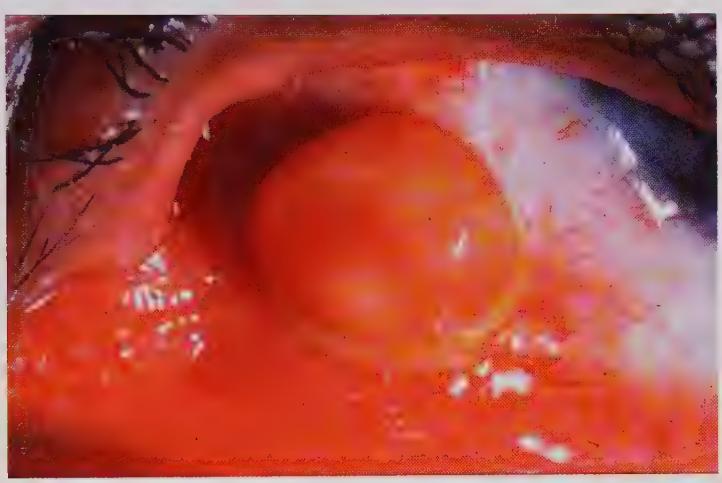


Fig. P-16a Conjunctival pyogenic granuloma following excision of a pterygium.

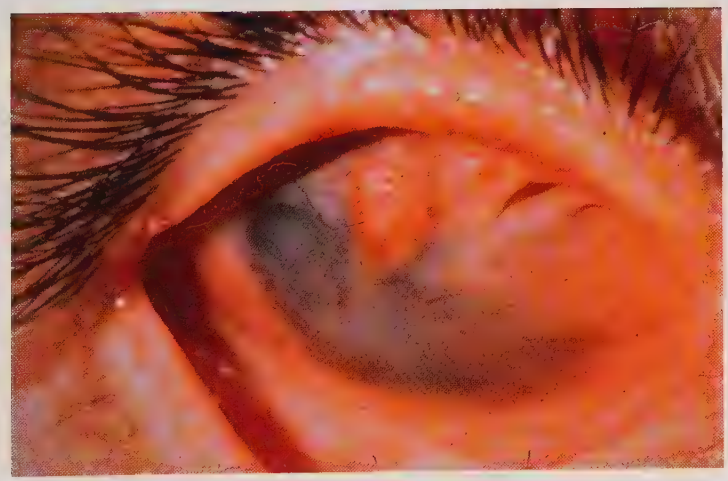


Fig. P-16b Corneal pyogenic granuloma over a pseudopterygium secondary to a severe alkali burn.

Recurrent corneal erosion syndrome (RCES)

DESCRIPTION

Instability of the attachment of the basal corneal epithelium to the underlying stroma through its normal adhesion structures results in a collection of signs and symptoms known as recurrent corneal erosion syndrome (RCES). It is a relatively common condition and may be classified as:

- dystrophic, when it occurs associated with an anterior corneal dystrophy (dystrophic RCES)
- non-dystrophic, when it occurs following superficial corneal trauma (post-traumatic RCES)
- unrelated to any pre-existing corneal disease or past history of trauma (idiopathic RCES).

Corneal dystrophies commonly associated with RCES include epithelial basement membrane dystrophy (Cogan's microcystic or map-dot-fingerprint dystrophy), Reis–Bückler's, lattice, and granular dystrophies. It may also be associated with posterior blepharitis or the rare, inherited, pemphigoid-like condition epidermolysis bullosa. Rarely, an epithelial defect caused by RCES may be complicated by the development of microbial keratitis.

SYMPTOMS

RCES causes an acute, sharp, ocular surface pain. If the epithelium lifts without ulcerating, the pain usually lasts seconds to minutes (microerosions) whereas, if the epithelium ulcerates, the pain and discomfort typically last hours to days (macroerosions). Symptoms often either occur on waking or can wake the patient in the middle of the night.

Patients often describe the initial sensation as being as if the eye is stuck to the eyelid. Attacks often occur in clusters of microerosions over several nights and will end in either resolution of the attacks for several nights or a macroerosion. Attacks can be precipitated by an alteration in normal sleeping patterns as occurs with travel and excessive alcohol intake.

During an attack, the vision is usually blurred, there is increased lacrimation, and upper lid swelling may occur. The pain is made worse with eye movement and blinking.

SIGNS

The area of epithelial disturbance is usually inferior but not involving the limbus. With a macroerosion, there is an area of ulceration, similar to that seen following an abrasion, with the adjacent epithelium not usually attached to the underlying stroma. After the ulcer has healed or with microerosions, microcysts can often be seen in the area of affected epithelium. Microerosions may also lead to an area of negative fluorescein staining, indicating that the epithelium is slightly elevated. Signs of an underlying anterior dystrophy or posterior blepharitis should also be sought in both eyes.

INCIDENCE

Common, usually affects people in their 30s to 50s.

SIGNIFICANCE

Leads to severe pain; vision may be blurred for days to a week or two following a macroerosion.

DIFFERENTIAL DIAGNOSIS

Meesmann's dystrophy; Dry eye syndromes.

SEE ALSO

Epithelial basement membrane dystrophy; Lattice dystrophy; Granular dystrophy; Blepharitis – posterior.

MANAGEMENT

Management falls into two categories: acute management of an ulcer and prevention of further attacks.

Acute management

Acute management of an ulcer complicating RCES is debridement of any ragged free-floating epithelium along the edge of the ulcer, followed by pain management with rest, ice packs, mydriasis, and analgesia. A broad-spectrum antibiotic ointment is administered qid until the epithelium is healed. Many clinicians also apply a pressure patch if the defect is

large. A therapeutic contact lens may be applied for several days to reduce pain and facilitate healing. However, it should be used in conjunction with a topical broad-spectrum antibiotic to reduce the risk of microbial keratitis.

Preventative measures

TEAR SUPPLEMENTS (OCULAR LUBRICANTS)

Topical lubricants, specifically with the nightly use of a lubricant ointment, can reduce the risk of an attack and lead to the resolution of symptoms. The ointment is usually used for 6 months after the last attack.

Therapeutics

Especially if there is associated posterior blepharitis, a course of oral doxycycline coupled with a weak topical corticosteroid such as fluorometholone may not only treat the blepharitis but also reduce the risk of further erosions.

Dosing: doxycycline 100 mg PO daily for 28 days followed by 50 mg PO daily for 50 days. Commenced at the same time G. fluorometholone 0.1% bd for 3 months.

Surgery

Several surgical interventions have been employed to stabilize the epithelium and prevent further attacks. The easiest technique is simple debridement of the affected area under topical anesthesia. Superficial micropuncture with a bent needle has also been tried and appears to be relatively effective. Several studies have also reported success with debridement coupled with alcohol delamination. There is a similar success rate of around 30–50% for debridement alone and micropuncture, slightly higher for debridement coupled with alcohol delamination.

Several studies have also demonstrated a superficial phototherapeutic keratectomy (PTK) (8–10 microns deep) to be about 80% successful. If successful, late recurrences are uncommon. Treatment failures tend to occur quickly, within the first few weeks to months following treatment, and can be treated with a repeat PTK with a high rate of success.

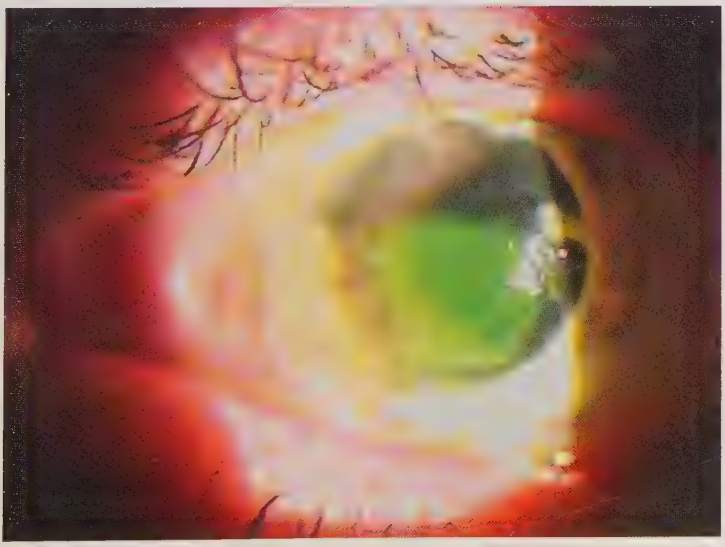


Fig. R-1 Typical large inferior corneal epithelial defect seen in RCES. Note the associated gross fluorescein leakage under the adjacent corneal epithelium.

R Refractive surgery signs – LASIK

DESCRIPTION

Corneal refractive surgery has evolved from radial and astigmatic keratotomy (RK and AK) through to current techniques, including photorefractive and photoastigmatic keratectomy (PRK and PARK), laser in situ keratomileusis (LASIK), laser epithelial keratomileusis (LASEK), and laser thermal keratoplasty (LTK). LASIK has now become easily the most common purely refractive surgery, with over one million procedures performed worldwide each year.

LASIK is an amalgam of two previous procedures: PRK and automated lamellar keratoplasty (ALK), with the excimer laser ablation being performed under a partially hinged, central, lamellar flap of corneal tissue approximately 160 microns thick created by a manual microkeratome or by a femtosecond YAG laser.

SYMPTOMS

If the central curvature becomes irregular, is too flattened or steepened, or if the treatment zone is too small, this can lead to a loss of best spectacle-corrected visual acuity (BCVA) or the induction of halos, especially around lights at night.

NORMAL POSTOPERATIVE SIGNS

The edge of the hinged incision is seen as a faint circumferential line in the anterior stroma with a diameter of between 8 and 9 mm. The line is discontinuous at the hinge, usually placed nasally or superiorly. Some interface debris may also be seen at the junction between the flap and the deeper stroma. The most commonly seen debris are small pieces of lint and meibomian secretions; these appear as small globular opacities resembling cysts. Small, faint, central, criss-cross lines (termed stria) may also be seen on close slit-lamp inspection of the flap with high magnification. These are common, especially with higher corrections (greater than 5D of myopia) and usually cause no problems. If marked, they may cause a reduction in the BCVA.

COMPLICATIONS

The following are some of the more common or important complications, and when they occur.

- Intraoperative: complete, displaced, buttonholed flaps (uncommon)
- Early postoperative
  - —Flap dislocation: rarely, the flap can become dislodged and slip, particularly within 24 hours following surgery, and leads to characteristic directional folds within the flap usually extending from the hinge
  - —Microbial keratitis (rare, approximately 1/5000)

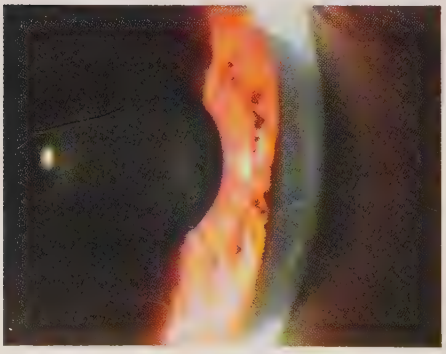


Fig. R-2a Normal postoperative appearance of a LASIK flap.

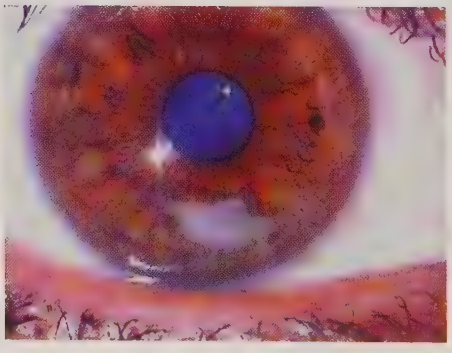


Fig. R-2b Epithelium growth under a LASIK flap.



—Extensive interface debris (rare)
—Diffuse lamellar keratitis (DLK): uncommon, and usually occurs in the first few days following surgery, with increased interface haze; it usually fully resolves but uncommonly may lead to both scarring and stromal loss
—Epithelial growth under the flap: uncommon, it appears as cystic areas extending centrally from the edge of the flap; a problem if marked, as it can lead to reduced vision and even loss of the flap

- Late postoperative
—Delayed DLK (rare)
—Flap dislocation (rare)
—Dry eye: keratoconjunctivitis sicca
—Iatrogenic keratoectasia (induced keratoconus): iatrogenic keratoectasia is rare, and may occur if the structural integrity of the central cornea is sufficiently compromised; strict exclusion criteria need to be in place to exclude at-risk patients from surgery; these include exclusion of young patients, patients with forme fruste keratoconus and thin corneas, and the preservation of a thick stromal bed

SIGNIFICANCE

Most complications of refractive surgery can lead to a loss of vision and induction of halos and glare, as detailed above.

SEE ALSO

Microbial keratitis; Refractive surgery signs – RK and PRK; Dry eye syndromes – keratoconjunctivitis sicca.

MANAGEMENT

Routine postoperative topical medication varies considerably between clinicians. However, a common regimen is a broad-spectrum antibiotic qid for 4 to 7 days and a tapering dose of a corticosteroid such as fluorometholone 0.1% for 2 weeks.

Interface debris

This rarely, if ever, causes visual problems. Meibomian secretions resolve spontaneously over weeks, while lint can still be observed years later.

Flap dislocation

The flap is relifted, hydrated with sterile water, occasionally stretched, and debrided of epithelium. This needs to be performed as early as possible (preferably within days). Occasionally the flap may even have to be sutured to remove all folds (stria).

DLK

The key to successful treatment is rapid diagnosis and treatment, preferably within a day of the onset of problems. Treatment is with intensive, potent topical steroids, usually prednisolone acetate 1% hourly day and night for several days, followed by a tapering dose over several weeks. A broad-spectrum topical antibiotic such as ciprofloxacin is usually added qid for the first few days to cover any possibility of a microbial keratitis.

Iatrogenic keratoectasia

Although often progressing rapidly over months, the treatment is similar to that for keratoconus with rigid contact lenses and occasionally a corneal graft. Prevention of progression has been reported with implantable ring segments, corneal collagen cross-linking, and a combination of both.

R Refractive surgery signs – RK and PRK

DESCRIPTION

Refractive surgery has become one of the most common ophthalmic operations worldwide, second only to cataract surgery. Refractive surgery aims to change the refractive state of the eye. Most commonly, it is used to treat myopia or myopic astigmatism. Less commonly, it is used to treat mixed astigmatism, hypermetropia, hypermetropic astigmatism, or presbyopia.

Radial keratotomy (RK) developed out of earlier techniques pioneered by Professor Sato in Japan in the 1920s to 1950s and was refined and popularized as the "Russian operation" by Dr Fyodorov in the 1970s and 1980s. It involves a series of deep, radial, peripheral corneal incisions to treat myopia. It is now rarely performed.

Photorefractive and photoastigmatic keratectomy (PRK and PARK, also termed surface laser) came into popular use in the early 1990s. It involves excimer laser ablation of the central anterior corneal stroma following removal of the corneal epithelium. It is still in common use.

SYMPTOMS

If the central curvature becomes irregular, is too flattened or steepened, or if the treatment zone is too small, this can lead to deterioration of the optics of the cornea. This may result in a loss of best spectacle-corrected visual acuity (BCVA), the induction of halos (especially around lights at night), glare sensitivity, decreased contrast sensitivity, and ghosting of vision. RK can also result in a long-term gradual hypermetropic shift from progressive central flattening of the cornea.

NORMAL POSTOPERATIVE SIGNS

RK

Multiple radial scars extending from the limbus or peripheral cornea to the mid-cornea, sparing the central cornea. These are over 90% deep and vary in number from 4 to 16. There may be associated circumferential or transverse astigmatic incisions usually at the 7-mm zone.

PRK

Some slight central reticular haze is common. This is restricted to the treatment zone and, as such, is usually 6–7 mm in diameter. The haze usually has its peak density at around 3 to 6 months following surgery and fades completely by 1 to 2 years.

COMPLICATIONS

The following are some of the more common or important complications, and when they occur.

	RK	PRK
EARLY	Microbial keratitis (very rare)	Microbial keratitis (very rare)
	Perforation into the anterior chamber (A/C)	Delayed epithelial healing
	Irregular astigmatism	Under- and over-correction Irregular astigmatism
LATE	Rupture of incisions	Stromal haze
	Microbial keratitis	Recurrent corneal erosion syndrome
	Hypermetropic shift (common)	Regression

INCIDENCE

RK was widely performed in the USA, South America, and parts of Europe, especially Russia. PRK is still performed throughout the world; however, it was largely replaced by LASIK in the mid to late 1990s. Due to increasing concerns with regard to iatrogenic keratoectasia following LASIK, there has been a recent resurgence in PRK.

SIGNIFICANCE

Most complications of refractive surgery can lead to a loss of vision, and induction of halos and glare, as detailed above.



SEE ALSO

Microbial keratitis; Refractive surgery signs – LASIK; Recurrent corneal erosion syndrome.

MANAGEMENT

Routine postoperative topical medication varies considerably between clinicians. However, a common regimen is a broad-spectrum antibiotic qid for 4 to 7 days and a tapering dose of a corticosteroid such as fluorometholone acetate 1% for 4 weeks.

Microbial keratitis

See "Bacterial keratitis" article for treatment.

Late hypermetropic shift following RK

Treatment of the refractive error with spectacles, a contact lens, or, rarely, an encircling (lasso) stromal suture.

Under- and over-correction following PRK

Treatment of any residual refractive error with repeat PRK is usually postponed until 18 to 24 months following the primary surgery.

Corneal haze following PRK

As explained above, this is usually transient and fades spontaneously over months to years. Rarely, may be treated with phototherapeutic keratectomy. Its incidence may be significantly decreased with the application of intraoperative mitomycin C.

Regression following PRK

A marked myopic shift, usually months following PRK, may be secondary to epithelial changes and frequently responds to a course of topical corticosteroids such as fluorometholone 1% qid tapering over several months.

Irregular astigmatism following PRK

This is usually treated with a rigid contact lens, although it may be amenable to treatment with further PRK. Newer excimer laser wavefront-guided ablations may be of particular benefit in the treatment of this problem.

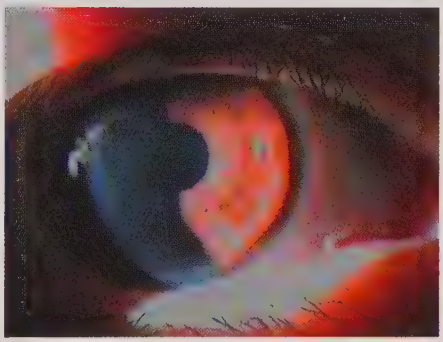


Fig. R-3a Normal postoperative appearance of RK.

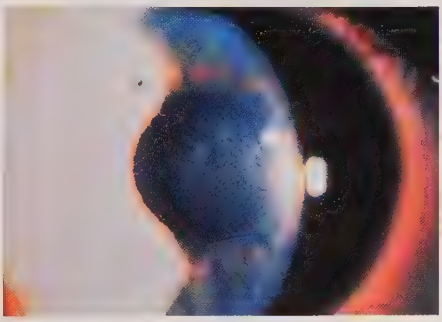


Fig. R-3b Grade 2 haze following PRK.

R

Reis–Bücklerʼs dystrophy

Honeycomb dystrophy, Thiel–Behnke dystrophy

DESCRIPTION

The corneal dystrophies are a group of progressive, usually bilateral, inherited (although non-congenital) corneal disorders with a typical age of onset of symptoms. Reis–Bückler's dystrophy affects the anterior cornea with degradation of the basal layers of the epithelium and the gradual development of subepithelial scarring and membrane formation. Irregular bands of collagen replace Bowman's layer in a honeycomb pattern. It is now known that Reis–Bückler's dystrophy, along with Avellino, lattice, and granular dystrophies, occurs secondary to a mutation in the kerato-epithelin gene *BIGH3* on the long arm of chromosome 5.

The severity of the corneal signs and symptoms in this disease can be quite varied; there are several named less severe variants of this dystrophy. A milder form of the disease said to be more common in central Europe is called Thiel–Behnke dystrophy.

SYMPTOMS

The onset of symptoms is usually in late childhood, with red, sore eyes and recurrent corneal erosion-type symptoms. Ocular surface discomfort often reduces over the years as increased scarring leads to reduced corneal sensation. Over the decades, this becomes worse, and blurring of vision increases to the point where, by about the fifth decade of life, surgical intervention becomes necessary to restore vision.

SIGNS

Although symptoms usually commence in late childhood, signs may frequently be first observed in the first decade of life. The signs are bilateral, symmetric, and central with the peripheral 1–2 mm of the peripheral cornea being uninvolved.

A reticular subepithelial "honeycomb" opacity develops over the years and is variously described as being like frosted glass, alveolar or polycystic. The densest opacities are usually in the mid-periphery. The overlying epithelium becomes irregular and, in advanced disease, the opacities may penetrate the epithelial surface. A reduction in corneal epithelial sensitivity is common.

INCIDENCE

Very rare.

SIGNIFICANCE

Leads to bilateral blurring of vision, ocular irritation, and recurrent erosions.

DIFFERENTIAL DIAGNOSIS

Granular dystrophy; Lattice dystrophy; Epithelial basement membrane dystrophy.

SEE ALSO

Macular stromal dystrophy; Recurrent corneal erosion syndrome; Corneal dystrophies, degenerations, and deposits.

MANAGEMENT

Genetic advice

This is an autosomal dominant disease with a high degree of penetrance, with symptoms becoming apparent early in life. As such, most patients present with a known family history of the disease.

Tear supplements

Ocular lubricants, with drops during the day and an ointment or gel at night, can aid in reducing both ocular surface irritation and recurrent corneal erosion symptoms.

Surgery

The most common treatment is a superficial keratectomy with removal of the epithelium and subepithelial fibrotic membrane. Rarely, a lamellar or penetrating keratoplasty is necessary to adequately clear the cornea.

Laser surgery

The use of a phototherapeutic keratectomy (PTK) with the excimer laser to remove the anterior opacities has been extensively reported. While it is effective in removing the opacity, it does lead to substantial

thinning of the cornea, with up to 100 microns of tissue commonly needing to be removed. This not only means that this treatment can only be repeated once or twice at the most, but also leads to a significant hypermetropic shift (up to 5 diopters) and may of itself lead to the development of significant corneal haze.

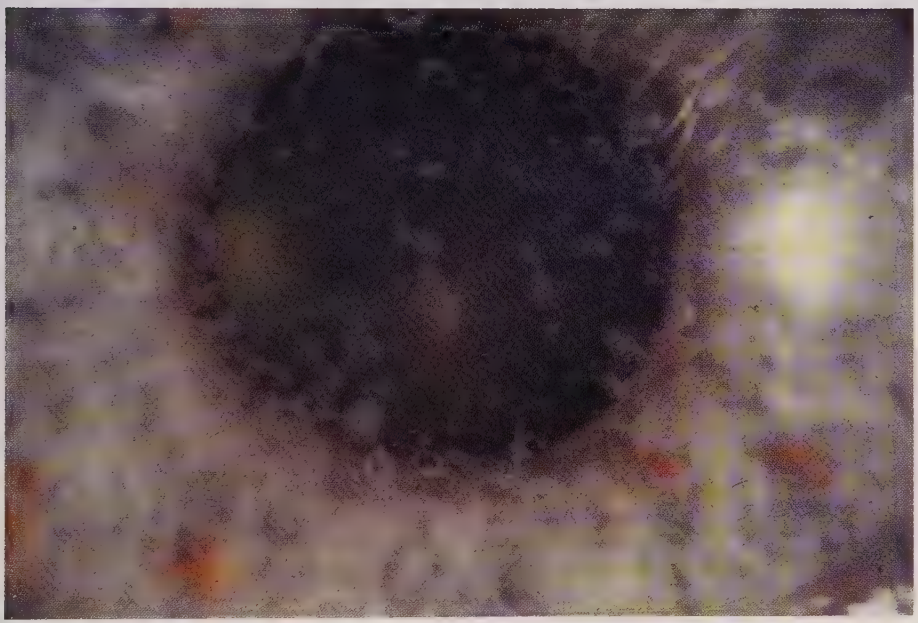


Fig. R-4 Typical central honeycomb subepithelial opacity seen with Reis–Bückler's dystrophy.

R

Retention cyst and lymphangiectasia

Conjunctival inclusion cyst

DESCRIPTION

A translucent "bubble" in the conjunctiva may arise for two reasons:

- A cyst within the conjunctiva, filled with fluid. This is termed a retention (or inclusion) cyst.
- A local dilation of the fine, thin-walled conjunctival lymphatic vessels, termed lymphangiectasia. These may be single or multiple.

These are not inflammatory, nor infectious, and seem to represent a mild irritation to the ocular surface. The conditions differ from the appearance of chemosis, in that they are smaller and more focal.

SYMPTOMS

Usually asymptomatic. There may be a mild foreign-body sensation or a cosmetic concern. Usually unilateral but may be multiple and bilateral.

SIGNS

A retention cyst appears as a raised, hemispherically shaped "bubble" on the conjunctiva. It is more commonly located in the fornices, on the palpebral conjunctiva, or at the inner canthus on the caruncle or plica. It commonly has a translucent, fluid-filled appearance, but may be white or yellow in color.

Lymphangiectasia is usually transparent, and there may be single or multiple lobulations, usually on the bulbar conjunctiva. The walls are only the dilated walls of the lymph vessels and hence are thinner and more transparent than those of an inclusion cyst.

INCIDENCE

Common (approximately 1/100).

SIGNIFICANCE

A retention cyst or lymphangiectasia is a relatively innocuous finding; it may be observed incidental to a general eye examination. Alternatively, it may be of mild cosmetic concern or cause mild discomfort.

DIFFERENTIAL DIAGNOSIS

Phlyctenulosis – conjunctival; Pingueculum; Conjunctival intraepithelial neoplasia; Conjunctivochalasis and chemosis.

MANAGEMENT

Advice

No treatment is required for this condition, unless the lesion is irritating or the patient requests it for cosmetic reasons.

Consulting room procedures

The lesion may be lanced with a 25-gauge needle (angled tangentially or away from the ocular surface). It may then be massaged through the closed eyelid to drain the fluid out. Permanent removal may require surgical excision of the whole cyst at the base. Following lancing and drainage, instillation of a single drop of a broad-spectrum topical antibiotic is appropriate, although the innate antibiotic properties of the tear film are probably adequate.

Review

Cysts may be recurrent and, therefore, the patient should be asked to re-present if the symptoms persist or recur.



Fig. R-5a Conjunctival retention cyst.

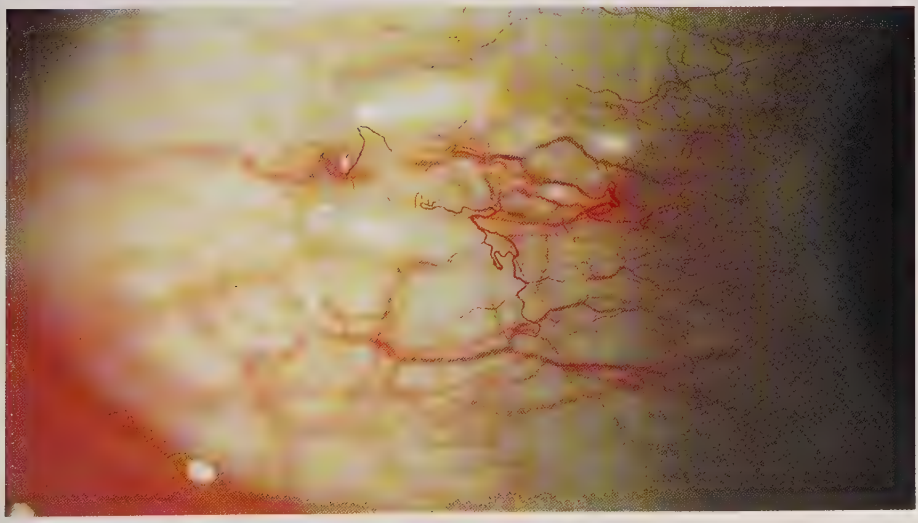


Fig. R-5b Lymphangiectasia.

Rheumatoid arthritis and the eye

DESCRIPTION

Rheumatoid arthritis (RA) is a relatively common systemic collagen vascular disorder affecting both articular cartilage and non-articular tissue. It is more common in women, but frequently follows a more aggressive course when it occurs in men. Ocular involvement is common, especially in advanced disease, usually occurring many years following the initial diagnosis. It is hoped that, with the treatment of RA increasingly being focused on disease modification rather than just suppressing articular inflammation, the incidence of ocular manifestations will decrease. The most common forms of ocular involvement are detailed here.

Dry eye

This is the most common ocular manifestation of RA and is usually secondary to a marked reduction in aqueous tear production from inflammatory damage to the acini in the lacrimal and accessory lacrimal glands.

Corneal stromal lysis

Acute loss of corneal stroma is also relatively common in advanced disease. This is often referred to as corneal "melting," and occurs either centrally with a small scalloped-out area or peripherally as a paralimbal gutter. Either may occur with active inflammation or without inflammation, when it is associated with ocular surface disease and dry eye. Perforation of the cornea is common with both forms of stromal lysis.

Episcleritis

This is usually quite mild, is non-destructive, and responds well to topical corticosteroids.

Scleritis

Anterior scleritis is a relatively uncommon manifestation of RA, and can occur either as an acute inflammatory necrotizing scleritis or slowly over years without inflammation, resulting in gradual scleral stromal loss (scleromalacia perforans). Perforation is very rare.

SYMPTOMS

The symptoms for each of these complications of RA are the same as with other causes of the conditions. Dry eye presents with typical symptoms of grittiness, dryness, and slight foreign-body sensation; corneal stromal lysis leads to photophobia, tearing, and blurred vision; episcleritis leads to redness, tearing, and slight ache; and scleritis is either asymptomatic if non-inflammatory or acutely painful if inflammatory.

