

AMERICAN ACADEMY OF OPHTHALMOLOGY®

# SUMMARY BENCHMARKS FOR PREFERRED PRACTICE PATTERN® GUIDELINES

#### Introduction

These are summary benchmarks for the Academy's Preferred Practice Pattern® (PPP) guidelines. The Preferred Practice Pattern series of guidelines has been written on the basis of three principles.

- Each Preferred Practice Pattern should be clinically relevant and specific enough to provide useful information to practitioners.
- Each recommendation that is made should be given an explicit rating that shows its importance to the care process.
- Each recommendation should also be given an explicit rating that shows the strength of evidence that supports the recommendation and reflects the best evidence available.

Preferred Practice Patterns provide guidance for the pattern of practice, not for the care of a particular individual. While they should generally meet the needs of most patients, they cannot possibly best meet the needs of all patients. Adherence to these Preferred Practice Patterns will not ensure a successful outcome in every situation. These practice patterns should not be deemed inclusive of all proper methods of care or exclusive of other methods of care reasonably directed at obtaining the best results. It may be necessary to approach different patients' needs in different ways. The physician must make the ultimate judgment about the propriety of the care of a particular patient in light of all of the circumstances presented by that patient. The American Academy of Ophthalmology is available to assist members in resolving ethical dilemmas that arise in the course of ophthalmic practice.

The Preferred Practice Pattern<sup>®</sup> guidelines are not medical standards to be adhered to in all individual situations. The Academy specifically disclaims any and all liability for injury or other damages of any kind, from negligence or otherwise, for any and all claims that may arise out of the use of any recommendations or other information contained herein.

For each major disease condition, recommendations for the process of care, including the history, physical exam and ancillary tests, are summarized, along with major recommendations for the care management, follow-up, and education of the patient. For each PPP, a detailed literature search of PubMed and the Cochrane Library for articles in the English language is conducted. The results are reviewed by an expert panel and used to prepare the recommendations, which are then given a rating that shows the strength of evidence when sufficient evidence exists.

To rate individual studies, a scale based on the Scottish Intercollegiate Guideline Network (SIGN) is used. The definitions and levels of evidence to rate individual studies are as follows:

- I++: High-quality meta-analyses, systematic reviews of randomized controlled trials (RCTs), or RCTs with a very low risk of bias
- I+: Well-conducted meta-analyses, systematic reviews of RCTs, or RCTs with a low risk of bias
- I-: Meta-analyses, systematic reviews of RCTs, or RCTs with a high risk of bias
- II++: High-quality systematic reviews of case-control or cohort studies; high-quality case-control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal
- II+: Well-conducted case-control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
- II-: Case-control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal
- III: Nonanalytic studies (e.g., case reports, case series)

Recommendations for care are formed based on the body of the evidence. The body of evidence quality ratings are defined by Grading of Recommendations Assessment, Development and Evaluation (GRADE) as follows:

- Good quality (GQ): Further research is very unlikely to change our confidence in the estimate of effect
- Moderate quality (MQ): Further research is likely to have an important impact on our confidence in the estimate of effect and may change the estimate
- Insufficient quality (IQ): Further research is very likely to have an important impact on our confidence in the estimate of effect and is likely to change the estimate; any estimate of effect is very uncertain



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#### Introduction (continued)

Key recommendations for care are defined by GRADE as follows:

- Strong recommendation (SR): Used when the desirable effects of an intervention clearly outweigh the undesirable effects or clearly do not
- Discretionary recommendation (DR): Used when the trade-offs are less certain—either because of lowquality evidence or because evidence suggests that desirable and undesirable effects are closely balanced

In PPPs prior to 2011, the panel rated recommendations according to its importance to the care process. This "importance to the care process" rating represents care that the panel thought would improve the quality of the patient's care in a meaningful way. The ratings of importance are divided into three levels.

- Level A, defined as most important
- Level B, defined as moderately important
- Level C, defined as relevant but not critical

The panel also rated each recommendation on the strength of evidence in the available literature to support the recommendation made. The "ratings of strength of evidence" also are divided into three levels.

- Level I includes evidence obtained from at least one properly conducted, well-designed randomized controlled trial. It could include meta-analyses of randomized controlled trials.
- Level II includes evidence obtained from the following:
  - Well-designed controlled trials without randomization
  - Well-designed cohort or case-control analytic studies, preferably from more than one center
  - Multiple-time series with or without the intervention
- Level III includes evidence obtained from one of the following:
  - Descriptive studies
  - Case reports
  - Reports of expert committees/organizations (e.g., PPP panel consensus with external peer review)

This former approach, however, will eventually be phased out as the AAO adopted the SIGN and GRADE rating and grading systems.

