The Battle of the Bulge...
Its All About the Backside

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Disclosure

• No Financial Interest
• Not the opinion of the VCE, DoD, VA or any other US Government Agency

Advanced KC Treatment Strategies
“Classic” Manifestations

• Progressive apical thinning with inferior conical protrusion
  — Non-vascularized?
  — Non-inflammatory?
• Bilateral but Asymmetric
  — True Unilateral KC?
• Irregular Cylinder & HOA
  — Vertical Coma
  — Spherical Aberrations

Advanced KC Treatment Strategies
“Classic” Manifestations

• Onset ~ 1st - 2nd decade of life and slows down ~ 4th decade
  — Stabilization trend ≠ absolute
• Incidence ≥ 1/2000?
• Multifactorial causes
  — Genetic?
  — Trauma?
  — IOP?
• LVC implications?
  — FFKC
  — Topo WNL but Family Hx

Video Courtesy Mike Tullo
Advanced KC Treatment Strategies

**Multifactorial Etiologies**

- Mechanical trauma in predisposed individuals
  - IOP Spikes
  - Increased surface temperature
    - Inflammatory mediators
      - IL-1/IL-6, MMPs, TNF-α
      - Proteolytic enzymes
    - Contact lens trauma?
    - Sleeping posture?
    - Keratocyte apoptosis

- Reduced biomechanics!

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**Advanced KC Treatment Strategies**

**Challenges In Conventional Mx**

- Biomechanical Weakening
- Irregular Optics
- Progression

**Growing Accumulations of Stress and Development of “PITA” Syndrome Due to:**

- Visual Frustration
- Physical Frustration
- Psychological Frustration

**Early Detection & Interdisciplinary Co-Management Are Keys to Optimizing Patient Outcome!!**
Advanced KC Treatment Strategies
Anatomical/Optical Signs: Examples
- Munson's Sign/Rizzuti's sign
- Fleischer Ring/Hydrops/Corneal scar
- Scissor Reflex/Oil droplet Reflex
- "Warped" Mires on keratometry/photokeratoscopy

Lower Specificity if Solely Using Color Pattern Recognition Via Anterior Corneal Topographical Profiles

Ultrasound Pachymetry (5262 Eyes)
- Avg. CT = 544 ± 34 um
- Suspect if < 476 um
- CCT is Least reliable indicator
  - Global Delphi Panel
  - KC with normal CCT

Optical Pachymetry
- CT Distribution and Elevation
- Epithelial masking of anterior curvatures
  - Color Pattern Recognitions!!
  - Posterior Profiles and HOAs (ie, V. Coma)
Advanced KC Treatment Strategies
Pachymetry/Tomography

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Corneal Biomechanics
• Dynamic Bidirectional Applanation

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Wavefront Aberrometry

Higher order aberrations make up approximately 17% of the total aberrations of normal eyes.

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High Frequency Ultrasound: Artemis


Advanced KC Treatment Strategies
Expanding Management Paradigm
1) Patient Monitoring Approach – Refractive Correction
2) Prophylaxis Approach – Stabilization
3) Functional Approach – Stabilization + Ref. Correction
Advanced KC Treatment Strategies
CXL/Corneal Cross-linking

- UV + Riboflavin (vitamin B2): 1st reported at U of Dresden; many other studies ongoing since 1994
  - Theo Seiler
  - Eberhard Spoerl
  - Gregory Wollensak

- Avedro - USA
- CXLUSA - USA
- Peshke
- IROC Innocross
- Sooft
- Vega X-Link

A Review of Global Consensus on Keratoconus and Ectatic Diseases

Cornea
April 2015
Purpose of the Project

- Desire to reach consensus
  - Keratoconus
  - Ectatic diseases
- Focus on
  - Definition
  - Concepts
  - Clinical Management
  - Surgical Treatments

Design and Organization

Conclusions

- Resulted in the diagnosis and management of keratoconus and other ectatic diseases via worldwide insight
- Conclusion resulted in
  - Definitions
  - Statements
  - Recommendations

Selection of Expert Panel

- Ophthalmologists with experience in the management of keratoconus and ectatic diseases
- Authorship of scientific publications in high-impact medical journals
- Wide recognition by the specialized medical community
- Willing to comply with the initial question rounds, face-to-face meeting, and project timelines