SIGNS

The signs of dry eye, episcleritis, and scleritis are covered in their individual articles. Central corneal stromal lysis occurs with an epithelial defect and an underlying scalloped-out area of stromal loss. There may be an area of associated infiltration and evidence of perforation with a positive Seidel's test (covered in the article on "Perforation – corneal"). Paralimbal stromal loss usually occurs as a gutter and, when acutely progressive, is associated with an overlying epithelial defect.

INCIDENCE

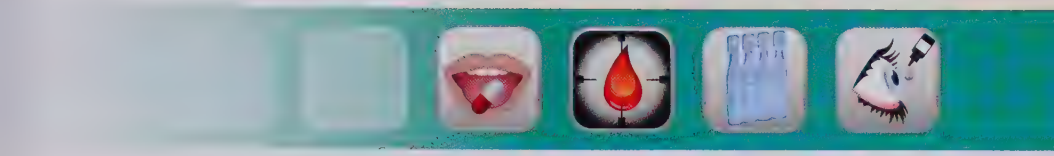
RA is a relatively common disease, and some form of ocular involvement is common. Most patients with advanced disease will develop some dry eye and, in a small subset, corneal stromal lysis will occur.

SIGNIFICANCE

Dry eye is common in advanced rheumatoid disease and, rarely, the ocular complications of RA can lead to loss of vision from ulceration or possibly corneal perforation.

DIFFERENTIAL DIAGNOSIS

Peripheral ulcerative keratitis; other causes such as Mooren's ulcer, polyarteritis nodosa; systemic lupus erythematosus; Wegener's granulomatosis.



SEE ALSO
Dry eye syndromes; Scleritis – overview; Scleritis – thinning and necrosis; Perforation – corneal.

MANAGEMENT

Adequate treatment of the underlying RA is essential. This requires monitoring of joint disease, non-articular manifestations of the disease, serum rheumatoid levels, and the erythrocyte sedimentation rate (ESR). The most commonly-used drugs to control the underlying disease process are hydroxychloroquine, non-steroidal anti-inflammatory agents, methotrexate, cyclosporin A, and the newer TNF-α antagonists.

Tear supplements

Adequate lubrication with tear supplements is essential to prevent corneal epithelial ulceration and its possible sequelae. Frequent unpreserved unit dose drops during the day and ointments at night are indicated. This may be supplemented with either silicone punctal plugs or punctal cautery.

Surgery

Ocular surface protection with severe dry eye may require a lateral tarsorrhaphy. If perforation of the cornea occurs, sealing with cyanoacrylate glue or even a lamellar or penetrating corneal graft may be required (see "Perforation – corneal").



Fig. R-6 Peripheral paralimbal gutter of stromal tissue loss in advanced rheumatoid arthritis.

Rosacea keratitis

DESCRIPTION

Acne rosacea is a relatively common inflammatory skin condition of unknown cause. It affects the face in a classic butterfly rash configuration across the cheeks and nose. While most involvement is fairly mild, it may rarely develop to a scarring and pustular stage. Patients sometimes report that alcohol, spicy foods, and even coffee intake can result in an exacerbation of the skin changes.

In about 5–10% of patients, there is significant ocular involvement. As with the skin disease, there is a wide range in the extent and severity of the disease, with posterior blepharitis, chalazion and internal hordeolum formation, keratitis, and possibly corneal perforation.possibly

SYMPTOMS

Aside from the facial rash and flush, patients describe ocular irritation, with gritty sore eyes, a slight foreign-body sensation, and photophobia. They may also complain of recurrent red eye and styes. The symptoms usually fluctuate significantly and are often chronic, lasting for years.

SIGNS

Rosacea commences with the typical facial butterfly rash. With increasing severity of the disease, the rash evolves from mild erythema with superficial telangiectasias through a papular and pustular stage to nodular disease, with the development of gross hypertrophy of the sebaceous glands of the skin of the nose. This leads to the development of a rhinophyma. Ocular rosacea is characterized by the development of posterior blepharitis with a typical purulent discharge from the meibomian gland orifices. If the patient develops corneal involvement, this may range from an inferior punctate keratopathy, consistent with active posterior blepharitis, through marginal keratitis, inferior vascularization with pannus formation, stromal inflammation, epithelial ulceration, stromal loss, and possibly corneal perforation.

INCIDENCE

While acne rosacea is common, rosacea keratitis is relatively uncommon, and severe rosacea keratitis with corneal perforation is very rare. It is more common in women and usually affects people in their 30s and 40s. The disease is more common in people of Anglo–Celtic background and may exhibit seasonal variation, being worse in harsh winter conditions.

SIGNIFICANCE

May rarely lead to significant ocular disease, even corneal perforation.

DIFFERENTIAL DIAGNOSIS

Posterior blepharitis; Herpes simplex keratitis.

SEE ALSO

Dry eye syndromes; Perforation – corneal; Marginal keratitis; Conjunctivitis – overview; Limbal stem cell deficiency.

MANAGEMENT

The skin disease is treated either topically with topical metronidazole gel or systemically with oral erythromycin or a tetracycline. Treatment of the ocular disease with an oral tetracycline obviously also treats the skin disease. When systemic antibiotic treatment is no longer required for the ocular disease, the patient can, at this stage, change over to cutaneous topical metronidazole gel for management of the skin disease.

Tear supplements

Because of the disturbance to the normal tear film lipid layer, tear film supplements may be helpful in stabilizing the tear film and improving the health of the ocular surface.

Therapeutics

A long-term, low-dose, weak topical corticosteroid, such as 1 drop of fluorometholone daily, may be quite useful in reducing ocular surface disease and keratitis associated with rosacea. As with



all topical corticosteroids used in the long term, care should be taken to assure there is no rise in intraocular pressure. If there is associated staphylococcal blepharitis, a short course of a broad-spectrum antibiotic is indicated.

Oral medication

The mainstay of treatment is a medium-term course of an oral tetracycline. Doxycycline has the advantage that it can be taken on a once-a-day basis, but does need to be taken with food to prevent gastric irritation. A typical course is 100 mg daily for 1 month, followed by 50 mg daily for 2 months. Occasionally the course needs to be extended if the condition relapses. It normally takes up to 1 month for the patient to respond to treatment. If tetracyclines are contraindicated, erythromycin can be substituted.

Surgery

Corneal perforation is very rare; its management is described under "Perforation – corneal."

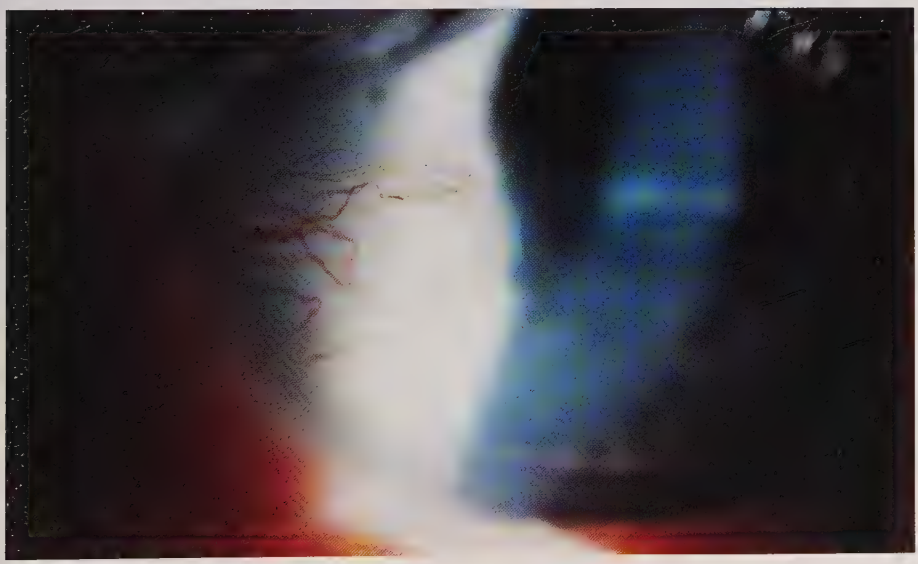


Fig. R-7 Peripheral corneal vascularization, corneal scarring and thinning in rosacea keratitis.

S

Salzmann nodular degeneration

Keloid formations

DESCRIPTION

Salzmann nodular degeneration is an uncommon anterior corneal condition usually seen secondary to chronic keratitis and often occurring in childhood. Common precipitating causes of this condition are phlyctenulosis, trachoma, vernal keratoconjunctivitis, and, classically, childhood infectious diseases such as measles and scarlet fever. It has become increasingly uncommon in developed countries, as childhood keratitis has become better controlled. A rare inherited autosomal dominant form of the disease has been reported.

Histopathologically, the epithelium may be thinned or grossly thickened with areas of both atrophy and hypertrophy; there is also marked subepithelial basement membrane scarring, with areas of dense collagen deposition (keloid) in the anterior stroma.

SYMPTOMS

The condition is usually bilateral and results in ocular surface discomfort with a foreign-body sensation, irritation, and increased lacrimation. It may also lead to photophobia and blurred vision.

SIGNS

The areas of degeneration are elevated and nodular and may be central or peripheral, singular or multiple. They occur either at the site of or along the edge of the area of previous inflammation. They are white to gray in color, and there is frequently associated corneal stromal scarring and thinning. Occasionally, they are surrounded by an iron deposition line.

INCIDENCE

Very rare.

SIGNIFICANCE

May lead to a bilateral decrease in vision and marked photophobia.

DIFFERENTIAL DIAGNOSIS

Phlycten; amelanotic melanoma; Pyogenic granuloma; Climatic droplet keratopathy; hypertrophic corneal scarring; intraepithelial corneal neoplasia; Dimmer's nummular keratitis; Marginal keratitis.

SEE ALSO

Phlyctenulosis; Vernal keratoconjunctivitis.

MANAGEMENT

Tear supplements

Ocular lubricants, especially gels and ointments, are helpful to relieve the ocular discomfort commonly seen in this condition.

Surgery

Surgical excision of the areas of degeneration with a surgical superficial keratectomy is occasionally indicated, to both reduce irritation and improve visual acuity. Occasionally a lamellar keratoplasty or even a penetrating keratoplasty is required to restore vision.

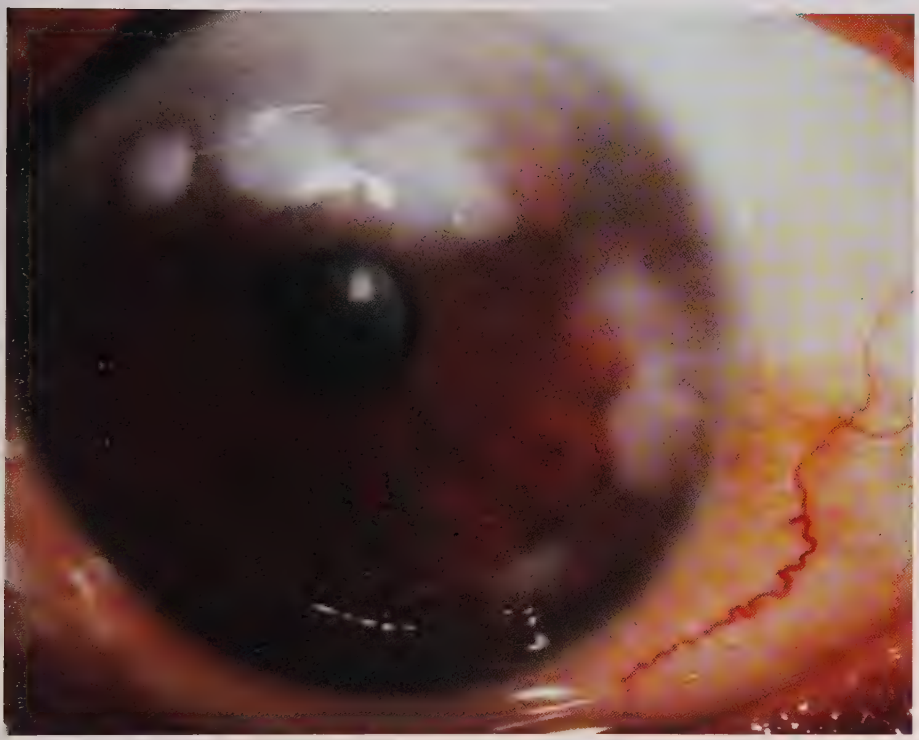


Fig. S-1 Multiple lesions of Salzmann nodular degeneration overlying areas of corneal scarring.

S

Scleritis – overview

DESCRIPTION

Scleritis is a protean and diverse group of clinical syndromes. It is much less common than episcleritis, but more commonly leads to significant ocular complications. Despite significant scleral thinning occasionally occurring, perforation is extremely rare.

Scleritis is commonly classified as below.

- Anterior
  - —Diffuse (most benign form)
  - —Nodular
  - —Necrotizing
    - – With inflammation (most severe form)
    - – Without inflammation
- Posterior

Scleritis is commonly associated with underlying systemic disease, most commonly rheumatoid arthritis, usually in elderly patients with established joint and extra-articular disease. Rheumatoid arthritis is associated particularly with necrotizing scleritis. Other connective tissue diseases associated with scleritis include Wegener's granulomatosis, polyarteritis nodosa, and systemic lupus erythematosus (SLE).

Approximately 10% of scleritis is infective with causative organisms including herpes zoster, fungi, Gram-negative bacteria, *Streptococcus pneumoniae*, and even *Acanthamoeba*. Other specific scleritis syndromes include surgically-induced necrotizing scleritis (SINS), post-keratoplasty sclerokeratitis, and post-irradiation scleral necrosis.

SYMPTOMS

Patients with anterior scleritis usually present with redness, discomfort, significant ocular ache (even boring pain), slight photophobia, and increased lacrimation. The more inflammatory the disease, the more marked the symptoms. Symptoms are usually unilateral and evolve over days.

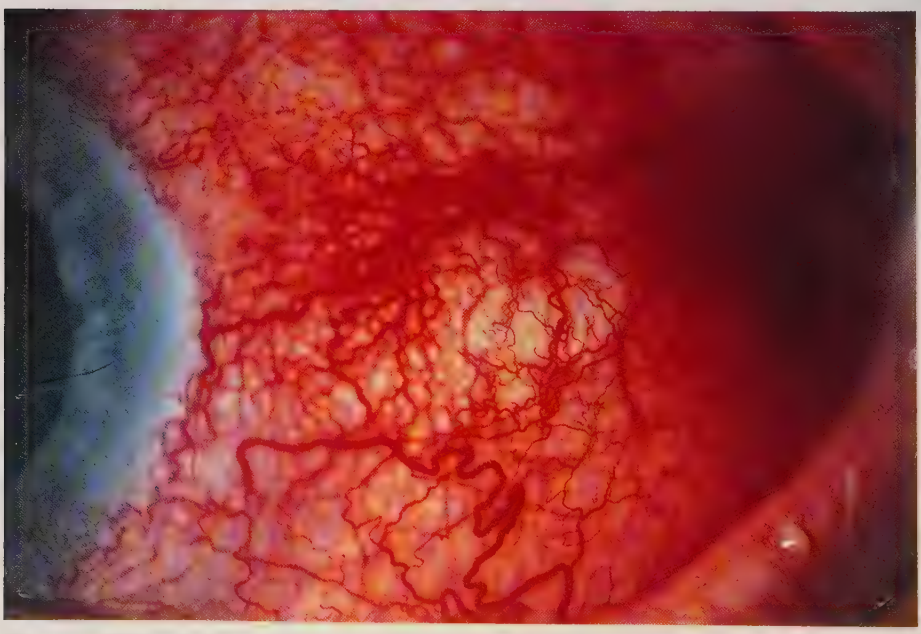
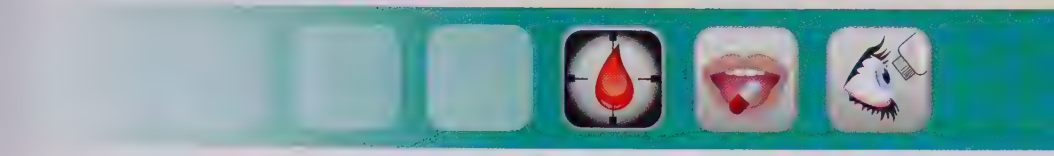


Fig. S-2 Diffuse anterior scleritis.



SIGNS

Signs common to all types of inflammatory anterior scleritis are scleral thickening, increased vascularity, distortion of the normal scleral vascular architecture, and tenderness over the affected area.

- Diffuse. It may be segmental or involve the entire anterior sclera. It is rarely associated with other significant diseases unless infective.
- Nodular. The nodules are deep red, immobile, and may be multiple. There may be associated uveitis.
- Necrotizing with inflammation. This is a very severe, though thankfully rare, form of the disease, and is frequently associated with a systemic, life-threatening vasculitis. As the disease progresses, blood vessel occlusion and drop-out is noted, followed by scleral thinning and exposure of the underlying uveal tissue. There is frequently associated intraocular inflammation.
- Necrotizing without inflammation. Non-inflammatory scleral thinning with blood vessel drop-out and exposure of underlying uveal tissue can occur over years, both in rheumatoid arthritis (scleromalacia perforans) and secondary to beta-irradiation following pterygium surgery.

Infective scleritis is usually diffuse and continuous with an old area of necrosis, corneal infection, or wound.

INCIDENCE

Rare.

SIGNIFICANCE

May be associated with significant underlying systemic disease and may lead to significant sight-threatening eye diseases.

DIFFERENTIAL DIAGNOSIS

Panophthalmitis; Episcleritis.

SEE ALSO

Rheumatoid arthritis and the eye; Herpes zoster ophthalmicus; Scleritis – thinning and necrosis.

MANAGEMENT

Infective scleritis is treated with both specific antibiotics and, frequently, early surgery to excise the affected area. Topical steroids are ineffective. Non-infective scleritis is treated as follows.

Blood tests

Laboratory tests should be performed to determine if there is an underlying cause of the scleritis. Appropriate tests are a full blood examination (FBE), rheumatoid factor, anti-neutrophil cytoplasmic antibodies (c-ANCA), antinuclear antibodies (ANA), and renal function tests.

Ocular investigations

A posterior segment examination should be made for signs of inflammation or retinal changes.

Oral medication

Diffuse and nodular scleritis can usually be managed with oral non-steroidal anti-inflammatory drugs (NSAIDs) such as flurbiprofen 100 mg tds, although occasionally oral prednisolone is also required especially for nodular scleritis. Inflammatory necrotizing scleritis usually requires oral corticosteroid and may also require other immunosuppressive agents such as cyclosporin, methotrexate, or cyclophosphamide if there is an associated underlying systemic disease. The newer TNF-α antagonists also have a useful role to play in refractory disease.

S Scleritis – thinning and necrosis

DESCRIPTION

The sclera is composed of criss-crossed lamellae of collagen fibers. It is of varying thickness, being much thinner immediately posterior to the muscle insertions (approximately 0.3 mm), and thickest around the optic nerve (approximately 1.0 mm). Interspersed between these lamellae are scattered resident fibroblasts termed sclerocytes. Although the metabolic rate of sclera is extremely low, there is a slow rate of collagen replacement sustained by the presence of sparse scleral blood vessels.

Loss of scleral tissue can occur via several mechanisms:

- the release of lytic enzymes by infective organisms, such as Gram-negative bacteria or invading neutrophils, as occurs in inflammatory necrotizing scleritis; this is an acutely sight-threatening disease
- the drop-out of scleral vessels, as occurs following the application of beta-radiation and mitomycin C or with scleromalacia perforans in rheumatoid arthritis and Wegener's granulomatosis
- spontaneously, especially in the very elderly; this usually occurs just anterior to the rectus muscle insertions.

Perforation of the sclera secondary to thinning is very rare. However, significant ocular surface problems can occur, especially with post-irradiation scleral necrosis.

SYMPTOMS

Unless inflammatory, scleral thinning itself is asymptomatic. If there is a focal depression, an epithelial defect, or calcium deposition, surface irritation with grittiness, burning, and a foreign-body sensation may occur.

SIGNS

The area of scleral loss may be diffuse, as occurs with scleromalacia perforans when the affected area is frequently raised, or quite focal, as occurs in post-irradiation necrosis when the area is usually depressed. The involved sclera itself is avascular and, in the case of post-irradiation necrosis, the surrounding sclera is also relatively avascular.

The overlying conjunctival epithelium is usually fused to the sclera due to loss of the subconjunctival space. The overlying epithelium may ulcerate, leading to infection (frequently fungal scleritis), or result in chronic ulceration with resultant calcification, with formation of a characteristic calcium plaque.

INCIDENCE

Uncommon.

SIGNIFICANCE

If there is an associated epithelial defect, infective scleritis may occur.

DIFFERENTIAL DIAGNOSIS

Ocular melanosis; eroded ciliary body melanoma.

SEE ALSO

Scleritis – overview; Pterygium.

MANAGEMENT

Senile scleral thinning does not require any treatment.

Review

If there is a significant area of thinning, especially following the application of beta-irradiation or mitomycin C, the patient should be reviewed annually because of the progressive nature of the lesion.

Tear supplements

If ocular surface irritation occurs, topical lubricants are usually quite helpful in reducing symptoms.

Surgery

Calcium plaques can be manually removed at the slit lamp with topical anesthesia and forceps if they are causing significant discomfort. Care should be taken because of the risk of infective scleritis, and patients should be placed on a broad-spectrum topical antibiotic qid for 1 week following the procedure.

If chronic ulceration occurs at the site of necrosis, surgical rehabilitation with excision of the necrotic tissue and overlying conjunctiva and replacement with a scleral patch graft can be performed. This is rarely needed.

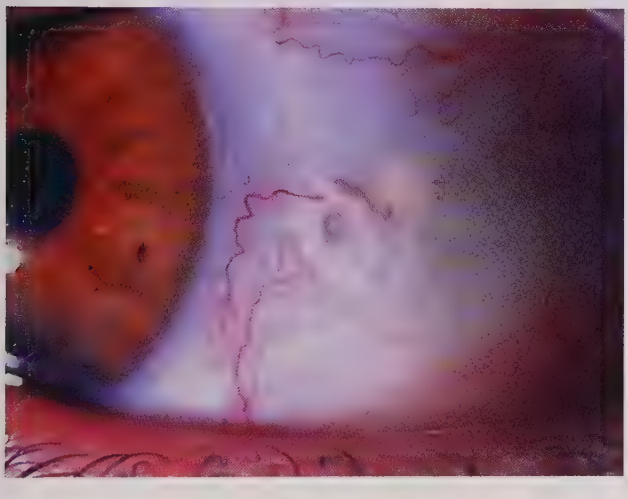


Fig. S-3a Scleral necrosis resulting from beta-radiation application following pterygium surgery. Note the central calcium plaque.

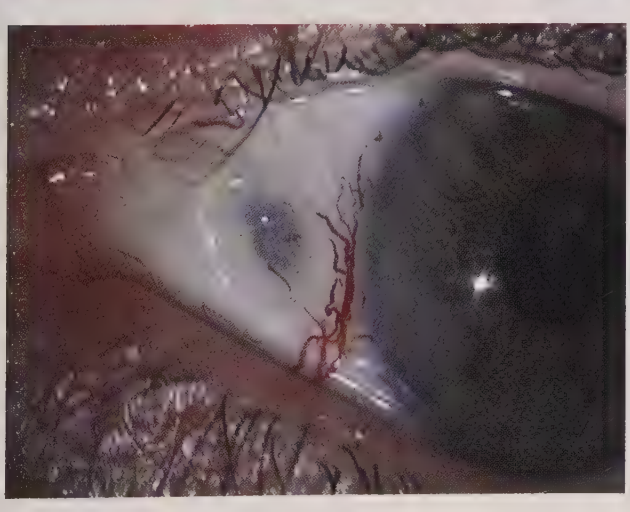


Fig. S-3b Scleromalacia from beta radiation following pterygium surgery. Note not only the marked thinning but also the surrounding relatively avascular area and markedly abnormal blood vessels just central to the area of maximal thinning.

S

Seasonal allergic conjunctivitis (SAC)

Allergic rhinoconjunctivitis, hay fever conjunctivitis

DESCRIPTION

Seasonal allergic conjunctivitis (SAC) is the most common form of ocular allergy. It is usually associated with allergic rhinitis (hay fever) and may be associated with other allergic diseases such as eczema and asthma. Pollens are the most common precipitating allergens and, as such, symptoms usually occur from spring through to mid-summer. If the patient is also allergic to a perennial allergen such as house dust mite, symptoms may persist throughout the year – perennial allergic conjunctivitis (PAC).

SYMPTOMS

The hallmark symptom of all ocular allergy is itch. The itch of SAC is recurrent, transient, and associated with increased lacrimation and slightly red eyes. Occasionally, there may be a mucus discharge or even mucus strands. As it typically occurs with hay fever there is associated sneezing, a watery nasal discharge, and nasal itch. Unlike with viral conjunctivitis there is no associated lymphadenopathy.

SIGNS

During an attack, the conjunctiva is slightly injected and edematous, with a papillary response leading to a velvety appearance most noticeable in the fornices.

INCIDENCE

Very common.

SIGNIFICANCE

Leads to symptoms of ocular irritation but does not lead to corneal scarring.

DIFFERENTIAL DIAGNOSIS

Other forms of ocular surface disease including Dry eye syndromes and Blepharitis. Other forms of ocular allergy including Atopic keratoconjunctivitis (AKC), Vernal keratoconjunctivitis (VKC), and Giant papillary conjunctivitis (GPC).

SEE ALSO

Allergic eye disease – overview; Conjunctivitis – overview.

MANAGEMENT

Allergen avoidance

Aside from the specific drug treatments listed below, allergen avoidance is an important component of treating SAC. Patients allergic to pollens should be counselled to avoid outdoor activities on high pollen count days. Cleaning around the eyes with a face washer and the topical application of ice packs can also reduce symptoms.

Therapeutics

Topical antihistamines such as naphazoline, antazoline, and levocabastine are quite useful in reducing ocular itch and discomfort. They are usually instilled when the symptoms occur and act rapidly, lasting for about 1 to 2 hours. They are often formulated with an ocular lubricant for added relief. Newer unpreserved unit dose preparations of these medications are now available.

Topical non-steroidal agents, especially ketorolac, are also very effective in reducing ocular itch.

Topical mast-cell stabilizers can also be effective in preventing the recurrence of symptoms, although they need to be taken prophylactically three to four times a day on a regular basis during the allergy season. It is not used as frequently in the treatment of SAC as PAC, VKC, and AKC. Topical corticosteroids are rarely used to treat SAC.

The most useful and commonly used class of topical agents are combined antihistamine and mast-cell stabilizers such as olopatadine and ketotifen.

Oral medication

Although oral antihistamines are often quite effective in treating allergic rhinitis, unfortunately they often have a minimal effect on ocular symptoms. Commonly used non-sedating oral antihistamines such as loratadine, cetirizine, and fexofenadine can be taken daily to treat other symptoms of hay fever.

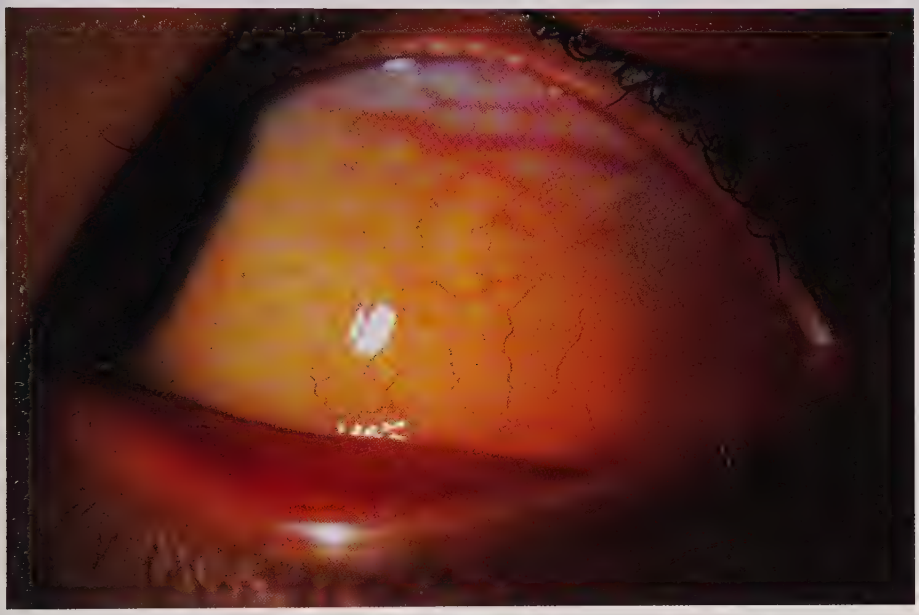


Fig. S-4 Velvety, edematous conjunctiva typical of acute SAC.

S

Sebaceous cell carcinoma

Sebaceous gland carcinoma, meibomian carcinoma

DESCRIPTION

The eyelids contain modified sebaceous glands, including the meibomian glands, which secrete the lipid layer of the tear film, and the glands of Zeis, which are associated with the lash follicles. There are also sebaceous glands in the caruncle, eyebrows, and elsewhere.

Neoplasms of the sebaceous glands may be benign, e.g. sebaceous gland adenoma, or malignant, such as sebaceous cell carcinoma. The benign lesions are more common on the non-ocular skin, whereas carcinoma more commonly involves the meibomian glands and other ocular sites.

Sebaceous carcinoma leads to local destruction of tissue and, if unchecked, can invade the globe, orbit, or sinuses. Alternatively, the tumor may spread in small clusters of cells within the epithelia of the conjunctiva or adjacent tissues, a pattern termed "pagetoid spread." Metastasis of the tumor can occur to the pre-auricular and sub-mandibular lymph nodes and, in a small proportion of patients, death can result from metastasis to the lung or liver.