The PPPs are intended to serve as guides in patient care, with greatest emphasis on technical aspects. In applying this knowledge, it is essential to recognize that true medical excellence is achieved only when skills are applied in a such a manner that the patients' needs are the foremost consideration. The AAO is available to assist members in resolving ethical dilemmas that arise in the course of practice. (AAO Code of Ethics)

- Ocular symptoms (e.g., degree of pain, redness, discharge, blurred vision, photophobia, duration of symptoms, circumstances surrounding the onset of symptoms)
- Contact lens history (e.g., wearing schedule, overnight wear, type of contact lenses, contact lens solution, contact lens hygiene protocol, tapwater rinse of contact lenses, swimming, using a hot tub, or showering while wearing contact lenses, method of purchase, such as over the internet, and decorative contact lens use.)
- Review of other ocular history, including risk factors such as herpes simplex virus keratitis, varicella zoster virus keratitis, previous bacterial keratitis, trauma, dry eye, and previous ocular surgery, including refractive and facial (including laser cosmetic) surgery
- Review of other medical problems, including immune status, systemic medications, and history of MRSA.
- Current and recently used ocular medications
- Medication allergies

# **Initial Physical Exam**

- Visual acuity
- General appearance of patient, including skin conditions
- Facial examination
- Globe position
- Eyelids and eyelid closure
- Conjunctiva
- Nasolacrimal apparatus
- Corneal sensation
- Slit-lamp biomicroscopy
  - Eyelid margins
  - Conjunctiva
  - Sclera
  - Cornea
  - Anterior chamber for depth and the presence of inflammation, including cell and flare, hypopyon, fibrin, hyphema
  - Anterior vitreous for the presence of inflammation
  - Contralateral eye for clues to etiology as well as possible similar underlying pathology

# **Diagnostic Tests**

- Manage majority of community-acquired cases with empiric therapy and without smears or cultures.
- Indications for smears and cultures:
  - Corneal infiltrate that is central, large, and/or associated with significant stromal involvement.

- Chronic or unresponsive to broad spectrum antibiotic therapy.
- History of corneal surgeries
- Atypical clinical features suggestive of fungal, amœbic, or mycobacterial keratitis.
- Infiltrates are in multiple locations on the cornea.
- The hypopyon that occurs in eyes with bacterial keratitis is usually sterile, and aqueous or vitreous taps should not be performed unless there is a high suspicion of microbial endophthalmitis, such as following an intraocular surgery, perforating trauma, or sepsis.
- Corneal scrapings for culture should be inoculated directly onto appropriate culture media to maximize culture yield. If this is not feasible, place specimens in transport media. In either case, immediately incubate cultures or take promptly to the laboratory.

# **Care Management**

- Topical antibiotic eye drops are capable of achieving high tissue levels, a preferred method of treatment in most cases.
- Single-drug therapy using a fluoroquinolone is as effective as combination therapy utilitizing fortified antibiotics (*I+, GQ, SR*) There is no difference found in corneal perforation rates across classes of topical antibiotics (*I+, GQ, SR*)
- Topical corticosteroid therapy may have a beneficial role, but much of the literature has not shown a difference in clinical outcome (*I+*, *GQ*, *SR*)
- Subconjunctival antibiotics may be helpful where there is imminent scleral spread or perforation or where adherence is questionable.
- For central or severe keratitis (e.g., deep stromal involvement or an infiltrate larger than 2 mm with extensive suppuration), use a loading dose (e.g., every 5 to 15 minutes), followed by frequent applications (e.g., every hour is recommended.) Severe cases should be followed daily initially, at least until stable or improvement is confirmed.
- Systemic therapy may be useful in cases of scleral or intraocular extension of infection of systemic infection such as gonorrhea.
- For patients treated with ocular topical corticosteroids at time of presentation of suspected bacterial keratitis, reduce or eliminate corticosteroids until infection has been controlled.
- When the corneal infiltrate compromises the visual axis, may add topical corticosteroid therapy following at least 2 to 3 days of progressive improvement with treatment with topical antibiotics typically after pathogen identification.
- Examine patients within 1 to 2 days after initiation of topical corticosteroid therapy and monitor intraocular pressure.
- In general, modify the initial regimen if there is lack of improvement or stabilization within 48 hours.

#### **Patient Education**

- Inform patients with risk factors predisposing them to bacterial keratitis of their relative risk, the signs and symptoms of infection, and to consult an ophthalmologist promptly if they experience such warning signs or symptoms
- Educate about the severe visual impairment from bacterial keratitis and need for strict adherence to the therapeutic regimen
- Discuss possibility of permanent visual loss and need for future visual rehabilitation
- Educate patients with contact lenses about increased risk of infection associated with contact lens, overnight wear, and importance of adherence to techniques to promote contact lens hygiene
- Refer patients with significant visual impairment or blindness for vision rehabilitation if they are not surgical candidates (see <u>www.aao.org/low-vision-</u> and-vision-rehab)

