A Vision Center of Excellence newsletter focused on ocular casualty care and readiness

FRONTLINES OF EYE CARE

SPRING 2019

IN THIS ISSUE

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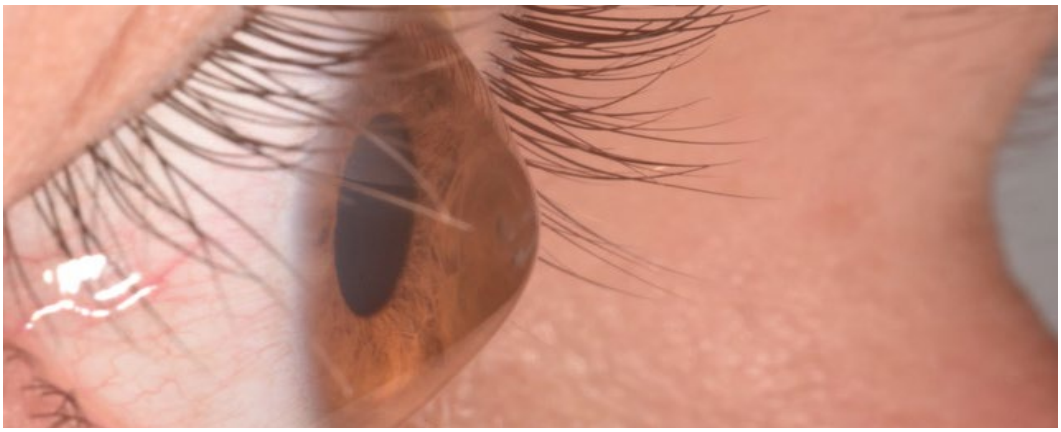


Keratoconic cornea. Note: Protrusion and irregular curvature. (Source: Thomas Arnold, OD, FSLS)

▸ FEATURE

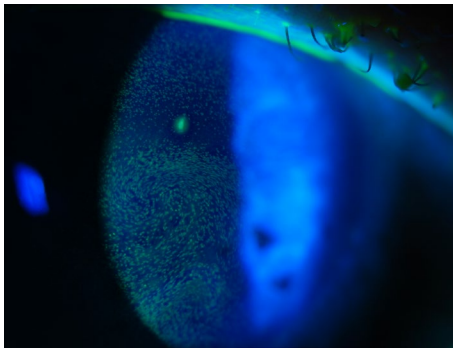
DISEASE: NON-BATTLE EYE INJURY – CORNEAL ECTASIA

Corneal ectasias are a category of eye diseases characterized by progressive steepening and thinning of the collagen-based corneal stroma.[1] Individually, they are part of their own unique primary disease process or can occur as result of refractive surgery. Conditions characterized as corneal ectatic diseases include keratoconus, pellucid marginal degeneration (PMD), keratoglobus, and post-refractive surgical ectasia. These disorders can be differentiated based on the pattern and location of corneal thinning, age of onset, and surgical history. Keratoconus usually presents around puberty, progresses until approximately 40 years of age, and is characterized by inferior thinning and protrusion of the cornea at its thinnest point.[2] PMD usually presents between 20-40 years of age and is characterized by corneal thinning inferiorly with protrusion of the cornea superior to the area of thinning.[3] Keratoglobus is most often congenital, though may be acquired secondary to systemic conditions and consists of generalized thinning of the entire cornea and globular protrusion.[3] Post-refractive corneal ectasia is corneal thinning with a clinical appearance similar to keratoconus, however, by definition, it occurs in patients with a history of either LASIK (laser-assisted in situ keratomileusis), PRK (Photorefractive Keratectomy), Radial Keratotomy (RK) or SMILE (Small Incision Lenticule Extraction).

Keratoconus is the most common corneal ectatic disease and is a

bilateral, asymmetric, multifactorial disease of the cornea. It affects both genders equally, and there are variable data supporting ethnicity as a factor. Some studies suggest that the prevalence is higher in Asians as compared to Caucasians.[4] Prior to the availability of more advanced diagnostic technologies such as corneal tomography, the incidence of keratoconus had been estimated as 1 in 2,000 in the general population[5] and a corresponding prevalence rate of 54.5 per 100,000 (per year).[6] When this data was collected in the mid to late 1980s, the diagnosis of keratoconus was based on using a retinoscope (scissors reflex) and keratometry (distorted mires);[6] both of which are inherently limited in their ability to evaluate changes in the corneal stroma or the cornea's anterior and posterior surface characteristics. In addition, the results from these tests are moderately subjective. Modern computerized corneal topography and tomography devices provide more accurate and repeatable information about the cornea and its structure, including progression analysis of the disease. As recently as 2017, and in conjunction with the advent of these newer and more sensitive computerized diagnostic methods, the annual incidence and the prevalence of keratoconus were shown to be 5-fold to 10-fold higher than previously reported.[7]

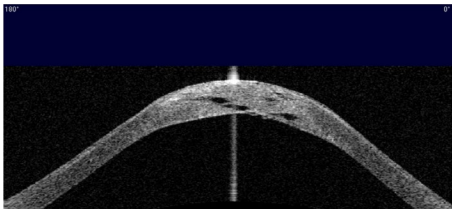
The diagnostic criteria of keratoconus include a loss of best corrected visual acuity, irregular corneal curvature patterns, tomographic front and back surface elevation changes greater than normative limits, corneal thinning, and the amount and location of corneal scarring.[1] These



Ocular surface disease in an individual with keratoconus. Note: Irregular whirl pattern of the corneal epithelial cells. In this image, the whirl is circling the apex of the cone. Note the superior corneal cells in a normal staining pattern not affected by the irregular corneal shape. (Source: Andrew S. Morgenstern, OD, FAAO, FNAP)

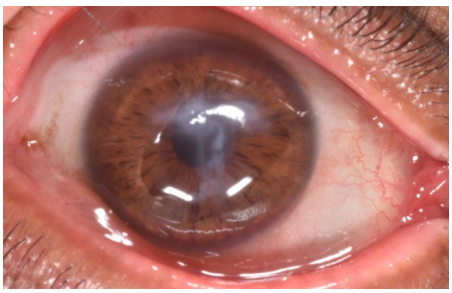
symptomatic changes are partly caused by the anterior movement of corneal tissue that is aberrant and unstable to varying degrees. The etiology of keratoconus is complex with many factors including genetics (including a positive family history), biomechanical influences and dysfunctions, numerous systemic disorders (such as connective tissue disorders), mechanical factors like eye rubbing and contact lens wear, and environmental causes. Allergic eye disease and atopic dermatitis have also been strongly associated with keratoconus.[8,9] Because of the irregular surface of the cornea in keratoconus, ocular surface disease and irregular corneal epithelial remodeling are common occurrences that may contribute to the loss of visual acuity and increased discomfort.[10] Commonly, it is the ocular surface discomfort that leads to the urge to rub the eyes. Unfortunately, eye rubbing has also been associated with corneal thinning. Therefore, overall and localized corneal thickness measurement play a significant role in the diagnosis, management, and monitoring of keratoconus.