SYMPTOMS

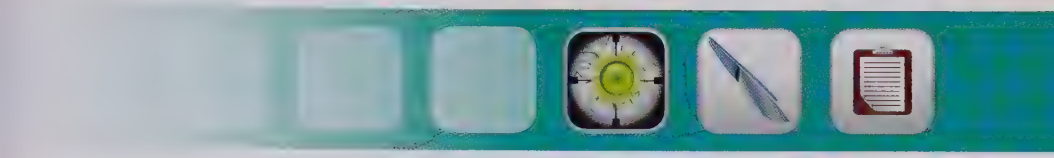
Typically, a painless, firm mass is noted; this may resemble a chalazion. It is almost always a unilateral condition.

SIGNS

- Nodular. While a sebaceous cell carcinoma may present as a slow-growing firm mass, it may have a variety of other presentations. It can appear similar to a recurrent chalazion, but the index of suspicion should be raised if the eyelid margin shows signs of tissue destruction, such as being irregular, vascularized, or with loss of eyelashes. In other cases, it may be confused with basal cell carcinoma (BCC) or squamous cell carcinoma (SCC).
- Spreading. Intraepithelial or pagetoid spread of sebaceous gland carcinoma may cause the condition to mimic more benign conditions such as blepharitis or



Fig. S-5 Nodular meibomian gland (sebaceous cell) carcinoma.



meibomianitis. It may also appear like a chronic conjunctivitis, superior limbic conjunctivitis, or sclerosing BCC.

The superior eyelid is most often affected by sebaceous gland carcinoma, and the lesion has a slow pattern of growth with an insidious onset. If the lipid secretions of the meibomian glands are affected, the tumor may have a yellowish color. Assess the preauricular or sub-mandibular lymph nodes for lymphadenopathy, since this may be an indication of metastasis.

INCIDENCE

Sebaceous cell carcinoma is very rare (approximately 1/100,000). It accounts for approximately 1% of eyelid malignancies, being the third most common after BCC (approximately 80–90%) and SCC (approximately 10%). As with the other malignancies, it more often occurs in patients who are in their 60s or 70s, although it occasionally occurs in younger patients as well.

SIGNIFICANCE

Sebaceous gland carcinoma is the most serious of the eyelid carcinomas. It is a malignant invasive lesion; untreated it is a fatal condition due to metastasis.

DIFFERENTIAL DIAGNOSIS

Basal cell carcinoma; Squamous cell carcinoma; Chalazion; Hordeolum; Preseptal cellulitis; Blepharitis.

SEE ALSO

Skin tumors – overview; Rosacea keratitis.

MANAGEMENT

Pathology

A full-thickness biopsy is indicated if there is doubt about the nature of the diagnosis. Suspicion should be raised with any chronic chalazion or an eyelid lesion with loss of eyelashes, irregular lid margin, or increased vascularity. Successful management depends upon early diagnosis and aggressive intervention.

Incisional surgery

Wide surgical excision is the mainstay of treatment. Eyelid reconstructive surgery is usually required. Additional possible treatments include radiation, cryotherapy, or carbon dioxide laser. Systemic treatments are likely to be required if there is a risk of metastasis.

Review

After surgery, review may initially be weekly to monthly to ensure full healing of the surgical wound and detect any possible recurrence.

S

Seborrheic keratosis

Basal cell papilloma, keratinocyte hyperplasia

DESCRIPTION

Seborrheic keratosis occurs due to an epidermal keratinocyte hyperplasia. Foci of keratinizing cells form cysts within the lesion. When the cysts coalesce and reach the surface of the skin, there is a discharge of the keratinized material causing the surface of the lesion to be irregular. The keratotic surface may be shed or removed, only to reform. Unlike actinic (solar) keratosis, a keratinocyte dysplasia that may transform into squamous cell carcinoma, the potential for malignant change with seborrheic keratosis is exceptionally low.

The term seborrheic refers to an excessively oily or greasy surface, due to the accumulation of the discharge from the sebaceous glands within the folds and fissures of the keratosis. The lesions also develop varying amounts of melanin, causing them to appear pigmented. For this reason, some seborrheic keratoses appear similar to malignant melanoma.

Seborrheic keratoses occur in any area where there are pilo-sebaceous follicles and, thus, do not occur in hairless areas of skin. They occur mainly on the arms and face, but can occur almost anywhere on the body, and can be in areas not always exposed to excessive sunlight. The cause is unknown, although sunlight may play a role.

SYMPTOMS

The patient may have noticed single or multiple slowly growing, superficial plaques; these have a characteristically greasy touch. Seborrheic keratoses are not usually painful or tender, but they may be inflamed or irritated if they have been subject to friction or manipulation.

SIGNS

Seborrheic keratoses are slow-growing, and typically reach a size of 1–2 cm in diameter. They form a flat, fissured, and slightly raised plaque, with a characteristic greasy texture and, most commonly, are an evenly pigmented brownish color.

Initially, seborrheic keratoses may only be slightly raised or dome shaped, but large ones may appear to be "stuck on" to the skin rather than growing from it. They may appear like viral warts or nevi (moles), although these have a smoother, dome-shaped appearance, or (infrequently) they may be pedunculated. On occasions, the lesion may become inflamed, enlarged, and itchy.

INCIDENCE

Uncommon around the eyelids (approximately 1/1000). They commence around the third decade of life, increase in frequency with increasing age, and are the most common pigmented lesion in people over 50.

SIGNIFICANCE

Seborrheic keratosis is a benign lesion, usually causing only mild cosmetic concern.

DIFFERENTIAL DIAGNOSIS

Melanoma – eyelid; Basal cell carcinoma (pigmented); Actinic (solar) keratosis; Nevus – eyelid; Skin tumors – benign (viral warts; keratoacanthoma).

SEE ALSO

Skin tumors – overview.

MANAGEMENT

Incisional surgery

Removal may be indicated to confirm diagnosis, for cosmetic purposes, or if they are becoming irritated with mild trauma from day-to-day activities. Liquid nitrogen cryotherapy is indicated if diagnosis is certain. Other possible treatments include curettage of small flat lesions or excision if pedunculated. While they can be removed for cosmetic reasons, they commonly recur following removal.

Pathology

Excised lesions should be submitted for histopathological examination.

Advice

Since seborrheic keratoses are benign, the majority of these lesions do not require treatment. The patient can usually be reassured with regard to the nature of the lesion and asked to re-present if any change is noticed.



Fig. S-6 Seborrheic keratosis (basal cell papilloma).

S

Skin tumors – benign

Viral wart, verrucae, papilloma, squamous cell papilloma, cutaneous horn, keratoacanthoma

DESCRIPTION

Viral wart

This condition is also known as a squamous cell papilloma, meaning a benign squamous proliferation, and is usually small in size (less than 1 cm). It is the most common benign tumor of the eyelids and is usually found in adults. Papillomas are caused by the human papilloma virus (HPV); the natural course of these infections is highly variable although many do eventually involute spontaneously.

Cutaneous horn

A superficial hyperkeratotic mass that may be associated with actinic keratosis or squamous cell carcinoma.

Keratoacanthoma

A benign, initially rapidly growing, but self-limiting epithelial neoplasm, keratoacanthoma shares some clinical and histological similarities with squamous cell carcinoma (SCC), but it appears to represent an abortive malignancy. Spontaneous involution can take up to a year.

The etiology of keratoacanthoma appears to be related to UV radiation exposure, although viral and other mechanisms have also been proposed. It is found in healthy patients, although more frequently in patients using immunosuppression therapy.

SYMPTOMS

A viral wart usually causes minimal symptoms unless it is chronically irritated or inflamed. With all conditions there may be concern about the cosmesis. Keratoacanthoma is most likely of concern to the patient because of the rapid change in the lesion. These conditions are usually unilateral or asymmetrical.

SIGNS

Viral wart

A pedunculated or broad-based sessile lesion, the surface is lobed but non-ulcerated. The mass is avascular and usually skin-colored.

Cutaneous horn

Hyperkeratotic lesion, which protrudes through the skin, appearing somewhat like a piece of fingernail.

Keratoacanthoma

This lesion has a number of appearances, depending upon its stage of development. Keratoacanthoma begins from normal skin, then becomes a single pink papule that can reach 2–4 cm in size, then remains dormant for 2 to 3 months. A key feature is the presence of a central keratinous plug or core. Later, with involution, the central core is expressed, and an atrophic scar remains.

INCIDENCE

Viral warts are very common (greater than 1/10); whereas cutaneous horn and keratoacanthoma are uncommon (approximately 1/1000). The latter are more common in the 50–70-year-old age group and fair-skinned individuals.

SIGNIFICANCE

These are benign lesions, although they may be of cosmetic concern to the patient, and accurate differentiation from malignant lesions is important.

DIFFERENTIAL DIAGNOSIS

Squamous cell carcinoma; Seborrheic keratosis; Actinic keratosis; Pyogenic granuloma; Nevus – eyelid.

SEE ALSO

Skin tumors – overview; Melanoma – eyelid.

MANAGEMENT

Review

Documentation and review are the mainstays of management of benign skin conditions. The advent of digital photography has made it easier to document a lesion and to promote self-assessment by the patient. Review is indicated if the patient feels there is any change to the lesion.

Incisional surgery
The lesion should be excised if it is causing cosmetic concerns or if there is any concern with regard to the diagnosis. Keratoacanthoma may be excised before it is fully grown in order to avoid subsequent scarring associated with a larger lesion.

Pathology
Excised lesions should be submitted for histopathological examination.

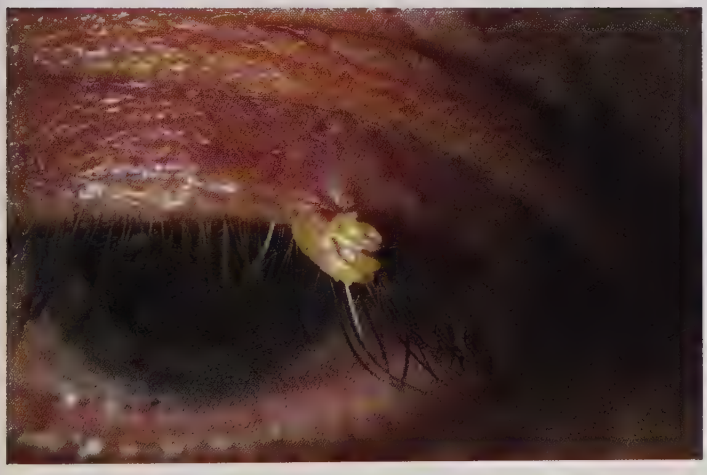


Fig. S-7a
Pedunculated viral wart (squamous cell papilloma).



Fig. S-7b
Keratoacanthoma after its growth phase, prior to involution and regression.

S

Skin tumors – overview

Dysplasia, metaplasia, neoplasia – bènign, neoplasia – malignant, skin cancer

DESCRIPTION

This text includes numerous conditions related to skin tumors. This entry is designed to explain the basis for the terminology and provide a framework for assessment. Depending on the histopathological changes within a tissue, various terms are used.

Metaplasia

Refers to the term used when one adult cell type is replaced by another. For example, conjunctival mucosa becoming keratinized in dry eye (squamous cell metaplasia).

Dysplasia

Refers to the abnormal growth of a tissue, but where normal tissue architecture is retained. For example, a small nevus or actinic (solar) keratosis.

Neoplasia

An abnormal growth of a tissue with uncontrolled multiplication of cells and loss of normal tissue architecture is called a neoplasia, also referred to as tumors or cancer.

A neoplasia signifies the irreversible loss of control of cell growth, frequently due to alteration of oncogenes. Oncogenes are a gene sequence that affect cell growth and signal transduction. Related genes in normal cells are called proto-oncogenes. There are also tumor suppressor genes, e.g. the absence of such a gene causes retinoblastoma.

Classification

There are three main forms of neoplasia:

- Benign. These lesions only grow by expansion, affecting the local tissue, and have limited growth potential. Examples are seborrheic keratosis, actinic (solar) keratosis, nevi, keratoacanthoma, viral wart (papilloma), cutaneous horn, pyogenic granuloma, strawberry nevus, port wine stain.
- Malignant. Malignancy signifies a lesion that grows and invades adjacent tissues. Examples of such lesions around the external eye include conjunctival intraepithelial neoplasia (CIN), basal cell carcinoma (BCC), squamous cell carcinoma (SCC), sebaceous gland carcinoma (SGC), melanoma, Kaposi sarcoma, and Merkel cell carcinoma.
- Metastatic. The cells of a neoplasm infiltrate the blood vessels and lymphatics, to be spread throughout the body, e.g. melanoma, SCC, SGC.

The terminology refers to the tissue affected by the neoplasm: "carcinoma" refers to tumors from epithelia, e.g. BCC; "adeno-" infers glandular; "sarcoma-" refers to neoplasms derived from connective tissue.

SYMPTOMS

The patient may have cosmetic concern for a mass or lesion. They may have noticed a change in its size or noted pain and tenderness. Skin tumors tend to be unilateral and asymmetrical. Posterior extension may lead to cranial nerve palsies with ptosis and eyelid anesthesia or paresthesia.

SIGNS

Common findings with malignant tumors of the eyelid include skin ulceration, inflammation, and distortion of the eyelid anatomy. Other findings suggestive of malignancy include abnormal color or texture, persistent bleeding, and madarosis (loss of eyelashes). Induration of the lesion signifies a feeling of hardness on palpation, as well as the impression that the lesion extends deeper than it appears to. Further signs that may be used in assessment include: rapidity of growth, pigmentation, telangiectasia, and skin surface texture including whether scaly, waxy/oily, or verrucous (wart-like).

INCIDENCE

Benign neoplasms are very common (approximately 1/10) in all age groups. Malignant neoplasms are less common.

SIGNIFICANCE

Malignant lesions can be locally invasive and cause significant tissue destruction and loss of function. Metastatic lesions are potentially life-threatening.

DIFFERENTIAL DIAGNOSIS

See the conditions under the subheading "Classification" above, listing benign and malignant neoplasms.

SEE ALSO

Skin tumors – benign; Hemangioma; Chalazion; Cysts of the eyelid; Hordeolum; Molluscum contagiosum; Preseptal cellulitis; Xanthelasma.

MANAGEMENT

The specific management depends upon the nature of the neoplasm – see individual conditions in this text for details.

Advice

Benign lesions may simply be documented and monitored. A photograph can provide more information than a drawing. The advent of digital photography has made it easier to document a lesion and can facilitate discussion with the patient.

Pathology

A biopsy is indicated if there is doubt about the nature of the diagnosis or the distinction between benign and malignant.

Incisional surgery

Excision is usually indicated for malignant lesions and may be indicated for larger benign lesions. Additional systemic treatments are likely to be required for lesions with a risk of metastasis.

Review

Review of the condition at 6- to 12-month intervals is indicated for patients with a history of multiple skin lesions, or as required if there is any concern by the patient. After surgery, review may be weekly to monthly to ensure full healing.



Fig. S-8 Early nodular basal cell carcinoma, showing the pearly smooth translucent surface and mild telangiectasia over the margin. The lesion is firm to palpate but asymptomatic.

Squamous cell carcinoma – lid

SCC, epidermoid carcinoma, prickle cell carcinoma

DESCRIPTION

Some possible histological stages in the development of squamous cell carcinoma (SCC) are (Reifler and Hornblass 1986):

- Intra-epidermal squamous dysplasia (actinic or solar keratosis). In this phase, epidermal cells are replaced by atypical cells, in an area sharply demarcated from the surrounding normal epidermis. The general stratification of the epithelium is still preserved.
- Intra-epidermal neoplasia (SCC in situ, Bowen's disease). This lesion may arise from a pre-cancerous dermatosis or may arise spontaneously. The epithelium shows complete disorganization, but the basement membrane remains intact.
- The invasion of the dermis by squamous tumor cells is the hallmark for the diagnosis of SCC. The tumor can evoke an inflammatory reaction, which can sometimes lead to its confusion with a stye. Compared to a basal cell carcinoma (BCC), an SCC grows more rapidly and has potential to metastasize or cause invasion of the orbit. If the SCC spreads along nerves or in blood or lymphatic vessels, then there is a risk of metastasis to various internal organs. The risk is probably around 1%, but could be higher if there is a delay in diagnosis. SCC is most common in elderly people with a fair complexion and a history of chronic sun exposure and skin damage. Patients with long-term immunosuppression are at increased risk of both primary SCC and metastases.

SYMPTOMS

The patient may have cosmetic concern regarding a unilateral mass or lesion. It may be irritated or itch. SCC may bleed easily, and is occasionally tender on palpation.

SIGNS

As with all malignant tumors of the eyelid, common findings include skin ulceration, distortion of the eyelid anatomy, and possible persistent bleeding. In the early stage of development, SCC is similar in appearance to BCC; it may present as a pearly indurated nodule or a flattish plaque with small vessels visible under the surface. Intra-epidermal neoplasia often presents as a slowly enlarging erythematous scaly patch of skin, with an irregular outline. Progression may be more aggressive if the lesion is at the lid margin. Cranial nerve involvement may occur, with cutaneous anesthesia.

There are three main clinical presentations of SCC:

- Plaque-like, which is characterized by a slowly growing erythematous, scaly, hyperkeratotic area of skin. This form may arise from a pre-existing intra-epidermal squamous dysplasia or neoplasia, as discussed above.
- Nodular, characterized by an indurated hyperkeratotic nodule that may develop crusting erosions and fissures. It may grow over a period of several months.
- Ulcerating, with sharply defined, indurated and elevated borders. Most SCCs will grow and tend to ulceration within a year.

Diagnosis may be made more difficult if SCC presents in combination with another lesion such as keratoacanthoma. Although metastasis is uncommon, with time it can occur to the regional lymph nodes, causing them to become firm and enlarged.

INCIDENCE

SCC is rare (approximately 1/10,000), although it is the second most common eyelid skin tumor.

SIGNIFICANCE

SCC is locally invasive; if untreated, it may threaten life by extending posteriorly to involve the middle cranial fossa. Rarely, it may metastasize.

DIFFERENTIAL DIAGNOSIS

Actinic (solar) keratosis; Basal cell carcinoma (BCC); keratoacanthoma; Molluscum contagiosum; Sebaceous gland carcinoma; Seborrheic keratosis.

SEE ALSO

Skin tumors – overview; Skin tumors – benign.

MANAGEMENT

Pathology

A biopsy should be performed if diagnosis is uncertain. Any persistent, non-healing sore located on or near the eyelids should be considered as SCC until proven otherwise.

Incisional surgery

Treatment modalities include surgical excision and radiotherapy. Surgical excision is the treatment of choice. The margin of excision should be 3–5 mm around the clinical signs of the tumor. Radiotherapy may be used should the patient be considered unsuitable for surgery.

Review

Follow-up should occur for at least 2 years after removal of the primary tumor. Clinical examination for signs of possible secondary tumor, e.g. in the regional lymph nodes, should be made at review.

REFERENCE

Reifler DM, Hornblass A. Squamous cell carcinoma of the eyelid. Surv Ophthalmol 1986; 30: 349–365.

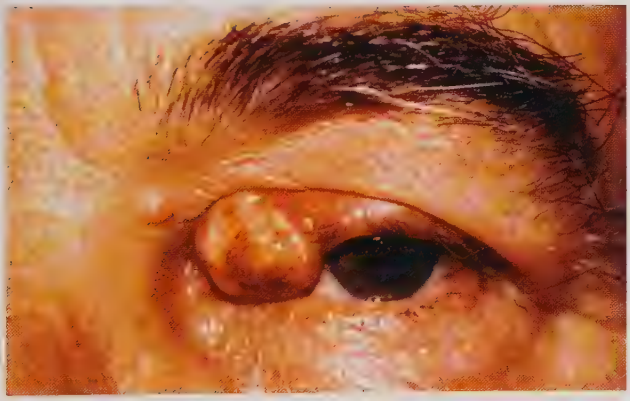


Fig. S-9a Nodular squamous cell carcinoma.



Fig. S-9b Ulcerating squamous cell carcinoma.

S

Stevens–Johnson syndrome (SJS)

Erythema multiforme, toxic epidermal necrolysis (TEN)

DESCRIPTION

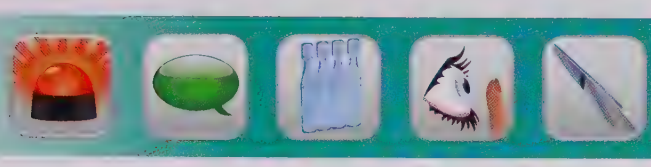
Stevens–Johnson syndrome (SJS) is a rare, occasionally fatal, acute mucocutaneous disorder and is probably an autoimmune disease. Although it is self-limiting, the sequelae of associated scarring and tissue destruction are often permanent. It most commonly occurs in children and young adults. Occasionally, patients also demonstrate features of other autoimmune diseases. The nomenclature surrounding this disease can be somewhat variable. However, in general the spectrum of disease is termed erythema multiforme. In erythema multiforme minor, involvement is restricted to the skin, with no or only minor mucous membrane involvement. In erythema multiforme major, also known as SJS, both the skin and two mucous membranes are affected. Severe skin involvement with extensive necrosis is termed toxic epidermal necrolysis (TEN). Drugs are the most common precipitating factor. Although a wide range of both topical and systemic drugs have been implicated, those most commonly involved are sulfonamides: penicillin, barbiturates, and phenytoin. Infections with various viruses, bacteria, and fungi, including herpes simplex virus, have also been implicated. Histopathologically, during the acute phase there is an increase in circulating immune complexes. Skin and mucosal biopsies of affected areas demonstrate a predominantly perivascular non-specific inflammatory reaction. Subepithelial bullae formation is common, eventually leading to scarring, with extensive goblet cell loss and epithelial keratinization.

SYMPTOMS

Initial presentation is with symptoms of fever, malaise, and upper respiratory tract infection. Patients often present because of concern over skin and mucosal membrane involvement. Ocular involvement leads to red, gritty eyes with increased lacrimation.



Fig. S-10 Focal, subepithelial conjunctival scarring following a relatively mild episode of SJS.



In the long term, dry eye symptoms are common as is a foreign-body sensation from eyelid scarring or ocular surface keratinization. Blurring of vision occurs if there is central corneal involvement from the ocular surface disease.

ACUTE PHASE SIGNS

The skin lesions are usually symmetric and occur most frequently on the extremities. Crops of skin lesions may develop every 2 to 3 weeks for 1 to 2 months. The primary lesion is a round, erythematous maculopapule termed a "target lesion." Desquamation may occur in severe disease (TEN).

The ocular acute phase usually lasts 2 to 3 weeks and involves bilateral conjunctival inflammation, with bullae, ulceration, and, occasionally, membrane formation. Macular lesions that subsequently become bullous and eventually ulcerated are common around the mouth and occasionally involve other mucous membranes including the mouth, nose, larynx, and pharynx.

LONG-TERM SEQUELAE

Ocular surface scarring is variable depending on the severity of the disease. Possible sequelae include severe dry eye from blepharitis, lacrimal ductule occlusion and goblet cell loss, conjunctival shrinkage with symblepharon formation, limbal stem cell deficiency, and lid dysfunction with ankyloblepharon and trichiasis. These can result in persistent epithelial defects, microbial keratitis, ocular surface keratinization, and corneal perforation.

INCIDENCE

Very rare.

SIGNIFICANCE

In severe cases may be extremely destructive of the ocular surface, leading to a marked bilateral loss of vision.

DIFFERENTIAL DIAGNOSIS

Ocular cicatricial pemphigoid; Chemical burns; scalded skin syndrome (Lyell's disease).

SEE ALSO

Conjunctivis–overview; Conjunctival scarring; Dry eye syndromes; Trachoma; Atopic keratoconjunctivitis; Limbal stem cell deficiency; Trichiasis; Membranous and pseudomembranous conjunctivitis.

MANAGEMENT

Advice

As a recurrence of the disease can occur with repeat challenge by the original drug, patients who have had SJS should avoid taking any medication, either topically or systemically, of the same class as a drug that was thought to lead to the initial attack.

The acute phase of SJS is treated in hospital with extensive care. Unfortunately, the use of systemic corticosteroids and other strategies such as plasmapheresis seem to have little benefit. Topical ocular treatment is with prophylactic antibiotics, lubricants, and mild corticosteroids.

The long-term sequelae of ocular surface scarring, dry eye, and eyelid disease are managed with a wide range of supportive and preventative measures, including medical (with lubricants) and surgical (with punctal occlusion and trichiasis surgery). Keratinization of the ocular surface, especially along the lid margins, can occur secondary to ocular surface scarring and dry eye. This can be treated with Oc. vitamin A 1% bd. Care should be taken in using topical vitamin A ointment in darker skinned individuals because of its potential to lead to skin depigmentation. Acute intervention is frequently required, even years after the initial episode, for specific complications such as microbial keratitis, persistent epithelial defects, limbal stem cell failure, corneal perforation, and ocular surface inflammation.

Contact lens

See "Contact lenses – gas permeable" for therapeutic treatment.

S

Sub-conjunctival hemorrhage

Ecchymosis

DESCRIPTION

A sub-conjunctival hemorrhage (ecchymosis) arises due to bleeding from a conjunctival blood vessel into the space between the sclera and conjunctiva. Idiopathic ecchymosis is a benign condition usually arising spontaneously, although it may occur secondary to mild ocular trauma or from coughing or straining.

While in the vast majority of cases the etiology is idiopathic, it may also be seen in patients with whooping cough, and, very rarely, there may be an underlying systemic condition such as hypertension or a bleeding disorder. A sub-conjunctival hemorrhage may also form part of the clinical presentation of blunt eye trauma.

SYMPTOMS

Sub-conjunctival hemorrhage usually occurs unilaterally with a red eye. It may be described by the patient as a "black eye." In addition, there may be mild ocular irritation. There is no history of discharge or ocular pain, and vision is unaffected. The patient may have experienced a sub-conjunctival hemorrhage previously.

SIGNS

The eye is red due to blood in the sub-conjunctival space. Classic signs are the clear ring around the limbus that is free of blood, as well as being able to locate the posterior limit to the hemorrhage. There are no blood vessel patterns, although the blood may show streaks as it spreads.

If there has been extensive trauma to the eye, the entire sclera may be obscured by blood, and there may be associated eyelid bruising. The patient should be assessed for any conjunctival lesion, and a check made of intraocular pressure, pupil reactions, and ocular motility.

INCIDENCE

Common (approximately 1/100).

SIGNIFICANCE

If idiopathic, the condition is benign, although there may be mild cosmetic concern. Rarely associated with significant systemic disease.

DIFFERENTIAL DIAGNOSIS

Conjunctivitis; Preseptal cellulitis; Glaucoma – acute angle closure; Iritis; Uveitis.

SEE ALSO

Eyelid trauma.

MANAGEMENT

This is essentially a benign condition, requiring no treatment. The patient should be advised that the condition will resolve spontaneously within 1 to 2 weeks (depending upon the initial density of the hemorrhage). Discourage use of aspirin unless on medical instruction.

Symptomatic treatment

- Cold compresses for 24 hours, to assist with cessation of the bleeding (if bleeding is still active at presentation).
- Hot compresses for 3 to 5 days, to assist with resolution of the hemorrhage.
- Topical ocular lubricants if there is mild ocular irritation present.

The patient should be advised to return for review if there is not substantial resolution within 2 weeks for idiopathic cases.

Systemic investigation with a full blood examination (FBE) and clotting profile is indicated if the sub-conjunctival hemorrhages are recurrent, if there is spontaneous bleeding or bruising elsewhere, if the hemorrhage fails to clear, or if both eyes are affected.

Traumatic

For management, see "Eyelid trauma." In brief, rule out a ruptured globe (internal traumatic signs), effects on the extraocular muscles, pupillary defects, or retrobulbar hemorrhage. The latter condition would have signs of proptosis and raised IOP, and the hemorrhage would extend posteriorly. Work-up would include referral for a CT scan of the orbit.

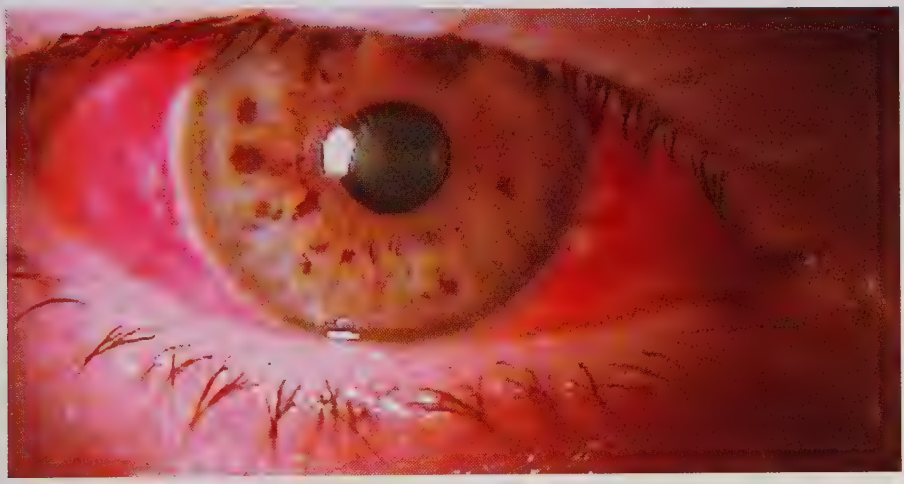


Fig. S-11a Appearance in primary gaze.



Fig. S-11b Limitation of hemorrhage, in the same patient. The clear para-limbal zone is evident in both images.

S

Superficial punctate keratopathy (SPK)

Punctate epithelial erosions (PEE)

DESCRIPTION

Superficial punctate keratopathy (SPK) results from a loss of superficial epithelial cells and associated defects in the overlying mucous layer. The level of epithelial involvement is usually very superficial, often not affecting the epithelial nerves; for this reason, the condition may be asymptomatic when mild.

The condition is not inflammatory, and corneal infiltrates are not present. Strictly speaking, a keratitis is only present if inflammation is present, and so the term "superficial punctate keratitis" should be reserved for when infiltrates or other signs of inflammation occur.

A word of caution on the nomenclature of superficial punctate keratopathy: in some centers the abbreviation SPK is used solely to refer to Thygerson's SPK, and the term punctate epithelial erosions (PEE) is used to describe non-inflammatory superficial epithelial disruption. In other centers, the abbreviation SPK is used synonymously with PEE to denote all causes of superficial epithelial disruption.