Organism	Topical Antibiotic	Topical Concentration	Subconjunctival Dose
No organism identified or	Cefazolin or Vancomycin with	25-50 mg/ml	100 or 25 mg in 0.5 ml
multiple types of organisms	Tobramycin or gentamicin or	9–14 mg/ml	20 mg in 0.5 ml
	Fluoroquinolones*	Various <sup>+</sup>	
Gram-positive Cocci	Cefazolin Vancomycin‡ Bacitracin‡ Fluoroquinolones*	50 mg/ml 10-50 mg/ml 10,000 IU Various†	100 mg in 0.5 ml 25 mg in 0.5 ml
Gram-negative Rods	Tobramycin or gentamicin Ceftazidime Fluoroquinolones	9–14 mg/ml 50 mg/ml Various†	20 mg in 0.5 ml 100 mg in 0.5 ml
Gram-negative Cocci <sup>s</sup>	Ceftriaxone Ceftazidime Fluoroquinolones	50 mg/ml 50 mg/ml Various†	100 mg in 0.5 ml 100 mg in 0.5 ml
Gram-positive Rods (Nontuberculous Mycobacteria)	Amikacin Clarithromycin Azithromycin <sup>I</sup> Fluoroquinolones	20-40 mg/ml 10 mg/ml 10 mg/ml Various†	20 mg in 0.5 ml
Gram-positive Rods (Nocardia)	Sulfacetamide Amikacin Trimethoprim/ Sulfamethoxazole: Trimethoprim	100 mg/ml 20-40 mg/ml 16 mg/ml	20 mg in 0.5 ml
	Sulfamethoxazole	80mg/ml	

#### Antibiotic Therapy of Bacterial Keratitis

Modified with permission from the American Academy of Ophthalmology Basic and Clinical Science Course Subcommittee. Basic Clinical and Science Course. External Disease and Cornea: Section 8, 2017-2018. Table 10-6. San Francisco: American Academy of Ophthalmology, 2017.

\* Fewer gram-positive cocci are resistant to gatifloxacin, moxifloxacin, and besifloxacin than other fluoroquinolones.

<sup>+</sup> Besifloxacin 6mg/ml; ciprofloxacin 3 mg/ml; gatifloxacin 3 mg/ml; levofloxacin 15 mg/ml; moxifloxacin 5 mg/ml; ofloxacin 3 mg/ml, all commercially available at these concentrations

<sup>‡</sup> For resistant *Enterococcus* and *Staphylococcus* species and penicillin allergy. Vancomycin and bacitracin have no gram-negative activity and should not be used as a single agent in empirically treating bacterial keratitis.

§ Systemic therapy is necessary for suspected gonococcal infection.

Data from Chandra NS, Torres MF, Winthrop KL. Cluster of *Mycobacterium chelonae* keratitis cases following laser in-situ keratomileusis. Am J Ophthalmol 2001; 132(6):819–30.

# **Blepharitis (Initial and Follow-up Evaluation)**

#### **Initial Exam History**

- Ocular symptoms and signs (e.g., redness, irritation, burning, tearing, itching, crusting of eyelashes, loss of eyelashes, eyelid sticking, blurring or fluctuating vision, contact lens intolerance, photophobia, increased frequency of blinking and recurrent hordeolum)
- Time of day when symptoms are worse
- Duration of symptoms
- Unilateral or bilateral presentation
- Exacerbating conditions (e.g., smoke, allergens, wind, contact lenses, low humidity, retinoids, diet and alcohol consumption, eye makeup)
- Symptoms related to systemic diseases (e.g., rosacea, atopy, psoriasis, and graft-versus-host disease [GVDH])
- Current and previous systemic and topical medications (e.g., antihistamines or drugs with anticholinergic effects, or drugs used in the past that might have an effect on the ocular surface [e.g., isotretinoin])
- Recent exposure to an infected individual (e.g., pediculosis palpebrarum [*Pthirus pubis*])
- Ocular history (e.g., previous intraocular and eyelid surgery, local trauma, including mechanical, thermal, chemical, and radiation injury, history of cosmetic blepharoplasty, history of styes and/or chalazia)

#### **Initial Physical Exam**

- Visual acuity
- External examination
  - Skin
  - Eyelids
- Slit-lamp biomicroscopy
  - Tear film
  - Anterior eyelid margin
  - Eyelashes
  - Posterior eyelid margin
  - Tarsal conjunctiva (everting eyelids)
  - Bulbar conjunctiva
  - Cornea

#### **Diagnostic Tests**

- Cultures may be indicated for patients with recurrent anterior blepharitis with severe inflammation as well as for patients who are not responding to therapy.
- Biopsy of the eyelid to exclude the possibility of carcinoma may be indicated in cases of marked asymmetry, resistance to therapy or unifocal recurrent chalazia that do not respond well to therapy.