Selection

- Worldwide geographic distribution
- Represent 4 Ophthalmological Cornea societies
  - Asia Cornea Society (Asia)
  - Cornea Society (USA and international)
  - EuCornea (Europe)
  - PanCornea (Latin America, USA and Canada)
- Each society had 4 experts plus coordinators

Flowchart of the Project
Definition and Diagnosis

Mandatory Findings to Diagnose Keratoconus

- Abnormal POSTERIOR Ectasia
- Abnormal Corneal Thickness Distribution
- Clinical Non-inflammatory corneal thinning
- Values and reference points vary based on device used for screening

Ectatic Disease List

- Keratoconus
- Pellucid Marginal Degeneration (PMD)
- Keratoglobus
- Post Refractive Surgery Ectasia

Non-Ectatic Corneal Diseases

- Terrien Marginal Degeneration
- Dellen
- Inflammatory Melts

Initial Consensus Statements

- Keratoglobus and Keratoconus are different clinical entities
- True unilateral keratoconus DOES NOT EXIST
- Thinning, location and pattern are aspects that distinguish Keratoconus, PMD and Keratoglobus

Best way to distinguish Keratoconus from PMD

- Full corneal thickness map
- Slit lamp exam
- Anterior curvature map
- Anterior tomographic elevation map
Least Reliable Indicator or Determinant

- Central Pachymetry

This is because Keratoconus can be present in a cornea of normal thickness

Consensus on Tests to Diagnose Early or Sub-Clinical Keratoconus

- Tomography (Scheimpflug or OCT)*

Posterior corneal elevation abnormalities must be present to diagnose mild or sub-clinical keratoconus

* Best and Most Widely Available tests

Definition of Ectasia Progression

- Consistent change in at least 2 of the 3 following parameters
  - 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
- Changes need to be consistent over time and above normal variability

Ectasia Progression

- A change in both UCVA and BSCVA is NOT required to document progression
- Testing for progression should be SHORTER for younger patients
- The same measurement platform should be used in sequential examinations

Ectasia Risk Factors

- Down Syndrome
- Relatives of affected individuals/patients
  - Especially in young
- Ocular allergy and systemic atopy
- Ethnic factors (Asian and Arabian)
- Mechanical factors
  - Eye rubbing and Floppy eyelid syndrome
- Connective tissue disorders
  - Marfan syndrome
- Ehlers-Danlos syndrome
- Lebers congenital amaurosis

Summary of Agreements Reached in the Definition/Diagnosis Panel

- The following findings are mandatory to diagnose keratoconus
  - Abnormal posterior elevation
  - Abnormal corneal thickness distribution
  - Clinical noninflammatory corneal thinning
- Keratoconus and FMD are different clinical presentations of the same disease
- The aspect that distinguishes keratoconus, FMD, and keratoglobus is “thinning location and pattern”
- Keratoconus and FMD are best differentiated by a combination of
  - Full tomographic corneal thickness map
  - Slit lamp examination
  - Anterior curvature map
  - Anterior tomographic elevation map
- As opposed to “thinning disorders”, the following are classified under “ectatic diseases”
  - Keratoconus
  - FMD
  - Keratoglobus
  - Postrefractive surgery progressive corneal ectasia
- Keratoglobus and keratoconus are different clinical entities
- True unilateral keratoconus does not exist
- The best current and widely available diagnostic test to diagnose early keratoconus is tomography (Scheimpflug or optical coherence tomography)
Summary of Agreements Reached in the Definition/Diagnosis Panel

• Currently, there is no clinically adequate classification system for keratoconus
• Posterior corneal elevation abnormalities must be present to diagnose early or subclinical keratoconus
• Secondary induced ectasia may be caused by a pure mechanical process (and can be unilateral)
• Central pachymetry is the least reliable indicator (determinant) for diagnosing keratoconus
• The pathophysiology of keratoconus is likely to include the following components
  – Genetic disorder
  – Biochemical disorder
  – Biomechanical disorder
  – Environmental disorder
• Placido-based topography analyzes the central anterior corneal surface, whereas tomography (Scheimpflug and/or optical coherence tomography) analyzes the anterior and posterior cornea and produces a near full corneal thickness map
• Keratoconus can be present in a cornea of normal central thickness