Possible causes of SPK

- Desiccation, e.g. dry eye/exposure keratopathy, blepharitis, ectropion, nocturnal lagophthalmos
- Toxicity and hypersensitivity, e.g. chemical injury, toxicity to topical drugs, contact lens solution preservatives
- Radiation, e.g. ultraviolet (snow blindness), flash burn
- Hypoxia, e.g. associated with the wearing of contact lenses with low oxygen transmissibility
- Mechanical, e.g. trichiasis, trauma
- Infection, e.g. *Chlamydia*, molluscum contagiosum
- Systemic disease, e.g. end-stage diabetic disease

SYMPTOMS

There may be none, or there may be discomfort, photophobia, blurring of vision, dryness, or foreign-body sensation.

SIGNS

Multiple microscopic epithelial defects, generally too numerous to count. Superficial punctate keratopathy is generally diagnosed using a slit lamp with cobalt blue light following the instillation of fluorescein dye into the eye. Contrast will be maximized if the staining is viewed though a yellow Wratten filter. While mild SPK may not be visible to the naked eye, if the punctate erosions are severe, coalescent, or confluent, the staining may be evident without magnification. It often extends onto the conjunctiva.

SPK can be a non-specific condition; however, in some cases a characteristic pattern of staining is present that can be indicative of the cause (see Figure S-13b). Other possible signs include conjunctival hyperemia or epiphora.

INCIDENCE

Very common (greater than 1/10).

SIGNIFICANCE

The significance of SPK is dependent upon the underlying cause. Usually SPK is only of mild significance although, if the underlying condition is untreated, the potential for corneal infection is increased.

DIFFERENTIAL DIAGNOSIS

Toxic keratopathy; Exposure keratopathy; Neurotrophic keratitis; Contact lens-related conditions; Flash burn.

SEE ALSO

Thygeson's superficial punctate keratitis; Bacterial keratitis; *Acanthamoeba* keratitis.

MANAGEMENT

By definition, SPK is a superficial condition. For this reason, management is primarily related to identifying the cause and facilitating the recovery of the ocular surface. Extensive amounts of SPK may be treated more aggressively and followed up more closely.

Tear supplements
Mild SPK due to desiccation may be treated with tear supplements bid to qid. Avoid tear supplements preserved with BAK.

Therapeutics
Broad-spectrum antibiotic drops are not usually required unless there is a full-thickness epithelial defect. Otherwise, the healing of superficial punctate defects may be retarded due to the effect of preservatives.

Cease contact lens wear
The patient should stop wearing contact lenses until the SPK has resolved and the underlying cause has been addressed (see "Contact lens complications"). If there are deposits on the lenses, the lenses should be replaced.

Review
Mild SPK usually heals within 24 hours. If there is concern about the condition, review after 3 days to ensure that the treatment is successful or that the underlying cause has been satisfactorily addressed.

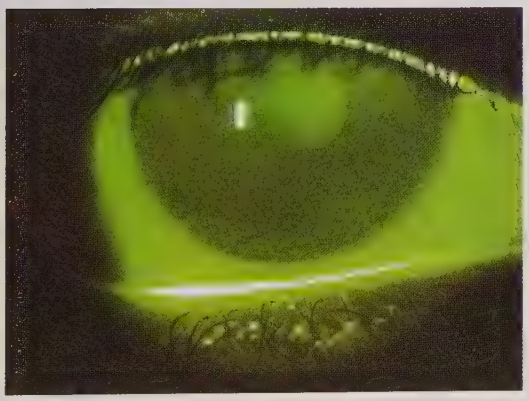


Fig. S-12a Moderate punctate epithelial erosions associated with contact lens solution toxicity.

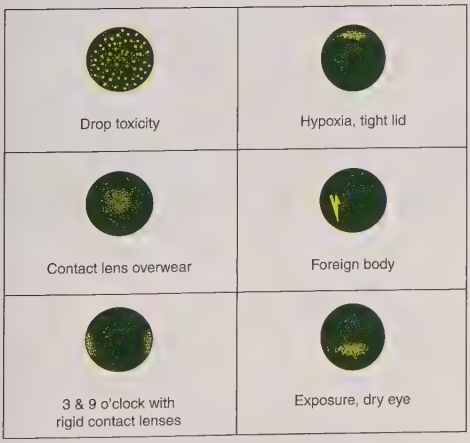


Fig. S-12b Some typical patterns of staining with contact lens usage.

S Superior limbic keratoconjunctivitis (SLK)

DESCRIPTION

Superior limbic keratoconjunctivitis (SLK) is a rare inflammatory condition usually affecting middle-aged women. It is thought to occur from rubbing of the superior bulbar and paralimbal conjunctiva against the conjunctival surface of the upper eyelid. This results in a vicious circle, with rubbing inducing conjunctival edema and more rubbing against the superior cornea. It may occur as an isolated process or associated with either dry eye (KCS) or thyroid eye disease. If associated with thyroid dysfunction, it may be the index eye sign occurring prior to other signs such as proptosis and conjunctival chemosis.

The diagnosis of SLK is frequently delayed due to many of the signs being hidden under the upper eyelid and the non-specific nature of the symptoms.

SYMPTOMS

The symptoms of SLK are usually fairly non-specific with ocular surface irritation with burning and a foreign-body sensation, photophobia, and slight blurring of vision. The eye often does not become red during an attack, but the patient may notice increased lacrimation or even a mucus discharge. Symptoms typically fluctuate significantly and are usually bilateral.

SIGNS

Signs occur at three sites – the marginal superior tarsal conjunctiva, superior bulbar conjunctiva, and superior cornea and limbus. The marginal superior tarsal conjunctiva is inflamed with a 2–3-mm strip of velvety papillary reaction parallel to the lid margin. There is a chemotic and injected 2 to 3 clock-hour band of superior bulbar conjunctiva. This extends for up to 10 mm peripheral to the limbus. This is often best appreciated by viewing the superior bulbar conjunctiva not on the slit lamp but with the naked eye with the patient looking down and manually elevating the upper eyelid. The conjunctiva in this area can easily be shown to be redundant. This is demonstrated by asking the patient to look down and manually pressing the upper eyelid both onto the globe and down toward the limbus. This results in the conjunctiva being thrown up into multiple deep folds. The superior cornea and limbus demonstrate pannus formation, superficial scarring, and thickening of the epithelium. Occasionally, there are also superior corneal filaments. There is characteristic and extensive superficial epithelial punctate staining with both fluorescein and rose bengal at all three of these sites.

INCIDENCE

Very rare.

SIGNIFICANCE

SLK itself does not lead to permanent ocular changes. However, it is important to identify any associated thyroid dysfunction.

DIFFERENTIAL DIAGNOSIS

Terrien's marginal degeneration; Trachoma.

SEE ALSO

Dry eye syndromes; Pannus; Filamentary keratopathy.

MANAGEMENT

Additional investigations

All patients with SLK should be examined for thyroid dysfunction with thyroid function tests.

Tear supplements

Tear supplements are useful, for both treating any underlying dry eye and reducing friction between the superior bulbar and tarsal conjunctiva. A superior silicone punctal plug is also often very useful and may result in total resolution of both symptoms and signs.

Therapeutics

A topical corticosteroid such as fluorometholone qid for several weeks may be useful in reducing inflammation of the superior conjunctiva.



Contact lens

The application of a soft contact lens may be useful as a temporary measure in protecting the superior cornea and reducing friction on the superior bulbar conjunctiva.

Surgery

Surgical management is directed at obliterating the subconjunctival space by fusing the superior conjunctiva to the underlying sclera. This removes the ability of the conjunctiva to roll over the limbus onto the superior cornea. Although various methods have been tried to fuse the conjunctiva, including cauterization, cryotherapy, and a localized chemical burn with sodium hydroxide, surgical resection of the conjunctiva and underlying episclera and Tenon's capsule appears to be the most effective. It is important that sufficient conjunctiva is resected to ensure that the process does not reoccur. As such, the conjunctiva should be resected for 8–10 mm posterior to the limbus.

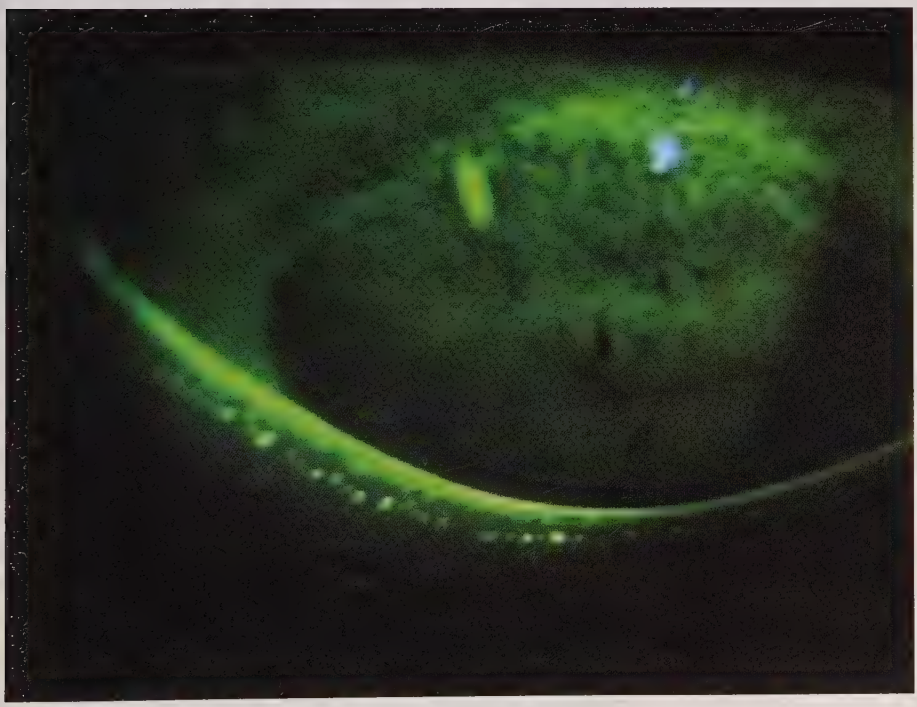


Fig. S-13 Inflamed and injected superior limbus and bulbar conjunctiva with associated pannus in a patient with SLK.

Synechia

Iris adhesions

DESCRIPTION

Adhesion of the iris to nearby tissue or an intraocular implant is called a synechia. Synechia may develop following iris inflammation of whatever cause: iritis, infection or surgical/accidental trauma. Synechia often have specific names depending on the tissue involved:

- crystalline lens (posterior synechia)
- capsular remnant following cataract surgery (capsular adhesion)
- peripheral cornea (peripheral anterior synechia [PAS])
- corneal wound (wound adhesion; if long and thin, is called an iris strand)
- corneal perforation (adherent leukoma)
- corneal graft–host junction (graft–host junction adhesion).

Congenital iris adhesions to an anteriorly displaced Schwalbe line (posterior embryotoxon) are seen in iridocorneal dysgenesis.

SYMPTOMS

The synechia themselves do not cause symptoms. Patients may be symptomatic from the cause of the synechia or secondary to complications of synechia formation such as glaucoma.

SIGNS

Synechia may not always be obvious on slit-lamp examination. Where suspected, a gonioscopic examination should be performed to assess the presence of PAS. A dilated examination should also be performed to assess posterior synechia not involving the pupillary margin. These may only be apparent with mydriasis. Posterior synechia may also lead to a distorted pupil.

Focal areas of pigment deposition on the anterior capsule may indicate a previous posterior synechia. The patient should be examined for evidence of active iris inflammation, such as anterior chamber cell and flare, and complications, such as secondary glaucoma.

INCIDENCE

Uncommon except in defined risks groups, especially recurrent anterior uveitis.

SIGNIFICANCE

PAS may lead to a form of closed-angle glaucoma; graft–host junction adhesions increase the risk of allograft rejection. Extensive capsular or lenticular adhesions may lead to significant pupil dysfunction. Rarely, uveitic posterior synechia can lead to pupil block glaucoma.

DIFFERENTIAL DIAGNOSIS

Posterior embryotoxon.

SEE ALSO

Iridocorneal dysgenesis; Iritis; Uveitis – anterior.

MANAGEMENT

Therapeutics

Recently formed posterior synechia, especially in uveitis, may be broken with medical mydriasis. A drop of homatropine should be instilled and the patient checked 30 minutes later to ensure that any synechia have broken. If any are still present a combination of mydriatics should be instilled, such as phenylephrine 10%, cyclopentolate 1%, and tropicamide 1%, in an attempt to break the synechia. If this is also ineffective, a subconjunctival injection of mydriatics can be tried.

Surgery

Although surgery is rarely performed solely to break established synechia, they are often broken with either visco-dissection or a surgical sweep at the time of intraocular surgery for other indications.

Laser surgery

If there are extensive posterior synechia, a YAG-laser peripheral iridectomy may be performed to prevent pupil block. Patients with uveitis should have at least two peripheral iridectomies performed because of the risk of one becoming blocked by subsequent synechia formation. Occasionally, a surgical iridectomy is required.

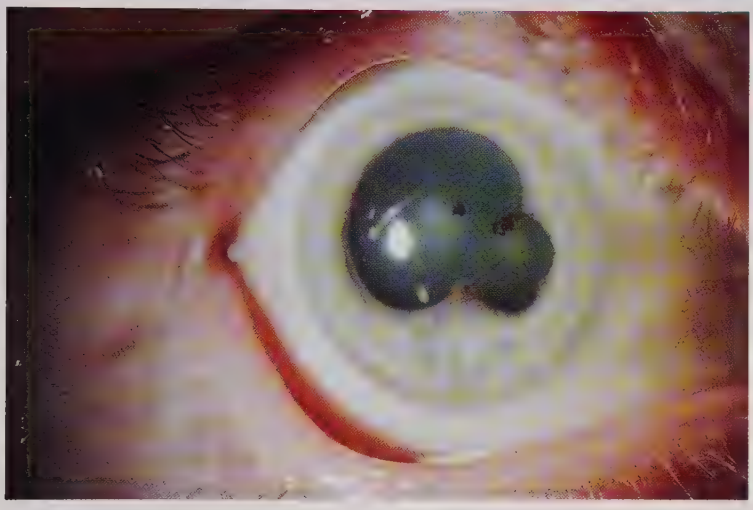


Fig. S-14a Pupillary distortion and pigment on the anterior surface of the crystalline lens secondary to posterior synechia formation in anterior uveitis.



Fig. S-14b Extensive peripheral anterior synechia.

Terrien's marginal degeneration

DESCRIPTION

Terrien's marginal degeneration is a rare peripheral corneal thinning condition of unknown etiology. The thinning leads to corneal stromal relaxation, resulting in induced irregular astigmatism. It may occur at any age, although it is uncommon before the 30s, and most common in older age. It predominantly affects men. Progression is characteristically quite slow. Histopathologically, the corneal collagen lamellae are disrupted and split with significant lipid deposition between collagen layers.

SYMPTOMS

Patients typically complain of slowly deteriorating vision or the need to progressively change their spectacles with increasing astigmatic correction. Both eyes are usually affected, although early in the disease the involvement may be quite asymmetric. Occasionally, patients complain of recurrent attacks of ocular irritation. These are usually mild, but may rarely be quite marked.

SIGNS

The signs are usually slowly progressive, often over years. The thinning usually occurs superiorly first, although it may involve both the superior and the inferior cornea. Rarely, involvement is nasal and temporal only. The initial lesions are fine, white, subepithelial, and paralimbal. These coalesce over time, and the affected cornea begins to thin. A superficial pannus forms, with a whitish, sclerotic area on the central edge of the involved cornea. This involvement then spreads around the limbus. At this stage pseudopterygia often develop. Corneal perforation may occur rarely, usually secondary to minor trauma. As this process progresses, the degree of astigmatism increases and the best spectacle-corrected visual acuity decreases due to the irregular nature of the induced astigmatism. It is not uncommon for cylindrical corrections to be as high as 10 to 15 diopters. The induced irregular astigmatism is usually against the rule, the cornea being steepest at 90° to the area of thinning. As such, if the involvement is nasal and temporal, the induced astigmatism is with-the-rule. There are no signs of ocular inflammation and the corneal epithelium is not ulcerated.

INCIDENCE

Very rare, mainly affects men, and has a bimodal distribution of incidence as described above. May lead to ocular irritation.

SIGNIFICANCE

Leads to induced irregular, usually against-the-rule, astigmatism.

DIFFERENTIAL DIAGNOSIS

Pellucid marginal corneal degeneration; Superior limbic keratoconjunctivitis; limbal Herpes simplex keratitis; senile peripheral cornea furrow; Peripheral ulcerative keratitis.

SEE ALSO

Pannus and pseudopterygia; Corneal opacities – peripheral; Mooren's ulcer.

MANAGEMENT

Additional investigations

Early on in the course of the disease, a corneal power map (corneal topography) may be helpful in making the diagnosis by demonstrating the irregular astigmatism. Corneal pachymetry can define the thinning although this is usually sufficiently marked to be observed on slit-lamp examination.

Therapeutics

Some clinicians believe that, by treating recurrent attacks of minor inflammation, progression of the underlying disease (and the induced astigmatism) can be arrested.

Contact lenses

Rigid gas-permeable lenses can be fitted,

except in the most advanced cases, and usually result in significant visual improvement.

Surgery

Occasionally, if the induced astigmatism is sufficiently marked such that spectacle and contact lens correction is unsuccessful, surgical intervention is required. Surgical rehabilitation is usually with a peripheral, crescentic, inlay lamellar corneal or corneoscleral graft.

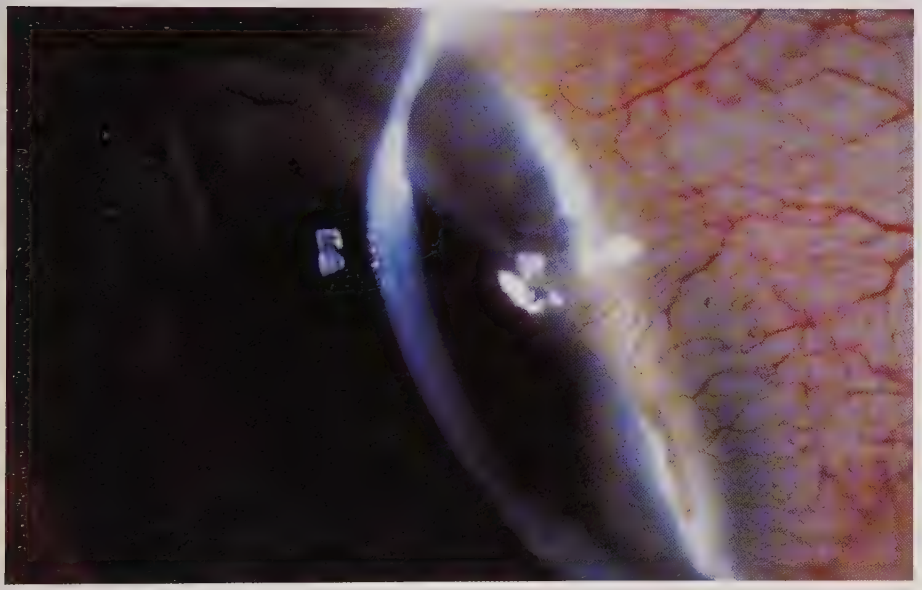


Fig. T-1 Terrien's marginal degeneration, demonstrating superior thinning and peripheral superficial vascularization.

Thygeson's superficial punctate keratopathy

DESCRIPTION

Thygeson's superficial punctate keratopathy (Thygeson's SPK) is a superficial corneal inflammatory condition. It is both rare and of unknown (although presumed viral) etiology. It characteristically affects young adults, and typically follows a relapsing course, affecting both eyes. One of its key features is that the underlying stroma is unaffected except for an occasional mild, isolated, evanescent, faint haze. It never causes stromal scarring. Attacks usually reduce in frequency and cease after several years.

It is not uncommon for other causes of SPK to be misdiagnosed as Thygeson's SPK. Care should be taken because of the somewhat conflicting meaning given to the term SPK by different clinicians. Some clinicians use the term SPK exclusively to refer to Thygeson's SPK, while many others use it synonymously with punctate epithelial erosions (PEE).

SYMPTOMS

Patients complain of recurrent episodes of ocular surface irritation and burning that fluctuate over time. It typically affects both eyes, but symptoms are often asymmetric at any one time. Untreated, the attacks typically occur every 6 to 8 weeks. Aside from ocular irritation, photophobia is quite typical as is increased lacrimation and slight blurring of vision.

SIGNS

The hallmark sign is that of multiple, grouped, raised epithelial lesions, probably epithelial infiltrates. These lesions are referred to as grouped SPK, although the term when applied to this condition describes a different pathological change from that usually referred to as SPK (see "Superficial punctate keratopathy"). They are quite small and have a characteristic "stuck-on" or "breadcrumb" appearance. The lesions are scattered across the whole of the epithelium; the number of lesions is quite variable, from several up to 20, but rarely more. They have a typical central dense area surrounded by a less dense area, termed an "areola."

The disease is typically bilateral but may be quite asymmetric at any one time, with many more lesions being present on one eye than the other. The underlying stroma is either clear or only slightly involved with a faint haze. The intervening epithelium is totally uninvolved. The eye itself is usually white and quiet, with only very mild conjunctival injection being present, if at all, and no anterior chamber reaction.

INCIDENCE

Very rare.

SIGNIFICANCE

While Thygeson's SPK causes recurrent photophobia and ocular irritation, it does not cause corneal scarring or predispose to microbial keratitis.

DIFFERENTIAL DIAGNOSIS

Other causes of SPK, including Dry eye, Blepharitis, toxicity, and Molluscum contagiosum; epidemic keratoconjunctivitis; adult inclusion conjunctivitis (Chlamydial conjunctivitis).

SEE ALSO

Superficial punctate keratopathy; Dry eye syndromes.

MANAGEMENT

Tear supplements

The regular use of ocular lubricants may lessen the ocular discomfort.

Therapeutics

The mainstay of treatment is the use of a moderate-strength topical steroid such as fluorometholone acetate several times a day during an acute attack, followed by maintenance therapy with a weak topical steroid such as fluorometholone qid. Typically, the response to topical steroids is rapid (within days) and complete. Rarely, the disease may become resistant

to weaker corticosteroids, and stronger maintenance steroids may be required. Alternatively, topical cyclosporin A has been reported as being effective.

Therapeutic contact lens
Occasionally, a therapeutic contact lens may be needed to control symptoms in a steroid-resistant case.



Fig. T-2 Multiple, grouped areas of epithelial punctate lesions typical of Thygeson's SPK.

Toxic keratopathy

Drop toxicity, medicamentosa, toxic epitheliopathy, preservative sensitivity, anesthetic abuse keratopathy

DESCRIPTION

Topical medication can adversely affect the ocular surface in several different ways. The drug, preservative, or vehicle (and other components within the formulation) can directly lead to surface toxicity. Alternatively, normal tear film function, important to the health of the ocular surface, can be adversely affected. Some drugs and preservatives can incite an inflammatory reaction. Apart from toxicity, an allergic reaction may also occur to the drug itself or other components within a particular formulation. Whereas allergies can occur from a single drop, toxicity is related to both the frequency of use and dosage. Multiple drugs can have a synergistic effect in leading to surface toxicity. Of particular concern is the preservative used within the formulation. The most common preservative used in ophthalmic drops is benzalkonium chloride (BAK; usually between 0.1 and 0.2 mg/mL). This can be quite toxic, and most people using drops containing this preservative more than four to six times a day encounter some toxicity. This is particularly a problem in people frequently using tear supplements. This concern has led to the development of both unpreserved unit dose lubricants and newer, less toxic preservatives such as purite and sodium perborate. Toxicity may also be exacerbated if the drop is applied during contact lens use as both the drug and the preservative can accumulate within the matrix of the lens, leading to prolonged exposure. Aside from the preservative, some drugs are more associated with surface toxicity than others. These include neosynephrine, dexamethasone alcohol, antiviral agents such as acyclovir ointment, and some anti-glaucoma medications such as pilocarpine and apraclonidine. Topical anesthetic abuse is a particularly potent form of drop toxicity leading to a persistent epithelial defect, stromal inflammation, and stromal lysis.

SYMPTOMS

Patients usually report stinging and burning of the affected eye or eyes and often have blurred vision if the corneal epitheliopathy is sufficiently severe. The eye is red and inflamed, and there may be photophobia and a watery discharge.

SIGNS

The ocular surface is inflamed with conjunctival injection. The corneal and conjunctival epithelium is incompetent with widespread superficial punctate staining. Fluorescein shows late leakage into and under the epithelium due to a breakdown of the normal cell–cell barrier function. There may be a slight ptosis. There is usually a papillary conjunctivitis, although sometimes a few follicles may be present in established cases. Drug toxicity can delay the healing of a pre-existing epithelial defect, leading to an indolent ulcer. Occasionally, the toxicity is so severe it can even lead to an epithelial defect or a filamentary keratopathy.

INCIDENCE

Common in people on multiple topical medications for more than a few weeks.

SIGNIFICANCE

Usually only causes ocular surface irritation but can threaten sight by leading to or maintaining an ulcer.

DIFFERENTIAL DIAGNOSIS

Dry eye; Blepharitis; Exposure keratopathy.

SEE ALSO

Acute allergic blepharoconjunctivitis; Persistent epithelial defects.

MANAGEMENT

Cease therapeutics

Surface toxicity may be the cause of a patient's ongoing problem. For this reason any patient presenting with ongoing ocular surface disturbance of unknown cause and frequently using topical medication should, as a first step, cease all medication (only on the advice of the prescribing practitioner). This must be accompanied by a thorough explanation

of why it is important to do this, so as to enhance compliance. If there are any medications that must be continued, preservative-free formulations should be substituted if available. If the patient is thought to be reacting to a vital anti-glaucoma medication, this should be changed to a different class of medication. It takes a few weeks for symptoms to improve, and a month or two to resolve. With toxic keratopathy, it is important the patient understands he/she was not allergic to the drug but that the medication was over-used.

Contact lens wear modification

If toxicity is related to contact lenses, a disinfection system, a fresh pair of lenses, and a disinfection solution based on peroxide system should be introduced. If unsuccessful, changing to daily disposable lenses is often helpful.

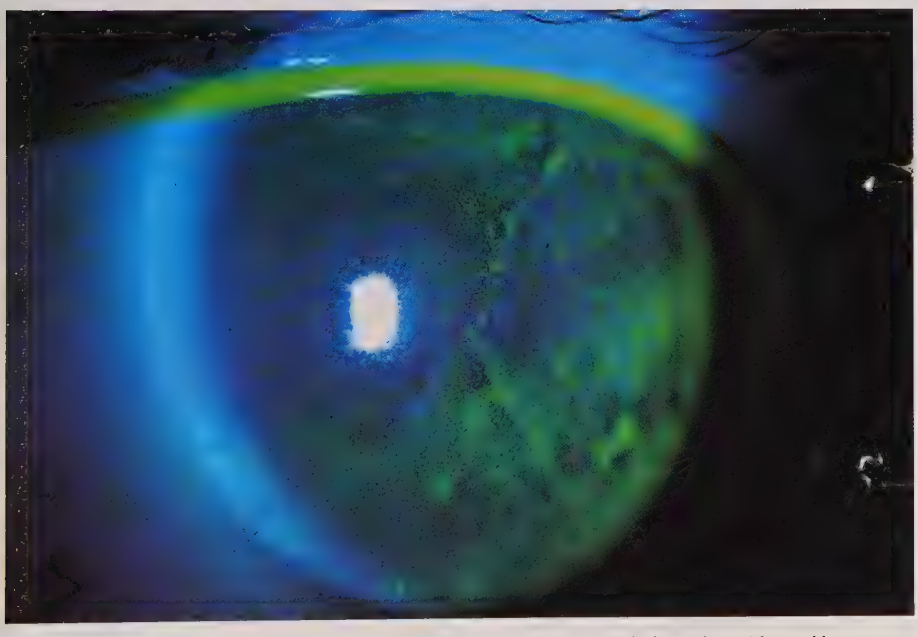


Fig. T-3 Typical changes of drug toxicity with widespread corneal epitheliopathy evidenced by punctate fluorescein staining.

Trachoma

Sandy blight

DESCRIPTION

Active trachoma is a chronic inflammatory, follicular conjunctivitis caused by infection with the bacterium *Chlamydia trachomatis*. Chronic active trachoma as a child can lead to adult scarring (cicatricial) trachoma, with extensive scarring of the conjunctiva and resultant upper eyelid shortening, distortion, and trichiasis. The epidemiology of trachoma is of immense importance, as it is the most common cause of preventable blindness in the world. It is estimated that approximately 150 million people worldwide have active trachoma, while approximately 5.5 million people are blind or at risk of blindness from cicatricial trachoma. Active trachoma almost exclusively affects the young and is a disease of people living in poor hygiene conditions. The chronic infection–reinfection process is probably caused by both the common fly and direct patient-to-patient contact. It can affect people in any geographical region but is prevalent in areas of Africa, India, the Middle East, southwest Asia, parts of Central and South America, and aboriginal communities in Australia. In developed countries, patients with evidence of old active trachoma almost exclusively have grown up in these areas.

SYMPTOMS

Symptoms of active trachoma are of recurrent conjunctivitis. Scarring trachoma

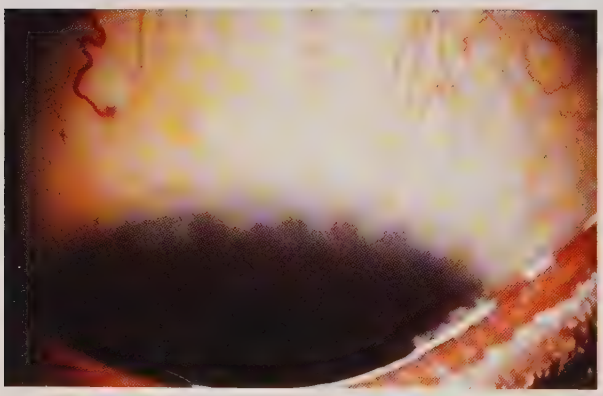


Fig. T-4a Herbert pits seen with inactive trachoma.