Three suspected factors in the development of keratoconus are thin corneas, mechanical fatigue, and increased biochemical activity. While the average normal corneal thickness ranges between 510-575 microns[11], corneas thinner than 470 microns are considered suspect for keratoconus.[12] The mechanical fatigue and thinning of the corneal tissue with or without eye rubbing can lead to more significant breakdown and can advance the disease. Biochemically, an increase in the activity of proteinase enzymes and a decrease in the activity of proteinase inhibitors in the cornea can also result in a breakdown and progressive thinning of the corneal stroma.[13] If the breakdown of tissue becomes significantly advanced, corneal hydrops, a rare, acute complication of keratoconus can occur. Corneal hydrops is characterized by corneal stromal edema leading to pain, decrease in visual acuity, and photophobia. In corneal hydrops, weakened collagen and corneal thinning leads to rupture of the posterior surface of the cornea (endothelium and Descemet's membrane). The rupture in tissue, in turn allows leakage of aqueous humor from the anterior chamber to penetrate into the corneal stroma. Treatment is commonly achieved with topical medications such as a hypotonic



Corneal hydrops. Note: Stromal swelling and rupture of posterior cornea. (Source: Edward Boshnick, OD)

solution to draw out excess fluid in the corneal stroma as well as a topical steroid and/or cycloplegic agent for control of pain, inflammation and photophobia. Rarely, an intracameral injection of air or gas can be performed to shorten the healing period. Upon resolution of corneal hydrops, corneal scar formation frequently occurs.[14] If the corneal scar is even remotely in the visual axis, significant and permanent vision loss can occur. Corneal transplantation (full or partial thickness) as a treatment to remove scarred corneal tissue occurs more frequently in this population, with age of onset of keratoconus having predictive value for rate and severity of disease to lead to corneal transplantation.[15]

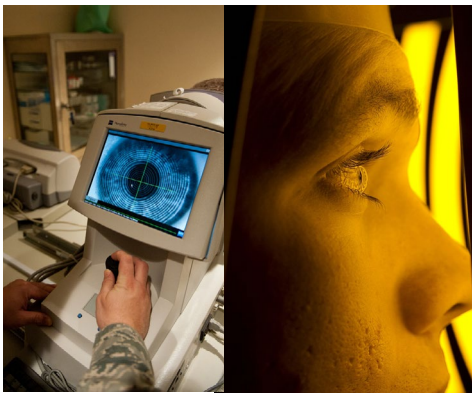


Corneal hydrops and resulting corneal scar. (Source: Edward Boshnick, OD)

DIAGNOSTIC TESTING

Corneal topography

(computerized analysis of Placido disk imaging) provides computerized evaluation of the anterior surface curvature or shape of the cornea and is a good method to evaluate for curvature changes related to corneal ectatic disease. However, the analysis does not assess the corneal thickness measurement (pachymetry) or the cornea's posterior surface curvature. In a cornea absent of disease, a normal reading for anterior corneal curvature is approximately



(L): Corneal Topographer with placido disc images on device screen.(R): Corneal Topographer with placido disc images reflecting off patient cornea. (Source L: U.S. Air Force photo by Airman 1st Class Kate Thornton/Released. Source R: U.S. Air Force photo by Staff Sgt. Perry Aston/Released)

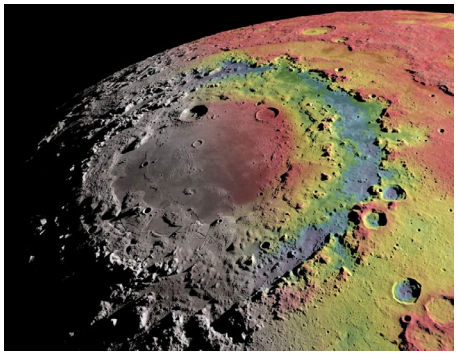
44.00-45.00 diopters (D). A topographic map showing the following are all suspicious for keratoconus:[16]

- Greater than 5 D of astigmatism
- Associated central corneal thickness of less than 470 microns
- Maximum corneal curvature (Kmax) value steeper than 48 D

Corneal Tomography

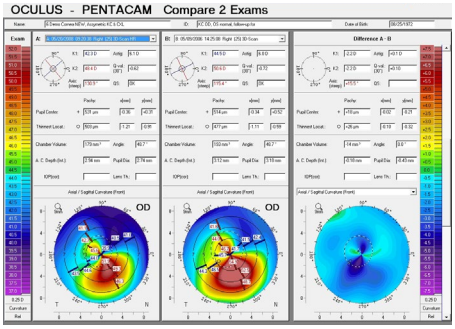
(Scheimpflug imaging) provides a comprehensive evaluation of the elevation and curvature of the anterior and posterior surfaces of the cornea, including pachymetry of the entire cornea. Elevation of the posterior surface via tomography provides evidence of the earliest changes in corneal ectasia.[14] As progressive corneal thinning is a hallmark of keratoconus, the only way thinning can occur is if the posterior surface of the cornea elevates at a more significant rate than the anterior surface. While curvature data is valuable in diagnosing keratoconus, tomography can better determine these elevation measurements where topography cannot. Tomography uses a peripheral, rotating camera that takes multiple images. Like a satellite collecting surface images while orbiting the moon, by comparing the multiple images from different locations, the device can develop elevation, curvature, and pachymetric maps of both the front and back surface of the cornea.

These tomographic maps can be compared against a normative database to assess for risk of progression and corneal ectasia.



Satellite image of elevation mapping of the moon. (Source: https://svs.gsfc.nasa.gov/4499)

Progression of disease can also be evaluated by difference maps that compare an individual's first examination to their subsequent examinations and create a third map which highlights differences or the amount of change over a certain time period.

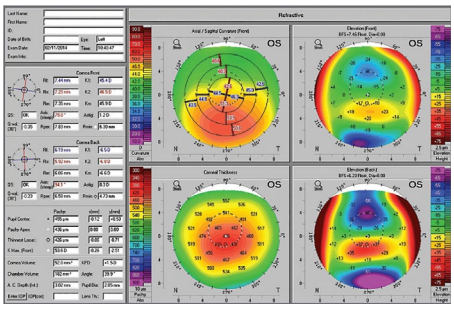


Difference tomographic map comparing two examinations for progression of keratoconus. (L) First image; (C) second image, (R) Difference map. (Source: Andrew S. Morgenstern, OD, FAAO, FNAP)

Other ways to look for keratoconic progression is pachymetric change (rate of corneal thickness change from the center of the cornea to the periphery), change in curvature, and anterior and posterior corneal elevation analysis. If these maps appear to have a decrease in pachymetry, abnormal pachymetric progression, or an increase in elevation or curvature, a diagnosis of progression can be made. A tomographic anterior elevation map of greater than +10 microns over the best fit sphere on the anterior surface combined with more than +15 microns of elevation over the best fit sphere on the posterior surface is commonly outside of normal limits and suspicious for corneal ectasia.[17] An abnormal finding for pachymetric progression is more than two standard deviations from the normative database and will be automatically identified on the pachymetric progression map.

