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The Missing Link: Earlier Keratoconus Diagnosis to Save Vision

Mitch Ibach, OD


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Host: Dr. Stephanie Woo

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Speaker Bio

Dr. Mitch Ibach is a residency trained optometrist at Vance Thompson Vision in Sioux Falls, SD. Dr. Ibach attended the Pacific University College of Optometry where he graduated summa cum laude. Mitch completed his residency training at Minnesota Eye Consultants with a concentration on cornea, refractive surgery, external disease and glaucoma. In September of 2014, he joined Vance Thompson Vision to focus on advanced anterior segment surgery care and pathology. Mitch is a fellow of the American Academy of Optometry, an Intrepid Eye Society member, a member with the American Optometric Association, Optometric Glaucoma Society (OGS), and the South Dakota Optometric Society. Mitch serves as the associate residency director at Vance Thompson Vision and is also an adjunct clinical faculty for the Illinois College of Optometry and the Pikesville College of Optometry.



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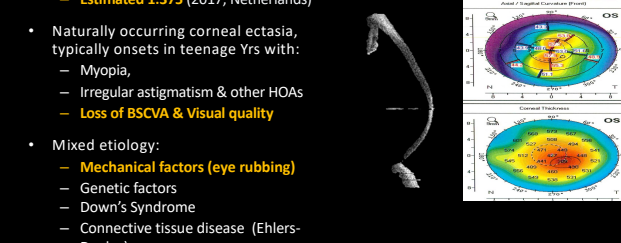
The Missing Link: earlier keratoconus diagnosis to save vision

Mitch Ibach, OD FAAO
Vance Thompson Vision
Co-Residency Coordinator

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Unstable Cornea: Keratoconus (KCN)

- Prevalence ~1:2000 (1986, US)
 - Estimated 1:375 (2017, Netherlands)
- Naturally occurring corneal ectasia, typically onsets in teenage Yrs with:
 - Myopia,
 - Irregular astigmatism & other HOAs
 - Loss of BSCVA & Visual quality
- Mixed etiology:
 - Mechanical factors (eye rubbing)
 - Genetic factors
 - Down's Syndrome
 - Connective tissue disease (Ehlers-Danlos)




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Unstable Cornea: Refractive Surgery Ectasia

Ectatic corneal degeneration

- Unilateral or bilateral
- Maybe asymmetric
- Manifests after corneal surgery

Higher risk in big corneal ablations, eye rubbers, RK patients, mildly abnormal pre-op topo/tomography



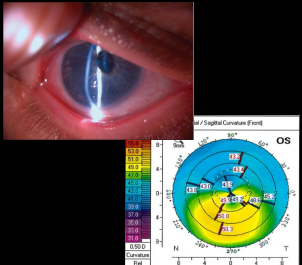
Randallman JB, Russell R, Ward MA, et al. Risk factors and prognosis for corneal ectasia after LASIK. Ophthalmology 2003; 110:267-275.
Paul AN, Johnson M, Sutti N. Progressive keratoconus after laser in situ keratomileusis. J Refract Surg 2004; 20:5718-5722.

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Other Corneal Ectasias

Pellucid Marginal Degeneration (PMD)

- Later life onset (2nd to 5th decade)
- Crescent shaped band of inferior thinning
- "Crab claw" topography pattern, "beer belly" corneal appearance



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Other Corneal Ectasias

Keratoglobus

- Most common form is congenital
 - A/W Ehler's Danlos
- Generalized thinning over whole cornea
- Diffuse rather than focal thinning



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Reported incidence & prevalence of keratoconus may be on the rise globally, largely due to improved diagnosis

Reference	Prevalence	Geography
Kennedy et al. 1986	0.05% or 1:2000	US
Jonas et al. 2009	2.3%	India
Milodot et al. 2011	2.3%	Israel
Xu et al. 2012	0.9%	China
Hashemi et al. 2014	2.5%	Iran
Godefrooij et al. 2017	0.26% or 1:375	Netherlands
Torres Netto et al. 2018	4.79%	Saudi Arabia
Chan et al. 2020	1.2% or 1:84	Australia
Hashemi et al. 2020*	0.14% or 1:700	Global Meta-Analysis

Incidence and prevalence of keratoconus in Denmark – an update

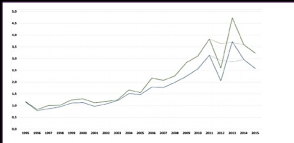


Fig. 1. Annual incidence rate per 100 000 person-years 1995–2015 (green) and annual incidence rate per 100 000 person-years 1995–2015 (including immigrants and descendants) (blue). The dotted lines indicate the incidence rate if the 28 persons (44 persons, excluding immigrants and descendants) recorded to be diagnosed on 1 January 2015 were at first diagnosed in 2012.