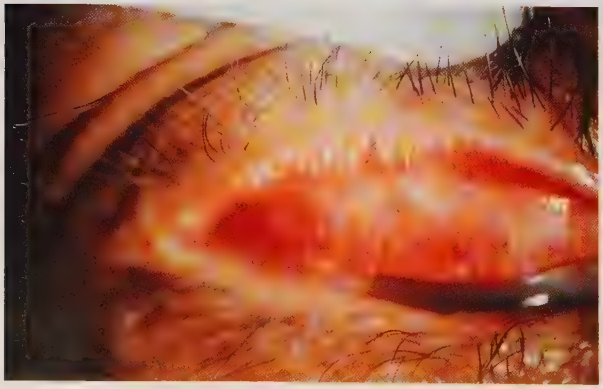
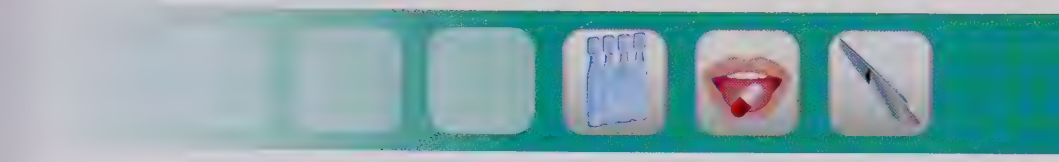


Fig. T-4b Superior tarsal scarring.



leads to ocular surface irritation, with foreign-body sensation from trichiasis, and dry eye and blurred vision from corneal opacities.

SIGNS

Active trachoma leads to a mainly superior tarsal conjunctival follicular response. There are often associated corneal signs, with superior pannus formation and central or peripheral corneal infiltrates. Trachoma was originally graded using the MacCallum classification, but this has been largely superseded by the World Health Organization (WHO) grading scale. This follows the disease from the active follicular stage through to conjunctival scarring and corneal opacification. It was developed for use by trained non-specialist personnel to allow the collection of epidemiological data:

- TF (trachomatous inflammation – follicular) – five or more follicles on superior tarsus
- TI (trachomatous inflammation – intense) – pronounced inflammation of the superior tarsal conjunctiva obscuring at least one-half of the normal deep vessels
- TS (trachomatous scarring) – superior tarsal scarring
- TT (trachomatous trichiasis) – at least one lash touching the ocular surface
- CO (corneal opacity) – easily visible central corneal opacity.

This grading scale may be remembered by the mnemonic "FISTO."

Specific signs of scarring trachoma include Arlt's line, a horizontal linear scarring near the lid margin of the superior tarsal conjunctiva, and Herbert pits, focal round depressions at the superior limbus from resolved limbal follicles.

INCIDENCE

Occurs in endemic areas in poor people with limited access to hygiene.

SIGNIFICANCE

Can lead to bilateral blindness.

DIFFERENTIAL DIAGNOSIS

Other causes of conjunctival cicatrization; for active trachoma, other causes of recurrent conjunctivitis, including bacterial infection.

SEE ALSO

Conjunctival scarring – overview; Conjunctivitis – overview; Chlamydial conjunctivitis; Ophthalmia neonatorum.

MANAGEMENT

The management of trachoma can be summarized by the WHO SAFE strategy: **S**urgery for trichiasis, community **A**ntibiotic programs, **F**acial cleanliness, and **E**nvironmental health.

Active trachoma

Active trachoma is essentially a public health problem and needs to be managed at this level; better hygiene, with access to clean water for such measures as face washing, is the main measure to implement. Fly reduction programs may also play a role in breaking the cycle of infection–reinfection. The specific drug treatment for an acute infection includes a single oral dose of the antibiotic azithromycin.

Scarring trachoma

Patients with inactive scarring trachoma commonly have dry eye symptoms and are aided by frequent topical lubricants. The most important specific measure is protection of the ocular surface by treating any trichiasis. Various methods of treatment have been investigated, with the most effective being bilamellar tarsal rotation surgery. Corneal grafting is rarely performed, even for significant corneal opacification, because of the poor survival rates for grafts in such a hostile environment with significant corneal vascularization and ocular surface disease.

Trichiasis, pseudotrichiasis, distachiasis, metaplastic lashes, and trichotillomania

Eyelash misdirection

DESCRIPTION

If eyelashes are misdirected posteriorly, they may rub against the globe, particularly the cornea, causing ocular surface damage.

There are several possible causes:

- Trichiasis is the common acquired form of eyelash misdirection. It can arise from scarring of the lid margin, causing misdirection of the lash follicles. Possible conditions include chronic blepharitis, herpes zoster ophthalmicus, trachoma, and basal cell carcinoma. However, trichiasis can also be idiopathic.
- Pseudotrichiasis refers to eyelash misdirection due to entropion. In other words, the eyelash follicles are normal, but the eyelid as a whole is misdirected and not in proper apposition to the globe. May be due to Stevens–Johnson syndrome, ocular pemphigoid, chemical burns, and other causes of conjunctival scarring.
- Distachiasis refers to the abnormal presence of a second row of lashes, located posterior to the mucocutaneous lid margin, near the meibomian gland orifices.
- Other conditions. Metaplastic lashes arise from the meibomian gland orifices. These can be due to late-stage cicatrizing conjunctivitis. Trichotillomania refers to an unconscious neurotic tendency to twist or twirl eyelashes or eyebrows.

SYMPTOMS

Regardless of the cause, the symptoms are usually the same. The patient will complain of ocular irritation or foreign-body sensation, worsened with blinking. In addition, there may be tearing and a red eye. Symptoms may be bilateral or unilateral, and the patient may be able to localize the sensation to a particular part of the eye(s).

SIGNS

The lashes abrading the ocular surface may at times be very fine and difficult to see, particularly in elderly patients. However, the changes to the ocular surface may point to the location of the trichiasis. There may be punctate epithelial erosions in one quadrant of the cornea, highlighted with fluorescein dye. Robust eyelashes can cause staining in a linear or irregular foreign-body pattern. If only the conjunctiva is affected, examination of surface staining with fluorescein will require both cobalt blue light and preferably a yellow filter to give adequate contrast.

More severe chronic ocular surface changes related to trichiasis include ulceration and pannus formation. The adjacent bulbar conjunctiva will be hyperemic and thereby indicate the affected area of cornea. In addition, the everted palpebral conjunctiva should be inspected for any foreign body.

INCIDENCE

Idiopathic or blepharitis-related trichiasis is very common (greater than 1/10) in the older patient group.

SIGNIFICANCE

The significance to the patient can be quite high due to the chronic persistent irritation. In chronic cases, and if corneal sensitivity is reduced, there can be a moderate threat to sight due to corneal ulceration or pannus formation.

DIFFERENTIAL DIAGNOSIS

Entropion; Foreign bodies – conjunctival and corneal.

SEE ALSO

Blepharitis; Ocular cicatricial pemphigoid (OCP); Stevens–Johnson syndrome; Conjunctival scarring.

MANAGEMENT

Consulting room procedures

Epilation with cilia forceps is the most common management. It may take several visits to locate all of the lashes, as well as deal with emerging ones.

Review
Since recurrence is common, review is important. In the elderly, corneal sensitivity may be reduced and in some cases the patient may not be aware of (or simply be tolerating) mild ocular surface trauma. Regrowth of lashes may take 1 to 2 months in adults.

Lid hygiene
Recommend daily lid hygiene measures if blepharitis is contributing to the trichiasis.

Therapeutic (bandage) contact lens
This may assist with ocular surface protection in the short term or assist in confirming the diagnosis if lashes cannot be found.

Therapeutics
Broad-spectrum antibiotic cover or tear supplements may be prescribed for corneal ulceration.

Surgical
Permanent surgical removal may be achieved with various methods, including electrolysis, laser ablation, cryotherapy, and incisional operations, for lashes resistant to other forms of treatment.

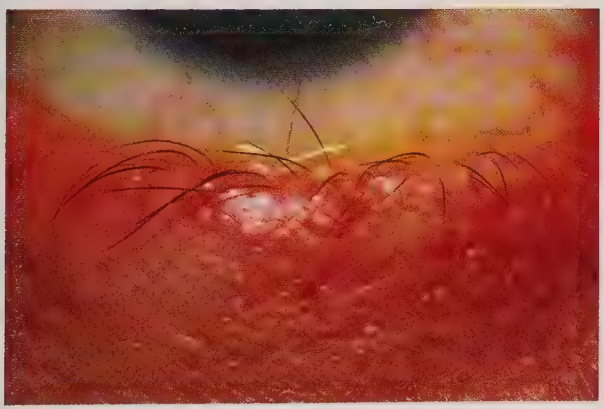


Fig. T-5a Trichiasis, showing fine aberrant lashes. Such lashes can sometimes be difficult to detect without careful examination under the slit lamp.



Fig. T-5b Pseudotrichiasis with associated corneal ulcer.

Uveitis: anterior – overview

Iritis, iridocyclitis

DESCRIPTION

Inflammation of the uveal tract. Anterior uveitis may be subdivided into:

- Iritis, inflammation affecting the iris. This is the most common type.
- Iridocyclitis, in which the ciliary body is also affected.

Causes of anterior uveitis include:

- External injury to the uvea or other external agents.
- Systemic diseases such as sarcoidosis. In addition, 25% of iritis is associated with a histocompatibility antigen (HLA-B27) positive systemic condition such as ankylosing spondylitis, Reiter's syndrome, psoriatic arthritis, or inflammatory bowl disease (Crohn's disease, but not celiac disease).
- Infections such as tuberculosis, candida, herpes zoster, toxoplasmosis.
- Idiopathic specific uveitis, such as Fuchs' uveitis syndrome.
- Idiopathic non-specific (about 25%).

SYMPTOMS

Acute uveitis is characterized by ocular pain, a red eye, and photophobia. There may be a history of recurrent episodes. Chronic anterior uveitis is associated with exacerbations and remissions, but with less marked inflammatory episodes.

SIGNS

The two critical signs are cells and flare in the anterior chamber.

- Aqueous cells. Cells in the aqueous are observed with a high intensity 1-mm × 3-mm slit-lamp beam. With anterior uveitis the number of cells in the aqueous greatly exceeds the number in the anterior vitreous. Aqueous cells may be graded as follows:
  - —1+ 5–10 cells
  - —2+ 11–20 cells
  - —3+ 21–50 cells
  - —4+ more than 50 cells
  - —Hypopyon
- Aqueous flare. Aqueous flare represents serum proteins that have leaked from damaged iris blood vessels into the aqueous, causing a clouding of the aqueous. It may be graded as:
  - —1+ trace, barely detectable
  - —2+ mild, with iris details clear
  - —3+ moderate, with iris details hazy
  - —4+ severe, with exudate
- Keratic precipitates (KP). These may be graded by size into:
  - —fine (suggesting acute uveitis)
  - —medium size (acute, or chronic uveitis)
  - —large "mutton fat" (typically in granulomatous conditions).

  The coloration of KP varies from white (if fresh) to pigmented or translucent (if old).
- Other signs.
  - —Ciliary injection (or ciliary flush): a circumcorneal hyperemia of the perilimbal blood vessels.
  - —Iris nodules; which can occur in granulomatous inflammation.
  - —Posterior synechia; adhesions between anterior lens and iris.
  - —Intraocular pressure is usually reduced, although is occasionally high, especially with herpetic conditions.

INCIDENCE

Uncommon (approximately 1/1000).

SIGNIFICANCE

Acute anterior uveitis can cause severe discomfort and pose a moderate threat to sight if left untreated. It requires prompt investigation and treatment.

DIFFERENTIAL DIAGNOSIS

See causes listed above. Fuchs' heterochromic iridocyclitis; Iritis; Glaucoma – acute angle closure.

SEE ALSO

Endothelial opacities (keratic precipitates); Iritis; Hypopyon; Synechia; Endophthalmitis.

MANAGEMENT

Blood test

While anterior uveitis may be idiopathic, all patients are investigated with a systemic

work-up, to evaluate for a possible systemic basis or association. Possible associations include inflammatory bowel disease, psoriatic arthritis, and Behçet's disease. If recurrent, granulomatous, bilateral, or posterior involvement is present, appropriate blood tests could include: HLA-B27 (human leukocyte antigen B27, testing for e.g. HLA-B27 uveitis without systemic disease), ESR (erythrocyte sedimentation rate, testing for e.g. ankylosing spondylitis, Reiter's syndrome), CXR (chest X-ray, e.g. TB or sarcoid), ACE (angiotensin-converting enzyme, test for sarcoidosis), FBE (full blood examination, any infection, e.g. TB), Mantoux test (for TB), ANA (antinuclear antibody, test for juvenile arthritis).

Ocular investigations

A posterior segment examination should be made for signs of inflammation related to uveitis or endophthalmitis.

Therapeutics

Cycloplegic, e.g. cyclopentolate 2% tid, topical steroid, e.g. prednisolone acetate 1% q1–6 hourly depending upon degree of inflammation. Dose is tapered.

Systemic medication

Very rarely, systemic steroids or periocular steroids may be required if the uveitis does not respond to q1h steroids.

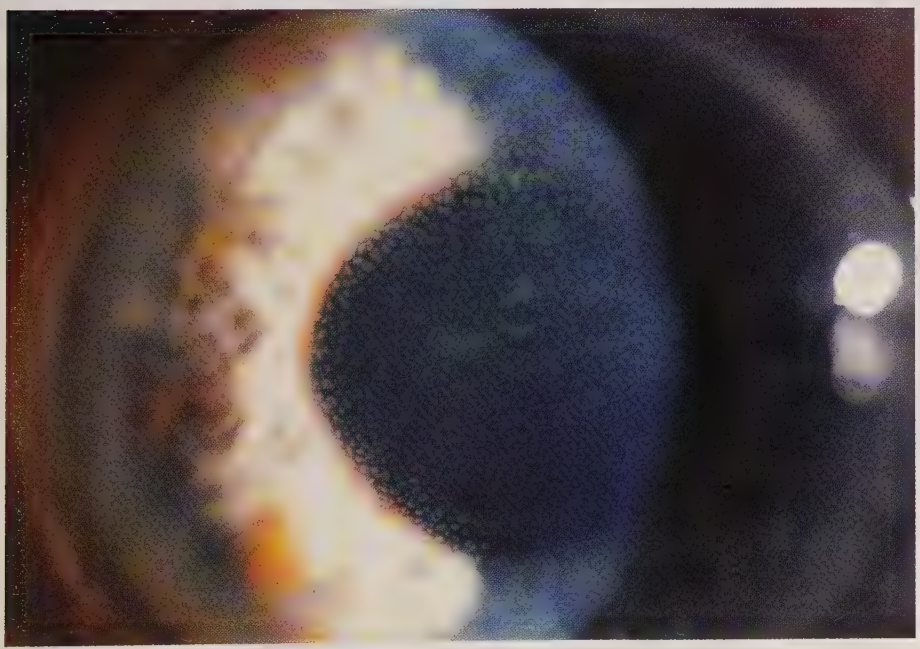


Fig. U-1 Endothelial dusting in acute anterior uveitis.

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Systemic medication

Very rarely, systemic steroids or periocular steroids may be required if the uveitis does not respond to q1h steroids.

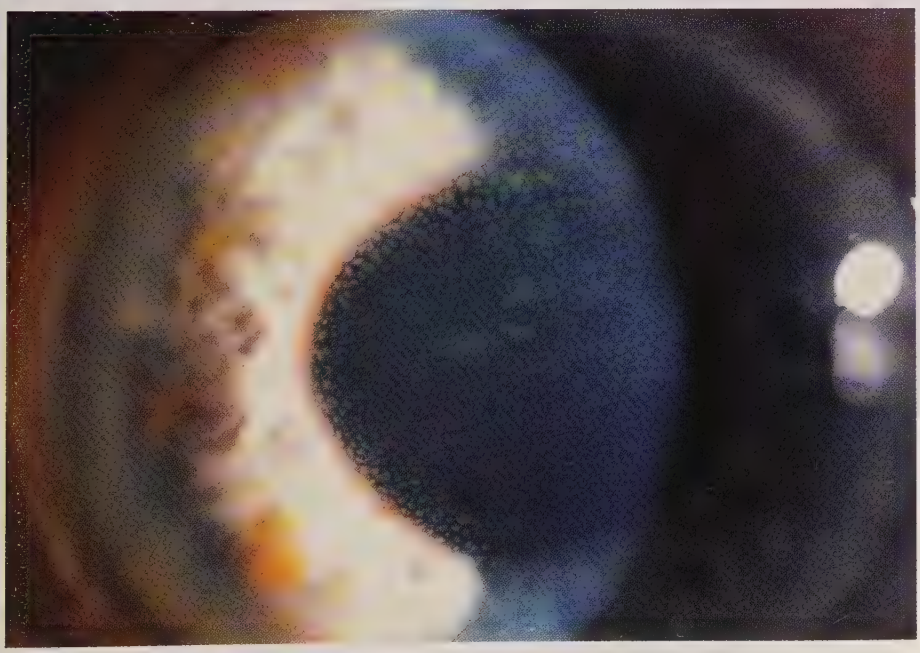


Fig. U-1 Endothelial dusting in acute anterior uveitis.

V

Vernal keratoconjunctivitis (VKC)

Spring catarrh

DESCRIPTION

Vernal keratoconjunctivitis (VKC) is a potentially severe ocular surface allergic condition seen exclusively in children and young adults, more common in boys. While most VKC primarily affects the superior tarsal conjunctiva (palpebral vernal) there is a less-common subtype that predominantly affects the limbus (limbal vernal). The latter is more common in people of African descent. The severity of the disease usually decreases over time, with total resolution in the teens being common. Unfortunately, in a significant proportion of patients, it evolves into atopic keratoconjunctivitis (AKC) as they become adults. The ocular inflammation is an IgE cell-mediated response, relying on mast cell degranulation and histamine release. It often has a seasonal component with exacerbations in early spring to mid-summer. Patients frequently have other allergic conditions such as hay fever, asthma, and, in particular, eczema. There may be a family history of atopy.

SYMPTOMS

VKC leads to intense bilateral itch, intermittent redness, increased lacrimation, and a foreign-body sensation. Mucous strands are also common with active disease and can be recovered from the inferior fornix. If the eyelids are chronically inflamed, a slight ptosis may develop. The presence of a shield ulcer leads to blurring of vision and marked photophobia.

SIGNS

Mild VKC leads to diffuse hyperemia with a velvety, papillary conjunctivitis. With more marked involvement, the superior tarsal conjunctiva develops large (up to 5-mm diameter) papillae with a cobblestone appearance. If there is active inflammation, the papillae are swollen and pink, with the intervening spaces between the papillae filled with a frothy, mucous exudate secondary to histamine activity. When uninflamed, the papillae are flat and whiter, secondary to extensive subepithelial fibrosis. In limbal vernal there is a fleshy white-to-pink, often relatively discrete, nodule at the limbus. In active disease Tranta's dots, small chalky white accumulations of eosinophils, may be present. The corneal epithelium is also affected in active disease, probably as a result of mechanical damage from tarsal changes and toxicity from pro-inflammatory mediators. Signs develop from grouped, usually superior, superficial punctate erosions through to frank ulceration – a shield ulcer. Shield ulcers are usually (but not always) superior vertically oval, and have a rolled indolent border.

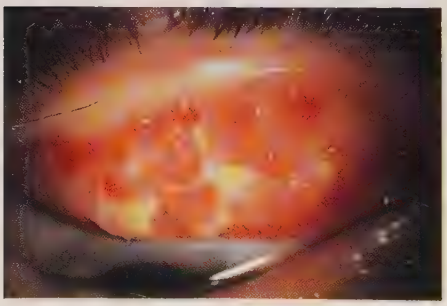


Fig. V-1a Giant papillae with an associated frothy mucus discharge involving the superior tarsal conjunctiva in active VKC.

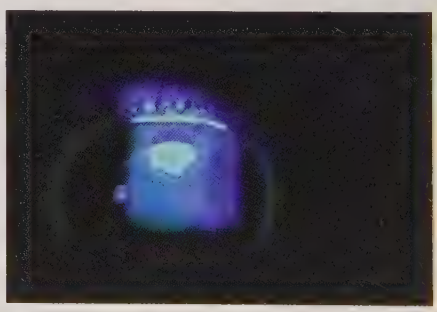


Fig. V-1b Fluorescein dye showing a superior shield ulcer in a young boy with VKC.



A mucous plaque may form at the base of the ulcer. Significantly, this is composed of a proinflammatory and angiogenic protein termed eosinophilic major basic protein. This stains intensely with rose bengal and is a significant impediment to healing of the ulcer.

INCIDENCE
Rare.

SIGNIFICANCE
Leads to significant ocular discomfort and irritation. Shield ulcers may lead to corneal scarring.

DIFFERENTIAL DIAGNOSIS
Atopic keratoconjunctivitis; Giant papillary conjunctivitis.

SEE ALSO
Allergic eye disease – overview; Seasonal allergic conjunctivitis; Conjunctivitis – overview; Follicular conjunctivitis; Persistent epithelial defect.

MANAGEMENT
An important principle of management is to primarily treat the corneal disease, not the conjunctivitis. It is not necessary to fully settle any conjunctival inflammation, as to do so may involve the excessive use of topical corticosteroids.

Allergen avoidance
Apart from the specific drug treatments listed below, allergen avoidance is an important component of treating VKC. Patients allergic to pollens should be counselled to avoid outdoor activities on high pollen-count days. Cleaning around the eyes with a face washer, flushing of the ocular surface with topical lubricants, and the application of ice packs to the closed eyelids can also significantly reduce symptoms.

Therapeutics
Topical antihistamines and non-steroidal anti-inflammatory agents are quite useful in reducing ocular itch and discomfort. However, they have no effect in controlling the underlying disease process. Topical mast-cell stabilizers are a mainstay of treatment. They need to be taken prophylactically 3 to 4 times a day on a regular basis, especially during the allergy season. They may be started prophylactically at the end of winter in anticipation of a flare-up over spring.

Corticosteroids, usually topically although occasionally by injection into the upper eyelid, are used to both suppress exacerbations of the disease and intensively heal a shield ulcer during an acute flare-up. When used over the long term it is imperative to check the intraocular pressure despite the increased difficulty of doing this in some children.

Although not currently available as a proprietary medication, topical cyclosporin A (1% or 2%) has been shown to be very useful.

Surgery
Rarely, a superficial keratectomy or stripping of a mucous plaque is required to promote healing of a shield ulcer. An excimer laser phototherapeutic keratectomy has also been reported for this purpose but appears to be of no advantage over surgical removal.

Vitamin A deficiency

Hypovitaminosis A, xerophthalmia

DESCRIPTION

Vitamin A deficiency is a significant cause of morbidity and mortality in developing countries worldwide. It can lead to the ophthalmic condition of xerophthalmia and is the leading cause of blindness among children in developing countries, accounting for thousands of new cases of blindness worldwide each year. Vitamin A is a fat-soluble vitamin stored in the liver. It occurs naturally in fish (especially cod liver), dairy products, egg yolk, tomatoes, and many other vegetables in precursor form as carotene. It normally takes many years for body stores to be depleted and, as such, deficiency does not occur in very young children born with adequate body stores. Onset of the disease is usually subacute over weeks to months, although may occur acutely if there is an associated febrile illness, typically measles, tuberculosis, or diarrheal disease. Children with vitamin A deficiency are at a very high risk of death, probably due to their marked susceptibility to infection.

Vitamin A deficiency may also rarely occur in people living in developed countries secondary to chronic liver disease, gut malabsorption syndromes, or self-imposed dietary restrictions. The most common causes are alcoholic liver disease with associated poor diet, cystic fibrosis, sprue, and severe bowel disease.

SYMPTOMS

Vitamin A is essential for the generation of the visual pigments and, therefore, deficiency leads to night blindness (nyctalopia), which is often one of the first symptoms. Visual loss increases with time, as do ocular surface irritation secondary to dry eye and epithelial keratinization.

SIGNS

Aside from night blindness, a reduction in visual acuity and scotomas are common. The whole conjunctiva (and later the cornea) becomes lusterless due to progressive keratinization and goblet cell loss. A Bitot's spot forms temporarily, a hyperkeratotic plaque colonized by the bacterium *Corynebacterium xerosis*. It appears as a foamy, silvery-gray plaque.

Corneal involvement is frequently bilateral and results in keratomalacia with stromal opacification epithelial defects and eventually perforation, usually inferiorly. If healed, they may cause an adherent leukoma.

INCIDENCE

Very rare in developed countries but a major cause of blindness worldwide.

SIGNIFICANCE

Acutely sight- and life-threatening.

SEE ALSO

Dry eye syndromes; Perforation – corneal.

MANAGEMENT

Blood tests

Serum vitamin A measurement is notoriously unreliable, in part due to the unstable nature of vitamin A, with regard to both UV light exposure and heat. Samples, if taken, need to be shielded from the light, by wrapping the tube in aluminum foil, and from heat, by storing in a fridge until tested. Aside from vitamin A, serum retinol-binding protein, albumin, and prealbumin are also reduced.

Oral medication

Treatment is with oral vitamin A 200,000 Units stat followed by another dose in 2 days and 1 week. Improvement in symptoms and signs usually occurs within days to weeks. The Bitot's spot may rarely persist despite treatment.



Fig. V-2a Corneal ulceration and stromal loss in an elderly lady with vitamin A deficiency secondary to chronic alcoholic liver disease and a poor diet.

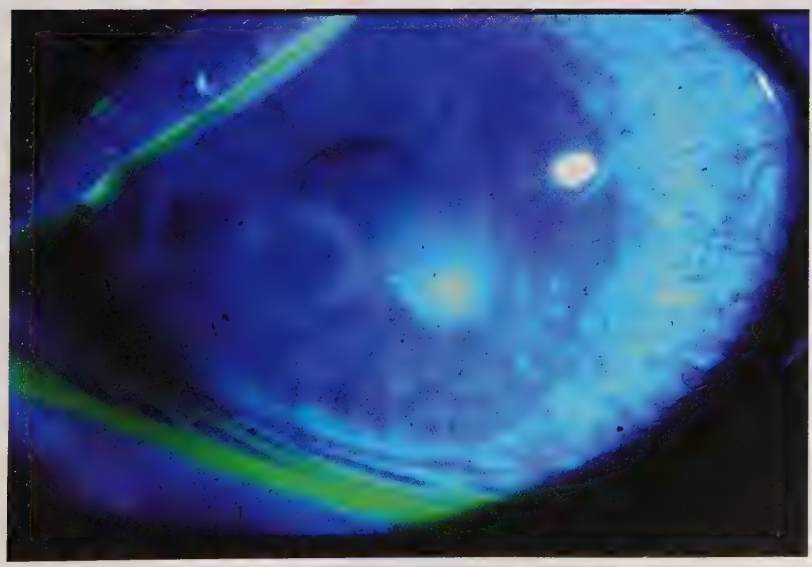


Fig. V-2b Conjunctival changes with formation of a Bitot's spot due to foamy keratinization of the conjunctival epithelium.

Vortex keratopathy

Corneal verticillata, amiodarone keratopathy, chloroquine keratopathy, Fabry disease

DESCRIPTION

Vortex keratopathy is due to deposition of a substance (usually a drug) from the tear film or limbal vasculature into the corneal epithelium. Reversible binding of the drug appears to occur within the epithelial cells. The vortex pattern appears to be caused by tear film circulation, and the resulting pattern of deposition varies somewhat between patients. The spiral epithelial patterns are also known as verticillata.

Causes include:

- Chloroquine and hydroxychloroquine. These drugs are used as antimalarial, amebicidal, and anti-inflammatory agents, and in the treatment of collagen vascular diseases, e.g. lupus erythematosus and rheumatoid arthritis. Patients may develop a range of systemic and ocular reactions including difficulty in accommodation, blurring of vision, corneal opacities, and, less often, retinal degeneration. Keratopathy may not occur with 400 mg/day hydroxychloroquine but occurs at 800 mg/day in most patients after several years. Keratopathy is more common with chloroquine.
- Amiodarone. This drug is used in severe cases of cardiac tachyarrhythmias not responding to other therapy. Patients are usually monitored for possible systemic and ocular toxicity during the course of therapy and for a period after discontinuation. Amiodarone keratopathy is common at high doses (400–1400 mg/day).
- Indomethacin. This non-steroidal anti-inflammatory is used to treat a range of conditions, including arthritis and gout.
- Fabry disease (sphingolipidosis). This disease is due to an inborn error of glycophospholipid metabolism that, being X-linked, primarily affects males. The ocular signs include corneal opacity, vortex keratopathy, and conjunctival and retinal vascular abnormalities. The corneal signs may also occur in female carriers of the disease. Rather than a deposition, the vortex pattern is due to opaque epithelial cells. A specific intravenous enzyme replacement therapy is available to prevent progression of the disease.

SYMPTOMS

Usually asymptomatic although, rarely, the patient may report visual disturbances such as blurring of vision, halos around lights, and photophobia.

SIGNS

Symmetrical, bilateral, grayish, or golden corneal deposits occur in a swirling pattern, concentric about a point below the pupil.

There are considered to be at least three stages of vortex keratopathy:

- Stage 1 – coalescence of fine punctate, grayish golden-brown opacities into a horizontal linear pattern in the inferior cornea. This line is similar to a Hudson–Stahli line and is located at the basal epithelial level.
- Stage 2 – additional arborizing and horizontal lines, extending toward the limbus.
- Stage 3 – with the extension of vertical lines, a verticillate, whorl-like pattern develops.

There appears to be no relation between the keratopathy of chloroquine/hydroxychloroquine therapy and the development of retinopathy. Possible retinal changes include abnormal macular pigmentation and depigmentation (sometimes described as a "bull's eye"), pallor of the optic disc, optic atrophy, and narrowing of the retinal arterioles.