• Consult with the pathologist prior to obtaining the biopsy if sebaceous cell carcinoma is suspected.

#### **Care Management**

- Treat patients with blepharitis initially with a regimen of warm compresses and eyelid cleansing.
- A topical antibiotic such as bacitracin or erythromycin can be prescribed to be applied one or more times daily or at bedtime on the eyelid margins for a few weeks.
- For patients with meibomian gland dysfunction, whose chronic symptoms and signs are not adequately controlled with eyelid cleansing or meibomian gland expression, oral tetracyclines and topical antibiotics may be helpful.
- Topical azelaic acid, topical ivermectin, brimonidine, doxycycline, and isoretinoin are effective treatments for patients with systemic rosacea. (*I+, GQ, SR*)
- A brief course of topical corticosteroids may be helpful for eyelid or ocular surface inflammation such as severe conjunctival infection, marginal keratitis, or phlyclenules. The minimal effective dose of corticosteroid should be utilized and long-term corticosteroid therapy should be avoided if possible.
- An eyelid tumor should be suspected in patients with atypical eyelid-margin inflammation or disease not responsive to medical therapy, and these patients should be carefully re-evaluated.

#### **Follow-Up Evaluation**

- Follow-up visits should include:
  - Interval history
  - Measurement of visual acuity
  - External examination
  - Slit-lamp biomicroscopy
- If corticosteroid therapy is prescribed, re-evaluate patient within a few weeks to determine the response to therapy, measure intraocular pressure, and assess treatment compliance

- Counsel patients about the chronicity and recurrence of the disease process.
- Inform patients that symptoms can frequently be improved but are rarely eliminated.
- Patients with an inflammatory eyelid lesion that appears suspicious for malignancy should be referred to an appropriate specialist.

- Ocular symptoms and signs (e.g., mattering and adherence of eyelids, itching, tearing, discharge, irritation, pain, photophobia, blurred vision)
- Duration of symptoms and time course
- Exacerbating factors
- Unilateral or bilateral presentation
- Character of discharge
- Recent exposure to an infected individual
- Trauma (mechanical, chemical, ultraviolet)
- Recent surgery
- Mucus fishing behavior (i.e., repetitive manipulation and wiping of the conjunctiva leading to mechanical irritation)
- Contact lens wear (lens type, hygiene and use regimen)
- Symptoms and signs potentially related to systemic diseases (e.g., genitourinary discharge, dysuria, dysphagia, upper respiratory infection, skin and mucosal lesions)
- Allergy, asthma, eczema
- Use of topical and systemic medications
- Ocular history (e.g., previous episodes of conjunctivitis and previous ophthalmic surgery)
- Compromised immune status (e.g., HIV, chemotherapy, immunosuppressents)
- Current and prior systemic diseases (e.g., atopy, SJS/TEN, carcinoma, leukemia, chickenpox, GVHD)
- Social history (e.g., smoking habits, exposure to second hand smoke, occupation and hobbies, exposure to air pollutants, travel, exercise habits, diet, use of illicit drugs, and sexual activity)

# **Initial Physical Exam**

- Visual acuity
- External examination
  - Regional lymphadenopathy, particularly preauricular
  - Skin (signs of rosacea, eczema, seborrhea)
  - Abnormalities of the eyelids and adnexae (swelling, discoloration, malposition, laxity, ulceration, nodules, ecchymosis, neoplasia, lateral flare, lash loss)
  - Orbits: fullness, asymmetry
  - Conjunctiva (laterality, type of conjunctival reaction, distribution, subconjunctival hemorrhage, chemosis, cicatricial change, symblepharon,

masses, discharge)

- Slit-lamp biomicroscopy
  - Eyelid margins (inflammation, edema, hyperpigmentation, meibomian gland dysfunction, ulceration, discharge, nodules or vesicles, bloodtinged debris, keratinization)
  - Eyelashes (loss of lashes, crusting, scurf, mites, nits, lice, trichiasis)
  - Lacrimal puncta and canaliculi (pouting, discharge, edema)
  - Tarsal and forniceal conjunctiva
  - Bulbar conjunctiva/limbus (follicles, edema, nodules, chemosis, laxity, papillae, ulceration, scarring, phlyctenules, hemorrhages, foreign material, keratinization)
  - Cornea
  - Dye-staining pattern (conjunctiva and cornea)
  - Anterior chamber/iris (inflammation reaction, synechiae, transillumination defects)

# **Diagnostic Tests**

- Cultures, smears for cytology and special stains are indicated in cases of suspected infectious neonatal conjunctivitis.
- Smears for cytology and special stains are recommended in cases of suspected infectious neonatal conjunctivitis, chronic or recurrent conjunctivitis, and gonococcal conjunctivitis in any age group.
- Confirm diagnosis of adult and neonate chlamydial conjunctivitis with laboratory testing.
- Biopsy the bulbur conjunctiva and take a sample from an eye with active inflammation when ocular mucous membrane pemphigoid is suspected.
- A full-thickness lid biopsy is indicated in cases of suspected sebaceous carcinoma.
- Thyroid function tests are indicated for patients with SLK who do not have known thyroid disease.