Acta Ophthalmol. 2019; 97: 752–755

*Hashemi H, Heydarian S, Hoshmand E, et al. The Prevalence and Risk Factors for Keratoconus: A Systematic Review and Meta-Analysis. *Cornea*. 2020;39(2):203-209


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Published in final edited form as:
Cornea. 2007 September ; 26(4): 223–232. doi:10.1016/j.jco.2007.03.001

Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study: Methods and Findings to Date


H. Wagner¹, J.T. Barr², K. Zadnik³, and the Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study Group
¹Nova Southeastern University, College of Optometry, Ft. Lauderdale, Florida
²The Ohio State University College of Optometry, Columbus, Ohio



Bilateral
96%



Eye Rubbing
50%



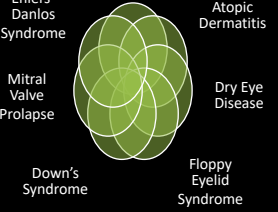
Genetic
13%

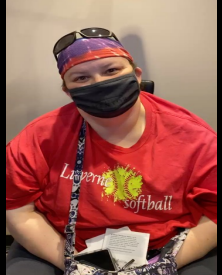
Wagner H, Barr J, & Zadnik K. (2007, September). Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study: Methods and Findings to Date. *Cornea*. 26(4), 223-232

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What does the KCN patient look like?

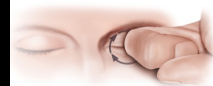





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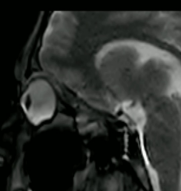
STOP the RUB



+50%



~90%



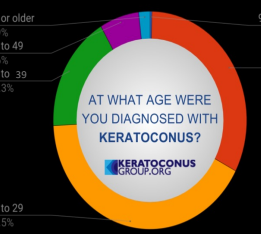
Hawkes E, Naravaty MA. Eye Rubbing and Keratoconus: A Literature Review. *Int J Kerat Ect Cor Dis*. 2014;3(3):118-121

Gattell D, & Sivakovskiy J. Novel Investigation of Eye Rubbing with Dynamic Medical Imaging (Video). *Koelchid Foundation*.

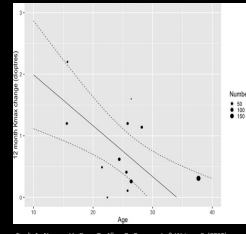
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Who Gets Keratoconus - Age



AT WHAT AGE WERE YOU DIAGNOSED WITH KERATOCONUS?
 KERATOCONUS GROUP.ORG




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Comprehensive Eye Exam

1. Patient Background/History
2. Autorefractor/Autokeratometer
3. Visual Acuity
4. Retinoscopy
5. Refraction
6. Eye Focusing and Eye Teaming Tests
7. Slit Lamp Exam
8. Tonometry
9. Pupil Dilation
10. Aberrometry
11. Eye Health




*Red indicates where KC may be detected

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Patient Reported Symptoms of Keratoconus

- Blurring of vision and/or loss of vision
- Decreased tolerance to contact lenses
- Increased sensitivity to light and glare
- Difficulty driving at night
- A halo around lights and ghosting (especially at night)
- Eye strain
- Headaches and general eye pain
- Eye irritation, redness, swelling,

Keratoconus vision symptoms can be hard to pinpoint until later stages



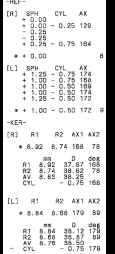

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Keratoconus Diagnosis: Tools in Every Office

- Objective measurement of the eye's prescription
- Passes light through the eye & measures reflection bounced back
- Great for pediatrics/intellectual delays
 - Requires no verbal response
 - Fast

Error Messages

Autorefractors are designed to measure a smooth prolate cornea. Other reasons beyond KC that an AR would provide an error message --> corneal graft, dry eye, blinking

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Which is the suspicious Auto-refraction?

- A. -1.00 -1.00 x 84
- B. +0.75 - 2.25 x 174
- C. +2.00 - 4.50 x 45
- D. -0.50 sph

3.00D refractive cylinder

Foth, A., Nguyen, V., Goss, D., Allen, B., Reisman, J., & Watson, S. (2019). Keratoconus Natural Progression: A Systematic Review and Meta-analysis of 11,529 Eyes. Ophthalmology. Wagner, H., Barr, J., & Zadnik, K. (2007, September). Collaborative Longitudinal Evaluation of Keratoconus (CLEK) Study: Methods and Findings to Date. Contact Lens. American Eye, 30(4), 223-232. Peters, D., Allen, J., Barnes, J., Mollnes, M., Tan, M., & Brannstrom, B. (2011). Medical Vector Analysis of Evolving Corneal Asymmetry: Changes in Keratoconus. Investigative Ophthalmology & Visual Science.