INCIDENCE

In patients taking these systemic medications at high doses for long periods, vortex keratopathy is very common (more than 1/10).

SIGNIFICANCE

Corneal opacities usually disappear completely if these drugs are stopped. However, the presence of keratopathy in hydroxychloroquine therapy or Fabry disease may highlight the need for additional investigations (see "Management").

DIFFERENTIAL DIAGNOSIS

Corneal dystrophies, degenerations, and depositions; Iron lines (Fleischer ring, Stocker's line, Hudson–Stahli line, Hemosiderosis); phenothiazine-related drugs, including chlorpromazine (antipsychotic drugs), cause general corneal endothelial pigmentation (see "Endothelial opacities"); Crystalline keratopathy.

MANAGEMENT

Additional investigations

Routine fundus examinations may detect retinal changes, especially pigmentary, due to hydroxychloroquine at an early and reversible stage when there is no apparent visual disturbance. Similarly, the presence of keratopathy in Fabry disease may highlight the need for systemic assessment.

Oral medication

Modify the level of medications, if symptoms or signs require it, and if the management of the systemic condition allows it. With hydroxychloroquine, any retinal changes or the appearance of scotoma, night blindness, or similar requires immediate discontinuation of the drug on the advice of the prescribing clinician; these patients should not subsequently receive any pharmacologically similar drugs.

Review

All patients on chloroquine and hydroxychloroquine should have an initial eye examination, repeated at least every 12 months during therapy. Fundus and visual field assessment should be performed for these drugs. High doses – 6 mg/kg ideal (lean) body weight per day – may indicate the need for more frequent review. Other indications include significant renal or hepatic impairment, complaints of visual disturbances, or treatment duration exceeding 8 years.



Fig. V-3 Vortex keratopathy in a 72-year-old male patient.

Xanthelasma

Hypercholesterolemia, xanthoma

DESCRIPTION

Xanthelasma is the most common form of cutaneous xanthoma, characterized by yellow subcutaneous plaques in the periorbital area. Histologically, these plaques are accumulations of foamy histocytes in the superficial layers of the dermis. Cholesterol from the blood is thought to accumulate within the xanthelasma, often due to anomalous blood cholesterol or lipoprotein levels. Rarely, it occurs due to an uncommon autoimmune disease, primary biliary cirrhosis, predominantly occurring in middle-aged women. The lesions are soft because of the lack of significant fibrosis.

SYMPTOMS

The lesions do not usually cause any symptoms, other than possibly some cosmetic concern.

SIGNS

The soft yellowish plaques are usually bilateral and on the medial aspect of the superior eyelids, as well as on the skin inferior to the inner canthi. The plaques follow the lines and folds of the skin in each area. They vary from 2–30 mm in length and are frequently symmetrical, with a tendency to be permanent and progressive.

INCIDENCE

Common (approximately 1/100), particularly in women and in older age groups.

SIGNIFICANCE

Patients with xanthelasma are at risk of premature cardiovascular disease if there is an underlying blood disorder. Plaques on other areas of the body may be indicative of diffuse planar xanthoma, a condition with serious possible systemic associations including multiple myeloma.

DIFFERENTIAL DIAGNOSIS

Skin tumors – benign (viral wart, papilloma, cutaneous horn, keratoacanthoma); Seborrheic keratosis; Actinic keratosis; Basal cell carcinoma; Squamous cell carcinoma; Cysts of the eyelid.

MANAGEMENT

Systemic

Treatment involves management of high cholesterol, if present. This would require professional dietary advice and oral medication. If there are any symptoms of obstructive liver disease, such as jaundice or itch, investigations for primary biliary cirrhosis are indicated.

If plaques are not limited to the periorbital area, but are found elsewhere on the body, a biopsy may be indicated for confirmation of diagnosis. In this situation, a more comprehensive systemic work-up for associated conditions is important.

Surgery

Surgical excision is the most widely used method for removal of the lesions, although alternatives include treatment with topical 35% trichloroacetic acid or use of the carbon dioxide laser. Patients should be aware that recurrence is possible. Surgical removal of large lesions may present issues in terms of avoiding changes in the habitual eyelid position.



Fig. X-1 Xanthelasma, showing the soft yellowish plaques superior to the inner canthi. Note the way the lesion is aligned with the contour of the tissues.

Appendix A: Therapeutics

Disclaimer
The following is a summary of some of the key points regarding topical ocular medications. For full details, consult the package inserts accompanying each product (or see the manufacturer's web sites), drug guides such as *MIMS*, or online Internet guides (e.g. DailyMed). Note that drug availability varies markedly between countries.

Overview
1 Antibiotics (antibacterials)
2 Antivirals
3 Antifungals (antimycotics)
4 Anti-amebics
5 Anti-glaucoma medications
6 Corticosteroids
7 Corticosteroid/antibiotic combinations
8 Non-steroidal anti-inflammatory drugs (NSAIDs, NSAIAs)
9 Cyclosporin A
10 Mast-cell stabilizers
11 Antihistamines
12 Vasoconstrictors
13 Ocular lubricants
14 Mydriatics and cycloplegics
15 Anesthetics
16 Stains
17 Aspects of the autonomic nervous system relevant to topical ocular medications

1 ANTIBIOTICS (ANTIBACTERIALS)
Contraindications
Risks of topical antibiotic usage include allergic reactions, toxicity, and the potential for the development of resistant strains of bacteria. For these reasons, the routine use of antibiotics for viral or allergic conjunctivitis is contraindicated.

Caution
Use in pregnancy, lactating mothers, and children under 1 year.

Dosage
Official prescribing information for topical antibiotics is available from the manufacturer and from regulatory agencies (e.g. dailymed.nlm.nih.gov). There are a variety of dosage regimens that may be applicable for various drugs and conditions; however, some typical examples are:

- post-surgical prophylaxis – qid for 1 week
- bacterial conjunctivitis – qid for 1 week
- severe bacterial conjunctivitis – 1 drop every 2–4 hours for 2 days; then, if there is improvement, 1 drop 4 times daily for 5 days
- bacterial keratitis – 1 drop every hour day and night for 48 hours, then decrease frequency according to clinical response, typically 2 hourly by day and 4 hourly by night for 2 days then qid for 1 to 2 weeks.

See also under "Dosage" for individual drug classes.

1A BACTERICIDAL ANTIBIOTICS

Quinolones

USES	Bacterial keratitis, severe bacterial conjunctivitis *Note:* Ciprofloxacin may form temporary white precipitates on the ocular surface, especially at the base of an epithelial defect. Ofloxacin is more soluble within the cornea and does not tend to precipitate
PEDIATRIC USE	Approved for use in patients down to age 1 year, except for Iquix, which is approved for 6 years and older
SENSITIVITY	Broad-spectrum antibiotic with some resistant Gram-positive cocci. Fluoroquinolones may be ineffective against infections with methicillin-resistant *Staphylococcus aureus* (MRSA), as shown in the Ocular TRUST study. Fluoroquinolones are widely available for systemic treatment and also used in the food chain in some countries. Alternatives include besifloxacin, a new chloro-fluoroquinolone that is ophthalmic only (no systemic equivalent), and other classes of drugs including vancomycin, trimethoprim, or chloramphenicol (see below)
PREPARATIONS	Ciloxan, 0.3% ciprofloxacin (Alcon) + generics Ocuflox, 0.3% ofloxacin (Allergan) + generics Zymar, 0.3% gatifloxacin (Allergan) Zymaxid, 0.5% gatifloxacin (Allergan) Quixin, 0.5% levofloxacin (Vistakon Pharmaceuticals) Iquix, 1.5% levofloxacin (Vistakon Pharmaceuticals). Unpreserved Vigamox, 0.5% moxifloxacin (Alcon). Unpreserved Besivance, 0.6%, besifloxacin suspension (Bausch & Lomb)
DOSAGE	General guidance is given above; see "Antibiotic Dosage." In addition: • Vigamox has approval for tid dosing for 7 days for bacterial conjunctivitis • Besivance has approval for tid dosing for 7 days for bacterial conjunctivitis. Since it is formulated as a viscous suspension, the Besivance bottle should be inverted and shaken once before use

Bactericidal Aminoglycosides

USES	Bacterial conjunctivitis and for prophylaxis following ocular surgery or ocular surface trauma (abrasions). Fortified preparation used in dual therapy with a cephalosporin for bacterial keratitis
SENSITIVITY	Broad-spectrum antibiotic with some resistant Gram-positive organisms. The aminoglycosides are seldom used systemically, due to issues with absorption and toxicity. The Ocular TRUST study showed aminoglycosides may be ineffective against infections with methicillin-resistant *Staphylococcus aureus* (MRSA)
PREPARATIONS	Genoptic, 0.3% gentamicin drops (Allergan) + generics Fortified gentamicin 1.3% (non-commercial) Tobrex 0.3% tobramycin drops, and ointment (Alcon) + generics Fortified 1.3% tobramycin (non-commercial)
DOSAGE	General guidance is given above, for "Antibiotic dosage"

Vancomycin

USES	Ophthalmic use is reserved for serious infections: infectious crystalline keratopathy, endophthalmitis, microbial keratitis. Only used systemically on-indication, as it is relatively toxic and must be administered intravenously
CLASS	Tricyclic glycopeptide (bactericidal)
SENSITIVITY	Broad Gram-positive coverage, although inactive against Gram-negative bacilli
PREPARATIONS	Vancomycin 2.5% (non-commercial)

Cephazoline (or Cefazolin)

USES	Fortified preparation used in dual therapy with an aminoglycoside for bacterial keratitis, endophthalmitis
CLASS	Cephalosporin (bactericidal)
SENSITIVITY	Broad Gram-positive sensitivity
PREPARATIONS	Cephazoline 5% (non-commercial)

10 BACTERIOSTATIC ANTIBIOTICS

Chloramphenicol

USES	Bacterial conjunctivitis Prophylaxis following ocular surgery or ocular surface trauma (abrasions)
SENSITIVITY	Broad sensitivity in Gram-negatives, and -positives, resistance common
PRECAUTIONS	Patients with a family history of blood disorders, using bone marrow suppressive agents, or needing prolonged topical treatment. See notes
PREPARATIONS	Minims chloramphenicol solution 0.5% (Bausch & Lomb) Chlorsig 0.5% drops, and 1.0% ointment (Sigma) Chloromycetin 0.5% drops, and 1.0% ointment (Pfizer, Parke-Davis) Other generics
NOTES	The safety of topical use of chloramphenicol has been supported by several large-scale scientific studies in the past 15 years. A link between chloramphenicol eye drops and aplastic anemia has not been supported. Systemic (oral) use of chloramphenicol is linked to an increased incidence of aplastic anemia and other potential complications; for this reason, systemic use is rare. Systemic complications relate to toxic derivatives in the gut and excessive prolonged serum concentrations. These issues do not appear to arise for topical ocular treatment in the normal short courses. Regulatory agencies in different countries have different approaches: • In the United States chloramphenicol eye drops are packaged with a warning that it should not be used when alternative treatments are available • In the United Kingdom (2005), and in Australia (2010), as well as in Belgium and Portugal, chloramphenicol eye drops have been classified as non-prescription medicine (Pharmacy-only)
DOSAGE	General guidance is given above for Antibiotic dosage, for bacterial conjunctivitis, and for post-surgical prophylaxis

Azithromycin

USES	Bacterial conjunctivitis, in children down to 1 year
CLASS	Macrolide
SENSITIVITY	*Haemophilus influenzae, Staphylococcus aureus* *Streptococcus mitis* group and *Streptococcus pneumoniae*. The Ocular TRUST study showed azithromycin to be largely ineffective against methicillin-resistant *Staphylococcus aureus* (MRSA)
PRECAUTIONS	The potential for anaphylaxis or other hypersensitivity reactions should be considered in patients with a known systemic hypersensitivity to azithromycin or erythromycin
PREPARATIONS	AzaSite, 1.0% azithromycin solution (Inspire Pharmaceuticals)

DOSAGE	AzaSite is dosed less frequently than other antibiotics. Since it is formulated as a viscous solution, the bottle should be inverted and shaken once before use. The recommendation for bacterial conjunctivitis is 1 drop every 8 to 12 hours for the first 2 days, then 1 drop daily for the next 5 days

Sulfacetamide

USES	Bacterial conjunctivitis, blepharitis
CLASS	Sulfonamide
SENSITIVITY	Broad-spectrum sensitivities, although staphylococcal and pseudomonal resistance common
PRECAUTION	Patients may be hypersensitive/allergic to sulfa-based drugs. A reaction may recur when a sulfonamide is readministered, irrespective of the route of administration. At the first sign of hypersensitivity, skin rash, or other serious reaction, discontinue use of the preparation. Rarely, hypersensitivity reactions are severe
PREPARATIONS	Bleph-10, 10% sulphacetamide sodium solution (Allergan) Cetamide, 10% sulphacetamide sodium ointment (Alcon) Sulf-10, 10% sulphacetamide sodium solution (Novartis) Other generics

Propamidine/dibromopropamidine

USES	Mild bacterial conjunctivitis, particularly in children (limited effectiveness in *Acanthamoeba* keratitis; see also under "4. Anti-amoebics")
CLASS	Diamidine
SENSITIVITY	Broad sensitivity in Gram-negatives and -positives, resistance common
PREPARATIONS	Brolene ointment, 0.15% dibromopropamidine isethionate (Sanofi-Aventis) Brolene Eye Drops, 0.1% propamidine isethionate (Sanofi-Aventis)

1C BACTERIOSTATIC COMBINATION ANTIBIOTICS

Trimethoprim and polymyxin B

USES	Surface ocular bacterial infections, including acute bacterial conjunctivitis and blepharoconjunctivitis, caused by susceptible microorganisms
SENSITIVITY	Broad sensitivity in Gram-negatives and -positives, resistance common
PREPARATIONS	Polytrim (polymyxin B and trimethoprim, Allergan) + generics
DOSAGE	In mild to moderate infections, instill 1 drop in the affected eye(s) every 3 hours (maximum of 6 doses per day) for a period of 7–10 days

Neomycin combinations

USES	Bacterial conjunctivitis and for prophylaxis following ocular surgery or ocular surface trauma (abrasions)
CLASS	Bacteriostatic aminoglycoside
SENSITIVITY	Broad-spectrum antibiotic with some resistant Gram-positive organisms
PRECAUTIONS	Neomycin has potential to cause a type IV delayed hypersensitivity reaction, affecting the treated area
PREPARATIONS	Neosporin Ointment: neomycin 0.5%, polymyxin B & bacitracin Solution: polymyxin B sulfate, gramicidin, neomycin sulfate (Pfizer) Solution: polymyxin B sulfate, bacitracin zinc Framycetin, 0.5% soframycin (Aventis Pharma)

1D TOPICAL ANTIBIOTICS AVAILABLE IN OINTMENT (Also note some antibiotics listed above are available as ointments, as well as eye drops)	
Erythromycin	
USES	Bacterial conjunctivitis and for prophylaxis following ocular surgery or ocular surface trauma (abrasions)
CLASS	Macrolide
SENSITIVITY	Broad sensitivity in Gram-negatives and -positives, resistance common
PREPARATIONS	Erythromycin ointment 0.5% (generic)
Polymyxin B combination	
USES	Bacterial conjunctivitis and for prophylaxis following ocular surgery or ocular surface trauma (abrasions)
CLASS	Macrolide
SENSITIVITY	Broad sensitivity in Gram-negatives and -positives, resistance common
PREPARATIONS	Polysporin, % polymyxin B/bacitracin (Monarch) + generics
Tetracycline	
USES	Bacterial conjunctivitis, inhibition of tissue collagenases. Oral tetracycline/doxycycline is used to treat meibomian gland dysfunction, rosacea keratoconjunctivitis, and chlamydial infections
SENSITIVITY	Broad sensitivity in Gram-negatives and -positives, resistance common
PREPARATIONS	1% ointment (non-commercial)

See also "7 Combination steroid, and antibiotic drops".

2 ANTIVIRALS

Uses

Topical antivirals are indicated for the treatment of acute epithelial herpes simplex keratitis (dendritic ulcers), usually with five times per day dosing. Topical antivirals are also used for prophylaxis (with bid dosing) when topical corticosteroids are used in a patient with herpes simplex keratitis (HSK).

Antivirals may be used in conjunction with debridement of dendritic lesions, to reduce the viral load. Oral antivirals (acyclovir, valaciclovir, and famciclovir) are used for both active herpes zoster ophthalmicus and HSK prophylaxis.

Precautions

All topical antiviral agents are relatively toxic and may lead to a punctate keratopathy (see "Toxic keratopathy").

Acyclovir (or Aciclovir)	
PREPARATIONS	Zovirax, 3% acyclovir ointment (GlaxoSmithKline)
DOSAGE	It is recommended that a 1-cm ribbon of ointment be administered to the inferior fornix 5 times per day or at approximately 4-hourly intervals during waking hours. Treatment should continue for 10 days or at least 3 days after healing is complete, whichever is shorter
Ganciclovir	
PREPARATIONS	Zirgan, 0.15% ganciclovir gel (Bausch & Lomb)

DOSAGE	The newest of the topical antiviral drugs, with lower corneal toxicity. The recommended regimen is 1 drop in the affected eye 5 times per day (approximately every 3 hours while awake) until the corneal ulcer heals (typically 4 to 5 days) and then 3 times per day for 7 days
Trifluridine	
PREPARATIONS	Viroptic, 1% trifluridine solution (Monarch Pharmaceuticals) + generics
DOSAGE	The recommended regimen is 1 drop in the affected eye every 2 hours while awake, for a maximum daily dosage of 9 drops until the corneal ulcer has completely re-epithelialized. Following re-epithelialization, recommended treatment is for an additional 7 days with a minimum of 5 times per day If there are no signs of improvement after 7 days of therapy or complete re-epithelialization has not occurred after 14 days of therapy, other forms of therapy should be considered. Avoid treatment periods exceeding 21 days because of potential ocular toxicity
Vidarabine	
PREPARATIONS	Vira-A, 3% vidarabine ointment (Monarch Pharmaceuticals)
DOSAGE	Apply approximately 1 cm of ointment at the lower lid margin 5 times per day for a week. If the lesion has healed, treat at a lower dosage, such as twice daily, over the next week to prevent recurrence. Other forms of therapy should be considered if healing has not occurred within 3 weeks
Idoxuridine	
PREPARATIONS	Stoxil, 0.5% idoxuridine
DOSAGE	The oldest of the antiviral drugs and superseded by those listed above. Penetration into the eye is poor. Administered hourly

3 ANTIFUNGALS (ANTIMYCOTICS)

Uses

Antifungals are used for fungal keratitis and scleritis. Topical preparations are often supplemented with oral antifungals.

Precautions

All are toxic to the corneal epithelium and lead to superficial punctate keratopathy. Polyenes and imadazoles may antagonize each other and reduce effectiveness if used together.

Amphotericin B			
CLASS	Polyene		
PREPARATION		0.15% (non-commercial)	
Natamycin			
CLASS	Imadozole		
PREPARATION	Natacyn	5%	Alcon
Voriconazole			
CLASS	Triazole		
PREPARATION		1% (non-commercial)	

Flucytosine	
CLASS	Fluorinated pyrimidines
PREPARATION	1% (non-commercial)

4 ANTI-AMEBICS

Both of the drugs listed below are polymeric biguanide antiseptics and are cystocidal and trophozoicidal. Other, less effective, anti-amebic drugs include propamidine and neomycin, both listed under antibiotics.

Phenyl-hexyl-methylene-biguanide (PHMB)	
USE	*Acanthamoeba* keratitis
SENSITIVITY	Resistance is uncommon
PREPARATIONS	0.02% (non-commercial)

Chlorhexidine	
USE	*Acanthamoeba* keratitis
SENSITIVITY	Resistance is uncommon.
PREPARATIONS	1% (non-commercial)

5 ANTI-GLAUCOMA MEDICATIONS

5A Prostaglandin analogs, prostamides, docosanoids, and combination drugs

Overview

This class of anti-glaucoma drugs was introduced in 1996 with latanoprost. They are particularly useful in low-tension glaucoma. More recently, these drugs have been made available with timolol in combination.

Precautions

As these drugs have a pro-inflammatory affect, caution should be exercised in the treatment of patients with a history of uveitis, cystoid macular edema or aphakia, herpes simplex keratitis, or a corneal graft. Latanoprost has been found, in a minority of patients, to cause iris darkening and darker, thicker lashes. A similar safety profile is likely with travoprost and bimatoprost. At this time, unoprostone isopropyl has no known systemic side effects.

Latanoprost			
TYPE	Prostaglandin F_2 alpha compound		
ACTIVITY	Increases aqueous outflow by uveoscleral pathway. Reduces IOP by about 25–30%		
PREPARATIONS	Xalatan	0.005%	Pfizer
	Xalcom	Latanoprost 0.005% and timolol 0.5%	Pfizer
DOSING	Once daily in evenings. Maximum effect is reached in about 2 weeks		

Travoprost			
TYPE	Prostaglandin F_2 alpha compound		
ACTIVITY	Increases aqueous outflow by uveoscleral pathway. Reduces IOP by about 25–30%		
PREPARATIONS	Travatan	0.004%	Alcon
	Travatan Z	0.004%	Alcon
	DuoTrav	Travoprost 0.004%, and timolol 0.5%	Alcon
NOTES	Travatan Z formulation is benzalkonium chloride (BAK) free; it contains the Sofzia preservative system (boric acid, propylene glycol, sorbitol, and zinc chloride), introduced in 2009		
DOSING	Once daily in evenings. Maximum effect is reached in about 2 weeks		

Bimatoprost			
TYPE	Prostaglandin F_2 alpha 1–thanolamide compound		
ACTIVITY	Probably increases aqueous outflow by uveoscleral pathway. Reduces IOP by about 25–30%		
PREPARATIONS	Lumigan	0.03%	Allergan
	Ganfort 0.3/5	Bimatoprost 0.03%, and timolol 0.5%	Allergan
DOSING	Once daily in evenings. Maximum effect is reached in about 2 weeks		

Unoprostone isopropyl			
TYPE	Docosanoid compound		
ACTIVITY	Probably enhances trabecular outflow. Reduces IOP by about 15%. As monotherapy, reduces IOP similarly to betaxolol. As adjunctive therapy, similar effect to beta-blockers, brimonidine, and topical carbonic anhydrase inhibitors		
PREPARATIONS	Rescula	0.15%	Sucampo Pharma
DOSING	Twice daily. Maximum effect is reached in about 1 week		

5B Beta-adrenergic receptor antagonists (beta-blockers)

Overview

Beta-blockers were introduced in 1978 and remain widely prescribed and cost-effective anti-glaucoma medications. Although very effective, the potential for systemic complications is the main contraindication/precaution. They are increasingly being used in combination drugs with prostaglandin analogs and others (see respective sections).

Activity

Decrease aqueous formation by blocking $beta_2$ receptors on non-pigmented ciliary epithelium. Non-selective beta-blockers reduce IOP by about 25%, and betaxolol reduces IOP by about 20%. It is estimated that 10–20% of patients do not respond to beta-blockers.

Contraindications

Asthma, or chronic obstructive pulmonary disease (COPD), significant heart disease, bradycardia, emotional depression.

Precautions

If the patient is taking a systemic beta-blocker, the effectiveness of the topical drug may be reduced. If systemic absorption is excessive, there is potential for central nervous system emotional depression.

Dosing

For the non-selective beta-blockers, first try the 0.25% concentration of these drugs, instilled once daily (qd) in a trial for 2 or 3 weeks. Studies have shown once-daily instillation in the morning is as effective as bid dosing. Immediate occlusion of the tear duct or gentle eye closure for 3 minutes will maximize efficacy and reduce systemic absorption.

Timolol maleate

TYPE	Non-selective beta-blocker
PREPARATIONS	Timoptic (0.25% and 0.5%) Timoptic XE (0.25% and 0.5% in gellan gumbased vehicle) Originally from Merck, are out of patent and available generically, e.g. Timoptol (Merck), Optimol (Alphapharm)

Timolol maleate and pilocarpine

TYPE	Non-selective beta-blocker and miotic
PREPARATIONS	Timpilo, timolol 0.5%, and pilocarpine 2%, or 4% (Merck)

Timolol hemihydrate

TYPE	Non-selective beta-blocker
PREPARATIONS	Betimol, 0.25% and 0.5% timolol hemihydrate (Novartis Ophthalmics)

Levobunolol

TYPE	Non-selective beta-blocker. Longer half-life than timolol
PREPARATIONS	Betagan, 0.25% or 0.5% Levobunolol (Allergan) + generics

Metipranolol

TYPE	Non-selective beta-blocker. Slight risk of uveitis
PREPARATIONS	OptiPranolol, 0.25% and 0.5% metipranolol (Bausch & Lomb)

Carteolol

TYPE	Non-selective beta-blocker, with reduced potential for causing depression or affecting cholesterol or lipid profiles. Reduced stinging
PREPARATIONS	Ocupress, 1% and 2% carteolol (Novartis Ophthalmics) + generics
DOSING	Must be used twice daily, unlike the other non-selective beta-blockers

Betaxolol and levobetaxolol

TYPE	Beta-1 selective adrenergic blocker. Improved safety profile with respect to systemic effect on pulmonary tissues; however, slightly less effective. Betaxolol reduces IOP about 20% and levobetaxolol about 22%
DOSING	Must be used twice daily, unlike the non-selective beta-blockers
PREPARATIONS	Betopic-S, 0.25% suspension (Alcon). As effective as the 0.5% solution, but stings less Betopic, 0.5% solution (Alcon) + generics

5C Alpha-adrenergic agonist (sympathomimetic) drugs

Overview

While non-selective sympathomimetics were used for many years, the selective alpha-2-adrenergic agonists have been introduced since the beta-blockers, with the advantage of an enhanced systemic safety profile.

Activity

Primarily reduce intraocular pressure by suppressing aqueous humor formation, although there may also be some increase in outflow. Alpha agonists reduce IOP by about 20–25%. Dipivefrin is less effective.

Contraindications

Avoid use in pregnancy, lactation (no data available). Avoid use in children as it can cause central hypotension, which could be fatal.

Precautions

Alpha-agonists can potentially cause fatigue, drowsiness, or decreased mental alertness in some patients.

Brimonidine tartrate	
TYPE	Selective alpha-2-adrenergic agonist. A neuroprotective role has been proposed
INDICATIONS	Chronic open angle glaucoma or ocular hypertension when beta-blockers are not tolerated or are contraindicated
CONTRAINDICATIONS	Patients using monoamine oxidase (MAO) inhibitors
PREPARATIONS	Alphagan, 0.2% brimonidine (Allergan) + generics, dosing bid Alphagan P, 0.15% brimonidine (Allergan) + generics, dosing bid Alphagan P, 0.1% brimonidine (Allergan), dosing bid Combigan, brimonidine tartrate 0.2% and timolol 0.5% (Allergan)
DOSING	One drop bid
NOTES	Alphagan P is preserved with Purite, a relatively non-toxic preservative, instead of BAK and is equally effective as Alphagan 0.2%
Apraclonidine	
TYPE	Selective alpha-2-adrenergic agonist. It is limited in application, since its effectiveness may decline after a month, and there is a significant incidence of allergic blepharoconjunctivitis after several months' use
INDICATIONS	Short-term adjunctive treatment, pre/postoperative. Also suited for adjunct treatment of acute angle closure, in conjunction with timolol
CONTRAINDICATIONS	Patients using monoamine oxidase (MAO) inhibitors, systemic sympathomimetics, or tricyclic antidepressants. Hypersensitivity to systemic clonidine
PREPARATIONS	Iopidine, 0.5% apraclonidine (Alcon) + generics Iopidine, 1% apraclonidine (Alcon)
DOSING	One drop bid–tid
Dipivefrin	
TYPE	Non-selective adrenergic agonist. It largely replaces the original adrenaline (epinephrine) eye drops. It is a pro-drug, which is rapidly metabolized to adrenaline by the cornea, and with greater penetration and fewer side effects than adrenaline

ACTIVITY	Limited role due to poor efficacy and frequent ocular adverse effects. Reported complications include allergic blepharoconjunctivitis and toxic follicular conjunctivitis in long-term use, rebound conjunctival injection, pigmented conjunctival deposits, mydriasis, and frontal headache
CONTRAINDICATIONS	Narrow anterior angles or closed-angle glaucoma, since mydriasis may provoke an intraocular pressure rise. Aphakia, since there is a 30% incidence of cystoid macular edema
PREPARATIONS	Propine, 0.1% dipivefrin (Allergan) + generics
DOSING	One drop bid

5D Carbonic anhydrase inhibitors (CAIs)

Overview

Can be used both systemically and topically to reduce IOP. Use inhibits bicarbonate production in the ciliary body epithelium, reducing the formation of aqueous humour. They are used in the management of acute angle closure glaucoma, as well as in chronic open-angle glaucoma when other treatments have failed. They have a metallic bitter taste.

Contraindications

Sulfonamide allergy, since CAIs are sulfonamide derivatives. Adrenal, hepatic, respiratory or renal failure, or metabolic acidosis, due to the risk of profound acidosis. Avoid concurrent use of aspirin, salicylates. Any sodium or potassium depletion is worsened.

Precautions

Since bicarbonate production is required in the corneal endothelium and epithelium, topical CAIs may lead to corneal edema in an already compromised cornea or a corneal graft.

Dorzolamide and brinzolamide	
ADMINISTRATION	Topical
ACTIVITY	Reduces IOP about 15–20% as monotherapy. May be used when beta-blocker is not tolerated or ineffective. As an adjunct to a beta-blocker, it may reduce the IOP about 30%
PREPARATIONS	Trusopt, dorzolamide 2% solution (Merck) + generics Azopt, brinzolamide 1% suspension (Alcon) Cosopt, dorzolamide 2%, and timolol 0.5% (Merck) + generics
DOSING	As monotherapy, bid–tid. As combination therapy, or as an adjunct, bid. In acute angle closure, 1 drop.
NOTES	Brinzolamide is similar to dorzolamide, but causes less stinging and burning, due to Alcon's carbopol suspension vehicle (which requires minimal shaking).