# **Conjunctivitis (Management Recommendations)**

#### **Care Management**

- The majority of cases in the adult popoulation are viral and self-limited, and do not require antimicrobial treatment. There is no proven effective treatment for eradication of adenovial infection; artificial tears, topical antihistamines, topical steroids, oral analgesics or cold compresses may mitigate symptoms. The use of antibiotics should be avoided because of potential adverse treatment effects.
- Allergen-specific immunotherapy is beneficial in reducing allergic conjunctivitis, more in children than in adults (*I+, GQ, SR*)
- Treat mild allergic conjunctivitis with an over-thecounter antihistamine/vasoconstrictor agent or second-generation topical histamine H1-receptor antagonists. (*I+, GQ, SR*) If the condition is frequently recurrent or persistent, use mast-cell stabilizers (*I++, GQ, SR*)
- Treatment for vernal/atopic conjunctivitis include modifying the environment and use of cold compresses and ocular lubricants. For acute exacerbations, topical corticosteroids are usually needed. Topical cyclosporine is shown to be effective for severe cases. (*I+, GQ, SR*)
- For contact lens-related keratoconjunctivitis, discontinue contact lens wear until the cornea returns to normal
- In severe cases, topical cyclosporine or tacrolimus can be considered (I+GQ, DR)
- Use systemic antibiotic treatment for conjunctivitis due to *Neisseria gonorrhoeae* or *Chlamydia trachomatis*
- Treat sexual partners to minimize recurrence and spread of disease when conjunctivitis is associated with sexually transmitted diseases and refer patients and their sexual partners to an appropriate medical specialist
- Refer patients with manifestation of a systemic disease to an appropriate medical specialist

#### **Follow-Up Evaluation**

- Follow-up visits should include
  - Interval history
  - Visual acuity
  - Slit-lamp biomicroscopy
- If corticosteroids are used, perform periodic measurement of intraocular pressure and pupillary dilation to evaluate for cataract and glaucoma

- Counsel patients with contagious varieties to minimize or prevent spread of disease and encourage minimization of contact with other people for 10 to 14 days after onset of symptoms (1+, GQ, SR) in the community
- Inform patients who may require repeat short-term therapy with topical corticosteroid of potential complications of corticosteroid use
- Advise patients with allergic conjunctivitis that frequent clothes washing and bathing/showering before bedtime may be helpful

- Disease onset and course
- Vision impairment
- Ocular, medical, and family history

### **Initial Physical Exam**

- Visual function assessment
- External examination
  - Eyelids and eyelid skin
- Slit-lamp biomicroscopy
  - Presence, extent, and location of corneal thinning or protrusion
  - Indication of previous ocular surgery
  - Presence of Vogt striae, prominent corneal nerves, Fleischer ring, or other iron deposition
  - Evidence of corneal scarring or previous hydrops, and presence of prominent corneal nerves
- IOP measurement
- Fundus examination: assessment of red reflex for dark area, and retina for tapetoretinal degenerations

# **Diagnostic Tests**

- Keratometry
- Corneal topography and tomography
  - Topographic power map
  - Topographic elevation map and tomography
- Optical coherence tomography (OCT)

# **Care Management**

- Therapy is tailored to the individual patient, depending on the visual impairment and a risk/ benefit analysis of each treatment option(s).
- Vision can be corrected with eyeglasses, but contact lenses may be required as keratoconus progresses to correct vision and reduce distortion.
- Rigid corneal gas permeable contact lenses can mask corneal irregularities. Hybrid contact lenses provide higher oxygen permeability and greater RGP/hydrogel junction strength. Piggyback contact lenses may be employed for greater comfort and less epithelial disruption. Scleral lenses may be indicated when RGP and/or hybrid contact lenses fail.
- Intrastromal corneal ring segment implantation can improve contact lens tolerance and BCVA for patients with corneal ectasia, a clear cornea, and contact lens intolerance.
- Cross-Linking (CXL) has long term data supporting its safety and stability and should be considered for patients with early Keratoconus and at risk of progression to arrest or slow progression in its earliest stage.

- Lamellar keratoplasty using DALK techniques can be considered for contact-lens-intolerant patients without significant scarring at Descemets membrane or persistent hydrops. Crescentic lamellar keratoplasty is an option when maximal thinning is in the cornea's periphery.
- Penetrating keratoplasty is indicated when a patient can no longer achieve functional vision with eyeglasses and contact lenses and CXL is contraindicated, or when persistent corneal edema occurs following hydrops. Descemet stripping endothelial keratoplasty cannot correct ectatic disorder.
- Penetrating keratoplasty (PK) is preferred over DALK in cases of deep stromal scarring. Overall, there is insufficient evidence to determine which tecnique offers better overall outcomes. (*I+, GQ, DR*)
- A lamellar graft can be performed for tectonic support when ectasia occurs in the far periphery of the cornea and additional PK can be performed for visual rehabilitation.