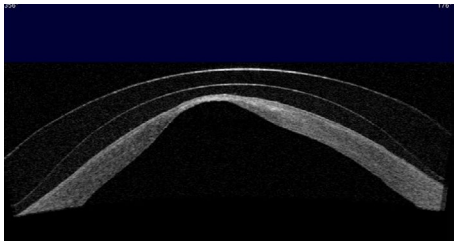
Anterior Segment Optical Coherence Tomography (AS-OCT)

provides a high-resolution, cross-sectional image of the anterior segment using optical light scatter. Most AS-OCT devices can image the cornea, corneal limbus, peri-limbal conjunctiva, anterior chamber, iris, and the anterior crystalline lens, and thereby can evaluate measurement of corneal pachymetry, anterior chamber depth, and anterior chamber angle.



Corneal Tomography. Note: Central anterior and posterior elevation values. (Source: https://www.ophthalmologymanagement.com/issues/2015/june-2015/corneal-topography-comes-of-age)

AS-OCT also provides the highest quality image to aid in the assessment of medical contact lens fitting of an ectatic cornea.[18] While AS-OCT can effectively measure corneal pachymetry in cases of corneal ectasia, the software does not have a normative database to compare against and currently does not have similar anterior surface map software for analysis similar to topography or tomography. However, assessing progression of corneal ectasia can be achieved by measuring corneal pachymetry on sequential exams and monitoring for corneal thinning.



AS-OCT image of keratoconic ectatic cornea and scleral contact lens. Note: Posterior corneal elevation. (Source: Edward Boshnick, OD)

CURRENT AND EMERGING TREATMENTS

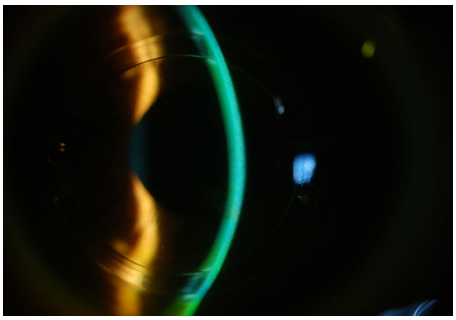
Spectacles/Contact Lenses

Mild or early ectatic disease can be corrected with spectacles and/or soft spherical toric contact lenses. Soft contact lenses are more comfortable and generally preferred to rigid contact lenses. If the disease progresses to the point where spectacles and commercially available soft contact lenses are not providing adequate vision, or if the contact lenses are uncomfortable, a variety of medical contact lenses are available to improve the patient's vision. Rigid gas permeable (RGP), hybrid soft skirt, piggyback, scleral contacts, and custom prosthetic molded lenses are options that may be designed for the individual cornea. There is a trend to fit ectatic corneas with scleral contact lenses because they can improve best corrected vision while elevating the lens over the keratoconic cornea and avoiding the development of scar formation from contact lens/anterior corneal surface contact. Contact lenses are indicated for all severity levels of corneal ectasia, however, the use of contact lenses does not slow or halt the progression of the disease.[19]

Intra Corneal Ring Segments (ICRS)

ICRS are polymethylmethacrylate (PMMA), clear, semi-circular rounded segments that are placed 2/3 of the depth of the stroma in the mid-periphery via a surgically developed channel created by either a circular microkeratome or femtosecond laser. By elevating the mid-periphery of the cornea, there is a corresponding flattening of the central cornea which is designed to reduce or eliminate the myopia and astigmatism from keratoconus.[24] ICRS are used to re-center the paracentral cone of an ectatic cornea, "regularize" refractive error and astigmatism, reduce higher order aberrations of the visual system, and improve the refractive performance of the eye. Like contact lenses, ICRS do not slow or halt the progression of the corneal ectasia. ICRS have been shown to reduce steepening and corneal astigmatism associated with keratoconus. Generally, results have been less

predictable than contact lenses for improving best corrected visual acuity. Additionally, there have been concerns about making an incision into a biochemically weak ectatic cornea for the purposes of refractive improvement. The indications for ICRS candidates are individuals with keratoconus and contact lens intolerance with no corneal scarring, and can be used with Corneal Cross Linking.



Intra Corneal Ring Segments in combination with CXL. (Source: Andrew S. Morgenstern, OD, FAAO, FNAP)

Corneal Cross-linking (CXL)

In April 2016, the U.S. Food and Drug Administration (FDA) approved CXL as the only known treatment to slow or halt progression of certain corneal ectatic diseases.[20] Currently, the only FDA-approved uses for CXL are treatment of progressive keratoconus and treatment of corneal ectasia following refractive surgery. Progressive keratoconus is diagnosed when at least two of the following three parameters are evident:

- Steepening of the anterior corneal surface
- Steepening of the posterior corneal surface
- Thinning and/or an increase in the rate of corneal thickness change from the periphery to the thinnest point.[14]

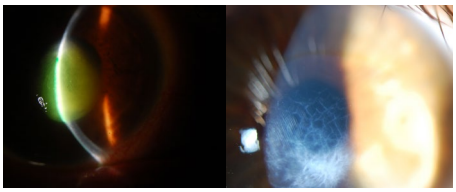
While it is common to have a decrease in best corrected visual acuity with progressive keratoconus, it is not required for the diagnosis.[14] Discussing post-CXL expectations with the patient prior to the procedure to manage expectations of pain and prognosis is very important. Further, the patient should understand that CXL is not a refractive improvement tool.

The current FDA-approved dosage and administration method is based on the Dresden Protocol,[21] or epithelium-off CXL (Epi-Off). In this protocol, a photo enhancer such as riboflavin 5'-phosphate 0.146% with or without 20% dextran (with and without are two different ophthalmic solution formulations), is used in combination with the UVA light source for 30 continuous minutes at 3mW/cm² at a wavelength of 365 nm, centered over the cornea.[22] This technique generates a singlet oxygen molecule which results in shortening of collagen fibrils and the development of new cross-linking of existing collagen.[23] This method results in strengthening of the corneal tissue, and thereby slows or halts the progression of corneal ectatic disease.