Acetazolamide	
ADMINISTRATION	Oral
ACTIVITY	Reduces IOP 20–30%
ADVERSE REACTIONS	Paresthesia of hands, feet, or mucocutaneous junctions, fatigue, depression, decreased libido, nausea, vomiting, abdominal cramps, diarrhea, polyuria, renal stones, transient hypokalemia
USE	Maximum of 250 mg qid. In acute angle-closure glaucoma, 500 mg
PREPARATIONS	Diamox, 250-mg tablet (Lederle)

5E Cholinergic (parasympathomimetic) drugs – miotics

Overview
Parasympathomimetics are the oldest form of glaucoma therapy, in use since 1876. They have been superseded with the advent of newer classes of medication (above) that lack the adverse effects on vision.

Activity
Cholinergic effect constricts the iris and contracts the ciliary muscle, which increases outflow through the trabecular meshwork.

Contraindications
Iritis/uveitis, secondary glaucomas associated with extensive outflow obstruction. Pregnancy (near-term).

Precautions
Topical ocular cholinergic drugs can potentially induce retinal detachment in patients with high myopia, aphakia, peripheral retinal degeneration, or a history of previous retinal detachment. More commonly, miotic therapy can induce fluctuating blurred vision, accommodative spasm, and frontal headaches in pre-presbyopic patients. Older patients are commonly affected by a loss of visual acuity, particularly if cataract is also present, reduced vision in low light, and constriction of the visual field. They may also have a slight pro-inflammatory affect.

Uses
1 to 2 drops per eye, up to 4 times daily. Usual concentration of pilocarpine is 2%; maximum is 4% for light iris, 6% for dark iris. For acute closed-angle glaucoma, 1 drop of 2% repeated every 15 minutes. Occlude the tear duct and close the eyes for 3 minutes to reduce systemic absorption. For reversal of mydriasis, use 1 drop of 1%.

Pilocarpine	
TYPE	Direct-acting cholinergic
PREPARATIONS	Generic pilocarpine 0.5%, 1%, 2%, 3%, 4%, 6%, e.g. Pilopt (Allergan), Isopto Carpine (Alcon), other companies, single-use minims (Chauvin Pharmaceuticals)
Carbachol	
TYPE	Direct-acting cholinergic, and also partially inhibits cholinesterase
PREPARATIONS	Isopto, carbachol 1.5% and 3.0% (Alcon)

6 CORTICOSTEROIDS

Absolute contraindications
- Acute epithelial (dendritic or geographic) herpes simplex keratitis
- Vaccinia and varicella viruses

Relative contraindications (precautions)
- May potentiate herpes simplex virus (HSV) replication.
- Acute untreated or purulent infection may be masked or enhanced by steroid medication.
- May potentiate fungal or amebic keratitis if used in the active phase of the disease.
- Mycobacterium (tuberculosis, leprosy).
- Caution is also indicated when there is a significant corneal epithelial defect due to the heightened risk of infectious keratitis. Healing of a significant epithelial defect may be slowed, and secondary bacterial infection may be potentiated.

PREPARATIONS	Flarex, 0.1% fluorometholone acetate suspension (Alcon) First FDA approval in 1986: generics also available
NOTES	Fluorometholone is a structural analog of progesterone, a naturally occurring steroid hormone. The acetate form of fluorometholone has greater anti-inflammatory effect than the alcohol form and is metabolized more slowly in the anterior chamber
DOSAGE	See dosing for prednisolone acetate 1% and also difluprednate, above. Also see general guidelines for corticosteroid dosage at top

Prednisolone sodium phosphate

GENERAL USES	Mild to moderate ocular surface, corneal, and intraocular inflammation
SPECIFIC USES	Corneal infiltrates, uveitis, post-intraocular surgery
CONTRAINDICATIONS, and PRECAUTIONS	See notes at the start of this section (5)
PREPARATION	Minims, 0.5% solution (Chauvin Pharmaceuticals) Inflamase Forte, prednisolone sodium phosphate 1% solution (Novartis) First FDA approval in 1993: generics also available
NOTES	Prednisolone sodium phosphate is less able to penetrate the cornea than prednisolone acetate (see above) and is less effective for intraocular inflammation
DOSAGE	Being a solution, this steroid will still be effective if not shaken prior to instillation. See general guidelines for corticosteroid dosage at top

Dexamethasone

GENERAL USES	Marked corneal or intraocular inflammation
SPECIFIC USES	Uveitis, corneal graft rejection
CONTRAINDICATIONS, and PRECAUTIONS	See notes at the start of this section (5)
PREPARATION	Maxidex, 0.1% suspension (Alcon) First FDA approval in 1962: generics available
NOTES	Like prednisolone, dexamethasone is a synthetic analog of hydrocortisone (cortisol), but may be more resistant to metabolism in the anterior chamber and increase IOP more rapidly. For this reason it is less commonly used than some other steroids

6B Corticosteroids with lower potency

Loteprednol etabonate 0.2%

GENERAL USES	Ocular surface inflammation, with minimal effect on intraocular pressure
SPECIFIC USES	Allergic eye disease, seasonal allergic conjunctivitis, Thygeson's SPK
CONTRAINDICATIONS, and PRECAUTIONS	See notes at the start of this section (5). However, in one study the increase in IOP with loteprednol 0.2% was no different to placebo
PREPARATION	Alrex, 0.2% suspension (Bausch & Lomb) First FDA approval in 1998: patent expires 2013
RECOMMENDED DOSAGE	Shake well before using. One drop instilled into the affected eye(s) 4 times daily. Improvement in signs and symptoms are expected within 2 days

Prednisolone acetate 0.12%	
GENERAL USES	Mild to moderate noninfectious allergic and inflammatory disorders of the lid, conjunctiva, cornea, and sclera (including chemical and thermal burns)
SPECIFIC USES	Corneal infiltrates, uveitis, corneal graft rejection
CONTRAINDICATIONS, and PRECAUTIONS	See notes at the start of this section (5)
PREPARATION	Pred Mild, 0.12% prednisolone acetate suspension (Allergan) First FDA approval in 1972: generics also available
DOSAGE	See dosing for loteprednol etabonate 0.2%, above. Also see general guidelines for corticosteroid dosage at top

Fluorometholone	
GENERAL USES	Mild ocular surface and corneal inflammation
SPECIFIC USES	Ocular allergy, Thygeson's SPK, adenoviral keratoconjunctivitis
PREPARATION	FML, 0.1% suspension, and ointment (Allergan) First FDA approval in 1972: generics also available
DOSAGE	See dosing for loteprednol etabonate 0.2%, above. Also see general guidelines for corticosteroid dosage at top

Hydrocortisone acetate	
GENERAL USES	Mild ocular surface and corneal inflammation
SPECIFIC USES	Ocular allergy, nonpurulent conjunctivitis or blepharitis, recurrent corneal marginal ulceration
PREPARATION	Hycor eye ointment, hydrocortisone acetate 0.5% and 1.0% (Sigma)
RECOMMENDED DOSAGE	Apply 1.0–1.5 cm into the lower conjunctival sac, then allow the eye to close

7 STEROID AND ANTIBIOTIC COMBINATIONS

When a decision is made to administer both a corticosteroid and an antimicrobial, a combination drop has several advantages:

- greater patient compliance and convenience
- appropriate dosage of both drugs is administered
- compatibility of ingredients is assured.

Contraindications and adverse reactions for combination drops are primarily those for the steroid component, in addition to possible hypersensitivity to components. For details, refer to the individual steroid drugs as listed above in Section 5, including possible intraocular pressure increases with steroids, particularly with dexamethasone. Antibiotic adverse reactions and warnings are listed above in Section 1, including possible hypersensitivity reactions to neomycin or sulfa drugs.

7A Corticosteroids with higher potency		
	COMBINATION	FORM
Zylet (Bausch & Lomb)	Loteprednol 0.5%, tobramycin 0.3%	Suspension eye drops
Pred-G (Allergan)	Prednisolone acetate 1%, gentamicin 0.3%	Suspension eye drops and ointment

Maxitrol (Alcon) + generics	Dexamethasone 0.1%, neomycin sulfate 0.35%, polymyxin B	Suspension eye drops and ointment
Tobradex (Alcon) + generics	Dexamethasone 0.1%, tobramycin 0.3%	Suspension eye drops and ointment

7B Corticosteroids with lower potency

	COMBINATION	FORM
Blephamide (Allergan) + generics	Prednisolone acetate 0.2%, sulfacetamide 10%	Suspension eye drops and ointment
Corticosporin (Monarch) + generics	Hydrocortisone 1%, neomycin sulfate 0.35%, polymyxin B	Suspension eye drops
FML-S (Allergan)	Fluorometholone 0.1%, sulfacetamide 10%	Suspension eye drops
NeoDecadron (Merck) + generics	Dexamethasone phosphate 0.1%, neomycin sulfate 0.35%	Solution eye drops
Poly-Pred (Allergan)	Prednisolone acetate 0.5%, neomycin 0.35%, polymyxin B	Suspension eye drops
Terra-Cortril (Roerig)	Oxytetracycline hydrochloride, and hydrocortisone acetate	Suspension eye drops
Vascodin (Novartis) + generics	Prednisolone phosphate 0.25%, sulfacetamide 10%	Solution eye drops

8 NON-STEROIDAL ANTI-INFLAMMATORY DRUGS (NSAIDS)

Topical NSAIDs have analgesic and anti-inflammatory effects, somewhat like aspirin.

Precautions

Topical NSAIDs have been occasionally implicated in the development of sterile corneal infiltrates and sterile keratolysis. Intended use is 2 weeks, or less. See package inserts (or online) for details.

Dosage

Typically used qid prior to and following surgery, unless otherwise noted below.

Bromfenac	
USES	Treatment of pain and inflammation associated with cataract surgery
PREPARATIONS	Xibrom, 0.09% Bromfenac (ISTA Pharmaceuticals)
DOSING	bid
Diclofenac	
USES	Postoperative inflammation, ocular pain secondary to epithelial defect
PREPARATIONS	Voltaren, 0.1% diclofenac sodium (Novartis) Generics also available
DOSING	qid
Flurbiprofen	
USES, and DOSING	Prevention of intraoperative miosis

PREPARATIONS	Ocufen, 0.03% flurbiprofen (Allergan) Generics also available
DOSING	qid

Ketorolac

USES	Seasonal allergic conjunctivitis (itch), postoperative inflammation
PREPARATIONS	Acular, 0.5% ketorolac tromethamine (Allergan) Acular LS, 0.4% ketorolac tromethamine (Allergan) Acuvail Unit-dose, 0.45% ketorolac tromethamine (Allergan) Generics also available
DOSING	qid

Nepafenac

USES	Treatment of pain and inflammation associated with cataract surgery
PREPARATIONS	Nevanac, 0.1% nepafenac suspension (Alcon)
DOSING	tid

9 CYCLOSPORIN (CICLOSPORIN)

Cyclosporin A

GENERAL USES	Chronic ocular surface inflammation	
SPECIFIC USES	VKC, AKC, moderate to severe dry eye	
PREPARATIONS	Restasis (Allergan)	0.05% cyclosporin emulsion
	Non-commercial	1 or 2%
DOSING	Shake unit dose vial to obtain uniform white emulsion before using. One drop per eye, twice per day approximately 12 hours apart	

10 MAST-CELL STABILIZERS

Overview

Mast-cell stabilizers prevent histamine release by blocking mast cell degranulation. They are most suited to chronic care of ocular allergy. Also note the combination antihistamine–mast-cell stabilizer (Section 11).

Uses

Long-term treatment and prevention of vernal keratoconjunctivitis, perennial and seasonal allergic keratoconjunctivitis. Mast-cell stabilizers are generally less effective in GPC, although some symptomatic improvement can occur. The newer products (pemirolast and nedocromil) may be more efficacious for GPC in contact lens wear. The main adverse reaction is irritation on instillation, more so with cromolyn sodium.

Dosing

Response is usually within a few days (lodoxamide) or 7–10 days (cromoglycate). Treatment is usually for several months although longer-term treatment is common. Contain benzalkonium chloride; patients should refrain from contact lens wear.

Sodium cromoglycate

NOTES	The first of the mast-cell stabilizers
INDICATIONS	Vernal keratoconjunctivitis, vernal conjunctivitis, and vernal keratitis

PREPARATIONS	Crolom, 4% solution (Bausch & Lomb) + generics
RECOMMENDED DOSING	Four times daily. Symptomatic relief may take 1–6 weeks. Adults and children greater than 4 years of age

Lodoxamide

INDICATIONS	Vernal keratoconjunctivitis, vernal conjunctivitis, and vernal keratitis
PREPARATIONS	Lomide/Alomide, 0.1% solution (Alcon)
RECOMMENDED DOSING	Four times daily. Adults and children greater than 2 years of age

Nedrocromil sodium

INDICATIONS	Treatment of itching associated with allergic conjunctivitis
PREPARATIONS	Alocril, 2% solution (Allergan)
RECOMMENDED DOSING	Twice a day, used at regular intervals. Treatment should be continued throughout the period of exposure (i.e. until the pollen season is over or until exposure to the offending allergen is terminated), even when symptoms are absent. Pediatric patients above the age of 3 years

Pemirolast potassium

INDICATIONS	Allergic conjunctivitis
PREPARATIONS	Alamast, 0.1% solution (Vistakon Pharmaceuticals)
RECOMMENDED DOSING	Four times daily. Symptomatic response to therapy (decreased itching) may be evident within a few days, but frequently requires longer treatment (up to 4 weeks). Pediatric patients above the age of 3 years

11 ANTIHISTAMINES/ANTIHISTAMINE–MAST-CELL STABILIZERS

Overview

These relatively selective histamine H-1 antagonists (second-generation antihistamines) are available without a vasoconstrictor drug included. Medications with antihistamine and mast-cell stabilizing activities are also covered in this section.

Other drugs that may be also considered are:

- loteprednol (Section 6)
- ketorolac (Section 8)
- for long-term (chronic) use, consider mast-cell stabilizers (Section 10)
- the first-generation antihistamines (antazoline and pheniramine) are covered below in "Combination drops" (Section 11).

Indication

Acute care of allergic eye conditions. Itchy eyes.

Levocabastine

CLASS	Histamine H-1 antagonist
USES	Mild to moderate ocular allergy
PREPARATIONS	Livostin, 0.05% levocabastine (Jansen-Cilag/Novartis)
DOSING	Up to qid dosing, for up to 8 weeks

Emedastine

CLASS	Histamine H-1 antagonist
USES	Mild to moderate ocular allergy
PREPARATIONS	Emadine, 0.05% emedastine (Alcon)
DOSING	Up to qid dosing, for up to 8 weeks

Olopatadine HCl	
CLASS	Histamine H-1 antagonist, with some mast-cell stabilizing properties
USES	Seasonal allergic conjunctivitis
PREPARATIONS	Patanol, 0.1% olopatadine HCl (Alcon) Pataday, 0.2% olopatadine HCl (Alcon)
DOSING	Patanol bid dosing, for up to 14 weeks, or qd for Pataday

Ketotifen fumarate	
CLASS	Histamine H-1 antagonist, with some mast-cell stabilizing properties
USES	Seasonal allergic conjunctivitis
PREPARATIONS	Zaditor, 0.025% ketotifen fumarate (Novartis) + generics
DOSING	Usually bid dosing (over-the-counter drug)

Azelastine HCl	
CLASS	Histamine H-1 antagonist, with some mast-cell stabilizing properties
USES	Seasonal allergic conjunctivitis
PREPARATIONS	Optivar, 0.05% azelastine HCl (Bausch & Lomb) + generics
DOSING	Usually bid dosing

12 VASOCONSTRICTORS

Overview

Topical vasoconstrictors have a mild alpha-adrenergic (sympathomimetic) effect, leading to mild vasoconstriction and mydriasis. Vasoconstrictors are available over-the-counter and often formulated with a lubricant, although most are preserved with benzalkonium chloride (BAK).

Uses

Mild ocular allergy, non-specific conjunctival irritations. Recommended for short-term use only (e.g. 2–3 days), as overuse may produce rebound hyperemia or medicamentosa.

Contraindications

Narrow-angle glaucoma, dry eye, contact lens wear; also caution in patients with elevated blood pressure or cardiac disorders, history of stroke, hyperthyroidism, diabetes, pregnancy, or lactation.

Precautions

Drowsiness may be experienced in some patients if there is excessive systemic absorption. Vasoconstrictors and/or the associated preservative can cause acute or chronic inflammatory conjunctivitis in susceptible patients or with longer term use.

Naphazoline HCl	
PREPARATIONS	Albalon, 0.1% (Allergan) Murine Clear Eyes, 0.012% (Abbott) Naphcon Forte, 0.1% (Alcon) Optazine, 0.05% (Whitehall) In-a-wink (Abbott Medical Optics) Naphazoline nitrate 0.005%, unpreserved unit dose (Novartis)

Phenylephrine HCl (all are 0.12% or 0.5%)	
PREPARATIONS	Albalon Relief, unpreserved unit dose (Allergan) Isoptofrin (Alcon) Prefrin Z, with zinc sulfate (Allergan) Zincfrin, with zinc sulfate (Alcon)

Tetrahydrozoline HCl (all 0.05%)

PREPARATIONS	Murine Sore Eyes (Abbott) Optazine Fresh (Whitehall) Visine Advanced Relief (Pfizer) Visine Original eye drops (Pfizer) Visine Relief (Pfizer)

COMBINATION DROPS

Uses

Mild ocular allergy. The first-generation topical ocular antihistamines (antazoline, pheniramine maleate) have been used topically for over 50 years and are available only with an adrenergic agonist vasoconstrictor.

Contraindications

As for the vasoconstrictors listed above.

Naphazoline (0.025%) and pheniramine (0.3%)

PREPARATIONS	Naphcon-A (Alcon)

Naphazoline and antazoline (0.5%)

PREPARATIONS	Albalon A (Allergan) Antistine-Privine (Novartis) Murine Allergy Eyes, antazoline (0.5%), and xylometazoline (0.05%) (Abbott)

Antazoline (0.5%) and tetrahydrozoline HCl (0.04%)

PREPARATIONS	In-a-wink Allergy Eyes (Novartis)

13 OCULAR LUBRICANTS (ARTIFICIAL TEARS, TEAR SUPPLEMENTS)

Newer longer-lasting lubricants

The more recent formulations of ocular lubricants use components that can adhere to the surface of the eye and provide longer-lasting relief of symptoms. They are particularly beneficial for patients with ocular surface damage (shown by staining with fluorescein or lissamine green). These modern formulations typically also include electrolytes and use mild non-toxic preservatives.

	CONSTITUENTS	FORMULATIONS
Blink Intensive Tears (AMO)	0.25% Polyethylene glycol 400 (PEG 400), boric acid, sodium hyaluronate, potassium chloride, sodium borate, calcium chloride, magnesium chloride, sodium perborate	Bottle 15 mL; Unit dose 0.4 mL (non-preserved)
Refresh Optive (Allergan)	0.5% Carmellose, and 0.9% glycerin, boric acid, calcium chloride, magnesium chloride, potassium chloride, sodium borate, sodium chlorite (Purite)	Bottle 15 mL; Unit dose 0.4 mL (non-preserved)
Systane (Alcon)	Polyethylene glycol 400 (PEG 400) 0.4%, and propylene glycol 0.3%, hydroxypropyl guar, boric acid, calcium chloride, magnesium chloride, potassium chloride, zinc chloride, Polyquad 0.001%	Bottle 15 mL; Unit dose 0.8 mL (non-preserved)
Systane Ultra (Alcon)	Similar to Systane (above), with addition of sorbitol and aminomethylpropanol. Faster equilibration in the eye	Bottle 10 mL; Unit dose 0.4 mL (non-preserved)

Soothe (Bausch & Lomb)	Glycerin 0.6%, propylene glycol 0.6%, boric acid, hydroxyalkylphosphonate, sodium alginate, sodium borate	Unit dose 0.6 mL (non-preserved)
Visine Pure Tears (Johnson & Johnson)	Glycerin 0.2%, hypromellose 0.2%, polyethylene glycol 400 1%, ascorbic acid, dextrose, disodium phosphate, glycine, magnesium chloride, potassium chloride, sodium citrate, sodium lactate, sodium phosphate	Single drop dispenser 9.5 mL; Unit dose 0.4 mL

Lipid drops

A newer category of dry eye treatment is aimed at meibomian gland dysfunction and an abnormal tear film lipid layer.

	CONSTITUENTS	FORMULATIONS
Soothe XP (Bausch & Lomb)	Light mineral oil 1.0% and mineral oil 4.5% (emollients), edetate disodium, octoxynol-40, PHMB (preservative), polysorbate-80, sodium hydroxide and/or hydrochloric acid (pH), sodium phosphate	Bottle 15 mL
Eye Logic Eye Spray	Soy lecithin 1.0% (phospholipid) liposomes. Sodium chloride 0.8%, ethanol 0.8%, vitamin A palmitate 0.025%, vitamin E (tocopherol) 0.002%, phenoxyethanol 0.5% preservative	Spray 10 mL
Tears Again (Optima Pharmaceutical)	Soy lecithin liposome eye spray. Sodium chloride, ethanol, vitamin A palmitate, vitamin E, phenoxyethanol preservative	Spray 10 mL
FreshKote (FOCUS Laboratories)	Polyvinyl pyrrolidone 2.0%, polyvinyl alcohol, amisol clear (phospholipid), potassium chloride, boric acid. Preservatives: disodium edetate dihydrate, polixetonium	Bottle 15 mL, Bottle 3.5 mL
Systane Balance (Alcon)	Mineral oil, dimyristoyl phosphatidylglycerol, propylene glycol 0.6%, hydroxypropyl guar, boric acid, polyoxyl 40 stearate, POLYQUAD® 0.001%, sorbitan tristearate, sorbitol	Bottle 10 mL
Visine Professional Intensive Dry Eye Liquid Gel	Povidone, carbomer, medium-chain triglycerides	Bottle 10 mL (multi-dose)

Older-type lubricants, with milder preservatives

These well-established lubricants are usually suited as initial therapy for most patients with mild to moderate dry eye. Milder preservatives are usually well tolerated for 3–4 times per day dosing. For more dosing (>4 per day), consider either the unit dose lubricants or the newer longer-lasting lubricants listed above.

Lubricant composition

(a) Hypromellose or hydroxypropyl methylcellulose (HPMC) has been in ophthalmic use since the 1940s. HPMC is based on methylcellulose (a polysaccharide) that is referred to as a mucilum (gum). Dextran (poly-D-glucose) is also a polysaccharide and is osmotically active.

(b) Carboxymethylcellulose (CMC, carmellose) is, like hypromellose, a polysaccharide and a formulation of methylcellulose. It is a polyanionic (negatively charged) polymer with mucoadhesive properties. Both CMC and HPMC are more viscous and give better surface wetting than polyvinyl alcohol (see below).

(c) Hyaluronanate is a naturally occurring glycosaminoglycan, found throughout the human body and eye. Sodium hyaluronate (1%) was developed for ocular surgery, as a vitreous replacement known as Healon (Abbott Medical Optics).
(d) Polyvinyl alcohol (PVA) is a water-soluble non-ionic synthetic polymer. It was first used in artificial tears in the 1960s and is still used, mainly in generic lubricants. PVA 1.4% concentration has low viscosity. Polyvinyl pyrrolidone (povidone) is a variation, and at 3–5% has higher viscosity. Polyacrylic acid is another variation. Carbomer is a polymerized carboxyvinylic acid (e.g. Viscotears, see below).

	CONSTITUENTS	FORMULATIONS
BionTears (Alcon)	Hypromellose 0.3%, and dextran 70 0.1%, bicarbonate buffer. Includes electrolytes of potassium, calcium, magnesium, and zinc	Unit dose 0.4 mL
Polytears (Alcon)	Hypromellose 0.3% and dextran 70 0.1%, Polyquad preservative. Also known as Tears Naturale	Bottle 15 mL
Cellufresh/ Refresh Plus (Allergan)	Carmellose 0.5%, calcium chloride, potassium chloride, magnesium chloride, and sodium lactate	Unit dose 0.4 mL
Refresh Tears Plus (Allergan)	Carmellose 0.5%, sodium chlorite (purite) preservative	Bottle 15 mL
Refresh (Allergan)	PVA 1.4% and povidone 0.6%	Unit dose 0.4 mL
Celluvisc (Allergan)	Carmellose 1.0%, calcium chloride, potassium chloride, and sodium lactate	Unit dose 0.4 mL
Aquify Comfort Drops (Ciba Vision)	Sodium hyaluronate 1%, sodium phosphate, and sodium perborate stabilized with phosphonic acid	Bottle 10 mL
Genteal (Novartis)	Hypromellose 0.3%, boric acid, potassium chloride, sodium perborate preservative	Bottle 15 mL
Genteal (Novartis)	Hydroxyethylcellulose and poloxamer 407	Unit dose 0.5 mL
Theratears (Advanced Vision Research)	Carmellose 0.25%, potassium, calcium, magnesium, and phosphate. Bicarbonate, boric acid, and borate buffer. Hypotonic, low viscosity	Unit dose 0.6 mL

Gels, ointments, and lid care

Gels are suited to day or nighttime use. Since the administration of ointments temporarily impairs vision, they are usually restricted to usage at night. Some are available as generics.

	CONSTITUENTS	FORMULATIONS
Genteal gel (Novartis)	Carbomer 980 0.25%, hypromellose 0.3% (3 mg mL), sodium perborate preservative. An improved formulation of Viscotears	Tube 10 g
Genteal Geldrops (Novartis)	Carbomer 980 0.2%, preservative-free	Unit dose 0.6 mL
Poly Gel (Alcon)	Carbomer 0.3% sorbitol, preservative-free	Unit dose 0.5 mL
Advanced Eye Relief™ Night Time Lubricant (Bausch & Lomb)	Eye ointment: mineral oil (20%), white petrolatum (80%), preservative-free	Ointment 3.5 g

Genteal PM (Novartis)	Mineral oil 15%, and white petrolatum 85%, preservative-free	Ointment 3.5 g
Systane ointment (Alcon)	Soft paraffin and wool fat, preservative-free. Also known as PolyVisc or Duratears Ointment	Ointment 3.5 g
Refresh Lacrilube (Allergan)	Paraffin 99.3%, preservative-free	Ointment 3.5 g
Lid care (Novartis)	Miranol-2 MCT 50 mg/mL, domiphen bromide preservative	100 cleansing pads

Ocular lubricants with older-type preservatives

Certain preservatives are potentially toxic to the corneal epithelium or may induce conjunctival inflammation. In particular, lubricants with benzalkonium chloride (BAK) should be avoided in moderate or severe dry eye with multiple daily dosing of qid or greater. Other older preservatives are chlorbutol and disodium edetate (EDTA). All are in 15-mL multi-use bottles.

	CONSTITUENTS	FORMULATIONS
Tears Plus	PVA 1.4%, povidone 6 mg/mL, chlorbutol preservative	Allergan
Teardrops	PVA 1.4%, povidone 6 mg/mL, chlorbutol preservative	Novartis
Liquifilm Tears	PVA 1.4%, BAK preservative	Allergan
Liquifilm Forte	PVA 3%, BAK preservative	Allergan
Murine Revital Eyes	PVA 1.4%, povidone 6%, BAK, and EDTA	Abbott
Murine Tears for Eyes	PVA 1.4%, povidone 6%, BAK, and EDTA	Abbott
Isopto Tears	Hypromellose 0.5%, BAK preservative	Alcon
Amcal Tears for Eyes	Hypromellose 0.5%, BAK, and EDTA	Amcal
Methopt	Hypromellose 0.5%, BAK and EDTA preservatives	Sigma
Methopt Forte	Hypromellose 0.5%, BAK and EDTA preservatives	Sigma

14 MYDRIATICS AND CYCLOPLEGICS

Contraindications

Mydriasis in patients with glaucoma or with narrow anterior-chamber angles may cause a rise in intraocular pressure or pupil-block glaucoma.

Precautions

Patients should be advised not to drive or engage in other hazardous activities while pupils are dilated. Patients may experience photophobia and should protect their eyes from bright light.

Administration

In order to minimize systemic absorption of eye drops, apply pressure to the tear duct immediately following administration of the drug. This particularly applies with higher doses of phenylephrine.

14A Antimuscarinics/anticholinergics

Action

This class of drugs antagonizes the muscarinic action of acetylcholine in the parasympathetic division of the autonomic nervous system. They reversibly block the acetylcholine receptors on iris sphincter and ciliary muscle, producing mydriasis as well as cycloplegia.