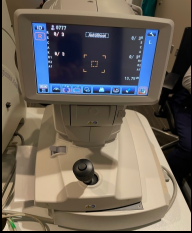
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Keratoconus Diagnosis: Tools in Every Office

Keratoconus Screening Using Values Derived From Auto-Keratometer Measurements: A Multicenter Study

TAKASHI KOHMA, TOMOYA NISHIDA, TOMOAKI NAKAMURA, AKENO TAMAKO, ASATO HASEGAWA, YUKI TAKAGI, HIROFUMI SATO, AND KAZUYO KUBIKAWA

- Auto-K parameters to predict KCN
- Absence of WTR cyl > Steep K > Flat K
- 93% normal cohort had WTR cyl
- 41% KCN cohort had ATR or oblique cyl
- 2/3 KCN cohort not correctable to 20/20
- ATR/Oblique cyl + BCVA <20/20 GET TOPO





Kohma, T., Nishida, T., Nakamura, T., Tamaki, A., Hasegawa, A., Takagi, Y., Ichikawa, K. (2020). Keratoconus Screening Using Values Derived From Auto-Keratometer Measurements: A Multicenter Study. American Journal of Ophthalmology, 215, 127-134.

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Keratoconus Diagnosis: Tools in Every Office

- Phoropter:
 - Unhappy with glasses / remakes
 - Reduced image quality
 - Patient ≠ 20/20
 - MRX increasing in myopic SEQ





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Keratoconus Diagnosis: Tools in Every Office

- Retinoscopy
- Available to ODs without advanced diagnostic equipment
- Sensitive and reliable test for detecting keratoconus, including early disease.
- Can be implemented for pediatric screening program.

During a 2019 Clinical Study:
 123 patients were screened for Keratoconus
 Eligible participants were initially examined by two independent retinoscopists
 Retinoscopy was found to have a sensitivity of **98%** and specificity of **78%** when compared with Pentacam's Belin and Ambrósio Display Final D index of ≥ 2.69 .



1) Al-Mohroqi, Hashim, et al. "Retinoscopy as a Screening Tool for Keratoconus." Cornea, U.S. National Library of Medicine, Apr. 2019. www.ncbi.nlm.nih.gov/pubmed/30640248

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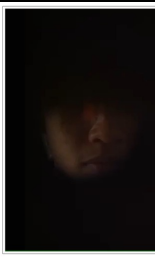
Do You Commonly Do Retinoscopy and could you recognize a scissoring reflex?

- A. Yes, Yes
- B. Yes, No
- C. No

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Retinoscopy

Normal



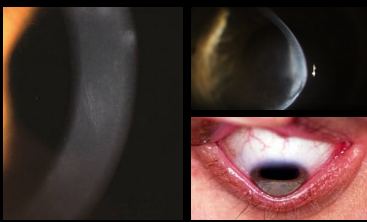
Scissor Reflex



https://www.youtube.com/watch?v=8RE-gCT4U Sahu, C. L. et al. Scissoring Reflex. (CD). Youtube. https://www.youtube.com/watch?v=ce6dMecsoo

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Keratoconus Diagnosis: Tools in Every Office



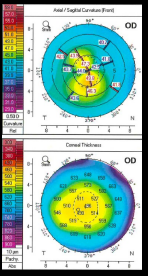
Slit lamp

- FLEISCHER RING
- VOGT'S STRIAE
- STROMAL THINNING
- STROMAL SCARS
- ENLARGED CORNEAL NERVES
- MUNSONS SIGN
- ACUTE HYDROPS

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KCN Clues that necessitate Topo/Tomo

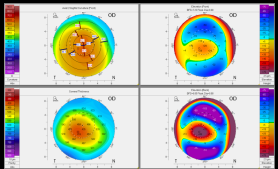
- Aggressive eye rubbing
- Auto K's/Keratometry > 48D
- Error Message Auto K's/Ref.
- Refractive cylinder > 3D
- Retinoscopy abnormal/scissor reflex
- Corneal striae or warped beam
- Family Hx: moderate +/- test



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Advanced Diagnostic Imaging

- Advanced imaging techniques enable earlier and more accurate diagnosis of keratoconus. **Gold Standard Topography/Tomography**
- If suspicious corneal findings → **refer for imaging**
- Early signs of keratoconus may include:
 - Stromal and epithelial thickness changes
 - Posterior elevation changes
 - Wavefront aberrations
 - Topographic changes
 - Inferior steepening
 - Irregularity indices



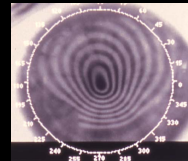
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Do you have Topo/Tomography in your practice?