Epi-Off CXL underneath UVA light source.[25] (Source: DoD photo by Reese Brown)

While the above Epi-Off method has shown to be safe and effective for the desired indications, CXL in an epithelium-on (Epi-On) method has shown to be effective as well. Though only the Epi-Off method is currently FDA approved, there is ongoing debate as to if one method is superior. Complications can occur with Epi-Off or Epi-On CXL; Epi-Off has been shown to have a likely higher risk of ulcerative keratitis, corneal infiltrate, and corneal opacity/haze caused by debridement of the epithelium. Currently, studies are underway for possible future FDA approval for Epi-On crosslinking.[14] As a result of CXL, the number of corneal transplant surgeries has decreased by approximately 25%.[24]

Emerging treatments that are less invasive and provide quicker recovery than current Epi-On CXL protocols are on the horizon. One example currently being investigated is a slit lamp-mounted CXL UVA irradiation source to make the procedure more portable because a smaller device is used.[21] Secondly, accelerated and targeted CXL using the Bunson-Roscoe law of reciprocity (increasing power of the light source and decreasing exposure time) will likely become



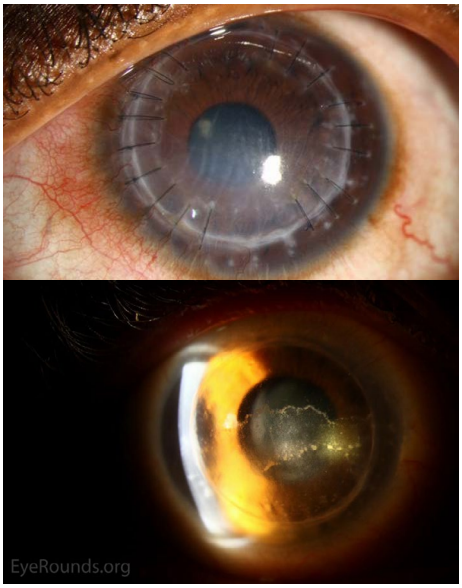
(L) Epi-Off post CXL cornea with central corneal infiltrate. (R) Same cornea after resolution of central corneal infiltrate. Note: Central scarring, Vogt Striae, and Fleischer's Ring are still observable. (Source: Andrew S. Morgenstern, OD, FAAO, FNAP)

more available.[22] Lastly, studies are underway on eye drops applied to the ocular surface to treat and halt progression of keratoconus absent of any UVA light source irradiation or procedure.

Lamellar Keratoplasty and Penetrating Keratoplasty

In cases of severe or advanced corneal ectasia, where vision cannot be corrected with spectacles, contact lenses, ICRS, CXL or combination therapy, or when less invasive treatments are not effective or are contraindicated, a corneal transplant (keratoplasty) may be necessary. Either full thickness penetrating keratoplasty (PK) or a partial thickness, deep anterior lamellar keratoplasty (DALK) corneal transplant may be indicated depending on the severity of disease.[14] After transplantation and initial recovery, medical contact lenses may be required for visual recovery. Surgical recovery for a corneal transplant patient for the treatment of keratoconus can take a minimum of 12 months with frequent follow-up visits and close monitoring of medications. Complications can occur with keratoplasty, including:[26]

- Graft rejection
- Graft infection
- Glaucoma
- Uveitis
- Retinal detachment
- Epithelial defect
- Choroidal hemorrhage
- Wound leak
- Endophthalmitis
- Suture break
- Suture migration
- Increase in refractive error
- Return of keratoconus in the graft cornea (very rare)



(Top) Corneal transplant decompensation and early graft rejection. Note: Inferior corneal edema. (Bottom) Corneal transplant rejection. Note: Line of white blood cells on corneal endothelium (Khodadoust line). (Source: https://webeye.ophth.uiowa.edu/eyeforum/atlas/pages/Khodadoust2/index.htm)

Waiver Guidelines for Active Duty Service Members

Eligibility for continued service for Service members with corneal ectasia: Military service with corneal ectasia may be considered on a case-by-case basis and may be waiverable. Examples of these waivers are listed below by service branch:

U.S. Air Force Waiver Guide

Keratoconus and Abnormal Corneal Topography

Last Update: April 17, 2019

Significant Change: Corneal collagen crosslinking is approved for all trained aircrew members.

Diagnosed corneal ectasia is a disqualifying condition for all flying classes and is not waiverable. Waivers for abnormal corneal mapping that is not diagnostic of keratoconus is possible and will be considered on a case-by-case basis. Trained aircrew members that are diagnosed with keratoconus must obtain frequent evaluations to ensure that they are adequately corrected to mitigate the optical side effects of the condition. In addition, aircrew members must be adequately corrected with spectacle back-ups. Evaluation of stereopsis with contact lenses and spectacles is required.[27]

U.S. Army Waiver Guide

Keratoconus Requirements and Aeromedical Concerns

Last Update: May, 2019

Aeromedical Concerns:

There are significant aeromedical concerns for those with any amount of keratoconus. Keratoconus is considered disqualifying for all classes of aviation duty. Blurred vision can interfere with flying. Initial flight applicants are not considered favorably for exception to policy. There is a long-term risk of corneal scarring. Keratoconus "suspects" who do not have definitive keratoconus will be reviewed on a case-by-case basis for waiver or exception to policy. Patients whose best corrected acuity falls below 20/20 or those requiring corneal transplant will be disqualified from flying. Aviators with rapidly increasing myopia or astigmatism may warrant keratoconus testing.

Rated and Non-Rated Aircrew to Include Class 2/3/4 Applicants:

A waiver may be possible for all other aviation classes in the early stages of keratoconus provided visual standards are met. For this group, an optometry or ophthalmology consult with corneal topography is required. In addition,

as there is an association with keratoconus and some systemic diseases, the exclusion of connective tissue as Marfan's or disorders such as Ehlers-Danlos syndromes may be indicated. At minimum, and dependent on the progression of the disease, an annual eye exam by an optometrist or ophthalmologist is required. Spectacles and/or hard contact lenses may be necessary to restore visual acuity to acceptable standards. Hard contact lens wearers must have in possession a pair of spectacles with corrected vision to 20/20.[28]

U.S. Navy Waiver Guide

Keratoconus, Pellucid Marginal Degeneration, or Corneal Ectasia

Last Update: May, 2019

Waivers will typically not be considered for applicants with suspected, forme fruste (early presence), frank keratoconus, or corneal disease, but may be considered in designated personnel if visual acuity is 20/20 or correctable to 20/20 with spectacles. History of CXL will be considered on a case-by-case basis for designated members. Contact lens use in any aviator requires specific authorization on the aeromedical clearance form and must comply with the naval aviation contact lens policy. However, contact lenses for therapeutic reasons such as keratoconus are not approved. *[Note: Student Naval Aviator (SNA) applicants must have corneal mapping performed (Topography or Pentacam) to rule out corneal disease (forme fruste keratoconus, other ectasias, or disqualifying conditions, etc.). Evaluation of stereopsis with contact lenses and spectacles is required.][29]*