Summary of Agreements Reached in the Definition/Diagnosis Panel

• Ectasia progression is defined by a consistent change in at least 2 of the following parameters where the magnitude of the change is above the normal noise of the testing system
  – Progressive steepening of the anterior corneal surface
  – Progressive thinning and/or an increase in the rate of corneal thickness change from the periphery to the thinnest point
• The changes need to be consistent over time and above the normal variability (ie, noise) of the measurement system (this will vary by system). Although progression is often accompanied by a decrease in BSCVA, a change in both uncorrected visual acuity and BSCVA is not required to document progression
• Risk factors for keratoconus:
  – Down syndrome, relatives of affected patients especially if they are young,
  – Ocular allergy and systemic atopy
  – Ethnic factors (Asian and Arabian),
  – Mechanical factors, eg, eye rubbing, floppy eyelid syndrome,
  – Connective tissue disorders (Marfan syndrome),
  – Ehlers–Danlos syndrome and Leber congenital amaurosis

Most Important Goals in Non Surgical Management

• Halting disease progression
• Visual rehabilitation

Non Surgical Management

Most Important Goals in Non Surgical Management

• Verbal Guidance to the patient to not rub one’s eyes
• Use of topical antiallergenic medication in patients with allergy
• Use of topical antiallergenic medications in patients with atopy or history of eye rubbing
• Use of topical lubricants

Dry Eye and Keratoconus

• There is no direct relationship between dry eye and keratoconus
• Use of eye drops without preservatives is preferable in keratoconic patients
  – PF agents reduce irritation and epithelial trauma...
Refraction

- Subjective refraction should be attempted in all patients with ectasia
- Aberrometry may help determine the refraction early in the disease process
- PAL’s are not contraindicated by rarely successful

Contact Lenses

- Contact lenses do not halt or slow the progression of corneal ectasias
- Cosmetic contact lenses should be discouraged due to difficulty in contact lens fitting and THE INCREASED RISK OF COMPLICATIONS FROM A POORLY FIT CONTACT LENS

“Rigid Contact Lenses”

- Should be used in cases of unsatisfactory vision with glasses or SCL’s
- Gas-permeable are preferred and should be tried initially in patients with keratoconus
- If failed in RGP’s then attempt
  - Hybrid
  - Toric or Bitoric
  - Specialty Keratoconic soft or rigid lens
  - Piggy-back
  - Scleral lenses

Special Situations

- Careful evaluation in cases of Down syndrome
- Pregnancy can contribute to the acceleration of ectasia progression
- In cases of acute hydrops non surgical or less invasive surgical management such as intracameral gas should be attempted before pregnancy

Summary of Agreements Reached in the Non Surgical Management Panel

- The 2 most important goals of management are halting disease progression and visual rehabilitation. Verbal guidance should be given to patients regarding the importance of not rubbing one’s eyes, use of topical antiallergic medication in patients with allergy, and use of topical lubricants (in the case of ocular irritation) to decrease the impulse to eye rub
- In cases of allergy or if there is any allergic component, patients should be treated with topical antiallergic medication and lubricants. Topical multiple-action antiallergic medications (ie, antihistamines, mast cell stabilizer, antiinflammatory) should be used in patients with keratoconus with atopy or history of eye rubbing
- There is no direct relationship between keratoconus and dry eye
- Preservative-free agents are preferred as they are associated with less irritation and epithelial trauma compared with agents with preservatives
- Subjective refraction should be attempted in all patients with corneal ectasia. Aberrometry may help to determine the optical correction in early disease
- Progressive-type glasses are not contraindicated in eyes with keratoconus or other ectasias, but they are rarely successful
- Contact and scleral lenses are extremely important for visual rehabilitation
- in patients with keratoconus and other corneal ectasias

Summary of Agreements Reached in the Non Surgical Management Panel

- Contact lens use does not slow or halt progression of corneal ectasias
- Rigid contact lenses should be used in cases of unsatisfactory vision with glasses or conventional soft contact lenses. Among the rigid contact lenses, gas-permeable lenses are preferred and should be tried initially in patients with keratoconus. In a patient with keratoconus who has failed conventional corneal gas-permeable lenses, alternative contact lens options would be: hybrid lens (rigid center, soft skirt); toric, bitoric, and keratoconus design soft contact lens; keratoconus design corneal rigid gas-permeable contact lens; piggy-back; corneoscleral, miniscleral, and semiscleral contact lens; and scleral lens
- A careful evaluation for keratoconus is strongly recommended in patients with Down syndrome and should be considered in patients with known risk factors for developing keratoconus (Table 2)
- Pregnancy could contribute to acceleration of the progression of ectasia
- In acute hydrops, nonsurgical management should be attempted before keratoplasty
Surgical Management