### **Follow-Up Evaluation**

- Follow-up visits should include:
  - Interval history
  - Visual acuity
  - External examination
  - Slit-lamp biomicroscopy
  - Assessment of corneal contour and thickness by topography and tomography
  - Measurement of corneal thickness
- With the advent of CXL, more frequent follow-up (i.e., 3-6 months) for progression is now indicated.

- Counsel all patients to avoid eye rubbing.
- Discuss the benefits and potential risks of early crosslinking in patients at high risk for progression or who historically have noted progressive loss of vision.
- Patients undergoing corneal transplantation should be made aware of the warning signs of rejection and should seek medical attention promptly if symptoms occur. The practitioner should be aware of the slit-lamp biomicroscopic findings of epithelial, stromal, and endothelial rejection.

# **Corneal Edema and Opacification (Initial Evaluation)**

#### **Initial Exam History**

- Symptoms and signs: blurred or variable vision often with a diurnial character; photophobia; redness; tearing; intermittent foreign body sensation; intense, disabling, or task-disrupting pain
- Recent history of other ocular surgery
- Age of onset
- Rapidity of onset: acute symptoms vs. gradual or fluctuating
- Persistence: transient or permanent
- Unilateral or bilateral presentation
- Moderating factors or situations
- Past ocular and medical history
- Topical and systemic medications
- Trauma: blunt or penetrating injury to eye or periocular region, forceps delivery, chemical injury
- Contact lens wear: rationale, type of lens, wear time, and cleaning routine
- Family and social history

#### **Initial Physical Exam**

- Visual function assessment
  - Comparison of visual acuity measurement and functional status
- Glare testing
- External examination
  - Evidence of proptosis, ptosis, lagophthalmos, or floppy eyelid syndrome
  - Eyelid or facial asymmetry, scarring, and malfunction
  - Miscellaneous (e.g., pupil responses, corneal diameter, dry eye evaluation)

- Slit-lamp biomicroscopy
  - Unilateral or bilateral signs
  - Diffuse or localized edema
  - Primarily epithelial or stromal edema
  - Evidence of epithelial breakdown, stromal infiltration, epithelial ingrowth, striae, focal thickening, thinning, scarring, interface haze, striae or inflammation, or stromal vascularization or deposits
  - Evidence of guttae, Descemet's membrane tear or detachment, endothelial vesicles, keratic precipitates (KP), pigment, peripheral anterior synechiae
  - Involvement of host tissue, if there is a corneal transplant
  - Evidence of sectoral corneal edema and a cluster line of KP, or an anterior chamber reaction
  - Status, shape, and position of the pupil and iris
  - Status and position of the crystalline lens or IOL and any other intraocular device
  - Evidence of past keratorefractive procedures
  - Healed or recent corneoscleral wounds, areas of scleral thinning associated with previous surgery, surgical devices, and signs of intraocular inflammation.
- IOP measurement
- Fundus examination
- Gonioscopy

#### Diagnostic Tests

- Potential acuity meter
- Rigid contact lens over-refraction
- Pachymetry
- Topography
- Specular microscopy
- Confocal microscopy
- Anterior segment optical coherence tomography
- Ultrasound biomicroscopy

# Corneal Edema and Opacification (Management Recommendations)

#### **Care Management**

- Therapeutic goal is to control the cause of corneal edema or opacity and enhance a patient's quality of life by improving visual acuity and comfort
- In most cases treatment starts with medical management, when this is insufficient, surgery may be considered
- Corneal edema: medical management
  - Lowering an elevated IOP is helpful
  - Topical carbonic anhydrase inhibitors should not be the first line of therapy when endothelial dysfunction is suspected
  - Topical corticosteroid can control inflammation once infection has been ruled out or controlled
  - Microcystic or bullous epithelial disease may produce discomfort or pain necessitating the placement of a bandage contact. Thin lenses with high water content and high oxygen diffusion coefficients may be the most advantageous.
  - Supportive management should be initiated to reduce inflammation and/or pain in cases of acute hydrops
- Corneal edema: surgical management
  - Patients with corneal edema and persistent discomfort, but limited or no visual potential, are generally better candidates for the following procedures:
    - Conjunctival flap
    - Amniotic membrane transplantation
    - A number of scarification procedures
    - Corneal transplantation
  - Endothelial keratoplasty
  - For patients with persistent corneal edema, a number of keratectomy and keratoplasty procedures can be considered.
- Corneal opacification: medical management
  - Corneal opacity treatment can be divided into two phases: a) management of the principal, initiating process (i.e., infection, trauma), and b) management of the resulting problems (i.e., surface erosions and irregularity, scarring, thinning, and vascularization)
  - Conventional treatment involves an antibiotic drop or ointment to protect against secondary bacterial infection
  - Temporary tarsorraphy with botulinum toxin, or suture can be helpful when blinking or lid closure is inadequate
  - A bandage contact lens or amniotic membrane may be useful in cases of delayed healing