Keratoconus of any degree or a history of a partial or full thickness corneal transplant is considered to be disqualifying from entering the military. If an Active Duty Service Member (ADSM) is on active duty in the military when diagnosed with corneal ectasia with distance visual acuity of any degree, and the condition cannot be corrected with spectacle lenses to at least 20/40 in each eye, at distance or near, the ADSM's condition could possibly be cause for a medical evaluation board for separation from service. Often, a diagnosis of corneal ectasia may require the use of soft or rigid contact lenses to achieve best corrected visual acuity. However, Army, Navy, and Marine personnel (except in accordance with policy) are prohibited from wearing contact lenses (soft or rigid) to achieve optimal vision while deployed. Non-Aircrew and Air Force personnel are also prohibited from wearing contact lenses while deployed, however, Air Force aircrew members are an exception to this rule (i.e., soft contact lenses only). As such, the diagnosis of corneal ectasia in an ADSM can negatively affect the individual, unit readiness, and quality of life.

Surveillance of Idiopathic Corneal Ectasias, Active Component

Data were requested from the Armed Forces Health Surveillance Branch (AFHSB) of the Defense Health Agency to characterize the incidence and prevalence of idiopathic corneal ectasias. Data were obtained from the Defense Medical Surveillance System (DMSS) on outpatient encounters of active component members of the Army, Navy, Air Force, and Marine Corps. The surveillance case definition used the ICD codes listed at the end of this report. Incident cases were counted if there was as at least one outpatient medical encounter with a qualifying diagnosis in any diagnostic position. Incident cases of corneal ectasias with any prior outpatient encounter that included a CPT code for refractive surgery (CPT: S0800 or S0810) in any CPT position were excluded, and person-time was censored at the time of the first occurring refractive surgery procedure. Annual lifetime prevalence of each type of disorder was calculated with prevalent cases (in active component service during the given year and diagnosed as an incident case on or prior to that year). The denominator for the prevalence rate is the number of Service members in active component service during each year.

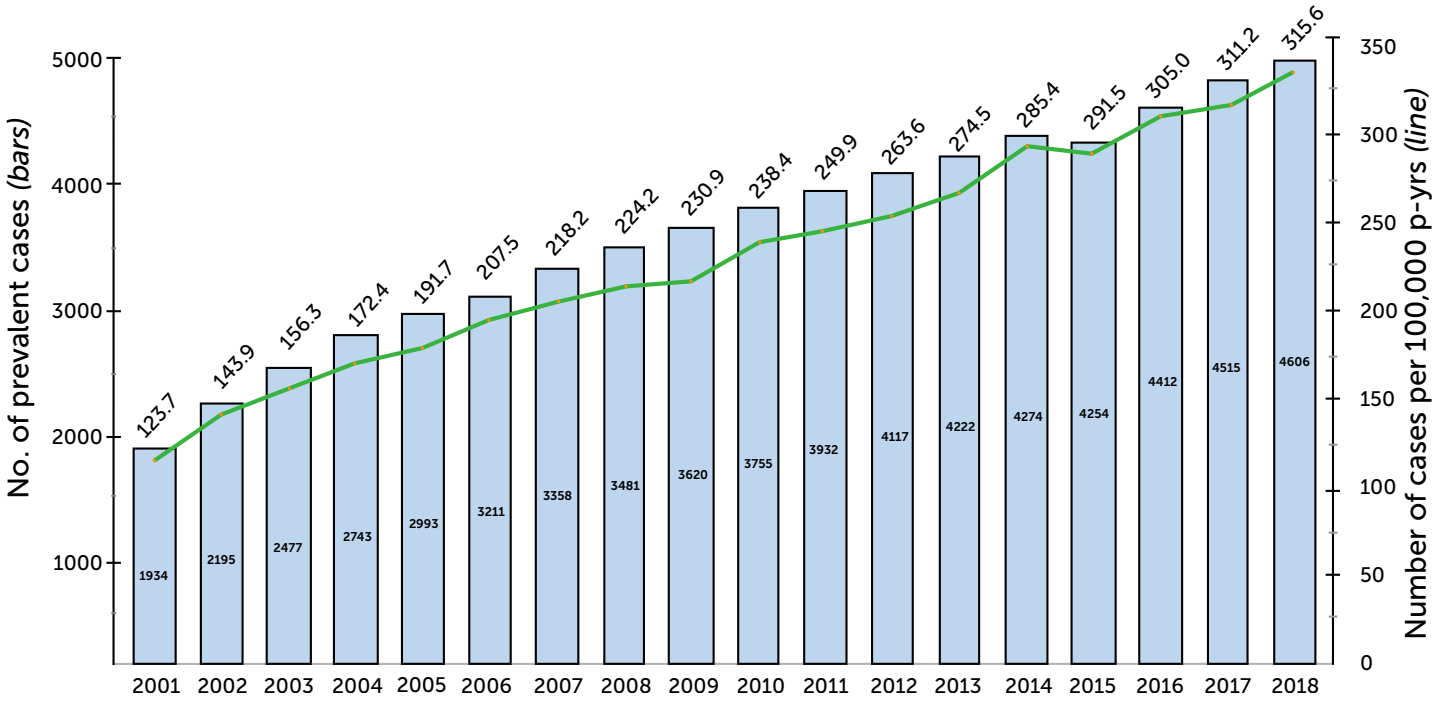
The table displays the incident cases and rates among different groups. Overall rates of corneal ectasias were 45.3 per 100,000 person-years with the above case definition. Rates were found to be fairly similar when comparing male to female Service members. Rates were highest among Service members 30-34 years of age and lowest among the < 20 years of age group. Incidence rates were highest among the O1-O3/W1-W3 rank group, and lowest among the E1-E4 group. Rates of corneal ectasias were highest among Army personnel and lowest among the Marines. Healthcare occupation groups had the highest rates, with the second highest rates being among pilot/aircrew occupational groups.

During the surveillance period, the incidence rates of idiopathic corneal ectasias followed a general upward trend, from 30.8 per 100,00 person-years in 2001 to 57.7 person-years in 2018. The rates peaked in 2016 at 65.1 per 100,000 person-years. Annual lifetime prevalence rates of idiopathic corneal ectasias increased steadily throughout the surveillance period, from 123.7 per 100,000 persons in 2001 to 315.6 per 100,000 persons in 2018.