• Surgery should be considered when patients were not fully satisfied with non surgical treatments
  — “Satisfied best-corrected”

CXL

• Available and performed by 83.3% of panelists
  — Other 16.7% would use it if available
• Variety of techniques
• Collagen Cross Linking is not a correct term
• Corneal Cross Linking is a correct term

CXL

• Extremely important for keratoconus with progression
• Very important for post-refractive keratoectasia (does not say progressive)
• Important for treatment of keratoconus with a perceived risk of progression (not confirmed)
• No consensus for sub-clinical keratoconus

CXL

• Consensus on No age restrictions
  — Should be used in eyes with demonstrated progression
• No consensus on age limit for treatment in keratoconic eye without evidence of progression
• Rarely indicated in patients over 40
• No consensus best UCVA that limits CXL treatment
  — i.e. Treat if better than 20/30?

Penetrating Surgery

• Anterior lamellar keratoplasty (ALK)
• Deep anterior lamellar keratoplasty (DALK)
• Penetrating keratoplasty (PK)
• Intra corneal ring segments (ICRS)
### Superficial Keratectomy
- Photo therapeutic keratoplasty (PTK)
- Photo refractive keratoplasty (PRK)
- Conductive keratoplasty (CK)
- Incisional keratoplasty (IK)
- Microwave corneal remodeling
- Clear lens extraction with IOL (spherical or toric) were uncommonly used by the panel

### Surgical Considerations
- **DALK**
  - Most important patient-related factor is CL intolerance
- **PK**
  - Most important factor is corneal scarring
  - Post hydrop scarring
  - CL intolerance
  - Low corneal pachymetry (below 200 microns)
    - High risk for hydrops
    - Previous failed corneal surgery

### Corneal Transplant
- Any form offered to 21%-60% eligible keratoconic patients
- In patients with no prior hydrops
  - Either ALK or DALK performed in more than 60% of patients
- In patients with prior hydrops
  - ALK or DALK performed in 0% to 20% of patients

### ALK Techniques
- No prior hydrops
  - Over 51% would perform DALK big bubble
- Microkeratome ALK is never performed
- Other ALK is performed less than 25%
  - Manual layer by layer predescemetic DALK (pdDALK) with viscodissection
  - pdDALK with Melles technique
  - Femtosecond laser assisted DALK

### PK
- Over half the panel have performed femtosecond laser-assisted PK for keratoconus
  - Of those who perform femto PK, only perform it on 1%-20% of their cases
- Majority of panelists currently perform a non laser technique

### Most Important Surgical Techniques for Keratoconus
- dDLAK
- PK
- ICRS
Most Important Surgical Technique for Post Op Rigid Lens Fit

- dDALK
- PK

Summary of Agreements Reached in the Surgical Management Panel

- Young (eg, 15-year-old) patient with stable KCN with satisfactory vision with glasses
  - Prescribe glasses only or in combination with contact lenses or CXL
- Young (eg, 15-year-old) patient with progressive KCN with satisfactory vision with glasses
  - Perform CXL and prescribe glasses & contact lenses
- Older (eg, 55-year-old) patient with stable KCN with satisfactory vision with glasses
  - Prescribe glasses only or with contact lenses
- Older (eg, 55-year-old) patient with progressive KCN with satisfactory vision with glasses
  - Perform corneal cross-linking only or with prescription of glasses/contact lenses
- Patient with stable KCN with unsatisfactory vision with glasses but satisfactory vision with rigid contact lenses and tolerates them well? This patient has a spherical equivalent of moderate myopia (eg, 5 diopters (D))
  - Prescribe contact lenses (including scleral lenses)
- Patient with stable KCN with unsatisfactory vision with glasses but good vision with rigid contact lenses, and tolerates them well? This patient has a spherical equivalent of high myopia (eg, 15 D)
  - Prescribe contact lenses (including scleral lenses)

Documenting Ectasia Progression

- Requires 2 of the following:
  - Steepening of the anterior surface
  - Steepening of the posterior surface
  - Thinning or changes in the pachymetric rate of change
- Magnitude of changes is unknown