- A. Yes
- B. No
- C. No, but I utilize anterior seg OCT or epithelial mapping

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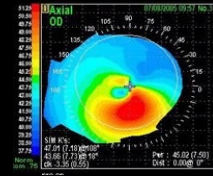
Corneal Topography



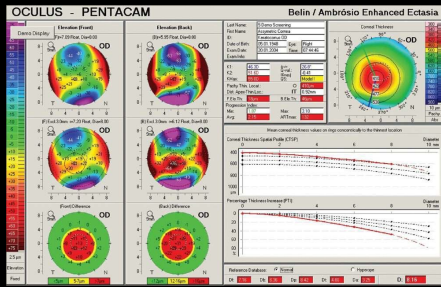
Placido Disc Topographer

- Concentric rings of light projected onto cornea.
- Reflection = mires

- Measures the anterior corneal elevation
- Reveals K1, K2, & corneal astigmatism.

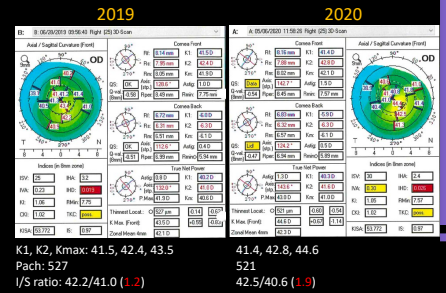


Belin-Ambrosio Enhanced Ectasia Display



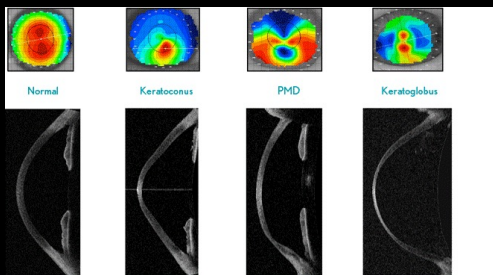
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Monitoring for Progression



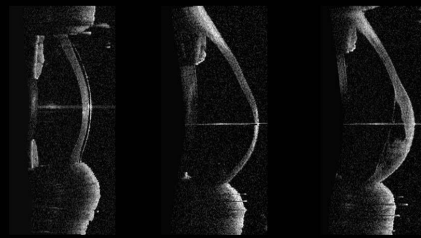
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Anterior Segment OCT



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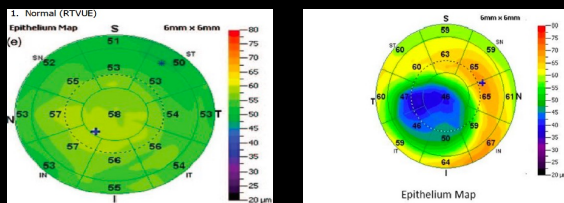
Evolution of KCN: Ectasia to Hydrops



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Epithelial Mapping

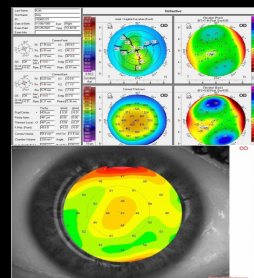
Remember the average epithelial thickness is 50um



Falkowich, Ella, and Linda Charles. "TDM Eases Diagnosis for Scissoring Reflective Sensors." *Ophthalmology Times*. 15 Sept. 2019.

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Normal vs. Form Fruste KCN vs. KCN



"Is this keratoconus or no?
 Would you recommend surgical treatment?"
 A.No, monitor
 B.Yes FFKCN, monitor
 C.Yes KCN, monitor
 D.Yes KCN, refer for treatment

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Normal vs. Form Fruste KCN vs. KCN

Corneal epithelial thickness mapping using Fourier-domain optical coherence tomography for detection of form fruste keratoconus

Cybil Temstet, MD, Osman Sandali, MD, Naciin Bouharroua, MD, Tawou Hamiche, OPTM, Alice Galan, Optm, Mohamed El Samharwi, MD, Elvina Badi, MD, PhD, Laurent Laroche, MD, Vincent Boudier, MD, PhD

- Thin total epithelium
- Thinner central epithelium
- Thinner minimum epithelial thickness
- Greater difference inferior to superior
- Thin epi over area of posterior change
- 52um epithelium at thinnest point

Pichot N, Schimziani F, Hozar S, et al. Distinguishing keratoconic eyes and healthy eyes using ultrahigh-resolution optical coherence tomography-based corneal epithelium thickness mapping. *Am J Ophthalmol*. 2018;199:47-54.