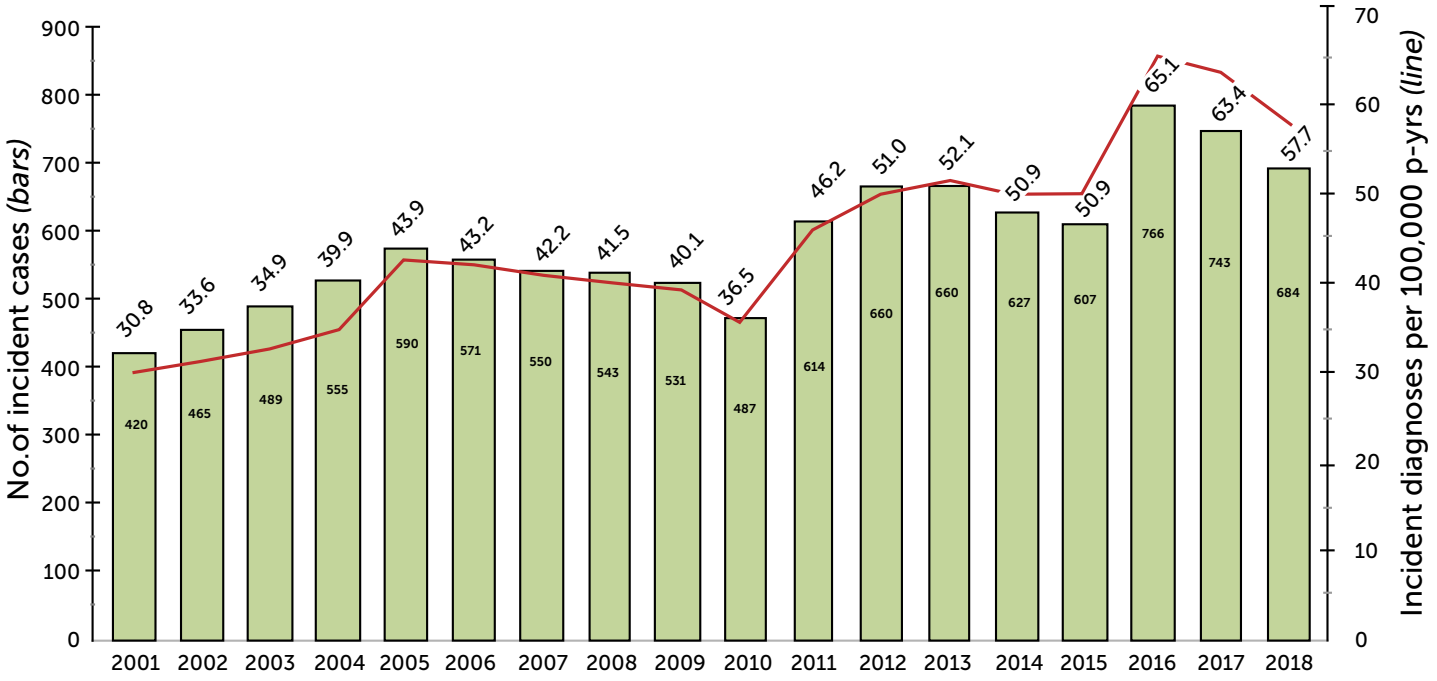
While rates of keratoconus among the U.S. population are available, there are no direct comparisons for the wider group of corneal ectatic conditions. Across the active component population, the increasing incident rates reported above may be caused by multiple factors including increased recognition and improved diagnostic capability. Steadily increasing annual prevalence is reflective of the chronic nature of these conditions once diagnosed. Due to the high potential impact on both readiness and retention of Service members with these conditions, health care providers (ophthalmic and non-ophthalmic) must be aware of the signs/symptoms and referral options for those suspected of having the disease.

Incident Cases	Count	Rate[a]
Total	10,562	45.3
Sex		
Males	9,132	46.0
Females	1,430	41.4
Age group (years)		
<20	1,000	30.3
20-24	2,518	39.6
25-29	2,726	52.1
30-34	1,845	54.2
35-39	1,386	51.3
40+	1,087	46.5
Service		
Army	4,504	52.6
Navy	2,335	40.2
Air Force	2,835	49.4
Marine Corps	888	27.6
Rank		
Junior Enlisted (E1-E4)	4,194	39.6
Senior Enlisted (E5-E9)	4,385	48.8
Junior Officer (O1-O3, W1-W3)	1,294	55.5
Senior Officer (O4-O9, W4-W5)	689	48.6
Military occupation		
Combat-specific	1,173	36.2
Motor transport	349	48.2
Pilot/air crew	250	27.9
Repair/engineering	2,826	41.0
Communications/intelligence	2,630	50.8
Health care	1,125	58.5
Other	2,209	49.5

Counts and rates of incident diagnoses of idiopathic corneal ectasias, by demographics and military characteristics, active component, U.S. Armed Forces, 2001–2018.



Numbers of prevalent cases and lifetime annual prevalance of idiopathic corneal ectasias diagnoses, active component U.S. Armed Forces 2001-2018.



Numbers of incident cases and incidence rates of idopathic corneal ectasias diagnoses, active component, U.S. Armed Forces, 2001–2018.

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DIAGNOSIS, MANAGEMENT, AND CODING OF IDIOPATHIC CORNEAL ECTASIA FOR THE EYE CARE PROVIDER

Irritants and/or pollutants causing a Service member to rub their eyes. (Photo by Sgt. Samuel Beyers, U.S. Marine Corps Forces, South)

Diagnosis

A comprehensive eye examination should be performed on any individual suspected of having corneal ectasia or keratoconus. Early detection and diagnosis is essential to direct early intervention. Diagnosis of keratoconus allowing interventions is a force readiness consideration. Delayed detection of the disease may lead to vision loss and affect a Service member's ability to fight, serve, and protect fellow Service members. Early detection, management, and proper treatment can halt disease in its early phase and maintain Service members' visual readiness on or off the battlefield.

Awareness of suspicious findings and clinical signs will help warrant early diagnosis.

Findings Suspicious for Keratoconus

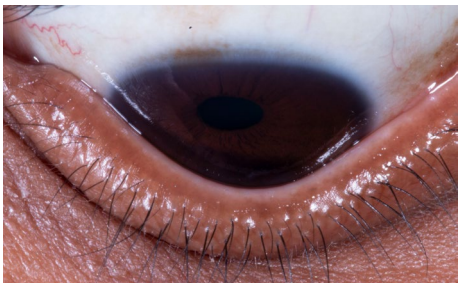
- Corneas steeper than 48 D.
- Corneas thinner than 470 microns.
- Corneal topographic maps with greater than 5 D of astigmatism.
- Increasing myopia absent of change in axial length.

> Diagnosis of keratoconus before overt clinical signs and symptoms will provide the highest chance of achieving best long-term visual outcome.