**YOUNGER PATIENTS SHOULD BE EXAMINED FOR CHANGE AT SHORTER INTERVALS AS ECTATIC CHANGE CAN PROGRESS RAPIDLY IN THIS GROUP**

Summary of Agreements Reached in the Surgical Management Panel

- Patient with stable KCN with unsatisfactory vision with glasses and contact and scleral lenses, or who does not tolerate contact or scleral lenses? This patient has a spherical equivalent of moderate myopia (eg, 25 D)
  - Perform dDALK. Consider ICRS in eyes with adequate corneal thickness and minimal to no scarring
- Patient with stable KCN with unsatisfactory vision with glasses and contact and scleral lenses, or who does not tolerate contact or scleral lenses? This patient has a spherical equivalent of high myopia (eg, 15 D)
  - Perform dDALK
- Patient with stable KCN with unsatisfactory vision with glasses and contact and scleral lenses? This patient has moderate anterior corneal scarring but no evidence of previous corneal hydrops
  - Perform dDALK
- Patient with stable KCN with unsatisfactory vision with glasses and contact and scleral lenses? This patient has moderate anterior and deep corneal scarring with evidence of previous corneal hydrops
  - PK alone or attempt pdDALK

Importance of Tomography

- Anterior and posterior views
- Significant importance of the posterior cornea as an early indicator of ectatic change
- Posterior corneal surface and alteration in the corneal thickness progression are necessary to diagnose keratoconus
Differentiate KC form PMD

- Items needed to differentiate:
  - Anterior and posterior tomography
  - Corneal thickness map
  - Slit lamp exam
  - Anterior surface measurements

KC and PMD Summary

- Different clinical presentations of the same basic disease process
- Ectatic diseases:
  - Keratoconus
  - PMD
  - Post refractive surgery ectasia
  - Keratoglobus

Pathophysiology of Ectasias

- Multifactorial
- Genetic
- Biochemical
- Biomechanical
- Environmental

Summary of CXL

All assuming good candidates

- Anyone with progressive ectasia should undergo CXL no matter what age level or vision
- OK to proceed with CXL even if patients were happy with their vision

Questions

- What is the new estimate or incidence of Keratoconus?
- Why weren’t Yaron Rabinowitz, MD Stephen Klyce, MD and Steven Wilson, MD included in the panel?
- Why is there no information on contact lens fitting, materials, design and follow-up?
- Why no Optometrists if they deal with a significant part of the non surgical and post operative patient?
- How is there is no direct relationship between dry eye and keratoconus?
- Contact lenses are just wrong...

Questions

- Why no information on placido disc technology?
- Why no discussion about biomechanical properties of the cornea?
- Why no information or recommendation of epi-on or epi-off
- If keratoconus is bilateral, should you treat a 20/20 eye non-keratoconic eye to prevent the progression?
- Is prophylactic treatment recommended?
Review of 
*The Association Between Sociodemographic Factors, Common Systemic Diseases, and Keratoconus* 
*Ophthalmology 2015*

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Executive Board Member  
International Academy of Keratoconus

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**Purpose**

- Determine if an association exists between
  - Common systemic diseases
  - Sociodemographic factors
  - Keratoconus

Among a large, diverse group of insured individuals in the United States

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**Design and Participants**

- Retrospective longitudinal cohort study
- 16,053 Keratoconic patients
- 16,053 non-Keratoconic patients

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**Results:**

Odds of being diagnosed with Keratoconus

- Compared to Whites - No Diabetes Mellitus (DM)
  - 57% higher in Blacks
  - 43% higher in Latinos
  - 39% lower in Asians
  - 20% lower in uncomplicated DM
  - 52% lower in DM with end organ damage
  - 35% lower in collagen vascular disease

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**Other Factors Increased Odds**

- Sleep Apnea
- Asthma
- Down Syndrome
Factors Not Associated

- Allergic Rhinitis
- Mitral Valve Prolapse
- Aortic Aneurysm
- Depression

Conclusions

- When caring for the keratoconic patient, clinicians should
  - Inquire about breathing or sleeping
  - Refer for sleep apnea or asthma evaluation
- Patients with Diabetic Mellitus have a potentially lower risk of Keratoconus due to glycosylation