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Higher Order Aberrations

Aberrations and Topography in Normal, Keratoconus-Suspect, and Keratoconic Eyes

- Corneal vertical Coma showed large difference between normal and KCN
- Ocular root mean square represents a viable way of identifying suspected KCN

Gordon-Shag, Antik, Millock, Michel, Hira, Reef, Shaver, Einar' Aberration and Topography in Normal, Keratoconus-Suspect and Keratoconic Eyes. *Optometry and Vision Science*. April 2012, Volume 89, Issue 4, p 411-418 doi: 10.1097/OPX.0b013e31829a7277

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Genetics in KCN

Wang Y, Rahvarzadeh N, Ruten JI, Wang H. Genetic epidemiological study of keratoconus: evidence for major gene determination. *Am J Med Genet*. 2000;95(3):403-409.

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Genetic Testing in KCN

Avagen (avellino labs)

- Genetic testing to determine risk of keratoconus and other K-dystrophies
- In office cheek swab
- Examines 75 genes with over 1000 variants to quantify a patient's keratoconus risk
- 0-100 Red-Green-Yellow score for each gene variant

Trotter, W., & Kramer, E. (2020, October). Identifying, Treating, and Monitoring the Progression of Keratoconus. *Current and Refractive Surgery Today*, 20(10), 36-39.

<https://www.avellino.com/products/avagen-test/>

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What's the Rub?

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Diagnostic patterns in keratoconus

Elke O. Krapp, Ben Claessens, Carina Koppen

Published: May 21, 2020 | DOI: <https://doi.org/10.1016/j.cmo.2020.05.002>

Abstract

Purpose: To investigate the current patterns of diagnosis and referral in keratoconus.

Methods: A retrospective chart review was performed of patients who had recently been diagnosed with keratoconus and attended an external clinic at Antwerp University Hospital, Belgium and Middelheim General Hospital, Ghent, Belgium between June 2013 and February 2018. Exclusion criteria included longstanding keratoconus diagnosis, reduced cognitive capabilities and prior surgical procedures (corneal crosslinking, penetrating keratoplasty or any type of refractive surgery).

Results: Three hundred and twenty-one patients (252 eyes) were included in this study. The mean age was 38.7 ± 8.6 years and the average maximal keratometry was 5.3 ± 0.2 D for the steepest eye and 5.8 ± 0.8 D for the same eye. Upon diagnosis, 223 eyes (20.2%) and 51 eyes (1.7%) had a thinnest pachymetry <450 and <400 µm, respectively. At 6-month follow-up, 58% of patients had been fitted with specialty contact lenses. During follow-up, 139 eyes (27.3%) underwent corneal crosslinking. One patient underwent corneal graft surgery of his worse eye due to contact lens intolerance and insufficient visual acuity.

Conclusion: Despite advances in diagnostic tools, keratoconus is often diagnosed at a relatively late stage. Earlier detection of keratoconus would increase the overall clinical benefit of corneal crosslinking. Further research into screening strategies is required to develop cost-effective screening programs.

Despite advances in diagnostic tools, keratoconus is often diagnosed at a relatively **late stage**.

- 722 Eyes
- Mean age 24.7 year
- Avg. Max K 58.4D
- 1/3 had thinnest pach < 450um

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Earlier Diagnosis and Earlier Intervention Improves QOL

Cornea's Archive for Clinical and Experimental Ophthalmology
 https://doi.org/10.1007/s40201-020-00480-1

Panther C et al. *Graefes Arch Clin Exp Ophthalmol*. 2020; ePub

CORNEA
Evaluation of vision-related quality of life in keratoconus patients, and associated impact of keratoconus severity indicators
 Christophe Panther^{1,2} · Sarah Moran³ · Jean Louis Bourges¹

- BCVA in the better eye is the most important factor contributing to patient's VR-QoL
- Consider targeting the better eye for the greatest impact on VR-QoL
- CXL contributed to higher VR-QoL scores
 - Early stages of KC: avoid deterioration of BCVA
 - In late stages of KC: decrease stress and anxiety concerning the progression of the disease

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KCN: New Mantra

Diagnose **Early** → Stop **Progression** → Maintain BCVA **Potential for Rehabilitation**

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THANK YOU!

mitch.ibach@vancethompsonvision.com

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