Slit Lamp and External Examination

If computerized corneal mapping is not available to detect early corneal ectasia, there may be common slit lamp and external findings in later stage disease:

- **Vogt Striae –** Vertical (rarely horizontal) stress/compression lines at the level of Descemet's membrane. Vogt Striae commonly lead to corneal stromal scarring.
- **Munson's Sign –** In downgaze, the lower eyelid will appear to have a "V" shaped pattern/deformation.
- **Fleischer's Ring –** An iron deposition in the basal epithelial corneal cells.
- **Rizzuti's Sign –** A bright reflection of the nasal corneal limbus when illuminating the temporal corneal limbus.
- In moderate to severe cases of keratoconus, it is possible to see the conic shape of the cornea from outside of the slit lamp. Observe the cornea from the side of the patient while the patient is in forward gaze to identify corneal shape irregularity.



Munson's Sign, "V" shaped lid formation in downgaze. (Source: Thomas Arnold, OD, FAAO, FSLS)

Management

Preservation and improvement of best corrected visual acuity is the goal in managing keratoconus. Improving visual acuity and being able to perform civilian daily tasks and successfully operate within Military Occupational Specialties (MOS) visual standards is paramount. While no two individuals with keratoconus will have exactly the same improvement, staging the

severity of disease and risk analysis will likely dictate which management option will improve visual acuity best. It is important to be aware that keratoconus is a dynamic disease and has the potential for progression; this progression of disease is common and in most cases can occur up to age 40. Therefore, over time, a patient can progress in their requirements of management.[1] Early in the disease, a patient may require spectacles and/or soft contact lenses for visual improvement. However, as the disease progresses, the cornea can get steeper and increase in astigmatic or spherical magnitude and require more advanced contact lens therapy. Any cornea can worsen enough as to require a corneal transplant. Management and follow -up practices vary depending on age of onset and severity of disease. Once a positive diagnosis has been made, the interval between testing and examinations should be shorter for younger patients and the same measurement platform should be used in sequential examinations.[14] In addition to the refractive, medical and surgical interventions, individuals with keratoconus should always be advised to:

- Discontinue eye rubbing.
- Address any ocular allergy or atopy issue.
- Lubricate the eyes with artificial tears throughout the day.
- Return for evaluation if there is a suspicion of refractive change or change quality of vision with the current refractive correction modality.

Corneal Cross Linking (CXL)

If you notice any signs of possible progression of keratoconus, consider CXL as a possible treatment to slow or halt the disease process.

- CXL is currently performed in an epithelium-off CXL fashion (Epi-Off).
- CXL is approved for progressive keratoconus and post-refractive surgery ectasia.
- CXL is not a refractive correction.
- Progression in keratoconus can occur in most cases up to age 40 and in rare cases, after CXL.
- Recovery and return to duty can take up to 3 months.

Progression of Keratoconus (with or without CXL)

Two of the following three parameters are required to document keratoconic progression:

- Steepening of the anterior corneal surface.
- Steepening of the posterior corneal surface.
- Thinning and/or an increase in the rate of corneal thickness change from the periphery to the thinnest point.[14]

Treatment considerations

Refractive and Clinical/Surgical Considerations (in order of severity of disease)

- **Mild**
  - Attempt to wear spectacles as a primary vision correction device in the early stages of the disease.
  - Use soft spherical or soft toric contact lenses.
  - Consider CXL if there is documented progression of corneal ectasia and/or young age.
- **Moderate** (after following the above if spectacles and soft contact lenses are not successful)
  - Scleral, rigid gas permeable, piggyback, hybrid/soft skirt or other medical contact lenses are all refractive options for the Service member with corneal ectasia.
  - Consider prosthetic scleral contact lenses.
  - Consider CXL if there is documented progression of corneal ectasia.
  - Consider ICRS if the patient is contact lens intolerant with a clear cornea.
  - Combine CXL and ICRS.
- **Severe**
  - Perform Corneal Transplant (full thickness or partial thickness).
  - Use medial contact lens post-operatively.

Possible Prescription and Non-Prescription Pharmaceutical Formulations for Chronic Corneal Ectasia and/or Corneal Ectasia Associated Complications

- Anti-allergy topical ophthalmic formulation
- Artificial tear and artificial tear gel
- Anti-inflammatory topical ophthalmic formulations
- Anti-infective topical ophthalmic formulations

Low Vision and Blind Rehabilitation Considerations

- Applies to rare cases where best corrected visual acuity is less than 20/70 in the better seeing eye even after surgical or non-surgical/refractive intervention for keratoconus.
- Advanced Vision Care Services are available to ADSM and Veterans through the Department of Veterans Affairs

(VA) Blind and Low Vision Rehabilitation Services (BRS).
- For more information on the VA BRS, please visit: https://www.va.gov/health-care/about-va-health-benefits/vision-care/blind-low-vision-rehab-services/

Documentation and Coding Guide

For proper monitoring of the keratoconus patient, specific measurements and clinical observations are critically important to identifying changes in disease status. Documentation and evaluation for progression in the following is suggested at all keratoconus follow up visits:
- Medical, ocular, and family history
- Corrected and uncorrected visual acuity
- Fit and centration of contact lens (if applicable)
- Intra ocular pressure
- Evaluation of eyelids
- Refraction
- Slit lamp evaluation
- Global corneal pachymetry
- Keratometric measurements
- Corneal topography
- Corneal tomography
- AS-OCT (as available)

In cases of prior surgical intervention (CXL, ICRS, or corneal transplantation), documentation and evaluation of the following is required:
- Stability of refraction and pachymetry
- Evidence of ring segment movement
- Missing or misplaced suture
- Intra ocular pressure
- Evidence of infection, inflammation, or rejection in cases of corneal transplant

Longitudinal analysis of clinical data provides valuable information to identify trends, treatment patterns, and formulate clinical practice recommendations. Uniform and consistent medical documentation and coding are essential to proper surveillance and research efforts.

ICD9	Description
371.60	Keratoconus, unspecified
371.61	Keratoconus, stable condition
371.62	Keratoconus, acute hydrops
371.71	Corneal ectasia
ICD10	**Description**
H18.601	Keratoconus, unspecified, unspecified eye
H18.602	Keratoconus, unspecified, left eye
H18.603	Keratoconus, unspecified, bilateral
H18.609	Keratoconus, unspecified, unspecified eye
H18.611	Keratoconus, stable, right eye
H18.612	Keratoconus, stable, left eye
H18.613	Keratoconus, stable, bilateral
H18.619	Keratoconus, stable, unspecified eye
H18.621	Keratoconus, unstable, right eye
H18.622	Keratoconus, unstable, left eye
H18.623	Keratoconus, unstable, bilateral
H18.629	Keratoconus, unstable, unspecified eye
H18.711	Corneal ectasia, right eye
H18.712	Corneal ectasia, left eye
H18.713	Corneal ectasia, bilateral
H18.719	Corneal ectasia, unspecified eye

Coding for Corneal Ectasia.