- Pressure patching used to be standard treatment, but a recent study found that this does not positively impact comfort or speed of healing (/+, GQ, DR)
- Progressive thinning of cornea or a small perforation usually requires structural support with application of a tissue adhesive.
- Topical corticosteroids are often used to reduce intraocular and corneal inflammation. IOP and cataract formation should be monitored with long-term topical corticosteroid use.
- A rigid gas permeable lens or hybrid or scleral lens when greater stability is needed — will often improve vision when surface irregularity is a factor; such lenses may preclude the need for more invasive procedures
- Corneal opacification: surgical management
  - Surgical strategy for managing corneal opacities depends on the tissue layer(s) involved:
    - Superficial keratectomy may be indicated for removal of superficial deposits
    - Lamellar keratoplasty may be indicated for removal of deeper deposits
    - Penetrating keratoplasty may be indicated for removal of even deeper multilevel opacities
    - Ethylenediaminetetraacetic acid (EDTA) may be used to remove calcific band keratopathy (III, IQ, DR)

# **Follow-Up Evaluation**

- In the management of corneal edema, the goal of follow up is to monitor endothelial dysfunction
- In the management of corneal opacification, follow up to monitor corneal clarity and degree of surface irregularity is necessary
- Coexisting problems, particularly intraocular inflammation and IOP, need regular reassessment

- Provide an understanding of balanced expectations of the amount of visual function that can realistically be preserved or recovered and risk of complications.
- Detailed discussion of the causes of edema or opacity, and various treatment options, is important.
- When the disease process or management is complex, every effort should be made to counsel the patient regarding such challenges to allow for appropriate expectations and informed decisionmaking.
- There is a commercially available point-of-care test to identify Avellino dystrophy in keratorefractive surgery candidates if either family history or clinical findings are inconclusive for this condition.

- Ocular symptoms and signs (e.g., irritation, tearing, burning, stinging, dry or foreign body sensation, mild itching, photophobia, blurry vision, contact lens intolerance, redness, mucous discharge, increased frequency of blinking, eye fatigue, diurnal fluctuation, symptoms that worsen later in the day)
- Exacerbating conditions (e.g., wind, air travel, decreased humidity, prolonged visual efforts associated with decreased blink rate such as reading and using the computer)
- Duration of symptoms
- Ocular history, including
  - Topical medications used and their associated preservatives (e.g., artificial tears, eyewash, antihistamines, glaucoma medications, vasoconstrictors, corticosteroids, antiviral medications, homeopathic or herbal preparations)
  - Contact lens history
  - Allergic conjunctivitis
  - Ocular surgical history (e.g., prior keratoplasty, cataract surgery, keratorefractive surgery)
  - Ocular surface disease (e.g., herpes simplex virus, varicella zoster virus, ocular mucous membrane pemphigoid, aniridia)
  - Punctal surgery
  - Eyelid surgery (e.g. prior ptosis repair, blepharoplasty, entropion/ectropion repair)
    Bell's palsy
- Medical history, including
  - Smoking or exposure to second-hand smoke
  - Dermatological diseases (e.g., rosacea, psoriasis, varicella zoster virus)
  - Technique and frequency of facial washing including eyelid and eyelash hygiene
  - Atopy
  - Systemic inflammatory diseases (e.g., Sjögren syndrome, graft-versus-host disease, rheumatoid arthritis, systemic lupus erythematosus, Stevens-Johnson syndrome, sarcoidosis, scleroderma)
  - Other systemic conditions (e.g., lymphoma, sarcoidosis)
  - Systemic medications (e.g., antihistamines, diuretics, hormones and hormonal antagonists, antidepressants, cardiac antiarrhythmic drugs, isotretinoin, diphenoxylate/atropine, betaadrenergic antagonists, chemotherapy agents, any other drug with anticholinergic effects)
  - Trauma (e.g., mechanical, chemical, thermal)
  - Chronic viral infections (e.g., hepatitis C, human immunodeficiency virus)
  - Nonocular surgery (e.g., bone marrow transplant, head and neck surgery, trigeminal neuralgia surgery)
  - Radiation of orbit
  - Neurological conditions (e.g., Parkinson disease, Bell's palsy, Riley-Day syndrome, trigeminal neuralgia)

• Nonocular symptoms (dry mouth, dental cavities, oral ulcers, fatigue, joint pain, muscle aches, menopause)