Atropine	
ACTIVITY	Atropine has a slower onset and more prolonged effect than other anticholinergics. Mydriasis peak effect in 30–40 min and lasting 7–10 days. Cycloplegia peak effect 3–6 h, with duration 7–14 days
PRECAUTIONS	Systemic absorption may cause toxic reactions, particularly in young children, including restlessness and excited behavior, tachycardia, dryness, and flushing of skin. In young children, long-term cycloplegia may cause amblyopia
USES	Atropine is an effective and long-acting cycloplegic. Its long-lasting effect is usually excessive for diagnostic cycloplegic refraction, when cyclopentolate is available
PREPARATIONS	Single-use minims, Atropine 1% (Chauvin Pharmaceuticals) Atropt 0.5% and 1.0%, 15 mL (Sigma)
Cyclopentolate	
ACTIVITY	Mydriasis peak effect in 30–60 min and lasting 1 day. Cycloplegia peak effect 25–75 min, with duration 6–24 h
USES	Useful for mydriasis and cycloplegia, particularly in children under 12 and infants. 0.5% concentration is recommended for infants under 1 year. Comfort may be assisted with proxymetacaine instilled beforehand
PREPARATIONS	Single-use minims, cyclopentolate 0.5% and 1.0% (Chauvin Pharmaceuticals) Cyclogyl 1%, 15 mL (Alcon)
Homatropine	
ACTIVITY	Mydriasis peak effect in 20–30 min and lasting 6 h to 4 days. Cycloplegia peak effect 30–90 min, with duration 10 h–2 days
USES	A moderately long-acting mydriatic and cycloplegic for cycloplegic refraction and in treatment of uveitis. Also for pre- and postoperative mydriasis
PREPARATIONS	Single-use minims, homatropine 2% (Chauvin Pharmaceuticals) Isopto Homatropine, 2% and 5%, 15 mL (Alcon)
Tropicamide	
ACTIVITY	Mydriasis peak effect in 20–40 min and lasting 6 h. Cycloplegia peak effect 30–40 min, with duration 2–6 h. Examine after 20 min or 30 min for darkly pigmented eyes and for cycloplegic refraction
USES	Tropicamide is an effective mydriatic with a weak cycloplegic action, making it most useful for ophthalmoscopy
PREPARATIONS	Single-use minims, tropicamide 0.5% and 1.0% (Chauvin Pharmaceuticals) Mydriacyl, 0.5% and 1.0%, 15 mL (Alcon)

14B Sympathomimetics/adrenergics

Action

These drugs directly or indirectly stimulate the pupil dilator. These include cocaine, ephedrine hydrochloride, adrenaline (epinephrine), and phenylephrine hydrochloride.

Precautions

Care is needed if these drugs are used in children or in patients with heart disease or high blood pressure, since high doses of this medicine may cause an irregular heartbeat and an increase in blood pressure. There have been rare reports associating the use of phenylephrine 10% with severe cardiovascular reactions, some ending fatally, usually in elderly patients with pre-existing cardiovascular disease.

Phenylephrine	
ACTIVITY	Mydriasis peak effect in 60–90 min and duration of action 5–7 h. Accommodation is not affected
USES	Phenylephrine 2.5% is commonly used as an adjunct if dilation is inadequate with an anticholinergic drug, such as tropicamide. Examples include patients with diabetes or darkly pigmented irises or where maximal view of the retinal periphery is needed
PREPARATIONS	Phenylephrine 2.5% and 10%

15 ANESTHETICS

General notes

Store anesthetics between 2°C and 8°C. Refrigerate; do not freeze. Protect from light. Do not use if anesthetic is discolored (amber).

Action

Topical anesthetics stabilize the neuronal membrane and prevent initiation and transmission of nerve impulses, thereby effecting local anesthesia.

Adverse effects

Commonly cause initial stinging, slight conjunctival redness, and superficial punctate erosions of the cornea. Allergic reactions with gross epithelial sloughing or systemic toxicity with CNS effects are rare. Prolonged use impairs corneal healing, prevents ocular protection, and masks progression of any keratitis.

Uses

Minor conjunctival and corneal surgery and for the removal of foreign bodies. Diagnostic procedures including tonometry, gonioscopy, and ultrasound. For simple procedures 1 drop is usually sufficient; however, for more invasive procedures requiring deeper or longer anesthesia, up to 1 drop every 5 minutes for 5 doses may be given.

Contraindication

Patient use for pain management is absolutely contraindicated because of the risk of anesthetic keratopathy (see "Drop toxicity") and corneal perforation.

Proxymetacaine (proparacaine) hydrochloride	
ACTION	The onset of anesthesia usually begins within 20 sec and lasts up to 15 min
PREPARATIONS	Ophthetic, 0.5%, 15-mL multi-use bottle (Allergan) Alcaine, 0.5%, 15-mL multi-use bottle (Alcon)

Oxybuprocaine (benoxinate) hydrochloride

ACTIVITY	Slightly more irritant than proparacaine, but less than amethocaine. Similar time of onset and duration of anesthesia to proparacaine
PREPARATIONS	Benoxinate, 0.4%, unit-dose minims (Chauvin Pharmaceuticals)

Amethocaine (tetracaine) hydrochloride

ACTIVITY	A 1% solution has an effect roughly equivalent to a 1% solution of cocaine, but its action is more prolonged
PREPARATIONS	Amethocaine, 0.5%, 1.0% unit-dose minims (Chauvin Pharmaceuticals)

Lignocaine (lidocaine)

ACTION	Lignocaine is used for injection, as well as topical anesthesia. It has the slowest onset of action (2–5 min), lasts 20–30 min, and has a longer initial sting, like amethocaine
PREPARATIONS	Xylocaine, 4%, 4-mL multi-use bottle (AstraZeneca)

Anesthetic and fluorescein combination solutions

ACTION	Useful for tonometry, offering a simple combination of the two required drops. While bacteria, particularly *Pseudomonas aeruginosa*, grow well in unpreserved fluorescein solution, combination solutions may be effectively preserved with chlorbutanol 1% or thimerosal
PREPARATIONS	Fluress Eye Drops (Allergan) Oxybuprocaine (benoxinate) hydrochloride and fluorescein sodium preserved with 1.0% chlorobutanol Unit-dose minims (Chauvin) Lignocaine hydrochloride 4% and fluorescein sodium 0.25%

16 STAINS

General notes

For diagnostic, not therapeutic, use. Preparations in combination with an anesthetic require storage below 8°C (refrigerate).

Fluorescein

ACTION	Fluorescein is an exclusion dye and does not stain a normal cornea, but corneal and conjunctival epithelial defects are stained a bright green. Fluorescein penetrates into the corneal stroma if there is a full-thickness or deep epithelial defect; foreign bodies are surrounded by a green ring; and there is an absence of fluorescence (negative staining) if there is a raised area of epithelium. Fluorescein also stains the tear film and is used for measuring the tear stability (tear break-up time), in applanation tonometry, in testing the patency of the lacrimal passage, and in rigid contact-lens fitting to show the degree of alignment with the corneal contour
NOTE	Maximum contrast is obtained under cobalt blue illumination and by viewing though a Wratten number 15 (yellow) filter
PREPARATIONS	Fluorets, sterile individually wrapped paper strips (Chauvin Pharmaceuticals) Minims, fluorescein sodium 1% (Chauvin Pharmaceuticals) Fluorescein sodium 0.25% and lignocaine 4% Ful-Glo Ophthalmic Strips (Allergan) Sterile individually-enveloped ophthalmic strips impregnated with 0.6 mg of sodium fluorescein Fluress Eye Drops, 5 mL (Allergan) Oxybuprocaine (benoxinate) hydrochloride and fluorescein sodium, preserved with 1.0% chlorobutanol

Rose bengal	
ACTION	Traditionally, it has been observed that rose bengal stains devitalized or necrotic cells but not healthy epithelial cells. More recently, it has been found that the dye will also stain healthy cells and have toxic effects on them if they are unprotected by the preocular tear film; thus, rose bengal is not truly a vital stain. Rose bengal staining can indicate a lack of a protective pre-ocular tear film or a lack of cell vitality or both. Rose bengal is most commonly used for ocular surface staining in dry eye (including keratoconjunctivitis sicca and Sjögren's syndrome). Rose bengal can also stain pressure marks from contact lenses, indicating that alteration of the lens may be necessary, and may be used to highlight the slight abrasion of a minute foreign body. With epithelial herpetic corneal ulcers, rose bengal stains the edges of the dendritic lesions, whereas fluorescein stains the center. Owing to its potent antiviral activity, rose bengal used on a suspected herpetic ulcer may render a viral culture negative
ADVERSE EFFECTS	Rose bengal commonly causes initial stinging and slight conjunctival hyperemia. It can produce significant stinging in more severe dry eyes, where it should be used with care. Anesthesia may be used beforehand to reduce irritation but may result in artifactual staining
PREPARATIONS	Minims, rose bengal 1% (Chauvin Pharmaceuticals)
Lissamine green	
ACTION	Similar to rose bengal, it stains degenerate cells, dead cells, and mucus, although it has the advantage of not causing ocular irritation. It has also been reported to stain membrane-damaged epithelial cells and corneal stroma, like fluorescein
PREPARATIONS	Minims, lissamine green 1% (Akorn)

17 ASPECTS OF THE AUTONOMIC NERVOUS SYSTEM RELEVANT TO TOPICAL OCULAR MEDICATIONS

DIVISION	PARASYMPATHETIC	SYMPATHETIC
Function	Dominates in normal sedentary life	Activated in stressful situations: "fight or flight" response
Fiber type	Cholinergic–muscarinic (and nicotinic)	Adrenergic
Neurotransmitter	Acetylcholine (ACh)	Noradrenaline (norepinephrine)
Inactivation	Acetylcholinesterase enzyme	COMT & monoamine oxidase (MAO)
Absence	Adie's tonic pupil (slight mydriasis) 0.125% pilocarpine	Horner's syndrome (slight miosis) 0.125% phenylephrine
Pupil	Miosis (sphincter contraction, α/β2). High parasympathetic drive (ACh) normally. ACh is an endogenous muscarinic receptor agonist	Mydriasis (dilator contraction, α), e.g. atropine, muscarinic receptor antagonist
Ciliary muscle	Accommodation (contraction, β2)	Far vision (minimal, or no effect, β2)
Lacrimal glands	Increased mucous and serous secretion	More viscous secretion (vasoconstriction)
CB aqueous production		Increase in active secretion (β2)

Aqueous drainage	Increased outflow (stimulates longitudinal ciliary muscle)	Increased outflow (increases active transport at trabecular meshwork)
Blood vessels of skin and mucosa	Vasodilation and conjunctival hyperemia	Blanching conjunctiva (constriction, α)
Palpebral fissure	Levator	Müller's muscle
Heart	Beats slower, vagal arrest (β1)	Beats faster (S–A node, β1)
Lung	Constricts airways (bronchi, β2)	Dilates airways (bronchi relaxation, β)
DRUGS		
Cycloplegia and mydriasis	Antimuscarinics (reversibly block the acetylcholine receptors): cyclopentolate tropicamide, atropine, homatropine	Cocaine (prolongs noradrenaline), phenylephrine 2.5% and 10% (sympathomimetic)
Miotic	Pilocarpine, carbachol (parasympathomimetic), physostigmine (anticholinesterase)	
Decongestants		Naphazoline, tetrahydrozoline, xylometazoline (adrenergic agonist vasoconstrictors), phenylephrine 0.12%
Anti-glaucoma (beta-blocker)		Non-selective beta-adrenergic blockers inhibit aqueous humour production, e.g. timolol maleate, timolol hemihydrate, levobunolol, metipranolol, carteolol Beta-1-selective adrenergic blockers, e.g. betaxolol and levobetaxolol
Anti-glaucoma (alpha agonist)		Selective alpha-2-adrenergic agonists: brimonidine, apraclonidine Non-selective adrenergic agonists: adrenaline (epinephrine), dipivefrin (pro-drug of adrenaline)

Appendix B: Laboratory investigations

Some inflammatory ocular conditions are associated with systemic inflammatory conditions such as chronic infections and collagen-vascular disorders. Specific serological investigations with an indicative normal reference range are given below.

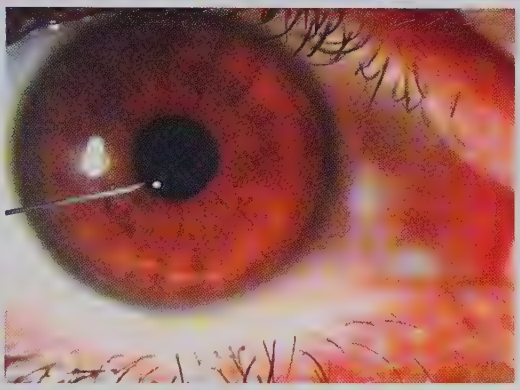
ACRONYM	TEST	INTERPRETATION
ACE	Angiotensin-converting enzyme	Raised in sarcoidosis during active stages of disease Normal range: 40–140 U/mL
ANA	Antinuclear antibodies	Usually raised in juvenile chronic (rheumatoid) arthritis Normal range: negative if titer <1/40
ANCA	Anti-neutrophil cytoplasmic autoantibodies	2 types of ANCA staining: cytoplasmic (c-ANCA) associated with Wegener's granulomatosis; perinuclear (p-ANCA) raised in polyarteritis nodosa Normal range: negative if titer of <1/10
CRP	C-reactive protein	Non-specific marker of systemic inflammation Normal range: <8 mg/L
CXR	Chest X-ray	May reveal focal granulomatosis of tuberculosis (TB), sarcoid, leprosy, and Wegener's granulomatosis
ESR	Erythrocyte sedimentation rate	Non-specific marker of systemic inflammation Normal range: <20 mm/h (normal value may be up to half the age in years)
FBE	Full blood examination	Changes may indicate chronic disease if white cell count (WCC) changes consistent with inflammation
HepC serology	Hepatitis C antigen	May be associated with peripheral ulcerative keratitis
HLA-B27	Human leukocyte antigen B27	Associated with anterior uveitis, ankylosing spondylitis, and Reiter's syndrome
Lyme serology	Lyme disease serology	Both IgM and IgG measured, may become negative in late disease
Mantoux test	PPD-s	Positive in most patients with active tuberculosis or in a patient with previous BCG vaccination Positive: >10 mm of induration at 48 or 72 hours; >20 mm of induration atypical of vaccination reaction
RTG	Renal test group, urea and electrolytes, renal function tests	Urea and creatinine may be raised with renal involvement from a systemic vasculitis
Rh.F	Rheumatoid factor	Raised in active rheumatoid disease Normal range: <30 IU/mL

Syphilis serology	Non-treponemal (VDRL, RPR): detect incidental cell-damage marker, cardiolipin	Rapid plasma reagin: titer usually <1/4, indicative of active disease, may become negative in late disease
	Treponemal (TPHA, EIA): detect antibody to, or antigen of *T. pallidum*	*Treponema pallidum* partial agglutination: titer usually <1/4

Appendix C: Office procedures

1 Foreign body removal
2 Excision of skin and conjunctival cysts
3 Corneal epithelial debridement
4 Lid hygiene
5 Meibomian gland expression
6 Taping of the lid
7 Pressure patching of the lid
8 Suture removal
9 Epilation
10 Lacrimal lavage
11 Punctal occlusion
12 Lacrimal sac massage and palpation

1 CORNEAL FOREIGN BODY REMOVAL



Instill a drop of topical anesthetic into both eyes. Superficial, non-embedded corneal and conjunctival foreign bodies can be irrigated away or manually wiped off with a cotton bud saturated in anesthetic.

Embedded corneal foreign bodies can be removed with the tip of a 25-gauge needle. The needle can be mounted on a cotton-tipped applicator ("cotton bud") or syringe to allow easier handling and, under slit-lamp control, is advanced to the foreign body with the needle tangential to the cornea (parallel to the iris plane). The tip of the needle may be partially bent-over to facilitate hooking out the foreign body; this can be achieved by pressing the tip of the needle against the inside of its protective plastic sheath. If there is an associated rust ring, it may be removed with a needle (if very superficial) or with a dental burr (if the rust has penetrated the superficial stroma).

2 EXCISION OF SKIN AND CONJUNCTIVAL CYSTS



This procedure allows the removal of the contents of a cyst, allowing the surrounding tissue to deflate. It is usually only required if the patient is symptomatic. If the cyst is on the bulbar conjunctiva, topical anesthetic should be instilled into both eyes; if the cyst is on the eyelid skin, the anesthetic can be applied to the skin using a cotton bud for 30 seconds; however, no anesthesia may be necessary for thin-walled cysts of Moll on the lid margin.

A sterile 25-gauge needle is used to lance the lesion and is held angled at a tangent or away from the ocular surface. Position the patient at the slit lamp or against the headrest of the patient chair in order to stabilize them. Simple incision and expression of the contents is usually sufficient to remove these lesions. To express the contents, place pressure on the lesion with a cotton bud or with two cotton buds on either side or use lid massage for a conjunctival cyst. If the cyst recurs, removal of the roof of the cyst or surgical excision of the whole cyst at the base may be required (see image). Following lancing and drainage, instillation of a single drop of a broad-spectrum topical antibiotic is appropriate, although the innate antibiotic properties of the tear film are probably adequate.

3 CORNEAL EPITHELIAL DEBRIDEMENT

In some conditions – notably with a corneal epithelial defect complicating recurrent corneal erosion syndrome and herpes simplex dendritic ulcer – corneal recovery is facilitated by removal of the affected epithelium.

Topical ophthalmic anesthetic is instilled into both eyes for the comfort of the patient and to enable them to keep as still as possible during the procedure. An anesthetic-soaked cotton bud is used at the slit lamp to gently wipe across the edges of the affected epithelium. The cornea is approached from the side and the cotton bud is kept in a tangential plane to the cornea. Loose epithelial cells are picked up by the cotton bud as it is stroked in one direction; avoid rolling the cotton bud or pressing directly on the cornea. After the procedure, the eye should be irrigated with sterile saline to remove the debris and a topical antibiotic or antiviral instilled, as required.

4 LID HYGIENE



It is important to give clear instructions regarding lid hygiene:

- Gentle cleansing of the lid margins. Scrubbing of the lashes and lid margin is performed with a mild detergent to remove built-up scale and oils. This can be done with either a commercially available product or diluted baby shampoo.
- Hot compresses at least 4 times daily. One method of applying heat is with microwaved wheat bags (commonly used for arthritis therapy). The heat is aimed at melting solidified sebum and expression of meibomian gland secretions.
- If massage of the lump is performed, it should be gentle to avoid gland rupture.
- Antisepsis may be required for staphylococcal overgrowth, using a non-alcohol-based antiseptic such as Betadine.

Positive results from local lid therapy should be seen over a 2–4 week period.

If there is mucopurulent discharge or accumulated exudate, lid hygiene may be assisted by irrigation with sterile saline. This procedure is designed to reduce ocular surface damage from the active lytic enzymes in the discharge and, in acute infections, this may need to be performed every hour. The procedure can also reduce ocular discomfort, inflammation, and membrane formation.

5 MEIBOMIAN GLAND EXPRESSION

Meibomian gland expression is an office procedure designed to relieve meibomianitis, a blockage or inflammation of the meibomian glands affecting the upper or lower eyelids. It is not indicated for acute internal hordeolum or in chalazion therapy. It can be uncomfortable for the patient, owing to the sensitivity of the eyelids.



Topical anesthetic drops should be instilled in both eyes, with repeat instillation after 5 minutes for deeper anesthesia. Use either the slit lamp or the headrest of the chair to stabilize the patient's head. Place a moistened cotton bud horizontally against the palpebral conjunctiva adjacent to the gland to be expressed and a second cotton bud in the corresponding position on the outside of the eyelid but from the opposite direction. Firmly press the two cotton buds together and roll them up toward the lid margin, expressing material out of the affected gland(s). While normal meibomian secretions are a clear fluid, in meibomianitis the expressed material will be a yellow-white semi-solid or a waxy solid. After expression, a regimen of lid hygiene should be prescribed for the patient (see above).

6 TAPING OF THE LID



If there is a need to close the eyelid for temporary corneal protection, simple taping of the eyelid can be attempted. Closure of the eyelid at night or even during the day may be achieved using 2-cm width hypoallergenic surgical tape (e.g. 3M™ Micropore™ Plus Surgical Tape). Ensure eyelid closure as, if the lid opens under the tape, the cornea may rub on the tape. To ensure maximum tape adherence, wipe the eyelids with an alcohol swab and allow to dry.

7 PRESSURE PATCHING OF THE LID



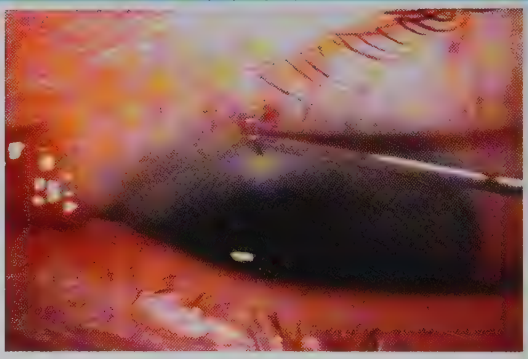
This technique was in widespread use for many years in the acute treatment of corneal abrasions or larger erosions. However, a number of recent scientifically controlled studies have shown no difference in the rate of corneal healing or patient comfort, whether or not pressure patching was applied. These studies found use of a broad-spectrum antibiotic (such as gentamicin, erythromycin, or polymyxin B), a cycloplegic, and a non-steroidal anti-inflammatory drug (such as ketorolac tromethamine 0.5%) was sufficient.

Pressure patching employs two eye patches taped over the closed eye, in such a way as to not only immobilize the eyelid in the closed position but also to cause the eyelid to exert gentle pressure on the cornea. As with the rationale for bandage contact lenses, the aim is to avoid the shearing effects of eyelid blinking on the cornea.

Pressure patching is contraindicated if there are signs of corneal infection or discharge, if the patient is a contact lens wearer, or if a penetrating injury is suspected.

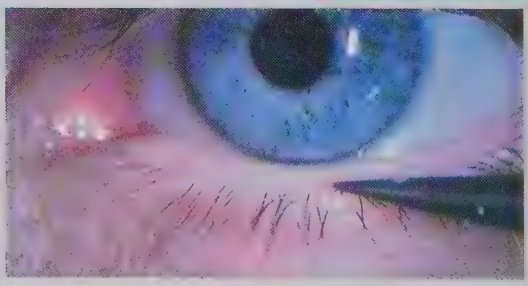
Before applying the patch, a drop of topical anesthetic may be instilled if required for patient comfort. If indicated because of possible traumatic iridocyclitis, instill a cycloplegic agent such as 5% homatropine and apply a prophylactic broad-spectrum topical antibiotic ointment. Use an alcohol swab to clean the skin around the eye, to ensure optimal tape adherence. A sterile gauze eye patch is folded in half and placed on the closed eyelid and taped in place, and another unfolded patch is placed on top. Tape is used to secure the patches in place, usually with the tape running from the middle of the forehead, across the patch and down to the cheek. Pulling the skin of the cheek up before adhering the tape to the skin ensures that the tape is taut. Add several more strips of tape in parallel to secure the entire eye patch.

8 SUTURE REMOVAL



Removal of corneal or conjunctival sutures is performed under slit-lamp or operating microscope control. The eye is anesthetized with a drop of topical anesthetic, the suture cut, and the ends raised with a 25-gauge needle (see image). The needle is advanced to the suture parallel to the iris plane with the bevel directed up. The suture is then removed with forceps in one sharp action. The patient is then prescribed a prophylactic topical antibiotic, usually qid for 4 days. When the suture is removed from a corneal graft, the patient is also prescribed a topical corticosteroid (such as prednisolone acetate) qid for 1 week.

9 EPILATION



Epilation with cilia forceps is the most common management for trichiasis and may also be useful in hastening drainage of pus from around an external hordeolum. It may take several visits to locate all of the lashes, as well as deal with emerging ones. Review is important since recurrence is common, and the patient may not be aware of, or simply be tolerating, the condition. Regrowth of lashes may take 1 to 2 months in adults. Permanent follicle destruction may be considered in recalcitrant cases.

Epilation may be made with or without a slit lamp, although finer lashes may only be visible under magnification. Assessment by fluorescein staining on the cornea may provide evidence of the location of lashes requiring removal. Topical anesthesia is usually not given, since the patient's subjective ocular comfort can provide a guide as to the success of the procedure. The patient should look away from the area of lid to be epilated to minimize the chance of accidental abrasion or aversion responses. Grasp the base of the lash with the forceps to ensure that the root of the lash is also removed. For epilation of the upper eyelid, it may be helpful to evert the lid first, which serves to both protect the cornea as well as make the lashes more accessible.

Other procedures using forceps are considered in their respective conditions, including "Lice," "Filamentary keratopathy," "Concretion," "Scleritis – thinning and necrosis," and "Membranous and pseudomembranous conjunctivitis."

10 LACRIMAL LAVAGE (DILATION AND IRRIGATION, SYRINGING)



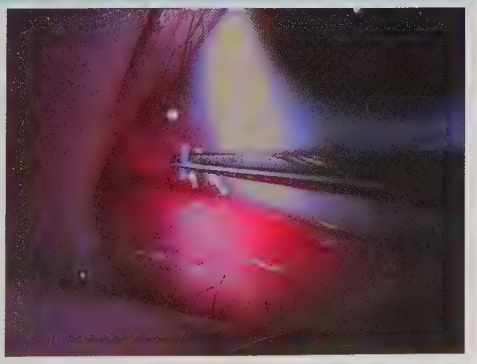


Instill a drop of local anesthetic to the eyes; deeper anesthesia of the punctum prior to the procedure can be achieved using a cotton-tipped applicator soaked with proparacaine. Dilate the punctum with a fine-tipped punctal dilator (e.g. Reudemann) and use punctal dilators of progressive diameters, until a lacrimal cannula can be inserted.

Sterile saline or water is then injected via syringe and the lacrimal cannula. If the canaliculus is blocked, fluid will reflux through the same punctum. Blockage to the common canaliculus or more distal structures will lead to efflux of fluid through the other punctum. If the drainage system is patent, fluid will pass through to the back of the throat.

The punctal dilation procedure may be only a temporary cure and, therefore, the patient should be asked to re-present after a few weeks if symptoms persist. Periodic repeat of the procedure may be beneficial if the stenosis is only mild. For moderate to severe stenosis, surgery may be the only permanent solution; alternatively, a review may be indicated at 6 to 12 months.

11 PUNCTAL OCCLUSION



Aqueous tear deficiency (keratoconjunctivitis sicca, KCS) may be treated with punctal occlusion if ocular lubricants are ineffective. Punctal occlusion is contraindicated in the presence of inflammation (dacryocystitis).

The first step in this treatment is usually a trial using temporary collagen plugs, implanted into the canaliculi (see image). Mild–moderate dry eye may only warrant occlusion of the inferior puncta, whereas both superior and inferior puncta may be blocked in severe dry eye. The initial step in the procedure is to locally anesthetize the puncta by direct application of a topical anesthetic agent. At the slit lamp, remove a collagen plug from the foam packaging

using fine forceps and insert the narrow end of the plug into the puncta of the patient. Use the forceps to tap the top end of the plug down into the canaliculus, until it is fully inside. Additional plugs may be inserted if required. Repeat the procedure for the other eye.

The collagen implants are temporary, since they dissolve after 7 to 10 days. If patients are reviewed after 2 weeks and the therapy has been successful, they should report alleviation of KCS symptoms for the first week or so, followed by a return of symptoms since. In that case, permanent punctal occlusion (via silicone punctal plugs or thermal cautery) can be considered. If the patient reports epiphora during the first week or so, further treatment with punctal occlusion is not indicated.

12 LACRIMAL SAC MASSAGE AND PALPATION



Palpation of the nasolacrimal duct may be used as a diagnostic test in dacryocystitis or as a treatment for nasolacrimal duct obstruction. In acute dacryocystitis, the swelling of the lacrimal sac, medial to the inner canthi, will be inflamed and tender, or painful. If the dacryocystitis is chronic, the lacrimal sac swelling is usually not inflamed. At the slit lamp, the lacrimal sac can be gently palpated, by using the forefinger or a cotton bud on the area between the inner canthus and the nose, while observing the punctum for discharge. Acute dacryocystitis is evidenced by the reflux of mucus or pus from the puncta, whereas a mucoid reflux would be more consistent with a chronic dacryocystitis. Acute dacryocystitis requires systemic antibiotic treatment and should not be massaged because of the risk of extending the surrounding area of cellulitis.

In contrast to the above, a congenital nasolacrimal duct obstruction may be treated with massage. It consists of applying gentle pressure with a finger over the canaliculi and stroking downward firmly to increase the pressure within the lacrimal sac. Massage should be repeated 5 times qid. The objective is to hasten rupture of the valve obstruction.

Appendix D: Drug administration abbreviations

BE (or OU)	both eyes
LE (or OS)	left eye
RE (or OD)	right eye
G, Gutt, or gtt	eye drops
Oc	eye ointment
Collyr or Coll	eyewash
ad or mdu	as directed
bid or bd	twice a day
tid or tds	three times per day
qid or qds	four times per day
m or mane	in the morning
n or nocte	at night
h	an hour
PO	by mouth
q	each or every (e.g. q1h)
qam	every morning
qd	every day
qod	every other day
qpm	every night
qqh or q4h	every 4 hours
PRN	when necessary
Rpt 2	supply 2 repeats
stat	immediately
sig	label
sol	solution
susp	suspension
ung	ointment

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(Note: CG preceding a page number refers to 'Condition grouping', located in the frontmatter of the book.)

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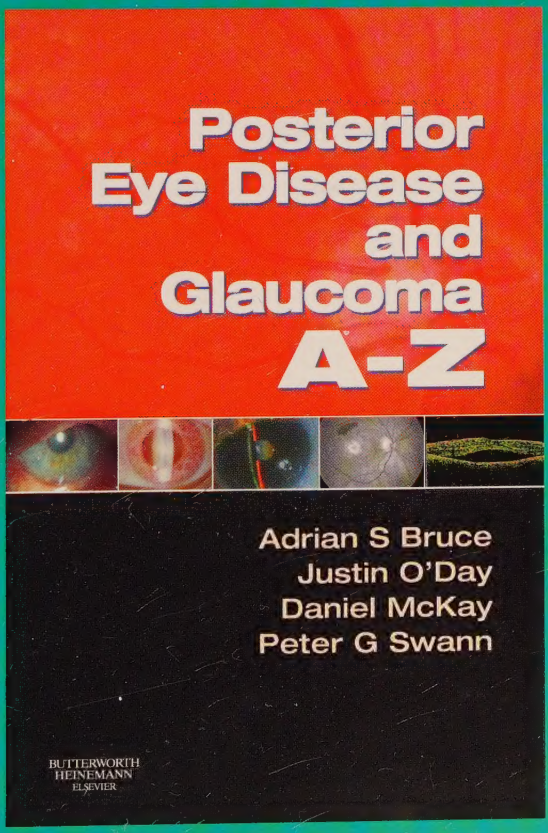
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