Operational and Readiness Considerations for All Providers

Awareness
- Unit providers must be aware that Service members with any type of corneal ectasia have an ocular disease that can affect quality of vision, specifically night vision. They require regular follow up examinations and are at an increased risk for damage to the cornea from trauma. These Service members may require contact lenses for best vision. Common visual complaints are blurry vision, glare, and sensitivity to light.
- Patients with corneal ectasia commonly have dry eyes that require using artificial tear eye drops or medicated eye drops frequently.
- If a Service member has worsening corneal ectasia, a new treatment, CXL has recently become available to slow or halt the disease. Facilitation of referral may be needed for the Service member to receive timely evaluation and treatment.
- If a Service member with a history of corneal ectasia reports that their glasses or contact lenses do not work as well as when they originally received them, or if there are any changes in vision, a comprehensive eye examination is warranted.
- Frequently, individuals with corneal ectasia wear contact lenses. The most common contact lens-associated complications include corneal ulcers, inflammation, permanent corneal scarring, ocular perforation, vision loss, and in worst cases, the loss of an eye.[30]
- Signs of a potential contact lens problem include pain, tearing, light sensitivity, and a decrease in vision. If a contact lens problem is suspected, an eye examination by an eye care provider is warranted.

- Post-CXL complications need to be addressed to determine the Service members' overall readiness.
- Delayed complications that can occur post CXL:
  - Haze formation
  - Infective keratitis
  - Dry eye/ocular surface disease
  - Progression of corneal ectasia

Prevention and Mitigation Considerations

Individuals with corneal ectasia should not rub their eyes because the physical forces from eye rubbing can make ectasia worse. Service members should alert team members of this concern, as subconscious eye rubbing frequently occurs.

- Environments with polluted airspace that could make the Service member rub their eyes more frequently should be avoided.
- Service members with a history of allergy or atopic dermatitis have a higher risk of corneal ectasia.
- Very rarely, an individual with corneal ectasia can develop a serious complication called corneal hydrops. New onset of pain and blurred vision in a Service member with a history of corneal ectasia requires prompt evaluation by an eye care provider.
- Using approved eye protection from the DoD Authorized Protective Eyewear List (APEL) is critically important, especially for those who have corneal ectasia.

As the understanding and the technology to detect keratoconus and other progressive corneal ectatic diseases continues to rise, so does the reported incidence and prevalence. This presents unique challenges to both the Service member and the Services as high quality vision is necessary to be most effective in operational environments. Also, if one were to receive a diagnosis of keratoconus or corneal ectasia, the most common form of visual improvement is gained via contact lenses. Contact lens use is prohibited in initial entry training, field exercises, gas chamber exercises, deployments, and/or most operational settings under current DoD policies DA PAM 40-506[31] (the Army Vision Conservation and Readiness Program) as well as the U.S. CENTCOM 021922Z[32] (December 2011 Mod 11 to U.S. CENTCOM individual protection and individual-unit deployment policy).[30] However, CXL has been identified to halt or significantly slow the progression of these visually disabling conditions. Therefore, eye care providers need to be well informed on how to best detect keratoconus and other corneal ectasias as early as possible with the goal that they can halt vision loss. Early intervention is critical to prevent a reduction in quality of life or vision loss that can impede in the ability to fight and perform any required military duties.

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Conference Presentations and Publications

The following references highlight contributions from VCE staff and collaborators.

Recent Conferences

National Capital Area TBI Research Symposium

4-5 March 2019, Bethesda, MD | https://www.abstractscorecard.com/cfp/submit/login.asp?EventKey=CHHZRJZM

Podium Presentation(s)

Felix M. Barker II OD MS, Natalya Merezhinskaya PhD, Rita K. Mallia OD MPA, DoHwan Park PhD, Daniel W. Bryden PhD, David A. Eliason MD, Mark E. Reynolds COL, MC, USA **Prevalence of Visual Deficits and Dysfunctions associated with Traumatic Brain Injury (TBI): A Meta-Analyses**

Association for Research in Vision Opthalmology

28 April - 2 May 2019, Vancouver Convention Centre Vancouver, B.C. | https://www.arvo.org/annual-meeting/

Podium Presentation(s)

Mark E. Reynolds COL, MC, USA, Vision Center of Excellence (VCE)/Defense Health Agency, J-9 **Military Relevant Priorities and Strategies for Injury Diagnostics and Treatments**

American Optometric Association

19-23 June 2019, St. Louis, MO | https://www.aoa.org/news/inside-optometry/optometrys-meeting-celebrates-st-louis-style

Poster Presentation

Felix M. Barker II OD MS, Natalya Merezhinskaya PhD, Rita K. Mallia OD MPA, DoHwan Park PhD, Daniel W. Bryden PhD, David A. Eliason MD, Mark E. Reynolds COL, MC, USA **Prevalence of Visual Deficits and Dysfunctions associated with Traumatic Brain Injury (TBI): A Meta-Analyses**

Prevent Blindness - Annual Focus on Eye Health National Summit

17 July 2019, Washington, D.C. | https://www.preventblindness.org/eighth-annual-focus-eye-health-national-summit

Podium Presentation

Mark E. Reynolds COL, MC, USA and David A. Eliason, MD, DoD/VA Vision Center of Excellence **Strategic Vision: Population Health and Vision in the Military**

Publications

Winegar JW, Justin GA, Caldwell MC, Calvano CJ, Cason JB, Hofmeister EM, Frazier TC, Mazzoli RA, Colyer MH, Davies BW. **Practice Patterns of United States Military Ophthalmologists in Iraq and Afghanistan: 2002-2018.** Ophthalmology. 2019 May 9. pii: S0161-6420(19)30359-8.

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World Wide Ocular Trauma Readiness Curriculum

Monthly Video Teleconference(s)

A monthly forum for both discussion of clinical cases as well as CEU-approved didactic lecture.

February 2019, CDR Eva Chou, MD USN, MD **Clinical Case Discussion Ophthalmic and OMFS**

March 2019, James Karesh, MD WRNMMC **Management of Canalicular Lacerations**

April 2019, LCDR Alfred Wolanin, MD USN NMCSD; CPT Matthew Cardinale, DO USA; CPT Maxine Harvey, DO USA AMC **Clinical Case Discussions of Open Globe Management**

May 2019, LTC Andrew Gregory, OD USA; Maj Justin Holbrook, OD USAF; Andrew S. Morgenstern, OD, FAAO, FNAP **The Medical Use of Contact Lenses**