### **Initial Physical Exam**

- Visual acuity
- External examination
  - Skin (e.g., scleroderma, facial changes consistent with rosacea, seborrhea)
  - Eyelids: incomplete closure/malposition, incomplete or infrequent blink, eyelid lag or retraction, erythema of eyelid margins, abnormal deposits or secretions, entropion, ectropion
  - Adnexa: enlargement of the lacrimal glands
  - Proptosis
  - Cranial nerve function (e.g., cranial nerve V [trigeminal], cranial nerve VII [facial])
  - Hands: joint deformities characteristic of rheumatoid arthritis, Raynaud phenomenon, splinter hemorrhages underneath nails
- Slit-lamp biomicroscopy
  - Tear film: height of the meniscus, debris, increased viscosity, mucus strands, and foam, break-up time and pattern
  - Eyelashes: trichiasis, distichiasis, madarosis, deposits
  - Anterior and posterior eyelid margins: abnormalities of meibomian glands (e.g., orifice metaplasia, reduced expressible meibum, atrophy), character of meibomian gland secretions (e.g., turbid, thickened, foamy, deficient), vascularization crossing the mucocutaneous junction, keratinization, scarring, eyelid margin hyperemia
  - Puncta: patency, position, presence, and position of plugs
  - Conjunctiva
    - Inferior fornix and tarsal conjunctiva (e.g., mucous threads, scarring, erythema, papillary reaction, follicle enlargement, keratinization, subepithelial fibrosis, foreshortening, symblepharon)
    - Bulbar conjunctiva (e.g., punctate staining with rose bengal, lissamine green, or fluorescein dyes; hyperemia; localized drying; keratinization, chemosis, chalosis, follicles)
  - Cornea: localized interpalpebral drying, punctate epithelial erosions assessed with fluorescein dyes, punctate staining with rose bengal or fluorescein dyes, filaments, epithelial defects, basement membrane irregularities, mucous plaques, keratinization, pannus formation, thinning, infiltrates, ulceration, scarring, neovascularization, evidence of corneal or refractive surgery

# Dry Eye Syndrome (Management Recommendations)

#### **Diagnostic Tests**

- Tear Break-up Time
- Ocular Surface Dye Staining
- Schirmer Test
- Fluorescein Dye Disappearance Test/Tear Function
  Index
- Tear Osmolarity Test

#### **Care Management**

- Treat any causative factors that are amenable to treatment as patients with dry eye symptoms often have many contributory factors
- Specific therapies may be chosen from any category (see Table) regardless of the level of disease severity, depending on physician experience and patient preference
- Artificial tears are safe and effective (I+, GQ, SR)
- Corticosteroids can decrease ocular irritation symptoms, decrease corneal fluorescein staining, and improve filamentary keratitis (*I+, GQ, SR*)
- Silicone plugs may provide symptomatic relief in patients with severe dry eye (*I+, GQ, DR*)
- Autologous serum tears may improve ocular irritation symptoms compared with artificial tears in the short-term
- For mild dry eye, the following measures are appropriate:
  - Education and environmental modifications
  - Elimination of offending topical or systemic medications
  - Aqueous enhancement using artificial tear substitutes, gels/ointments
  - Eyelid therapy (warm compresses and eyelid hygiene)
  - Treatment of contributing ocular factors such as blepharitis or meibomianitis
  - Correction of eyelid abnormalities
- For moderate dry eye, in addition to above treatments, the following measures are appropriate:
  - Anti-inflammatory agents (topical cyclosporine and corticosteroids, systemic omega-3 fatty acids supplements)
  - Punctal plugs
  - Spectacle side shields and moisture chambers
- For severe dry eye, in addition to above treatments, the following measures are appropriate:
  - Systemic cholinergic agonists
  - Systemic anti-inflammatory agents
  - Mucolytic agents
  - Autologous serum tears

- Contact lenses
- Correction of eyelid abnormalities
- Permanent punctal occlusion
- Tarsorrhaphy
- Monitor patients prescribed corticosteroids for adverse effects such as increased intraocular pressure, and cataract formation

#### **Follow-Up Evaluation**

- Purpose is to assess response to therapy as a basis for altering or adjusting treatment as necessary, to monitor for ocular surface damage, and to provide reassurance.
- Frequency and extent will depend on the severity of disease, therapeutic approach and response to therapy.

- Patient education is an important aspect of successful management
- Counsel patients about the chronic nature of dry eye and its natural history.
- Set and discuss realistic expectations for therapeutic goals
- Provide specific instructions for therapeutic regimens.
- Reassess periodically the patient's compliance and understanding of the disease, risks for associated structural changes and realistic expectations for effective management, and reinforce education.
- Refer patients with manifestation of a systemic disease to an appropriate medical specialist.
- Caution patients with pre-existing dry eye that keratorefractive surgery, particularly LASIK, may worsen their dry eye condition.